Abstract N°: 28

Effects of Semaglutide Treatment on the Psoriatic Lesions in Obese Patients with Type 2 Diabetes Mellitus: An Open-Label, Randomized Clinical Trial

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Introduction & Objectives:

Psoriasis is a chronic inflammatory skin disease with relapsing nature. Estimates are that approximately 2-3% of the world's population suffers from this disease. More severe forms of psoriasis are conditions of high inflammation, which is confirmed by the clinical picture and numerous inflammatory parameters such as C-reactive protein (CRP), cytokines and homocysteine which with disease activity. The objective of this clinical study was to investigate the effect of a GLP-1 receptor agonist semaglutide therapy on pro-inflammatory factors in the serum and the severity of the clinical picture of psoriasis in obese patients with type 2 diabetes mellitus (T2DM) on chronic metformin therapy.

Materials & Methods:

This randomized clinical study was conducted on 31 psoriatic patients with T2DM that were randomized into two groups; one that received semaglutide during the 12-week trial (n = 15), while the second was control (n = 16).

Results:

The results demonstrated that the severity of the clinical picture of psoriasis, determined by the Psoriasis Area and Severity Index (PASI) score, was significantly better after the administration of semaglutide. Also, the quality of life in the group of patients who received the drug, measured by the Dermatology Life Quality Index (DLQI), improved significantly after 3 months. The use of semaglutide led to a significant decrease in pro-inflammatory cytokines in the serum (IL6), as well as a significant decrease in CRP values. A significant decrease in the body mass index (BMI) value in the semaglutide-treated group was also identified, as well as a significant decrease in the level of low-density cholesterol (LDL)

Conclusion:

In conclusion, semaglutide, based on its systemic anti-inflammatory characteristics, could contribute to the treatment of psoriatic obese patients with T2DM.

Abstract N°: 144

Lupus Profundus in a Patient with Multiple Autoimmune Diseases: A Diagnostic Challenge

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Introduction:

Lupus erythematosus panniculitis (LEP), or lupus profundus, is a rare form of chronic cutaneous lupus erythematosus (CCLE) affecting the subcutaneous fat, presenting as painful, indurated nodules and plaques. Due to its nonspecific clinical presentation, it is often misdiagnosed as panniculitis, vasculitis, or morphea, leading to delayed treatment and complications such as lipodystrophy and ulceration. Histopathological findings play a key role in differentiating LEP from other autoimmune and inflammatory conditions.

Case Report:

A 68-year-old woman presented with a 2-year history of painful nodules on the upper extremities and chest. Her medical history included Rheumatoid Arthritis, Sjögren Syndrome, Autoimmune Hypothyroidism, and Arterial Hypertension. Physical examination revealed multiple erythematous, indurated nodules with overlying well-demarcated, scaly, hyperpigmented plaques on the upper extremities and a solitary nodule on the right chest. No lesions were noted on the lower extremities or face.

Histopathology revealed interface dermatitis with necrotic keratinocytes, pigment incontinence, and a dense lymphoplasmacytic infiltrate involving periadnexal and perineural structures. Deep dermis and subcutaneous fat showed fibrinoid necrosis, hyaline changes, and interstitial lymphocytic infiltrate. Small-caliber vessels exhibited intraluminal eosinophilic deposits without true vasculitis. These findings support a diagnosis of cutaneous lupus erythematosus with panniculitis.

The patient was treated with Rituximab, systemic prednisolone, and intralesional triamcinolone injections, leading to symptom resolution and no new lesions at follow-up.

Discussion:

This case illustrates the diagnostic complexity of LEP, particularly in patients with multiple autoimmune diseases, where clinical findings may overlap with vasculitis, scleroderma, and other panniculitides. Histopathological confirmation remains essential for an accurate diagnosis. The combination of lobular lymphocytic panniculitis, hyaline fat necrosis, interface changes, and plasma cells helps distinguish lupus panniculitis from other entities such as erythema nodosum, subcutaneous panniculitis-like T-cell lymphoma, and vasculitic processes. Also, immunofluorescence studies (IgG and C3 deposition at the dermoepidermal junction) can further aid in differentiation (*Table 1*).

Table 1. LEP Clinical Presentation and Diagnosis

Category	Findings in LEP
Clinical Features	Painful, indurated subcutaneous nodules/plaques, erythematous, hyperpigmented, may ulcerate in severe cases
Common Sites	Upper extremities, chest, face; spares lower extremities
Histopathology	Lobular lymphocytic panniculitis, mucin deposition, hyaline fat necrosis, periadnexal/perivascular lymphocytic infiltrates
Direct Immunofluorescence (DIF)	IgG and C3 granular deposition at the dermoepidermal junction (Lupus band test)
Serologic Markers	ANA (+/-), Anti-dsDNA (-/low), Anti-Ro/SSA (+ in some cases), Normal complement levels
Differential Diagnoses	Erythema nodosum, panniculitis, morphea, lymphoma, vasculitis

Therapeutic strategies in LEP depend on disease severity and response to treatment. First-line therapies include antimalarials (hydroxychloroquine) and corticosteroids, while refractory cases benefit from immunosuppressants such as methotrexate or mycophenolate mofetil. The use of Rituximab in this case highlights the potential role of B-cell depletion therapy in severe or refractory LEP cases (*Table 2*).

Table 2. Treatment of LEP Based on Disease Severity

Disease Severity	First-Line Treatment	Second Line / Refractory Cases
Mild (Few nodules, no systemic involvement)	Hydroxychloroquine (HCQ) ± Corticosteroids	Methotrexate, Mycophenolate Mofetil
Moderate (Multiple nodules, progressive disease)	HCQ + Systemic Corticosteroids	Azathioprine, Dapsone
Severe / Refractory (Ulceration, widespread disease, treatment failure)	Systemic Corticosteroids + HCQ	Rituximab, Cyclophosphamide, IVIG

Conclusion:

LEP remains a diagnostic and therapeutic challenge, especially in patients with multiple autoimmune comorbidities. A high index of suspicion, early histopathological evaluation, and multidisciplinary management are crucial for accurate diagnosis and optimal treatment. This case underscores the importance of recognizing LEP in autoimmune patients and the potential benefits of B-cell-targeted therapy in refractory cases.

Abstract N°: 154

Content validation of patient-reported outcome (PRO) measures for fatigue, physical function impacts, and skin symptoms in adult Dermatomyositis (DM)

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Introduction & Objectives:

Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy (IIM) characterized by muscle weakness and characteristic rashes.

The aim of this study was to evaluate the content validity of patient-reported outcome (PRO) measures used in adult DM, i.e., whether the content of these measures 1) captures symptoms or impacts that are important to adults with DM and 2) is comprehensible to respondents.

Materials & Methods:

Individual, semi-structured, 90-minute online interviews were conducted with n=15 adults with DM in the US between January to March 2024. The IRB-approved guide included open-ended questions to evaluate the content validity and perceptions of meaningful changes to scores on 8 PROs: the Functional Assessment of Chronic Illness Therapy – Fatigue (FACIT-Fatigue), the Patient Reported Outcome Measurement Information System – Physical Function Short Form 20a (PROMIS-PF), the 5-D Pruritus (Itch) Scale, the Patient Global Activity Assessment – Verbal Rating Scale (PGA VRS), the Patient Global Impression of Change – Physical Activity (PGIC-PA), and Patient Global Impression of Severity – Physical Activity (PGIS-PA), and the PGIC – Skin Symptoms (PGIC-SS) and PGIS – Skin Symptoms (PGIS-SS). Interviews were audio-recorded, transcribed, anonymized and analyzed using qualitative framework analysis in ATLAS.ti v9.

Results:

Participants were mostly female (n=10, 67%) and/or white (n=9, 60%), with a mean age of 48.6 years (range 28 – 64 years). Participants had completed high school education (n=6, 40%) or other degrees or diplomas, including bachelor's degree (n=4/15, 27%), graduate degrees or college diplomas (each n=2/15, 13%) or associate's degree (n=1/15, 7%). Due to time constraints, not all PROs were debriefed by all participants.

An overview of findings is presented in Table 1. The instructions and item wording were interpreted as intended by the majority of participants for all PROs debriefed. The measures were generally conceptually relevant to the majority of the sample. Most participants correctly interpreted the specified recall periods. The response options/scales were correctly interpreted by participants. Measures were generally considered appropriate, with no suggested changes deemed critical.

Meaningful change was explored at the item level for multi-item PRO s. Generally, 1- or 2-point changes on 5-D Pruritus (Itch) (improvement), PROMIS-PF SF-20a and FACIT-Fatigue (improvement or worsening) items were considered meaningful.

On the PGA-VRS, PGIS-SS, and the PGIS-PA single-item measures, a 1-point change (improvement or worsening) was considered by most participants to be meaningful. Most participants felt that improving on the PGIC-PA to 'A little better' or 'Much better' after taking a new medication would be meaningful, or to 'A little better' on the PGIC-SS.

Conclusion:

All 8 PROs were demonstrated to be content valid for use in research with adults who have DM. For most multi-item PROs, a 1 or 2-point change was considered meaningful, and a 1-point change on single item PROs was considered meaningful. The meaningfulness of these small changes to specific aspects of DM demonstrate the substantial impact of DM on feeling and functioning. These insights are valuable for interpreting quantitative within-patient changes when evaluating treatments for DM.

Table 1. Overview of findings from cognitive debriefing interviews in patients with DM

PRO (n participants who debriefed the measure) and response options	Instruction and item interpretation	Relevance	Recall	Meaningful change
FACIT-Fatigue (n=14) Participants selected from five response options for each item ranging from 'Not at all' (0) to 'Very much' (4). All participants were able to use the response options correctly.	All participants interpreted the instructions, and most items (13/14 items) correctly.	All items were relevant to the majority of participants.	Most participants correctly understood the recall period.	Meaningful change was explored at the item-level. An item-level change of 1- or 2-points was considered meaningful to most participants.
PROMIS-PF (n=12) Participants selected from five response options for each item ranging from 'Without any difficulty' or 'Not at all' (5) to 'Unable to do' or 'Cannot do' (1). All participants were able to use the response options correctly.	The version debriefed did not include instructions. All participants interpreted most items (17/20 items) correctly.	Most items were relevant to more than half of participants.	A range of timeframes were reported (this measure does not specify a timeframe).	Meaningful change was explored at the item-level. An item-level change of 1- or 2-points was considered meaningful to most participants.
5-D Pruritus (Itch) Scale (n=15) Participants selected from the following five-point response scales: Duration: 'Less than 6 hrs/day' (1) - 'All day' (5) Degree: 'Not present' (1) – 'Unbearable' (5) Direction: 'Completely resolved' (1) – 'Getting worse' (5) Disability domains: 'Never' (1) – 'Always'	All participants interpreted the instructions and all items (8 items) correctly.	All items were relevant to the majority of participants.	Almost all participants correctly understood the recall periods.	Meaningful change was explored at the item-level. An item-level change of 1- or 2-point change was considered meaningful to most participants.

PRO (n participants who debriefed the measure) and response options	Instruction and item interpretation	Relevance	Recall	Meaningful change
(5) Distribution: Check list of body locations Most participants were able to use the response options correctly.				
PGA-VRS (n=13) Participants selected from five response options ranging from 'None' (0) to 'Very severe' (4). All participants were able to use the response options correctly.	All participants interpreted the item correctly.	Item was relevant to all participants.	All participants correctly understood the recall period.	A 1-point change was considered meaningful.
PGIS-PA (n=12) Participants selected from five response options ranging from 'None' (0) to 'Very severe' (4). All participants were able to use the response options correctly.	All participants interpreted the item correctly.	Item was relevant to all participants.	All participants correctly understood the recall period.	A 1-point change was considered meaningful.
PGIC-PA (n=11)[1] Participants selected from five response options ranging from 'Much better' to 'Much worse.' All participants were able to use the response options correctly.	All participants interpreted the item correctly.	*N/A*	All participants correctly understood the recall period.	Most participants indicated that improving to 'A little better' or 'Much better' would be meaningful after taking new medication.
PGIS-SS (n=9) Participants selected from five response options ranging from 'None' (0) to 'Very severe' (4). All	All participants interpreted the item correctly.	Item was relevant to all participants.	All participants correctly understood the recall period.	A 1-point change was considered meaningful to most participants.

PRO (n participants who debriefed the measure) and response options	Instruction and item interpretation	Relevance	Recall	Meaningful change
participants were able to use the response options correctly.				
PGIC-SS (n=10) † Participants selected from five response options ranging from 'Much better' to 'Much worse.' All participants were able to use the response options correctly.	All participants interpreted the item correctly.	*N/A*	All participants correctly understood the recall period.	Most participants indicated that improving to 'A little better' would be meaningful after taking new medication.

Abbreviations: FACIT-Fatigue, Functional Assessment of Chronic Illness Therapy-Fatigue; PGA-VRS, Patient Global Activity assessment-Verbal Rating Scale; PGIC-PA, Patient Global Impression of Change – Physical Activity; PGIC-SS, Patient Global Impression of Change-Skin Symptoms; PGIS-PA, Patient Global Impression of Symptoms-Physical Activity; PGIS-SS, Patient Global Impression of Symptoms-Skin Symptoms; PROMIS-PF, Patient-Reported Outcome Measurement Information System-Physical Function. †Participants were asked to imagine they were taking part in a clinical trial of a new treatment while completing the patient global impression of change items.

Abstract N°: 159

Very delayed onset of facial, radiation-induced bullous pemphigoid

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Introduction & Objectives: Among the cutaneous adverse effects of radiotherapy (RT), the occurrence of bullous dermatosis remains very rare.

We report a case of radiation-induced bullous pemphigoid (BP) localized to the face of late onset in a patient treated for ENT cancer.

Observation : A 72-year-old patient with a history of melanoma of the right cheek treated in 1990 with brachytherapy is followed up for an ENT squamous cell carcinoma treated in 2014 with surgery and in 2016 with local radiotherapy at a dose of 64 Gray combined with chemotherapy. The patient is in good partial response, undergoing maintenance treatment with cetuximab.

In October 2023, he developed an erythematous, indurated plaque on the right cheek, progressively extending to the neck and shoulders. Various classes of antibiotics were prescribed without success. The appearance of asymmetrical erosive plaques (later crusted) led to suspicion of erosive pustulosis, and treatment with doxycycline and dermocorticoids led to improvement. An overlap infection caused the transient discontinuation of dermocorticoids, with a rapid worsening of the lesions: appearance of tense, serosanguineous bullae on an erythematous base, prompting the patient’s hospitalization. Nikolsky’s sign was negative in peri-bullous skin. There was no mucosal involvement.

The etiological work-up revealed significant IgG and C3 deposits along the dermal-epidermal junction and high levels of anti BP 180 antibodies, supporting the diagnosis of bullous pemphigoid (BP). The initial aspect, strictly localized to the 30-year-old irradiation field, was in favor of radiation-induced BP.

Treatment with topical steroids was resumed with good results, but secondary extension beyond the irradiation field (trunk and limb) prompted the prescription of general steroids therapy.

Discussion : Very rarely, BP can be induced by radiation. In most reported cases, radiation-induced BP occurred in patients treated for breast cancer, with a highly variable onset time (a few months to several years). The long-delayed onset (30 years from the irradiation of the area, 7 years from the second radiotherapy but at a different localization) and the facial localization make our case so special.

The average lesion-inducing radiation dose was 50Gy, with a minimum threshold of 29Gy. Only a few patients had lesions extending beyond the irradiated region.

Asserting the radiation-induced nature of BP is not easy. Paraneoplastic BP or an incidental association are also possible. In our case, the dissociated evolution of the cancer (in remission) and the bullous disease, and the initial localization of the lesions in the irradiated zone are arguments in favor of the radiation-induced nature of the BP.

Conclusion: We report a rare case of radiation-induced bullous pemphigoid, remarkable for its very long onset

time as well as the facial localization. Radiotherapists and oncologists should be aware of the association between BP and irradiation, so that it can be promptly investigated and treated.

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Abstract N°: 170

KELOIDAL SCLERODERMA WITH NO SYSTEMIC INVOLVEMENT: A Case Report

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Introduction & Objectives:

KS is a chronic autoimmune disease characterized by single or multiple sclerotic plaques in the skin and subjacent tissues. It is distinct from SSc, an autoimmune disease in which diffuse cutaneous sclerosis is accompanied by systemic manifestations1.

We present an unusual case of a 46-year-old female patient with coexistence of two distinct subtypes of morphea (keloidal and linear) and exclusive skin involvement, which contrasts with the typical presentation of nodular/keloidal scleroderma, often associated with organ-specific disease. However, recent publications have diverged from previous ones regarding systemic sclerosis, with no systemic involvement reported between 2018 and 2024, which we evaluated in our descriptive literature review2. With less than 50 cases reported in total, our case underlines the importance of recognizing this rare disease, ensuring appropriate evaluation, treatment, and follow-up.

Materials & Methods:

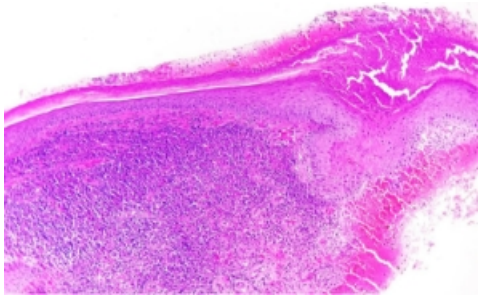
A case of Nodular Sclerodermia is reported, from a patient seen at our service the past year.

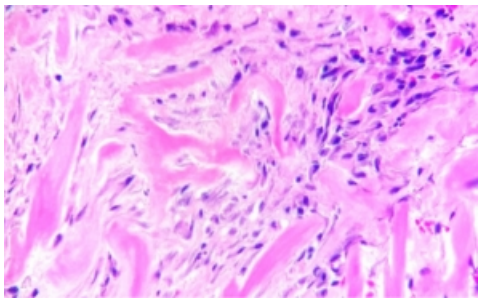
Results:

A 46 years old hispanic female with multiple linear keloids that appeared since she was 25 years old in the form of multiple asymptomatic linear keloids on her chest and arms, slowly increasing in size and number. There was no history of trauma, burns, acne or surgery in any affected site and no personal or family history of keloid formation. (**Fig.A)**

At physical examination multiple violaceous indurated linear keloid-like lesion in the chest, measuring up to 10 × 12 cm (Figure A), as well as a violaceous indurated linear lesion located on the dorsal part of both arms, with an approximate length of 30 x 7cm, were observed.

A 4mm punch biopsy from an indurated linear keloid revealed dermal fibrosis with haphazardly arranged thickened and compact collagen bundles with loss of interfibrillar space, sclerotic appearance, and loss of adnexal structures in the deep dermis with epidermal hyperplasia, orthokeratosis and lymphocytic infiltrates. **(Fig.B)**





Subsequent laboratory investigations revealed normal full blood count, electrolytes, and renal and liver function tests. Anti-Borrelia burgdorferi antibodies were not detected, ANA y eNAs was negative, chest X ray normal, EKG and holter and cardiologist exams without alteration.

These findings confirmed the diagnosis of NODULAR SCLERODERMA.

Conclusion:

KS is a rarely reported morphologic variant where nodular lesions resembling keloids appear within the background of sclerodermatous skin.

Since keloidal lesions are seen in patients with system sclerosis, one must be cautious before classifying these patients as keloidal scleroderma, especially when there are multiple lesions3.

As observed in our patient, keloidal scleroderma seems to occur more likely in patients with dark skin and tend to occur in areas prone to gel keloids2,4.

While nodules appearing on a background of sclerodermatous skin make diagnosis easy, purely nodular or linear lesions may pose diagnostic difficulties 5,6 .

In such instances histopathology become useful in differentiating this from a keloid or a hypertrophic scar.

Minimal atrophy of the epidermis absence of vertically placed bloods vessels in the dermis and preserved elastin fibers within the area of scleroderma may be helpful in differentiating scleroderma from a keloid scar.6

Our patient responded well to topic treatment with corticosteroids and calcineurin inhibitors. She also received methotrexate and oral steroids6.

Abstract N°: 353

Clinical Challenges in the Diagnosis of Psoriasis Vulgaris and Small Vessel Vasculitis

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Introduction & Objectives:

Psoriasis vulgaris (PV) is a chronic inflammatory, T-cell-mediated autoimmune skin disease characterized by keratinocyte hyperproliferation and driven by proinflammatory cytokines. Clinically, it manifests as erythematous, scaly plaques with sharply demarcated margins. While the diagnosis of PV is usually straightforward, coexisting conditions with overlapping clinical features can pose significant diagnostic challenges. We present a unique case of PV occurring concurrently with clinically suspected cutaneous small vessel vasculitis (CSVV), a condition characterized by inflammation of the blood vessels, leading to purpura and ulcerations. It may present alongside PV as a separate pathology rather than a mere clinical mimic.

Materials & Methods:

A 74-year-old patient with a 4-year history of PV and psoriatic arthropathy presented with a worsening skin condition for 2 weeks. He had an ankle joint infection before the rash exacerbation. PV was previously well controlled with topical corticosteroids, and systemic corticosteroids along with phototherapy for episodic flare-ups. Clinical examination showed scaly erythematous plaques on the trunk with widespread reddish-purple non-blanching macules and plaques on the lower extremities. Affected body surface area exceeded 80 percent, psoriasis area and severity index was 25. Laboratory tests revealed elevated CRP levels (142 mg/l) and leukocytosis (11,7 x109/l) with neutrophilia (9,1x109/l). Due to the unclear diagnosis, a skin punch biopsy was performed to differentiate between CSVV and PV. Under these conditions, triggers such as infections, trauma, and some drugs may lead to disease flare-ups, further complicating the diagnosis.

Results:

The recent ankle joint infection may have contributed to immune dysregulation and vascular inflammation, potentially exacerbating PV and triggering CSVV. Therefore, a thorough assessment of the skin lesions during clinical examination is essential to identify any concurrent conditions. Despite the clinical suspicion of vasculitis, histology findings were consistent with PV, showing features such as parakeratosis, acanthosis with elongated rete ridges and prominent granular layer, additionally, the presence of telangiectasia and perivascular lymphocytic infiltration.** Direct immunofluorescence was negative. The diagnosis of PV was made based on history and clinical examination, supported by histopathological results. Treatment with Methotrexate 7.5 mg/weekly was initiated. The skin condition showed mild improvement. Due to the positive response, treatment was continued with oral Methotrexate 10 mg/weekly. After 3 months, a positive clinical response was observed.

Conclusion:

This case highlights the need to reconsider the diagnosis when there is suspicion of coexisting conditions, such as

PV and CSVV. A potential infection may exacerbate both conditions and further complicate the clinical presentation. A thorough re-evaluation ensures the most accurate diagnosis and allows the optimal treatment approach, ultimately improving patient outcomes.

Abstract N°: 362

A rare case of recalcitrant Pemphigus vulgaris with co-existing Behcet's disease treated successfully with Adalimumab

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Introduction: The coexistence of pemphigus vulgaris (PV) and Behcets disease (BD) is extremely rare and may be resistant to therapy. We herein report an interesting case of recalcitrant pemphigus vulgaris with co-existing BD treated successfully with Adalimumab.

Case: A 59-year-old female presented with intermittent low-grade fever, multiple painful crusted erosions over lips/oral/nasal/genital mucosa, and redness/burning in both the eyes for 4-months. During hospitalization, she developed similar lesions, predominantly over the venepuncture sites and a few over the trunk (pathergy phenomenon). Anti-Dsg 1/3 antibody titres were 1:300/1:400 respectively and HLA-B*51 was positive. Histopathology of the oral mucosa showed ulceration of the epidermis with dense mixed inflammatory infiltrate with features of leukocytoclastic vasculitis and intraepidermal collection of neutrophils. DIF revealed IgG and C3 deposits in the intercellular spaces of the epidermis in a fish-net pattern. She did not respond to oral prednisolone (1mg/kg/day), injection dexamethasone pulse, and azathioprine. The therapy was shifted to infliximab infusion at 5mg/kg (300mg) at 0, 2, and 4-week without any significant healing of the lesions. Subsequently, injection Adalimumab was started with a loading dose of 80mg followed by 40mg subcutaneous every 2-weeks which resulted in significant healing of the lesions starting within the first two weeks. The oral steroids could be tapered rapidly over next four weeks and she is maintained on injection Adalimumab. The last follow-up at 6-month revealed no new lesions and the existing lesions had healed significantly.

Conclusion: A strong pathergy phenomenon, poor response to high-dose oral and intravenous steroids and HLA*B51 positivity pointed towards a co-existing BD which is an extremely rare association and may be resistant to multiple immunosuppressants. A high degree of suspicion is required in any case of recalcitrant pemphigus vulgaris to rule out a coexisting or an independently existing BD.

Abstract N°: 457

Comparative Efficacy and Safety of 500 mg versus 1000 mg Rituximab in Pemphigus Vulgaris

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Introduction:

Pemphigus Vulgaris (PV) is the most prevalent chronic autoimmune blistering disorders, which is driven by pathogenic autoantibodies targeting desmosomal adhesion proteins, specifically desmogleins 1 and 3 (Dsg1 & Dsg3).

Rituximab, a chimeric human-mouse monoclonal IgG1 antibody directed against the CD20 antigen on B lymphocytes, has emerged as a first line therapy for PV. This study evaluates the long-term clinical outcomes of

low-dose (500 mg) versus high-dose (1000 mg) Rituximab regimens in PV management, focusing on four key endpoints:

1. complete remission off therapy (CR off), defined as disease resolution without any need for maintenance treatment;
2. complete remission on therapy (CR on), requiring minimal adjuvant therapy; (3) partial remission (PR), marked by persistent but controlled disease activity; (4) relapse or disease flare.

Objectives:

1. Compare the efficacy of 500 mg vs. 1000 mg Rituximab in achieving complete remission off therapy (CR off) in PV patients.
2. Evaluate the rate of complete remission on therapy (CR on) with 500 mg vs. 1000 mg Rituximab.
3. Assess partial remission (PR) and disease control in both groups.
4. Investigate relapse rates and treatment failures in low-dose vs. high-dose groups.
5. Examine reductions in Pemphigus Area and Activity Scores (PAAS) for skin and mucous membranes.
6. Analyze changes in Dsg1 and Dsg3 titers with low-dose vs. high-dose Rituximab.
7. Evaluate the safety and side effects of 500 mg and 1000 mg Rituximab.
8. Determine the potential of 1000 mg Rituximab as first-line therapy for PV.

Materials & Methods:

A total of 68 PV patients (mean age 31–45 years, 69.12% female) were enrolled. Patients were randomly assigned to receive either 500 mg (low-dose) or 1000 mg (high-dose) Rituximab. Clinical outcomes, including complete remission off therapy (CR off), complete remission on therapy (CR on), partial remission (PR), relapse, and treatment failure, were evaluated at 12 months. Pemphigus Area and Activity Scores (PAAS) for mucous membranes and skin were measured. Serum levels of Dsg1 and Dsg3 antibodies were assessed at 3, 6, and 12 months. Statistical analysis was performed to compare the outcomes between the two treatment groups.

Results:

This prospective study evaluates the efficacy and safety of low-dose (500 mg) versus high-dose (1000 mg)

Rituximab in 68 PV patients of mean age: 31–45 years having female predominance (69.12%). At 12 months, patients on high-dose Rituximab (n=21), 7 (33.3%) achieved CR off, 9 (42.8%) CR on, and 5 (23.8%) PR, with no relapses or treatment failures. In contrast, low-dose Rituximab (n=47) showed lower CR off with 9 patients (19.1%), CR on in 21 patients (44.6%), relapse in 5 patients (10.6%), and treatment failure in 6 patients (12.7%). Mucous membrane PAAS reduction was significant in both groups ($p<0.05$), though high-dose therapy (1000mg) achieved faster normalization (0.52 vs. 0.59 at 12 months post infusion). Cutaneous PAAS improvements also showed similar results with high-dose treatment (0 vs. 1.49 at 12 months) but it lacked statistical significance ($p>0.05$).

Serologically, high-dose Rituximab induced earlier Dsg1/Dsg3 titre declines as compared to low dose (Dsg1: $p<0.05$ at 3 and 6 months; Dsg3: showed non-significant trend)

Conclusion:

These findings clearly show high-dose Rituximab's (1000mg) superiority in achieving prolonged and faster remission, preventing relapse, and enabling steroid minimization, aligning with its role as first-line therapy.

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Abstract N°: 523

Bullous pemphigoid and neoplastic comorbidities: fact or myth?

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Introduction & Objectives:

Bullous pemphigoid (BP) is the most prevalent autoimmune bullous disease, with a prevalence that is not negligible despite being considered rare. In recent decades, there has been a significant increase in its incidence. Several factors have been associated with this raise; malignancy has been classically, yet controversially, linked to BP and is considered one of the possible explanations for this increase. The aim of this study was to analyze the neoplastic comorbidities of patients with BP in our setting.

Materials & Methods:

A retrospective observational study was conducted; all patients with a diagnosis of BP, attended in our department between 2000 and the first semester of 2020 were included. The epidemiological, clinical, immunological, histopathological and therapeutic characteristics of all patients, including the presence of malignant comorbidities, were collected. Cases of BP related to, triggered by or developed during any immune checkpoint inhibitor therapy were excluded from this study.

Results:

Out of the total of 257 patients with BP, 47 (18.3%) had at least one malignancy diagnosis during the study period. The mean age of this subgroup of patients was 79.52 years (± 10 SD) and 68.1% were men. Prostate cancer was the most prevalent, affecting 17 patients, followed by gastrointestinal and gynecologic cancers (including breast cancer) in 7 patients each and hematological malignancies in 6 patients. In 42 cases, a temporal relationship between diagnoses could be established; bullous pemphigoid preceded the onset of malignancy in 12 patients, while both diagnoses occurred within a period of 12 months or less in 9 cases. Solid and hematological malignant neoplasms have been classically associated to BP and the risk for developing BP has been estimated to be relatively high within the first 2-5 years after a malignancy diagnosis; however, this remains a subject of debate. The existence of BP as a paraneoplastic syndrome, the systemic inflammation caused by neoplasms and the use of immunosuppressive medications have been postulated as possible connections between BP and malignant diseases.

Conclusion:

Neoplastic diseases are among the main comorbidities described in patients with BP. This finding may be relevant in establishing the need for cancer screening after BP diagnosis. However, studies investigating this association, including large meta-analysis, have reported conflicting results. Further research is needed ¡and neoplasm screening should be guided by patient age, sex, genetic predisposition and other risk factors.

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Abstract N°: 560

Corymbiform lichen planus- A Rare Presentation

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Introduction & Objectives:

Lichen planus (LP) is an idiopathic inflammatory skin disease involving skin and mucosa. It has a chronic course associated with relapses and periods of remission. It is seen in patients of all ages, but most commonly seen in adults between third and sixth decade of life with no gender predominance, but it affects females more commonly. The classical lesion of Lichen planus includes shiny, polygonal, firm, pink to purple colored, flat topped papules. Clinical variants of LP include hypertrophic, eruptive, linear, annular, atrophic, actinic, nail, oral, vulvovaginal, inverse, lichen planus pemphigoides, bullous, lichen planus- lupus erythematosus overlap syndrome.

Case report:

A ten-year-old female patient presented to our department, with itchy dark lesions over the trunk, upper limbs, lower limbs and scalp for four months. Cutaneous examination revealed, multiple violaceous, flat topped, smooth papules surrounded by smaller skin colored to violaceous minute papules over upper limbs, lower limbs, abdomen and back. Dermoscopic examination revealed red- brown background, starburst pattern of Wickham's striae and brown dots. Histopathology of the lesion showed moderately dense lichenoid infiltrate of lymphocytes at the dermoepidermal junction. The epidermis showed saw tooth acanthosis, wedge shaped hypergranulosis and orthohyperkeratosis. Several colloid bodies were seen at the dermoepidermal junction and in the papillary dermis. Based on the clinical, dermoscopy and histopathological findings, a diagnosis of lichen planus with corymbiform lesions was made.

Discussion:

In Botany, the word corymb refers to racemose inflorescence in which flowers are located at the same level due to gradual shortening of length of the pedicels along the axis. Corymbiform arrangement of skin lesions in dermatology is characterized by presence of central larger papule which is surrounded by small satellite papules. Corymbose lesions have been described in literature in different dermatological disorders such as secondary syphilis, Lichen sclerosus et atrophicus, nodular amyloidosis, sarcoidosis. In syphilis, corymbose lesions are associated with recurrences, relapses and have evidence of increased severity.

Conclusion:

In Lichen planus, papules arranged in a corymbiform pattern is not yet reported in literature. In our patient, the lesions had corymbiform arrangement at the time of presentation which makes it an interesting clinical morphology. Thus, Corymbiform lichen planus is a novel, distinct and previously unreported variant of LP in literature. Further observation and research regarding correlation of corymbose lesions with increased severity, relapse or poor prognosis is warranted.

Abstract N°: 652

Generalised bullous pemphigoid in a young man after exposure to haemodialysis membrane: a case report

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Introduction & Objectives:

Bullous pemphigoid (BP) is the most common disorder within the group of sub epidermal immunobullous disorders. BP has multifactorial origin like genetic predisposition, immune dysregulation, environmental triggers and several factors have been implicated in the development and exacerbation of BP, including advanced age, certain medications, ultraviolet light, radiation, infections, vascular graft replacement, dialysis and other autoimmune diseases.

Case report:

A 23-year-old male with a longstanding history of chronic kidney disease (CKD), progressing to end-stage renal disease (ESRD), presented to our hospital in 2022 for hemodialysis (HD). It was initiated using a polysulfone membrane dialyzer (Fresenius, Germany), with a regimen of three sessions per week. Approximately six hours after the first HD session, the patient reported a burning sensation on his skin, which rapidly progressed to the development of fluid-filled blisters. Dermatological examination revealed multiple tense, fluid-filled blisters distributed primarily on photo-exposed areas of the body. Multiple small vesicles and bullae were seen on the trunk and extremities, which revealed a negative Nikolsky's sign but a positive bulla spread sign. Erosions, primarily over the trunk and lower limbs, healed with areas of hypopigmentation and atrophy. A Tzanck smear showed no acantholytic cells. Biopsy from an intact bulla revealed subepidermal clefting with eosinophils and neutrophils present in the blister cavity and in perivascular areas of the dermis. Direct immunofluorescence (DIF) showed linear staining of the basement membrane zone with IgG and C3, confirming a diagnosis of bullous pemphigoid. Given the findings, the patient was transferred to the dermatology ward for further evaluation and management. Hemodialysis was temporarily discontinued, and the patient was started on injectable furosemide, oral nifedipine, and prazosin for management of his ESRD-related complications. Prednisolone at a dose of 1 mg/kg/day had a significant improvement in the itching, tense blisters, and burning sensation after one week. Despite the improvement, symptoms recurred shortly after HD was resumed with the previous parameters. Within four hours of resuming HD, the patient reported worsening of symptoms. Due to the severity of his symptoms, HD was again discontinued, and the dose of oral prednisolone was increased to 1.5 mg/kg/day. The patient was advised to follow up after two weeks, but he did not report back for further evaluation.

Conclusion:

This case illustrates an atypical presentation of bullous pemphigoid in a young male with ESRD, triggered following exposure to a polysulfone membrane during hemodialysis. Prior studies have suggested that BP in HD patients may be triggered by materials used in the dialysis process. In our case, the polysulfone membrane may have played a significant role by activating eosinophils in the bloodstream and skin, subsequently promoting the formation of autoantibodies against the BP180 and BP230 antigens. Clinicians should be mindful of allergic reactions to dialyzer membranes, as potential triggers for bullous pemphigoid in patients on hemodialysis. Further research into the immunological responses to dialysis materials could aid in understanding the pathophysiology of BP in HD patients, ultimately leading to better prevention and management strategies for this subset of patients.

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Abstract N°: 764

Development of a Multivariate Predictive Model for Diagnosing Oral Lichen Planus

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Introduction & Objectives:

Approximately 25% of adults experience oral ulcers, which can be caused by various factors, including lichen planus—a chronic inflammatory and autoimmune disorder affecting the oral mucosa. According to the literature, direct immunofluorescence (DIF) can aid in the differential diagnosis of oral lichen planus. Diagnostic criteria for oral lichen planus, first published in 1978, have been continuously refined, with the most recent diagnostic algorithm proposed in 2019, incorporating clinical signs and medical history. The aim of this study was to develop a new multivariate predictive model combining medical history and DIF results to improve the diagnosis of oral lichen planus.

Materials & Methods:

The study included patients who were presented to the Dermatology Clinic between 2019 and 2022 with erosive lesions in the oral cavity or were referred by their dentists for further evaluation. The following variables were collected: DIF IgG, DIF IgA, DIF IgM, DIF C3, DIF F1, DIF F2, histopathology, gender, age at lesion onset, stress at the time of examination, stress at lesion onset, location of white patches and erosions, previous treatments, use of supplements, herbs, or medications, dental health status, smoking, and use of mouthwash. Statistical analysis was conducted using Statistica 13.

Results:

The study group consisted of 80 patients: 63 women (78.8%) and 17 men (21.2%). Lichen planus was confirmed histopathologically in 4 patients (5.0%), not confirmed in 57 patients (71.2%), inconclusive in 30 patients (37.5%), and excluded in 31 patients (38.8%).

There were no significant differences in the levels of DIF IgG, DIF IgA, DIF IgM, DIF C3, DIF F1, and DIF F2 between patients with confirmed or unconfirmed lichen planus, nor between those with excluded or non-excluded lichen planus. However, data mining analysis identified four significant predictors for the outcome “lichen planus” not excluded by histopathology, but none for lichen planus confirmed. The significant predictors included: stress at lesion onset ($p = 0.017$), erosions under the tongue ($p = 0.049$) and white patches under the tongue ($p = 0.029$).

Conclusion:

In some patient populations, DIF is not a significant indicator for diagnosing lichen planus, regardless of whether strict diagnostic criteria were used during histopathological examination or whether the findings were suggestive of lichen planus. The identification of stress at lesion onset, white patches under the tongue, and erosive lesions in specific oral regions as significant predictors highlights the importance of integrating clinical and medical history data into diagnostic models.

Abstract N°: 833

A Novel GPCR19 Agonist Reverses Hair Loss by Suppressing NLRP3 Inflammasome Activation in Alopecia Areata

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Introduction & Objectives:

Alopecia areata (AA) is an autoimmune hair loss disorder caused by the collapse of immune privilege in hair follicles, leading to aberrant immune attacks on the follicular epithelium. Current treatments are often limited by safety concerns or lack of sustained efficacy, emphasizing the need for safer and more effective therapies.

Several studies have highlighted the pivotal role of innate immunity and inflammasome signaling in AA. NLRP3 inflammasome activation contributes not only to the initiation but also to the progression of the disease, and its inhibition with the selective inhibitor MCC950 has demonstrated therapeutic benefit in the C3H/HeJ mouse model of AA.

G protein–coupled receptor 19 (GPCR19) is an emerging target due to its unique ability to suppress both inflammasome priming and activation. In a recent study, GPCR19 deletion aggravated hair loss, while GPCR19 agonist treatment restored hair growth and mitigated JAK1-STAT3 and downstream inflammatory signaling.

In this study, we introduce a novel GPCR19 agonist, identified through AI-driven drug discovery, with anti-inflammatory and hair-regenerative properties, suggesting a first-in-class therapeutic strategy for AA.

Materials & Methods:

GPCR19 agonist activity was confirmed by HTRF cAMP assay in THP1-GPCR19 cells. Anti-inflammatory effects were assessed under LPS, IFN-γ, and BzATP stimulation in THP1-GPCR19 cells and human PBMCs by ELISA (TNF-α, IL-1β). Treg induction in PBMCs was analyzed by flow cytometry under resting conditions. Ex vivo efficacy was evaluated in murine vibrissa follicle organ cultures by measuring hair shaft elongation under poly(I:C) and IFN-γ exposure. In vivo efficacy was evaluated in C57BL/6 mice (topical application for anagen induction) and in C3H/HeJ mice with poly(I:C)- and IFN-γ-induced alopecia areata, with hair regrowth assessed visually and by histological analysis.

Results:

The novel GPCR19 agonist, SH1010337, demonstrated potent receptor activation, with a cAMP EC_{50} of 11.3 nM in THP1-GPCR19 cells. In THP-1 cells stimulated with LPS and IFN-γ, the compound inhibited TNF-α and IL-1β secretion with IC_{50} values of 0.9 and 49.4 nM, respectively. In human PBMCs, SH1010337 reduced LPS-induced TNF-α secretion (IC_{50}: 0.56 μM) and induced a dose-dependent increase in regulatory T cells under non-stimulated conditions, with an 11.2% increase in the $CD25^{+}Foxp3^{+}$ fraction of $CD4^{+}$ T cells at 10 μM.

In ex vivo organ culture of murine vibrissa follicles, hair shaft elongation was markedly suppressed by poly(I:C) and IFN-γ treatment. Co-treatment with SH1010337 effectively rescued this suppression, restoring hair shaft

elongation to 97% of the vehicle control level.

In vivo, SH1010337 promoted robust anagen induction in C57BL/6 mice, as evidenced by macroscopic hair regrowth. In the C3H/HeJ AA model induced by poly(I:C) and IFN-γ, topical treatment of SH1010337 improved hair coverage and attenuated inflammatory features in affected skin.

Collectively, these findings demonstrate that SH1010337 promotes hair regrowth and immune homeostasis by modulating innate immunity and enhancing Treg differentiation, demonstrating robust efficacy in murine models of AA.

Conclusion:

This study demonstrates that SH1010337, a first-in-class GPCR19 agonist, modulates innate immune dysregulation and promotes anagen induction in alopecia areata. These results support its advancement toward clinical translation.

Abstract N°: 924

Gender-based disparity in pemphigus severity

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Introduction & Objectives:

Pemphigus is a rare autoimmune blistering disease characterized by the loss of keratinocyte adhesion due to autoantibodies targeting desmogleins. Although this condition affects both sexes, some studies suggest epidemiological differences, with a slight female predominance.

However, the influence of sex on the clinical severity and progression of pemphigus remains poorly explored. This study aims to compare the clinical and evolutionary profiles between men and women to assess a potential link between sex and disease severity.

Materials & Methods:

We conducted a retrospective case series study spanning a period of 8 years, from May 2017 to February 2025. All patients hospitalized in the dermatology department of the Mohammed VI University Hospital Center of Marrakech, who met the diagnostic criteria for pemphigus based on clinical, histological, and immunological findings, were included.

Data collection was carried out from the department's archives as well as digitized records in the HOSIX system. Statistical analysis was performed using IBM SPSS Statistics version 25, applying Fisher's exact test and the student's t-test for the comparison of variables.

Results:

Among the 73 patients included (46 women, 27 men), the mean age was 53 years (51 years for women, 56 years for men), with a median disease duration of 12 months for both sexes. Vulgar pemphigus (44%) was the predominant form in both sexes. The hospitalization was significantly longer for men (42 days) compared to women (15 days).

Clinical symptoms such as asthenia, anorexia, pruritus, pain, and dysphagia were significantly more frequent in men. Mucosal involvement was more common in women (60%), while hair and nail involvement was more common in men (56%).

Men had a higher severity, with 52% having a PDAI (Pemphigus Disease Area Index) >45 and 89% having a skin involvement >5%. The use of high doses of corticosteroids (1.5mg/kg/day) was also more frequent in men (58% vs 13% in women).

In the short term, disease-related complications were more frequent in men, including malnutrition, dehydration, and superinfections. Complete healing was achieved in 47% of women compared to 19% of men.

In the long term, 60.5% of women achieved complete remission compared to 20% of men. Relapses were more frequent in the male group (72% vs 33%). The mortality rate was 7% in men, while no deaths were reported in women.

Conclusion:

Our results show statistically significant differences between sexes in the severity of pemphigus. Men presented a more severe form, with a higher PDAI and a larger affected skin surface, requiring higher doses of corticosteroids. In contrast, women showed better healing and a higher rate of complete remission. Our findings are consistent with the few studies published in the literature, although limited, suggesting a more severe form of the disease in men. These results emphasize the importance of tailoring treatments based on sex. Further large-scale studies are needed to better understand the biological factors underlying these differences.

Abstract N°: 944

The Presence of Purpuric Lesions on the Upper Extremities is Associated with Systemic Involvement in Leukocytoclastic Vasculitis

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Introduction & Objectives:

Leukocytoclastic vasculitis (LCV) is a cutaneous, small-vessel vasculitis of the dermal capillaries and venules, typically presenting with purpura, usually on the lower extremities. Most cases of LCV are self-limited, with 90 percent of cases resolving within weeks to months of onset. LCV is usually confined to the skin, with rare extracutaneous manifestations in less than 30% of cases. In this study, we aimed to predict systemic involvement in patients who presented with a diagnosis of LCV based on histopathological findings.

Materials & Methods:

Adult patients diagnosed with LCV based on histopathological findings, who presented to our clinic between December 2018 and December 2024, were included in this study. The patients were categorized into two groups: those with systemic involvement, as determined by laboratory or imaging findings, and those with isolated cutaneous involvement. These groups were compared based on demographic, clinical, laboratory, histopathological, immunofluorescence characteristics, and skin manifestations.

Results:

A total of 88 patients were included in the study, with 47 having isolated cutaneous involvement and 41 having systemic involvement. No significant differences were observed between the two groups in terms of age and gender (p=0.882 and p=0.906, respectively).

The mean (±SD) body mass index (BMI) was 28.29 ± 4.7 in the isolated cutaneous involvement group and 25.7 ± 5.9 in the systemic involvement group (p=0.03). The mean (±SD) lymphocyte count was 2.05 ± 0.9 $\times 10^9$/L in the isolated cutaneous involvement group and 1.6 ± 0.84 $\times 10^9$/L in the systemic involvement group. Median (Range) C-reactive protein (CRP) was 0.85 (0 – 20) mg/L in the isolated cutaneous involvement group and 2.68 (0 – 34.0) mg/L in the systemic involvement group. Univariate analysis revealed that in patients with systemic involvement mean BMI and mean lymphocyte count were significantly lower compared to the patients without systemic involvement (p=0.03 and p=0.035, respectively). Additionally, IgA deposition in direct immunofluorescence (DIF), the presence of purpuric lesions on the upper extremities, and active smoking were significantly associated with systemic involvement (p=0.028, p<0.01, and p=0.009, respectively).

Multivariate logistic regression analysis identified that the presence of purpuric lesions on the upper extremities increased the risk of systemic involvement 7.6 times compared with those without such lesions (p=0.06, OR: 7.6, 95% CI: 1.7–32.7).

Conclusion:

Our study highlights that while the majority of patients with LCV present with isolated cutaneous involvement,

certain clinical and laboratory parameters may help predict systemic involvement. Specifically, the presence of purpuric lesions on the upper extremities appears to be a significant clinical predictor for systemic involvement, which could guide early identification and more tailored management strategies. Further research is needed to validate these findings and explore additional predictive factors for systemic involveöment in LCV.

Abstract N°: 960

Efficacy and Safety of Low-Dose Rituximab in Pemphigus Vulgaris and Foliaceus: A Case Series in Routine Clinical Practice

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Introduction & Objectives:

Rituximab (RTX) is currently considered the first-line treatment for pemphigus vulgaris (PV), as it is a B-cell-mediated acquired autoimmune disease. Traditionally used at high doses to treat lymphoproliferative diseases, the initial regimen for PV has consisted of 1,000 mg on day 1, followed by a second dose after two weeks. However, there is no consensus on the optimal dosage, as lower doses appear to be equally effective while potentially reducing the risk of toxicity.

Our aim was to describe the outcomes of treatment with a low dose of rituximab (500 mg) for autoimmune blistering diseases in a real-world clinical practice setting.

Materials & Methods:

Retrospective descriptive analysis of patients with PV or pemphigus foliaceus (PF) treated with 500 mg of RTX in the dermatology department of a tertiary hospital. Quantitative variables are expressed as median (minimum - maximum), while qualitative variables are reported as number (percentage).

Results:

We included 10 patients: 4 (40%) women and 6 (60%) men, median age of 55 years (24-77). Of these, 7 (70%) had PV and 3 (30%) had PF; 6 (60%) had moderate disease and 4 (40%) severe disease.

All patients had received a median of 2 (1-3) prior treatments including azathioprine, mycophenolate, methotrexate (MTX), or dapsone, before receiving rituximab 500 mg, and all had been treated with oral prednisone (PDN). The maximum and median doses of prednisone before RTX treatment were 70 mg/day (30-120) and 15 mg/day (5-100), respectively.

Eight patients (80%), including those with PF, received a single dose of 500 mg of RTX, while 2 (20%) received 500 mg on day 1 and 500 mg after 14 days. PDN treatment could be discontinued in all but one patient after a median of 7 months (0-26) of RTX treatment. Other immunosuppressive treatments could be stopped in all but 2 (20%) patients, in whom rituximab was administered in combination with MTX in one case and with MTX and dapsone in another case.

All patients responded to RTX, with 9 (90%) achieving complete remission after a median of 4.2 months (1-18). Seven patients (70%) received maintenance doses of 500 mg RTX. Maintenance RTX was administered when patients showed first signs of relapse. The median interval between RTX doses was 7 months (5-18).

No serious adverse events or emergency department visits were observed.

The median follow-up period was 28 months (2-82). At the end of follow-up, the response to RTX was sustained, and all patients continued on maintenance RTX treatment.

Conclusion

In this series, all patients with PV or PF treated with RTX 500 mg responded to this dosage, with almost all achieving complete remission.

These results suggest that this dosage is sufficient to treat autoimmune blistering diseases by targeting autoreactive B cells while preserving a portion of the normal B-cell population. This partial depletion may reduce the risk of infections and allow for faster immune system recovery.

Our main limitation is the small number of included patients, due to the low prevalence of these diseases, and its retrospective nature, which made it impossible to follow a homogeneous treatment and follow-up protocol. However, our findings reflect the conditions that clinicians face in real-world clinical practice.

To conclude, low-dose rituximab (500 mg) is effective in treating pemphigus vulgaris and pemphigus foliaceus while preventing serious adverse events. However, further studies are needed to establish the optimal treatment regimen for these patients.

Abstract N°: 1076

Skin involvement in large-vessel vasculitis - a report of three cases

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Introduction & Objectives:

Mucocutaneous manifestations in large-vessel vasculitis (LVV) are uncommon, and literature on this topic remains scarce. However, these dermatological findings may represent the initial presentation of the disease, leading patients to seek dermatological consultation. This report describes three cases in which cutaneous manifestations prompted suspicion of LVV.

Case reports:

Case 1: An 84-year-old woman presented to the Dermatology clinic with blisters and erosions in the frontotemporal region, initially without systemic symptoms. She later developed unilateral headache, and laboratory findings revealed an elevated erythrocyte sedimentation rate. Doppler ultrasound of the temporal arteries demonstrated a positive halo and compression sign, confirming the diagnosis of giant cell arteritis (GCA). The patient was initiated on high-dose corticosteroids, leading to rapid symptom resolution and progressive normalization of inflammatory markers without relapse during tapering.

Case 2: An 83-year-old woman with a history of polymyalgia rheumatica in remission presented to the Dermatology clinic with a chronic lingual ulcer unresponsive to prior treatments, accompanied by a persistent holocranial headache. Physical examination revealed indurated, pulseless temporal arteries. Urgent rheumatological assessment and Doppler ultrasound confirmed GCA. Intravenous methylprednisolone was initiated, followed by tocilizumab as a steroid-sparing agent. The patient experienced complete ulcer resolution and remains in remission on low-dose prednisone and tocilizumab.

Case 3: A 50-year-old woman with a prior diagnosis of Takayasu arteritis (TA) was referred to the Dermatology clinic for painful nodular lesions on both legs. A skin biopsy demonstrated lobular panniculitis with ischemic changes. Treatment was intensified with increased corticosteroid dosage and the addition of tocilizumab, resulting in resolution of dermatologic symptoms.

Discussion:

Cutaneous manifestations in LVV can be classified into ischemic and inflammatory lesions. In GCA, ischemic findings such as erythema, purpura, induration, and blistering in the temporal region may serve as early indicators of disease, with progression to necrosis and ulceration in severe cases. Mucosal involvement, including tongue and lip necrosis, has also been described. Inflammatory lesions such as panniculitis and breast nodules have been reported in extracranial GCA. In TA, lower limb nodules, occasionally progressing to ulceration or mimicking pyoderma gangrenosum, are among the most frequently observed cutaneous manifestations. These lesions have been associated with active TA and require treatment escalation.

Conclusion:

These cases highlight the relevance of dermatologic findings in LVV and the importance of prompt recognition and interdisciplinary management to ensure timely diagnosis and intervention, ultimately reducing morbidity and

preventing complications.

Abstract N°: 1094

Pemphigus Vegetans: A Clinical Rarity Detected from the Emergency Department

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Introduction & Objectives:

Pemphigus Vegetans is a rare variant of pemphigus vulgaris (PV), accounting for 1–2% of cases. Early diagnosis is crucial due to its clinical implications and associated morbidity and mortality. Its acute progression may lead patients to present to Emergency Departments rather than to dermatology outpatient clinics.

Clinical Case:

A 31-year-old male with no relevant medical history presented to the Emergency Department with a three-week history of evolving skin lesions. The Emergency Department requested a consultation with our Dermatology Department. The lesions initially appeared during a trip to Germany, where the patient was diagnosed with bullous impetigo and treated with oral cloxacillin. Physical examination revealed extensive erosive and crusted lesions on the scalp, face, trunk, and extremities, in addition to large, macerated, and foul-smelling vegetating plaques in the inguinal folds, beard area, occipital region, and chest. He also exhibited erosive cheilitis and erosions on the buccal mucosa and tongue, without involvement of other mucosal sites. The suspicion of Pemphigus Vegetans was confirmed by histopathology and direct immunofluorescence, along with serological positivity for anti-desmoglein 1 and 3 antibodies. Treatment was initiated with prednisone and clindamycin to address bacterial superinfection. Following diagnostic confirmation, rituximab was administered with progressive tapering of prednisone, achieving complete remission.

Discussion:

Pemphigus Vegetans is characterized by vegetating plaques predominantly located in flexural areas, although other regions may also be affected, as illustrated in this case. Traditionally, two variants are described: the Hallopeau type and the Neumann type. In both, oral mucosal involvement is common. The disease is primarily driven by autoantibodies against desmoglein 1 and 3; however, other factors such as bacterial or fungal superinfection, semi-occlusion, and genetic predisposition may contribute to the development of vegetating lesions. Rituximab combined with prednisone is considered the first-line treatment for moderate to severe cases.

Conclusion:

Multidisciplinary management and good coordination between services are essential when treating severe dermatological conditions that, as in this case, may require urgent medical attention.

Abstract N°: 1111

Lupus erythematosus tumidus simulating Morbihan Disease, a clinical case.

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Introduction & Objectives

Morbihan syndrome is a rare and chronic dermatological condition characterized by persistent solid facial edema, primarily affecting the upper two-thirds of the face, including the forehead, eyelids, nose, and cheeks. It is often associated with rosacea, leading to diagnostic challenges due to its symptom overlap with other inflammatory skin diseases.

On the other hand, lupus tumidus (LT) is a rare and distinct subtype of cutaneous lupus erythematosus (CLE). It is characterized by photosensitive erythematous plaques without scaling, ulceration, or scarring, predominantly appearing on sun-exposed areas. Histologically, it is distinguished by superficial and deep perivascular and periadnexal lymphocytic infiltration with abundant dermal mucin deposition, notably lacking epidermal involvement. This differentiates it from other lupus variants such as discoid lupus erythematosus (DLE) and subacute cutaneous lupus erythematosus (SCLE). While LT is generally considered benign and self-limiting, its diagnosis is often delayed due to its clinical similarity to other dermatological conditions.

Materials & Methods

A 52-year-old female with a known history of rosacea presented with persistent erythema and non-pitting edema of the upper face, including the eyelids, forehead, and cheeks. These findings were consistent with Morbihan syndrome, a rare complication of rosacea-associated lymphedema.

Her past medical history was unremarkable for autoimmune diseases, and she denied any history of photosensitivity, systemic symptoms (such as joint pain, fever, or fatigue), or previous episodes of facial edema. There was no recent history of infections, allergic reactions, or new medication use that could explain her condition.

Despite initial medical therapy (antibiotics, corticosteroids, and antihistamines), the patient’s symptoms persisted. Consequently, a skin biopsy was performed.

Results

Due to the lack of response to standard treatments for rosacea and Morbihan syndrome, a skin biopsy was performed to evaluate alternative causes of chronic facial edema. The histopathological analysis revealed:

- Superficial and deep perivascular and periadnexal lymphocytic infiltrate.
- Prominent dermal mucin deposition.
- Absence of epidermal involvement (i.e., no atrophy, vacuolar degeneration, or interface dermatitis).

These findings were not consistent with Morbihan syndrome but were instead characteristic of lupus tumidus (LT), a subtype of cutaneous lupus erythematosus (CLE).

Conclusion

This case emphasizes the importance of considering lupus tumidus in patients presenting with chronic facial edema initially thought to be Morbihan syndrome. The diagnosis relies on histopathological confirmation, as the clinical overlap between rosacea-associated lymphedema and cutaneous lupus can be significant. Early recognition and targeted treatment, including antimalarials and photoprotection, may improve patient outcomes.

Abstract N°: 1112

Unilateral heliotrope rash and 'erysipelas-like' facial eruption as a rare but distinctive manifestation of anti-MDA5 dermatomyositis: case report and review of the literature

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Introduction & Case Presentation

A 30-year-old male with a history of alopecia areata was admitted for a four-week duration of a progressive left-sided facial rash with left periorbital swelling. He denied the use of contactants or new medications. On examination, thee was a violaceous erythema and oedema unilaterally affecting his left cheek and eyelids. A computed tomography (CT) of the face was done and was suggestive of a soft tissue infection. He was treated for facial erysipelas and pre-septal cellulitis with one week of antibiotics but failed to improve.

One week later, he developed a dry cough and Gottron's papules on his knuckles. Power examination was normal. A skin biopsy performed over the Gottron's papule revealed interface dermatitis with dermal mucin. Further investigations revealed anti-MDA5 antibodies and raised aldolase. A computed tomography scan of the thorax showed interstitial lung disease. He was diagnosed with hypomyopathic anti-MDA5 dermatomyositis (DMS) with ILD and was treated with prednisolone, tofacitinib and intravenous rituximab with improvement of his rashes and cough.

Conclusion:

We describe this rare but distinctive manifestation of anti-MDA5 DMS featuring a unilateral heliotrope and "erysipelas-like" facial rash. Anti-MDA5 DMS is commonly associated with rapidly progressive ILD, portending high mortality rates despite aggressive immunosuppressive therapy, necessitating the need for early diagnosis and intervention.

In our literature review of previous DMS cases with a similar presentation, 77% of patients had MDA-5 positivity and more than 50% had ILD. With a strong propensity for rapidly progressive life-threatening ILD, physicians need to be cognisant of this peculiar manifestation and have a high index of suspicion for anti-MDA5 DMS to facilitate early diagnosis and aggressive intervention.

Abstract N°: 1160

The Role of T Cell subsets and Related Cytokines in the pathogenesis of Bullous Pemphigoid

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Introduction & Objectives:

Bullous pemphigoid (BP) is an autoimmune blistering disease that significantly compromises the quality of life and can be life-threatening. Our study aims to conduct a comprehensive comparison of the roles of T cell subsets in BP.

Materials & Methods:

Skin immunofluorescence, serum magnetic Luminex assay kits, and peripheral flow cytometry were utilized to identify the distribution characteristics of T cell chemokine receptors and cytokines in patients with BP, AD (Atopic Dermatitis), and PV (Pemphigus Vulgaris) .

Results:

Skin immunofluorescence revealed that, compared to AD and PV patients, Th1, Th2 and Tfh, but not Th17 related chemokine receptors and cytokines were elevated in BP patients. Plasma levels of Th1-related chemokine receptors or cytokines (e.g., CXCL9, CXCL10, CXCL11), Th2-related chemokine receptors or cytokines (IL4, CCL17, CCL18), and Tfh-related chemokine receptors or cytokines (CXCL13) were elevated in BP patients. Peripheral flow cytometry demonstrated that CD4+CXCR5+ Tfh cells were elevated in BP patients compared with healthy controls, whereas CD4+CCR6+ Th17 cells were lower than that in healthy controls.Multivariate analysis showed that BP180 antibodies and CCL17 had a positive correlation with BPDAI (Bullous Pemphigoid Disease Area Index), while CCL20 was negatively correlated with BPDAI.

Conclusion:

BP is a disease characterized by the involvement of multiple immune cells. Our results indicate that Th1, Th2, and Tfh cells, but not Th17 cells, play a significant role in the pathogenesis of BP.

Abstract N°: 1209

Vascular Purpura Revealing Fetal Growth Restriction and Chronic Inflammatory Placental Disorders: A Mandatory Etiological Investigation!

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Introduction & Objectives:

Vascular purpura is a medical emergency requiring rapid etiological diagnosis, especially in pregnant women, where maternal and fetal prognosis may be compromised. Pregnancies complicated by vasculitis linked to systemic diseases are high-risk, with outcomes varying according to the affected vessel size.

Observation:

A 34-year-old gravida 3, para 3, with no significant history, was admitted at 35 weeks of amenorrhea for acral infiltrated petechial purpura evolving over five days, associated with photosensitivity, in an afebrile context with preserved general condition.

Biological investigations ruled out thrombocytopenia and infection. Serologies for TORCH, HIV, and CMV were negative. There was discordance between ESR and CRP, with a negative 24-hour proteinuria test. Immunological tests revealed positive ANA (1/160 titer) and negative anti-DNA antibodies.

Histopathology of a skin biopsy with direct immunofluorescence showed vasculitis with IgG and C3 deposits at the dermo-epidermal junction, forming a lupus band pattern, suggesting cutaneous lupus. A complementary immunological workup was ordered, with follow-up six months postpartum.

Obstetric ultrasound revealed oligohydramnios and intrauterine growth restriction (IUGR), with fetal weight below the 10th percentile. Umbilical Doppler showed a high resistance index (0.77), indicating vascular-related IUGR.

The patient underwent cesarean delivery, followed by postpartum gynecological and dermatological monitoring.

Discussion:

IUGR risk factors include **maternal, placental, and fetal** elements. These factors are not mutually exclusive, and their presence alone **does not always necessitate intervention**.

Women with **systemic vasculitis** can achieve term pregnancies but face **higher risks of late preterm delivery**. Small-vessel vasculitis increases susceptibility to **IUGR flares** and **exacerbation of vascular disease**.

Autoimmune diseases like **lupus** are strongly associated with **adverse obstetric outcomes**, including **preeclampsia, miscarriage, stillbirth, IUGR, and preterm birth**. These risks are **higher in undiagnosed cases**, with **vascular purpura** as a possible **initial manifestation**.

Delayed recognition of **vascular purpura** can significantly impact **maternal and fetal prognosis**. Given its diverse etiologies, particularly **systemic and infectious diseases**, clinicians must **go beyond leukocytoclastic vasculitis diagnosis** and **perform a thorough workup**, especially in pregnant patients.

Conclusion:

This case underscores the **diagnostic significance of early dermatological signs**, such as **vascular purpura**, in detecting **vasculitis during pregnancy**. Recognizing such manifestations enhances **preconception counseling** and optimizes **antenatal, perinatal, and postpartum care**, improving overall outcomes.

Abstract N°: 1269

The complex role of IL-13 in Bullous Pemphigoid: new insights from a retrospective cohort study

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Introduction & Objectives:

Bullous pemphigoid (BP) is the most prevalent autoimmune blistering skin disease, predominantly affecting the elderly. Th2 cytokines, including interleukin-13 (IL-13), are implicated in BP pathogenesis through immune cell recruitment and eosinophil activation. While IL-13 is known to play a role in local inflammation, its systemic relevance and potential as a therapeutic target remain uncertain. This study aimed to compare serum IL-13 levels between idiopathic and DPP4-inhibitor–induced BP, as well as with controls, and to explore associations with disease severity, mucosal involvement, and prognosis.

Materials & Methods:

We conducted a retrospective cohort study of 42 adult BP patients diagnosed between 2008–2023 at a dermatology referral center. Diagnosis was based on clinical features and direct immunofluorescence. Patients were classified as idiopathic or DPP4-inhibitor–induced BP. The control group included 12 healthy individuals and 4 pemphigus vulgaris patients. Serum samples were collected at diagnosis before treatment and analyzed for IL-13 levels using ELISA. Clinical parameters included eosinophil counts, mucosal involvement, adjuvant therapy requirement, and mortality.

Results:

Serum IL-13 levels were significantly lower in BP patients (mean 62.46 pg/mL ±16.53) compared to healthy controls (87.83 pg/mL ±8.87, $p<0.0001$) and pemphigus patients (87.6 pg/mL ±5.16, $p=0.013$). DPP4i-induced BP patients showed lower IL-13 levels than idiopathic BP patients (57.36 pg/mL ±21 vs. 67.01 pg/mL ±14.3, $p=0.0104$). IL-13 levels did not significantly correlate with mucosal involvement ($p=0.338$), eosinophil counts ($r=0.18$, $p=0.253$), need for adjuvant therapy ($p=0.32$), or mortality ($p=0.45$). However, elevated eosinophil levels were associated with a higher likelihood of requiring adjuvant therapy ($p=0.027$).

Conclusion:

Serum IL-13 levels are reduced in BP patients, particularly in those with DPP4i-induced disease, suggesting immunologic differences between idiopathic and drug-induced BP subtypes. The lack of association between IL-13 levels and clinical outcomes limits its utility as a biomarker. These findings underscore the need for further investigation into IL-13's mechanistic role in BP and may inform future personalized therapeutic approaches based on cytokine profiles.

Abstract N°: 1375

Drivers of Health in Adults with Alopecia Areata, Atopic Dermatitis, Hidradenitis Suppurativa, and Psoriasis

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Introduction & Objectives: Research on drivers of health in alopecia areata (AA), atopic dermatitis (AD), hidradenitis suppurativa (HS), and psoriasis (PsO) is lacking. We sought to explore drivers of health and influences on healthcare seeking behaviors in these patient groups.

Materials & Methods: Adults living with self-reported AA, AD, HS, or PsO were recruited through the AmeriSpeak panel, a national sample of US adults, and disease-specific patient advocacy groups. Patients completed an electronic one-time electronic survey which included questions on race/ethnicity, overall disease severity (past month), drivers of health, influences on seeing a healthcare provider (HCP), and HCP type. Data are reported descriptively.

Results: Overall, 1,284 adults (n=156 AA, n=260 AD, n=583 HS, n=285 PsO) responded to the survey. Self-identified race included 52.6% White, 32.9% Black/African American, 4.0% Asian, 10.4% American Indian/Alaskan Native, Native Hawaiian/Other Pacific Islander, or multiple races; 14.1% identified as Hispanic/Latino ethnicity. Disease severity (mild, moderate, and severe) was reported for AD (49.6%, 43.1%, 7.3%), HS (16.1%, 49.1%, 34.8%), and PsO (53.0%, 36.8%, 10.2%). Severity of AA patients was reported as percent of the scalp affected: 6% to 20% (16.7%), 21% to 49% (19.2%), ≥50% (64.1%). 54.5% AA/ 49.2% AD/ 47.9% HS/ 56.8% PsO indicated that a dermatologist was the HCP they primarily saw for medications to treat their condition, followed by either primary care providers (33.1% AD/ 26.9% of HS/ 21.8% PsO), or no provider (35.3% AA). When queried about drivers of health, 24.4% AA/ 22.6% AD/ 36.8% HS/ 26.6% PsO reported they or a family member were unable to get medicine or healthcare services in the past year, 13.5% AA/ 17.7% AD/ 33.2% HS/ 17.4% PsO reported they or a household member were unable to get food in the past year, and 12.8% AA/ 13.1% AD/ 17.8% HS/ 20.0% PsO reported being worried about losing their housing. Almost one in five HS and PsO patients reported social media/online groups influenced them to see a HCP for their condition, while < 4% of AA and AD patients reported influence from social media. Of those, 79.6% HS/96.2% PsO reported being influenced by Facebook, and 25.0% HS/98.1% PsO were influenced by TikTok.

Conclusion: We found multiple factors, including drivers of health related to food, healthcare access, and housing stability, can affect patients with AA, AD, HS, and PsO, and likely contribute to varying health outcomes. The study highlights a need for individual patient-centered care integrated with an understanding of drivers of health to optimally reach and treat patients.

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Abstract N°: 1409

Dupilumab in pemphigus: case series and literature review

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Introduction & Objectives:

Pemphigus vulgaris (PV) and pemphigus foliaceus (PF) are rare autoimmune blistering diseases. The standard treatment for PV includes systemic corticosteroids, immunosuppressants, intravenous immunoglobulins, and rituximab. In PF, dapsone can also be used.

Materials & Methods:

We present three cases of PV and PF treated with dupilumab.

Case 1. A 68-year-old male with a history of type II diabetes mellitus and recently diagnosed invasive urothelial carcinoma was diagnosed with PV. He was initially treated with oral prednisone (1 mg/kg/day in a tapering regimen), combined with dupilumab (600 mg loading dose, followed by 300 mg every two weeks). After one month, most lesions were residual or in a re-epithelialization phase. At six months of follow-up, the patient remained free of new PV flares.

Case 2. A 63-year-old female with a history of dyslipidemia, minor thalassemia, and Lynch syndrome was diagnosed with PV. She had previously been treated with systemic corticosteroids, azathioprine, methotrexate, and rituximab, all discontinued due to inefficacy. Dupilumab was initiated (600 mg loading dose, followed by 300 mg every two weeks). After two months, she exhibited marked clinical improvement, although isolated active lesions persisted on the upper gingival margin and soft palate. At four months, she continued to show progressive improvement with reduced disease activity.

Case 3. A 57-year-old male with a history of past hepatitis B infection and obesity was diagnosed with PF. He had previously received systemic corticosteroids, azathioprine, mycophenolate mofetil, and dapsone, all of which were discontinued due to inefficacy or adverse effects. Treatment with dupilumab (600 mg loading dose, followed by 300 mg every two weeks) was initiated. However, after six months, he continued to experience recurrent flares with extensive lesions in seborrheic areas, leading to treatment discontinuation.

Results:

Th2-driven immune mechanisms, particularly interleukin-4 (IL-4), play a crucial role in the pathogenesis of PV and PF. Recent reports have documented nine cases of patients with pemphigus treated with dupilumab: six with PV and three with PF.

Among the six PV cases, five showed rapid improvement of the erosions and blisters. Two remained disease-free for over a year, while one patient continued to develop lesions after four weeks, leading to treatment discontinuation.

Regarding the three PF cases, all demonstrated an initial response to dupilumab. One patient remained in sustained remission for over a year.

Dupilumab regimens varied among patients, with some following dosing schedules similar to those used in atopic dermatitis.

These findings suggest that Th2-mediated pathways contribute significantly to pemphigus activity and may explain the favorable response to dupilumab observed in some patients.

Conclusion:

Dupilumab appears to be a promising therapeutic alternative for PV and PF in patients refractory to conventional immunosuppressive therapies or in those with contraindications to standard treatments, including rituximab. Further studies are needed to evaluate its efficacy.

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Abstract N°: 1427

Cutaneous manifestations of thymoma-associated multi-organ autoimmunity (TAMA) – case report and literature review

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Cutaneous manifestations of thymoma-associated multi-organ autoimmunity (TAMA) – case report and literature review

Introduction & Objectives:

We report a case of papulosquamous eruption secondary to thymoma associated multi-organ autoimmunity (TAMA). The patient is an elderly Chinese male who was diagnosed with seropositive myasthenia gravis since November 2022. He was treated with systemic prednisolone, pyridostigmine, intravenous immunoglobulin and rituximab. His Computed tomography (CT) scan showed the presence of an anterior mediastinal mass, consistent with a thymoma. He underwent an open thymectomy in December 2024 but there was resident tumour adherent to his CABG graft implantation site. He was admitted for pneumonia in December 2024 and Dermatology was referred for itchy generalised rashes of two months duration. There is no personal or family history of atopy and no contactants were elicited. On examination, there were multiple well circumscribed scaly erythematous to hyperpigmented papules, predominantly over the limbs, and scattered on the anterior trunk and back. Palms, soles, scalp and the genital region were clear.

Materials & Methods:

Results:

A skin punch biopsy revealed epidermal acanthosis, parakeratosis and multiple scattered cytoid bodies but there was no separation of the epidermis from the dermis. VDRL and TPPA returned negative. The patient was diagnosed with papulosquamous eruption secondary to TAMA.

Conclusion:

He was continued on prednisolone 15mg OM for his myasthenia gravis, and started on adjunctive topical corticosteroid therapy. The patient was subsequently lost to follow-up as he returned to his home country for further treatment.

TAMA is a rare paraneoplastic syndrome with a total of 47 cases reported thus far. Skin involvement is the most frequent manifestation of TAMA, but can present variably as papulosquamous eruptions, lichenoid dermatitis, pemphigus-like lesions or erythroderma. The cutaneous manifestations are often accompanied by other systemic autoimmune symptoms, such as liver dysfunction and enteritis. Histopathologically, TAMA is characterized by features such as epidermal acanthosis, parakeratosis, individual cell keratinization, liquefaction degeneration, and intraepidermal infiltration of CD8-positive lymphocytes. The pathogenesis of TAMA involves a breakdown of immune tolerance, likely due to inadequate T cell selection in the thymic tumor environment, leading to the activation of autoreactive T cells. This immune dysregulation results in the characteristic The pathogenesis of TAMA involves a breakdown of immune tolerance, likely due to inadequate T cell selection in the thymic tumor environment, leading to the activation of autoreactive T cells. Treatment options of TAMA include thymectomy, systemic corticosteroids, immunosuppressive agents such as ciclosporin, intravenous immunoglobulin, and

biologics such as rituximab and tocilizumab. For patients with localized skin involvement, topical corticosteroids and phototherapy can be used to reduce inflammation and manage symptoms.

Abstract N°: 1446

A Retrospective Analysis of Clinical Presentations in Anti-SAE Subtype Dermatomyositis: A Case Series from India

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Introduction & Objectives:

Anti-small ubiquitin-like modifier activating enzyme (SAE) antibody, one of the recently described autoantibodies, is rare in dermatomyositis (DM). Studies from North America, Europe, and China have highlighted a predominance of cutaneous features with relatively mild muscle involvement. This study aims to delineate the cutaneous, muscular and systemic manifestations of anti-SAE positive DM in an Indian population.

Materials & Methods:

A retrospective study was conducted between January 2014 and December 2024 on clinically suspected adult DM patients who attended the Dermatology outpatient department of a tertiary care hospital in South India. Demographic, clinical, laboratory, and treatment data of patients with anti-SAE positive DM were analyzed.

Results:

Of the 194 patients, 134 of them underwent antibody testing for myositis via the line immunoassay. Among the 101 patients who tested positive for myositis-specific or associated antibodies, six (5.9%) patients were strongly positive for anti-SAE antibodies. The mean age of onset was 46.6 years (SD ±8.7), with a female predominance (66.6%). All patients presented initially with cutaneous symptoms such as rash, redness, itching and photosensitivity. All patients had periorbital oedema and heliotrope rash. Gottron's sign was seen in four patients; malar rash, 'V' sign, shawl sign, Samitz sign, and diffuse alopecia in three patients; Gottron's papules in two patients; holster sign and mechanic's hands in one patient. Proximal muscle weakness was observed in five patients (83.3%), while one patient had clinically amyopathic DM. Systemic involvement included interstitial lung disease and dysphagia seen in one patient each. No malignancies were detected. Electromyography (n=4) showed myopathic changes in three cases. Only one out of the six patients had elevated creatinine phosphokinase levels at the time of presentation, while all the five patients tested for lactate dehydrogenase had elevated levels. Skin biopsies (n=3) were done which revealed interface changes with dermal mucin deposition. All patients were treated with corticosteroids; methotrexate (5/6) and hydroxychloroquine (3/6) were used as adjunctive therapies.

Conclusion:

In this study, anti-SAE positive DM showed predominant cutaneous features with variable myopathy, aligning with international data. These findings highlight a distinct clinical phenotype that merits further investigation in diverse populations.

Abstract N°: 1573

Tattoo as a cause of ASIA syndrome

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Introduction & Objectives:

The autoimmune/inflammatory syndrome induced by adjuvants (ASIA) is a rare condition caused by an immune response associated with overreactivity of the immune system triggered by adjuvants. The most common adjuvants are aluminium salts, but they can also be bioimplants or infectious agents. ASIA may lead to the development of various autoimmune diseases.

Materials & Methods:

I want to present two female patients who developed ASIA syndrome after tattoo and hyaluronic acid procedures.

The first patient, a 26-year-old woman, developed SLE, likely induced by ASIA syndrome, after hyaluronic acid procedures. She was admitted because of arthralgia and fever with butterfly-shaped erythema on her face and erythematous and infiltrative skin lesions on the posterior surface of the thighs and buttocks. The laboratory tests revealed leucopenia and lymphopenia, normocytic anaemia, positive ANA in a titer of 1:5120 granular, 1:320 cytoplasmic, and homogeneous 1:5120, anti-U1RNP++, anti-Ro-52+, anti-SS-A+, anti-dsDNA++, anti-nucleosome antibodies++, and anti-histone antibodies++, reduced levels of complement components C3 and C4. The patient fulfilled the ACR/EULAR 2019 classification criteria of the SLE. Due to exposure to numerous adjuvants like tattoo ink, hyaluronic acid, and piercing and the development of the delayed inflammatory reaction (DIR) to hyaluronic acid (HAF), the patient also fulfilled the criteria of ASIA. The patient was treated with non-steroidal anti-inflammatory drugs (NSAIDs), steroids, hydroxychloroquine, and cyclosporine, which resulted in an improvement in the general condition, resolution of swelling and joint pain, and improvement in skin lesions.

The second patient, a 46-year-old woman, has had numerous coloured tattoos for 7 years and had an overwent lip correction with HA 7 years ago. Three months after the last colored tattoo with a contact reaction to the violet dye, she suddenly noticed Raynaud’s phenomenon, low-grade fever, muscle pain, significant weakness, sleep and memory problems, symptoms of dry mouth and conjunctiva, symptoms of paresthesia in the lower limbs and rapid fatigue. CT revealed signs of lung fibrosis. An uncharacterised connective tissue disease was diagnosed as a result of ASIA syndrome, for which the patient met the criteria.

Results:

Both patients fulfilled the criteria of ASIA syndrome. In addition, as in these cases, it can develop several years after the procedure, making it difficult for both patient and physician to become aware of the connection.

Conclusion:

ASIA syndrome after tattoo and aesthetic procedures is still underdiagnosed, probably due to ignorance or diagnostic difficulties, as symptoms are uncharacteristic and there is no immunological marker for this syndrome. Early diagnosis requires a multidisciplinary approach and may require immunosuppressive treatment in specific cases.

Abstract N°: 1636

Genetic Signatures in Pemphigus Vulgaris: Decoding Immune Susceptibility through HLA Patterns and FCGR2B Variants

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Introduction & Objectives:

Pemphigus vulgaris (PV) is a rare autoimmune blistering disorder with a global annual incidence of 0.5–32 per million and 4.7 per million in Turkey. Though, being a relatively well understood disease, the initiation of the autoimmunity still warrants elucidation. It is generally accepted that polymorphic residues in the MHC class II β-chain play an important role in the selective binding of desmoglein epitopes, which are presented by antigen presenting cells to Dsg3-specific $CD4^+$ T cells, subsequently promoting class switching in Dsg3-specific B cells. Among Fcγ-receptors, the FcγRIIB was the only inhibitory one which is mainly expressed on B cells and primarily suppresses BCR signaling. In transgenic mice model, FcγRIIB was shown to inhibit pathogenic class switching of Dsg-3-specific B cells. Vice versa, attenuated FcγRIIB function may be a predisposing factor for PV, corresponding to the previous evidence for an association between *FCGRIIB*-I232T (rs1050501) polymorphism and systemic lupus erythematosus. Thus, this study aimed to investigate the effects of the rs1050501 variant in the *FCGR2B* gene and the *HLA-DRB1* gene allelic polymorphisms on the genetic etiology of PV.

Materials & Methods:

This single-center genetic epidemiology case-control study included adult PV patients diagnosed via clinical, histopathological, direct immunofluorescence microscopy and/or ELISA methods, and age, sex- and ethnicity-matched healthy controls without known autoimmunity, with power set at 80% (ES=1.5). The Hardy-Weinberg (H-W) equilibrium test was performed on controls using rs1050501 SNP alleles. Analysis of rs1050501 was performed through a two-step approach involving long-PCR followed by nested PCR, with the final PCR product analyzed via Sanger sequencing. *HLA-DRB1* genotyping was conducted using the PCR-SSOP-Luminex technique and analyzed by the LABScan3D platform. Demographic, clinical data and comorbidities were recorded. All statistical analysis performed by Chi-square and Fisher's exact test on open source RStudio (v.2024.04.2) platform using dplyr, tidyr, ggplot2, writexl and openxlsx packets. A Benjamini-Hochberg (FDR) corrected p-value ($p\ adj < 0.05$) was considered statistically significant.

Results:

A total of 91 PV and 200 healthy controls were analyzed. The H-W equilibrium test showed that rs1050501 variant alleles frequency was in equilibrium ($p > 0.05$). Thirty-three different HLA-DRB1 alleles were observed between groups. HLA-DRB1*04:02 and *14:01 alleles were significantly more frequently detected in PV patients compared

to the controls (**Table 1, Figure 1**). A significantly higher of the HLA-DRB1*16:01 allele was observed in controls as protective from PV. It was a very interesting observation for us that the HLA-DRB1*04:02/14:01 (compound heterozygous) genotype was seen only in PV patients (**Table 2**). Between the patients and controls, there was no statistically significant difference in terms of rs1050501 variants and genotypes (adj.p=0.12 and adj.p=0.35, respectively).

Conclusion:

HLA-DRB1*04:02 and 14:01 alleles and* HLA-DRB1*04:02/14:01* genotype were significantly more frequent in PV patients. To the best of our knowledge, this study is the first to demonstrate that the HLA-DRB1*04:02/14:01 genotype is significantly more frequent in PV patients compared to healthy individuals, based on a relatively large cohort. However, no association was found between rs1050501 and PV.

Table 1. HLA-DRB1 alleles and Chi-square test statistics

DRB1*	Controls (n)	PV (n)	p_value	p_adj	Odds_Ratio (OR)	95% CI
04:02	18	70	2.03×10^{-24}	**6.71×10^{-23}**	12.95	7.40-24.57
14:01	17	31	7.51×10^{-07}	**1.23×10^{-05}**	4.55	2.35-8.74
16:01	26	0	9.75×10^{-05}	**0.001**	0.03	0-0.31
11:01	39	6	0.00	**0.037**	0.33	0.10-0.75
14:04	0	4	0.00	0.06	20.19	1.46-Inf
15:01	33	6	0.02	0.10	0.40	0.12-0.91
13:03	12	0	0.02	0.10	0.08	0-0.77
07:01	37	7	0.03	0.13	0.41	0.17-0.97
09:01	8	0	0.06	0.22	0.12	0-1.27
04:01	16	1	0.07	0.23	0.19	0.029-1.14
11:04	39	9	0.07	0.23	0.50	0.22-1.09
08:01	0	1	0.09	0.25	6.61	0.40- Inf
11:11	0	1	0.09	0.25	6.61	0.40- Inf
03:01	26	5	0.12	0.28	0.43	0.16-1.23
04:05	6	0	0.18	0.37	0.16	0-1.86
04:08	5	0	0.18	0.37	0.19	0-1.86
15:02	12	1	0.24	0.44	0.25	0.03-1.62
10:01	11	2	0.24	0.44	0.46	0.03-1.64
04:04	4	4	0.26	0.46	2.22	0.40-12.06
04:03	9	7	0.30	0.49	1.76	0.59-5.10
04:07	3	0	0.31	0.49	0.31	0-3.34
13:02	12	7	0.46	0.69	1.3	0.51-3.98
13:01	19	6	0.51	0.74	0.72	0.21-1.72
01:01	23	7	0.55	0.76	0.68	0.27-1.67
08:04	2	1	0.59	0.78	1.31	0.15-30.33
01:02	3	2	1	1	1.57	0.1-7.80
04:06	1	0	1	1	0.72	0-11.78
13:05	1	0	1	1	0.72	0-11.78
16:02	1	0	1	1	0.72	0-11.78
08:02	2	0	1	1	0.43	0-11.70
08:03	7	3	1	1	1.02	0.24-4.15
11:03	6	1	1	1	0.50	0.07-4.01
12:01	2	0	1	1	0.43	0-11.71

Table 2. Six representative genotypes observed in study groups

DRB1* Genotype	Controls (n)	PV (n)	a*	b*	c*	d*	p	p_adj	OR	95% CI
04:02 / 14:01	0	20	20.5	71.5	0.5	200.5	1.2×10^{-11}	**2.62×10^{-9}**	115.0	13.1-inf*
01:01 / 04:02	0	5	5.5	86.5	0.5	200.5	0.0	0.003	25.5	2.7-inf
04:02 / 04:03	0	6	6.5	85.5	0.5	200.5	0.0	0.003	30.5	2.7-inf
04:02 / 13:02	0	6	6.5	85.5	0.5	200.5	0.0	0.003	30.5	2.7-inf
04:02 / 04:04	0	3	3.5	88.5	0.5	200.5	0.0	0.200	15.9	1.5-inf
04:02 / 13:01	0	4	4.5	87.5	0.5	200.5	0.0	0.200	20.6	1.5-inf

* Correction was made by adding 0.5 to the cell frequencies (Haldane-Anscombe correction). Therefore, the upper limit of CI is infinite.

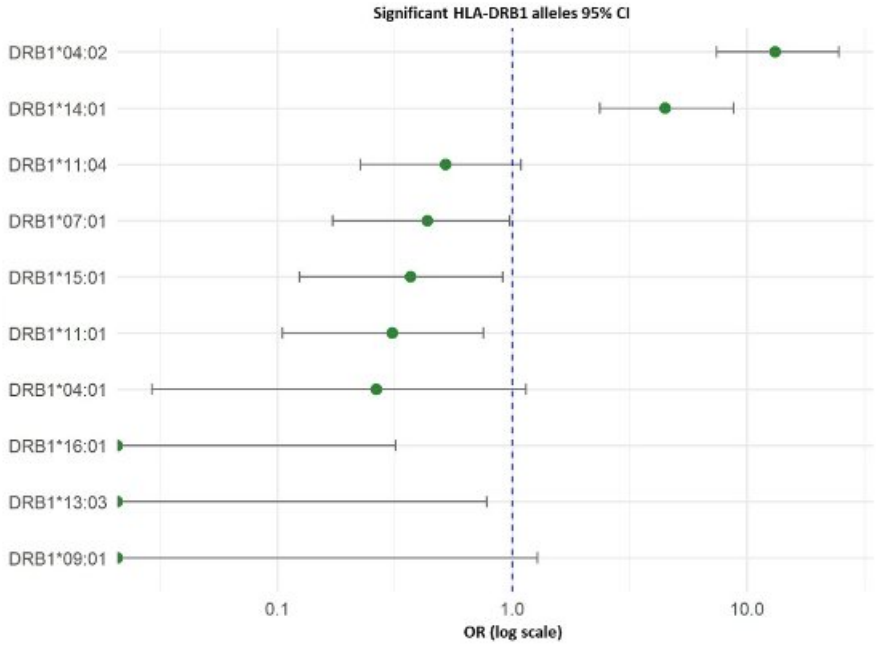

Figure 1. Ten most significant HLA-DRB1 alleles between groups

Abstract N°: 1821

Systemic lupus erythematosus onset with purpura fulminans: a rare presentation. Literature review and case report.

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Systemic lupus erythematosus onset with purpura fulminans: a rare presentation. Literature review and case report

Introduction and Objectives: Purpura fulminans is a rare, life-threatening condition characterized by rapidly progressive skin necrosis and thrombotic vasculopathy. Its occurrence as the first manifestation of systemic lupus erythematosus is extremely uncommon. When associated with the antiphospholipid antibody syndrome, the clinical course is often more severe and its management more complex. We report a case of purpura fulminans as the initial clinical presentation of systemic lupus erythematosus, associated with class IV lupus nephritis and secondary antiphospholipid antibody syndrome, emphasizing the diagnostic approach, histopathological findings, and immunosuppressive treatment that led to patient recovery.

Materials and Methods: A clinical, immunological, histopathological, and therapeutic evaluation was conducted in a 23-year-old woman presenting with rapidly evolving necrotic cutaneous lesions. The diagnosis was based on the criteria for systemic lupus erythematosus, skin and kidney biopsy results, and the response to multidisciplinary treatment.

Results: A previously healthy 23-year-old woman presented with fever, general malaise, and painful purpuric lesions on her limbs. Within 48 hours, these lesions progressed to retiform purpura, hemorrhagic blisters, and skin necrosis. Skin biopsy revealed thrombotic vasculopathy with extensive epidermal necrosis. During hospitalization, she met multiple clinical and immunological criteria for systemic lupus erythematosus, including malar rash, non-scarring alopecia, oral ulcers, cytopenias, hypocomplementemia, high-titer antinuclear antibodies, and positive anti-double-stranded DNA and anti-Smith antibodies. Kidney biopsy confirmed diffuse proliferative glomerulonephritis, corresponding to class IV lupus nephritis. The presence of anticardiolipin antibodies confirmed a secondary antiphospholipid antibody syndrome. Treatment included intravenous pulses of methylprednisolone, followed by oral prednisone and intravenous cyclophosphamide following the Euro-Lupus protocol. Supportive measures and prophylactic anticoagulation were added. The patient showed clinical improvement, with resolution of cutaneous lesions, hematologic recovery, and stabilization of kidney function.

Discussion: Purpura fulminans is an extremely rare dermatological presentation of systemic lupus erythematosus. Its pathogenesis involves immune complex deposition, endothelial activation, consumption of coagulation factors, and a prothrombotic state, particularly when antiphospholipid antibodies are present. In this case, the coexistence of acute cutaneous lupus and antiphospholipid antibody syndrome contributed to the severity of cutaneous and renal manifestations. Early diagnosis and initiation of aggressive immunosuppression and anticoagulation are crucial.

Conclusion: Purpura fulminans may represent the first clinical manifestation of severe systemic lupus erythematosus, especially when associated with antiphospholipid antibody syndrome. Early recognition, histopathological confirmation, and prompt immunosuppressive therapy are essential to prevent life-threatening

complications.

Abstract N°: 1837

Epidemiological and clinical characteristics of cutaneous lupus erythematosus

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Introduction & Objectives:

Lupus erythematosus (LE) is a chronic disease of connective tissue, with heterogeneous clinical manifestations and production of a wide variety of autoantibodies. It can be confined to the skin, without systemic involvement, or involve any organ of the body, and in severe cases become life-threatening. Cutaneous forms of LE appear in 72-85% of patients with systemic LE (SLE), and are the first expression in 25% of them, although they can occur at any stage of the disease.

Objectives: To determine epidemiological and clinical characteristics of different subtypes of cutaneous lupus erythematosus (CLE). To analyze the prevalence and factors associated with the presence of systemic lupus erythematosus (SLE).

Materials & Methods:

Descriptive cross-sectional prevalence study of 260 patients with CLE, April 2013-August 2016. The European Society of Cutaneous Lupus Erythematosus (EUSCLE) Core Set Questionnaire was used to characterize the patients and the illness.

Results:

Of a total of 260 patients with CLE, 183 were female (70.4%) and 77 were male (29.6%), with a female:male (F:M) ratio of 2.4:1. Most patients had a single subtype of CLE (76.2%) while the remainder had more than one subtype. The most prevalent CLE subtypes were discoid LE (DLE) (36.5%), with localized form in almost 2/3 of the cases, and intermittent (ICLE)/tumidus (LET) (28.5%). Subacute cutaneous LE (SCLE) occurred in 14.4% and acute cutaneous LE (ACLE) in 7.1%. The 82.7% of patients had only CLE and 17.3% also had SLE criteria. Patients with CLE without SLE Vs. CLE with SLE had a decade later disease onset, and a lower F:M ratio (2:1 Vs. 8:1). The odds of SLE in women was 4 times higher than in men (OR: 4.03; 95% CI: 1.15-10.65; p=0.003). Compared with ICLE/LET, the odds of developing SLE in ACLE was 200-fold higher (OR: 206.33; 95% CI: 32.02-1335.87); 8.5-fold higher in SCLE (OR: 8.52; 95% CI: 1.67-43.38) and 6-fold higher in patients with DLE (OR: 6.36; 95% CI: 1.44). Non-specific lesions were 12 times more likely to be observed in patients with SLE than in non-SLE. SLE was associated with antinuclear antibodies (ANA) > 1/160 (OR: 10.58) and anti-RNP (OR: 32.84), while body mass index (BMI) was protective (OR: 0.76).

Conclusion:

The most prevalent subtypes of CLE subtypes were DLE and ICLE/LET. The F:M ratio was lower, and the disease onset was later in CLE patients without SLE criteria. The presence of SLE in CLE patients was associated with ANA and anti-RNP positive antibodies, and inversely with BMI.

Abstract N°: 1890

Topical gluconolactone induces immune tolerance in cutaneous lupus erythematosus in human and mice

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Introduction & Objectives:

Dysfunctional regulatory T (Treg) cells are a hallmark of systemic (SLE) and cutaneous lupus erythematosus (CLE), yet therapies targeting Treg stability and function remain scarce. Here we report a translational study employing murine models (MRL.lpr and IMQ), human in vitro experiments, and clinical applications in patients identifying a novel approach to restore immune tolerance.

Materials & Methods:

Phosphoproteomic and metabolomic profiling identified gluconolactone (GDL), a pentose phosphate pathway metabolite, as a key promoter of Treg differentiation. GDL was tested prophylactically and therapeutically in short-term imiquimod-induced skin mouse models, topically and systemically in MRL.lpr lupus-prone mice, in vitro human SLE T cells, and topically in CLE patients.

Results:

In vitro, GDL enhanced murine (i)Treg differentiation and suppressive function by upregulating Foxp3 and promoting p-Stat5 while suppressing Th17 differentiation. In IMQ and MRL.lpr models, topical GDL significantly reduced cutaneous inflammation, boosted Treg suppressive function, and inhibited Th17 responses. This was confirmed in ex vivo experiments using PBMCs from SLE patients including significant reduction of IFNg and IL17A mRNA expression in CD4+ T cells. In therapy-refractory CLE patients, topical GDL improved significantly clinical and histological skin features within two weeks.

Conclusion:

GDL enhances Treg differentiation and suppresses Th17-driven inflammation in vitro and in vivo, in humans and mice. These findings highlight the therapeutic potential of topical GDL in addressing immune dysregulation in CLE and other autoimmune diseases.

Abstract N°: 1901

Low-Dose Interleukin-2 Restores Immune Tolerance and Drives Rapid Repigmentation in Vitiligo by Enhancing Regulatory T Cell Function

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Introduction & Objectives:

Regulatory T (Treg) cell dysfunction is implicated in vitiligo pathogenesis, yet no therapies specifically targeting Treg stability and function have been established. We hypothesize that restoration of immune tolerance by low-dose interleukin-2 (IL-2LD) is effective in treating vitiligo.

Materials & Methods:

Here we report a translational study involving 7 patients with therapy-refractory generalized non-segmental vitiligo. Each patient received four cycles of IL-2LD (1.5 million international units of aldesleukin daily for 5 days) at weeks 0, 3, 6, and 9. A multi-omics approach was applied to analyze blood (PBMCs and serum) and skin biopsies taken at baseline and after cycle 4 (week 9). PBMCs were assessed using CITE-seq (single-cell RNA sequencing and proteomics) and TCR sequencing, while skin samples underwent single-cell RNA sequencing and spatial transcriptomics. Functional Treg suppression assays were conducted.

Results:

All patients exhibited significant repigmentation at 12 weeks, confirmed by Wood-light photography. In blood, Treg cells expanded in all patients, with a concurrent reduction in $CD8^+$ T cells in 6 of 7 patients, leading to an increased Treg/$CD8^+$ cell ratio. Treg cells showed upregulation of stability and suppressive markers, including FoxP3, CD25, CTLA-4, and IKZF2. In skin, Treg cells also increased in number and functionality.

Conclusion:

These findings underscore the pivotal role of Treg cells in vitiligo pathogenesis and demonstrate that IL-2LD can restore immune tolerance by enhancing Treg cell function and stability in both blood and skin. This study highlights the promising therapeutic potential of IL-2 in vitiligo treatment.

Abstract N°: 1923

Pemphigoid with autoantibodies against laminin 332 without mucosal involvement: A Case Report.

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Introduction & Objectives:

Pemphigoid with positive autoantibodies against laminin 332, without mucosal involvement, is a rare, autoimmune, bullous dermatosis characterized by subepidermal blisters and the presence of autoantibodies against laminin 332. Laminin 332 is a glycoprotein which plays a key role in the adherence of the lamina lucida and lamina densa. The disease is of particular clinical significance, as it is often associated with underlying malignancies, necessitating patient's thorough investigation to exclude neoplastic disease.

Materials & Methods:

An 85-year-old woman with a history of type 2 diabetes mellitus, hypertension, and chronic kidney failure was admitted to our hospital due to the appearance of both intact and cracked bullae on an erythematous base. Lesions were mainly on edematous extremities of the hands and feet accompanied by itching. Bullae were also scarcely present on forearms, trunk, thighs, and genitals. The appearance of lesions was reported to begin six months ago, and there was no involvement of the mucous membranes.

Results:

To investigate the disease, a biopsy was obtained and sent for histopathological examination and direct immunofluorescence (DIF). Findings we in accordance to subepidermal bullous and linear deposition of IgG(+) and C3(+++) along the basement membrane zone respectively, with a negative profile of autoimmune bullous dermatopathies (Elisa technique), while salt split skin revealed linear choroidal fixation (floor pattern) of IgG globulin along the basement membrane zone with positive antibodies against laminin 332 on the Biochip Dermal Mosaic using indirect immunofluorescence (IIF). Above findings confirmed the nosological entity of pemphigoid against laminin 332. A gross biochemical test revealed impaired renal function, while imaging - including chest, upper and lower abdomen CT scans, mammography, pap smear, and thyroid ultrasound - revealed no evidence of neoplastic disease.

Conclusion:

Patient was treated with both systemic and local corticosteroids, starting with an initial dose of 0.5 mg/kg per day using a cortisone tapering regimen, antihistamines, and Dupilumab. Diagnosis and treatment in each case must be individualized, as the disease may have an aggressive course, due to the presence of a coexisting malignancy. Advances in immunological and molecular techniques have contributed significantly towards improving diagnostic accuracy, while newer treatment options facilitate a more targeted and effective management.

Abstract N°: 2008

When Urticarial-like Lesions Do Not Represent Common Urticaria: A Case Report of McDuffie Syndrome

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Introduction & Objectives:

McDuffie syndrome, also known as hypocomplementemic urticarial vasculitis (HUV), is a rare disorder characterized by persistent urticarial skin lesions, hypocomplementemia, and often systemic involvement. It is a small-vessel vasculitis mediated by immune complexes that trigger vascular inflammation and activation of the complement system, leading to depletion of C1q, C3, and C4. First described by McDuffie et al. in 1973, HUV is distinct from chronic idiopathic urticaria (CSU).

Materials & Methods:

A 36-year-old female with an 11-year history of recurrent cutaneous lesions was referred with a diagnosis of systemic lupus erythematosus. She experienced daily episodes of erythematous, slightly elevated plaques with mild pruritus, lasting over 24 hours and leaving residual hyperpigmentation. The lesions affected the upper trunk and proximal limbs. Laboratory tests revealed hypocomplementemia (C3: 26 mg/dL; C4: <8 mg/dL; C1q: <0.58; CH50 <12.5 U/mL) and positive ANA at a titer of 1:320 with a coarse speckled pattern. Urinalysis showed mild proteinuria and microscopic hematuria. Serological tests for hepatitis B, C, syphilis, and HIV were negative. Skin biopsy revealed vascular dilation, endothelial swelling, erythrodiapedesis, and a subtle neutrophilic and lymphocytic infiltrate consistent with urticarial vasculitis. Unfortunetly, anti-C1q antibodies was not avaliable in our institution.

Results:

McDuffie syndrome is marked by urticarial lesions differing from common urticaria in duration and severity. Systemic symptoms such as fever, malaise, arthralgia, and myalgia are frequent. Hypocomplementemia—especially of C1q, C3, and C4—indicates classical complement pathway activation. Anti-C1q antibodies are often present and support diagnosis. ANA may be positive without meeting full criteria for systemic lupus erythematosus (SLE), though overlap or progression may occur. Systemic involvement may include non-erosive arthritis, COPD, and renal manifestations such as hematuria or glomerulonephritis. Gastrointestinal and neurological symptoms may occur, and rare ocular signs include episcleritis or uveitis.

Conclusion:

Hypocomplementemic urticarial vasculitis is a rare but potentially severe condition that can mimic benign dermatoses and delay diagnosis. Early recognition of persistent urticarial lesions, associated hypocomplementemia, and histological signs of leukocytoclastic vasculitis is key. Long-term follow-up is essential due to systemic risk and possible evolution into autoimmune disease.

Abstract N°: 2063

Pemphigus Foliaceus with Rituximab-Induced Anaphylaxis Successfully Managed with Drug Desensitization

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Introduction & Objectives:

Pemphigus foliaceus (PF) is an intraepidermal autoimmune bullous disease with potentially severe and fatal complications. Refractory cases may be successfully treated with rituximab (RTX). However, significant adverse reactions to RTX including anaphylaxis may pose a therapeutic challenge. Rapid drug desensitization (RDD) has been employed in such scenarios where the immunobiologic is crucial for disease control. We report a case of PF with RTX-induced anaphylaxis that was successfully managed with RDD.

Materials & Methods:

A 40-year-old male presented with a 3-year history of vesicles, erosions, and erythematous-squamous-crusted plaques on the face, trunk and thighs. Histopathology and direct immunofluorescence confirmed the diagnosis of PF.

PF remained active after prednisone up to 1 mg/kg/day and mycophenolate mofetil 3g/day were introduced. A first cycle of RTX was administered in two 1 g infusions, leading to partial disease control. The patient received a second RTX cycle after 6 month, and developed generalized wheals, hypotension and bronchospasm. RTX was discontinued, and methylprednisolone and diphenhydramine were administered with complete resolution of anaphylaxis. Mycophenolate mofetil was replaced with azathioprine 200 mg/day without improvement of lesions within six months.

Due to refractoriness of PF to immunosuppressive therapy and previous efficacy of RTX, RDD was performed. Intradermal and prick tests were initially performed and resulted negative. Pre-medication included metamizole 1 g and diphenhydramine 100 mg, followed by RTX infusion using three increasing concentrations - 0.005, 0.1 and 4 mg/mL - with clinical evaluations between doses. Diphenhydramine 50 mg/mL was repeated between the second and third doses. The patient received the total dose of RTX 1 g without adverse events. Three additional infusions of RTX 1 g were administered, each preceded by RDD, with significant improvement.

Results:

PF is an autoimmune blistering disease caused by IgG autoantibodies against Dsg1, with potential severity. Therefore, achieving remission through effective therapy is imperative.

First-line treatment includes systemic corticosteroids, and immunosuppressants in severe cases. RTX, a chimeric anti-CD20 monoclonal antibody, is recommended in refractory PF. While generally well tolerated, RTX can cause anaphylaxis, which is an extreme form of type I hypersensitivity.

RDD is an individualized approach that allows reintroduction of essential drugs in patients with prior hypersensitivity reactions. It is performed in stages, such as risk stratification and eligibility assessment; premedication; and administration of the drug at increasing doses within defined intervals, with close monitoring.

Indications for RDD include the hypersensitivity reaction types I, IV and due to cytokine release, in cases where the

drug is irreplaceable or has a unique mechanism of action. Contraindications include type II and III hypersensitivity reactions, drug reaction with eosinophilia and systemic symptoms, acute generalized exanthematous pustulosis, Stevens-Johnson syndrome and toxic epidermal necrolysis.

Conclusion:

Given the severity of PF and limited therapeutic response to immunosuppressive drugs in our case, RTX remained a critical treatment despite the occurrence of anaphylaxis. RDD was successfully implemented, enabling continued use of RTX and disease control.

Abstract N°: 2089

Real-World Impact of Disease Severity on Disease Progression and Mental Health Among Patients with Alopecia Areata in the United States

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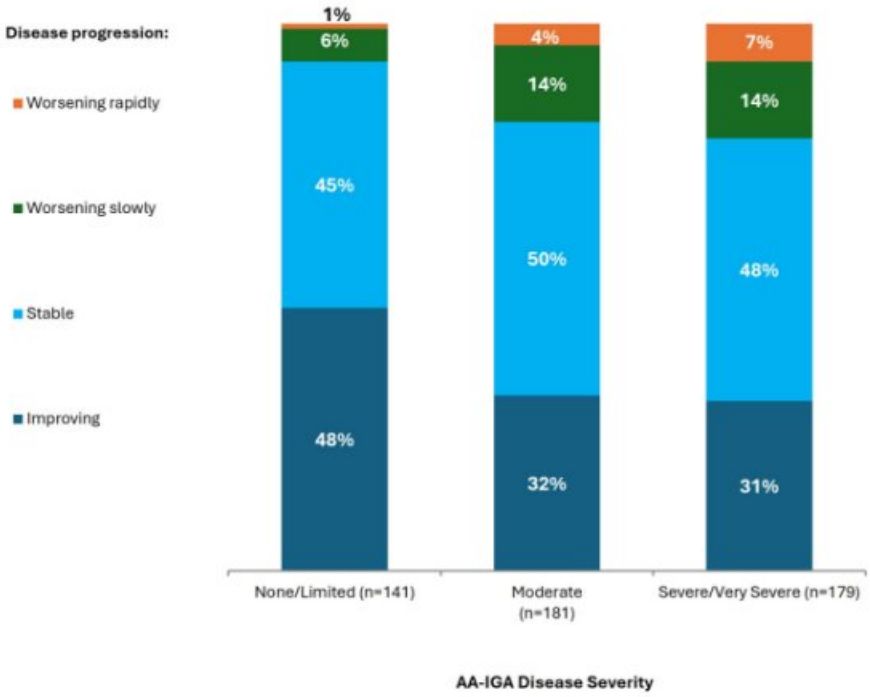
Introduction & Objectives: Alopecia areata (AA) is an autoimmune disease characterised by nonscarring hair loss on the scalp, face and body, which may reduce patients' self-esteem and cause substantial psychological burden. Previous studies investigated the impact of AA severity on these outcomes, with severity defined subjectively by physicians. This study investigated the relationship between AA severity – rigorously defined by the AA-Investigator Global Assessment (IGA) score – and disease progression/patient mental health outcomes.

Materials & Methods: Data were drawn from the Adelphi Real World AA Disease Specific Programme™, a cross-sectional survey, with retrospective data collection, of dermatologists and their patients with AA in the United States from November 2023 to June 2024. Patients were ≥18 years with a physician-confirmed AA diagnosis. Dermatologists treating ≥7 patients with AA per month (≥1 mild, ≥3 moderate and ≥3 severe/very severe) reported patient demographics, clinical characteristics and anxiety/depression status. Data were stratified by disease severity according to the AA-IGA score (≤20% scalp hair loss=none/limited, 21%–49%=moderate and ≥50%=severe/very severe AA). AA severity and changes in disease status were reported at AA diagnosis, initiation of current treatment and at the time the survey was conducted. Analysis was descriptive.

Results: Overall, 65 physicians reported data for 501 patients with AA, of whom 51% were female and mean (SD) age of 37.8 (12.4) years. Mean (SD) time since diagnosis (n=379) was 2.2 (3.4) years. At the time of the survey, 28.1% (n=141), 36.1% (n=181) and 35.7% (n=179) of patients had none/limited, moderate and severe/very severe AA severity, respectively. According to physician surveys, overall AA severity worsened in 15% (n=55/366) of patients from diagnosis to initiation of current treatment and in 7% (n=26/366) of patients from initiation of current treatment to time of survey. Disease worsening was seen in 21% [n=38] of patients with severe/very severe AA (**Figure 1**). For patients with severe/very severe AA, 39% [n=70] had eyebrow, 25% [n=44] eyelash and 20% [n=35] body hair loss; among patients with moderate and none/limited severity, 14% [n=26] and 8% [n=11]; 9% [n=16] and 6% [n=8]; and 9% [n=16] and 3% [n=4] had eyebrow, eyelash and body hair loss, respectively. Physicians reported dissatisfaction with disease control for 40% [n=72] of patients with severe/very severe AA, 37% [n=67] of patients with moderate AA and 21% [n=30] of patients with none/limited AA. Anxiety and depression were observed in patients across all severity levels. Severe anxiety and depression were reported by 20% [n=35] and 16% [n=29] of patients with severe/very severe AA; 4% [n=8] and 4% [n=8] of patients with moderate AA; and 9% [n=13] and 8% [n=11] of patients with none/limited AA, respectively. Anxiety and depression were a direct result of AA in 68% (n=21) of patients with severe/very severe AA, 40% (n=8) of patients with moderate AA, and 46% (n=6) of patients with none/limited AA.

Conclusion: AA has a high psychological burden across all disease severities; therefore, physicians should continue to prioritise mental health support for patients with AA, especially for patients with more severe disease. AA severity is also linked to worsening disease progression, thus further research should evaluate the impact of novel AA treatments on disease severity and subsequent progression.

Figure 1: Current physician-reported AA-IGA–assessed disease severity and disease progression



AA, alopecia areata; IGA, Investigator Global Assessment

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Abstract N°: 2092

Treatment Patterns and Physician Satisfaction Among Patients With Alopecia Areata in the United States in the JAK Inhibitors Era

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Introduction & Objectives: Alopecia areata (AA) is an autoimmune disease characterised by nonscarring hair loss on the scalp, face and body. Janus kinase inhibitors (JAKi) are promising new treatments for patients with AA; however, limited studies have investigated current treatment patterns and the impact of disease severity since the approval of JAKis for AA in 2022. This study investigated real-world treatment patterns and physician satisfaction stratified by AA disease severity in the JAKi era.

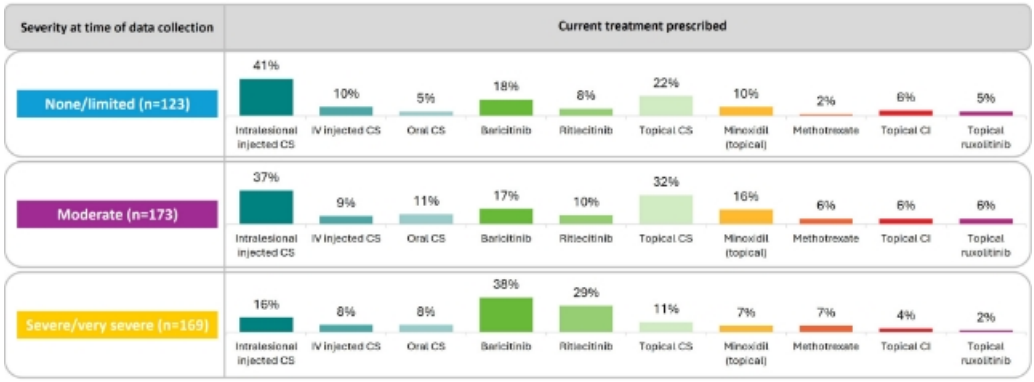
Materials & Methods: Data were drawn** from the Adelphi Real World AA Disease Specific Programme™, a cross-sectional survey, with retrospective data collection, of dermatologists and their patients with AA in the United States from November 2023 to June 2024. Patients were aged ≥18 years with a physician-confirmed diagnosis of AA. An oversample of 224 patients receiving baricitinib or ritlecitinib was also collected. Dermatologists treating ≥7 patients with AA per month (≥1 mild, ≥3 moderate and ≥3 severe/very severe) reported patient demographics, clinical characteristics, treatment data and disease severity using the AA-Investigator Global Assessment score (≤20% scalp hair loss = none/limited, 21%–49% = moderate, and ≥50% = severe/very severe AA). Analyses were descriptive.

Results: Overall, 65 physicians reported data for 501 patients with AA, of whom 51% were female and mean (SD) age of 37.8 (12.4) years. Mean (SD) time since diagnosis (n=379) was 2.2 (3.4) years. At the time of the survey, 28.1% (n=141), 36.1% (n=181) and 35.7% (n=179) of patients had none/limited, moderate and severe/very severe AA severity, respectively. Across disease severities, top physician-reported treatment goals overall were reducing scalp hair loss (88%) and improving quality of life (39%). Physicians also cited reduced eyebrow loss for 34% of patients with severe AA (n=60) and 14% with moderate AA (n=26). Physicians (n=65) cited increased or continued hair loss (72% [n=47]) and lack of long-term control of AA (71% [n=46]) as the top triggers for treatment change, followed by concerns over long-term use of corticosteroids (66% [n=43]). Oral JAKis (baricitinib/ritlecitinib) were the most prescribed AA treatment (40% [n=192/475]) – commonly administered as monotherapy (80% [n=154/192]) – closely followed by intralesional injected corticosteroids (30% [n=143/475]), most often prescribed in combination with other drugs (65% [92/141]). Oral JAKis were prescribed to 67% of patients with severe/very severe AA and 26%-27% of patients with moderate/limited severity (**Figure 1**). Physician dissatisfaction with patient's disease control on their current AA treatment was reported for 34% overall, ranging from 21% (n=30/141) in patients with none/limited AA to 41% (n=72/179) in patients with severe/very severe AA. The top three reasons for dissatisfaction (n=168) were slow onset of action for 38% (n=63), lack of overall efficacy for 33% (n=55) and patchy hair regrowth for 22% (n=37). In the JAKi oversample, physician dissatisfaction was reported as 16% (n=24/148) for patients currently receiving oral JAKis and 27% (n=28/105) for patients not currently receiving oral JAKis among patients receiving treatment for >6-months.

Conclusion: Since their approval, use of JAKis in the treatment of AA has increased dramatically, especially in patients with severe disease; however, there remains a need to improve satisfaction to meet treatment goals set

by physicians for their patients with AA.

Figure 1: Current AA treatment stratified by disease severity at the time of data collection



AA, alopecia areata; CI, calcineurin inhibitor; CS, corticosteroids; IV, intravenous

Abstract N°: 2182

Systemic lupus erythematosus flare following mRNA COVID-19 vaccination in a patient with long-standing subacute cutaneous lupus erythematosus

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Introduction & Objectives: Subacute cutaneous lupus erythematosus (SCLE) is a chronic autoimmune skin disorder that may remain clinically stable for years but carries the potential to evolve into systemic lupus erythematosus (SLE). Known triggers for disease exacerbation include infections, medications, and immunizations. With widespread administration of mRNA COVID-19 vaccines, isolated cases of autoimmune disease activation have been reported. We describe a 45-year-old Caucasian woman with a long-standing history of untreated SCLE who developed severe systemic lupus flares following vaccination, who developed severe systemic lupus flares following vaccination, ending in a fatal outcome during her last admission despite intensive care.

Materials & Methods: The patient was diagnosed with SCLE in 2003 and remained untreated with systemic therapy. One month after receiving her second dose of a mRNA COVID-19 vaccine, she developed a generalized erythematous rash. Despite initiation of hydroxychloroquine, her condition progressively worsened, leading to multiple hospitalizations over the course of a year. On her final admission, she presented with fever, weight loss, mucocutaneous ulcerations, and extensive inflammatory skin lesions. Laboratory testing during this final hospitalization revealed normocytic normochromic anemia (hemoglobin 73 g/L), hypoalbuminemia (24 g/L), elevated C-reactive protein (173.2 mg/L), urea (33.1 mmol/L), and creatinine (123 µmol/L). Autoimmune serology showed ANA positivity at 1:320, anti-dsDNA antibodies at 32.97 IU/mL, anti-SSA at 26.42 AU/mL, and persistently low complement levels (C3: 0.334 g/L; C4: 0.082 g/L). Compared to a previous hospitalization, ANA and complement consumption persisted, and a decline in anti-dsDNA titers was noted, possibly reflecting immune complex deposition or effects of prior immunosuppressive treatment.

Results:

The patient fulfilled the 2015 ACR/SLICC classification criteria for systemic lupus erythematosus with 10 criteria met. She was treated with high-dose corticosteroids, hydroxychloroquine, intravenous immunoglobulin, immunosuppressants, antibiotics, and comprehensive supportive care. Despite these interventions, her condition progressed to multiorgan failure, and she died during the final admission.

Conclusion: Conclusion

This case illustrates an aggressive and progressive systemic lupus flare temporally associated with COVID-19 vaccination in an immunologically predisposed individual. The close timing between vaccination and the onset of symptoms, along with observed immunologic changes, has prompted ongoing discussion about the potential impact of vaccines on immune function in individuals with underlying autoimmune conditions. Several reports in the literature have also described similar patterns, suggesting a possible association that warrants further scientific exploration. Close monitoring and early systemic intervention may be warranted in such patients when exposed to novel immunologic stimuli.

Abstract N°: 2235

The German National Registry of Vitiligo (VitiBest): objectives, methodology and baseline data

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Introduction & Objectives:

There is a need of systematic research investigating the long-term outcomes of Vitiligo in dermatological routine care. Patient registries represent a methodology that enables the observation and analysis of long-term outcomes in the real world, within the context of medical practice. The German National Registry of Vitiligo (VitiBest) records and assesses the long-term efficacy, safety, patient benefit and treatment regimens of Vitiligo. It was founded in 2023 and successfully completed the pilot phase with start of recruitment in August 2024.

We introduce the VitiBest registry, a non-interventional, prospective cohort study that documents the long-term effects of various treatment options for Vitiligo in routine clinical practice in Germany.

Materials & Methods:

A total of 1000 adult patients diagnosed with Vitiligo will be enrolled, with follow-up assessments scheduled at 0, 6, 12, 18 and 24 months. The follow up period will be extended to 5 years. Clinical and patient-reported outcomes are collected through standardized questionnaires administered by dermatologists. The registry's organizational structure includes collaboration between the German Dermatological Society, the Professional Association of German Dermatologists and the Working Group Neuroendocrinology of the Skin, ensuring high methodological standards while utilizing a web-based data collection system. The first data sets will be published as part of the baseline paper with a recruitment of 30 participants.

Results:

The target parameters include Vitiligo-related assessments, clinical outcome measures such as the Vitiligo Extent Score (VES) and patient reported outcomes such as the Vitiligo Noticeability Scale (VNS), and quality of life evaluations using validated instruments. Data will be analyzed to assess the efficacy, safety, and tolerability of treatments.

Conclusion:

The VitiBest registry provides real-time data and thus improves the care of patients.

Abstract N°: 2329

Dermatomyositis Revealing a Gynecological Cancer : a case report

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Introduction & Objectives:

Dermatomyositis (DM) is a rare disease, predominantly affecting women, characterized by an inflammatory condition of the skin and striated muscles, resulting in skin lesions and muscle weakness. The pathophysiology and etiology remain unknown. Adult-onset DM is associated with neoplasia in 20 to 30% of cases, which may appear before, simultaneously with, or after the diagnosis of the disease. This association worsens the prognosis and necessitates the systematic search for an underlying cancer.

The objective of this case report is to highlight the diagnostic approach and clinical implications of a paraneoplastic dermatomyositis revealing an underlying gynecological adenocarcinoma in an elderly female patient.

Materials & Methods:

We report the case of a 78-year-old woman, who presented with a six-month history of dermatological and muscular symptoms suggestive of dermatomyositis. A complete clinical evaluation was performed, along with laboratory investigations including muscle enzymes and myositis-specific antibody testing. Skin and muscle biopsies were obtained for histopathological confirmation. Electromyography (EMG) was performed. Given the patient's age and the presence of certain risk indicators, a full paraneoplastic work-up was undertaken, including tumor markers and thoraco-abdominopelvic computed tomography (CT), followed by targeted tissue biopsy.

Results:

Clinically, the patient exhibited heliotrope rash with facial edema, diffuse erythema involving the neck, chest, and upper limbs, purpuric lesions on the forearms, Gottron's papules over the metacarpophalangeal joints, and a positive "holster sign." She also presented with proximal muscle weakness involving the scapular and pelvic girdles, as well as intermittent total dysphagia. Laboratory tests revealed elevated levels of muscle enzymes. The autoimmune profile was notable for isolated anti-TIF-1γ antibody positivity, a marker strongly associated with cancer-associated dermatomyositis. Histological analysis of the skin revealed interface dermatitis, while muscle biopsy confirmed inflammatory myositis. EMG showed no abnormalities. Tumor markers CA125 and CA15-3 were elevated. CT imaging demonstrated peritoneal carcinomatosis, and biopsy of peritoneal tissue confirmed a well-differentiated infiltrating adenocarcinoma involving fibro-adipose tissue, most likely of gynecological origin. The patient was treated with systemic corticosteroids at 1 mg/kg/day and referred to oncology for specialized management.

Conclusion:

This case underscores the importance of a thorough malignancy screening in all adult patients presenting with dermatomyositis. The identification of a paraneoplastic etiology has crucial therapeutic and prognostic implications. Early diagnosis and multidisciplinary management are essential to improve outcomes in patients with cancer-associated dermatomyositis.

Abstract N°: 2399

Sequelae of Mucous Membrane Pemphigoid in a Global Clinical Database

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Introduction & Objectives:

Mucous membrane pemphigoid (MMP) is a rare, chronic autoimmune blistering disease affecting mucosal surfaces. Scarring can lead to functional impairment and sequelae such as blindness, tracheal and esophageal strictures, as well as comorbid neuropsychiatric diseases and neoplasms significantly impact patient morbidity. Given the rarity of MMP, risk assessment in large-scale clinical studies is challenging, necessitating alternative research approaches.

Materials & Methods:

We leveraged the TriNetX global electronic health record database to analyze the burden of MMP sequelae. Patients with MMP were identified and propensity-score matched to control groups. Kaplan-Meier survival analysis and Cox proportional hazards models were used to assess the risks of non-melanoma skin cancer, esophageal strictures, and blindness compared to controls, adjusting for relevant confounding factors.

Results:

Patients with MMP exhibited a significantly increased risk of squamous cell carcinoma (HR 1.89, 95% CI: 1.21–2.95, $p = 0.004$) and basal cell carcinoma (HR 1.50, 95% CI: 1.06–2.13, $p = 0.021$). The incidence of esophageal strictures tripled from 2016 to 2023, with an overall prevalence rising from 1.43% to 3.51%. Young patients with MMP (20–30 years) exhibited the highest risk. Patients with MMP also had a 3.1-fold increased risk of blindness (HR 3.1, 95% CI: 2.2–4.2, $p < 0.0001$), with an absolute 5-year risk of 5.5%.

Conclusion:

These large-scale cohort studies highlight underrecognized long-term complications of MMP, emphasizing the need for interdisciplinary management. Early dermatologic, gastroenterologic, and ophthalmologic screening is crucial to mitigate severe outcomes. Future studies incorporating autoantibody profiling and treatment effects are warranted.

Abstract N°: 2407

Cutaneous Neonatal Lupus in a Male Newborn: Diagnostic Clues and a 30-Year Retrospective Review

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Introduction & Objectives:

Neonatal lupus erythematosus (NLE) is a rare alloimmune condition that occurs in infants born to mothers with autoimmune diseases, particularly Sjögren's syndrome and systemic lupus erythematosus (SLE). Cutaneous manifestations are among the most frequent presentations, though atypical forms may delay diagnosis. Cardiac involvement is the most feared complication and must be excluded. This study aims to report a case of cutaneous NLE with an atypical presentation and to review biopsy-confirmed NLE cases diagnosed at our institution over the past 30 years.

Materials & Methods:

We describe the case of a 28-day-old male newborn with erythematous circular papules and plaques on the scalp, trunk, lower limbs, and soles, referred to dermatology for evaluation. The work-up included skin biopsy, serological testing, and cardiac assessment. A retrospective chart review of all histologically confirmed NLE cases in our hospital between 1993 and 2023 was also conducted.

Results:

The skin biopsy showed histopathological features consistent with cutaneous lupus. Serology was positive for anti-Ro/SSA and anti-La/SSB antibodies; no hematologic, hepatic, or electrocardiographic abnormalities were found. The mother had a history of Sjögren's syndrome and was treated with hydroxychloroquine. A diagnosis of NLE with atypical cutaneous involvement and no systemic disease was made. In the retrospective review, four biopsy-confirmed NLE cases were identified. Half were male, with a mean age at diagnosis of 48 days. Two-thirds were positive for both anti-SSA and anti-SSB antibodies, and none had cardiac involvement. In 50% of cases, maternal autoimmune disease had been diagnosed prenatally.

Conclusion:

This case highlights the importance of considering NLE in newborns presenting with unusual cutaneous findings, especially when maternal autoimmune disease is known. Despite its rarity, NLE should be included in the differential diagnosis of neonatal dermatoses. Our 30-year institutional review reflects the low incidence of biopsy-confirmed NLE and reinforces the need for clinical suspicion and multidisciplinary evaluation to ensure early diagnosis and management.

Abstract N°: 2434

Quality of Life in Patients with Lupus

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Introduction & Objectives:

Systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory disease that encompasses a wide spectrum of clinical manifestations, ranging from isolated cutaneous lupus to potentially severe systemic forms. Cutaneous involvement, present in the majority of cases, often serves as the first noticeable sign of the disease. Due to its frequent localization on exposed areas (face, scalp, limbs), cutaneous lupus is classified as one of the visible dermatoses, which can significantly alter patients' body image and psychological well-being. This visibility, often stigmatizing, can have a considerable impact on patients' social, familial, and professional lives.

Materials & Methods:

A 15-question questionnaire was created on Google Forms and distributed to patients followed for systemic lupus erythematosus in the dermatology department. Quality of life was assessed using the validated Arabic dialect version of the DLQI (Dermatology Life Quality Index) questionnaire; the total score ranges from 0 to 30, divided into five categories: 0-1 (no effect), 2-5 (minor effect), 6-10 (moderate effect), 10-20 (significant effect), 20-30 (extremely important effect).

Results:

A total of 30 patients, aged from 11 to 71 years, with a mean age of 43.8 ± 15.1 years, were included in our study. Among them, 80% were female, reflecting the typical demographic of lupus patients, and 73.3% had been diagnosed for more than 5 years, indicating a deep understanding of the disease. 63.3% of the patients had isolated cutaneous lupus, while 36.7% had cutaneous involvement as part of systemic lupus. Additionally, 63.3% of the patients had a history of photosensitivity. The mean DLQI score of the patients included in this study was 10.6 ± 4.3, indicating a moderate to significant impact on the quality of life of patients with cutaneous lupus. The distribution of quality of life impairment based on the DLQI scores was as follows:

- An extremely important impact in 13.3%
- A significant impact in 43.3%
- A moderate impact in 33.3%
- A mild impact in 10%

The impairment of quality of life was particularly marked in women and younger patients, as well as in those with isolated cutaneous lupus. Photosensitivity played a key role in limiting outdoor activities. These results emphasize the importance of a comprehensive approach to management, taking into account both the dermatological and psychological aspects of the disease.

Conclusion:

In conclusion, lupus, especially cutaneous lupus, significantly affects patients' quality of life due to the visibility of

skin lesions, leading to psychological distress and social challenges. Women, younger patients, and those with isolated cutaneous lupus are particularly impacted. Photosensitivity further restricts outdoor activities. These findings emphasize the need for a holistic treatment approach, addressing both the clinical and psychological aspects of the disease. Comprehensive care is essential to improve the overall well-being of lupus patients.

EA DV CONGRESS
PARIS 17-20 SEPTEMBER 2025
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Abstract N°: 2477

Bullous Pemphigoid in a Patient with Generalized Morphea: Coincidence or Pathogenic Link?

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Introduction & Objectives:

Generalized morphea is a rare form of localized scleroderma characterized by widespread indurated plaques. Like other scleroderma types, it is of unknown etiology, possibly autoimmune, affecting both the microvasculature and connective tissue. Bullous pemphigoid (BP) is the most common autoimmune blistering disease, involving autoantibodies against BP180 and/or BP230. The association of bullous lesions with scleroderma is rare. Here, we report a case of generalized morphea complicated by BP 13 years later.

Materials & Methods:

A 50-year-old woman with a history of diabetes and hyperthyroidism had generalized morphea since 2011, confirmed by skin biopsy. A month before admission, she developed post-bullous erosive plaques with erythematous borders on the back and clavicles. Biopsy and direct immunofluorescence confirmed BP. She was treated with corticosteroids (0.7 mg/kg/day), resulting in good clinical improvement.

Results:

This case represents a rare association between BP, an autoantibody-mediated skin disorder, and morphea, a sclerosing skin disease. Interestingly, the BP lesions were confined to areas previously affected by morphea, suggesting that the autoimmune mechanisms underlying both conditions might overlap. BP is an immunobullous dermatosis primarily triggered by factors such as trauma, burns, UV radiation, drugs, and infections, but in this case, the patient's long-term use of metformin and gliclazide was unlikely to be a trigger, as these drugs had been used for more than two years before the onset of BP.

While BP and morphea typically have distinct inflammatory patterns, both conditions share common immunological pathways. For instance, both diseases are influenced by T-helper (Th) 2 cell-mediated cytokines, which play a central role in the fibrosis observed in morphea. Additionally, these cytokines are overexpressed in both the skin and blood of BP patients. Studies have shown that genetic factors, such as IL-13 polymorphisms, may predispose individuals to both BP and systemic sclerosis, further supporting the idea of shared immunological mechanisms. The presence of the HLA-DQ7 haplotype, a known risk factor for BP, has also been observed in patients with localized scleroderma and linear scleroderma, suggesting a common genetic predisposition that may contribute to the development of BP in morphea patients.

Conclusion:

We report a rare case of BP arising in a patient with longstanding morphea. The coexistence of these two diseases may be explained by overlapping immune mechanisms and a shared genetic background. Morphea may predispose individuals to the development of BP, possibly triggered by an external factor that induces immune dysregulation, leading to the production of autoantibodies against BP180.

Abstract N°: 2495

Cell-free DNA profiles of dermatomyositis and its potential role in discriminating phenotypes

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Introduction & Objectives:

Cell-free DNA (cfDNA) functions in the early-detection and monitoring of autoimmune diseases including systemic lupus erythematosus and rheumatoid arthritis. However, investigations into cfDNA profiles in dermatomyositis and their potential clinical implications remain scarce. Hence, we aim to explore the overall landscape of cfDNA profiles in dermatomyositis and investigate potential roles in discriminating subtypes.

Materials & Methods:

Following informed consent, 24 patients diagnosed with dermatomyositis and 16 healthy controls were enrolled. We examined cfDNA concentrations, fragment distribution patterns, 5’-end motif frequencies and genetic variation profiles in all participants. We also investigated the correlation between extracellular DNases levels and cfDNA profiles. Lastly, we investigated the differences of cell-free DNA profiles among patients with or without malignancies.

Results:

Compared to healthy controls, dermatomyositis patients exhibited elevated cfDNA concentrations, with significantly longer cfDNA fragments, primarily centered around 180-360 bp. The A-end predominated the 5’-end motif, whereas the C-end was underrepresented, contrasting with the patterns observed in healthy controls. Plasma levels of extracellular DNase appeared to correlate with cfDNA levels and end motif profiles. In addition, genetic variations in several genes, including PDE4DIP and BRCA2, were commonly detected in cfDNA from dermatomyositis patients. Notably, end-motif profiles and cfDNA fragment length exhibited variations between anti-transcription intermediary factor 1-gamma positive patients with and without malignancies.

Conclusion:

This study presents the first comprehensive depiction of cfDNA profiles in patients with dermatomyositis. Furthermore, extracellular DNases may be involved in the synthesis of cfDNA, warranting further investigation. Finally, cfDNA features exhibit variability across some sub-phenotypes and may serve as discriminatory indices.

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Abstract N°: 2507

A Case of a Pemphigus Vulgaris and Foliaceus Overlap Presenting with Malar Rash and Crusted Eroded Lesions

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Introduction & Objectives:

Pemphigus comprises a spectrum of rare autoimmune mucocutaneous disorders in which autoantibodies target desmosomal adhesion proteins, resulting in intraepidermal blistering and acantholysis. Subclassification of pemphigus relies on clinical presentation, histopathology, and antibody profiles, with pemphigus vulgaris (PV) and pemphigus foliaceus (PF) being the most common forms. We present a rare case of overlapping PV and PF in a 70-year-old man who exhibited malar erythema, crusted erosions, and tense bullae—remarkably without mucosal involvement.

Materials & Methods:

We reviewed the patient's clinical records and gathered relevant medical history. A literature search was performed using the terms "pemphigus vulgaris," "pemphigus foliaceus," "malar rash," and "pemphigus erythematosus."

Results:

A 70-year-old male developed an erythematous, eroded rash over the nasal tip that, over two weeks, extended bilaterally across the malar regions. Topical therapies provided no relief. Soon thereafter, crusted erosions appeared on his abdomen and lower extremities, and flaccid bullae formed on his back. On examination, both direct and indirect Nikolsky signs were positive; mucosal sites, including oral and genital regions, were spared.

Laboratory evaluation revealed high titers of anti-desmoglein (Dsg) 1 and Dsg 3 antibodies, with a negative antinuclear antibody (ANA) screen. The patient was admitted and started on systemic corticosteroids (oral prednisolone 56 mg/day). Biopsies from the malar and lower limb lesions underwent histopathological and direct immunofluorescence analysis: the malar biopsy showed intraepidermal acantholysis consistent with PF, whereas the lower-limb specimen demonstrated suprabasal clefting characteristic of PV with intercellular IgG and C3 deposition. Based on high titers of anti-desmoglein (Dsg) 1 and Dsg 3 antibodies, histopathological and direct immunofluorescence findings, a diagnosis of PV–PF overlap was made and azathioprine was added to the treatment regimen. After three weeks of combined immunosuppressive therapy, the patient experienced marked resolution of both facial and body lesions.

Conclusion:

Overlap between PV and PF manifesting as an erythematous malar rash without mucosal lesions is exceedingly uncommon. This presentation may mimic pemphigus erythematosus—a milder PF variant confined to the malar area and nasal bridge that clinically resembles lupus erythematosus. Awareness of such atypical overlaps is vital, as it can influence both the diagnostic workup (including targeted biopsies and serologic assays) and therapeutic decisions. Clinicians should consider pemphigus variants in the differential diagnosis of malar eruptions, even

when classical mucosal involvement is absent.

EADV CONGRESS
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Abstract N°: 2522

Assessment of efficacy of a combination of low dose rituximab and low dose prednisolone vs low dose prednisolone alone in inducing repeat remission in rituximab treated pemphigus vulgaris patients who develop disease relapse: A randomised controlled trial

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Introduction & Objectives: There are no evidence-based treatment guidelines for managing relapse in Pemphigus Vulgaris (PV) patients previously treated with the RA protocol of rituximab. Current strategies include rituximab infusion every 6 months, periodic immunological monitoring with infusion of low-dose rituximab on immunological relapse, and infusion of standard RA protocol on clinical relapse. These approaches may be impractical because of cost constraints and unavailability of tests like flow cytometry and anti-desmoglein antibody ELISA at many places across the globe. Patient feedback suggested us that prednisolone at a low dose (0.3 mg/kg/day) may be effective in mild, early relapses after rituximab. This study was carried out to evaluate the efficacy of low-dose rituximab (500 mg) combined with low-dose prednisolone (0.3 mg/kg/day) versus low-dose prednisolone alone (0.3 mg/kg/day) in achieving repeat remission in relapsed PV patients previously treated with rituximab (RA protocol).

Materials & Methods: This is a randomized, hospital-based, non-blinded ongoing trial aiming to achieve a target sample size of at least 15 patients in each group and has currently recruited 21 participants. These patients were divided into two groups: Group A (rituximab + low-dose prednisolone, n =10) and Group B (low-dose prednisolone alone, n =11). The primary outcome was the time to achieve repeat remission. The secondary outcomes were to assess time to clinical remission, remission on minimal and no therapy, cumulative corticosteroid dose, frequency of relapse, adverse effects observed and effect on desmoglein 1 and 3 titres. Patients were followed for up to 9 months. Fifteen participants completed at least 3 months (for whom the data has been evaluated) and six completed 6 months of follow-up.

Results: At 3 months, all Group A patients (7/7) achieved disease control compared to 75% in Group B (6/8). Clinical remission was observed in all patients in Group A while only 37.5% patients of Group B achieved this outcome. Complete remission off therapy (Croff) was achieved by 71.4% of Group A patients, but none in Group B. The cumulative prednisolone dose was significantly lower in Group A (median: 1350 mg vs. 1800 mg in Group B; p=0.022). Adverse events were fewer in Group A (1 vs. 8 in Group B; p=0.0014), highlighting a corticosteroid-sparing effect and better safety profile.

Conclusion: In the interim analysis of the ongoing non-blinded randomized controlled trial, we observed that low-dose rituximab (500 mg) combined with low-dose prednisolone (0.3 mg/kg/day) is highly effective in managing relapsed mild-to-moderate PV, achieving faster disease control, higher remission rates, reduced corticosteroid usage, and fewer adverse effects. This regimen bridges the gap in existing treatment strategies, providing a cost-effective alternative for management of mild to moderate PV relapse. **

Abstract N°: 2600

Refractory subacute cutaneous lupus in the context of myelodysplastic syndrome: exceptional response to anifrolumab.

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Introduction & Objectives: subacute cutaneous lupus erythematosus (SCLE) may present as an isolated condition or in association with systemic lupus erythematosus. It has also been described as a paraneoplastic phenomenon, primarily linked to solid organ malignancies. Cutaneous manifestations associated with myelodysplastic syndrome (MDS) encompass a wide range of dermatologic presentations, which may be secondary to the hematologic disorder itself, related cytopenias, or paraneoplastic phenomena. Recent studies support the use of anifrolumab, a monoclonal antibody targeting the type I interferon receptor, as a fast and effective therapeutic option for cutaneous manifestations of lupus.

Materials & Methods: presentation of a clinical case and review of the literature

Clinical case: we report the case of a 70-year-old female with a history of sigmoid adenocarcinoma in remission since 2014, followed by the Dermatology Department since 2023 for biopsy-confirmed SCLE with negative antinuclear antibodies (ANA). Initial systemic treatment included prednisone, hydroxychloroquine, and methotrexate, achieving only partial disease control. Methotrexate was discontinued due to the onset of cytopenias. A subsequent workup confirmed low-risk MDS, with no specific treatment indicated by the Hematology Department. The patient experienced a flare-up of cutaneous lesions along with systemic symptoms such as asthenia, weakness, and fatigue, leading to hospital admission. A comprehensive evaluation, including skin biopsy, PET-CT, and extended autoimmune blood testing, ruled out internal organ involvement and other underlying conditions. Given the refractoriness of the cutaneous disease, treatment with intravenous anifrolumab 300 mg monthly was initiated. The patient experienced a marked improvement from the first doses, with complete resolution of skin lesions and systemic symptoms, and no significant adverse events.

Conclusion: this case highlights the potential role of SCLE as a facultative paraneoplastic phenomenon and underscores the efficacy of anifrolumab in patients refractory to conventional therapies or with contraindications to them. It also supports the central role of type I interferon in the disease pathophysiology through the promotion of aberrant immune activation.

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Abstract N°: 2658

Demographic and Clinical Characteristics of Pemphigus Patients Influencing Treatment Outcomes: A Retrospective Cohort Study

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Introduction & Objectives: In pemphigus, the course of the disease, prognosis, and rates of remission and relapse vary individually. This research aims to investigate the relationship between pemphigus patients' demographic, clinical, and laboratory characteristics and their remission and relapse statuses.

Materials & Methods: This study was a single-center, cross-sectional, retrospective study including seven years and reviewing the demographic and clinical data, treatment details, laboratory results, and information on remission and relapse of patients with pemphigus.

Results: Our study included 55 male and 45 female patients. Pemphigus vulgaris was diagnosed in 92% of the patients, and mucocutaneous involvement was observed in 71%. Mild disease was present in 32% of patients, moderate in 51%, and severe in 17%. During the treatment process, 53% of the patients were hospitalized, and 38% experienced treatment-related side effects. Remission was achieved in 83% of the patients, with an average time to remission of 11.2±8.1 months. Among those who achieved remission, the proportion of patients with mild to moderate disease at initial assessment and the length of hospital stays were significantly higher than those who did not achieve remission (p=0.042 and p=0.036, respectively). Disease severity and length of hospital stay were independent factors affecting remission (HR=0.125, p=0.027 and HR=1.402, p=0.011, respectively). After remission, relapse was observed in 54.2% of the patients, with an average remission period of 12.5±11.7 months prior to relapse. The most common cause of relapse was infections (38.8%). Compared to those without relapse, patients who experienced relapse had a higher prevalence of chronic diseases and treatment-related side effects (p=0.023 and p=0.007, respectively), and their age at admission was younger (p=0.017). Male gender and younger age at disease onset were identified as risk factors for relapse (HR=0.044, p=0.018 and HR=0.807, p=0.039, respectively). The duration of remission was significantly longer in those without chronic diseases (p=0.028). The month of disease in which rituximab (RTX) was applied was identified as an independent factor influencing the duration of remission (HR=0.792, p=0.043). Each 1-month delay in RTX administration shortened patients' remaining in remission before relapse by 1.26 times.

Conclusion: Age at disease onset, initial disease severity, the month of initiation of rituximab therapy, duration of hospitalization, and accompanying comorbidities are closely linked to remissions and relapses in pemphigus. In particular, our findings highlight the importance of considering RTX therapy in appropriate patients during the early stages of the disease, especially when assessing the risk of relapse. Furthermore, they underscore the significance of treatment planning in the follow-up and management of patients.

Abstract N°: 2711

Anifrolumab as a treatment for cutaneous manifestations in refractory Dermatomyositis: A Case Series

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Introduction & Objectives:

Dermatomyositis (DM) is an autoimmune connective tissue disease with clinical manifestations at the cutaneous and/or systemic level. Although multiple treatments are used for its management, cutaneous symptoms are often refractory to them.

Anifrolumab is a monoclonal antibody targeting the type I interferon-alpha receptor, approved for the treatment of moderate-to-severe and refractory systemic lupus erythematosus. We present a case series of DM with refractory cutaneous involvement that responded to treatment with anifrolumab.

Materials & Methods

Observational retrospective study that involves 5 patients with dermatomyositis treated with anifrolumab. We assessed previous treatments, demographic data, and clinical data before and after treatment.

Results:

A 90-year-old woman with amyopathic dermatomyositis (DM) of the antisynthetase syndrome type, refractory to multiple treatments (methotrexate, azathioprine, corticosteroids, hydroxychloroquine, and immunoglobulins). Anifrolumab (300 mg/month), prednisone (5 mg/day), and immunoglobulins were initiated, with rapid improvement and lesion resolution by the third month.

A 53-year-old woman with DM characterized by panniculitis and calcinosis. After failure of previous treatments (methotrexate, corticosteroids, hydroxychloroquine, immunoglobulins, and tofacitinib), anifrolumab (300 mg/month) was started in combination with hydroxychloroquine, immunoglobulins, and prednisone (5 mg/day), with improvement noted at 3 months.

A 68-year-old woman with paraneoplastic amyopathic DM who had received methotrexate, corticosteroids, hydroxychloroquine, tofacitinib, and immunoglobulins with partial response. Anifrolumab (300 mg/month) was initiated while maintaining immunoglobulins, leading to lesion clearance from the first month. As a side effect, she presented with bilateral trochanteric-type pain.

Conclusion:

The treatment of DM is varied, and disease activity is often difficult to control. Therefore, management should be personalized based on comorbidities, disease activity, and patient tolerance.

The involvement of type I interferon (IFN) in the pathogenesis of DM has been proposed due to the overexpression of IFN-regulated genes in these patients. Furthermore, this overexpression appears to correlate with increased disease severity. Given the role of IFN in DM pathogenesis, it seems reasonable to consider anifrolumab as a potentially effective treatment. The cases we present responded quickly and effectively, with good tolerance. However, further studies are needed to confirm these findings.

Abstract N°: 2743

Correlation of clinical and histopathological features with anti-desmoglein antibody profile in pemphigus patients

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Introduction & Objectives: The desmoglein compensation hypothesis (DCH) proposes that clinical and histopathological features of pemphigus are determined by the distribution of desmoglein 1 (Dsg1) and desmoglein 3 (Dsg3) in skin and mucosa, and the presence of corresponding autoantibodies. While DCH explains many classical presentations, emerging evidence shows discrepancies between serology, histology, and clinical phenotype, suggesting the model may be oversimplified. In this study of pemphigus vulgaris (PV) and pemphigus foliaceus (PF) patients, we aimed to correlate clinical presentation, level of histopathological split, and anti-Dsg1/3 profiles to evaluate the fidelity of these relationships to DCH.

Materials & Methods: In this bicentric retrospective study, 106 pemphigus patients (93 PV, 13 PF) were analyzed based on clinical presentation, lesional histopathology, and serum Dsg1/Dsg3 IgG levels. Concordance among serology, histology, and phenotype was assessed against the predictions of DCH.

Results: Among PF patients, subcorneal acantholysis was seen in 46.2%, while 53.8% showed unexpected suprabasal or dual-level splits. Only 61.5% had isolated anti-Dsg1 antibodies; 30.7% showed dual positivity, and 1 patient had isolated anti-Dsg3. In PV, 76% of mucocutaneous patients showed suprabasal acantholysis, with 74.7% demonstrating dual antibody positivity—both findings consistent with DCH. However, among mucosal PV patients, only 37.5% had isolated anti-Dsg3, while 43.8% showed dual positivity and 18.7% lacked detectable antibodies. Notably, 7.5% of all PV patients had no serological reactivity despite active disease. The Dsg3:Dsg1 ratio was significantly higher in PV than PF ($p=0.001$) but did not correlate with histological split level ($p=0.861$).

Conclusion: While DCH explains findings in a subset of patients, particularly those with mucocutaneous PV, a significant proportion of cases deviate from its predictions—demonstrating dual-level splits, discordant antibody profiles, or seronegative disease. These findings highlight the limitations of a desmoglein-only model and underscore the need for broader frameworks that incorporate antibody affinity, subclass, epitope specificity, and non-Dsg targets.

Abstract N°: 2765

Efficacy and safety of intradermal hyaluronidase injections in the treatment of microstomia in systemic sclerosis.

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Introduction & Objectives:

Systemic sclerosis (SSc) is an autoimmune disease characterized by cutaneous and visceral fibrosis. Microstomia is a common complication in patients with SSc, presenting as a reduction in oral aperture that hinders feeding and oral hygiene, negatively impacting quality of life. Current therapeutic options for this condition are limited.

Materials & Methods:

Two patients with SSc and microstomia were treated. Each received two perioral intradermal injections of 150 IU of hyaluronidase, administered 4 to 7 weeks apart. Oral aperture (in mm), perioral fibrosis, and patient-reported subjective improvement were assessed before and after treatment.

Results:

The first patient demonstrated a 5 mm increase in lateral opening and a 20 mm increase in vertical opening. The second patient improved by 5 mm in both dimensions. Both reported a significant improvement in quality of life, including fewer limitations in feeding and oral hygiene, and less frequent episodes of angular cheilitis. No significant adverse effects were observed, and the treatment was well tolerated.

Conclusion:

Intradermal hyaluronidase injections appear to be a promising therapeutic option for microstomia in systemic sclerosis, with potential to enhance patients' quality of life. Further studies are warranted to confirm these preliminary findings and optimize treatment protocols.

Abstract N°: 2784

Drug-Induced Pemphigus Erythematosus Presenting As Vasculitis In A 27-Year Old Filipino Female With Pulmonary Tuberculosis: A Case Report

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Introduction:

Pemphigus erythematosus (PE), also known as Senear-Usher syndrome, is a rare autoimmune disorder exhibiting overlapping clinical and histopathological features of both pemphigus and lupus erythematosus. It is considered a variant of pemphigus foliaceus presenting with crusted erosions in a seborrheic distribution, at times concurrent with more lupus-like discoid lesions with “carpet-tack” scale. Flaccid, small vesicles that easily rupture may also be present. While pemphigus erythematosus usually presents with crusted erosions, some cases have been reported of PE mimicking different inflammatory conditions. PE may be misdiagnosed as seborrheic dermatitis, contact dermatitis, or small plaque parapsoriasis. It may also appear as tumor lesions, presenting as multiple seborrheic keratosis-like papules and plaques. With various atypical presentations, overlapping features, and complexity of its nature, pemphigus erythematosus remains to be a diagnostic challenge to this day.

Case Discussion:

This is a case of a 27-year-old Filipino female with bacteriologically confirmed pulmonary tuberculosis, who developed angioedema and generalized, erythematous, non-blanching papules, patches, and plaques following initiation of first-line antituberculosis therapy (HRZE regimen: isoniazid, rifampicin, pyrazinamide, ethambutol). An initial diagnosis of a drug-induced hypersensitivity reaction was made, and she was treated with systemic corticosteroids and antihistamines. However, minimal clinical improvement was observed, prompting further diagnostic evaluation. Serological testing revealed positive antinuclear antibody (ANA), anti-double-stranded DNA (anti-dsDNA), anti-histone IgG, and decreased serum complement C3 levels, findings supportive of drug-induced systemic lupus erythematosus (DI-SLE). Histopathology showed epidermal atrophy, focal interface vacuolar dermatitis, mucin deposition in between collagen bundles, and features of leukocytoclastic vasculitis. Direct immunofluorescence (DIF) studies, on the other hand, showed findings indicative of a pemphigus disease etiology, revealing granular intercellular deposits of IgG and IgA, along with strong granular vascular fibrinogen deposition. Given the coexistence of lupus-specific serologic markers and immunopathologic findings characteristic of pemphigus, a diagnosis of pemphigus erythematosus was established. The patient was started on high-dose intravenous methylprednisolone pulse therapy. One month following treatment initiation, she demonstrated marked clinical improvement, with resolution of active lesions and leaving behind only few slightly erythematous to hyperpigmented patches.

Conclusion:

This case provides additional evidence to the various atypical presentations of pemphigus erythematosus. While vasculitis is an uncommon finding in pemphigus, its presence may be the primary manifestation of pemphigus erythematosus. Therefore, clinicians should maintain a high index of suspicion for pemphigus erythematosus presenting with vasculitic lesions, particularly when such lesions do not respond to standard therapeutic interventions.

Abstract N°: 2791

Differential risks of venous thromboembolic events in pemphigus and pemphigoid diseases: A large-scale retrospective cohort study

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Introduction & Objectives:

Venous thromboembolic events (VTE) are serious sequelae of bullous pemphigoid (BP) and pemphigus. However, the VTE risk in other autoimmune blistering diseases (AIBD), including mucous membrane pemphigoid (MMP) and epidermolysis bullosa acquisita (EBA), remains unclear.

Materials & Methods:

We used the TriNetX Global and US Collaborative Networks to conduct retrospective cohort studies using electronic health records (EHRs). Patients with BP, pemphigus, MMP, or EBA were identified using ICD-10-CM codes and compared to non-AIBD controls (ICD-10-CM: Z00). VTE was defined as pulmonary embolism (I26) or deep vein thrombosis (I82.4). Propensity-score matching (PSM) accounted for age, sex, neoplasms, and surgery. Sensitivity analyses (S1–S5) used alternative outcome windows and PSM strategies.

Results:

After PSM, cohorts included 8,837 pemphigus, 14,220 BP, 3,997 MMP, and 971 EBA patients with matched controls. BP and pemphigus were associated with increased VTE risks (HR range: 1.46–2.13). In MMP, no significant difference in VTE risk was observed (HR 0.67, 95% CI: 0.43–1.11, p=0.08). In contrast, EBA patients showed increased VTE risk (HR 3.19, 95% CI: 1.35–7.49, p=0.005), confirmed in two sensitivity analyses.

Conclusion:

Our study confirms increased VTE risks in BP and pemphigus and points towards a possible elevated VTE risk in EBA, but excludes this for MMP. Differences in systemic inflammation and body surface area involvement may underlie these findings. These results suggest the need for VTE monitoring in selected AIBD patients, particularly those with extensive disease.

Abstract N°: 2807

Treatment with DPP4-inhibitors does not increase circulating autoantibodies for autoimmune bullous diseases in patients with type 2 diabetes

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Introduction & Objectives:

Dipeptidyl-peptidase 4-inhibitors (DPP4i) are widely used in managing type 2 diabetes mellitus (T2DM). However, several studies have linked DPP4i to an increased risk of autoimmune bullous diseases (AIBD), particularly bullous pemphigoid (BP), with reported risk increases between 1.4- and 4.4-fold. Prior studies have shown slightly elevated - but not statistically significant - levels of full-length BP180 autoantibodies in DPP4i users, while levels of BP180-NC16A and BP230 remained unchanged. To date, only BP180 and BP230 have been studied in this context. Our study expands this by assessing additional autoantibodies.

Materials & Methods:

This study, approved by the University of Lübeck IRB (#20-154), included 101 T2DM patients. Blood samples were collected from Praxiszentrum Diabendo (Rostock) and the Department of Medicine I, UKSH Lübeck. Using commercial ELISA kits (Euroimmun, Germany), we measured antibodies to BP180, BP230, type-VII collagen (COL7), desmoglein 1 (Dsg1), desmoglein 3 (Dsg3), gliadin, and tissue transglutaminase (tTG).

Results:

Of the 101 patients, 44 were on DPP4i therapy, 36 with sitagliptin and 8 with saxagliptin. Most had been on treatment for over five years. DPP4i users were older on average (68 ± 13.3 years) than non-users (62.5 ± 11.9 years), but sex distribution was similar across groups. Antibody levels did not differ significantly between DPP4i users and non-users. Gliadin and tTG levels were strongly correlated ($\rho = 0.66$ for controls; 0.53 for DPP4i users), consistent with existing literature. Applying the recommended diagnostic cut-off value of 20 RU/ml, few samples tested positive. Three patients tested positive for two antibodies: one DPP4i user (BP180 and BP230), and two non-users (BP230+Dsg1; COL7+Dsg1).

Conclusion:

Our findings align with previous studies showing no significant increase in AIBD-related autoantibodies in DPP4i-treated T2DM patients without a prior AIBD diagnosis. Limitations include older age among DPP4i users and limited DPP4i types (only sitagliptin and saxagliptin were prescribed). These results support the view that serologic testing should be reserved for patients with clinical suspicion of AIBD. Overall, our study suggests that DPP4i therapy does not elevate circulating autoantibodies to skin antigens in T2DM patients.

Abstract N°: 2859

Rowell Syndrome: A diagnostic challenge in a patient with overlapping autoimmune disorders

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Introduction & Objectives:

Rowell Syndrome is a rare and controversial clinical entity, characterized by the coexistence of lupus erythematosus (LE), erythema multiforme (EM)-like lesions, and the presence of immunological abnormalities, including positive antinuclear antibodies (ANA) in a speckled pattern, positive anti-La/SS-B or anti-Ro/SS-A antibodies, and positive rheumatoid factor. This syndrome was first described by Dr. Neville Rowell et al. in 1963, and the majority of cases occur in middle-aged women, given the fact that this is an immune-mediated condition. The precise etiology remains ambiguous, posing a challenge for clinicians and requiring multidisciplinary collaboration. Therapy schemes for Rowell Syndrome include systemic corticosteroids in medium and high doses, immunosuppressive agents and individualized strategies based on the patient's comorbidities.

Materials & Methods:

We report a case of a 57-year-old female patient who developed multiple generalized erythema multiforme lesions over two years, including three severe episodes with extensive blistering, possibly triggered by herpes simplex infection. Initially, she was misdiagnosed with impetigo and was prescribed antibiotics, but no biopsy was performed. Two years later, she underwent a punch biopsy with histopathological evaluation, which established the diagnosis of discoid lupus erythematosus. Laboratory investigations revealed positive rheumatoid factor and antinuclear antibodies (ANA), with positive anti-Ro and anti-La antibodies, followed by the appearance of anti-Sm and anti-dsDNA antibodies. Furthermore, she was also diagnosed with Sjögren's syndrome, autoimmune thyroiditis, and IgA deficiency. The combination of cutaneous and serological findings supported the diagnosis of Rowell Syndrome.

Results:

The patient was prescribed antimalarial therapy along with a corticosteroid cream. Sun protection is crucial, as exposure to ultraviolet radiation can worsen the skin lesions, leading to hyperpigmentation and photosensitivity, and may also trigger lupus flares. Although improvement was observed, the lesions did not completely resolve, highlighting the chronic and relapsing nature of the condition.

Conclusion:

Our aim in presenting this case is to highlight the importance of considering Rowell Syndrome in patients with lupus and EM-like lesions, especially when characteristic serologic markers are present. Accurate and timely diagnosis—ideally from the first presentation—is essential in order to reduce unnecessary treatments, minimize psychological distress, and preserve the patient's quality of life. The absence of an identifiable trigger in most cases further complicates the diagnostic process, reinforcing the need for a high index of suspicion and multidisciplinary evaluation.

Abstract N°: 2866

A Case of Chronic Axonal Polyneuropathy in Patient with Pemphigus Vulgaris

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Introduction & Objectives:

Peripheral neuropathies can manifest with muscle weakness, sensory disturbances, altered reflexes, and neuropathic pain. Chronic axonal polyneuropathy (CAP) is a subtype characterized by progressive axonal degeneration, often presenting with distal sensory deficits and motor weakness. While CAP has been associated with autoimmune diseases, such as Sjögren's syndrome, Behçet's disease, systemic lupus erythematosus, and rheumatoid arthritis. To our knowledge, no clinical cases linking pemphigus vulgaris (PV) with CAP of the lower limbs have been previously reported. The closest known case involves chronic inflammatory demyelinating polyneuropathy (CIDP) in a patient with PV, described by Spanish researchers in 2001, which affected the upper limbs and was demyelinating rather than degenerative in nature. This report aims to present the first documented case of PV associated with CAP affecting the lower limbs.

Materials & Methods:

We report the case of a 58-year-old female with a confirmed diagnosis of PV and concurrent lower limb CAP.

Results:

The patient, originally from Ukraine, was diagnosed with PV three years prior, following a COVID-19 infection, and had been receiving methylprednisolone 24 mg/day. She presented with new-onset lower back pain, bilateral lower limb weakness, and inability to stand.

Dermatological examination revealed erythematous macules, plaques, vesiculobullous lesions, oral erosions, and post-inflammatory hyperpigmentation. Nikolsky sign was positive, and the Pemphigus Disease Area Index (PDAI) score was 22. Serological testing confirmed PV with elevated anti-desmoglein 1 and anti-desmoglein 3 antibodies. Skin biopsy showed intraepidermal blistering with perivascular lymphocytic infiltration, and direct immunofluorescence demonstrated IgG and C3 deposition in intercellular spaces.

Electroneuromyography (ENMG) revealed partial axonopathy involving both motor and sensory fibers. Neurological evaluation showed proximal (2/5) and distal (1–2/5) muscle weakness, absent patellar reflexes, and impaired deep sensation—consistent with CAP and an overlapping myopathic syndrome.

Treatment included azathioprine 100 mg/day and prednisolone 30 mg/day (tapered to <5 mg/day), alongside topical therapies (Octenisept, zinc cream, paraffin gauze) and physiotherapy. Follow-up of patient after 18 months demonstrated marked improvement in PV (PDAI score 15) and neurological symptoms, including restoration of ambulation.

Conclusion:

This case highlights chronic axonal polyneuropathy as a rare, under-recognized neurological condition associated

with pemphigus vulgaris. Early diagnosis through clinical evaluation and ENMG, along with azathioprine therapy, can lead to significant recovery, restoring function and quality of life for patients with PV and CAP.

Abstract N°: 2910

Onychomadesis As a Predictor of Disease Severity in Pemphigus Vulgaris: A Case Report

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Introduction & Objectives:

Pemphigus vulgaris (PV) is an autoimmune mucocutaneous bullous disorder characterised by intraepidermal blistering with suprabasal acantholysis and presence of autoantibodies directed against the adhesion proteins desmoglein 1 and 3 (Dsg1 and Dsg3)1,2. It typically affects individuals between the fourth and sixth decades of life and may be life-threatening if not treated promptly2. Although uncommon, nail involvement may occur and is associated with severe disease and poor therapeutic response1-4.

Materials & Methods:

We report a case of a 35-year-old female, Fitzpatrick phototype IV, with no previous comorbidities, who initially presented with odynophagia and fever (40ºC), without visible oral lesions. She was repeatedly evaluated in primary care and prescribed several antibiotics without improvement. Simultaneously with the onset of oral lesions, she developed paronychia affecting all fingernails with purulent discharge. Two months later, she developed painful, eroded lesions on the buccal mucosa, tongue, gingiva and lips. She sought an urgent care facility, where she was hospitalised for 19 days for the treatment of presumed oral candidiasis with intravenous fluconazole. One week after discharge, she re-presented with vesiculobullous skin lesions and was then referred to a hospital with dermatology service.

On admission, she presented with extensive cutaneous involvement, disseminated blisters and erosions on the skin, mouth, and genital mucosae, in addition to onychomadesis of both halluces. Skin biopsy revealed suprabasal acantholysis with preservation of the basal layer. Direct immunofluorescence showed intercellular IgG deposition in a honeycomb pattern and ductal staining of eccrine glands, confirming PV.

Initial treatment with systemic corticosteroids (1 mg/kg/day) showed no clinical response. Human immunoglobulin (2 g/kg over five days) led to partial improvement, but azathioprine was discontinued due to gastrointestinal intolerance and liver enzyme abnormalities. Immunosuppression and compromised skin barrier contributed to septic shock, precluding further immunoglobulin cycles. After recovery, mycophenolate was introduced with partial response. Complete mucocutaneous re-epithelialisation was achieved following rituximab infusion (1000 mg, repeated after 14 days).

Results:

Nail changes occur in up to 22% of PV cases, more frequently involving thumbs and index fingers3. Although paronychia and onychomadesis are the most common ungual findings, PV may also cause Beau’s lines, trachyonychia, onycholysis, subungual haemorrhages, nail plate discolouration, onychoschizia, and dystrophy4. Local expression of Dsg1 and Dsg3 in the nail is low, and only high titles of desmosomal autoantibodies appear to result in nail involvement. Thus, nail alterations are believed to be markers of severe disease and poor response to standard therapies1-4.

The rarity of nail involvement in autoimmune diseases may be explained by the concept of immune privilege in the nail unit, which limits antigen presentation4. However, thumb and hallux nails are most frequently affected, possibly due to greater susceptibility to trauma and subsequent immune activation3-4.

Conclusion:

Nail involvement may precede, coincide with, or be the sole manifestation of PV. Its presence should prompt thorough evaluation and may serve as a predictor of disease severity and treatment resistance, as illustrated in this case.

Abstract N°: 2978

Bullous pemphigoid induced by Celecoxib

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Introduction:

Bullous pemphigoid (BP) is a rare autoimmune subepithelial blistering dermatosis affecting mostly elderly persons. It can be triggered by certain medications. We report a case of generalized PB secondary to celecoxib.

Case presentation:

A 55-year-old man with moderate mental retardation presented with a generalized, pruritic bullous eruption on the 8th day of treatment with celecoxib, prescribed for osteoarthritis. Clinical examination revealed multiple tense bullae, some with hemorrhagic content, located on erythematous plaques disseminated across the body, including the face, with mucosal involvement in the oral and genital regions. The Nikolsky's sign was negative. Laboratory tests indicated eosinophilia at 2450/mm^3. Histopathological examination showed a subepidermal blister and direct immunofluorescence revealed linear deposits of IgG and C3 along the dermal-epidermal junction. Indirect immunofluorescence testing for serum anti-basement membrane antibodies was positive. A diagnosis of BP was made.** The French drug reaction causality assessment method indicated the responsibility of celecoxib. Given the extent of the lesions, the patient was started on corticosteroid therapy at a dose of 0,5 mg/kg/day, with a rapidly favourable evolution.

Conclusion:

It is important to consider the possibility of a drug-induced in any case of bullous pemphigoid, especially in the presence of mucosal and/or cephalic involvement.

Abstract N°: 2980

Dermatomyositis-like Lesions in a Patient with Still's Disease: An Uncommon Clinical Manifestation

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Introduction:

Still's Disease (SD) is a rare, multisystemic autoinflammatory disease characterized by the involvement of the innate immune.[1–3]

Case Report:

A 42-year-old female patient, previously healthy, presented to the emergency department with a 4-month history of a fixed, pruritic, diffuse maculopapular rash that worsened with sun exposure; daily fever (39°C–40°C) for 3 weeks, more intense at night; chronic polyarthritis; bilateral pleuritic chest pain; periorbital edema and erythema; and lower limb edema. Complementary tests revealed pericarditis with mild pericardial effusion, hepatomegaly, left pleural effusion, left axillary and external iliac chain lymphadenopathy. Bone marrow biopsy showed reactive marrow with no evidence of hemophagocytosis. Skin biopsy revealed epidermis without significant histological changes and a mild superficial and interstitial perivascular inflammatory infiltrate with neutrophil participation, consistent with SD. Laboratory tests showed mild microcytic, normochromic anemia; thrombocytosis; leukocytosis (>10,000/mm^3); negative ANA and rheumatoid factor; elevated transaminases, ferritin, and inflammatory markers. Based on clinical and laboratory findings, the patient met Yamaguchi's criteria for the diagnosis of SD. Dermatomyositis was ruled out.

The patient was refractory to methotrexate, corticosteroids, and cyclosporine, but achieved complete remission for two years with tocilizumab. However, after a 3-month shortage of this medication on the market, she experienced a disease flare, presenting with a fixed, pruritic, diffuse maculopapular rash predominantly in photoexposed areas; flagellate erythema on the knees, back, and inframammary region; bilateral periorbital edema and erythema (heliotrope-like lesions); odynophagia; maculopapular rash on the chest (V-sign); and arthritis in elbows, ankles, and knees. Gottron-like lesions were not observed.

Discussion:

SD typically presents with quotidian spiking fevers, arthralgia or arthritis, evanescent salmon-colored rash, leukocytosis with neutrophilia, hyperferritinemia, and absence of autoantibodies. The pathogenesis involves a dysregulated innate immune response, with prominent roles for cytokines such as IL-1β, IL-6, and IL-18.[1–3]

Typical cutaneous lesions in SD present as a non-pruritic, evanescent, salmon-pink maculopapular rash associated with febrile episodes, which improve with defervescence.[1] However, atypical rashes have recently been described in the literature, with the dermatomyositis-like rash, characterized by heliotrope-like lesions, Gottron-like lesions, flagellate erythema, and the V-sign.[2] Dermatomyositis-like lesions in SD have been associated with higher disease activity and an increased risk of life-threatening complications such as disseminated intravascular coagulation and macrophage activation syndrome. [1–2]

Patients with atypical cutaneous phenotypes may demonstrate suboptimal response to conventional immunosuppressive therapies, as observed in our case. Biologic agents targeting IL-6, such as tocilizumab, have

shown efficacy in refractory SD.

Conclusion:

Dermatomyositis-like skin lesions represent an uncommon but clinically significant manifestation of SD. Recognizing this presentation is crucial for timely diagnosis, appropriate therapeutic escalation, and prevention of potentially fatal complications.

Abstract N°: 2981

Use of dupilumab in refractory bullous pemphigoid: a case series in a tertiary hospital

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Introduction & Objectives:

Bullous pemphigoid (BP) is the most common autoimmune blistering disease, occurring mainly in the elderly. Until now, the main treatment options have been topical or systemic corticosteroids and classic immunosuppressants, with their corresponding side effects, especially considering the age and comorbidities of these patients. In recent years, several cases treated with potentially effective therapeutic alternatives and with a lower associated risk, including dupilumab, have been published.

Material and methods:

We retrospectively collected data on 10 patients with bullous pemphigoid treated with dupilumab. Before starting treatment with dupilumab, 9 of the 10 patients had received methylprednisolone pulses; 5 of them had also received classic immunosuppressants (methotrexate, azathioprine) and one of them had previously received omalizumab. An initial dose of 600 mg was administered to all patients, followed by doses of 300 mg every two weeks.

Results:

In 4 of the 10 patients, dupilumab was used as the sole systemic therapy, thereby achieving disease control. In one patient, methotrexate 12.5 mg weekly had to be associated. In another two patients, a new round of methylprednisolone pulses was needed, and one of them, as well as the 3 most recent cases, continue with associated oral systemic corticosteroid therapy. No patient has presented adverse effects so far.

Discussion and Conclusions

There are many studies indicating that AP is a disease characterized by a predominance of Th2 type immune response, resulting in an overproduction of cytokines such as IL-4, IL-5 and IL-13. In fact, it is considered an autoallergic as well as an autoimmune disease. Dupilumab is a monoclonal antibody that inhibits IL-4 and IL-13 cytokine signaling, whose application in autoimmune blistering diseases was raised in 2019. Currently, the European Academy of Dermatology and Venereology includes it as an optional treatment for moderate-severe BP refractory to conventional therapies. In our experience, dupilumab has proven to be an effective and safe therapy in the treatment of bullous pemphigoid.

Abstract N°: 3043

Linear IgA bullous dermatosis mimicking exuberant erythema multiforme in a woman

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Introduction: Linear IgA bullous dermatosis (LABD) is a rare autoimmune subepidermal blistering disorder that frequently presents with the abrupt onset of tense vesicles and bullae associated with different of varying intensity.

Case report: Female patient, 55 years old, cook, with a history of hypertension presented to the Dermatology outpatient clinic with a 2-months history of pruritic eruption involving the abdomen, arms e legs. This eruption remained unchanged after low doses of Prednisone and Antihistamine therapy. Her medications included Losartan 50mg/day and Hydrochlorothiazide 25mg/day for 5 years. There was no history of antibiotics, nonsteroidal anti-inflammatory drugs or sulfa therapy; recent infection; or known allergy. The dermatological examination revealed confluent erythematous papules and edematous annular plaques with central erosions, suggestive of excoriation, on the abdomen, back, legs and arms. No vesicles or blisters were evident. The oral mucosa was not affected. A diagnosis of Erythema Multiforme was suspected, and biopsy specimens were obtained. The Direct Immunofluorescence (DIF) revealed a linear band of IgA at the dermoepidermal junction.

Conclusion: LABD is a rare, idiopathic or drug-induced autoimmune mucocutaneous disorder characterized by subepidermal bullae and a linear deposit of IgA1 antibodies along the basement membrane zone, which target several different antigens of the dermoepidermal junction. This disorder is immunologically and clinically heterogeneous, showing different clinical features and distribution when in adults or children. Most adults present an abrupt onset of tense vesicles and bullae on sites of inflamed or noninflamed skin, annular lesions demonstrating peripheral vesiculation develop less frequently. Pruritus is common and may be severe, resulting in excoriated papules. The trunk, extensor extremities, buttocks, and face are common sites for lesion development, mucosal sites can be affected. The intensity of the skin lesion is variable and simulate most conditions in the spectrum of autoimmune blistering. Clinical suspicion is essential to guide diagnosis confirmation through biopsy and direct immunofluorescence.

Abstract N°: 3112

A case of Generalized Bullous Pemphigoid with concomitant Irritant Contact Dermatitis in an 18-year-old Filipino male

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Introduction & Objectives:

Rarely seen in children and adolescents, bullous pemphigoid (BP) is the most prevalent autoimmune blistering condition among the elderly. Widespread eczematous plaques, tight bullae, and intense pruritus—occasionally with mucosal involvement—are common symptoms of pediatric BP. BP should be taken into account in the differential diagnosis of vesiculobullous illnesses because of its unusual and diverse appearance in younger people. This case emphasizes important clinical and immunopathologic findings and a unique presentation of bullous pemphigoid in an 18-year-old Filipino male.

Materials & Methods:

We present a case of an 18-year-old male who developed intermittent fever followed by the appearance of multiple erythematous, pruritic plaques on the extremities, later spreading to the trunk and head. Tense vesicles and bullae subsequently developed over these plaques, accompanied by intense pruritus. Nikolsky and Asboe-Hansen signs were negative. The lesions spread, affecting the oral mucosa as well. Self-medication with herbal concoctions worsened the condition, causing erosions and thickening of plaques. Leukocytosis with eosinophilia and significantly increased serum IgE were found in laboratory tests. Histopathology showed a subepidermal split with eosinophilic infiltrate. Direct immunofluorescence revealed linear 1+ IgG and 2+ C3 deposition along the basement membrane zone. Anti-BP180 ELISA was positive, confirming the diagnosis.

Results:

Initial treatment with intravenous hydrocortisone was shifted to oral prednisone (40 mg/day), which was tapered gradually. The patient achieved complete resolution of vesiculobullous lesions with no recurrence at follow-up. There was were post-inflammatory hyper- and hypopigmentation, as well as mild scarring noted.

Conclusion:

Although bullous pemphigoid is predominantly seen in the elderly, it can uncommonly present in adolescents, often with extensive cutaneous and mucosal involvement. High serum IgE may serve as a biomarker for disease activity in pediatric BP. This case reinforces the need for clinicians to maintain a high index of suspicion for BP in young patients presenting with blistering dermatoses and to initiate timely immunosuppressive therapy for effective disease control.

Abstract N°: 3151

The impact of pregnancy on the course of vitiligo – a systematic review

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Title: The impact of pregnancy on the course of vitiligo – a systematic review

Introduction & Objectives:

Vitiligo is a chronic autoimmune skin disorder affecting 0.2-1.8% of the global population. Previous research has shown that there is an interaction between pregnancy and autoimmune disorders. However, evidence regarding the interaction between pregnancy and vitiligo is limited. Pregnant women with vitiligo may face uncertainty about disease progression during pregnancy. A better understanding of the impact of pregnancy on the course of vitiligo can enable healthcare providers to provide more accurate information and offer improved care.

The objective is to systematically examine the available literature with regard to the impact of pregnancy on the course of vitiligo.

Materials & Methods:

A systematic review was conducted in accordance with the PRISMA guidelines. Comprehensive searches were performed in MEDLINE, EMBASE, the Cochrane Library, and Scopus from inception to December 2024. Original studies reporting on the course or onset of vitiligo during pregnancy were included.

Results:

Of the seven included studies, only two cross-sectional survey studies specifically addressed the course of vitiligo during pregnancy. The other studies mentioned it merely as an incidental finding within broader cohorts. The two cross-sectional studies indicate that for the majority of women (approximately 65%), the disease remains stable during pregnancy, with improvement and exacerbation of vitiligo occurring at comparable rates (approximately 12% and 20%, respectively). New onset of vitiligo during pregnancy was rarely reported across all studies. Due to the limited number of studies and lack of prospective data, these findings should be interpreted with caution.

Conclusion:

This systematic review seems to indicate that vitiligo remains stable during pregnancy in the majority of women. The occurrence of new-onset vitiligo during pregnancy appears to be rare. These findings indicate that excessive concern regarding potential disease exacerbation during pregnancy may be unnecessary. Importantly, the overall certainty of the evidence is limited by methodological shortcomings in the available studies. High-quality, prospective research is needed to better understand the course of vitiligo during pregnancy.

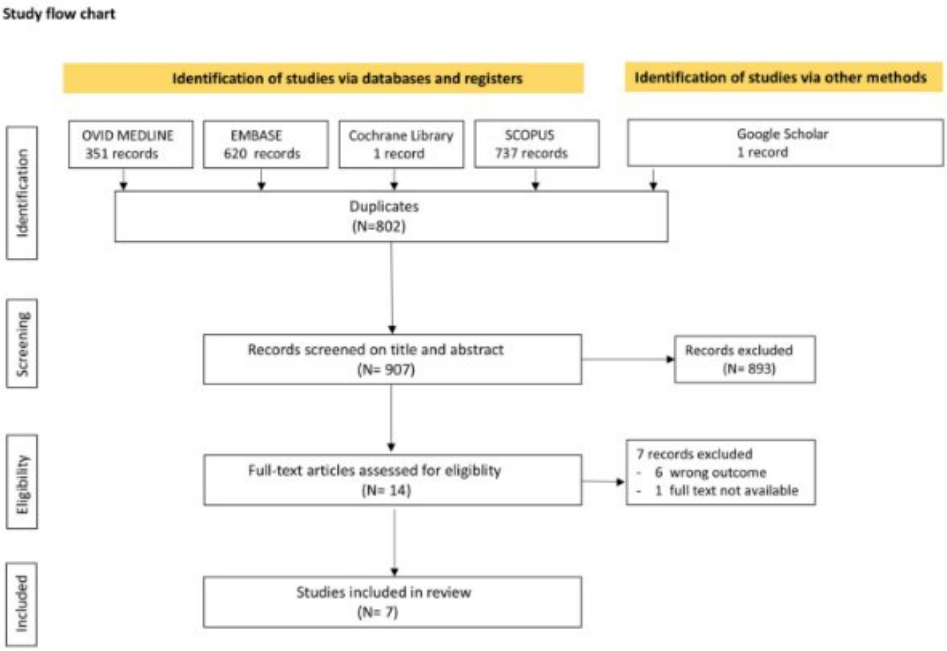

Figure 1. Study flow chart

Abstract N°: 3203

Efficacy and safety of first-in-class oral small molecule toll-like receptor 7/8 inhibitor enpatoran in patients with lupus erythematosus and active cutaneous manifestations: Results of the Phase II WILLOW study

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Introduction & Objectives: Cutaneous lupus erythematosus (CLE) may occur in the presence or absence of systemic lupus erythematosus (SLE) and has high unmet treatment needs. Elevated type I interferon (IFN) gene transcripts in skin lesions highlight the importance of the IFN signalling pathway in disease pathogenesis. Enpatoran is an oral small molecule toll-like receptor (TLR)7/8 inhibitor with potential to modulate IFN activation central to CLE and SLE development. WILLOW (NCT05162586) is a large Phase II randomised double-blind placebo-controlled dose-finding parallel adaptive study evaluating the efficacy, safety and effect on type I IFN gene signature (IFN-GS) of enpatoran in adults with CLE and/or SLE receiving standard of care.

Materials & Methods: Cohort A enrolled patients (pts) with Cutaneous Lupus Disease Area and Severity Index-Activity (CLASI-A) score ≥8 and/or SLE [British Isles Lupus Assessment Group [BILAG] 2004 ≤1B, C, D]) with predominantly active lupus rash. Pts were randomised 1:1:1:1 to one of three doses of enpatoran or placebo for 24 weeks. The primary objective was to evaluate the dose-response relationship of enpatoran in reducing disease activity, based on change from baseline (BL) in CLASI-A score at Week 16, by Multiple Comparison Procedure-Modelling (MCP-Mod). Safety was a secondary endpoint. Cohort B enrolled pts with moderate/severe SLE (BILAG 2004 ≥1A or 2B). Exploratory endpoints included CLASI-A improvement ≥50%/70% (CLASI-50/70; Cohort A and Cohort B subgroup with CLASI-A score ≥8 at BL) and change from BL in type I IFN-GS level (both cohorts) through Week 24.

Results: Pts were randomised (N=102) to Cohort A (safety analysis set) and 100 were evaluated for efficacy (full analysis set; placebo n=26; enpatoran low dose n=23; mid dose n=25; high dose n=26); 77.0% of pts were female, and 58.0% had CLE only. At BL, 59.0% of pts were receiving systemic corticosteroids, 38.0% immunosuppressants and 76.0% antimalarials; 71.0% had moderate-to-severe disease (CLASI-A score ≥10). The primary objective was met: at Week 16, a significant dose response for enpatoran in reducing CLASI-A score from BL was detected (p=0.0002) (Table 1). In Cohort A, up to 87.0%/69.6% of pts receiving enpatoran achieved CLASI-50/70 responses

vs. 30.8%/23.1% for placebo, respectively at Week 24. In the CLASI-A ≥8 subgroup of Cohort B (n=162/354; placebo n=41, enpatoran low dose; n=29; mid dose; n=38; high dose n=54), up to 81.5%/60.5% of pts receiving enpatoran had CLASI-50/70 responses vs. 41.5%/26.8% for placebo, respectively at Week 24. In both cohorts, enpatoran reduced type I IFN-GS level from BL as early as Week 2, maintained to Week 24 (Figure 1). High-dose enpatoran had a higher rate of treatment-emergent adverse events (TEAEs) (80.8%) than mid-dose (57.7%) or low-dose (62.5%) enpatoran, or placebo (46.2%); the most frequent TEAEs were in the system organ class of infections and infestations. Treatment-related TEAEs were reported in 26.9%, 30.8%, 33.3% and 7.7% and serious TEAEs in 3.8%, 0%, 8.3% and 3.8% of pts, respectively.

Conclusion: Cohort A of WILLOW showed a significant dose response with enpatoran in change from BL in CLASI-A vs placebo at Week 16 in pts with CLE and/or SLE with predominantly active lupus rash. In both cohorts, more pts receiving enpatoran than placebo had CLASI-50/70 responses. Reduced IFN-GS levels support TLR7/8 involvement in IFN pathway activation, a hallmark of lupus. Enpatoran was well tolerated.

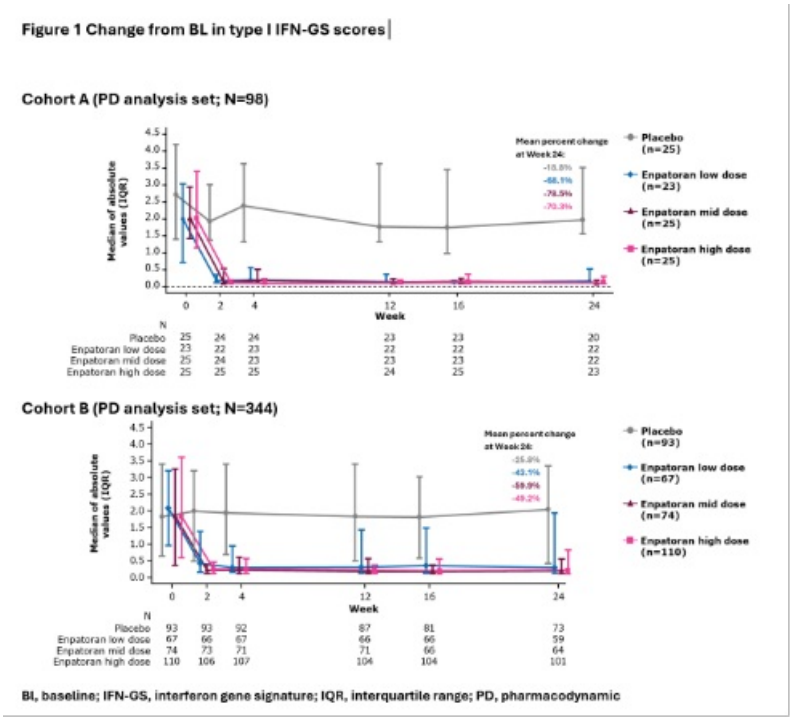
Table 1 Dose-response relationship of enpatoran in reducing disease activity based on change from BL in CLASI-A score at Week 16 (Cohort A FAS; N=100)

	Placebo	Enpatoran dose		
	(n=26)	Low (n=23)	Mid (n=25)	High (n=26)
Primary analysis: Based on MCP-Mod				
Detection of a dose-response relationship		P=0.0002		
Selected model		Log-linear (E0=−44.3, Delta=−6.0)		
Adjusted means in change from BL in CLASI-A score at Week 16, % (95% CI)	−44.3 (−55.1, −33.4)	−63.9 (−70.0, −57.7)	−67.9 (−74.8, −61.1)	−72.0 (−80.1, −64.0)

MCP-Mod adjusted for CLASI-A at BL, region and disease diagnosis (CLE only vs CLE + SLE).

BL, baseline; CI, confidence interval; CLE, cutaneous lupus erythematosus; CLASI-A, Cutaneous Lupus Disease Area and Severity Index-Activity; FAS, full analysis set; MCP-Mod, Multiple Comparison Procedure-Modelling; SLE, systemic lupus erythematosus.

Figure 1 Change from BL in type I IFN-GS scores



BL, baseline; IFN-GS, interferon gene signature; IQR, interquartile range; PD, pharmacodynamic

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Abstract N°: 3239

Mortality and prognostic factors in patients with bullous pemphigoid: A hospital-based retrospective cohort study in China

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Mortality and prognostic factors in patients with bullous pemphigoid: A hospital-based retrospective cohort study in China

AuthorShuzhen Kong,Haoru Niu, Yanhong Gui, Baibing Mi,Bin Peng,songmei Geng

Introduction & Objectives:

Introduction :Bullous pemphigoid (BP) is the most prevalent subepidermal blistering disorder, driven by autoantibodies and predominantly affecting the elderly. It has a profound impact on quality of life and is associated with significant morbidity and mortality. Data on mortality rates in BP remain limited.

Objectives :To summarize the clinical characteristics of BP and assess the cumulative survival rate and prognostic factors affecting overall survival.

Materials & Methods:

This retrospective cohort study included BP patients hospitalized at our institution from March 2017 to March 2024. Data were extracted from electronic medical records, with follow-up conducted by telephone until August 2024.

Results:

The cohort included 404 patients, with a median age at diagnosis of 71.5 years (range: 18-102); 251 (62.1%) were male. Among them, 310 patients (76.7%) had body surface area (BSA) involvement >30%, and 277 (68.6%) were treated with conventional therapies such as glucocorticoids and/or immunosuppressants, while 127 (31.4%) received biological agents or JAK inhibitors during severe disease phases. The median survival time was 18.2 months. The 1-, 3-, and 5-year mortality rates were 16.09%, 29.70%, and 33.66%, respectively. There was no significant difference in survival rates across treatment modalities. Neurological comorbidities and hypertension were associated with higher mortality. Lower albumin (<35 g/L), elevated D-dimer (>1000 ng/mL), and lower hemoglobin (<115 g/L) were linked to increased mortality risk. Multivariate analysis showed patients over 70 years old had a 3.06-fold higher hazard ratio (HR) for mortality compared to younger patients. Neurological disease (HR 2.24, 95% CI [1.55–3.22]) and low albumin (HR 1.96, 95% CI [1.27–3.03]) were also significant predictors of mortality.

Conclusion:

Age, neurological comorbidities, and markers such as albumin, D-dimer, and hemoglobin levels influence prognosis in BP patients, providing essential insights for improving management strategies.

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Abstract N°: 3248

A Rare Case of Juvenile Dermatomyositis in a 2-Year Old Filipino Female with Pulmonary Tuberculosis and Ascariasis

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Introduction:

Juvenile Dermatomyositis (JDM) is a rare type of idiopathic inflammatory myopathy affecting children, characterized by symmetric proximal muscle weakness and pathognomonic cutaneous manifestation such as heliotrope rash and gottron papules. In population-based studies, juvenile dermatomyositis has a reported global annual incidence ranging from two to four cases per one million children. In the Philippines, there were only 40 cases from 2011 to 2022. Juvenile dermatomyositis is known to be an autoimmune disease, but several studies have suggested that systemic infections may play a role in triggering its onset. In cases complicated by systemic infection, early initiation of comprehensive treatment is essential in order to achieve remission.

Case Discussion:

This is a case of a 2-year-old female presenting with a 2-month history of erythematous macules over the metacarpophalangeal (MCP), proximal interphalangeal (PIP), distal interphalangeal joints (DIP), and knees, nailfold changes, and facial erythema on sun exposure. This was associated with decreased activity, inability to walk continuously, and symmetric proximal muscle weakness. Histopathologic findings revealed interface vacuolar dermatitis, perivascular infiltrates composed of lymphocytes and histiocytes, and widened spaces in between collagen bundles with a positive Alcian blue stain. Laboratories revealed elevated ANA, aldolase, LDH, and SGPT which were all consistent with dermatomyositis. Patient was started on oral prednisone (2 mg/kg/day), hydroxychloroquine (200 mg/day) and midpotency topical corticosteroids. Further complicating the clinical presentation, the patient was concurrently diagnosed with pulmonary tuberculosis and ascariasis, presenting with diffuse rales across all lung fields and a distended, globular abdomen, respectively. Chest radiography demonstrated streaky opacities in the bilateral inner lung zones with a nodular opacity probably an adenopathy noted on the right hilar region. Stool examination identified *Ascaris lumbricoides ova* while* whole abdominal ultrasound revealed intestinal parasitism and hepatomegaly with fatty infiltration. Thus, she was also started on anti-Kochs (Isoniazid 10 mg/kg/day, Rifampicin 15 mg/kg/day, and Pyrazinamide 35 mg/kg/day) regimen and mebendazole (100 mg/day, 2x/day for 3 days). After 2 months of steroid therapy, hydroxychloroquine, and anti-Kochs therapy, there was marked improvement in her cutaneous lesions with significant increase in activity and mobility.

Conclusion:

If left untreated, juvenile dermatomyositis can result in a range of complications, including progressive muscle weakness, joint contractures, and cutaneous ulcerations. Timely initiation of treatment is essential to enhance the patient's quality of life at an early stage in the disease course. In countries where pulmonary tuberculosis and ascariasis are highly prevalent, prompt and thorough investigation of underlying infections is imperative, as these may not only coexist with but also potentially trigger the onset of juvenile dermatomyositis.

Abstract N°: 3364

Cardiac Involvement in Dermatomyositis: Successful Management using a Novel IVIg Preparation

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Introduction & Objectives:

Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy characterized by cutaneous, muscular, and systemic involvement. Cardiac involvement, while uncommon and diagnostically challenging, significantly contributes to morbidity and is increasingly recognized as a key determinant of mortality in DM. Despite immunosuppressive therapies such as high-dose corticosteroids (GC) and steroid-sparing agents, managing cardiac involvement remains challenging, and optimal therapeutic strategies are currently still a subject of research. The aim of this case report is to evaluate the clinical efficacy and safety of a novel intravenous immunoglobulin (IVIg) formulation in a multimorbid patient with DM complicated by refractory cardiac manifestations.

Materials & Methods:

A 72-year-old female presented with classic features of DM, including a heliotrope rash, Gottron's papules, and marked muscle weakness, accompanied by dysphagia and exertional dyspnea. Serological testing revealed positivity for TIF-1γ antibodies. In light of the known association with malignancy, a thorough tumor screening was performed, which showed no evidence of an underlying neoplasm. A comprehensive diagnostic work-up further revealed elevated cardiac biomarkers, a peripheral pulmonary embolism, and ECG and cardiac MRI findings consistent with DM-related cardiac involvement. Though the embolism was managed, cardiac biomarkers remained elevated despite high-dose corticosteroids (prednisolone 80 mg/day) and mycophenolate (1 g, 3×/day). Due to insufficient clinical response and considering the patient's history of pulmonary embolism, we initiated treatment with a novel IVIg formulation at a dosage of 2 g/kg body weight over four days, repeated every four weeks. This preparation differs from conventional IVIg by reducing shear stress via vibromixing and eliminating properdin, a complement activator. Thrombogenic factors are removed through caprylic acid and low-pH treatment. In vitro assays confirmed absence of thrombogenic activity and measurable coagulation potential.

Results:

Treatment with IVIg led to a rapid and sustained decline in cardiac enzyme levels, accompanied by significant clinical improvements in muscle strength, exertional dyspnea, and dysphagia. Serial echocardiographic assessments confirmed preserved cardiac function, while long-term ECG monitoring showed a reduction in arrhythmic episodes. Within three months, corticosteroids were tapered to 20 mg/day; mycophenolate to 2 g/day due to lymphopenia, with lymphocytes normalizing thereafter. No adverse events, including thromboembolic or infusion-related complications during IVIg therapy, were observed. The patient has remained clinically stable for ten months under continued IVIg treatment and the current immunosuppressive regimen.

Conclusion: Rapid symptom resolution, combined with a favorable safety profile, supports further investigation into this novel IVIg formulation as part of a personalized treatment strategy, particularly in patients with severe DM manifestations or complex multimorbidity. This is the first documented case of DM-related cardiac involvement with TIF-1γ antibody positivity. These findings underscore the need for individualized treatment strategies in this subgroup.

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Abstract N°: 3397

An atypical presentation of bullous pemphigoid revealing a liposarcoma

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Introduction & Objectives:

Bullous pemphigoid accounts for 70% of subepidermal autoimmune bullous dermatoses, mainly affecting elderly subjects. Its association with cancer remains controversial. We report here a case of bullous pemphigoid occurring simultaneously with a liposarcoma, raising the question of a possible link between these two conditions.

Materials & Methods:

Case report and bibliographic research.

Results:

A 58-year-old man with no significant medical history presented with a generalized pruritic bullous dermatosis that had been progressing for one and a half months. Dermatological examination revealed tense bullae and post-bullous erosions on a pseudo-urticarial background, along with patchy dyschromic and atrophic scars. Nikolsky's sign was negative. Mucosal involvement was limited to a few labial erosions. The affected skin surface was estimated at 80%.Histopathological analysis showed subepidermal detachment accompanied by a polymorphic dermal infiltrate. Direct immunofluorescence demonstrated linear junctional deposits of IgG, C3, IgM, and IgA, while indirect immunofluorescence revealed double positive staining, more pronounced at the roof level. ELISA confirmed the presence of anti-BP180 and BP230 antibodies, establishing the diagnosis of bullous pemphigoid.

During hospitalization, the patient reported right hip pain associated with functional impairment. Clinical examination identified an indurated, tender swelling in the right thigh, adherent to deep tissues. Soft tissue ultrasound revealed a heterogeneous mass suggestive of an aggressive lesion. A surgical biopsy confirmed the diagnosis of liposarcoma. The patient was referred to the orthopedic department for specialized management.

Conclusion:

Our case illustrates an atypical presentation of bullous pemphigoid. The atypical features identified were: the patient's relatively young age, involvement of the head and neck, and double indirect immunofluorescence (IFI) staining. These atypia suggest an induced form of the dermatosis. However, our patient had no previous medical history and had not received any drug treatment in the months preceding the eruption. In this context, the potential role of neoplasia, diagnosed concomitantly with bullous pemphigoid, was evoked. The association between bullous pemphigoid and neoplasia remains debated in the literature. A meta-analysis has suggested a possible association between bullous pemphigoid and certain haematological malignancies, while no significant link with solid tumours has been demonstrated. In some cases the paraneoplastic course of the disease can be an additional argument in favour of a causal link. In a case of bronchopulmonary cancer, the skin lesions completely resolved within two days following lobectomy, without any specific treatment, which strongly suggests a paraneoplastic mechanism. These case reports suggest the value of exploring this association, particularly in the face of atypical presentations.

Abstract N°: 3408

Lupus/ lichen overlap syndrome: a case report

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Introduction & Objectives:

Lupus erythematosus (LE) /lichen planus (LP) overlap syndrome is a rare condition that combines clinical, histological, and immunopathological features of both diseases, with a lack of well-defined diagnostic criteria.

Materials & Methods:

We report a new observation of this rare syndrome.

Results:

A 75-year-old patient with no prior medical history presented to the dermatology department with chronic polymorphic cutaneous lesions on the back and trunk.

Physical examination revealed well-demarcated pink to violaceous plaques, with an annular configuration. The lesions presented central depigmentation or atrophy with a scarring appearance. The borders were hyperpigmented, elevated, and, in some areas, covered with fine scales. The lesions were predominantly located on the trunk and back. Otherwise, there was no mucosal, scalp, or nail involvement, and no gastro-intestinal signs or joint pain.

Dermoscopy revealed a pink to violaceous background with a well-defined brownish border, radial vessels and Wickham's striae-like pattern. Additionally, rosettes were present, along with a central erythematous to violaceous zone and surface scaling**.**

Based on the clinical presentation and the dermoscopic signs, we suspected the diagnosis of lichen/lupus overlap syndrome. A 4mm punch biopsy was performed, consisting with lichen planus. Lab tests revealed positive ANA (antinuclear antibodies) and negative anti-DNA and anti-neutrophilic cytoplasmic antibodies. The renal and hepatic function, c-reactive protein and the complete blood count were normal. Given the clinical presentation, the histological features and the positive ANA, the diagnosis of lichen/lupus overlap syndrome was made and the patient was started on topical corticosteroids and hydroxychloroquine 200mg daily with a significant improvement at 3 months.

Conclusion: LE and LP, two distinct dermatoses, rarely coexist as an overlap syndrome despite the relative prevalence of both conditions. There is ongoing debate on whether LP-LE overlap syndrome constitutes a distinct disorder, an intermediate form, or merely the coexistence of LE and LP lesions in the same patient. While some cases present with separate lesions characteristic of each disease, at least one report describes true overlap within a single lesion. Cutaneous lesions predominantly affect the distal extremities, face, and trunk, with palmoplantar involvement considered a hallmark of the condition. The plaques are typically large, painful, centrally atrophic, and exhibit a bluish red to hypopigmented coloration with a scaly surface.

When evaluating such lesions, dermoscopy is a useful non-invasive technique that can help with diagnosis by showing overlapping dermoscopic patterns. In our case, we found dermoscopic features of both conditions. This

LP-LE overlap syndrome is often underdiagnosed due to its variable presentations. Early recognition is crucial to avoid iatrogenic worsening, as treatments for lichen planus, such as narrowband UVB and PUVA, may worsen cutaneous lupus erythematosus. Treatment of this variant often requires systemic therapy. However, remarkable clinical improvement has been reported with high-potency topical corticosteroids and acitretin. Topical tacrolimus ointment produced a moderate effect without eradicating the lesions. Cyclosporine and hydroxychloroquine have also been reported to be effective.

Abstract N°: 3417

Interesting Associations Between Alopecia Areata and Iron Deficiency Anemia

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Introduction & Objectives:

Alopecia areata (AA) is a chronic autoimmune disorder causing non-scarring hair loss in well-defined patches. Its development is influenced by genetic predisposition, environmental factors, and immune dysregulation, with Th1 and Th17 immune responses playing significant roles. These immune responses disrupt the normal hair cycle, leading to hair follicle damage.

AA is commonly associated with other autoimmune conditions such as autoimmune thyroid diseases, vitiligo, lupus, rheumatoid arthritis, type 1 diabetes, and celiac disease, supporting the idea that AA is part of a broader systemic autoimmune process. These comorbidities can complicate disease management.

The association between iron deficiency anemia (IDA) and AA remains debated. While some smaller studies found no link, recent larger cohort studies and meta-analyses suggest a potential association. Biological explanations include chronic inflammation in AA leading to disrupted iron metabolism, coexistence of other autoimmune conditions that are linked to IDA, and immunosuppressive treatments such as corticosteroids potentially contributing to anemia.

Additionally, iron deficiency itself may impair immune function, particularly T-cell activity, which may worsen AA. Disrupted iron homeostasis may further contribute to immune dysregulation and sustain inflammation, potentially worsening hair follicle damage leading to a vicious cycle, a cycle that reinforces both conditions.

Given the conflicting evidence, this study aimed to clarify the relationship between AA and IDA using a large population-based cohort.

Materials & Methods:

A retrospective case-control study was conducted within a large HMO covering about 2.35 million individuals, including all AA patients diagnosed from 2005 to 2019. AA cases were matched 1:2 with healthy controls based on age and gender. Diagnoses of AA were confirmed by board certificated dermatologists using clinical codes; IDA was identified using ICD-9 codes (280.0, 280.1, 280.8, 280.9) via primary care physicians.

Statistical Analysis :

Analyses were performed using SPSS version 28.0 (IBM, Armonk, NY, USA). The Kruskal–Wallis test was used for continuous variables, while Pearson's $\chi 2$ test or Fisher's exact test were used for categorical variables.

The protocol was approved by an institutional review board.

Results:

The cohort included 33,401 AA patients and 66,802 controls. The average age of onset was 29.9 years, with a

gender distribution of 56.5% males and 43.5% females.(Table 1)

Overall, 11% (11,789 individuals) had IDA: 15% of AA patients (5,080) versus 10% of controls (6,709), showing a significant increase in IDA prevalence among AA patients (Odds Ratio = 1.61, 95% CI: 1.55–1.67, p < 0.01). This association was significant across all age groups. (Table 2)

In terms of diagnosis timing: IDA preceded AA in 40% of cases (2,032 patients),AA preceded IDA in 58% (2,946 patients), both were diagnosed within a 3-month window in 2% of cases (101 patients).

Conclusion:

While IDA has traditionally been viewed as a consequence of AA or its treatments, emerging evidence, suggests IDA may also contribute to the development or worsening of AA by impairing immune regulation.

This study reinforces a strong association between AA and IDA, though causality cannot be established due to the retrospective design.

It is important to recognizing such comorbidities as it may lead to improved management strategies for AA patients.

TABLES:

Table 1 Clinical and demographic characteristics of patients with alopecia areata and controls

		Total (*N* = 100,203)		Control (*N* = 66,802)		Case (*N* = 33,401)		*P*-value
		N	%	*n*	%	*n*	%	
Age (years) at diagnosis (mean +/- standard deviation)		29.9 +/- 16.9		29.9 +/- 16.9		29.9 +/- 16.9		1.000
Gender	Female	43,638	43.5	29,092	43.5	14,546	43.5	1.000
	Male	56,565	56.5	37,710	56.5	18,855	56.5	
Country of birth	Israel	82,873	82.7	53,829	80.6	29,044	87.0	< 0.001
	Other	17,330	17.3	12,973	19.4	4,357	13.0	
Socioeconomic status (scale 1–10 classes)	1–4 (low)	20,711	20.7	13,517	20.2	7,194	21.5	< 0.001
	5–7 (medium	55,184	55.1	36,448	54.6	18,736	56.1	
	8–10 (high)	24,308	24.3	16,837	25.2	7,471	22.4	

SES (socioeconomic status) was defined according to the Israeli Central Bureau of Statistics (scale 1-10), combining geographic and socioeconomic information for each neighborhood in Isael.

Table 2: Odds ratios for Iron deficiency anemia of Alopecia Areata patients and controls in different age groups

	Alopecia areata	**Iron deficiency anemia**				**OR (95%CI)**	**p-value**
		No		**Yes**			
		N	**%**	**N**	**%**		
All (N=100,203)	Control 66802	60093	89.96	6709	10.04	1.61 [1.55-1.67]	<0.001
	Case 33401	28321	84.79	5080	15.21		
Age <18 (N=27363)	Control 18242	16476	90.32	1766	9.68		
	Case 9121	7787	85.38	1334	14.62	1.60 (1.48-1.73)	<0.001
Age 18-40 (N=44169)	Control 29446	26931	91.46	2515	8.54		
	Case 14723	12756	86.64	1967	13.36	1.65 (1.55-1.76)	<0.001
Age > 40 (N=28671)	Control 19114	16686	87.3	2428	12.7		
	Case 9557	7778	81.39	1779	18.61	1.57(1.47-1.58)	<0.001

Abstract N°: 3524

Epidemiological, Clinical, and Immunological Profile of Bullous Pemphigoid with Double Staining Patterns Assayed by Indirect Immunofluorescence (IIF): A Study of 19 Cases

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Introduction & Objectives:

Bullous pemphigoid (BP) is a subepidermal autoimmune blistering disease characterized by autoantibodies targeting hemidesmosomal proteins, leading to junctional separation. In its typical form, indirect immunofluorescence (IIF) on salt-split skin demonstrates IgG deposition exclusively on the epidermal side (roof). In this study, we focus on patients diagnosed with BP according to the Vaillant criteria, who presented with an atypical IIF pattern showing double staining patterns assayed on both the epidermal (roof) and dermal (floor) sides. The objective of our study is to describe the epidemiological, clinical, and immunological characteristics of these patients with this dual IIF pattern.

Materials & Methods:

We conducted a retrospective descriptive study of BP cases with double staining patterns assayed by IIF in the Tunis region between 2020 and 2025.

Results:

A total of 19 patients were included, with a mean age of 69.8 years and a male-to-female ratio of 0.42. A neuropsychiatric history was present in 31% of cases. Pruritus was reported in 76% of patients. Limb involvement was a constant feature, predominantly affecting flexural surfaces (89%) compared to extensor surfaces (32%). Truncal involvement was observed in 83% of cases, while head and neck involvement was noted in 58%. Mucosal involvement was present in approximately 40% of cases, and skin appendage involvement was observed in 16%. Clinically, the disease presented with tense blisters on non-inflamed skin in 68% of cases, which evolved into dyschromic scars (95%) and atrophic scars (37%). Direct immunofluorescence (DIF) showed IgG and C3 deposition in 100% of cases, with additional IgA deposits in 47% and IgM in 37% of cases. ELISA testing revealed anti-BP180 antibodies in 72% of patients, while anti-BP230 antibodies were positive in only 17%. Notably, 21% of patients had neither anti-BP180 nor anti-BP230 autoantibodies.

Conclusion:

Comparing our findings to the literature on classic BP (which exhibits exclusive roof staining on IIF), no significant differences were observed in epidemiological features: the mean age remained around 70 years, neuropsychiatric comorbidities were common, and pruritus was a frequent symptom. However, the clinical presentation diverged from classic BP. In our series, blisters predominantly arose on intact skin, whereas in conventional BP, blisters typically develop on urticarial or erythematous bases. Additionally, head and neck involvement, which is uncommon in classic BP, was present in over half of our cases. Mucosal involvement, reported in 10–20% of BP cases in the literature, was markedly higher in our study (exceeding 40%). Immunologically, DIF findings were consistent with those of classic BP, showing C3 and IgG deposition with variable presence of IgA and IgM. However, a notable discrepancy was observed in serological profiles: while anti-BP180 was detected in the

majority of cases, anti-BP230, which is positive in approximately 60% of classic BP cases, was infrequent in our cohort. These results suggest that there is a relationship between the presence of anti-BP180 antibody and combined patterns of IIF-SSS. The combined pattern may be due to the presence of antibodies in the patients' sera against the multiple antigenic epitopes along the BP180.

Abstract N°: 3545

Epidemiological and Clinical Profile of Bullous Pemphigoid: A Retrospective Series of 45 Cases

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Introduction & Objectives:

Bullous pemphigoid (BP) is an autoimmune subepidermal blistering disorder affecting the dermo-epidermal junction. Although it remains a rare condition, BP is the most common subepidermal autoimmune bullous disease. The objective of this study is to analyze the epidemiological and clinical profile of BP.

Materials & Methods:

We conducted a bicentric, retrospective, descriptive study including all BP cases managed at the dermatology departments of La Rabta Hospital and Charles Nicolle Hospital (HCN) in Tunis between 2019 and 2024.

Results:

A total of 45 patients were included. The mean age was 75 years, with 69% of patients being over 70 years old. The male-to-female ratio was 0.44. A history of neuropsychiatric disorders was present in 17% of cases (n = 8), with stroke accounting for half of these cases. An association with other autoimmune diseases was found in 9% of cases. Pruritus was reported in 80% of cases (36 patients). Clinically, 100% of patients presented with tense blisters, which were located on erythematous skin in 75% of cases, urticarial in 34%, and eczematous in 16%. Prurigo-like lesions were associated in 20% of cases. Post-inflammatory hyperpigmentation was the predominant mode of healing, observed in 93% of cases, followed by atrophic scars (32%) and milia formation (5%). The distribution of lesions was as follows: Flexural limb surfaces (93%), upper and lower limbs (87%), trunk (77%), acral (51%), head and neck (22%) and scalp (13%). Mucosal involvement was observed in 31% of cases, mainly affecting the oral mucosa, while 9% had genital mucosal lesions.

Conclusion:

Our findings align with the existing literature: BP predominantly affects individuals over 70 years old, with no clear gender predominance. Neurological disorders, particularly stroke, are recognized risk factors. Pruritus is a frequent symptom, and flexural limb surfaces remain the most commonly affected sites.

Abstract N°: 3646

Immunophenotypic and Transcriptomic Analysis of Peripheral Blood Mononuclear Cells in Bullous Pemphigoid

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Introduction & Objectives:

Bullous pemphigoid (BP) is an autoimmune blistering disease driven by autoantibodies against BP180 and BP230. While type 2 inflammation plays a key role, the precise immune cell alterations and transcriptomic changes remain unclear. We examined the immune cell composition and transcriptomic changes in BP patients using fluorescence-activated cell sorting (FACS)-based immunophenotyping and RNA sequencing.

Materials & Methods:

A case-control study was conducted on 10 newly diagnosed, treatment-naive BP patients and six healthy controls. Disease activity was assessed using the Bullous Pemphigoid Disease Area Index (BPDAI). Peripheral blood mononuclear cells were isolated for FACS analysis to determine immune cell subsets. RNA sequencing was performed to identify differentially expressed genes (DEGs) and enriched pathways. Statistical analyses included t-tests, Mann-Whitney U tests, and correlation analysis.

Results:

FACS analysis revealed a reduction in CD4+ T cells, Th2, and B cells in BP patients ($p < 0.05$), alongside an increase in M2a-like monocytes ($p < 0.001$). RNA sequencing identified 262 DEGs, with secretory leukocyte peptidase inhibitor (SLPI) and transmembrane protein 237 (TMEM237) being the most significantly upregulated. Proline-serine-threonine phosphatase interacting protein 2 (PSTPIP2) and SAM domain, SH3 domain, and nuclear localization signals 1 (SAMSN1) positively correlated with BPDAI ($p < 0.001$). Gene ontology analysis highlighted enrichment in inflammatory responses and neutrophil degranulation pathways.

Conclusion:

BP patients show distinct immune dysregulation, including decreased CD4+ T cells, Th2, and B cells, increased M2a-like monocytes, altered gene expression profiles, and correlations between PSTPIP2, SAMSN1 and disease activity. These findings provide insights into pathogenesis and potential therapeutic targets of BP.

Abstract N°: 3720

Ulcerative Dermatomyositis with Anti–NXP2 Positivity: A Case Report Highlighting Therapeutic Challenges

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Introduction & Objectives:

Dermatomyositis (DM) is an idiopathic inflammatory myopathy marked by cutaneous findings and proximal muscle weakness . Among the various autoantibodies associated with DM, anti–NXP-2 (nuclear matrix protein 2) is linked to severe muscle disease, malignancy risk, and occasionally calcinosis or distal ulcers . On the other hand, anti-MDA5 (melanoma differentiation–associated protein 5) antibodies are typically associated with the clinically amyopathic subtype of DM and are associated with skin ulcers and rapidly progressive interstitial lung disease (ILD). Although ulcerative lesions are more commonly linked to anti-MDA5 antibodies, ulcerative skin involvement secondary to vasculopathy may also occur in anti–NXP-2–positive DM. Calcinosis is a known feature of anti-NXP2–positive DM, with soft tissue calcium impairing perfusion or causing local reactions, thereby contributing to ulcer formation. Skin biopsies from anti-NXP2–positive DM cases with cutaneous ulceration generally do not reveal specific histopathological findings . We report a rare anti–NXP-2 positive, anti–MDA5 negative DM case with therapy-resistant ulcers and discuss clinical and therapeutic implications.

Materials & Methods:

The clinical records of a patient diagnosed with DM presenting with therapy-resistant ulcers were reviewed.

Results:

A 50-year-old male with DM presented with painful ulcers on the extremities and back. Dermatologic examination revealed a milk-white atrophic plaque with perifollicular erythema and scale on the left parietal scalp, subungual telangiectasias, shawl sign, V sign, Gottron’s sign, and pruritus. CK was elevated; ANA, anti-Ro52, and anti–NXP-2 antibodies were positive. EMG showed myopathic changes. Biopsy demonstrated ulceration, pseudoepitheliomatous hyperplasia, vacuolar alteration, perineural lymphoplasmacytic infiltration, fat necrosis with dystrophic calcification, and dermal fibrosis. X-ray showed soft tissue calcifications. Chest CT ruled out ILD. Hematologic and gastrointestinal malignancies were excluded via peripheral smear, endoscopy, colonoscopy, and abdominal ultrasound. The ulcers were unresponsive to colchicine, corticosteroids, azathioprine, and rituximab. Marked improvement was observed after IVIG (2 g/kg), hydroxychloroquine, and a calcium channel blocker. Clinical follow-up confirmed progressive healing

Conclusion:

In conclusion, ulcer development in anti–NXP2–positive, anti–MDA5–negative DM is rare but clinically significant. In the presence of ulcers, the severity of underlying vasculopathic processes should be considered, and patients should be screened for multisystem complications with reassessment of treatment intensity [. Early recognition of atypical cutaneous manifestations and individualized treatment strategies are critical to reducing morbidity. Timely transition to multidisciplinary care and the use of therapies such as biologics and intravenous immunoglobulin (IVIG) may improve patient outcomes.

Abstract N°: 3791

Lewis blood group antigens and ABH secretor status in patients with Pemphigus

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Introduction & Objectives: Pemphigus forms a group of chronic autoimmune blistering diseases of the skin and mucosal surfaces. Ethnic clustering, familial aggregation and autoimmune comorbidity, suggest a genetic contribution to the etiopathogenesis of Pemphigus. The majority of identified genes belong to the HLA locus, while there is limited data regarding the association of non-HLA genes. Human histo-blood group antigens as haematological markers have been assessed in autoimmune and mucocutaneous diseases; however, conflicting conclusions exist regarding the linkage between susceptibility to Pemphigus and Lewis blood group antigens co-associated secretor status. The genes we inherit at the ABO, Lewis and Secretor loci control the glycoconjugate profile of our sialylated antigens, which are hypothesized to influence the risk, development and severity of various medical conditions. This study aimed to investigate and evaluate Lewis blood group phenotypes and secretor status in relation to the clinico-immunopathological profile of Pemphigus for the first time in the Greek population, one of the protagonists in the disease' global map distribution.

Materials & Methods: Thirty-six patients and 36 age and gender - matched control subjects were recruited to the study. Patients were followed up at the Center of Expertise on Autoimmune Bullous Diseases within the 2nd Dermatology Department of the Aristotle University of Thessaloniki. 2ml blood and 3ml saliva were collected from each subject, in order to identify Lewis phenotypes and determine secretor status, using monoclonal gel cards and hemagglutination inhibition technique respectively.

Results: All Pemphigus Foliaceus (PF) and Pemphigus Vulgaris (PV) cutaneous type patients were Lea(−) secretors. Statistically significant differences were observed between Lewis phenotypes and (i) Pemphigus type, (ii) clinical involvement. Rare Le(a−b−) phenotype was higher in PF compared to PV patients, as well as in cutaneous compared to mucocutaneous Pemphigus, while Le(a+b−) presented dominantly in PV patients with mucocutaneous lesions. A negative correlation between Lea and Leb antigen expression was observed.

Conclusion: The relationship between blood groups and Pemphigus remains unclear and contentious. Rare Lewis phenotype Le(a−b−), especially Lea(−) negative antigen appears to be a predisposing risk factor to the development of Pemphigus, characterized exclusively by skin lesions. The pathogenic heterogeneity of anti-desmoglein antibodies, along with blood group carbohydrate antigens, may reinforce an individual's susceptibility to a cutaneous subtype. Further research should be conducted with a larger sample size to determine whether these findings, along with the routine use of a Lewis and secretor status test, may improve the initial diagnostic and prognostic evaluation of Pemphigus patients.

Abstract N°: 3798

Diagnostic Value of Cutaneous Direct Immunofluorescence Intensity for Renal Involvement in IgA Vasculitis: A Retrospective Cohort Study

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Introduction & Objectives: Renal involvement, occurring in 20-54% of IgA vasculitis (IgAV) patients, remains a critical determinant of long-term prognosis. Current predictive models lack reliable biomarkers for early identification of high-risk patients. This study investigated the clinical utility of cutaneous vessel immunofluorescence intensity as a novel predictor of nephropathy progression.

Materials & Methods: In this retrospective cohort study, we analyzed 104 biopsy-confirmed IgAV patients (2012-2024) from Northwest China who underwent standardized direct immunofluorescence (DIF) testing. Immunofluorescence parameters (deposition area, mean intensity, and integrated density of IgA complexes) were quantified using digital pathology systems. Renal involvement was defined as ≥0.5g/24h proteinuria and/or ≥5 RBCs/HPF. Multivariable logistic regression and ROC analyses were performed to evaluate diagnostic performance.

Results: DIF characterization revealed IgA deposition in 81% (84/104), C3 in 65% (68/104), with complete IgG negativity. Quantitative DIF parameters demonstrated moderate diagnostic value for renal involvement: IgA deposition area (AUC 0.72, 95%CI 0.60-0.86, *P*=0.004)**,** mean intensity (AUC 0.70, 95%CI 0.56-0.84, *P*=0.011), and integrated density (AUC 0.67, 95%CI 0.53-0.82, *P*=0.028).** The combination model with abdominal pain/arthritis showed limited improvement (AUC 0.77 vs 0.72 for IgA area alone, *P*=0.12 for comparison). The observed AUC ranges (0.67-0.77) suggest potential as supplementary indicators rather than standalone diagnostic tools.

Conclusion: While cutaneous IgA deposition patterns show promising associations with renal outcomes in IgAV patients, their clinical utility requires confirmation in prospective cohorts.

Abstract N°: 3901

When the immune system doesn't follow the script: Clinical-immunological mismatch in autoimmune dermatology

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Introduction & Objectives:

Systemic sclerosis is a chronic connective tissue disease characterized by autoimmune systemic dysregulation centered by vasculopathy, with Raynaud phenomenon as main onset clinical sign and progressive organ fibrosis.

Regarding the immune profile, not less one hundred antibodies were described in autoimmune diseases, of which three were included in the SSc classification criteria (namely, anti-centromere or ACA, anti-topoisomerase I or anti-Scl-70, and anti-RNA polymerase III) due to their increased specificity for this pathology phenotype. After screening for ANA by indirect immunofluorescence assay (IFA) on Hep-2 cells, confirmation by immunoblot-based is used for confirmation.

Materials & Methods:

A 57-year-old male patient, a heavy smoker with no history of occupational exposure to toxic agents, presented with morning stiffness in the small joints of the hands. Clinical examination revealed scleroderma-like skin changes, with a modified Rodnan skin score of 32, and facial telangiectasias.

The patient reported exertional dyspnea, and high-resolution chest computed tomography (HRCT) was suggestive of newly diagnosed with non-specific interstitial pneumonia (NSIP) interstitial lung disease (ILD), while the pulmonary function tests revealed a mildly reduced diffusing capacity for carbon monoxide (DLCO 54%). Echocardiographic and electrocardiographic evaluation revealed evidence of ischemic heart disease, with associated segmental wall motion abnormalities. A muscle biopsy performed on the deltoid muscle revealed a necrotizing-regenerative myopathy, with overexpression of MHC class I and minimal inflammatory changes, while skin biopsy was suggestive of scleroderma.

Laboratory investigations showed positive ANA at a high titer (1:2560) by IIF and two ICAP patterns AC-4 and AC-18. Both ACA and anti-Scl-70 antibodies were negative, as were immunoblot profiles for both myositis and scleroderma, suggesting the positivity of a rare ANA. Infectious disease screening, including HIV and viral hepatitis, was negative. Vitamin D level was markedly reduced (8 ng/mL), while the rest of the nutritional panel was within normal limits. Additionally, serum levels of creatine kinase (CK), CK-MB, and high-sensitivity troponin T (hs-TnT) were elevated (while normal hs-cTnI).

Results:

A diagnosis of an overlap connective tissue disease was supported, combining clinical features of SSc and idiopathic inflammatory myopathy (IIM), with multisystem involvement including skin, muscle, and interstitial lung disease. The workup was completed with malignancy screening and a comprehensive cardiological evaluation for ischemic heart disease. Treatment was initiated with low-dose corticosteroids, mycophenolate mofetil, nintedanib, a calcium channel blocker, an antiplatelet agent, a statin, and vitamin D supplementation.

Conclusion:

This case illustrates a rare and complex presentation of an overlap connective tissue disease positive for ANA IIF in high titer but negative for specific SSC or IIM antibodies, with clinical features consistent with SSc and IIM.

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Abstract N°: 3949

Successful Management of Severe Refractory IgA Pemphigus with Adalimumab: A Case Report.

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Title: Successful Management of Severe Refractory IgA Pemphigus with Adalimumab: A Case Report.

Introduction & Objectives: Immunoglobulin A (IgA) pemphigus is a rare autoimmune blistering disease characterized by vesiculopustular lesions resulting from IgA autoantibodies targeting epidermal components and subsequent neutrophilic infiltration. Treatment can be challenging due to the lack of standardized guidelines and frequent refractoriness to conventional therapies, including corticosteroids and dapsone. This report aims to present a case of severe, treatment-refractory IgA pemphigus successfully managed with adalimumab.

Materials & Methods: A 76-year-old woman presented with a three-month history of extensive, pruritic vesiculopustular plaques and erosions. Diagnosis of intraepidermal neutrophilic IgA pemphigus was confirmed by histology (subcorneal/intraepidermal pustules with neutrophils) and direct immunofluorescence (intercellular IgA deposition). Indirect immunofluorescence was positive for anti-desmoglein 1 and anti-desmoglein 3 antibodies. The patient had previously failed treatments with acitretin, prednisone, dapsone, cyclosporine, and rituximab, leading to hospitalization with erythroderma: Body Surface Area (BSA) 90%, Pemphigus Disease Area Index - Activity (PDAI-A) 128, Pruritus Visual Analogue Scale (VAS) 10/10. Adalimumab was initiated (80 mg loading dose, then 40 mg every two weeks) adjunctively to prednisone 10 mg/day and dapsone 100 mg/day.

Results: Two months after initiating adalimumab, the patient demonstrated a dramatic clinical improvement, achieving near-complete remission with BSA <1%, PDAI 1, Pruritus VAS 1/10, and Quality of Life VAS 0/10. This significant response was maintained throughout a 6-month follow-up period. No adverse effects related to adalimumab were observed during this time.

Conclusion: Adalimumab, targeting Tumor Necrosis Factor-α (TNF-α), which plays a crucial role in neutrophil recruitment and survival, proved highly effective in inducing rapid and substantial remission in this case of severe, refractory IgA pemphigus. The patient experienced dramatic improvements in skin lesions, symptomatology, and quality of life within two months, a response maintained over the follow-up period with no observed adverse effects. This case contributes to the limited but growing body of evidence suggesting that adalimumab may represent a valuable therapeutic option for challenging IgA pemphigus cases unresponsive to conventional treatments. However, given the rarity of the disease and scarcity of large-scale data, further investigation, including comparative studies, is warranted to confirm these findings, establish its precise role, optimal dosing regimens, long-term efficacy and safety, and to explore strategies for potential treatment de-escalation or discontinuation in the management algorithm for IgA pemphigus.

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Abstract N°: 3969

Rapid Improvement of Cutaneous Dermatomyositis with Brepocitinib: A 12-Week Open-Label Trial

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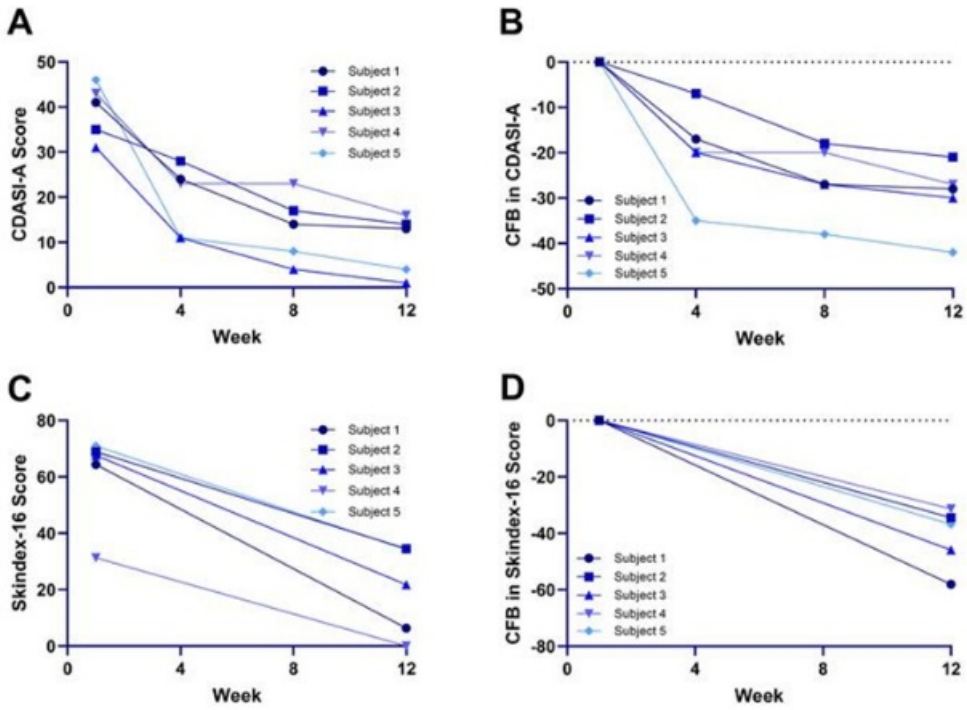
Introduction & Objectives: Dermatomyositis (DM) is an idiopathic, multisystem autoimmune disease associated with significant morbidity for which effective therapies are limited. Cutaneous involvement in DM can be particularly challenging to treat and often persists despite adequate control of muscle symptoms. Brepocitinib is a once-daily, oral inhibitor of TYK2/JAK1 that is thought to antagonize proinflammatory cytokines central to DM immunopathogenesis, including type I/II interferon [IFN], interleukin (IL)-6, IL-12, and IL-23. The objective of this trial was to evaluate the efficacy and safety of oral brepocitinib in adults with recalcitrant, skin-predominant DM.

Materials & Methods: This 12-week, open-label, single-arm trial was conducted at a single center in the United States from August 2024 to February 2025 (NCT06433999). Participants were required to have moderate-to-severe cutaneous DM, defined as a Cutaneous Disease Activity Severity Index Activity (CDASI-A) score ≥14 despite treatment with systemic corticosteroids, non-steroidal immunosuppressives, and/or antimalarial agents, and skin-predominance, defined as no more than minimal muscle disease. Participants received oral brepocitinib 30 mg once daily for 12 weeks. The primary outcome was the change from baseline in CDASI-A score at 12 weeks, where the minimal clinically important difference has been reported as a decrease of 4-7.8 points or 40% from baseline. Secondary endpoints included change in patient reported outcomes (including Patient Skin Activity Visual Analogue Scale [PtGA-skin] and Skindex-16) and safety and tolerability.

Results: N=5 participants with recalcitrant skin-predominant DM (4 female; mean [SD] age, 53 [10] years; mean [SD] CDASI-A, 40.2 [6.1]) were enrolled. All participants demonstrated rapid and substantial improvement in CDASI-A and other measures of skin disease activity (**Figure 1**). Mean change [SD] and mean percentage change [SD] in CDASI-A scores from baseline were -20.8 [8.4] and -51.8% [17.8%] at week 4, and -30.6 [6.4] and -77.0% [15.8%] at week 12, respectively. Similar reductions from baseline in PtGA-skin (mean [SD], 5.1 [1.8]) and Skindex-16 (mean [SD], 41.3 [10.8]) scores were observed at week 12. Brepocitinib was well tolerated with no serious adverse events (SAEs) or discontinuations due to AEs.

Conclusion: In a population with recalcitrant, severe cutaneous DM, brepocitinib was associated with rapid, clinically meaningful improvement in CDASI-A in all participants during 12 weeks of treatment. Consistent benefits were observed in other clinician- and patient-reported measures of skin activity. The results of a global phase III, randomized, placebo-controlled, dose-ranging, 52-week trial of brepocitinib in adult DM patients with skin and muscle disease (VALOR, NCT0543726) are forthcoming and will further clarify the potential of brepocitinib for the treatment of DM.

Figure 1: Clinical Response to Brepocitinib in Skin-Predominant Dermatomyositis Absolute change (A, C) and change from baseline (B, D) in CDASI-A and Skindex-16 scores during treatment.



Abbreviations:

CFB = change from baseline; CDASI-A = Cutaneous Dermatomyositis Disease Area and Severity Index – Activity

Abstract N°: 4037

Evaluating the Connection Between Vitiligo and the Severity of Major Depressive Disorder

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Introduction & Objectives: One prevalent dermatological condition that has a substantial psychological impact is vitiligo. This study examines the connection between the severity of major depressive disorder (MDD) and vitiligo, emphasizing the wider effects on the mental health of those who are impacted. Our objectives are to determine the prevalence and determinants of depression in adult vitiligo patients and investigate the connection between vitiligo and the severity of MDD.

Materials & Methods: The study employed a cross-sectional design and measured the degree of depression and vitiligo using the Patient Health Questionnaire-9 and the Vitiligo Area Severity Index, respectively. 60 vitiligo patients with diagnoses from different healthcare settings participated in this study. The impact of sociodemographic factors was assessed using logistic and ordinal regression analysis with factors and forms of vitiligo on the severity of MDD.

Results: Among the subjects, 51.6% had MDD. Significant differences existed in the severity of depression: 25% of patients had mild depression, 20% had moderate depression, 16,6% had moderately severe depression, and 11.66% had severe depression. Compared to male patients, female patients were more likely to experience severe depression . Depression severity was negatively correlated with age; patients over 60 had much lower odds . Higher depression severity was linked to lower income . Types of vitiligo also affected the degree of depression; acrofacial vitiligo and vulgaris were significantly linked to higher depression levels than focal vitiligo.

Conclusion: According to the research, vitiligo may raise the chance of developing severe depression, underscoring the necessity of combined dermatological and psychological therapy strategies to address the condition’s effects on both physical and mental health.

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Abstract N°: 4039

Mortality and prognostic factors in patients with bullous pemphigoid: A hospital-based retrospective cohort study in China

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Introduction & Objectives:

Introduction :Bullous pemphigoid (BP) is the most prevalent subepidermal blistering disorder, driven by autoantibodies and predominantly affecting the elderly. It has a profound impact on quality of life and is associated with significant morbidity and mortality. Data on mortality rates in BP remain limited.

Objectives :To summarize the clinical characteristics of BP and assess the cumulative survival rate and prognostic factors affecting overall survival.

Materials & Methods:

This retrospective cohort study included BP patients hospitalized at our institution from March 2017 to March 2024. Data were extracted from electronic medical records, with follow-up conducted by telephone until August 2024.

Results:

The cohort included 404 patients, with a median age at diagnosis of 71.5 years (range: 18-102); 251 (62.1%) were male. Among them, 310 patients (76.7%) had body surface area (BSA) involvement >30%, and 277 (68.6%) were treated with conventional therapies such as glucocorticoids and/or immunosuppressants, while 127 (31.4%) received biological agents or JAK inhibitors during severe disease phases. The median survival time was 18.2 months. The 1-, 3-, and 5-year mortality rates were 16.09%, 29.70%, and 33.66%, respectively. There was no significant difference in survival rates across treatment modalities. Neurological comorbidities and hypertension were associated with higher mortality. Lower albumin (<35 g/L), elevated D-dimer (>1000 ng/mL), and lower hemoglobin (<115 g/L) were linked to increased mortality risk. Multivariate analysis showed patients over 70 years old had a 3.06-fold higher hazard ratio (HR) for mortality compared to younger patients. Neurological disease (HR 2.24, 95% CI [1.55–3.22]) and low albumin (HR 1.96, 95% CI [1.27–3.03]) were also significant predictors of mortality.

Conclusion:

Age, neurological comorbidities, and markers such as albumin, D-dimer, and hemoglobin levels influence prognosis in BP patients, providing essential insights for improving management strategies.

Abstract N°: 4104

Anifrolumab and deucravacitinib as novel therapeutic options for cutaneous lupus erythematosus: utility in multi-refractory patients and safety regarding latent tuberculosis infection

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Introduction & Objectives: Anifrolumab is a biologic drug that blocks the 1 subunit of the type 1 interferon receptor. Deucravacitinib blocks TYK2-mediated inflammation. Boths treatments have increasing evidence for the treatment of cutaneous lupus erythematosus (CLE). We present a case series of cutaneous lupus in patients without systemic lupus criteria, multirefractory of with comorbidities (latent tuberculosis infection), who were successfully treated with anifrolumab or deucravacitinib.

Materials & Methods: Presentation of a case series. Effectiveness data were collected using the Cutaneous Lupus Erythematosus Disease Area and Severity Index -CLASI- scale, as well as treatment safety data.

Results: Three female patients were included. Two patients were treated with anifrolumab off-label after obtaining their consent for treatment. The age of patients was of 53 and 43 years old, and both had chronic cutaneous lupus with CLASI-A>10. In the first case, the patient had been refractory to treatment with methotrexate, acitretin, hydroxychloroquine and rituximab. In the second case, a latent tuberculous infection was detected in the patient and she was treated with rifampicin 600mg per day for 4 consecutive months. Anifrolumab was initiated one month after starting treatment with rifampicin. In both cases a dose of 300mg intravenous every 4 weeks was administered, observing a striking and rapid improvement of the skin involvement after the first infusion, decreasing significantly the CLASI-A to 3 after 1 dose in both cases. No relevant side effects were observed during treatment, except headache in one of the patients. Another 44 year-old woman was treated with 6mg daily deucravacitinib because of concurrence of cutaneous discoid lupus, positive antinuclear antibodies and psoriasis. The patient had been unresponsive to methotrexate. After three months of treatment basal CLASI-A score decreased from 7 to 2, showing also an improvement in psoriasis skin lesions.

Conclusion: Anifrolumab is a drug currently approved for use in systemic lupus. In our experience, its use in CLE without associated systemic lupus is effective, even in multirefractory cases. The cutaneous improvement seems to be rapid from the first infusions. In our case, no alarming safety data have been observed when treating a patient with latent tuberculosis infection treated. Deucravacitinib seems to be effective to treat CLE, specially in those cases with concurrent psoriasis or psoriatic arthritis.

Abstract N°: 4111

Association of Bimekizumab for refractory pemphigus foliaceus. A report of two cases.

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Introduction & Objectives:

Pemphigus foliaceus (PF) is a rare autoimmune blistering disease mediated by autoantibodies against desmoglein-1 (Dsg1). While rituximab remains a cornerstone therapy, a subset of patients remains refractory to treatment. Interleukin-17 (IL-17) is an important pro-inflammatory cytokine related to many immune-mediated diseases. In the last years, in vitro studies have demonstrated an overexpression of CD4+ tissue-resident memory T (TRM) cells in pemphigus skin lesions with upregulated blood levels of IL17 in patients with active disease. Bimekizumab, a monoclonal antibody targeting IL-17A and IL-17F, has shown efficacy in psoriasis but has not been previously reported in PF. We present two cases of treatment-resistant PF successfully managed with off-label bimekizumab.

Materials & Methods:

We present two male patients (ages 57 and 43) with longstanding PF refractory to corticosteroids, immunosuppressants, IVIG, and rituximab. They were initiated on off-label bimekizumab (320 mg subcutaneously every 8 weeks) with clinical and serological response.

Results:

Patient 1: A 57-year-old man with PF since 2018, previously treated with multiple cycles of rituximab, IVIG, mycophenolate, and prednisone, remained clinically active with undetectable peripheral B cells. Bimekizumab was initiated in July 2024. Anti-Dsg1 titers decreased from 252 IU/ml to undetectable levels by August 2024. Clinical remission allowed cessation of prednisone and tapering of mycophenolate.

Patient 2: A 43-year-old man with PF since 2010, resistant to azathioprine, mycophenolate, dapsone, and rituximab (January 2024), began bimekizumab in July 2024. He experienced early clinical improvement after induction and received 4 doses.

No adverse events were reported in either case.

Conclusion:

These two cases highlight the potential role of IL-17 blockade in PF, particularly in rituximab-refractory patients. The rapid clinical response and decline in anti-Dsg1 antibody titers suggest that bimekizumab may offer a novel therapeutic option in selected cases. Further studies are warranted to explore its efficacy and safety in autoimmune blistering diseases.

Abstract N°: 4255

Atypical Cutaneous GVHD: When Keratosis Pilaris Is More Than It Seems

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Introduction & Objectives: Graft-versus-host disease (GVHD) is a severe complication of haematopoietic stem cell transplantation, resulting from a deleterious immune response in which donor T lymphocytes attack host tissues. The skin is often the first and most visibly affected organ, with a wide spectrum of clinical presentations. While acute GVHD (aGVHD) and chronic GVHD (cGVHD) have well-described typical forms, atypical cutaneous variants may delay diagnosis. We report an unusual case of cGVHD initially mimicking keratosis pilaris, highlighting the importance of recognizing rare clinical patterns to ensure early management.

Materials & Methods:

A 37-year-old man with no prior medical history was diagnosed with bone marrow aplasia and received an allogeneic peripheral blood stem cell transplant from his HLA 10/10 matched brother. Eight months post-transplant, he developed a pruritic skin rash and was referred to dermatology. Examination revealed diffuse keratosis pilaris-like lesions with generalized xerosis. There was no history of atopy. A skin biopsy showed follicular plugs, mild epidermal hyperkeratosis, and a subtle perifollicular lymphocytic infiltrate, consistent with keratosis pilaris. He was treated with a urea-based emollient.

Two months later, new violaceous, scaly patches appeared on the trunk, back, and arms. A second biopsy revealed mild epidermal acanthosis, orthokeratosis, and a lymphocytic infiltrate with minimal exocytosis, confirming lichenoid cGVHD. Treatment with topical corticosteroids, tacrolimus 0.1%, and oral prednisolone led to stabilization within three months.

Results:

Cutaneous GVHD typically manifests as a morbilliform rash in its acute form and as lichenoid or sclerotic lesions in chronic stages. However, atypical forms are increasingly recognized. In this case, the patient initially presented with a keratosis pilaris-like eruption—a rare manifestation of cGVHD. Keratosis pilaris is usually characterized by follicular papules with surrounding erythema or pigmentation. It has been proposed as a possible early sign of sclerotic cGVHD. Our patient's lesions progressed into a lichenoid variant, representing, to our knowledge, the first reported case of such evolution.

Other rare presentations include follicular hyperkeratosis. Kim et al. reported a similar keratosis pilaris-like biopsy-proven cGVHD. The 2014 NIH consensus defines such lesions—including ichthyosis, hypopigmentation, and hyperpigmentation—as "unclassified" features of cGVHD. While they cannot establish the diagnosis alone, they are part of the broader clinical picture once cGVHD is confirmed.

Recognizing these atypical signs is crucial for early diagnosis and avoiding progression to more severe or irreversible forms.

Conclusion: This case highlights a rare form of cutaneous cGVHD initially presenting as keratosis pilaris, later evolving into a lichenoid pattern. It emphasizes the need to consider atypical variants of cGVHD in post-transplant dermatological assessments to ensure timely and appropriate intervention.

Abstract N°: 4278

Esophageal pemphigus: a rare manifestation in pemphigus vulgaris – A case report.

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Introduction & Objectives:

Pemphigus vulgaris (PV) is an autoimmune intraepidermal blistering disorder of the skin and mucous membranes. The pathogenesis involves IgG1 and IgG4 autoantibodies targeting intercellular adhesion molecules, Desmoglein 1 and 3 located in Desmosomes of keratinocytes. This results in disrupted keratinocyte adhesion, leading to acantholysis and the formation of intraepidermal blisters. Esophageal pemphigus is a rare occurrence in a setting of PV and is often underdiagnosed. Here we report a case of a 46-year-old male with PV presenting with esophageal involvement.

Objective: To describe the clinical and histological characteristics of esophageal pemphigus.

Materials & Methods:

A 46-year-old male with a history of PV presented with odynophagia and dysphagia for 2 months. There were superficial crusted erosions on the scalp and trunk. Oral ulcers, lip erosions and white non adherent plaques on the tongue were noted. He was treated with Itraconazole and Nystatin with the clinical suspicion of esophageal candidiasis due to long-term immunosuppression, however, the patient's symptoms persisted despite treatment.

Results:

The full blood count revealed a hemoglobin level of 8.2 g/dL. Further evaluation indicated hypochromic microcytic anemia, and stool occult blood testing was positive. Esophagogastroduodenoscopy (EGD) revealed esophageal erosions and ulcerations. Biopsies were taken from the esophageal ulcers. The histology was in keeping with esophageal pemphigus with features of acantholysis and tomb stone appearance. Fungal stains, cytomegalovirus PCR, herpes simplex PCR and HIV serology were negative. Patient was commenced on Rituximab and both cutaneous and gastrointestinal symptoms resolved within one month.

Discussion:

The true prevalence of esophageal involvement is unknown. A study performed by Calka et al in 26 patients with PV revealed esophageal involvement in 46.15% of the cases. Patients with esophageal involvement of PV commonly present with dysphagia , odynophagia and rarely with hematemesis, though majority of individuals are asymptomatic. In the background of chronic immunosuppression infections such as esophageal candidiasis, herpetic esophagitis and cytomegalovirus esophagitis should be considered. Stool occult blood testing and hemoglobin levels can be used as surveillance tools in patients with pemphigus who present with gastrointestinal symptoms, prior to proceeding with endoscopy. Characteristic features of esophageal pemphigus vulgaris in EGD are diffuse exfoliation of the mucosal surface, erosions and ulcerations with or without mucosal erythema. Cytology, histology, direct and indirect immunofluorescence of mucosal biopsies can confirm the diagnosis. Corticosteroids, rituximab, and intravenous immunoglobulin (IVIG) are therapeutic options for the management of esophageal pemphigus.

Conclusion:

Esophageal involvement in pemphigus is often overlooked, and a high index of clinical suspicion is essential for the early identification of lesions. Endoscopic evaluation should be considered in patients with pemphigus vulgaris presenting with gastrointestinal symptoms such as odynophagia and dysphagia. Timely detection of esophageal pemphigus is crucial in preventing serious outcomes like upper gastrointestinal bleeding.

Abstract N°: 4298

Acquired epidermolysis bullosa: From diagnostic challenge to unexpected and cost-effective therapeutic success with colchicine

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Introduction & Objectives: Acquired epidermolysis bullosa (EBA) is a rare, chronic autoimmune subepidermal blistering disease caused by autoantibodies against type VII collagen. It can manifest in two major clinical forms: the mechanobullous (classical) variant, characterized by trauma-induced blisters and scarring, and the inflammatory variant, which may resemble other autoimmune blistering or inflammatory dermatoses. The clinical heterogeneity often makes diagnosis particularly challenging. EBA is typically recalcitrant to treatment, and while first-line therapies include systemic corticosteroids and dapsone, more severe or refractory cases may require biologics such as rituximab, infliximab, or intravenous immunoglobulin (IVIG). However, therapeutic response remains variable.

Materials & Methods: Clinical data and histopathological findings were retrospectively reviewed from the patient's medical records. Relevant images and complementary tests were included.

Results: We present the case of a 68-year-old man with no relevant medical history, who initially developed asymptomatic erythematous-squamous plaques on photo-exposed areas. Over the following weeks, the patient developed fragile bullous lesions predominantly on distal areas of the extremities. A biopsy showed subepidermal blistering with linear IgG and C3 deposits at the basement membrane zone on direct immunofluorescence, leading to a presumptive diagnosis of bullous pemphigoid. Despite systemic oral corticosteroids and high-dose intravenous methylprednisolone, there was minimal clinical improvement. Due to the trauma-prone distribution of the blisters and poor therapeutic response, EBA was suspected. A NaCl separation immunofluorescence revealed IgG and C3 deposits on the dermal side of the split, confirming the diagnosis of EBA. Given the patient's chronic hepatitis B (anti-HBc+) and latent tuberculosis, biological therapies were contraindicated. Colchicine (0.5 mg BID) was initiated with rapid and sustained clinical improvement. A mild relapse was effectively controlled by increasing the dose to 0.5 mg TID. The treatment was well tolerated, and the patient remains well controlled after 12 months.

Conclusion: This case illustrates the diagnostic complexity of EBA, particularly its inflammatory form, and highlights the importance of repeated biopsies and immunofluorescence studies in unclear presentations. Although many cases of EBA are refractory and may require expensive biologic agents, colchicine — a safe, low-cost anti-inflammatory — proved surprisingly effective in this patient. Colchicine should be considered a viable therapeutic alternative in EBA, especially in patients with contraindications to immunosuppressants or biologics.

Abstract N°: 4426

Anifrolumab for adolescent systemic lupus erythematosus with cutaneous involvement: description of two cases

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Introduction & Objectives: The wide range of clinical manifestations in systemic lupus erythematosus (SLE) is due to the heterogeneity of the disease in which different etiopathogenic factors and immunological abnormalities intervene. Adolescents with cutaneous lupus often present with a higher number of SLE criteria at diagnosis compared to younger children, indicating more severe disease activity.

Materials & Methods: We present two cases of SLE with predominant cutaneous affection in two female teenagers of 14 and 17 years, in follow-up in dermatology and rheumatology department for SLE with articular and cutaneous involvement so as serologic activity with different lines of treatment required. As for cutaneous lesions, subcutaneous lupus lesions on the face and extremities were present with no control with methotrexate, prednisone, and hydroxychloroquine. For the first case, belimumab was also used but due to irregular compliance was interrupted. Anifrolumab was initiated in both cases, with a rapid response in the second case after only two infusions and maintained afterwards, and to be evaluated in the first case, as it has only been used for two months.

Results: Anifrolumab is a fully human monoclonal antibody that targets the type I interferon receptor subunit 1 (IFNAR1), and it has been approved for the treatment of moderate to severe SLE in adults. Its use in teenagers is less well-documented but emerging evidence suggests potential benefits. A case report described the use of anifrolumab in two pediatric patients with refractory cutaneous lupus lesions associated with SLE and *Shawn et al* describe seven adolescent patients with discoid lesions treated with anifrolumab. These patients showed significant clinical improvement after receiving anifrolumab therapy, indicating its potential efficacy in this population**.**

Conclusion: Rapid and effective treatment is critical to minimize duration of lesions that may affect selfesteem, particularly among adolescent patients, so anifrolumab is an option to be considered in these cases.

Abstract N°: 4534

“Natural Nutrients in Dermatology: Evidence and Applications”

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Introduction & Objectives:

There is growing scientific and clinical interest in the role of natural nutrition in dermatology. Dietary factors, especially naturally derived nutrients, can significantly influence the development, severity, and progression of various skin conditions. Chronic inflammatory dermatoses, premature skin aging, and barrier dysfunction are increasingly linked to lifestyle and nutritional patterns. This abstract aims to highlight current evidence on the dermatological impact of natural nutrients, including their mechanisms of action and therapeutic potential in managing common skin disorders.

Materials & Methods:

A narrative literature review was conducted using studies published in the last 10 years from PubMed, Scopus, and Google Scholar. The focus was on randomized controlled trials, clinical reviews, and meta-analyses involving key natural nutrients such as omega-3 fatty acids, vitamins C and E, polyphenols, probiotics, and plant-based antioxidants. Skin conditions explored include acne, atopic dermatitis, psoriasis, and age-related changes. The main outcomes reviewed were improvements in inflammation, skin barrier function, oxidative stress, and microbial balance.

Results:

Several natural nutritional compounds have demonstrated positive dermatologic effects. Omega-3 fatty acids were found to reduce inflammation in conditions like psoriasis and eczema. Antioxidant vitamins, particularly C and E, enhanced skin healing and photoprotection by neutralizing free radicals and supporting collagen formation. Polyphenols from sources such as green tea and berries showed anti-inflammatory and antimicrobial properties, improving acne symptoms and skin tone. Probiotics contributed to skin barrier restoration and immune regulation through modulation of the gut-skin axis. Overall, the integration of specific dietary elements was associated with measurable benefits in both inflammatory and degenerative skin

Conclusion:

Natural nutrition offers a safe and effective adjunct to conventional dermatologic treatments. The inclusion of targeted dietary components can improve clinical outcomes, enhance skin resilience, and promote overall skin wellness. A personalized nutritional approach, grounded in scientific evidence, may represent an important evolution in holistic dermatologic care.

Abstract N°: 4542

Association of statins, gliptins, and antipsychotics with bullous pemphigoid: A case-control study in the Cretan population

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Introduction & Objectives: Bullous pemphigoid (BP) is an autoimmune blistering disorder predominantly affecting the elderly. Recently, many studies have shed light on the effect of specific drug intake and comorbidities on the development of BP. The purpose of this study was to investigate the association of specific drug class intake and comorbidities with the development of BP in the Cretan population.

Materials & Methods: We conducted a retrospective 1:3 case–control study at the University General Hospital of Heraklion on the island of Crete. The study included 64 cases with BP diagnosis and 191 sex- and age (±2 years)-matched controls. Inclusion criteria for both cases and controls were Cretan descent (up to two generations) and admission to hospital during the period 2012–2020. The diagnosis of BP in cases was determined by clinical features, histopathological diagnosis, and at least one of the following immunological examinations: positive direct immunofluorescence or positive enzyme- linked immunosorbent assay (ELISA) serum test. Controls must have visited the hospital the same year the case was admitted to the hospital because of a BP diagnosis. Exclusion criteria for all participants were the presence of another autoimmune bullous disease and age under 40 years.

Results: Significant associations with BP were found for statins (odds ratio [OR] = 4.06, 95% confidence interval [CI] 1.99-8.27, P < 0.001), gliptins (OR = 4.27, 95% CI 2.33-7.83, P < 0.001), and antipsychotics (OR = 3.33, 95% CI 1.36-8.11, P = 0.006). Higher proportions of use in the BP group vs. control group were found for atorvastatin (OR = 1.86, 95% CI 1.04-3.32, P = 0.035), linagliptin (OR = 6.63, 95% CI 2.17-20.23, P < 0.001), vildagliptin (OR = 3.20, 95% CI 1.73-5.91, P < 0.001), alogliptin (OR = 5.11, 95% CI 1.19-22.04, P = 0.016), and quetiapine (OR = 4.21, 95% CI 1.5-11.85, P = 0.004). The presence of diabetes mellitus in the absence of gliptins did not show any significant effect on BP (OR = 1.60, 95% CI 0.79-3.23, P = 0.188). Metformin intake showed no significant association with BP (OR = 0.48, 95% CI 0.18-1.28, P = 0.143).

Redarding gliptins, similar odds ratios (ORs with 95% CI) were found during the first year of gliptin intake (OR = 9.76, 95% CI 6.55–19.95) and between the first and second year (OR = 9.52, 95% CI 6.00–16.22) of gliptin intake. Use of gliptins for a period of 2–4 years still shows an increased OR, but lower compared to those of the previous shorter time periods (OR = 4.26, 95% CI 2.73–7.57). The same was true for longer than 4 years of gliptin intake (OR = 2.93, 95% CI 1.69–3.99). Using Kaplan–Meier analysis, the median time for developing BP was 52.0 (95% CI 34.2–69.8) months or 4.3 (95% CI 2.9–5.8) years. Multivariate logistic regression model of BP with gliptin intake duration, drug categories, and patients' medical history revealed similar OR magnitudes for BP development for any duration of gliptin intake except for gliptin intake period longer than 4 years (OR = 1.76, P = 0.260). In BP patients there were fewer cases with CAD (14.1%) than in controls (30.0%), resulting in OR = 0.38 (95% CI 0.18–0.83, P = 0.012). Similarly, lower OR for functional thyroid disorder was found in BP patients (OR = 0.37, 95% CI 0.19–0.73, P = 0.003).

Conclusion: Our findings confirm and extend previous studies reporting the association of gliptins and antipsychotics on BP in other European populations. The association found for statins is new, thus more studies

are needed to corroborate its validity.

Abstract N°: 4570

Efficacy and safety of deucravacitinib in patients with discoid lupus erythematosus (DLE) and/or subacute cutaneous lupus erythematosus (SCLE): results from PAISLEY CLE, a global, phase 2 randomized, double-blind, placebo-controlled trial

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Introduction & Objectives:

Deucravacitinib, an oral, selective, allosteric TYK2 inhibitor approved for moderate to severe plaque psoriasis, has demonstrated significant improvements in cutaneous manifestations in the phase 2 PAISLEY SLE trial. We report data from the week 16 (W16) placebo-controlled period of the phase 2 PAISLEY CLE trial (NCT04857034) of deucravacitinib vs placebo in patients with discoid lupus erythematosus (DLE) and/or subacute cutaneous lupus erythematosus (SCLE).

Materials & Methods:

Adults were randomized 1:1:1 to placebo or deucravacitinib (3 or 6 mg twice daily [BID]). Patients with or without SLE were enrolled, with SLE participation limited to ≤50%. The primary endpoint was mean percent change from baseline (CFB) in Cutaneous Lupus Erythematosus Disease Area and Severity Index-Activity (CLASI-A) score at W16. Mean CFB in CLASI-A and ≥50% reduction from baseline CLASI-A score (CLASI-50) were secondary endpoints; CLASI-70 was a post hoc analysis. $P<0.1$ represents statistical significance. Safety was also assessed.

Results:

Deucravacitinib provided significantly improved mean percent CFB in CLASI-A vs placebo (placebo [n=24], -28.4%; deucravacitinib 3 mg BID [n=25], -47.5%, $P=0.0670$; 6 mg BID [n=25], -50.0%, $P=0.0385$), mean CFB in CLASI-A (5.3 vs -9.3 [$P=0.0425$] and -8.7 [$P=0.0805$]), and CLASI-50 response (19.0% vs 56.7% [$P=0.0092$] and 52.3% [$P=0.0193$]). Deucravacitinib 3 mg BID was associated with a higher CLASI-70 response vs placebo (15.9% vs 49.5% [$P=0.0184$] and 29.5% [$P=0.2713$]). No new safety signals were observed. Adverse event (AE) rates were 50.0% vs 68.0% and 79.2%; most AEs with deucravacitinib were mild to moderate and consistent with the known safety profile.

Conclusion:

In patients with DLE/SCLE, deucravacitinib showed statistically significant and clinically meaningful improvements

in CLASI-A outcomes and was well tolerated, supporting further evaluation in the ongoing phase 3 POETYK SLE trials (NCT05617677, NCT05620407).

Abstract N°: 4584

Efficacy and safety of deucravacitinib up to week 52 from POETYK PsA-2: a multicenter, randomized, double-blind, placebo-controlled, phase 3 study in patients with psoriatic arthritis

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Introduction & Objectives:

Deucravacitinib, a first-in-class, oral, selective, allosteric TYK2 inhibitor, was investigated for psoriatic arthritis (PsA) in the randomized, double-blind, phase 3 POETYK PsA-1 (NCT04908202) and POETYK PsA-2 (NCT04908189) trials. Deucravacitinib has >4 years of long-term data and is approved in multiple countries for moderate to severe plaque psoriasis (PsO). We investigated deucravacitinib efficacy and safety up to week 52 in POETYK PsA-2 adults with active PsA who were naive to biologic DMARDs, were TNF inhibitor (TNFi) naive, or had experienced failure of or were intolerant to ≤2 prior TNFis for PsA or PsO.

Materials & Methods:

Patients met CASPAR criteria and had PsA diagnosis ≥3 months, active/documented history of plaque PsO, active arthritis (≥3 swollen/≥3 tender joints), and hsCRP level ≥3 mg/L at screening. Primary endpoint was ACR 20 at week 16. Key secondary endpoints included musculoskeletal system, skin, and PsA disease activity measures and quality of life (QoL) vs placebo at week 16. Patients were randomized 3:3:1 to deucravacitinib 6 mg QD, placebo, or apremilast 30 mg BID through week 16. From weeks 16-52, those receiving deucravacitinib or apremilast continued treatment and those receiving placebo were switched to deucravacitinib. Multiplicity-controlled hierarchical testing was used to assess all key endpoints (week 16). Nonresponder imputation was used for

missing binary data and control-based pattern imputation for missing continuous data. Safety was evaluated up to week 52.

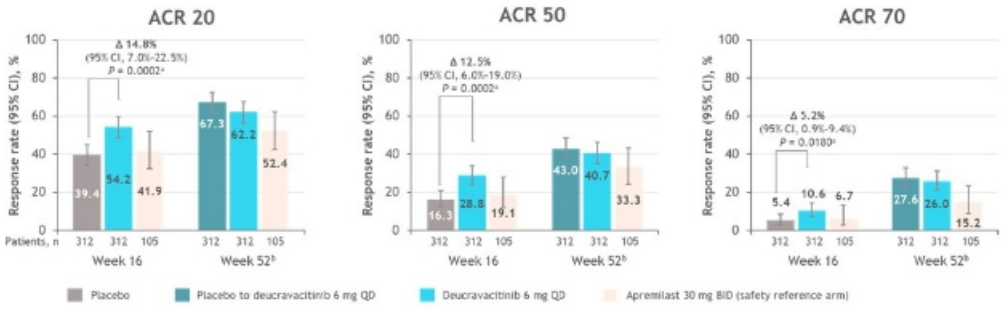
Results:

In all, 729 patients were randomized (deucravacitinib, n=312; placebo, n=312; apremilast, n=105). Baseline patient demographics/disease characteristics were balanced across groups. At week 16, significantly more patients receiving deucravacitinib vs placebo achieved ACR 20 (54.2% vs 39.4%; *P*=0.0002), with similar trends for ACR 50 (28.8% vs 16.3%) and ACR 70 (10.6% vs 5.4%) (**Figure**). Response rates were maintained through week 52. Apremilast efficacy data were reported (**Figure**). Achievement of key secondary endpoints was significantly greater with deucravacitinib vs placebo (PASI 75, 40.9% vs 15.4%, *P*<0.0001; minimal disease activity, 25.6% vs 14.7%; *P*=0.0007). Deucravacitinib-treated patients had greater mean change from baseline in HAQ-DI (−0.3246 vs −0.2120; *P*=0.0013) and SF-36 PCS (5.838 vs 3.796; *P*=0.0002). POETYK PsA-1/PsA-2 prespecified pooled analyses showed enthesitis resolution rates of 50.3% (deucravacitinib) vs 45.1% (placebo) (*P*=0.1781); higher dactylitis resolution rates were 57.6% vs 44.1% (nominal *P*=0.0100). Numerical improvement in FACIT-Fatigue score was seen with deucravacitinib vs placebo (2.5 vs 1.8; *P*=0.2017); mean change from baseline in DAS28-CRP was −1.2781 vs −0.8038 (nominal *P*<0.0001). Few serious AEs (placebo, 1.0%; deucravacitinib, 1.9%; apremilast, 3.8%) and discontinuations due to AEs (1.3%; 2.2%; 10.5%) occurred through week 16. Deucravacitinib was well tolerated through week 52 (**Table**). Similar rates of MACE, VTE events, opportunistic infections, and malignancies were observed with each treatment arm throughout the trial.

Conclusion:

Deucravacitinib showed superior efficacy vs placebo in PsA across multiple endpoints at week 16. Clinical responses were maintained through week 52. Safety was consistent with the established deucravacitinib safety profile; no new safety signals were identified. Deucravacitinib is efficacious and well tolerated in PsA.

Figure. ACR 20/50/70 at weeks 16 and 52



No prespecified statistical comparisons were performed for the apremilast arm.

All randomized patients were assessed. Nonresponder imputation was used to handle missing data. Treatment discontinuations prior to week 16 were considered treatment failures (composite variable strategy). All rescue medication–related intercurrent events were treated with a treatment policy–estimand strategy. The Clopper-Pearson estimation method was used to estimate CI. [a]A Cochran-Mantel-Haenszel test stratified by TNF inhibitor (yes/no), screening hsCRP concentration (< 10 mg/L vs ≥ 10 mg/L), and csDMARD use at baseline (yes/no) was used to compare the response rates with deucravacitinib 6 mg QD to placebo; [b]Exploratory endpoint.
ACR 20, American College of Rheumatology 20% improvement in response; ACR 50, American College of Rheumatology 50% improvement in response; ACR 70, American College of Rheumatology 70% improvement in response; BID, twice daily; csDMARD, conventional synthetic disease-modifying antirheumatic drug; hsCRP, high-sensitivity C-reactive protein; QD, once daily; TNF, tumor necrosis factor.

Table. Most frequent AEs (≥5% in any arm) by preferred term: cumulative treatment period weeks 0-52[a]

Most frequent AEs (≥5% in any arm) by preferred term	Total deucravacitinib 6 mg QD[b] (n=604)		Apremilast 30 mg BID (n=105)	
	n (%)	IR/100 PYs	n (%)	IR/100 PYs
Total patients with an event	451 (74.7)	220.8	92 (87.6)	366.5
Nasopharyngitis	71 (11.8)	16.0	8 (7.6)	9.9
COVID-19	70 (11.6)	15.8	13 (12.4)	16.9
Upper respiratory tract infection	66 (10.9)	15.0	11 (10.5)	13.6
Diarrhea	27 (4.5)	5.8	20 (19.0)	26.5
Hypertension	23 (3.8)	5.0	7 (6.7)	8.8
Psoriatic arthropathy	23 (3.8)	4.9	7 (6.7)	8.4
Nausea	20 (3.3)	4.3	9 (8.6)	11.0
Headache	20 (3.3)	4.3	13 (12.4)	16.7
Urinary tract infection	16 (2.6)	3.4	7 (6.7)	8.4

PY is defined as exposure based on time to first onset. Incidence rate per 100 PYs of exposure (IR/100 PYs) is calculated as event count×100/PYs of exposure. MedDRA v27.1 was used to define preferred terms for AEs.

[a]AEs in the treated population are shown. AEs are treatment-emergent AEs with an onset on or after the first dose date of study treatment up to 30 days after the last treatment dose; [b]The total deucravacitinib group includes patients who received deucravacitinib from baseline through week 52 as well as patients who switched from placebo to deucravacitinib at week 16 and continued through week 52.

AE, adverse event; BID, twice daily; IR, incidence rate; MedDRA, Medical Dictionary for Regulatory Activities; PY, person-years; QD, once daily.

Abstract N°: 4780

IMACS Guidelines: An External Validation in Dermatomyositis Patients in Dermatology and Rheumatology Clinics in a Metropolitan Academic Center

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Introduction & Objectives:

Dermatomyositis (DM) is an idiopathic inflammatory myopathy (IIM) characterized by proximal muscle weakness, distinct skin findings, and increased cancer risk. Cancer risk is greatest in the 3 years before or after DM onset, in which case it is called paraneoplastic DM. Because of this increased cancer risk, appropriate post-diagnostic guidelines should be followed to screen for malignancy. The International Guideline for Idiopathic Inflammatory Myopathy-Associated Cancer Screening (IMACS) set forth evidence-based guidelines for the risk-stratification and cancer screenings for newly diagnosed IIM patients. This study seeks to provide an external validation to the guidelines set forth by the IMACS in a metropolitan academic center.

Materials & Methods:

Patient charts were reviewed retrospectively for patients with a known DM diagnosis between Jan 2019 and December 2024. Patients were included if they had a rheumatology or dermatology visit for DM and if they had ≥2 follow-up visits. Demographics, myositis autoantibody panels, physical exam findings, clinical history, laboratory results, and cancer screening/diagnosis data were collected. Patients were stratified based on the risk factors determined by the IMACS.

Results:

We analyzed 413 patients with a diagnosis of DM. 12.6% (n=52) of patients had a cancer diagnosis at any time in their life. Patients that developed cancer within 3 years before or after their DM diagnosis were categorized as paraneoplastic. 6.5% (n=27) of DM patients had a diagnosis paraneoplastic DM. Patients were stratified as paraneoplastic and non-paraneoplastic. Most patients were female, comprising 88.9% of paraneoplastic patients and 77% of non-paraneoplastic patients. The median age at disease onset for paraneoplastic patients was 60 ± 13.3, and for non-paraneoplastic patients was 48 ± 17.2. Patients were stratified by the risk factors outlined in the IMACS guidelines into high-, intermediate-, and low-risk factors. 70.4% of paraneoplastic DM patients were categorized as high risk, compared to 51% of non-paraneoplastic patients. In our sample, age ≥40 at IIM diagnosis was statistically significant for an increased risk of paraneoplastic DM (p=0.001) [Table 1].

Of the 27 paraneoplastic DM cases, the most common diagnosis was breast cancer (n=10), followed by lung cancer (n=5), uterine cancer (n=2), Non-Hodgkin lymphoma (n=2), cervical cancer (n=1), and other (n=7) [Table 2]. 33% (n=9) of paraneoplastic DM patients were diagnosed with cancer within 3 years prior to their DM diagnosis. 80% of paraneoplastic lung cancer patients were diagnosed prior to their DM diagnosis, via chest x-rays and confirmed with CT. All 10 breast cancer patients were detected using routine age-appropriate mammography screenings. Both uterine cancer patients were detected via abdominal CT.

Conclusion:

Our results provide a validation for the guidelines set forth by the IMACS, with most paraneoplastic DM patients

being found using the IMACS screening guidelines. Nearly one-third of paraneoplastic DM patients presented with a cancer diagnosis within three years prior to their DM diagnosis, and two-thirds were diagnosed following their DM diagnosis. These findings support the IMACS screening guidelines for the identification of paraneoplastic DM, leading to more targeted and risk-based screening.

Table 1. Risk Factors for Paraneoplastic and Non-Paraneoplastic Dermatomyositis

	Non-Paraneoplastic (n=386) N(%)	Paraneoplastic (n=27) N(%)	P-Value	Odds Ratio	95% CI
High Risk Factors					
Dermatomyositis	191 (49)	18 (67)	0.110	2.04	[0.83-5.29]
Anti-TIF1γ antibodies	16 (4)	3 (11)	0.119	2.88	[0.50-11.1]
Anti-NXP2 antibodies	18 (5)	0 (0)	0.620	0	[0-3.31]
Age ≥40 at IIM onset	246 (64)	25 (93)	0.001*	7.09	[1.72-62.7]
Persistent high disease activity despite therapy	9 (2)	0 (0)	1	0	[0-7.49]
Dysphagia (moderate to severe)	89 (23)	10 (37)	0.106	1.96	[0.77-4.72]
Cutaneous necrosis	24 (6)	3 (11)	0.406	1.88	[0.34-6.90]
≥ 2 'High risk' factors	197 (51)	19 (70)	0.071	2.27	[0.92-6.16]
Intermediate Risk Factors					
Amyopathic Dermatomyositis, Polymyositis, IMNM	126 (33)	9 (33)	1	1.03	[0.40-2.50]
Anti-SAE1 antibodies	10 (3)	0 (0)	1	0	[0-6.59]
Anti- HMGCR antibodies	2 (1)	0 (0)	1	0	[0-77.1]
Anti-Mi2 antibodies	21 (5)	0 (0)	0.383	0	[0-2.78]
Anti-MDA5 antibodies	24 (6)	0 (0)	0.389	0	[0-2.39]
Male Sex	89 (23)	3 (11)	0.229	0.418	[0.08-1.42]
≥ 2 'Intermediate risk' factors	144 (37)	9 (33)	0.837	0.841	[0.32-2.03]
Low Risk Factors					
Antisynthase syndrome	40 (10)	0 (0)	0.0938	0	[0-1.33]
Anti-SRP antibodies	2 (1)	0 (0)	1	0	[0-77.1]
Anti-Jo1 antibodies	40 (10)	1 (4)	0.500	0.333	[0.008-2.15]
Non-Jo1 ASSD antibodies	22 (6)	1 (4)	1	0.637	[0.014-4.28]
MAA (PM-Scl, Ku, RNP, SSA/Ro, or SSB/La)	116 (30)	7 (26)	0.828	0.815	[0.28-2.08]

Table 2. Paraneoplastic DM Cancer Diagnosis

Cancer Type	Number of patients N(%)	Imaging Modality for Diagnosis	Clinical Cancer Symptoms	Mean Time between DM Diagnosis and Cancer Diagnosis
Breast	10 (37)	Mammogram	None	23.3 months
Lung	5 (19)	Chest CT, Chest X-ray, PET scan	Cough, dysphagia	18.4 months
Uterus/Endometrial	2 (7)	Abdominal CT	None	20.0 months
Cervix	1 (4)	N/A	Bleeding after starting blood thinner prescription	0.2 months
Non-Hodgkin Lymphoma	2 (7)	PET scan	Lymphadenopathy in R axilla (1) and L neck (1)	3.2 months
Other (Tonsillar squamous cell carcinoma, chondrosarcoma, multiple myeloma, testicular cancer, thymoma, thyroid, colon)	7 (26)	MRI, Ultrasound, Abdominal CT	Weight loss (2)	22.6 months

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Abstract N°: 4788

Anti-p200 pemphigoid diagnosed by biochip array technology with BP180 ELISA positivity: A Case Report

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Introduction & Objectives:

Anti-p200 pemphigoid (anti-laminin-γ1 pemphigoid) is a rare subepidermal bullous disease characterized by antibodies targeting a 200 kDa protein known as laminin gamma-1, localized in the lamina lucida of the basement membrane. Its clinical presentation is similar to that of bullous pemphigoid, but with certain distinct epidemiological, clinical and immunological features that we will highlight in this case.

Results:

A 98-year-old woman with no medical history presented with a two-week history of pruritus and the development of bullous lesions. Examination revealed tense blisters extending over 40% of the body surface area, involving the head, neck, upper and lower extremities, with marked acral involvement. Dyschromic and atrophic scars were observed. There was no involvement of mucous membranes or appendages. Laboratory findings showed no evidence of hypereosinophilia. Anatomopathological examination showed subepidermal cleavage associated with a dermal inflammatory infiltrate composed of eosinophilic polymorphonuclear cells and lymphocytes. Direct immunofluorescence showed IgG and C3 deposition. Indirect immunofluorescence on salt-split human skin showed double staining on the dermal and epidermal sides. Anti-BP180 was positive on one cross in the ELISA test. Bioship analysis showed labelling of the P200 protein. The diagnosis of anti-P200 pemphigoid was made.

Discussion & Conclusion:

Several patients diagnosed with anti-P200 pemphigoid were initially misdiagnosed with bullous pemphigoid due to the significant overlap between the two entities. However, herein are some features that differentiate anti-P200 pemphigoid from bullous pemphigoid according to the literature: History of psoriasis or use of gliptins, younger age at onset. The medical history was not consistent with our patient’s presentation. However, she had several clinical criteria that supported the diagnosis of anti-P200 pemphigoid: involvement of the head and neck and both upper and lower limbs, with an acral predilection. No mucosal involvement was observed in our patient. Although mucosal involvement is reported in 20% of cases in the literature, when present, it tends to be less extensive and less severe than in cicatricial pemphigoid. Indirect immunofluorescence, which showed staining of the blister floor, was a key diagnostic clue suggesting anti-P200 pemphigoid. Although the ELISA was positive for anti-BP180 antibodies, their presence remains possible in anti-P200 pemphigoid and does not exclude the diagnosis. The presence of BP180 antibodies is most likely due to an epitope spreading phenomenon. Finally, detecting a 200 kDa band on the biochip confirmed the diagnosis.

In cases with atypical presentations of bullous pemphigoid, whether anamnestically, clinically or immunologically, anti-P200 pemphigoid should be considered a potential diagnosis.

Abstract N°: 4790

Autoimmune blistering diseases at the Clinic of Dermatology and Venereology, University Clinical Center of Serbia

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Introduction & Objectives:

Autoimmune bullous dermatoses (AIBD) represent rare organ-specific diseases that affect the skin and mucous membranes. They include two main groups: intraepidermal and subepidermal blistering diseases. The clinical picture is characterized by the appearance of blisters or erosions on the skin and/or mucous membranes. The diagnosis of AIBD is based on clinical features and histopathological findings, confirmed by immunofluorescent tests (direct and indirect) and enzyme-linked immunosorbent assays. The frequency of AIBD differs significantly depending on the geographic region and population evaluated. The average age of patients is 50- 80 years, depending of the AIBD subtype.

The objective of this study was to examine differences in the frequency of AIBD subtypes in relation to sex and age and the trend of changes of hospitalization frequency during the four years of this study.

Materials & Methods:

A retrospective study was conducted at the Clinic of Dermatology and Venerology of the University Clinical Centre of Serbia for the period 2020-2024. Patients' data were collected from medical histories and the electronic database. The type of AIBD was analyzed. Demographic data (age and sex) were used.

Results:

From 2020-2024, 230 patients with various AIBD were hospitalized (111 men, 48.3% and 119 women, 51.7%). The average age of patients was 64±18.4 years, lower in pemphigus group (54.11±16.02 years). In pemphigus group, pemphigus vulgaris was the most common, in 88 (38.3%) patients (52.3% women and 47.7% men). In the group of subepidermal AIBD, bullous pemphigoid was the most common, in 96 (41.7%) patients (52.1% women and 47.9% men). The rarest form of AIBD observed were IgA pemphigus, paraneoplastic pemphigus, pemphigus herpetiformis (one case each) and pemphigoid gestationis (in two pregnant women). The largest number of newly diagnosed hospitalized patients with pemphigus vulgaris was in 2023, 24/88 (27.3%) and in 2021 for newly diagnosed pemphigoid patients (24/96, 25%). Probably due to Coronavirus disease 2019 (COVID-19) pandemic, there was a decrease in a number of hospitalized patients during 2020-2022, the largest number of all hospitalized patients with AIBD was in 2023 (64% of all pemphigus cases and 31.3% of all pemphigoid cases).

Conclusion:

We confirmed that the most common AIBD were pemphigus vulgaris and bullous pemphigoid. Although female predominance of AIBD is suggested in various studies, we observed an equal distribution between sexes. The average age of the onset of the disease in different AIBD subtypes in our study was similar to literature data. An increased tendency of frequency of AIBD during the study period was observed. This increase (especially of newly diagnosed pemphigus patients) could be explained by a possible influence of various environmental factors

(COVID-19 infection and/or vaccines, multiple drug administration during COVID-19 pandemic and emotional stress).

Abstract N°: 4944

Atypical Presentation of Systemic Lupus Erythematosus in Children: A Case Report

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Introduction & Objectives:

Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disease that can affect multiple organs and has a relatively high mortality rate in children. The clinical manifestations of childhood-onset SLE are often atypical and diverse, making it difficult to diagnose due to its resemblance to other diseases. It is essential to recognize the specific clinical features of SLE in children to improve early detection and diagnosis.

Materials & Methods:

A 14-year-old female presented to the Outpatient Clinic of the Dermatology and Venereology Department at Surabaya Islamic Hospital–Ahmad Yani with complaints of reddish spots, scaling with black cores, and wounds on the scalp, cheeks, and nose that had persisted for one month. Dermatological examination of the face and scalp revealed erythematous plaques with multiple well-defined scales, partially coalescing plaques, crusts, and erosions. Additionally, petechiae and purpura were observed on the palms and soles. Laboratory investigations showed anemia and pancytopenia. Immunological screening was positive for antinuclear antibodies (ANA) and anti–double-stranded DNA (anti-dsDNA). Complement levels (C3 and C4) were markedly decreased.

Results:

A diagnosis of SLE was established and being treated in collaboration with Pediatric Department. The patient was given blood transfusions, systemic corticosteroids, topical corticosteroid twice a day along with topical antibiotic during hospitalization.

Conclusion:

Recognizing the varied manifestations of SLE in children is critical. Early diagnosis allows for prompt and appropriate treatment, which can significantly reduce morbidity and mortality.

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Abstract N°: 5128

From Diclofenac to lichen planus pemphigoid: Exploring drug-induced pathogenesis

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Introduction & Objectives:

Lichen planus pemphigoid (LPP) is a rare condition, typically idiopathic, that combines the clinical, histological, and immunological features of lichen planus and bullous pemphigoid. Through this case report, we highlight the clinical features that differentiate LPP from other autoimmune blistering disorders and classical lichen planus, and we discuss the possible pathogenic mechanisms underlying drug-induced LPP.

Materials & Methods:

We report the case of a patient who developed lichen planus pemphigoid one month after initiating oral diclofenac therapy. Diagnosis was based on clinical examination, histopathological evaluation of lesional skin, and direct immunofluorescence of perilesional tissue. A causality assessment was performed to evaluate the likelihood of Diclofenac as the inducing agent.

Results:

A 63-year-old woman with diabetes presented with a generalized pruritic eruption that appeared one month after taking Diclofenac for lumbago. Dermatological examination revealed multiple purple, scaly papules and plaques on the skin, with Wickham's striae on dermoscopy. Tense blisters with hemorrhagic content and post-bullous erosions were present on the lichenoid plaques as well as at distance from them. Nikolsky sign was negative. A pigmented slate-gray patch was noted on the forehead, showing a granular pattern on dermoscopy. Dermatological assessment revealed diffuse onychorrhexis and a scaly scalp without setback of the frontal hairline or mucosal involvement. Paraclinical tests were unremarkable. Biopsy of a blister and a purple plaque showed features of bullous pemphigoid and lichen planus, respectively. Pharmacovigilance investigation yielded an imputability score of I3B4. The diagnosis of drug-induced lichen planus pemphigoid was established, and the patient was treated with topical corticosteroids, leading to significant improvement.

Conclusion:

Lichen planus pemphigoid is a rare entity that may represent a distinct disease rather than an association of lichen planus and bullous pemphigoid. It is characterized by the appearance of subepidermal blisters on normal skin and on lichen planus lesions as well. In most published cases, lichen lesions precede the development of blisters. The pathogenesis of this condition is not fully understood, but it is likely due to damage to the basement membrane caused by a lichenoid inflammatory process, leading to the release of antigens, subsequent development of autoantibodies, and blister formation. Although this entity is generally idiopathic, drug-induced cases have been reported, particularly with angiotensin-converting enzyme inhibitors, with a delay period ranging from 15 to 120 days. In contrast, cases of lichenoid reactions and bullous pemphigoid have been separately reported with Diclofenac use.

Abstract N°: 5265

When the airway speaks: Mucous membrane pemphigoid unmasked in a teenager post tracheostomy

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Introduction & Objectives:

Mucous membrane pemphigoid is a subepidermal autoimmune bullous dermatosis primarily directed against BP 180 and Laminin-332 basement membrane antigens. It is associated with a broad spectrum of clinical disorders, making diagnosis difficult for many clinicians, including dermatologists. Its severity is mainly due to ocular, laryngeal and esophageal involvement, which are often diagnosed late at the stage of life-threatening respiratory distress.

Materials & Methods:

We describe the case of a 17-year-old female who underwent emergency tracheostomy due to acute respiratory distress. Initial evaluation revealed mucosal inflammation and scarring involving the larynx and oropharynx. A detailed clinical examination was followed by endoscopic assessment of the upper airway. conjunctival mucosa biopsies were obtained and examined histopathologically and by direct immunofluorescence, confirming the diagnosis of mucous membrane pemphigoid. The patient was managed with systemic immunosuppressive therapy in coordination with a multidisciplinary team involving dermatology, ENT, pulmonology, and ophtalmology teams.

Results:

A 17-year-old girl with no previous history, was admitted to the emergency department with severe respiratory distress requiring emergency tracheostomy. She reported 2 years prior to her admission ocular lacrimation, a sensation of nasal obstruction, dysphonia with gingivorrhagia. She was put on several treatments without improvement. The evolution was marked by the appearance of blisters on the body, so her dermatologist put her on Dapsone, with good improvement of cutaneous and oral lesions, but worsening of ocular and respiratory symptoms. Dermatological assessment revealed bilateral symblepharon, atrophic scarring pigmented macules, with no vesiculobullous lesions, and no oral or genital involvement. A biopsy + direct immunofluorescence of the conjunctival mucosa was performed, confirming the diagnosis of Mucous membrane pemphigoid. The patient was put on prednisone, Dapsone and Rituximab with good improvement

Conclusion:

Mucous membrane pemphigoid is characterized by synechial lesions of the mucosa, with or without skin involvement, with linear deposits of immunoglobulins and complement in the basement membrane. The oral mucosa is the most frequently affected site, followed by the ocular, anogenital, then the laryngeal and esophageal mucosa. Therapeutic management is based on site, severity and speed of disease progression. Prevention of synechiae remains the most important aspect of treatment. In contrast to the overall epidemiology of mucosal pemphigoid, which is considered a disease of the elderly, most cases reported in the literature with tracheobronchial involvement involve young female patients. Therefore, it is appropriate to think that age and sex may play a role in predicting laryngeal involvement, imposing early and aggressive treatment with close follow-up in search of respiratory symptoms.

Abstract N°: 5287

Discovery and characterization of SIM0711, a potent and selective IRAK4 PROTAC with Enhanced Efficacy and Safety

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Introduction & Objectives:

IRAK4 plays a pivotal role in the innate immune response, acting downstream of TLRs and IL-1R It exhibits both kinase activity and scaffolding functions, making it a critical target for modulating inflammatory signaling pathways. Traditional IRAK4 kinase inhibitors have shown limited efficacy due to their inability to fully inhibit the scaffolding function of IRAK4. IRAK4 PROTAC offers a novel approach by selectively degrading the entire protein, thereby achieving broader pathway inhibition.

This study aimed to discover and characterize a potent and selective oral IRAK4 PROTAC, and evaluate its therapeutic potential for treating autoimmune and inflammatory diseases.

Materials & Methods:

We developed and extensively characterized SIM0711, an oral IRAK4 PROTAC, *in vitro* and *in vivo*. Its activity was compared with either the benchmark IRAK4 PROTAC (KT-474) or the benchmark IRAK4 inhibitor (PF-06650833). Key assays included:

\1. IRAK4 degradation in human PBMCs and fibroblast/keratinocyte cell lines.

\2. IRAK4 degradation kinetics in THP-1 cells or primary monocytes.

\3. Inhibition of pro-inflammatory cytokines (IL-6 and IL-8) in human PBMCs stimulated by LPS or IL-33.

\4. Efficacy in a mouse model of IL-33-induced skin inflammation.

\5. PK/PD, bioavailability, and safety profiles across multiple species.

Results:

In comparison to benchmark PROTAC KT-474, SIM0711 induced near-complete IRAK4 degradation in diverse primary human immune cells and stromal cell lines, demonstrating faster degradation kinetics in THP-1 cells and primary human monocytes *in vitro*. In the meantime, SIM0711 also led to deeper inhibition of the pro-inflammatory cytokines IL-6 and IL-8 following the stimulation of LPS, or IL-33 compared to either PF-06650833 or KT-474. *In vivo*, in a mouse model of IL-33-induced skin inflammation, SIM0711 dose-dependently reduced skin inflammation, nearly abolishing downstream IL-5 secretion at the inflammation site. Furthermore, SIM0711 demonstrated improved bioavailability, favorable PK/PD characteristics, and good safety profiles across multiple species.

Conclusion:

These findings establish SIM0711 as a best-in-class IRAK4 PROTAC with the potential to treat a wide range of

autoimmune and inflammatory diseases. Its robust IRAK4 degradation, superior anti-inflammatory efficacy, and favorable drug properties support its further development, with an IND submission anticipated in H2 2025.

Abstract N°: 5336

Characterization of Mucous Membrane Pemphigoid in Childhood: A Multicenter Study of 12 Cases

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Introduction & Objectives:

Mucous membrane pemphigoid (MMP) ,rarely described in children, is an autoimmune blistering disease (AIBD) of the dermal-epidermal junction with predominant mucosal involvement and a tendency to scare. The aim of our study was to characterize features of the disease and its management.

Materials & Methods:

A descriptive, retrospective, multicenter study was conducted, including children under 18, with a clinical diagnosis of MMP confirmed by histology, with or without immunological confirmation.

Results:

Twelve patients were included; 11 were girls. The median age of onset was 7 years and 8 months [4.5-14 years]. The time between diagnosis and histological or immunological confirmation was 4 months [0-12 months] and 6 months [0-31 months], respectively.

Mucosal involvement was predominantly genital (n=7/12): erosive vulvitis (n=5/7), clitoral blisters (n=1/7), balanitis (n=1/7). Other mucosal involvements included oral (erosive gingivitis n=5/12, ENT blisters n=1/12), nasal (crusting n=2/12), ophthalmic (cicatrizing conjunctivitis n=1/12) and pulmonary (bronchial stenosis n=1/12) changes. Three children had multiple mucosal involvements. Three had skin bullae. Five had scarring involving genital and pulmonary localization. (Table 1)

This clinical suspicion was supported by a skin biopsy for histopathology, which revealed a subepidermal blister with mixed inflammatory cell infiltrates in the dermis in all patients. The diagnosis was confirmed by positive direct immunofluorescence (10/12) and supported by ELISA (1/1) and immunoblotting (1/1). Two patients had negative DIF results. In one of these, the diagnosis was supported by indirect immunofluorescence; in other, immunological tests were all negative but clinicopathological correlation allowed us to retain the diagnosis of “most likely MMP”.

Regarding treatment, topical steroids (very potent) were used in 7 patients; they were effective as monotherapy in one patient with localized vulval involvement. Systemic treatment included dapsone (n=8), doxycycline (n=5), oral steroids (n=3), rituximab (n=2), intravenous immunoglobulin therapy (n=1), omalizumab (n=1), sulfasalazine (n=1) and erythromycin (n=1). After a median follow-up of 4 years the patients had received an average of 4 treatments; remission was partial in 7 cases and complete in 5.

Conclusion:

MMP is rare in childhood (27 cases reported in the literature). Diagnosis is difficult and often requires repeated skin biopsies. Serum immunological confirmation is rare. For genital involvement: the main clinical differential diagnosis is lichen sclerosus. We report the first case of pediatric bronchial involvement. Scarring is not always present, but early diagnosis is necessary to prevent it.

In mild/moderate cases, first-line treatment with dapsone or doxycyclin. Topical steroid treatment alone is rarely sufficient. Maintenance treatment is sometimes necessary because of relapses.

Our study provides a better characterization of this rare disease, often misdiagnosed, which can be serious in children.

Patient	Sex/ Age at onset (years and months)	Clinical features	DIF	IIF	ELISA	Immunoblot	Treatment	Outcomes
1	F/ 12y 8M	- **Clinical**: Erosive gingivitis of the incisors; neck, oral and vulvar blisters - **FS**: Pain - **Skin involvement**: tense blisters	-	Marking roof of the blister with salt-split skin	NEG	NEG	1. Oral corticosteroids (1 month) 2. Topical corticosteroids (ongoing)	1. Improvement 2. Improvement
2	M/8Y	- **Clinical**: Tense blisters of the prepuce, erosive balanitis, severe erosions of the glans (>50%) and the penis - **FS**: urinary burning or dysuria, pain, urinary retention - **Skin involvement**: ?	Linear C3, IgG	NEG	NEG	NEG	1. Topical corticosteroids + oral (1 month) corticosteroids 2. Dapsone (5 years) 3. Topicycline (1 year) + urethral dilation	1. Improvement 2. Improvement but recurring on treatment 3. Recovery
3	F/ 13Y 9M	- **Clinical**: Erosive gingivitis, gingival hypertrophy - **FS**: Pain, odynophagia, dysphagia - **Skin involvement**: 0	NEG	NEG	NEG	NEG	1. Doxycycline (5 year)	1. Improvement
4	F/ 6Y 11M	- **Clinical**: Erosive vulvitis, sclerosis, synechia, blisters - **FS**: dryness, tingling, pain, pruritus, urinary burning - **Skin involvement**: ?	Linear IgG	NEG	NEG	Not carried out	1. Dapsone (ongoing) 2. Topical corticosteroids (3 months) 3. Topical tacrolimus (7 months) 4. Doxycycline (ongoing)	1. Improvement 2. Ineffective 3. Stabilisation, 4. Improvement
5	F/ 13Y	- **Clinical**: Erosive vulvitis, synechia - **FS**: Urinary burning, pain - **Skin involvement**: ?	Linear C3	NEG	NEG	NEG	1. Topical corticosteroids (ongoing) 2. Dapsone (6 months)	1. Stabilisation 2. Stabilisation
6	F/ 7Y 4M	- **Clinical**: Erosive vulvitis - **FS**: Urinary burning, blood on wiping - **Skin involvement**: ?	Linear C3, IgG	NEG	NEG	Not carried out	1. Topical corticosteroids (4 months)	1. Recovery
7	F/ 12Y 9M	- **Clinical**: Fibrotic conjunctivitis, bronchial stenosis, nasal crust, epistaxis - **FS**: dryness, tingling eyes, pain, pruritus, dysphonia, dyspnoea - **Skin involvement**: tense blisters	Linear C3, IgG	NEG	NEG	200kDa	1. Cyclosporine eye drops 2. Autologous serum + doxycycline + dapsone (ongoing) + rituximab + ofatumumab + intravenous immunoglobulin	1. Poor tolerance 2. Stabilisation
8	F/ 4Y 8M	- **Clinical**: Erosive gingivitis, erosive vulvitis - **FS**: pain - **Skin involvement**: ?	Linear C3, IgG	NEG	NEG	Not carried out	1. Dapsone (ongoing)	1. Recovery
9	F/ 14Y	- **Clinical**: Erosive gingivitis - **FS**: Pain - **Skin involvement**: 0	Linear C3, IgG	NEG	BP 230-180	NEG	1. Doxycycline (6 months) 2. Dapsone (on going) + sulfasalazine	1. Stabilisation 2. Recovery
10	F/ 7Y	- **Clinical**: Erosive vulvitis, no synechia - **FS**: Pain, pruritus - **Skin involvement**: ?	Linear C3, IgG, IgA	NEG	NEG	NEG	1. Topical corticosteroids (ongoing) 2. Dapsone (9 months) 3. Erythromycin (18 months)	1. Stabilisation 2. Stabilisation 3. Improvement
11	F/ 5Y	- **Clinical**: Erosive gingivitis with oral detachment - **FS**: Odynophagia, dysphagia, pain - **Skin involvement**: ?	Linear C3, IgG	NEG	NEG	Not carried out	1. Dapsone + corticosteroid mouthwash cure (ongoing for dapsone, corticosteroid mouthwash stopped after 2 months)	1. Recovery
12	F/ 13Y	- **Clinical**: Pharyngeal blisters and post-blister erosions, nasal crusts - **FS**: Pain - **Skin involvement**: Blisters at the root of the scalp and at the nape	Linear C3, IgG, IgA	NEG	NEG	NEG	1. Colchicine + colchicoside (1 month) 2. Oral corticosteroids (1 month) 3. Rituximab (6 months)	1. Inefficacy 2. Improvement 3. Improvement

Table 1: Clinical, histological and immunological features and treatment of paediatric mucous membrane pemphigoid

Legend: FS: Functional sign. DIF: direct immunofluorescence. IIF: indirect immunofluorescence. NEG: negative. ELISA: enzyme-linked immunosorbent assay.

Abstract N°: 5468

Interstitial Granulomatous Dermatitis: a retrospective clinicopathological analysis of 47 cases

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Introduction & Objectives:

Interstitial granulomatous dermatitis (IGD) is a reactive condition to many diseases, with a clinical and pathological heterogenicity.

The aim of this study is to describe the epidemiological, clinical and histopathological features of patients with a diagnosis of IGD seen in our department from 2002 to 2025.

Materials & Methods:

We present a retrospective analysis of 47 cases, previously diagnosed as IGD and identified from the histopathological file of the Dermatopathology Unit in our Dermatology Department, between January 2002 and March 2025.

The clinical and epidemiological data was obtained from the patient's medical records, and the histopathological findings were revised.

Results:

Forty-seven patients with IGD were included (16 males, 31 females), aged between 16 and 89 years (mean 61.2 years).

The most frequent clinical features found were plaques (22 cases, 46.81%), followed by macules (21 cases, 44.68%) and papules (8 cases, 17.02%). In most cases the lesions were either erythematous (28 cases, 59.57%) or erythematous to violaceous (15 cases, 31.91%), and less frequently violaceous (3 cases, 6.38%). One patient presented with erythroderma (1 case, 2.13%).

Itching was absent in most of our patients (35 cases, 74.47%), and present in a quarter of them, being mild in the majority of the cases (11 cases, 23.40%), with one presentation referring intense itching (1 case, 2.13%).

The most frequent affected areas were the thighs (25 cases, 53.19%), trunk (23 cases, 48.4%), upper limbs (15 cases, 31.91%) and armpits (12 cases, 25.53%).

Our most frequent differential diagnoses were granuloma annulare (20 cases, 42.55%), followed by morphoea (14 cases, 29.79%) and mycosis fungoides (12 cases, 25.53%).

Associated systemic diseases were observed in 29 cases (61.70%), TNF inhibitor treatments were found in 4 cases (8.51%).

Out of all patients who attended follow-up visits, the lesions disappeared in 29 of them (74.36%), starting at 2 weeks up to 2 years (mean 4.12 months). One patient died of an unrelated condition shortly after the diagnosis.

On histological examination, diffuse and interstitial infiltrate was present in all cases, being dense in 19 cases (40.43%), moderate in 26 cases (55.32%) and mild in 2 cases (4.26%). In all cases histiocytes were found on the infiltrate, which was located from superficial to deep dermis. Histiocytes around collagen fibres (“floating sign”) were found in 36 cases (76.6%). Collagen degeneration was found in 35 cases (74.47%). Perivascular infiltrate was found in 36 cases (76.6%).

Conclusion:

We present 47 cases of IGD, a condition with unspecific clinical manifestations in which the finding of this histological pattern might be the key to initiate further studies and the early diagnosis of systemic diseases. Furthermore, frequently used treatments like TNF inhibitors should also be considered as a potential cause, with its suppression being crucial to resolve this dermatosis.

Abstract N°: 5489

A Case of Wolf's Isotopic Response presenting as a linear IgA bullous dermatosis

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Introduction & Objectives:

Wolf's isotopic response refers to the emergence of a new, unrelated skin disorder at the site of a previously healed cutaneous disease. The most frequently implicated initial condition is herpes zoster, while subsequent lesions have commonly included granulomatous disorders or cutaneous malignancies. We present a rare case of linear IgA bullous dermatosis (LABD) developing at the site of a previously resolved herpes zoster infection.

Materials & Methods

A 71-year-old male patient presented with a painful, erythematous, vesiculobullous eruption localized to the left T7 dermatome. The lesions had been present for approximately two months, with progressive extension to adjacent sites. The patient reported a episode of herpes zoster involving the same dermatome six months prior to presentation. Histopathological examination of a lesional skin biopsy, along with direct immunofluorescence, was consistent with a diagnosis of linear IgA bullous dermatosis (LABD). Based on the clinical history, dermatomal distribution of the lesions, and immunopathological findings, a diagnosis of LABD occurring as a manifestation of Wolf's isotopic response was established.The patient was initiated on systemic corticosteroid therapy in combination with oral dapsone. Significant clinical improvement was observed within four weeks of initiating treatment

Results:

To our knowledge, this is only the second reported case of linear IgA bullous dermatosis presenting as a Wolf's isotopic response following herpes zoster infection. In the latest review, Ruocco et al. analyzed previously documented cases of isotopic responses. While a wide spectrum of secondary dermatoses has been described in this context—including granulomatous, lichenoid, and neoplastic conditions—bullous autoimmune disorders remain exceedingly rare. Notably, only a single case of linear IgA bullous dermatosis following herpes zoster has been reported in the literature to date, published in 2008.

The precise pathophysiological mechanisms underlying Wolf's isotopic response remain incompletely understood. However, emerging evidence supports a multifactorial etiology involving viral persistence, neuronal damage, immune dysregulation, and localized vascular alterations. Some authors have hypothesized that remnant viral particles within the affected dermatome may act as immunologic triggers, thereby initiating the bullous response.

Conclusion:

Wolf's isotopic response is a rare and underrecognized phenomenon characterized by the development of a new, unrelated skin disorder at the site of a previously healed dermatosis. We report a new case of linear IgA bullous dermatosis occurring at the site of a prior varicella-zoster virus (VZV) infection. This case reinforces the concept of the **"immunocompromised district"**, wherein neurotropic viral infections such as VZV may induce localized immune dysregulation, rendering the affected dermatome susceptible to subsequent immunologic or inflammatory skin diseases.

Abstract N°: 5562

Cancer During Treatment with Biologics in Immune-Mediated Inflammatory Diseases: A Systematic Review and Meta-Analysis

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Introduction & Objectives:

Research on the carcinogenic properties of biologics is conflicting. While some studies report an increased risk of cancer following treatment with biologics for immune-mediated inflammatory diseases (IMIDs) such as psoriasis, rheumatoid arthritis and inflammatory bowel disease, others report no or decreased risk. A lack of an association between treatment with biologics and risk of cancer might be attributed to the generally low number of included patients and rarity of cancer, thus requiring larger trials or a meta-analysis to investigate such an association. We therefore conducted a meta-analysis, with the aim of assessing the incidence of cancer during treatment with biologics in chronic IMIDs and assessing the risk of cancer associated with biologics compared with other treatments or placebo.

Materials & Methods:

This systematic review and meta-analysis searched PubMed, Embase, Cochrane Library, and Web of Science up to January 13, 2025, for studies assessing the incidence and risk of cancer in patients with IMIDs treated with biologics compared to controls (PROSPERO; CRD42023394492). IMIDs included patients with psoriasis, psoriatic arthritis, rheumatoid arthritis, ankylosing spondylitis, axial spondylarthritis, and inflammatory bowel disease. Controls included patients receiving placebo or non-biological treatment. Pooled risk estimates were calculated using the exact Mantel-Haenszel method to account for cancer being a rare event.

Results:

Overall, 299 studies comprising 195,880 patients (n=141,816 adults; n=54,064 children) were included. In randomized controlled trials, no significantly increased or decreased crude risk of cancer was found for Tumor Necrosis Factor-α inhibitors (TNFi; risk ratio (RR) 1.30 [95%confidence interval (CI): 0.90-1.89],p=0.16), interleukin-6 inhibitors (RR: 1.46 [95% CI: 0.62-3.45], p=0.38), interleukin-23 inhibitors (RR: 1.33 [95% CI: 0.38-4.69], p=0.65) or any other assessed biologics. In observational studies reporting adjusted hazard ratios (HRs), no association was found comparing TNFi with conventional therapies (HR: 0.90 [95% CI, 0.43-1.86], p=0.71) for cancer overall. However, there was a decreased crude risk of cancer overall (RR: 0.55 [95% CI: 0.52-0.58], p<0.0001; incidence rate ratio: 0.69 [95% CI: 0.65-0.73], p<0.0001) and several individual cancers following treatment with TNFi compared with conventional therapies. In observational studies in children with IMIDs treated with TNFi compared to conventional therapies, the crude RR of cancer overall was 1.60 (95%CI: 0.96-2.67, p=0.069).

Conclusion:

The study supports the notion of no increased risk of cancer following treatment with for any of the assessed biologics. However, most studies were conducted on TNFi and are subject to potential confounding limitations. Nevertheless, these results give reassurance to patients and physicians prescribing biologics.

Abstract N°: 5626

Analysis of Paraneoplastic Pemphigus Patients' Age, Gender, Cancer Types, and Interval Between Cancer Diagnosis and Pemphigus Onset: A 30-Year Retrospective Cohort Study

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Introduction & Objectives:

Paraneoplastic pemphigus (PNP) is a rare autoimmune blistering skin disease, most commonly associated with underlying malignancies, particularly hematologic neoplasms. However, data on the prevalence of cancer types in PNP and the temporal relationship between cancer diagnosis and the onset of pemphigus symptoms remain limited. This study aims to investigate the spectrum of malignancies in patients with PNP and to analyze the timing of cancer diagnosis in relation to the onset of pemphigus symptoms, along with age and gender distribution.

Materials & Methods:

This is a retrospective cohort study. In this study, medical records of patients who presented to the Department of Dermatology at Ondokuz Mayıs University Faculty of Medicine between 1995 and 2025 were reviewed. Patients diagnosed with paraneoplastic pemphigus based on clinical and/or histopathological findings were included. Demographic characteristics, types of malignancies, and the time interval between cancer diagnosis and the onset of pemphigus symptoms were analyzed using descriptive statistical methods.

Results:

Among 524 patients diagnosed with pemphigus (234 males, 289 females), 24 (4.58%) were found to have at least one associated malignancy. Of these, 15 were male (62.5%) and 9 were female (37.5%). The mean age at onset of pemphigus symptoms was 58 years (range: 33–80), and the mean age at cancer diagnosis was 62 years (range: 37–80). In 19 patients (79.1%), pemphigus symptoms preceded the cancer diagnosis, while in 5 patients (20.9%), cancer was diagnosed before the onset of pemphigus. The average time from pemphigus onset to cancer diagnosis was 85 months, whereas the average time from cancer diagnosis to onset of pemphigus was 114 months.

The spectrum of malignancies included both solid tumors and hematologic cancers. The most frequently observed malignancy was lung cancer (n=4, 16.6%), followed by breast cancer and bladder cancer (each n=3, 12.5%). Other malignancies included colorectal cancer (n=2), glioblastoma (n=2), and single cases of laryngeal cancer, malignant pituitary neoplasm, chronic lymphocytic leukemia, malignant melanoma, prostate cancer, esophageal squamous cell carcinoma, renal cell carcinoma, micropapillary thyroid carcinoma, pancreatic cancer, and non-Hodgkin lymphoma. The most common malignancy among female patients was breast cancer (n=3, 33.3%), while among male patients, it was lung cancer (n=4, 26.6%).

Conclusion:

This study demonstrates that, in addition to hematologic malignancies, solid tumors may also be frequently observed in patients with PNP. Lung cancer in males and breast cancer in females were the most commonly identified malignancies. Contrary to previous reports, these findings suggest that solid tumors may be more prevalent in PNP than previously thought. Furthermore, the finding that pemphigus symptoms often precede cancer diagnosis highlights the importance of malignancy screening in patients presenting with pemphigus and

may help earlier detection and management of underlying neoplasms.

Abstract N°: 5634

Reverse Koebner Phenomenon in vitiligo patients treated with topical application of TCA 80%

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Introduction & Objectives:

Vitiligo is a chronic depigmenting disorder characterized by well-demarcated white macules resulting from melanocyte loss in the epidermis and hair follicles. While its precise pathogenesis remains incompletely understood, various triggering mechanisms have been proposed.

The Koebner phenomenon—first described by Heinrich Koebner in 1876—is the appearance of new lesions at sites of skin trauma and is observed in vitiligo, psoriasis, lichen planus, among others.

Conversely, the Reverse Koebner phenomenon (RKP) refers to lesion resolution or improvement following trauma, a much rarer and poorly understood process. Reports of RKP following the application of trichloroacetic acid (TCA) in vitiligo are exceedingly rare.

Materials & Methods:

We selected vitiligo patients exhibiting a positive Koebner response—regardless of disease activity—who were resistant to conventional therapies.

An 80% concentration of trichloroacetic acid (TCA) was applied using a toothpick, directed from the center of the vitiliginous patches outward, maintaining a 2 mm margin from the edges. The procedure endpoint was the development of uniform frosting over the treated area. Sessions were repeated every 4 weeks with consistent technique.

Results:

Peripheral repigmentation was observed in nearly all cases, leading to a reduction in lesion size. Marginal repigmentation appeared more prominently than central or diffuse repigmentation patterns.

Conclusion:

Our findings suggest that controlled topical application of 80% TCA may induce a Reverse Koebner phenomenon and serve as an effective adjunct in vitiligo patients with Koebner-positive lesions unresponsive to standard treatments. This method could represent a promising direction in the management of refractory vitiligo.

Abstract N°: 5644

Epidemiological and clinical profile of gestational pemphigoid: a retrospective 11-year study (2014–2025)

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Introduction & Objectives:

Gestational pemphigoid (GP) is a rare autoimmune subepidermal blistering disorder that occurs specifically during pregnancy. It typically manifests in the third trimester, although it can also appear in the second trimester or postpartum (PP). Treatment strategies are based on the severity of the condition, and recurrence in subsequent pregnancies tends to occur earlier and with increased severity. This study aims to assess the epidemiological, clinical, therapeutic, and evolutionary aspects of gestational pemphigoid.

Materials & Methods:

A retrospective descriptive study was conducted over an 11-year period (2014–2025) in the dermatology department. All cases of gestational pemphigoid diagnosed clinically, histologically, and by direct immunofluorescence were included.

Results:

The average age of onset was 27 years (range 20-40 years). The condition predominantly affected multiparous women, with 64% of cases occurring in this group. It was most commonly observed in the third trimester (10 cases), followed by the second trimester (2 patients), and in a few postpartum cases (2 patients). All patients reported intense pruritus preceding the skin eruption. Clinical examination revealed urticarial plaques in 13 patients and bullous lesions in 12 patients. The lesions typically began in the peri-umbilical region and then spread to the trunk (100% of cases) and upper and lower limbs (7 cases). The face was spared in all but two cases. No post-bullous erosions were observed. The diagnosis was confirmed by skin biopsy and direct immunofluorescence (DIF), which showed linear deposition of C3 at the dermo- epidermal junction. Treatment included systemic corticosteroids, dermocorticoids, and antihistamines in all cases. The outcome was favorable, with remission achieved in all patients except one, who experienced fetal death.

Conclusion:

Our study aligns with the existing literature on gestational pemphigoid (GP), especially regarding its higher prevalence in multiparous women and the frequent onset during the second and third trimesters. However, our findings also highlight clinical variability, as we observed cases in primiparous women and postpartum, as well as facial involvement in 14.2% of patients and lesions affecting the extremities. These variations suggest a broader clinical spectrum than previously reported. The maternal prognosis is generally favorable, although recurrences are common in subsequent pregnancies, often occurring earlier and with increased severity, regardless of the partner. Fortunately, no cases of fetal malformation or intrauterine mortality were observed. Treatment with dermocorticoids and antihistamines proved beneficial, especially for localized forms of the condition, while gradual reduction of systemic corticosteroids postpartum is recommended to avoid exacerbations. In conclusion, gestational pemphigoid, although rare, has a relatively favorable maternal prognosis when managed appropriately. Recurrences can occur in future pregnancies, but early intervention with corticosteroids and antihistamines can lead to favorable outcomes. Early management is essential to prevent complications.

Abstract N°: 5677

Evaluation of Factors Associated with Remission and Relapse in Patients with Bullous Pemphigoid: a retrospective study

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Introduction & Objectives:

Bullous pemphigoid (BP) is a chronic, autoimmune blistering disease predominantly affecting the elderly. While multiple studies have explored mortality risk factors, limited data exist on predictors of remission and relapse. This study aimed to assess these outcomes in a cohort of BP patients.

Materials & Methods:

In this retrospective study, 131 patients with bullous pemphigoid referred to Razi Hospital between 2001 and 2022 were evaluated, each with a minimum follow-up of 4 months. Data collected included demographic characteristics (age, sex), disease severity, mucosal involvement, initial systemic and topical corticosteroid doses, history of rituximab administration, hospitalization history, number of admissions, presence of circulating autoantibodies, erythrocyte sedimentation rate (ESR), and comorbidities. These variables were analyzed for associations with remission and relapse.

Results:

Of the 131 patients, 104 (79.3%) achieved clinical remission after a mean treatment duration of 461 days (SD = 239.4). Significant prognostic factors for remission included absence of diabetes, absence of neurological diseases, and history of rituximab use. Patients who had received rituximab and those with fewer hospital admissions achieved remission faster. Conversely, 66 patients (50.4%) experienced at least one relapse, with a mean time to first relapse of 193 days (SD = 103.6). A history of Parkinson's disease was significantly associated with relapse; however, notably, none of the four patients with Parkinson's experienced a relapse during the follow-up. The first relapse occurred sooner in patients without a history of rituximab use or Alzheimer's disease.

Conclusion:

This study identified several factors potentially influencing the remission and relapse of BP, including comorbidities and treatment history. Absence of certain comorbidities and use of rituximab were linked with better remission outcomes. Further large-scale, prospective studies are recommended to validate and expand upon these findings.

Abstract N°: 5706

An analysis of the safety and effectiveness of treating vitiligo with microneedling alone and in combination with N-acetylcysteine

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Introduction & Objectives: The selective breakdown of melanocytes results in vitiligo, a condition affecting skin pigmentation. In patients with restricted and persistent vitiligo, this study evaluates the therapeutic benefits of microneedling both with and without N-acetylcysteine (NAC). The aim is to evaluate the effectiveness of microneedling in treating individuals with limited and persistent vitiligo, both with and without N-acetylcysteine (NAC).

Materials & Methods: A clinical trial design with double-blind randomization was used in this study. Two distinct treatment groups were created for the study. In addition to applying 4.7% NAC cream once daily to the designated area, 24 afflicted areas received meso-microneedling with 5% NAC ampoules over the course of six sessions in the intervention group. On the other hand, the control group, which included 22 lesions, received six sessions of microneedling with distilled water. The Modified VETI Score was used to assess the degree of repigmentation and the severity of lesions. Both patient feedback and physician evaluations were used to assess the effectiveness of the treatment.

Results: 40 patients were included to the study, and their average age was 36.4 years. Two weeks following the third session and one month following the conclusion of the treatment, there was a substantial improvement in the mean percentage of lesions and their intensity ($p < 0.01$). The intervention and control groups did not vary in any way that was statistically significant. Patients' treatment outcomes were not statistically correlated with gender, age, family history, length of disease, duration of disease stability, or history of hypothyroidism ($p > 0.05$).

Conclusion: For persistent vitiligo lesions, microneedling, either with or without the use of NAC, seems to be a successful therapy strategy. Nevertheless, the difference was not statistically significant, even though the use of NAC resulted in a higher improvement rate.

Abstract N°: 5725

Association between mitochondria-associated endoplasmic reticulum membranes and cellular senescence in morphea

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Introduction & Objectives:

Cellular senescence is observed in morphea. The mitochondria-associated endoplasmic reticulum membrane (MAM) serves as a key regulatory structure of senescence. However, the relationship between MAM and senescence in morphea remains poorly understood.

Materials & Methods:

Morphea dataset GSE153011 was obtained from GEO for analysis. Differentially expressed genes (DEGs) were analyzed with limma package. These DEGs were intersected with MAM-related genes to derive MAM-Related and Morphea-Related Genes (MAM-MRGs). Furthermore, an intersection was performed between the DEGs and senescence-related genes to ascertain cellular senescence-related and morphea-related genes (CS-MRGs). Senescence-associated hub genes were identified using the CytoHubba plugin in Cytoscape. The correlation between senescence-associated hub genes and MAM-MRGs was evaluated using the corrplot package in R. The distance between the ER and mitochondria was quantified through transmission electron microscopy (TEM). The relationship between the protein expression of MAM gene and senescence-associated β-galactosidase (SA-β-gal) was assessed via immunohistochemistry (IHC).

Results:

We have identified nine MAM-MRGs. The majority of senescence-related hub genes demonstrate significant correlations with MAM-MRGs. Transmission electron microscopy (TEM) revealed structural abnormalities in MAMs within morphea lesions. IHC analysis demonstrated significant upregulation of ERO1L (MAM-MRGs) and SA-β-gal protein in skin lesion of morphea patients compared to control. Spearman correlation analysis revealed that expression levels of endoplasmic reticulum oxidoreductase 1L (ERO1L) protein showed a strong positive correlation with β- SA-β-gal protein.

Conclusion:

Our study reveals a potential link between MAMs and cellular senescence in morphea. Targeting MAM regulation may represent a novel therapeutic strategy for morphea.

Abstract N°: 5885

Lichen Planus Pemphigoides: A Distinct Autoimmune Blistering Disease with Clinical and Histological Overlap

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Introduction & Objectives: Lichen Planus Pemphigoides (LPP) is a rare autoimmune subepidermal blistering disorder characterized by features of both Lichen Planus (LP) and Bullous Pemphigoid (BP). Though once considered a clinical variant of these conditions, increasing evidence supports its classification as a distinct disease entity. Accurate diagnosis relies on a comprehensive assessment of clinical, histological, and immunological characteristics.

Materials & Methods: To illustrate diagnostic challenges, we include a representative clinical case. We conducted a selective literature review on LPP, with a focus on its epidemiology, pathogenesis, diagnostic criteria, and treatment approaches.

Results: We present the case of a 68-year-old female with a 1.5-month history of scaly plaques on the trunk, who had received a short course of oral corticosteroids, discontinued 10 days prior to presentation. She developed scaly papules on the palms and soles, along with tense bullae. Three skin biopsies were taken for histopathological and immunofluorescence evaluation. Her medical history included antihypertensive therapy, amlodipine, over the past 3 years. Family history revealed psoriasis in a sibling and discoid lupus erythematosus (DLE) in her mother. Differential diagnosis included psoriasis, dyshidrotic eczema, paraneoplastic dermatoses, autoimmune blistering diseases, and others. Histology and immunofluorescence supported the diagnosis of LPP. Literature data indicate that LPP can be triggered by drugs, with antihypertensive agents among reported suspects. The disease typically manifests with lichenoid plaques and blistering lesions, often in non-overlapping areas. Direct immunofluorescence reveals linear IgG and/or C3 deposition along the basement membrane. Treatment typically involves systemic corticosteroids and immunosuppressants, with emerging evidence supporting the role of biologics in refractory cases.

Conclusion: LPP is a diagnostically challenging but distinct autoimmune blistering disease. The coexistence of lichenoid and bullous lesions, along with the immunopathological findings, underscores the importance of a multidisciplinary diagnostic approach. Greater awareness of LPP and its triggers, including drug-induced cases, may lead to earlier diagnosis and more effective management strategies. Understanding its pathogenesis may also provide insights into related autoimmune blistering conditions.

Abstract N°: 5932

The Relationship Between Clinical Activation and CRP/Albumin and Fibrinogen/Albumin in Behçet's Disease

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Introduction & Objectives:

Behçet's Disease (BD) is a chronic, multisystemic vasculitis characterized by recurrent oral and genital ulcers, skin lesions, and systemic involvement. Diagnosing and monitoring disease activity remain challenging due to the lack of specific biomarkers. This study aimed to evaluate the role of two novel inflammatory markers—CRP-to-Albumin Ratio (CAR) and Fibrinogen-to-Albumin Ratio (FAR)—in distinguishing BD patients from healthy controls and assessing disease activity.The primary objectives were to compare CAR and FAR levels between BD patients and healthy controls, investigate the association of these ratios with clinical manifestations and disease activity and determine cutoff values for CAR and FAR to aid in BD diagnosis

Materials & Methods:

This prospective study included 62 BD patients (30 active, 32 inactive) diagnosed per ISG criteria and 28 matched healthy controls, recruited from Necmettin Erbakan University (June–October 2021). Exclusion criteria: pregnancy, diabetes, liver/kidney disease, other inflammatory conditions. Demographic and clinical data were collected; disease activity was assessed via BDCAF. Blood samples were analyzed for hematological and biochemical parameters. CAR and FAR were calculated. Data were analyzed using SPSS 22; normality tests and group comparisons were performed, and diagnostic value assessed via ROC analysis.

Results:

Mean age of BD patients was 38.62 ± 10.48 years; 53.3% were male.Active BD patients had a lower mean BMI (24.09 ± 2.31) compared to inactive BD (27.02 ± 5.37, *p* = 0.021).Oral ulcers (64.5%), arthralgia (54.8%), and papulopustular lesions (45.2%) were the most common manifestations. Active BD showed higher frequencies of oral/genital ulcers, skin lesions, and arthritis (*p* < 0.01). BD patients had significantly higher CRP, fibrinogen, WBC, neutrophil, monocyte, and RDW levels than controls (*p* < 0.05). Albumin levels were lower in BD patients (*p* = 0.009). CAR and FAR were elevated in BD patients (*p* = 0.015 and *p* = 0.001, respectively). Active BD had higher CAR than controls (*p* = 0.013), but no difference was observed between active and inactive BD. **CAR ≥ 0.051**: Sensitivity 64.52%, specificity 64.29%, AUC 0.660. BD risk increased 3.273-fold (*p* = 0.011).**FAR ≥ 6**: Sensitivity 67.74%, specificity 60.71%, AUC 0.749. BD risk increased 3.245-fold (*p* = 0.011).

Conclusion:

CAR and FAR are promising biomarkers for distinguishing BD patients from healthy controls, with moderate sensitivity and specificity. However, they did not differentiate between active and inactive disease, highlighting the need for more specific markers. Elevated CAR and FAR correlated with skin and joint involvement, suggesting their potential role in monitoring disease severity. Further studies with larger cohorts are warranted to validate these findings and explore their clinical utility in BD management.CAR and FAR may aid in BD diagnosis but not in assessing disease activity. Active BD patients had lower BMI and albumin levels, reflecting systemic inflammation.

Cutoff values (CAR ≥ 0.051, FAR ≥ 6) could serve as adjunct tools in clinical practice. This study contributes to the search for accessible, cost-effective biomarkers in BD, though challenges remain in defining disease activity markers.

Abstract N°: 6060

Type I interferon and type I interferon induced biomarkers in dermatomyositis

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Introduction & Objectives:

Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy characterized by progressive symmetric proximal skeletal muscle weakness, with pathognomonic skin manifestations including Gottron papules and heliotrope rash. Type I interferons (IFN) and type I IFN-inducible genes have been shown to be uniquely upregulated in DM compared to other muscle diseases, and may be important in DM pathogenesis. Given the potential of biomarkers to improve understanding of DM pathogenesis and provide therapeutic targets, and the unique upregulation of type I IFN in DM compared to other types of myositis, this systematic review aims to identify potentially clinically relevant type I IFN (IFN-α, IFN-β, IFN-ω, IFN-ε, IFN-δ, and IFN-κ) or type I IFN-inducible biomarkers in DM.

Materials & Methods:

This systematic review (PROSPERO: CRD42025637247) was conducted according to the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) guidelines. Medline and Embase databases were search from database conception to December 2024 for studies on DM, IFN, and biomarkers. Eligible studies included English-language case-control or cohort studies on type I IFN biomarkers in active DM. Two reviewers independently screened and extracted data on patient demographics, disease and biomarker characteristics, and outcomes.

Biomarkers were categorized by medium and assay type, and associations with disease severity were assessed. Risk of bias was evaluated using the Newcastle-Ottawa Scale, and evidence certainty was graded using GRADE, considering bias, indirectness, imprecision, and inconsistency.

Results:

Of the 690 articles retrieved, 38 articles with 794 DM patients (mean age 35.9 years; 73.1% female) reporting on 29 biomarkers were included for analysis. Subtypes included adult DM (48.1%), juvenile DM (38.4%), and clinically amyopathic DM (13.5%).

The most well-studied mRNA and protein biomarkers included IFN-α, IFN-β, C-X-C motif chemokine ligand 10 (CXCL10), myxovirus resistance 1 (MX1), interferon-stimulated gene 15 (ISG15), monocyte chemoattractant protein 1 (MCP-1) and interferon-inducible T cell alpha chemoattractant (I-TAC). All of these biomarkers were overexpressed in DM patients in skin, muscle, serum and peripheral blood mononuclear cells (PBMCs) compared to healthy subjects, with fold increases up to 408. They also correlated with disease severity, with the exception of IFN-α. CXCL10 and MCP-1 showed strong correlations with cutaneous and global disease activity scores, including CDASI and VAS. MX1, ISG15, and I-TAC also demonstrated positive, albeit limited, correlations with muscle disease activity. Correlations with functional muscle scores (e.g., CMAS, MMT) were limited and inconsistent. Evidence certainty varied, with CXCL10 supported by high-certainty evidence and others by low-to-moderate certainty.

Conclusion:

The results of this systematic review suggest the type I IFN activity in DM is primarily driven by IFN-β. IFN-β, CXCL10, MX1, ISG15, MCP-1, and I-TAC may have utility as a biomarker panel for both DM diagnosis and tracking of disease progression or response to treatment. Future clinical trials investigating the efficacy of anti-IFN-β in DM with measurement and correlation of these biomarkers with disease severity are warranted.

Abstract N°: 6094

LP Pemphigoides

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Introduction & Objectives: Lichen Planus Pemphigoides (LPP) is a rare autoimmune subepidermal blistering disorder characterized by features of both Lichen Planus (LP) and Bullous Pemphigoid (BP). Though once considered a clinical variant of these conditions, increasing evidence supports its classification as a distinct disease entity. Accurate diagnosis relies on a comprehensive assessment of clinical, histological, and immunological characteristics.

Materials & Methods: To illustrate diagnostic challenges, we include a representative clinical case. We conducted a selective literature review on LPP, with a focus on its epidemiology, pathogenesis, diagnostic criteria, and treatment approaches.

Results: We present the case of a 68-year-old female with a 1.5-month history of scaly plaques on the trunk, who had received a short course of oral corticosteroids, discontinued 10 days prior to presentation. She developed scaly papules on the palms and soles, along with tense bullae. Three skin biopsies were taken for histopathological and immunofluorescence evaluation. Her medical history included antihypertensive therapy, amlodipine, over the past 3 years. Family history revealed psoriasis in a sibling and discoid lupus erythematosus (DLE) in her mother. Differential diagnosis included psoriasis, dyshidrotic eczema, paraneoplastic dermatoses, autoimmune blistering diseases, and others. Histology and immunofluorescence supported the diagnosis of LPP. Literature data indicate that LPP can be triggered by drugs, with antihypertensive agents among reported suspects. The disease typically manifests with lichenoid plaques and blistering lesions, often in non-overlapping areas. Direct immunofluorescence reveals linear IgG and/or C3 deposition along the basement membrane. Treatment typically involves systemic corticosteroids and immunosuppressants, with emerging evidence supporting the role of biologics in refractory cases.

Conclusion: LPP is a diagnostically challenging but distinct autoimmune blistering disease. The coexistence of lichenoid and bullous lesions, along with the immunopathological findings, underscores the importance of a multidisciplinary diagnostic approach. Greater awareness of LPP and its triggers, including drug-induced cases, may lead to earlier diagnosis and more effective management strategies. Understanding its pathogenesis may also provide insights into related autoimmune blistering conditions.

Abstract N°: 6120

a case report:h syndrome

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Introduction & Objectives: H syndrome is a rare autosomal recessive genodermatosis caused by loss-of-function mutations in the *SLC29A3* gene, which encodes the equilibrative nucleoside transporter ENT3. The syndrome is characterized by cutaneous hyperpigmentation, hypertrichosis, hepatosplenomegaly, heart abnormalities, hearing loss, hypogonadism, short stature, hallux valgus, sclerodermatous skin thickening, hyperglycemia, and fixed flexion contractures of the toe joints. To date, only 100–120 cases have been reported in the literature, predominantly among individuals of Indian, North American, and Arab descent.

Materials & Methods: A 30-year-old female presented to our outpatient clinic with bilateral inner thigh hyperpigmentation and induration on palpation. She had a history of hearing loss, hallux valgus, inner thigh hyperpigmentation since birth, and underdeveloped secondary sexual characteristics. Laboratory tests, a skin biopsy, ultrasound, ECG, and audiometric examination were performed.

Results: Laboratory tests revealed elevated blood glucose and C-reactive protein levels, along with microcytic anemia. Liver enzymes and erythrocyte sedimentation rate were within normal limits. Growth hormone and thyroid function tests were normal. Audiometry showed bilateral sensorineural hearing loss. Histopathological examination revealed hyperpigmentation of the basement membrane and an irregular, acanthotic epidermis. Lymphoid aggregates and plasma cells were noted in the dermis, along with fibrosis. Immunohistochemical analysis showed diffuse staining of mast cells with CD117 and CD68, and S-100 positivity. Based on these findings, the patient was diagnosed with H syndrome.

Methotrexate treatment was initiated in November 2023. At follow-up, both hyperpigmentation and induration had significantly improved.

Conclusion: H syndrome is an extremely rare autoinflammatory disorder that requires further research for improved identification and a better understanding of its pathophysiology. The diagnosis should be considered in patients presenting with hyperpigmentation and hypertrichosis in characteristic anatomical regions.

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Abstract N°: 6143

Reticular erythematous mucinosis and lupus erythematosus tumidus in the same patient – can we distinguish them using dermoscopy?

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Introduction & Objectives: The relationship between reticular erythematous mucinosis (REM), a rare condition traditionally classified as a primary idiopathic cutaneous mucinosis, and lupus erythematosus tumidus (LET) has yet to be fully clarified. Due to clinical and histological overlap, some authors consider them as closely related conditions or manifestations of chronic or intermittent cutaneous lupus erythematosus (LE). To date, no reports in the literature describe the simultaneous occurrence of both conditions in the same patient. Additionally, dermoscopic descriptions of these diseases remain scarce.

We present a patient with concurrent REM and LET, highlighting dermoscopic findings and criteria that may aid in their differentiation.

Materials & Methods: The patient was evaluated using clinical examination, dermoscopy and histopathology.

Results: A 51-year-old man presented as an outpatient due to recurrent cutaneous lesions and severe pain in nearly all joints. Clinical examination revealed reddish, smooth annular plaques and succulent papules on the back, forearms, and upper arms, along with maculopapular reticular erythema on the central chest. He had been diagnosed with cutaneous LE eight years earlier, based on two biopsies – one consistent with LET, and the other with LE due to focal vacuolar alteration of basal epidermal cells. Cutaneous symptoms worsened after sun exposure, with relapses typically occurring in spring. Topical corticosteroids, photoprotective agents, and systemic treatments including chloroquine, hydroxychloroquine, prednisone, and methotrexate had limited efficacy and failed to prevent disease relapses. Over eight years of clinical and laboratory follow-up, the patient did not fulfill the criteria for systemic lupus erythematosus (SLE) or show signs of inflammatory arthropathy. An autoimmune workup revealed a positive ANA titer and borderline positive anti-U1-RNP antibodies, with normal FBC, complement components C3 and C4, and general biochemistry results. For the past 20 years, the patient has suffered from diabetes, significant degenerative joint changes, cervicobrachial syndrome, C5–C6 disc protrusion, lumboischialgia, Charcot arthropathy, and polyneuropathy. He has also been a smoker for the past 20 years. A dermoscopy of the annular plaque revealed subtle orange-reddish globules on a pinkish-reddish background. A dermoscopy of the maculopapular erythema on the chest showed dotted and linear vessels, along with uniform, structureless yellowish-white oval areas. Histopathological findings of the annular plaque revealed dense perivascular and periadnexal lymphocytic infiltrates with mucin deposits in the mid and deep reticular dermis, without epidermal changes – findings consistent with a diagnosis of LET. In contrast, the biopsy of the reticular erythema on the chest showed sparse perifollicular and superficial dermal lymphocytic infiltrates with mucin deposits in the upper dermis, consistent with a diagnosis of REM.

Conclusion: The simultaneous presence of REM and LET lesions in the same patient suggests a shared pathogenesis and raises the question of whether they represent a continuum of the same disease. The dermoscopic finding of yellowish structureless areas – corresponding to more superficial mucin deposits between collagen tissue – along with dotted and linear blood vessels, may help differentiate REM from LET, in which dermoscopic features are generally non-specific.

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Abstract N°: 6155

Juvenile pemphigus vulgaris in a patient with intestinal helminth infection

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Introduction & Objectives:

Pemphigus vulgaris (PV) is a rare autoimmune disease that manifests clinically with blistering of the skin and/or mucous membranes. Although PV is most commonly seen in adults, it is also reported in children with an estimated incidence of eight cases per million per year. Juvenile PV (JPV) typically affects adolescents between the age of 10-18 years and usually manifests benign course and good prognosis. The exact etiology of pemphigus in all age groups remains unclear, considering the role of genetic predisposition and a wide range of environmental triggering factors, only occasionally including parasitoses. We observed a case of fatal JVP coinciding with pinworm infection.

Materials & Methods:

An 11-year old girl presented with a widespread vesiculo-bullous eruption of two years duration. Multiple vesicles and bullae with clear contents, followed by painful erosions covered with serohaemorrhagic crusts involved the scalp, face, trunk and extremities. All mucous membranes, including those of the eyes, oral cavity, nose, genitals, perianal zones, as well as the mucosa of the pharynx and esophagus were affected. In addition, the child was complaining of nausea, vomiting and occasional abdominal pain, but being institutionalized, family history or other comorbidities were poorly documented.

Results:

The routine complete blood count detected only mildly elevated levels of erythrocytes and platelets. The histology of a skin lesion revealed suprabasal epidermal acantholytic blisters filled with eosinophils and acantholytic cells. Direct immunofluorescence on perilesional skin showed intercellular deposition of IgG (++) and C3(+) in the epidermis. Serum ELISA detected positive anti-desmoglein (Dsg) 1 and 3 antibodies. The child's gastro-intestinal complaints were proven to be related to concomitant *Enterobius vermicularis* infestation. High-dose systemic corticosteroids and antibiotics along with topical antiseptics and potent dermocorticoids achieved slow and unstable improvement. Antihelminthic therapy with albendazole for 3 days was added to treat the pinworm infection. Soon after discharge, while on corticosteroid tapering doses, the patient's gastro-intestinal complaints worsened abruptly with significant deterioration of her general condition, followed by a fatal outcome.

Conclusion:

PV in children and adolescents, if detected on time, demonstrates better long-term prognosis than in adults and only rarely exhibits a malignant course. The presence of intestinal parasitosis in the reported JPV might be a mere coincidence, reflecting the poor socio-economic status of the patient. However, a causative relationship may also be suspected, given the reported PV onset in individuals with gut microbiome abnormalities or exacerbation of gastro-intestinal infections by the immunosuppressive pemphigus therapy. Future experiments are required to elucidate the causal relationship between the helminth infection, gut microbiome, and PV.

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Abstract N°: 6182

a case of subacute cutaneous lupus erythematosus induced by terbinafine, presenting with erythema multiforme-like lesions

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Introduction & Objectives: Cutaneous lupus erythematosus (CLE) is a collagen vascular disease categorized into three subtypes: acute, subacute (SCLE), and chronic. SCLE typically manifests with annular or papulosquamous lesions; however, erythema multiforme-like lesions are rarely seen. Druginduced SCLE has been documented in the literature, with terbinafine recognized as a highrisk agent. In this report, we present a rare case of terbinafine-induced SCLE with an atypical erythema multiforme-like presentation.**

Materials & Methods: A 51-year-old female patient was admitted to our dermatology inpatient unit due to widespread cutaneous eruptions. She had been prescribed systemic terbinafine for tinea corporis one month prior. Two weeks after initiating terbinafine, a rash first appeared on the trunk and subsequently spread to involve the entire body.

Dermatological examination revealed erythematous plaques on the scalp and face, painful atypical macular targetoid lesions on the bilateral forearms, and violaceous erythematous tender plaques covering approximately 50–60% of the body surface area, with a tendency to coalesce.

Results: : Histopathological examination showed vacuolar interface dermatitis with follicular involvement, widespread parakeratosis, and mild dermal mucin deposition. Direct immunofluorescence testing was negative. Laboratory analyses revealed positive antinuclear antibody (ANA) at 1:100 titer (2+, fine speckled), positive anti-SSA, and mildly elevated extractable nuclear antigen (ENA) at 5.09+.

The patient was treated with oral cyclosporine (200 mg/day) and methylprednisolone (60 mg/day). Full resolution of cutaneous lesions was observed after 15 days of treatment.

Conclusion: : Although SCLE most often presents with annular or papulosquamous plaques, drug-induced variants may manifest with widespread, bullous, vasculitic, or erythema multiforme-like lesions. In this case, the presence of painful, generalized targetoid lesions initially raised suspicion of a drug reaction. However, histopathological findings did not demonstrate necrosis or dyskeratotic keratinocytes as expected in erythema multiforme. The combination of ANA positivity, erythema multiforme-like lesions, and a history of terbinafine use prompted consideration of Rowell syndrome; however, the absence of systemic lupus erythematosus (SLE) features and discoid lesions ruled out this diagnosis. Based on clinical morphology, serologic profile, histopathology, and drug history, a diagnosis of terbinafine-induced SCLE with an erythema multiforme-like presentation was made. This rare variant highlights the need for thorough differential diagnosis in patients presenting with unusual cutaneous eruptions following drug exposure.

Abstract N°: 6209

Pediatric pemphigus foliaceum

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Introduction & Objectives:

Pemphigus foliaceum is an exceptionally rare autoimmune bullous disease in children, accounting for less than 5% of pediatric pemphigus. It manifests as superficial skin lesions, without mucosal involvement, due to autoantibodies directed against desmoglein-1. Its rarity and diversity of clinical presentations mean that diagnosis is often delayed. We report an illustrative case highlighting these challenges.

Materials & Methods:

A 6-year-old boy presented for six months with annular erythemato-vesiculocrustal lesions affecting the face, scalp, trunk and limbs. Pruritus was moderate, the superinfected lesions were painful, and retroauricular and inguinal adenopathies were noted. Biological tests were normal overall, apart from moderate anemia. The initial histological study was non-specific, showing suprabasal intraepidermal cleavage, while a second biopsy suggested junctional bullous dermatosis. Direct immunofluorescence revealed intercellular IgG and C3 meshwork deposits, confirmed by positive indirect immunofluorescence for anti-desmoglein 1 antibodies, leading to the diagnosis of superficial pemphigus foliaceum of childhood, Treatment with prednisolone and dapsone resulted in rapid improvement of the PDAI score. However, an early relapse necessitated an increase in corticosteroid doses, leading to severe side effects including a cushingoid facies and significant weight gain. Despite satisfactory clinical control with intensive corticosteroid therapy, toxicity prompted consideration of alternative therapies. **Results:**

Pediatric pemphigus foliaceum remains an exceptional pathology, with around thirty sporadic cases reported outside endemic areas. Diagnostic difficulties are compounded by the clinical similarity with other common childhood dermatoses, and by the frequency of non-contributory biopsies, especially if samples are taken from eroded or infected lesions. Confirmation relies on a rigorous immunopathological approach, combining histology and immunofluorescence.

From a therapeutic standpoint, although systemic corticosteroids are the mainstay of treatment, their efficacy as monotherapy remains limited and their toxicity significant, particularly in children. Combination with dapsone, as in our case, sometimes enables corticosteroid doses to be reduced, but does not always prevent relapses. Today, rituximab appears to be a promising therapeutic option, offering better disease control with reduced toxicity, and its early introduction is recommended in cases of failure or complications associated with conventional treatments. New alternatives, such as apremilast, are also being explored, although no pediatric data are yet available.
Conclusion:

Pemphigus foliaceum in children requires early and rigorous diagnosis, based on clinical, histological and immunological comparison. Multidisciplinary management is essential to limit the sequelae of the disease and the complications of treatment.

Abstract N°: 6282

Bullous pemphigoid associated with canagliflozin intake

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An 85-year-old Caucasian male was admitted to our inpatient clinic showing tense blisters on erythematous itchy skin, without mucosal involvement. The patient was also affected by ulcerative colitis, Hashimoto's thyroiditis, hypertension, and type 2 diabetes (T2D), treated with sulfasalazine 500 mg three times per day, L-thyroxin 100 µg/die, bisoprolol 5 mg/die, and canagliflozin 100 mg/die. He started complaining about pruritus and tense blisters on erythematous skin three months after starting canagliflozin after the interruption of linagliptin, which failed to control serum glucose levels. Pathologically, a subepidermal cleft with eosinophilic infiltrate was detected. In addition, a linear deposition of C3 and IgG along the dermo-epidermal junction was shown by direct immunofluorescence. However, indirect immunofluorescence on monkey esophagus was negative and ELISA for bullous pemphigoid (BP) 180 and BP230 (Mesacup, MBL Co.) only showed a weak positivity to NC16A domain of BP180 (10.0 U/ml; cut-off ≥ 9.0). Interestingly, elevated IgG levels were detected with in-house ELISAs based on BP180 entire ectodomain (ECD-BP180) (pemphigoid index units (PIV)=70.9; cut-off ≥10.02)1 and on its mid-portion (E-1080) and C-terminus (E-1331) epitopes (PIV= 367.6, cut-off ≥14.9; and PIV=21.2, cut-off ≥4.5, respectively). According to Naranjo's adverse drug reaction algorithm, a score of 6 was obtained. Therefore, the diagnosis of BP induced by canagliflozin was made. Canagliflozin was immediately discontinued, and the patient was treated with clobetasol propionate ointment 0.05% two times per day and prednisone 40 mg i.v., leading to a massive improvement of the clinical picture. Canagliflozin is a sodium-glucose linked transport protein 2 (SGLT2) inhibitor used to treat T2D, as monotherapy or in combination with other drugs, including metformin and pioglitazone. Common side effects of SGLT2 inhibitors are increased incidence of urinary tract infections, elevations in LDL cholesterol, increased urination, and reduction of blood pressure. However, to the best of our knowledge, canagliflozin has never been reported as a trigger for BP.

Abstract N°: 6306

Dupilumab for the treatment of Bullous Pemphigoid: a retrospective single center study

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Introduction & Objectives:

Bullous Pemphigoid (BP) is a chronic autoimmune blistering disease mainly affecting the elderly, characterized by tense blisters, itching, and redness. It is commonly associated with multiple comorbidities such as hypertension, diabetes, stroke, and neurological conditions, and significantly reduces quality of life, often leading to depression. Standard treatment involves topical and oral corticosteroids, but in severe or resistant cases, corticosteroid-sparing agents are used. However, treatment choices can be limited by comorbid conditions. In this single-center retrospective study, data on BP patients treated with dupilumab are presented, evaluating the drug's effectiveness and safety in a real-world clinical setting.

Materials & Methods:

This retrospective study analyzed data from a Dermatology Clinic in Greece. It included adult patients (>18 years) with a confirmed diagnosis of BP, verified by at least two diagnostic methods. All participants had received dupilumab treatment for at least three months prior to enrollment, using a dosing regimen similar to that for atopic dermatitis. Clinical and demographic data—including age, sex, comorbidities, diagnostic methods, and prior treatments—were collected. Disease activity was assessed using BPDAI scores at baseline, 3, 6, and 12 months. Anti-BP180 antibody levels were measured via ELISA at baseline and, where available, at 3 and 6 months to evaluate treatment response.

Results:

The study included 40 patients with Bullous Pemphigoid (BP), with a mean age at diagnosis of 76 years. All patients initiated dupilumab treatment while receiving oral corticosteroids (mean prednisolone dose: 26.7 mg/day), and four were also on azathioprine. On average, patients had been treated with oral corticosteroids for nine months prior to starting dupilumab (mean: 9 months, SD: 4.52). The mean baseline BPDAI score was 27.8, which showed substantial improvement at 3, 6, and 12 months, declining to 5.9, 2.0, and 1.5, respectively. At the time of assessment, 26 patients had achieved complete response, 10 had disease control with minimal corticosteroid use (<10 mg/day), and 4 were lost to follow-up. All four patients on azathioprine successfully discontinued its use. The average time to achieve complete response with cessation of oral corticosteroids was 3,85 months (mean: 3.5, SD: 1.3). Dupilumab was generally well tolerated, with patients also reporting reduced pruritus. Available data indicated a trend toward decreasing anti-BP180 antibody levels over time.

Conclusion:

Dupilumab appears to be an effective treatment option for BP, demonstrating clinical improvement, reduced corticosteroid dependence, and good tolerability. These findings support its potential role as a steroid-sparing agent in the management of BP, particularly in patients with multiple comorbidities.

Abstract N°: 6379

Off-Label Treatment of Bullous lupus Erythematous with Upadacitinib Leading to Clearance

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Introduction & Objectives:

Bullous lupus is a rare blistering eruption in patients with SLE. It presents as tense bullae on extremities, trunk, head and neck, which heals without scars. The clinical criteria include diagnosis of SLE, bullae, histopathology compatible with dermatitis herpetiformis, negative or positive indirect immunofluorescence (IIF) for circulating basement membrane autoantibodies, and direct IF showing IgG and/or IgM and often IgA at BMZ.1 Treatment options include dapsone, cyclophosphamide, azathioprine, mycophenolate mofetil, systemic steroids, rituximab, ankinra or IVIG.2 JAK inhibitors may improve SLE/CCLE by modulating IFN-alpha.3 Tofacitinib, a broad JAK inhibitor has been reported in phase I trialto be effective in SLE.3 Baricitinib has also been reported in a phase II trial to improve both skin disease and arthritis in SLE.4

Materials & Methods:

A 63 year-old female presented with a history of recurrent pruritic bullae on the dorsum of the hands that progressed to erosions and ulcers. These were worse with UV exposure. She had a history of rheumatoid arthritis and gout, treated with allopurinol, hydroxychloroquine and leflunomide, and had failed methotrexate. Topical high potency steroids and sunscreen did not help. She developed larger ulcers from coalesced bullae. A biopsy was performed which was compatible with CTD and IF was positive for IgG and IgA along the DEJ; workup for autoantibodies and porphyria were negative except mildly positive ANA. She had a history of CVA and herpes zoster, but no history of cancer. Based on a suspected diagnosis of bullous lupus and in discussion with her rheumatologist, her treatment was switched to Upadacitinib, a JAK1/2 inhibitor.

Results:

She started Upadacitinib 15mg daily. Within three months, the hand ulcer closed and she had no further bullae. She had no pruritus. Her RA symptoms were also improved.

Conclusion:

JAK inhibitors, including upadacitnib show promise in treating cutaneous manifestations of lupus. JAKs can be used to treat bullous lupus as a single agent and in those patients that are resistant to treatment. Future phase III trials are ongoing.

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Abstract N°: 6488

Flagellate Dermatitis: A Series of 4 Cases of Different Etiologies and Literature Review

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Introduction & Objectives: Flagellate dermatitis (FD) is an uncommon rash characterized by linear lesions on the trunk and limbs, often itching and self-limited. Nonetheless, it may serve as a clinical clue to underlying systemic diseases. The aim of this study was to present a case series of FD with different etiologies and emphasize its diagnostic value as a potential marker of autoimmune disorders and malignancies.

Materials & Methods: A retrospective analysis of four clinical cases treated at the Dermatology Department of Hospital Universitari Sagrat Cor was performed. Clinical data, patient history, progression of cutaneous findings, diagnostic procedures, and final diagnoses were collected. A literature review was also conducted to explore reported causes and histopathological features of FD.

Results: The series included:

1. A patient with Hodgkin's lymphoma who developed FD after 8 months of bleomycin treatment.
2. A woman who presented with pruritic FD 24 hours after ingesting Shiitake mushrooms.
3. A patient diagnosed with adult-onset Still's disease following a flare of FD.
4. A man in whom FD, along with generalized edema and erythema, led to the diagnosis of anti-NXP2 dermatomyositis. In all cases, FD was a key indicator that guided further investigation and diagnosis

Conclusion: Flagellate dermatitis, while rare, should be recognized as a significant clinical indicator pointing to potential pharmacologic toxicity, food-related reactions, or systemic conditions such as connective tissue diseases or malignancies. Early identification and thorough evaluation can support accurate diagnosis and tailored treatment with important prognostic implications.

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Abstract N°: 6516

HnRNPC Lactylation Regulating Fibroblast TRIM24 Alternative Splicing and Drives Fibrosis Activation in Systemic Sclerosis.

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HnRNPC Lactylation Regulating Fibroblast TRIM24 Alternative Splicing and Drives Fibrosis Activation in Systemic Sclerosis.

Introduction & Objectives:

Systemic sclerosis (SSc) is characterized by progressive fibrosis, yet the molecular mechanisms driving this pathology remain unclear. Emerging evidence suggests that lactylation, a novel post-translational modification, may play a role in fibrotic processes. This study aimed to investigate the involvement of lactylation in SSc pathogenesis, particularly its effects on fibroblast activation and the regulation of fibrosis-related signaling pathways.

Materials & Methods:

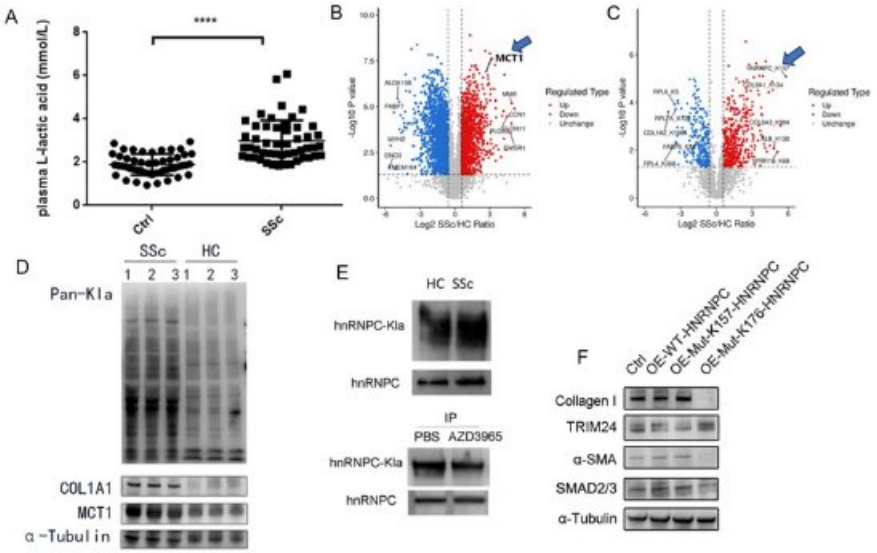
Plasma lactate levels were measured in SSc patients. Expression of the lactate transporter MCT1 was assessed in skin lesions. Fibroblasts isolated from SSc skin were analyzed for lactylation of fibrosis-related proteins. Lactylation proteomics and transcriptomic sequencing were conducted to identify target proteins and splicing changes. The regulatory effect of hnRNPC lactylation on TRIM24 expression and its impact on the TGF-β/Smad pathway were evaluated. A bleomycin-induced skin fibrosis mouse model was used to validate the findings in vivo.

Results:

SSc patients exhibited elevated plasma lactate levels and increased MCT1 expression in lesional skin. Lactate promoted lactylation of fibrosis-associated proteins and fibroblast activation. Proteomic analysis identified hnRNPC as a highly lactylated protein in SSc fibroblasts. Lactylation of hnRNPC led to alternative splicing and downregulation of TRIM24, a negative regulator of the TGF-β/Smad pathway via ubiquitination. TRIM24 expression was significantly reduced in SSc fibroblasts. In the bleomycin-induced fibrosis model, increased MCT1 expression, TRIM24 downregulation, and TGF-β/Smad activation were observed, consistent with in vitro findings.

Conclusion:

MCT1-mediated hyper-lactylation of hnRNPC in SSc fibroblasts suppresses TRIM24 expression through alternative splicing, impairing ubiquitination and promoting fibrosis via sustained TGF-β/Smad signaling. These findings reveal a novel mechanistic link between lactylation and fibrosis in SSc and suggest potential molecular targets for therapeutic intervention.


A
plasma L-lactic acid (mmol/L)
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Collagen I
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SMAD2/3
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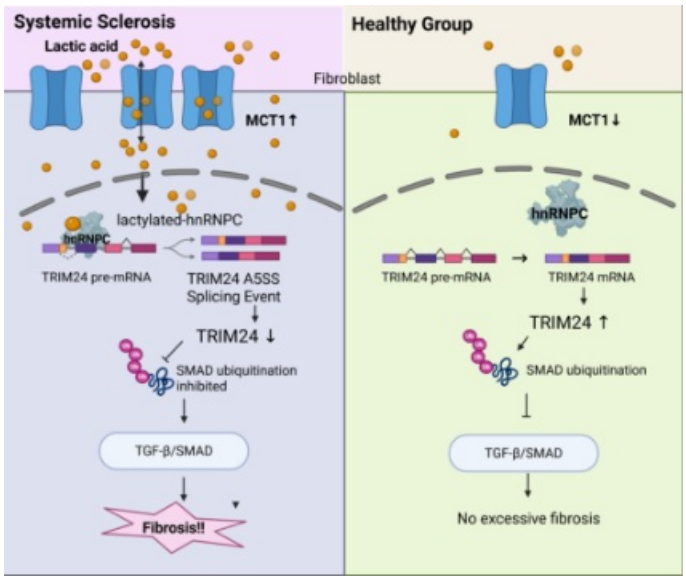
Systemic Sclerosis
Lactic acid
Fibroblast
MCT1↑
lactylated-hnRNPC
hnRNPC
TRIM24 pre-mRNA
TRIM24 A5SS
Splicing Event
TRIM24 ↓
SMAD ubiquitination
inhibited
TGF-β/SMAD
Fibrosis!!
Healthy Group
MCT1↓
hnRNPC
TRIM24 pre-mRNA
TRIM24 mRNA
TRIM24 ↑
SMAD ubiquitination
TGF-β/SMAD
No excessive fibrosis

Abstract N°: 6557

Efficacy and safety of rituximab therapy in immunobullous diseases: Experience from a cohort treated at a tertiary care center in ..

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Introduction & Objectives:

Studies on the long-term safety and efficacy of rituximab for immunobullous diseases are limited. The objective of this study was to assess the efficacy and safety of rituximab in patients with immunobullous diseases.

Materials & Methods:

This was a retrospective study of patients who received rituximab for immunobullous disease from January 2018 to January 2025, in the Department of Dermatology and Venereology……. All the patients had received rituximab as per the Rhenumatoid arthritis protocol and subsequent pulses were decided according to the disease activity. Definitions of disease outcome parameters were kept as per the international consensus. The study variables included age, sex, type of immunobullous disease, duration of disease, type of remission, relapse, and adverse effects. Descriptive analysis was performed using frequencies and percentages. Predictors of remission and relapse such as-age, sex, disease duration, timing of treatment (early vs late) and concomitant use of steroids, or azathioprine treatment)- were analyzed with general linear model in SPSS version 26.

Results:

Out of 63 patients who received rituximab for immunobullous diseases, there were 54 patients of pemphigus vulgaris, four patients of pemphigus foliaceous, and five patients of bullous pemphigoid. The mean age of the patient was 41.75 years (+/-14.9), with 21 males (33.3%) and 32 females (66.7%). Median duration of follow-up was 18 months (range:1 to 72 months). Remission data was retrieved for 50 patients, out of which 27 patients (54%) achieved complete remission, 19(38%) achieved partial remission and 4(8%) were refractory to treatment at five years follow-up. Median time to remission was 4 weeks (range :1-56 weeks). Data of 45 patients were available for relapse out of which 18 patients relapsed (40%). Adverse effects included infusion reactions in 16 patients, would infection, fever in one patient each and two deaths. Age, sex, duration of disease, timing of treatment (early vs late), concomitant steroids, and azathioprine treatment were not statistically significant predictors of remission and relapse in this study($P>0.05$).

Conclusion:

Rituximab therapy is safe and efficacious in majority of patients with immunobullous diseases. Age, sex, duration of disease, early /late treatment, concomitant treatment with steroid and azathioprine were not predictors of relapse or remission in this study.

Abstract N°: 6676

Characterizing the effect of clobetasol treatment on the oncogenic and immune signalling pathways in patients with vulvar lichen sclerosus.

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Introduction & Objectives

Little is known about the mechanistic effect of clobetasol in the treatment of vulvar lichen sclerosus (vLS). vLS is a chronical inflammatory dermatitis with a predilection for the anogenital area, vLS is clinically diagnosed and has a 4-6.7% risk for progression into vulvar cancer (1). The aim of this study was to determine the effect of clobetasol treatment in vLS patients. More precise, we compared inflammatory and oncologic pathways in vLS before and after treatment with clobetasol.

Materials & Methods

The study consisted of an observational and an interventional part in 10 vLS patients. Inclusion criteria were women aged 25-95 years with a body mass index <30 kg/m2, in general good health, and with a clinical confirmed vLS diagnosis. In the observational part, a 4 mm non-lesional and lesional vulvar biopsy were taken. Seven days later, during the interventional part, the patients applied clobetasol ointment 0.05% on the vulvar lesional area for 28 days once daily, after which two 4 mm lesional vulvar biopsies were taken. Based on patient-reported and clinical characteristics, severity of the vLS was determined pre- and postdose using the Günthert score (2). Formalin-fixed paraffin-embedded tissue sections were stained with hematoxylin and eosin for histopathological characteristics of vLS and for HPV genotyping. Immunohistochemistry (IHC) was performed for p16INK4a, p53 and PD-L1 expression. Bulk RNA was isolated from snap-frozen biopsies, quality checked using Agilent Bioanalyzer and subjected to the PanCancer IO360 and Cancer Pathway nCounter panel (Nanostring, Bruker). After quality control and normalization, gene expression, cell type profiling and pathway analysis were performed.

Results

In 3 out of 10 vLS patients, histopathological characteristics of vLS were seen. At baseline, all vLS biopsies were HPV negative. IHC staining revealed p16INK4a positive cells in 4 vLS patients, PD-L1 positivity in 3 vLS patients and wildtype p53 expression in 7 vLS patients. Clobetasol treatment compliance was confirmed for all patients. After treatment, one vLS biopsy was HPV positive and PD-L1 staining became absent in all vLS patients. Moreover, there was a decrease on average Günthert score from 5.0 to 3.7 ($p = 0.0071$) after treatment that indicates a reduction in vLS severity. The RNA transcriptomic data of both nCounter panels resulted in 39 overlapping genes and a total of 1441 non-shared genes. After adjustment for multiple testing, differentially expressed genes (DEGs) were identified between the different biopsies. Compared to non-lesional, the lesional biopsies resulted in 357 DEGs predose and 392 DEGs after treatment. Compared to predose, clobetasol treatment resulted in 148 DEGs in lesional vLS biopsies. Upregulated DEGs were associated with myeloid cells, wnt signaling, matrix remodeling, whereas the downregulated DEGs were mainly involved in antigen presentation, cytokine and chemokine signaling. Analysis of the cell type scores showed lower infiltration of cytotoxic cells and exhausted CD8+ T cells

and dendritic cells after treatment, whereas an increase in macrophages was observed compared to baseline vLS biopsies.

Conclusion

Our data on clobetasol treated vLS clearly indicate that clobetasol treatment inhibits inflammation via the downregulation of antigen presenting pathways and an influx of macrophages. Validation of the effect on immune cells in vLS sections at the protein level is required to confirm these findings.

Abstract N°: 6682

Mechanistic Study of YKL-40-Mediated Mast Cell Activation via the NLRP3 Inflammasome Signaling Pathway

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Introduction & Objectives:

To investigate the expression and role of YKL-40 in mast cell activation, its effects on inflammatory cytokine release and the NLRP3 signaling pathway, and to validate its role in chronic spontaneous urticaria (CSU) using an NLRP3 inhibitor.

Materials & Methods:

Human mast cell line HMC-1 was used in a four-part study. In Part I, cells were divided into control and urticaria model groups (stimulated with 25 ng/ml PMA and 1 μg/ml A23187). YKL-40 and NLRP3 inflammasome-related proteins were assessed by RT-qPCR and Western blot (WB). In Parts II and III, control, model, YKL-40 overexpression, and silencing groups were established via lentiviral transfection and RNA interference. After PMA/A23187 stimulation, expression of tyrosine kinases (Lyn, Fyn, Syk) and cytokines (IL-4, IL-6, IL-17, TNF-α, IFN-γ) were measured. In Part III, NLRP3, Caspase-1, and IL-1β expression were evaluated. Part IV included a group treated with an NLRP3 inhibitor on top of YKL-40 overexpression. Expression of tyrosine kinases, cytokines, and NLRP3-related proteins was analyzed.

Results:

In Part I, YKL-40 and NLRP3, Caspase-1, IL-1β expression were significantly increased in the model group ($P<0.01$). In Part II, YKL-40 overexpression led to increased expression of Lyn, Fyn, Syk and cytokines (IL-4, IL-6, IL-17, TNF-α) ($P<0.01$ or $P<0.001$), while silencing had the opposite effect. In Part III, overexpression of YKL-40 enhanced NLRP3, Caspase-1, and IL-1β expression ($P<0.05$–0.001); silencing reduced their expression ($P<0.05$–0.01). In Part IV, the NLRP3 inhibitor reversed YKL-40-mediated increases in tyrosine kinases and cytokines ($P<0.01$), restoring levels to that of the model group. IFN-γ was undetectable in all groups.

Conclusion:

YKL-40 is upregulated during mast cell activation and promotes inflammatory responses via the NLRP3 inflammasome pathway. Inhibition of NLRP3 attenuates these effects, indicating YKL-40/NLRP3 signaling as a key mechanism in CSU pathogenesis and a potential therapeutic target.

Abstract N°: 6683

Cutaneous Sarcoidosis and Thyroid Tuberculosis: A Coincidental Association or Linked Diseases?

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[2]Mohammed V Military Training hospital, dermatology, rabat, Morocco

Introduction & Objectives:

Sarcoidosis and tuberculosis are chronic granulomatous diseases that share clinical and histological similarities. Distinguishing between these two conditions can be complex, especially when they occur sequentially in the same patient. Cases of coexisting sarcoidosis and tuberculosis have been reported, but remain rare. We present a case of cutaneous sarcoidosis with mediastinal-pulmonary involvement, followed by thyroid tuberculosis after one year of disease progression.

Materials & Methods:

A 56-year-old woman with a history of antiphospholipid syndrome on anticoagulant therapy presented with progressive bilateral supra-eyebrow plaques. Physical examination revealed erythematous-violaceous, non-pruritic plaques with telangiectasia. Dermoscopy showed homogeneous yellow-orange lesions with branched vascular patterns. Skin biopsy demonstrated non-necrotizing tuberculoid granulomatous dermatitis, consistent with sarcoidosis. Staging workup confirmed stage II mediastino-pulmonary sarcoidosis. The patient was treated with systemic corticosteroids and intralesional steroid injections. One year later, she developed cervical pain, and further investigations identified a TIRADS V thyroid nodule. Histopathological examination confirmed thyroid tuberculosis

Results:

The patient was treated with systemic and intralesional corticosteroid therapy for pulmonary sarcoidosis with cutaneous involvement, as well as with antituberculous treatment for tuberculosis, with good clinical improvement.

Conclusion:

The coexistence of tuberculosis and sarcoidosis, two granulomatous diseases, poses significant diagnostic challenges, especially in tuberculosis-endemic regions. Although rare, their association warrants heightened clinical vigilance to ensure timely diagnosis and management.

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Abstract N°: 6729

New onset recalcitrant pemphigus vulgaris following coronavirus vaccination

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Introduction & Objectives:

Pemphigus represents a group of autoimmune blistering dermatoses primarily characterized by the formation of flaccid bullae and mucocutaneous erosions. These manifestations arise due to the immune-mediated disruption of desmosomal adhesion within the epidermis, typically driven by circulating autoantibodies against desmoglein (Dsg)1 and/or Dsg3. Numerous exogenous factors, including pharmacological agents and immunizations, have been implicated in the initiation or exacerbation of pemphigus. We report a case of pemphigus vulgaris (PV) following the first administration of a SARS-CoV-2 vaccine and subsequently exacerbating by a booster dose.

Materials & Methods:

A 73-year-old Caucasian man was admitted to the hospital with a new flare of a previously documenetd PV of three years duration. Flaccid blisters and painful, progressively enlarging erosions on the scalp, face, trunk and extremities resulted in vast denuded areas. Notably, the initial onset of the cutaneous symptoms, as reported by the patient, has occurred shortly after the first administration of the SARS-CoV-2 vaccine series and was markedly exacerbated following a booster dose several months later. Prior therapeutic regimens included prolonged use of systemic and topical corticosteroids, antibiotics, and an experimental neonatal Fc-receptor antagonist, without evident therapeutic benefit.

Results:

Routine laboratory investigations, including complete blood count and biochemical profile, revealed no clinically significant abnormalities. Histopathological evaluation demonstrated suprabasal acantholysis with the formation of intraepidermal blisters, as well as a sparse perivascular lymphocytic infiltrate in the superficial dermis. Direct immunofluorescence revealed intense intercellular deposition of IgG (+++) and complement C3 (++) throughout the epidermis. ELISA testing detected markedly elevated titers of anti-Dsg 1 and 3 autoantibodies. A concurrent mycological examination revealed oral candidiasis. Treatment with intravenous dexamethasone, ceftriaxone, oral nystatin, topical antiseptics and potent corticosteroids resulted in only partial clinical improvement. Given the refractory nature of the disease, the patient was evaluated as a candidate for rituximab therapy.

Conclusion:

The reported case adds to the growing body of evidence suggesting a potential temporal link between SARS-CoV-2 immunization and the onset or exacerbation of PV in predisposed individuals. Although causality remains controversial, several plausible immunopathogenic mechanisms, including molecular mimicry, polyclonal B cell activation, and innate immune system stimulation by vaccine-derived epitopes have been suggested. This may lead to a breakdown of tolerance against desmosomal proteins such as Dsg 1 and 3, thus acting as a PV trigger in genetically susceptible individuals.

Abstract N°: 6794

Blistering mystery – an unusual presentation of mucous membrane pemphigoid

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[2]The Mater Misericordiae University Hospital, Dublin, Ireland

Introduction & Objectives:

Mucous membrane pemphigoid (MMP) is a group of chronic, autoimmune subepithelial blistering diseases with a predominant mucosal involvement. We present a unique case of mucous membrane pemphigoid (MMP) presenting in in a 51-year-old gentleman with no previous medical history. This case of MMP was intriguing due to the acute onset of the symptoms presenting similarly to Steven-Johnson syndrome with extensive cutaneous involvement that is not usually seen in MMP.

Materials & Methods:

This patient presented to the emergency department with a blistering rash on his back, hands and feet with mucosal erosions and conjunctival erythema. The rash was associated with pain and a burning sensation. On examination there were small papulovesicular lesions on the palms of the hands and soles of the feet. In addition he had developed superficial mucosal erosions and vesiculobullous eruption around the mouth. There was also evidence of conjunctival injection. He had received oral antibiotics from the GP the previous week for a sore throat.

Following assessment in the emergency department there were multiple differential diagnosis considered with primary diagnosis including Stevens-Johnson syndrome, erythema multiforme major or an immunobullous disorder. We also considered infectious causes such as syphilis due to the palmoplantar distribution. The patient was admitted to hospital and treated with oral prednisolone 1mg/kg, intravenous acyclovir, prednisol mouthwash and dermovate ointment topical.

Within 24 hours following admission the patient developed worsening eye pain and burning urethral pain. There was progression of the blisters and he began to develop large tense bullae on the feet, hands and toes which resulted in degloving of his hands and feet. The scaly patches on his back and chest began to develop small bullae with a string of pearl appearance. Treatment was initiated with dapsone 50mg, doxycycline 100mg twice daily, prednisolone 1mg/kg continued along with topical treatments. At this point, the patient required morphine based analgesia. He required assistance with self-care and was unable to mobilise due to the severity of the skin loss/

Results: Histology was reviewed within the first 24 hours of admission due to the severity of the mucosal involvement and the clinical concern for Stevens-Johnson syndrome. Figure 1 (a) Skin biopsy revealed skin with a subepidermal split containing numerous neutrophils and occasional eosinophils with secondary epidermal necrosis and regenerative changes. There is moderate superficial perivascular lymphocytic infiltration with rare eosinophils. Importantly, there were no necrotic keratinocytes noted on the biopsy.

Direct immunofluorescence (IF) microscopy showed linear deposits of IgG and C3 at the DEJ. Serum autoantibodies localized to the base by indirect IF microscopy on salt-split skin but no reactivity with BP180 and BP230 was detected by various ELISA and immunoblotting analyses.

Conclusion:

These findings are in keeping with a diagnosis of mucous membrane pemphigoid. This patient had a prolonged hospital admission as treatment was challenging. The combined use of oral prednisolone, dapsone 100mg, and rituximab were ultimately successful in controlling his disease.

Abstract N°: 6885

Successful Use of Dupilumab in the Management of Recalcitrant Pemphigoid Gestationis: A Case Report

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Introduction & Objectives: Pemphigoid gestationis (PG) is a rare autoimmune blistering disease of pregnancy, typically emerging in the second or third trimester. It poses risks to both mother and fetus, including preterm delivery and intrauterine growth restriction. Standard treatment with systemic corticosteroids can be limited by safety concerns. Dupilumab, an IL-4Rα antagonist approved for atopic dermatitis, has shown efficacy in bullous pemphigoid. We present a case of steroid-refractory PG successfully managed with dupilumab.

Materials & Methods: A 31-year-old primigravida at 28 weeks gestation developed intensely pruritic, erythematous, vesiculobullous lesions beginning on the abdomen. Diagnosis was confirmed by clinical features, histopathology, direct immunofluorescence showing continuous C3 deposition along the basement membrane zone, and elevated BP180 autoantibodies via ELISA.

Results: Initial treatment with oral prednisolone (40 mg/day) and topical corticosteroids resulted in initial improvement, but symptoms recurred during tapering. To minimize fetal exposure to high-dose steroids, dupilumab was introduced at 34 weeks gestation (600 mg loading dose, then 300 mg every two weeks). Corticosteroids were tapered concurrently. Marked clinical improvement followed, with resolution of lesions and pruritus. The patient delivered a healthy infant at 37 weeks with no neonatal complications. Dupilumab was continued for four weeks postpartum to reduce relapse risk, with sustained remission and no adverse events.

Conclusion: This case supports dupilumab as a potentially safe and effective steroid-sparing option for refractory PG during pregnancy. Its role in modulating type 2 inflammation may offer therapeutic benefit where conventional therapies are inadequate or contraindicated. While current data are limited, emerging reports suggest dupilumab may be considered in select PG cases under close multidisciplinary supervision. Further studies are needed to establish safety, dosing, and long-term outcomes in this population.

Abstract N°: 6900

Alopecia Areata: Epidemiological, Clinical, Dermoscopic, Therapeutic, and Prognostic Aspects

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Introduction & Objectives:

Alopecia areata is a non-scarring autoimmune disease that presents as alopecic patches on the scalp or other hair-bearing areas. It results from an immune-mediated attack on hair follicles by cytotoxic T lymphocytes, with a genetic predisposition playing a key role. The condition can affect individuals at any age, and its course is chronic, relapsing, and unpredictable, making effective management particularly challenging.

This study aimed to assess the epidemiological and clinical profile, dermoscopic features, prognostic factors, and therapeutic outcomes in patients with alopecia areata.

Materials & Methods:

This prospective, descriptive, and analytical study was conducted from November 2019 to April 2025 in the Department of Dermatology at our University Hospital Center. A total of 167 patients diagnosed with alopecia areata were included.

Results:

There was a female predominance, with a male-to-female ratio of 0.46. The mean age at onset was 30.64 years. A family history of alopecia areata was noted in 13% of patients. A personal history of atopy was found in 25% of cases, and a family history of atopy in 10%. Additionally, 13% of patients had a history of autoimmune diseases, including thyroiditis, vitiligo, psoriasis, diabetes, and rheumatoid arthritis. The patchy form of alopecia areata was the most frequently observed. Dermoscopic features were diverse, most commonly including yellow dots (83%), black dots (57%), vellus hairs (46%), exclamation mark hairs (41%), circle hairs (15%), cadaverized hairs, and broken hairs. Alopecia severity was greater among female patients. Disease extent was associated with early onset, longer disease duration, and the presence of nail abnormalities. Therapeutic choices depended on patient age, disease extent, and disease duration. When involvement was less than 40%, a favorable therapeutic response was observed in over 50% of cases. In cases with more than 40% involvement, therapeutic failure was reported in over 70% of patients. Among patients with alopecia totalis, 45% achieved a cosmetically acceptable response, and 30% experienced complete regrowth.

Conclusion:

Alopecia areata remains a complex and unpredictable autoimmune disorder with significant clinical variability. This study highlights a female predominance and a relatively young age of onset, with the patchy form being the most prevalent. Dermoscopic examination proved valuable in identifying characteristic features, aiding in diagnosis and evaluation. The severity and extent of the disease were strongly associated with early onset, prolonged duration, and nail involvement. Despite the therapeutic challenges, especially in extensive forms, favorable outcomes were achieved in cases with limited involvement. These findings underscore the importance of early diagnosis, individualized treatment strategies, and long-term follow-up to improve prognostic outcomes in patients with alopecia areata.

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Abstract N°: 6914

Blister Fluid Analysis: Identifying Key Biomarkers for Pemphigus Severity

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Introduction & Objectives:

Pemphigus is a serious autoimmune disorder that can be life-threatening, leading to the formation of blisters within the epidermis on the skin and in the oral mucosa. Blister fluid contains antibodies, chemokines, cytokines, mRNA, exosomes, and other components, making it a potential source for biomarkers that could indicate the severity, activity, and treatment response of blistering diseases. The aim of our study was to assess chemokine and cytokine profiles in both serum and blister fluid from patients with pemphigus.

Materials & Methods:

We conducted a cohort-study involving 43 patients diagnosed with pemphigus vulgaris, consisting of 33 women and 11 men. The mean age of patients was 55 years. Disease severity was assessed by PDAI index. All the patients underwent collection of blister fluid (0,2-0,5 ml) and serum (5 ml) using a sterile syringe. The samples were subsequently placed in Eppendorf tubes and stored at -800C prior to treatment. The concentrations of cytokines, including TNF-α, IL-4, IL-15, IL-10, granulysin and chemokines CXCL8 and CCL11, were analyzed by Cytometric Bead Array (CBA) and enzyme-linked immunosorbent assay (ELISA) methods.

Results:

IL-15 (14.5 (13.25; 25.5 pg/ml)), IL-10 (Median - 66 pg/ml), CCL11 (Median - 94.5 pg/ml), and CXCL8 (77.5 pg/ml) were statistically significantly higher in patients with severe pemphigus compared to those with mild and moderate condition, respectively ($p<0.0306$, $p<0.0102$, $p<0.0122$, $p<0.0007$). Notably, that in patients with moderate form, the serum levels of IL-4 were significantly elevated at (9 pg/ml (8; 11 pg/ml)) compared to those with mild (6 pg/ml (5; 7 pg/ml)) and severe pemphigus (7.5 pg/ml (6.25; 8 pg/ml)) respectively ($p<0.0020$). In blister fluid from patients with severe pemphigus vulgaris, the levels of TNF-α (median - 91.5 pg/ml), CCL11 (median - 192.5 pg/ml), and granulysin (10739.5 ng/ml (10391.25; 15930.5 ng/ml) ($p<0.0028$, $p<0.0027$, $p<0.0005$) were significantly higher compared to mild and moderate forms. In comparison, patients with mild and moderate pemphigus exhibited statistically significant increases in IL-15 and IL-10 levels, respectively ($p<0.0082$, $p<0.0003$).

Conclusion:

The significant disparity in chemokine levels between serum and blister fluid suggests that these molecules are likely produced locally by cells at the site of blister formation, rather than being influenced by systemic circulation. Cytokines and chemokines can bind to cell receptors which commonly results in lower concentrations in serum compared to those found in blister fluid. We found that higher levels of TNF-α, CCL11 and granulysin in blister fluid correlated with the disease severity. Thus, it may serve as a valuable source of potential biomarkers that could predict both the disease progression.

Abstract N°: 6955

Superficial Pemphigus Associated with Systemic Lupus Erythematosus and Primary Biliary Cholangitis: A Case Report

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Introduction & Objectives:

Superficial pemphigus (SP) is an autoimmune bullous dermatosis. Its association with other autoimmune diseases has been described in the literature. We report a case of SP associated with systemic lupus erythematosus (SLE).

Materials & Methods:

Case Report:

A 43-year-old man was being followed for moderate SP, treated with azathioprine 100 mg/day and prednisone at 1 mg/kg/day. He initially responded well, but relapsed during corticosteroid tapering. Clinical examination revealed fixed urticarial lesions on the trunk, a malar erythema (“butterfly rash”) with photosensitivity, chilblain on the left great toe, and patchy alopecia. Laboratory tests showed lymphopenia (250/mm^3), mixed hepatic cytolysis and cholestasis (gamma-GT 353 IU/L, ALT 4× normal), and 24-hour proteinuria of 2.77 g/24h. Immunological workup revealed positive antinuclear antibodies (ANA) at 1:320, along with anti-gp120 and anti-LKM1 antibodies. A diagnosis of primary biliary cholangitis (PBC) was made. Renal biopsy was consistent with lupus nephritis, stage III. Histological analysis of the fixed urticarial skin lesions revealed leukocytoclastic vasculitis. The diagnosis of SLE with renal, hematological, and cutaneous involvement was established. The patient received corticosteroid pulse therapy and was referred to nephrology for possible treatment with mycophenolate mofetil.

Results:

Discussion:

The coexistence of SP and SLE is rare. Serological studies show that about one-third of patients with SP have positive ANA, often with a homogeneous pattern associated with active SLE. A recent genome-wide association study identified new CD4+ T-cell pathways linking pemphigus and SLE, and highlighted IRF8 and STAT1 as key regulatory genes, supporting the biological plausibility of this clinical association. In our patient, three autoimmune diseases coexisted—SP, SLE, and PBC—supporting the theory of autoimmune clustering.

Conclusion:

We report a rare case of superficial pemphigus associated with systemic lupus erythematosus and primary biliary cholangitis. Further research is needed to clarify the molecular basis of this association.

Abstract N°: 7005

Atypical Bullous Pemphigoid Following SARS-CoV-2 Infection: A Case Report

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Introduction & Objectives: Bullous pemphigoid (BP), the most common subepidermal autoimmune blistering disease, typically affects the elderly. Atypical presentations of BP, possibly triggered by external factors such as infections, are increasingly being recognized.

Materials & Methods: We report a case of BP with unusual clinical features temporally associated with SARS-CoV-2 infection, highlighting its potential role in autoimmune dysregulation.

Results: A 55-year-old man with no significant medical history presented with a generalized bullous

eruption and concurrent flu-like symptoms of two weeks' duration. Cutaneous examination

revealed tense bullae, erosions, and dyschromic scarring predominantly involving the head, neck, and palmoplantar regions, the latter showing a dyshidrosiform pattern. The Nikolsky sign was negative. Oral mucosal involvement included erosions and blisters. Laboratory investigations revealed peripheral eosinophilia (5,310 cells/mm^3). Histopathology confirmed subepidermal blistering with eosinophil-rich infiltrates. Direct immunofluorescence showed linear IgG and C3 deposition along the basement membrane zone. Systemic corticosteroid therapy (prednisone 0.75 mg/kg/day) was initiated. Days later, the patient developed a persistent dry cough and dyspnea, prompting SARS-CoV-2 RT-PCR testing, which was positive. The infection was treated supportively. Cutaneous disease activity resolved within six weeks, allowing corticosteroids to be tapered over 10 months. There were no relapses during 12 months of follow-up.

Conclusion: This case illustrates an atypical BP phenotype characterized by younger age of onset, mucosal involvement, dyshidrosiform palmoplantar distribution, and predominant head/neck involvement. The temporal association with SARS-CoV-2 infection raises the question of viral triggering of autoimmunity; although causality remains unproven, several factors support this hypothesis. The bullous eruption preceded the confirmed COVID-19 diagnosis, consistent with reported delays in autoimmune manifestations after infection. SARS-CoV-2 may induce molecular mimicry (shared epitopes between BP180 and viral proteins) or bystander T-cell activation, as proposed in recent literature on COVID-19-associated autoimmune phenomena. The rapid disease control with corticosteroids and the sustained remission suggest transient immune activation rather than classical idiopathic BP.

Abstract N°: 7036

Drug-Induced Bullous Pemphigoid: Diagnostic Challenges and Therapeutic Insights — A Literature Review

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Introduction & Objectives:

Drug-induced bullous pemphigoid (DIBP) is a rare autoimmune blistering skin disorder that develops in response to certain medications. Its clinical and histopathological similarity to idiopathic bullous pemphigoid complicates timely diagnosis. In recent years, the number of reported cases has increased, along with an expanding list of potential drug triggers. This trend highlights the need for updated, systematized data on DIBP.

Materials & Methods:

A literature review was conducted using PubMed, Scopus, ScienceDirect, and Elsevier databases covering the period from 2019 to 2024. Included sources were original studies, systematic reviews, clinical guidelines, and case reports confirming an association between drug intake and pemphigoid onset. Clinical presentation, diagnostic modalities, proposed pathogenic mechanisms, and therapeutic strategies were analyzed. Particular attention was given to the clinical course following drug discontinuation.

Results:

The most commonly implicated medications include diuretics (e.g., furosemide), antibiotics (penicillins, cephalosporins), ACE inhibitors, anticoagulants, and psychotropic agents. Clinically, the disease manifests as tense bullae, erythematous plaques, and severe pruritus, primarily on the trunk and extremities. Histopathology typically reveals a subepidermal blister with eosinophilic infiltration. Direct immunofluorescence demonstrates linear IgG and/or C3 deposition along the basement membrane zone. Diagnostic challenges arise in cases of polypharmacy and delayed onset. In most patients, drug withdrawal leads to resolution of lesions; however, systemic therapy is often required, ranging from corticosteroids to immunosuppressants and, in refractory cases, rituximab.

Conclusion:

DIBP is a clinically relevant but frequently underrecognized variant of bullous dermatoses. Accurate diagnosis requires a comprehensive approach including clinical assessment, histopathology, immunofluorescence studies, and detailed drug history. Prompt discontinuation of the offending agent remains the cornerstone of effective management. Given the lack of standardized diagnostic criteria and the increasing number of reported cases, further research is needed to unify diagnostic and treatment strategies. Systematic reviews such as this one may provide a foundation for developing future clinical guidelines.

Abstract N°: 7049

Lichen Sclerosus: Clinical, Laboratory, and Histopathological Evaluation of 168 Patients

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Introduction & Objectives: Lichen sclerosus is a chronic dermatosis characterized by inflammation and epidermal atrophy. Its prevalence is higher in women. In women, two peak periods are observed, between the ages of 8-13 and 40-59. Although the exact etiopathogenesis has not been fully elucidated, it is thought that genetic, immunological, and hormonal factors, as well as external factors such as trauma and infection, contribute to its development. In cases of clinical suspicion, a detailed investigation for accompanying autoimmune diseases is recommended.

Materials & Methods: : This study included 168 patients who were clinically and histopathologically diagnosed with lichen sclerosus between 2008 and 2024. The patients' data were accessed retrospectively. In addition to clinical and demographic data, patients were evaluated using autoantibody and thyroid function tests.

Results: A total of 168 patients were evaluated in our study, consisting of 150 women and 18 men. Of these patients, 27 (16%) belonged to the pediatric group. The mean age of the patients was 46±20 years (ranging from 6 to 91). Among the patients, 77 (45.8%) had extragenital lesions, 62 (36.9%) had genital lesions, and 29 (17.3%) had both genital and extragenital lesions. The most common sites of extragenital lesions were the trunk (29.5%), lower extremities (13.6%), and intertriginous areas (12.4%).

During the initial examination of the 91 patients with genital lesions, 10 patients (11%) had erosions, 2 patients (2.1%) had ulcers, and 1 patient (1%) had fissures. As complications, 17 patients (18.6%) had labial resorption, 2 (2.1%) had labial fusion, and 1 patient (1%) was diagnosed with squamous cell carcinoma following a biopsy. The average time between the onset of lesions and hospital admission was 2.5±4 years (ranging from 1 month to 30 years), while the average time to benefit from treatment was 3.5±2.5 months.

A total of 59 patients (35.1%) had at least one autoimmune disease, with the most commonly reported conditions being hypothyroidism (17.2%), morphea (8.9%), and vitiligo (4.7%). Among the 15 patients with concomitant morphea, 14 (93%) were women, showing a significant gender difference ($p<0.05$). In autoantibody test results, elevated anti-thyroid peroxidase (TPO) levels were found in 20 of 60 patients (33.3%), elevated anti-thyroglobulin (TG) levels were found in 9 of 40 patients (18.4%), and positive antinuclear antibody (ANA) results were found in 13 of 61 patients (21%).

Conclusion: Our study indicates a higher rate of extragenital lichen sclerosus in our clinic compared to data in the literature. This finding emphasizes the importance of conducting full-body dermatological examinations. Additionally, the presence of thyroid disease and positive autoantibodies in our patients highlights the necessity of thoroughly evaluating individuals with lichen sclerosus for autoimmune diseases. The complication risk in lichen sclerosus patients is relatively high, around 20%. Patients are at risk for vulvar anatomical abnormalities and the development of squamous cell carcinoma. Therefore, initiating early treatment is crucial for improving the quality of life and preventing serious complications.

Abstract N°: 7134

Lupus tumidus following allergen immunotherapy: A rare injection-site reaction.

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Introduction & Objectives:

Emerging evidence suggests that environmental triggers, including certain medications and vaccines, may contribute not only to the onset of systemic lupus erythematosus, but also to cutaneous variants such as cutaneous lupus erythematosus (CLE). Although rare, cases of lupus flares or new onset following immunisation have been documented. When skin involvement is reported, it typically presents as subacute CLE, with lesions predominating on the trunk and limbs.

Our aim is to report a rare case of lupus tumidus potentially triggered by long-term allergen-specific immunotherapy (ASIT) in a patient with antiphospholipid antibody (APL) positivity, highlighting histopathological and molecular correlations.

Materials & Methods:

A 28-year-old man was evaluated for a two-year history of intermittently pruritic skin lesions on the posterior aspect of both arms, corresponding to prior ASIT injection sites. Clinical examination revealed well-defined, annular, erythematous, non-scaly plaque-nodular lesions on the right arm, and smaller lesions on the left. Two temporally separated punch biopsies were performed. Histopathology showed perivascular and periadnexal lymphoplasmacytic infiltrates in absence of epidermal involvement; dermal mucin deposition (Alcian blue-positive) and folliculotropism of T-cells. Molecular analysis demonstrated polyclonal Ig/TCR gene rearrangements, ruling out cutaneous lymphoma. Laboratory tests revealed fluctuating APL and anti-β2GP1 antibody titres, with no evidence of systemic lupus erythematosus.

Results:

A diagnosis of lupus tumidus was established. The patient commenced oral hydroxychloroquine (200 mg/day) and continued for nine months with limited improvement, ultimately leading to self-discontinuation. He is currently managed with topical clobetasol propionate and has been referred to haematology due to persistent triple positivity for APL. Thromboprophylaxis with low-dose acetylsalicylic acid (100 mg/day) has been initiated.

Conclusion:

Lupus tumidus has previously been associated with pharmacological agents such as Bortezomib and anti-TNF-α therapies. There have been reported cases of either new-onset or relapse of previously stable lupus erythematosus following administration of RNA- or adenovirus-based COVID-19 vaccines, including a documented instance of progression from cutaneous to systemic LE. Our case suggests that ASIT may similarly act as an immunological trigger in susceptible individuals, potentially via shared pathways with vaccine-induced cutaneous lupus. Nonetheless, it is crucial to emphasise that vaccination remains a safe, effective, and beneficial intervention for patients with lupus, including those with systemic forms.

Abstract N°: 7177

Discoid Lupus Erythematosus and Lichen Planus Overlap Syndrome: A Case Report Highlighting the Role of Dermoscopy

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Introduction & Objectives:

Lupus erythematosus (LE) and lichen planus (LP) are two distinct and well established dermatoses which occasionally can occur as an overlap syndrome. it is characterized by the combination of clinical, histopathological and/or immunopathological features of both diseases in the same patient and/or at the same lesion of one patient. The majority of cases occurs between the ages of 25 to 45 with a slight female predominance.

Materials & Methods:

We report a case of overlap syndrome involving discoid lupus erythematosus and mucosal lichen planus, in which the diagnosis was guided by dermoscopic findings

Results:

We report the case of a 44-year-old female who presented with spontaneously occurring erythematous scaly plaques on the face, evolving over the past year. She had no prior medical history and exhibited no systemic symptoms. Clinical examination revealed three well-defined erythematous scaly plaques located on the upper lip, nose, and left pretragal area. Dermoscopic evaluation showed yellowish and white scales, keratotic plugs, rosettes, chrysalis structures, and hairpin vessels. Additionally, she exhibited erosive cheilitis, with dermoscopic findings including central erosions, Wickham striae, rosettes, punctate, linear and hairpin vessels, and peripheral pigmentation.

Histopathological examination of two cutaneous biopsies was performed. The first biopsy, taken from the upper lip plaque, confirmed discoid lupus erythematosus (DLE) with direct immunofluorescence (DIF) revealing a junctional deposition of C3 and IgG. The second biopsy, obtained from the erosive cheilitis lesion, was consistent with lichen planus (LP).

Based on these findings, a diagnosis of overlap syndrome of lupus and lichen planus was established. The patient was treated with topical tacrolimus, leading to significant clinical improvement.

Conclusion:

The clinical presentation of LE/LP overlap syndrome varies widely, ranging from classic violaceous papules of LP to erythematous scaly plaques seen in discoid LE. Some patients may exhibit mucosal involvement, like our patient, where distinguishing LP from LE can be difficult due to similar atrophic and ulcerative changes. Nail involvement, seen in both diseases, can present as longitudinal ridging, onychodystrophy, or pterygium formation. Dermoscopy can be a valuable tool in differentiating overlapping features, revealing Wickham striae in LP and telangiectatic vessels in LE. Histopathology remains the gold standard for diagnosis, showing a lichenoid inflammatory infiltrate with basal layer degeneration, vacuolar interface dermatitis, and colloid bodies.

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Abstract N°: 7184

Pemphigus : étude épidémiologique, clinique et thérapeutique rétrospective (2017-2025)

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Introduction & Objectives:

Pemphigus is a rare autoimmune blistering disease, divided into superficial and deep forms. Despite advances in understanding its pathogenesis, the disease's clinical presentation remains heterogeneous, posing diagnostic challenges. This study aims to describe the epidemiological, clinical, and therapeutic profiles of pemphigus in our clinical setting.

Materials & Methods:

We conducted a retrospective and prospective case series over an 8-year period (May 2017 – February 2025), including 73 patients hospitalized for pemphigus at the University Hospital of Marrakech. Diagnosis was based on clinical, histological, and immunological criteria.

Results:

A total of 73 patients were included over the 8-year study period. There was a marked female predominance, with a sex ratio of 0.62. The mean age was ≥60 years, ranging from 20 to 85 years, reflecting the disease's prevalence in middle-aged and older adults. Most patients (48 of 73) came from urban areas, and nearly 80% had a low socioeconomic status, potentially contributing to diagnostic delay and limited access to specialized care.

Clinically, the disease most often began with mucosal involvement, particularly of the oral mucosa, seen in 23 patients. The most common symptoms were pruritus (61 patients), pain (39), dysphagia (19), anorexia (22), and fatigue (25). Nikolsky's sign was positive in 57 patients, indicating epidermal fragility. Nail and hair involvement, especially of the scalp, was noted in 51 cases. Disease severity ranged from mild (26 patients) to moderate (36) and severe (11), with cutaneous involvement varying from <5% to >50% of body surface area.

Histopathological analysis confirmed the diagnosis in all cases. Two main forms were identified: deep pemphigus (61.6%) and superficial pemphigus (38.4%). Direct immunofluorescence on skin or mucosal biopsies showed clear intercellular staining within the epidermis, consistent with autoantibodies targeting desmosomal proteins.

Most patients received systemic corticosteroids, mainly oral prednisone. This was commonly combined with immunosuppressants: azathioprine (41 patients) and cyclophosphamide (15 patients). Rituximab, a targeted anti-CD20 monoclonal antibody recommended as first-line treatment in moderate to severe cases according to international guidelines, was administered to 14 patients with refractory or severe disease. Other treatments included dapsone (4 patients) and intravenous methylprednisolone pulses (3 patients) in acute or severe presentations.

The clinical outcome was favorable in many cases: 30 patients achieved complete healing within three months. However, complications were common. Over 40 patients experienced superinfections, mainly bacterial or herpetic. Malnutrition and dehydration were also frequent, particularly in extensive or severe mucosal forms. In the long term, 32 patients experienced relapses, often linked to poor adherence or premature treatment discontinuation. Two patients died from severe complications, and two others were lost to follow-up.

Conclusion:

Pemphigus, more prevalent in the Maghreb with a predominance of deep forms, remains a severe condition associated with serious complications such as infections and relapses. Although rituximab is internationally recommended as first-line therapy, its limited accessibility in our setting underscores the importance of early detection and individualized care strategies to improve patient outcomes.

Abstract N°: 7218

When lupus isn't lupus: unmasking pemphigus erythematosus

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Introduction & Objectives:

Pemphigus erythematosus (PE) is a rare autoimmune bullous disease that presents with overlapping features of both lupus erythematosus and pemphigus foliaceus (PF). We report a case of PE, illustrating its clinical presentation, diagnostic challenges and therapeutic approach.

Materials & Methods:

A 63-year-old man presented with a 5-month history of pruritic, scaly plaques initially localized to the scalp, progressively spreading to the face and upper trunk. Prior outpatient treatments for presumed psoriasis, seborrheic dermatitis, superficial fungal infection were ineffective. Topical 5-fluorouracil (5-FU) for suspected actinic keratosis led to aggravation of the facial lesions. He reported a major stressor related to the recent death of his wife from a rare genetic disease, later identified also in their daughter. Examination revealed a symmetric eruption consisting of erythematous-violaceous, well-demarcated, round-to-oval plaques and patches with surface erosions, yellow crusts and scaling. The lesions followed a malar distribution and also involved the seborrheic areas of the trunk and the scalp. Nikolsky's sign was positive on the trunk. Mucosae were spared. General exam was unremarkable. A clinical diagnosis of PE was considered. Differentials included bullous lupus erythematosus, pemphigus vulgaris (PV) and paraneoplastic pemphigus. Skin biopsy was performed for histopathological (HP) examination and direct immunofluorescence (DIF), along with a work-up for systemic lupus erythematosus (SLE), autoimmune bullous diseases and malignancy.

Results:

HP revealed superficial epidermal acantholysis. DIF showed intercellular IgG and C3 deposition throughout the epidermis, with no immune deposits at the basement membrane zone (BMZ). Serology revealed high anti-desmoglein 1 (anti-Dsg1) and anti-desmoglein 3 (anti-Dsg3) antibodies titers, positive ANA and anti-dsDNA with no further SLE evidence. These findings supported the diagnosis of PE. High-dose systemic corticosteroids led to partial improvement, but relapse occurred on tapering. Azathioprine was added, inducing clinical remission. However, anti-Dsg1 antibodies remained elevated at 3 months. Given their predictive value for cutaneous relapses, rituximab was initiated. Follow-up is planned 3 months post-rituximab.

Conclusion:

PE is a relatively benign and localized form of PF, but evolution to PV may occur. It does not meet SLE criteria, but often shows ANA positivity. Typically, it presents with seborrheic lesions on the face, resembling the lupus malar rash. Our patient presented all the clinical, HP, DIF and serological features of PE, except for BMZ immune deposition. This case illustrates the diagnostic challenges of early PE and prompts reflection on potential

aggravating or triggering factors, such as topical 5-FU and emotional stress. It also underlines the need to bear in mind the hypothesis of progression to PV, particularly in the context of elevated anti-Dsg3 titers.

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Abstract N°: 7233

Trichoscopy and nailfold capillaroscopy in dermatomyositis

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Introduction & Objectives:

Dermatomyositis is an idiopathic inflammatory myopathy characterized by a combination of muscle weakness and distinctive skin manifestations. It is a clinically heterogeneous condition that can be challenging to diagnose. The hallmark features include symmetrical proximal muscle weakness and characteristic skin rashes such as Gottron's papules, heliotrope rash and shawl sign. Scalp involvement is a common manifestation, with prevalence rates as high as 82%. The main hair-related findings include diffuse erythema and scaling, pruritus, burning sensation, non-scarring alopecia, poikiloderma, brittle or fragile hair. Trichoscopy may show perifolicular erythema and scaling, dilated and tortuous blood vessels, reduced hair density, broken and miniaturized hair, poikilodermatous background – especially in chronic cases, and absence of scarring, which helps differentiate from scarring alopecias. These findings are nonspecific but supportive when correlated with clinical and histopathological features. They also help in monitoring disease activity and treatment response. Likewise, nail manifestations are commonly observed. The most prevalent findings include increased vascular diameter, decreased vascularity, and microhemorrhages, which are often observed through nailfold capillaroscopy. They are indicative of microangiopathy and can correlate with disease activity and systemic involvement. Capillaroscopic patterns are characterized by elongated capillaries, avascular areas, disorganized vessel architecture, and tortuous and dilated capillaries. The objective of this report is to describe less common trichoscopic features and nail fold capillaroscopy in a case of dermatomyositis.

Case report:

Female, 46 years old, with Hashimoto's thyroiditis, started with arthralgia, symmetrical proximal muscle weakness, heliotrope rash, Gottron's papules and shawl sign. Autoantibodies were negative, except for anticardiolipin. Skin biopsy revealed interface dermatitis, hydropic degeneration of the basal cell layer, sparse perivascular lymphocytic infiltrate, with increased dermal mucin. Five months later, the patient presented with painful patchy alopecia. Scalp trichoscopy exhibited reduced hair density, abscence of scarring, bushy capillaries, perifollicular pigmentation and markedly interfollicular pigmentation. Capillaroscopy showed giant loops, hairpin capillaries and haemorrhages. The patient is now under one year treatment with hydroxychloroquine, methotrexate and prednisone, with significant improvement and complete hair regrowth, associated with oral minoxidil.

Conclusion:

Dermoscopy has been increasingly used in the assessment of inflammatory dermatoses, expanding its role beyond the traditional scope of pigmented lesions and skin cancer detection. It is a valuable technique in the clinical management of dermatomyositis, offering insights into disease activity, aiding diagnosis and potentially guiding therapeutic decisions.

Abstract N°: 7286

Bullous pemphigoid and hematologic malignancies: Clarifying the debatable association by a large-scale population-based study

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Introduction & Objectives:

The current literature is inconsistent regarding the association of bullous pemphigoid (BP) with hematologic malignancies (HM). In the current study, we aimed to assess the bidirectional association between BP and HM.

Materials & Methods:

A population-based retrospective cohort study was conducted to compare BP patients (n=3,924) with age-, sex- and ethnicity-matched control subjects (n=19,280) regarding the risk of 6 HMs: acute leukemia (AL), chronic leukemia (CL), Hodgkin lymphoma (HL), multiple myeloma (MM), non-Hodgkin lymphoma (NHL), and polycythemia vera (PV). A case-control design was additionally adopted to estimate the odds of BP in individuals with a preexisting diagnosis of each of the six HMs.

Results:

In the retrospective cohort design, the overall incidence rates of AL, CL, HL, MM, NHL, and PV in patients with BP were estimated at 4.3 (95% CI, 1.6-9.4), 27.3 (95% CI, 18.9-38.1), 7.7 (95% CI, 3.7-14.1), 35.1 (95% CI, 25.5-47.1), 27.7 (95% CI, 19.2-38.6), and 4.3 (95% CI, 1.6-9.5) cases per 1,000 person-years, respectively. Patients with BP did not experience an elevation in the risk of any of the six aforementioned HMs. The development of subsequent BP was not significantly associated with a history of any of the investigated HM, as evidenced by the case-control study design. A significant increase in the probability of subsequent BP was only observed among males with a history of NHL (OR, 1.43; 95% CI, 1.00-2.05).

Conclusion:

In conclusion, our study revealed that patients with BP do not experience an elevated risk of HM and that a history of HM is not associated with a significant increase in the odds of subsequent BP. Specific awareness should be given to the slight increase in the odds of subsequent BP among males with NHL. Overall, the current data do not lend weight to the performance of routine screening for HM among patients with BP.

Abstract N°: 7421

plasma cell storm in discoid lupus erythematosus: unveiling a rare reactive cutaneous plasmacytosis

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Introduction & Objectives:

Discoid lupus erythematosus (DLE) is a chronic cutaneous autoimmune condition characterized by localized inflammation and scarring. Although lymphocytic infiltrates are typical, the presence of reactive plasmacytosis is exceedingly rare and poorly documented. This report aims to highlight a unique case of DLE associated with a dense plasma cell-rich infiltrate, emphasizing the diagnostic implications and potential immunopathological connections.

Materials & Methods:

A 63-year-old female presented with a 7-year history of painful ulcerated plaques involving the scalp, face, oral mucosa, trunk, and lower limbs. No history of systemic autoimmune diseases or hematologic malignancy was reported. Skin biopsy revealed interface dermatitis with basal layer vacuolization, subepidermal clefting, dermal fibrosis, and a dense perivascular and periappendageal lymphoplasmacytic infiltrate. Mucin deposits were noted. Immunohistochemistry confirmed the presence of reactive plasma cells (CD79a+, Kappa+, Lambda+). Hematologic evaluation excluded clonal proliferation, confirming reactive cutaneous plasmacytosis.

Results:

The diagnosis of DLE with associated reactive cutaneous plasmacytosis was established based on clinical, histological, and immunophenotypic features. The patient was managed with hydroxychloroquine and topical calcineurin inhibitors, leading to progressive clinical improvement. The unusually rich plasma cell infiltrate raised awareness of underrecognized immunologic responses in DLE.

Conclusion:

This case underscores a previously unreported association between DLE and reactive cutaneous plasmacytosis, expanding the known histopathological spectrum of lupus erythematosus. Recognition of such findings is crucial to avoid misdiagnosis with plasma cell neoplasms. Further studies are warranted to elucidate the immunological mechanisms driving such exaggerated plasmacytic responses in chronic autoimmune inflammation.

Abstract N°: 7430

Cutaneous Myiasis Leading to Diagnosis of Pemphigus Vulgaris in a Young Adult Female

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Title: Title: Cutaneous Myiasis Leading to Diagnosis of Pemphigus Vulgaris in a Young Adult Female

Introduction & Objectives: Pemphigus vulgaris (PV) is an autoimmune intraepidermal blistering disorder caused by IgG autoantibodies against desmoglein 3 and, less frequently, desmoglein 1. While the classical presentation includes mucocutaneous erosions and flaccid bullae, atypical or masked presentations can complicate diagnosis. Cutaneous myiasis, a parasitic infestation by Diptera larvae, is rare and typically occurs in neglected wounds. Its coexistence with undiagnosed autoimmune skin disease is extremely uncommon. This case aims to highlight how a parasitic infestation may obscure or unmask an underlying immune-mediated dermatosis.

Materials & Methods: A 30-year-old female presented with a one-month history of a malodorous scalp wound infested with live maggots, alongside excoriated papules, erosions and crusts on the trunk and limbs. There was no history suggestive of mucosal involvement. Cutaneous myiasis was managed by oral ivermectin, manual extraction and surgical debridement. During evaluation, vesicular lesions were biopsied for histopathology and direct immunofluorescence (DIF); indirect immunofluorescence (IIF) was also performed.

Results: Histopathology revealed suprabasal acantholysis with sparse dermal lymphocytic and neutrophilic infiltration. DIF showed intercellular IgG and C3 deposits in a fishnet pattern; IIF confirmed circulating antibodies against desmoglein 1 and 3 consistent with the diagnosis of pemphigus vulgaris. The patient was initiated on systemic corticosteroids and azathioprine with subsequent resolution of skin lesions and healing of the scalp wound.

Conclusion: This case highlights an atypical presentation of pemphigus vulgaris, with cutaneous myiasis serving as the initial clinical manifestation. In resource-limited tropical regions, myiasis may mask underlying autoimmune blistering skin diseases. This case underscores the need for vigilance and thorough evaluation of new or unexplained skin lesions, particularly in patients with secondary infections or infestations, as these may obscure underlying autoimmune diseases requiring prompt recognition and treatment.

Abstract N°: 7521

Herpes zoster overlying malignant melanoma: a case of Wolf's isotopic response

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Introduction & Objectives:

Malignant melanoma (MM) and herpes zoster (HZ) represent two distinct entities, yet common mechanisms of their association may be suspected. MM is an aggressive malignant tumour originating from melanocytes. HZ is an epidermoneurotropic viral infection in the elderly caused by reactivation of the varicella-zoster virus during periods of impaired immunity. HZ has been reported to precede various co-localized neoplasms due to altered local immunity in the spectrum of the so-called Wolf isotopic response (WIR). We observed a patient with inverse chronology of the two conditions.

Materials & Methods:

We present an 87-year-old man with a one-week history of clustered clear-contents vesicles on erythematous and edematous base, as well as scattered erosions, covered by thick haemorrhagic crusts affecting the right half of the face but mainly confined to the right fronto-maxillar zone. Within the evolution of the herpetiform rash, a solitary pigmented nodular lesion, previously uninvestigated and untreated, became apparent in the same area. The patient's other comorbidities included chronic heart failure and stroke.

Results:

Routine laboratory revealed mild anaemia, as well as elevated inflammatory markers. Dermoscopy of the pigmented lesion was compatible with MM. Based on the clinical presentation the patient was diagnosed with maxillary HZ co-localized with MM. Treatment of HZ with systemic acyclovir 4g/day along with topical antibiotics resulted in resolution of the herpetic lesions. Surgical excision of MM was advised after the complete healing of the viral infection.

Conclusion:

HZ typically affects the elderly but its occurrence at the site of an already existing malignancy reveals the major role of the local immune response. The skin, the nervous system and immunity share common neuromodulators and cytokines in the so-called neuroimmunocutaneous system. The topical destabilization of the latter by various factors, including infection or trauma, makes the area vulnerable for a wide range of immunity-related disorders such as opportunistic infections, primary or metastatic tumours, etc. This concept of "the immunocompromised district" may explain the co-localization of the infection and malignancy. WIR, also named Wolf's postherpetic isotopic response, as HZ is usually the preceding pathology, illustrates this interpretation. The intriguing in our patient is the reverse chronoly and the development of HZ at the site of an already existing untreated MM.

Abstract N°: 7527

Morphea of the eyelids

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Introduction & Objectives: Morphea, also known as localized scleroderma, is a rare benign autoimmune connective tissue disorder mainly affecting women. It is typically characterized by asymmetrical indurated skin patches with ivory-coloured center and peripheral lilac ring. Any of the body skin areas may be involved but previous case series demonstrated highest frequency of the thorax adjacent to the breasts, followed by the trunk including the neck, lower extremities, upper extremities, face, and genitalia. Eyelid involvement is only occasionally recognized. The first clinical observation of unilateral eyelid morphea dated back to 1895, and in 1982 the first histologically confirmed case of symmetrical eyelid morphea was added. We report a patient with rare presentation of bilateral eyelid morphea associated with other sites of involvement.

Materials & Methods: A 48-years-old woman presented with six months history of well-demarcated round-shaped indurated patches measuring up to several centimeters in diameter, overlayed by atrophic epidermis and surrounded by the typical “lilac ring”. Zones of hyper- and hypopigmentation within the mentioned plaques could also be detected. The lesions involved symmetrically the upper eyelids of the patient and were accompanied by mild bilateral eyelid oedema without any subjective complaints. Similar isolated plaques were observed on the breasts and trunk. The patient reported negative serological tests for Lyme disease, performed within the past year.

Results: Routine laboratory results were within the reference ranges, apart from elevated C-reactive protein and erythrocyte sedimentation rate. Histothatological investigation of a skin biopsy specimen was compatible with morphea. Treatment with penicilline 2x5mln IU/day, antihistamines and potent dermocorticoids resulted in slow favorable evolution.

Conclusion: Eyelid involvement in morphea represents an exceptionally rare clinical manifestation. When present, it may lead to significant functional and cosmetic complications, including eye retraction, impaired ocular closure, and potential vision disturbances. Due to its atypical localization, it may remain unrecognized but needs to be differentiated from other potentially debilitating diseases, namely linear scleroderma “en coup de sabre”, Parry-Romberg syndrome or extragenital lichen sclerosus (LS) which may require more aggressive therapies. The reported cases help to trigger awareness among dermatologists and ophthalmologists.

Abstract N°: 7532

Development of a Humanized Mouse Model of Vitiligo

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Introduction & Objectives:

Vitiligo involves a multifaceted interplay of genetic, environmental, metabolic, and immune factors, which have been challenging to mimic in preclinical vitiligo research models. Yet these factors are essential for therapy development and full understanding of vitiligo. To better capture human disease pathobiology and support therapeutic development, we aimed to establish a humanized mouse model that closely mimics key features of human vitiligo.

Materials & Methods:

SCID/beige mice grafted with healthy human dark skin were topically treated with H_2O_2-NaN_3/catalase inhibitor to induce epidermal oxidative stress. Autologous PBMCs were Th1polarized, and melanocytes were menadionestressed and incubated with short synthetic peptides derived from MART1, gp100 and tyrosinase to boost extracellular melanocyteantigen availability for crosspresentation by PBMCderived dendritic cells. Co-cultured Th1-skewed PBMCs and MC were injected intradermally. Mice also received intradermal IgG4 from vitiligo patients and intravenous HSP70, a stress protein known to enhance antigen presentation. Xenotransplants were evaluated for vitiligo features and therapeutic response.

Results:

Under the above protocol, vitiligo-like depigmented lesions developed in 80% of the human skin xenotransplants . qIHC decreased epidermal melanin content and depletion of MC numbers and markers (Melan-A, gp100, c-KIT) . This was associated with increased keratinocyte secretion of vitiligo-associated cytokines (IFN-γ, IFN-α, IL-15, IL-18), enhanced gp100/NKG2D/MICA and CD8/NKG2D/gp100 interactions, and elevated CD11c+ and pDC cell numbers, and a significantly increased number of human epidermal TRM cells (CD8+/ CD103+/CD49a+) in vitiligo compared to control xenotransplants treated only with activated CD8/NKG2D cells. Preceding depigmentation, both lesional human keratinocytes and MC showed elevated senescence markers (P16INK4A, SIRT1, p-S6), alongside reduced antioxidant and mitochondrial markers (e.g., NRF2, MTCO1, Porin/VDAC, PGC1α,). FACS analysis of the epidermal sheet demonstrated an increased number of “stressed” melanocytes, i.e., MICA+, MCs compared to controls and confirmed the presence of TRM cells (CD8/CD69+/CD103+/CD49a) with increased expression of TNFα and IFN-γ in lesional, but not in control xenotransplants. Topical Tacrolimus and ruxolitinib effectively promoted repigmentation of 30% and 70% of experimentally induced vitiligo lesions, respectively.**

Conclusion:

We present the first humanized mouse model of vitiligo that recapitulates key disease features—including the characteristic immune-mediated MC loss and senescence, cytokine dysregulation, TRM cell accumulation, and responsiveness to known repigmentation-promoting therapeutics. Thus, this model—offers a clinically relevant, instructive preclinical research platform for studying vitiligo pathogenesis and evaluating candidate therapeutics directly in human skin in vivo.

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Abstract N°: 7563

Pemphigus foliaceus with erythema annulare centrifugum-like flare pattern

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Introduction & Objectives:

Pemphigus foliaceus (PF) is a rare autoimmune blistering disorder characterized by superficial epidermal blistering due to autoantibodies targeting desmoglein (Dsg) 1. Erythema annulare centrifugum (EAC) is a reactive dermatosis that presents with annular, erythematous, migrating lesions and is associated with a variety of triggers, including infections, malignancies, or autoimmune conditions. The concomitant presentation of PF and EAC is exceptionally uncommon and may reflect various scenarios. This case aims to highlight the diagnostic complexity and therapeutic considerations in a patient presenting with overlapping features of both conditions.

Materials & Methods:

We present a 48-year-old man with a 4-year history of previously confirmed PF, who was admitted to our Dermatology Department due to a disease flare over the past 2-3 weeks. The patient presented with diffuse erythema and scaling on the scalp and face, along with new onset of multiple annular erythematous lesions on the trunk and upper extremities, occurring while on maintenance therapy with triamcinolone following a tapering protocol. Subjective symptoms included pruritus and pain. No potential triggering factors preceded the new lesions. A comprehensive clinical, histopathological, immunofluorescent and immunoserological evaluation was conducted to assess both the PF activity and the nature of the new annular eruption.

Results:

Laboratory findings revealed leukocytosis, neutrophilia, elevated serum urea, and hypercholesterolemia. Histopathological examination of a biopsy specimen from EAC-like lesion could not detect apparent acantholysis. Direct immunofluorescence on perilesional skin demonstrated intercellular IgG deposition (+++). ELISA testing for anti-Dsg antibodies showed high levels of anti-Dsg1 antibodies, while anti-Dsg3 antibodies were negative. Based on the clinical presentation and supporting investigations, we considered EAC-like PF flare. Systemic treatment with dexamethasone 8 mg daily along with clobetasol propionate cream, fusidic acid cream, and emollients resulted in rapid healing of the EAC-like lesions.

Conclusion:

To the best of our knowledge, we report for the second time EAC-like relapse of previously confirmed PF. Our case follows immediately the first observation published in April 2025 of a new EAC-like PF in a patient with former localized PF. Previously described EAC-like clinical rash with immunofluorescence pattern of pemphigus has been initially termed “erythema annulare centrifugum-like acantholytic dermatosis (EAAD)” and was promoted as a novel acantholytic disease. Later, it was interpreted as a morphological variant of PF even in the absence of histologically detectable acantholysis. Thus, a new rare EAC-like clinical variant of pemphigus may be defined, like the already existing EAC-like bullous pemphigoid and EAC-like linear IgA dermatosis.

Abstract N°: 7622

Humoral response of immunotherapy-induced bullous pemphigoid patients is characterized by autoantibodies against non-NC16A epitopes: BP180 ectodomain ELISA can reduce their diagnostic delay

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Introduction & Objectives: Immune checkpoint inhibitors (ICIs) are monoclonal antibodies that bind to cytotoxic T-lymphocyte antigen-4 (CTLA-4) and programmed cell death-1 or its ligand (PD-1/PD-L1), resulting in the activation of the T-cell immune response which in turn will fight tumor cells. Their use significantly improved cancer treatment; however, the immune response activation is often associated with immune-related adverse events that most commonly affect the skin. Among these, although relatively rare (the estimated overall incidence is less than 5%), several cases of autoimmune bullous diseases following anti-PD1/L1 therapy have been reported in literature, with bullous pemphigoid (BP) being the most prevalent. BP is characterized by subepidermal

blistering caused by autoantibodies targeting hemidesmosomal proteins BP180 and BP230.

Materials & Methods: In this multicenter study, 53 patients who developed BP during ICI treatment or within 1 year from its discontinuation (ICI-BP) were demographically, clinically and serologically characterized. The humoral response was delineated by both commercial and in-house ELISA assays detecting IgG, IgE and IgA autoantibodies targeting the immunodominant domain of BP180 (NC16A) and BP230, as well as BP180's whole ectodomain (ECD), mid-portion (E-1080) and C-terminal (E-1331) epitopes. The results were compared to 87 idiopathic BP patients (IBP).

Results: ICI-BP had a mean age of 74.9 years and were predominantly males (M/F ratio = 7.8). The comparison with IBP revealed a characteristic IgG humoral response in ICI-BP: while no major differences were found toward NC16A, ICI-BP sera had higher reactivity than IBP to other BP180 epitopes (ECD: 93.6% vs 78.2%, p=0.032; E-1080: 55.3% vs 33.7%, p=0.026; E-1331: 72.3% vs 39.5%, p=0.0005), and also had higher mean titers to ECD and E-1331 (p=0.048 and p=0.002, respectively). Moreover, ICI-BP were less reactive to BP230 and had lower mean titers (24.0% vs 54.0%, p=0.001; 25.8 vs 65.4 U/ml, p=0.0009). Some differences were also found in IgE and IgA responses, where ICI-BP had higher IgE titers (p=0.030) and higher IgA reactivity (p=0.008) to NC16A compared to IBP.

Of note, in-house ELISAs based on the ECD of BP180 and on its mid-portion and C-terminus epitopes improved the combined sensitivity of commercial BP180 and BP230 ELISAs: in fact, of 8 patients who did not react to BP180 commercial kit, 7 were also negative to BP230, while all of them were positive in these alternative approaches.

Conclusion: Our findings show that ICI-BP have a specific humoral response characterized by the increased reactivity to non-NC16A epitopes, such as the C-terminal one. This distinctive response suggests that ELISAs based on the ECD of BP180 and on its C-terminus epitope may prove useful in reducing the diagnostic delay in ICI-BP patients, in whom a timely diagnosis might be crucial to appropriately manage the disease and ultimately avoid the discontinuation of cancer therapy.

Abstract N°: 7636

A Systematic Review of Case Series and Clinical Trials Investigating Regenerative Medicine for the Treatment of Vitiligo

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Introduction & Objectives:

The aim of this study is to examine the efficacy and safety of various regenerative medicine treatments, such as cell therapy, platelet-rich plasma (PRP), plasma-poor platelet (PPP), plasma-rich fibrin (PRF), mesenchymal stem cells, stromal vascular fraction (SVF), exosomes, adipose-derived stem cells (ADSC), and stem cell-conditioned media (SC-CM), for treating vitiligo.

Materials & Methods:

We conducted a thorough search of major databases such as PubMed, Scopus, and Web of Science, and selected 48 articles based on specific criteria. We used EndNote X8 and Google Sheets to review and extract data from the articles. After analyzing the studies, we categorized them accordingly.

Results:

This systematic review analyzed 48 articles involving 2,186 patients with vitiligo to assess the effectiveness of regenerative medicine treatments. Key findings revealed that methods such as autologous noncultured melanocyte-keratinocyte transplantation and platelet-rich plasma (PRP) injection exhibited significant repigmentation, particularly when combined with modalities like NB-UVB phototherapy and laser treatments. Notably, the autologous melanocyte-keratinocyte transplantation achieved over 50% repigmentation within 9 months, while PRP demonstrated an average repigmentation of 58.7%, especially effective with CO2 laser treatment. Hair follicle-derived cell transplantation also showed impressive response rates, achieving good to excellent results in up to 93.8% of patients. Side effects were noted in 21 of 28 studies, primarily involving pain, with no serious adverse events reported. The risk of bias assessment indicated that 37.21% of studies were low risk, while 48.84% had high risks overall. These findings suggest that while regenerative medicine holds promise for vitiligo treatment, further clinical trials are necessary to explore additional methods like stromal vascular fraction and exosomes.

Conclusion:

We have concluded that regenerative medicine plays an effective role in the treatment of vitiligo lesions. Furthermore, this treatment method is safe and does not cause serious complications. It can be used alone or in combination with other methods for treating vitiligo. To advance the treatment of vitiligo, we recommend conducting clinical trials on the unexplored branches of regenerative medicine.

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Abstract N°: 7647

Brunsting-Perry pemphigoid – a distinct phenotype in the spectrum of pemphigoid diseases

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Introduction & Objectives:

Brunsting-Perry pemphigoid (BPP)** is a rare, chronically persistent subepidermal autoimmune blistering dermatosis predominantly affecting middle aged or elderly men. The blistering eruption is peculiarly localized in plaques confined mainly to the photoexposed skin of the head and neck. Healing occurs with superficial scarring and milia formation. BPP was initially considered a variety of cicatricial or mucous membrane pemphigoid (MMP), but differs significantly from the latter by the absence of significant mucosal involvement. Currently, BPP's nosologic place remains uncertain between classical MMP, bullous pemphigoid (BP), and epidermolysis bullosa acquisita (EBA), as autoantibodies may target different components of the dermo-epidermal junction (DEJ), namely BP180, BP230, laminin 332, collagen VII, or another unidentified antigen.

Materials & Methods:

An 85-year-old man was admitted to the clinic for the first time with complaints of a mildly pruritic vesiculo-bullous eruption of a few weeks duration. The clinical observation revealed scattered vesicles and occasionally hamorrhagic bullae on erythematous patches, predominantly affecting the lateral parts of the face and temporal areas, the fronto-parietal scalp, and the shoulder area. Rupturing of the blisters resulted in different sized erosions some of which healed with scarring and milia formation. The other parts of the trunk, extremities and mucous membranes were spared. The patient didn't report any concomitant medical conditions and had negative drug history.

Results:

Laboratory tests detected hyperuricemia and elevated erythrocyte sedimentation rate. Histopathological evaluation revealed subepidermal blistering with an inflammatory infiltrate rich in eosinophils. Direct immunofluorescence analysis of perilesional skin demonstrated linear immunoglobulin (Ig) G (+++), IgA (++), and complement C3 (++) at the DEJ, with n-serration pattern of immune deposits. ELISA BP180, BP230 and Col VII were all negative. Based on the clinical findings and laboratory results, an autoimmune subepidermal blistering disorder consistent with BPP was considered. Systemic therapy with methylprednisolone 30mg/24h, doxycycline 100mg/24h, along with topical high-potency corticosteroids and emollients resulted in rapid improvement.

Conclusion:

The reported case of BPP** is typical in its presentation,** corresponding to the clinical disease phenotype originally described by Brunsting and Perry. Diagnostic difficulties may arise due to BPP's clinical, histopathologic and immunologic overlap with the other entities in the group of pemphigoid diseases, but it is characterized by a better overall prognosis. Some authors regard BPP as a localized variant of EBA, while others report on BPP's possible transitioning from previous BP due to epitope spreading. A comprehensive diagnostic approach is necessary to further elucidate BPP's exact nature.

Abstract N°: 7664

Successful Remission of Refractory Generalized Morphea with Low-Dose Tofacitinib: A Case Report

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Introduction & Objectives:

Morphea is a chronic autoimmune skin disease characterized by inflammation and excessive collagen deposition, leading to dermal and, at times, deeper tissue sclerosis. Generalized morphea is a severe subtype of plaque morphea, defined by the presence of more than four lesions across at least two anatomical sites. It predominantly affects females and often leads to joint mobility restriction due to deeper tissue involvement. While topical therapies and phototherapy may be effective in localized cases, systemic immunosuppression—such as methotrexate or mycophenolate mofetil—is typically required for generalized disease. Recently, Janus kinase (JAK) inhibitors, particularly tofacitinib, have gained attention for their ability to inhibit key cytokines such as IFN-γ, TGF-β, and IL-6, which are implicated in the fibrotic processes of morphea. This report presents a female patient with progressive generalized morphea unresponsive to multiple conventional therapies, who achieved remission with tofacitinib.

Materials & Methods:

The clinical records of a patient with generalized morphea who was treated with tofacitinib were reviewed.

Results:

A 58-year-old female patient presented to our clinic with a three-month history of progressive skin hardening. Dermatological examination revealed indurated plaques on the intermammary area of the chest and the upper abdomen. Repeated skin biopsies showed orthokeratosis, perivascular lymphohistiocytic infiltrates in the reticular dermis, and thickened collagen bundles on Masson’s trichrome staining. Laboratory investigations—including ANA, ENA, and RF—were negative, with all other parameters within normal limits. Magnetic resonance imaging (MRI) demonstrated no involvement of the deep fascia.She had previously received 70 sessions of narrowband UVB phototherapy, weekly subcutaneous methotrexate (cumulative dose: 1.34 g), hydroxychloroquine (200–400 mg daily for 78 months), and mycophenolate mofetil (1.5–3 g daily for 75 months), without achieving disease control. Despite these interventions, new sclerotic plaques continued to appear, particularly on the back, outer quadrant of the left breast, periumbilical region, bilateral lumbar area, lateral side of the left thigh, and bilateral pretibial regions—where longitudinal deep linear atrophic plaques were noted. At baseline, the Localized Scleroderma Severity Index (LoSSI) was 25, and the modified Localized Scleroderma Damage Index (LoSDI) was 15. All previous therapies were discontinued, and tofacitinib 5 mg daily was initiated. By the first month of treatment, a notable reduction in sclerosis was observed, with the LoSSI improving to 4; LoSDI remained at 15. The patient remained in remission at the eighth month of therapy.

Conclusion:

Promising outcomes with tofacitinib in juvenile scleroderma and generalized deep morphea suggest its potential as a treatment option in refractory cases. In many reports, a daily dose of 10 mg tofacitinib was administered. However, considering the patient’s body mass index (which exceeded 25) and her active smoker status, the dosage was not increased. Despite the use of a relatively low dose, the patient exhibited a rapid and favorable therapeutic response, which was well tolerated. JAK inhibition may offer a valuable alternative for patients who do

not respond to standard treatments; however, further studies are needed to confirm its long-term efficacy and safety.

Abstract N°: 7682

The Presence of Immunoglobulin A autoantibodies is Related to a More Severe Mucous Membrane Pemphigoid Phenotype

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Introduction & Objectives: Mucous membrane pemphigoid (MMP) presents with predominant mucosal blisters, erosions, and subsequent scarring, which can lead to serious complications, such as blindness, strictures, and airway obstruction. Next to the clinical presentation, MMP is characterized by IgG and/or IgA autoantibodies mainly against structural proteins of the basement membrane zone. Data on the clinical significance of IgA autoantibodies are scarce. The objective of this study to understand the impact of IgA autoantibodies on the clinical phenotype of MMP.

Materials & Methods: In this retrospective study (2002-2024), we included 189 patients with predominant mucosal blistering disease showing linear IgG, IgA, and/or C3c deposits in the basement membrane zone in direct immunofluorescence microscopy (DIF). Baseline diagnostic tests, and clinical findings at presentation, treatment information were extracted from patient files.

Results: Of 189 patients, 60.3% (n=114) showed IgA deposition in DIF. The patients with IgA reactivity in biopsy were significantly older (median [interquartile range]: 68 [60-77] vs. 61 [49-70], p<0.001). DIF IgA-positive patients had significantly higher rates of ocular involvement (37.7% vs.12.0%, p<0.001), and a higher number of affected mucosal sites (median [IQR]: 2 [1-3] vs. 1 [1-2], p=0.002). The presence of IgA in DIF was associated with an increased risk of 3 or more adjuvant treatment need (odds ratio [95% confidence interval]: 2.21 [1.07–4.55]) and rituximab (OR [95% CI]: 2.66 [1.23–5.76]), (p=0.032 and p=0.013, respectively).

Conclusion: The presence of IgA in DIF is associated with a more severe MMP phenotype. Therefore, IgA-targeted therapies might become important in this subgroup of patients.

Abstract N°: 7686

Liquefied Calcinosis in Dermatomyositis: A Rare Case Report and Therapeutic Challenges

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Introduction & Objectives:

Dermatomyositis (DM) is a rare autoimmune inflammatory myopathy with typical cutaneous and muscular manifestations. It may also present with systemic involvement, including pulmonary and gastrointestinal complications. One of the most debilitating cutaneous complications is calcinosis cutis, which often develops in chronic or refractory cases and is associated with prolonged disease activity and delayed treatment initiation. Among its variants, liquefied calcinosis—commonly referred to as "milk of calcium"—is exceedingly rare and underreported. It is characterized by subcutaneous calcium deposits that undergo central liquefaction, mimicking abscesses and posing diagnostic challenges. Early identification is essential to avoid unnecessary surgical interventions and to guide targeted therapy. This report aims to present a rare case of liquefied calcinosis in DM, highlighting its clinical significance, diagnostic approach using advanced imaging techniques, and therapeutic management in a real-world setting.

Materials & Methods:

We describe a 72-year-old female patient with a diagnosis of DM since 2019, who initially presented with proximal muscle weakness, Gottron's papules, heliotrope rash, Raynaud's phenomenon, and elevated muscle enzymes (CPK 1650 U/L). Laboratory findings included positive antinuclear antibodies (ANA >1:640) and a sclerodermiform pattern on capillaroscopy. The patient developed painful, ulcerated calcified plaques on the lower extremities, causing pruritus and functional impairment. After suspension of methotrexate due to an inguinal abscess, the cutaneous lesions evolved into fluctuant masses. MRI revealed deep calcinosis with associated panniculitis and diffuse muscle edema. Dermatologic ultrasound confirmed the presence of heterogeneous hyperechoic calcifications with posterior acoustic shadowing and areas of fluid collection consistent with "milk of calcium." Ultrasound-guided puncture yielded a white, milky material. Cultures were negative for infection, ruling out secondary bacterial involvement.

Results:

Management included high-dose corticosteroids, monthly intravenous immunoglobulin (IVIG) infusions for six months, and reintroduction of methotrexate once infectious complications resolved. Supportive care included local wound management, analgesia, and physical therapy. The patient showed marked clinical improvement, with progressive healing of skin ulcers, reduction in subcutaneous fluctuance, and partial recovery of muscle strength. Serial imaging confirmed stabilization of calcified lesions and reduction of associated inflammatory signs.

Conclusion:

Liquefied calcinosis is a rare but important complication of dermatomyositis that must be considered in the differential diagnosis of soft, fluctuant subcutaneous lesions in patients with long-standing or refractory disease. This case illustrates the importance of high-resolution imaging for early and accurate diagnosis, enabling conservative medical treatment and avoiding unnecessary surgical procedures. The combination of IVIG and

immunosuppressive therapy effectively reduced disease activity and improved quality of life. Given its association with a worse prognosis, close monitoring and individualized treatment strategies are essential. Further studies are needed to define standardized therapeutic protocols and improve outcomes in such rare presentations.

Abstract N°: 7723

Paraneoplasic dermatomyositis :

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Introduction & Objectives:

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by an inflammatory infiltrate primarily affecting the skeletal muscle and skin. It can also affect other organs such as lungs and joins. DM is a rare disease, it's more common among females and in low geographical latitude countries. The association between malignancy and dermatomyositis has been widely described and confirmed by numerous epidemiological studies.

The aim of this study is to describe the prevalence and types of cancer among patients with dermatomyositis and the clinical features of paraneoplasic dermatomyositis.

Materials & Methods:

We retrospectively reviewed the case records of all patients diagnosed with dermatomyositis from June , 2014 to June, 2022 in the Dermatology and Venereology Derpartement of Mohammed VI University Hospital in Oujda city. All patients with a diagnosis of DM (with or without associated malignancy) fulfilling Bohan and Peter's criteria and/or the European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) were included. Information obtained included patient demographics and clinical data.

Results:

Twenty seven patients were included, with a mean age of 49,5 ± 21 years (range: 12–82 years). Twenty patients were female (74,1%) and seven patients were male (25,9 %).

The diagnosis of cancer was retained in 8 patients (29.6%) of our series. The mean age of patients with paraneoplastic dermatomyositis was 57.7 years (+/- 16.6) (range: 36-80 years). We noted a female predominance with a sex ratio F/H of 1.6. All our patients presented general symptoms , dominated by asthenia which was present in all patients, followed by significant weight loss in 6 patients (75%) and prolonged fever in 4 patients (50%).

The most frequent cancer in our study was nasopharangeal cancer in 3 patients (37.5%), followed by breast cancer in 2 patients (25%), larynx, endometrium and stomach cancer in 1 case each . Nasopharangeal cancer was the most frequent cancer among men, while breast cancer was the most frequent type of cancer among women. The diagnosis of cancer preceded dermatomyositis in 3 cases (37.5%), was concurrent with dermatomyositis in 3 cases (37.5%), and followed the diagnosis of DM in 2 cases (25%). The median time from cancer diagnosis to dermatomyositis was 3 months [15days , 47 months].

The association between malignancy and dermatomyositis has been widely described and confirmed by numerous epidemiological studies. Approximately 13-42% of patients with DM may develop cancer , which is consistent with the results of our study.

A wide variety of malignancies have been reported in patients with DM. Nasopharyngeal cancer was the most common cancer in men, while breast cancer was the most common in women in ou study. Our results are closest

to the Asian series in singuapoure , Taiwan , and China where nasopharyngeal cancer was the most frequent. This could be explained by common environmental factors.

Conclusion:

The association of dermatomyositis with cancer is frequent and represents the main cause of mortality. We recommend a systematic nasofibroscopy in all patients with dermatomyositis in view of the frequency of nasopharyngeal cancer in our context, especially in male patients of advanced age.

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Abstract N°: 7725

Development of psoriasis (initially scalp psoriasis, followed by inverse psoriasis) and psoriatic arthritis in a patient with antisynthetase syndrome: a rare case report

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Introduction & Objectives:

Antisynthetase syndrome (ASS) is a rare autoimmune disorder serologically characterised by the presence of anti-tRNA synthetase (ARS) antibodies and clinically manifests as interstitial lung disease (ILD), myositis, arthritis, fever, Raynaud phenomenon and mechanic's hands. ASS is thought to result from a breakdown of immune tolerance, resulting in self-reactivity of the immune system. Psoriasis is a chronic systemic inflammatory disease affecting not only the skin but also various tissues and organs including joints, gut, eye, and metabolic and cardiovascular systems.

Here, we present a rare case involving the development of psoriasis- initially scalp psoriasis, followed by inverse psoriasis- and psoriatic arthritis (PsA) in a patient with ASS.

Materials & Methods:

A 44-year-old female was admitted to our department with a 2-year history of erythematous scaly plaques on the scalp and a 2-week history of erythematous lesions in the flexural areas accompanied by joint pain.

Six years earlier, she was diagnosed with ASS (anti-EJ +++, anti-Ro-52 +++, along with ILD). Over the course of her illness, she received various treatments in response to changes in her condition and was currently being treated with oral methylprednisolone (16 mg QD) and nintedanib (100 mg BID). Two years ago, she developed erythematous scaly plaques on the scalp, without similar lesions elsewhere on the body. Dermoscopic examination revealed features consistent with scalp psoriasis. Topical treatment with calcipotriol and halometasone resulted in partial improvement of the lesions. Two weeks before admission, she developed symmetric, mildly thickened, smooth erythematous plaques in the flexural areas, including the neck, antecubital fossae, axillae, inframammary folds, vulva, and perianal area. Concurrently, she experienced pain in the left knee, heel, and left index finger joint, without noticeable joint swelling or deformity.

Laboratory tests: CRP 12 mg/L, CK-MB 31 U/L, ANA 1:80 (++), anti-EJ +++, anti-Ro-52 +++, anti-SS-A +++, anti-SS-B ++, and RF, anti-CCP, ANCA, and HLA-B27 were negative. Skin biopsy of lesions in the antecubital fossae showed mild hyperkeratosis and perivascular lymphocytic infiltration in the dermis. MRI of the left knee showed mild soft tissue edema and minimal joint effusion. The patient was additionally treated with oral tofacitinib (5 mg BID), and two weeks later, the flexural erythema and joint pain had significantly improved.

Results:

The patient was diagnosed with ASS, scalp psoriasis, inverse psoriasis, and PsA. She was additionally treated with oral tofacitinib (5 mg BID), resulting in significant improvement of scalp and flexural lesions, as well as joint pain within two weeks.

Conclusion:

The overlapping of psoriasis and connective tissue diseases has been occasionally reported. However, coexistence

of psoriasis (especially inverse psoriasis and PsA) and ASS is extremely rare. In addition to the genetic background, shared pathogenesis including pDC, type I interferon, and IL-23/Th17 axis, may contribute to the development of both psoriasis and ASS. Furthermore, in this case, the addition of the JAK inhibitor tofacitinib to baseline treatment with corticosteroids and nintedanib appeared to be effective.

Abstract N°: 7732

Palisaded Neutrophilic and Granulomatous Dermatitis in ANA-Negative Lupus

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Introduction & Objectives:

Palisaded neutrophilic and granulomatous dermatitis is a rare entity but is reported to be associated with autoimmune connective diseases, lymphoproliferative disorders and infections. Although it has been reported mostly with rheumatoid arthritis, it can be observed in association with systemic lupus erythematosus and systemic vasculitides.

Materials & Methods:

A 50-year-old woman presented to our clinic with a one-month history of painful eruption on her thighs unresponsive to systemic antibiotics and topical corticosteroids. She had a known history of type 2 diabetes mellitus, for which she was receiving metformin therapy. Dermatological examination bilateral, symmetrical, erythematous plaques measuring approximately 30 × 30 cm, circumferentially distributed around both thighs. The patient reported intermittent arthralgia and lower extremity pain that improved with activity, for approximately six years prior to the cutaneous eruption. Still, no definitive rheumatologic diagnosis had been established. A skin biopsy was performed, followed by direct immunofluorescence (DIF) analysis.

Results:

Skin biopsy demonstrated collagen necrobiosis surrounded by palisading neutrophilic infiltrates in the dermis and mild mucinous material between collagen fibers. Direct immunofluorescence revealed fibrinogen and C3 deposition in superficial dermal blood vessel walls. These features were consistent with PNGD, and the patient was evaluated for possible underlying systemic disease and malignancy.

Initial laboratory workup including ANA, anti-dsDNA, RF, anti-CCP, ANCA, thyroid autoantibodies, and ACE were negative. Serum protein electrophoresis was consistent with polyclonal pattern. Spot urine protein/creatinine ratio was mildly elevated at 197 mg/g. Due to the histopathologic findings and the worsening joint symptoms, the patient was later referred to rheumatology, where an extended autoantibody panel revealed anti-Sm/RNP (+++), anti-histone (+), and anti-PCNA (+) confirming the diagnosis of systemic lupus erythematosus.

The patient was initiated on hydroxychloroquine (200 mg twice daily) and methylprednisolone (10 mg daily). This treatment resulted in complete remission of both cutaneous and joint symptoms.

Conclusion:

This case is notable for its seronegative ANA profile, which may delay diagnosis. According to the 2023 EULAR/ACR classification, Anti-Sm/RNP positivity can substitute for the immunologic domain in the absence of ANA. The histologic diagnosis of PNGD prompted a systemic workup, leading to diagnosis of systemic lupus erythematosus. With appropriate treatment, both the skin and systemic manifestations resolved. This case underscores the critical role of skin biopsy in the diagnostic workup of atypical dermatoses. It highlights how dermatopathologic findings can serve as early indicators of systemic autoimmune disease, even in the absence of classic serologic markers.

Abstract N°: 7769

Causes of mortality in dermatomyositis

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Introduction & Objectives:

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by an inflammatory infiltrate primarily affecting the skeletal muscle and skin. It can also affect other organs such as lungs and joins. DM is a rare disease, it's more common among females and in low geographical latitude countries. The aim of this study is to describe the prevalence and causes of mortality among patients with dermatomyositis.

Materials & Methods:

We retrospectively reviewed the case records of all patients diagnosed with dermatomyositis. All patients with a diagnosis of DM (with or without associated malignancy) fulfilling Bohan and Peter's criteria and/or the European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) were included

Results:

Death occurred in 25.92% in our series (7 cases), the cause of death was cancer in 42.8% (3 cases), respiratory distress due to respiratory muscle failure in 28.6% (2 cases), an infectious cause in 28.6% (2 cases) including 1 case by septic shock with a pulmonary starting point to the germ Pseudomonas Aeruginosa and another case by a pulmonary infection to SARS-COV2. Death occurred within a mean time of 124 days+ /- 121 days after the diagnosis of DM with extremes ranging from 45 days to 1 year.

With earlier diagnosis and the use of immunosuppressive agents in recent decades, survival of patients with DM has gradually improved worldwide. Cancer, pulmonary and cardiac complications, and infections are generally cited as the most common causes of death in DM.

In our study, death occurred in 25.9%. DM-related mortality varies between 18.1% and 58.1% in different series in the literature . Our results are closest to the Japanese study by Yamasaki et al. and the Spanish study by Torres et al. where the mortality rate was 26.9% and 27.5% respectively.

In our series, the first cause of death was cancer in 42.8%, followed by respiratory distress due to respiratory muscle failure in 28.6% and infection in 28.6%. These results are consistent with the results of the literature where the main causes of death were cancer, respiratory failure, infection and cardiac injury.

Conclusion:

Dermatomyositis remains a potentially life-threatening condition despite therapeutic advances. Our findings confirm that cancer, respiratory complications, and infections are the leading causes of death in patients with DM. Early diagnosis, systematic cancer screening, and vigilant monitoring of pulmonary and infectious complications are crucial to improve prognosis.

Abstract N°: 7770

Two patients with refractory dermatomyositis treated with Anifrolumab: A new hope?

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Introduction & Objectives:

Dermatomyositis is an idiopathic, immune-mediated inflammatory myopathy characterized by proximal muscle weakness and characteristic skin lesions, with potential systemic involvement and a risk of association with malignancy. Traditionally, this connective tissue disorder has been treated with systemic corticosteroids or other immunosuppressants such as methotrexate or mycophenolate. Recently, biologic drugs or small molecules are being studied for the treatment of complex and refractory cases. We present two cases of dermatomyositis refractory to classic immunosuppressors treated with intravenous Anifrolumab.

Clinical Cases:

Our first patient is a 53-year-old woman diagnosed in 2018 with amyopathic dermatomyositis with negative myositis-specific autoantibodies. She had received multiple treatments, which were discontinued due to ineffectiveness or adverse effects. In December 2024, treatment with intravenous Anifrolumab 300 mg every 4 weeks was initiated. From the first dose, there was resolution of the skin lesions, and to date, she has not experienced any complications. The second patient is an 83-year-old woman diagnosed with amyopathic dermatomyositis anti-SAE. After the ineffectiveness of methotrexate, toxicodermia from hydroxychloroquine, and a cytomegalovirus infection triggered by mycophenolate, she started treatment with intravenous Anifrolumab 300 mg every 4 weeks, with an excellent response and no associated adverse events

Discussion/ Results:

Anifrolumab is a monoclonal antibody targeting subunit I of the type I interferon receptor. This drug has shown great efficacy in treating skin lesions in refractory dermatomyositis safely in isolated clinical cases and case series. We present our experience with this drug in two patients who did not respond or had adverse effects with various immunosuppressors.

Conclusion:

We highlight the promising role of intravenous Anifrolumab in the treatment of dermatomyositis.

Abstract N°: 7795

Isotretinoin-Induced Dermatomyositis: A Case Report

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Introduction & Objectives:

Isotretinoin is widely prescribed for moderate-to-severe acne vulgaris and has a well-established safety profile. However, its rare association with autoimmune phenomena has raised concerns about immune dysregulation. We report a rare and possibly first histologically confirmed case of isotretinoin-induced dermatomyositis (DM) in a previously healthy young woman, highlighting a novel link between isotretinoin and connective tissue disease.

Materials & Methods:

A 24-year-old woman with no autoimmune history began isotretinoin 20 mg/day for nodulocystic acne. Within two months, she developed fatigue, joint pain, and facial and truncal erythema. The dose was increased to 30 mg/day but discontinued due to symptom worsening. Clinical exam showed malar erythema sparing the nasolabial folds, periocular discoloration, and erythematous patches over the dorsum of the hands and chest. ANA was positive; anti-dsDNA and myositis-specific antibodies were negative. A skin biopsy revealed interface dermatitis with dermal mucin deposition. Rheumatological assessment, clinico-histopathological correlation, and serology all concurred early DM. Treatment included prednisolone 5 mg, azathioprine 100 mg, and hydroxychloroquine 200 mg daily.

Results:

Following cessation of isotretinoin and initiation of immunosuppressive therapy, the patient's fatigue and joint discomfort notably subsided, with gradual resolution of facial and truncal erythema. The temporal sequence, clinical manifestations, histopathologic findings, and subsequent response to treatment collectively support a drug-induced autoimmune etiology. To date, dermatomyositis remains an unreported adverse reaction in large-scale isotretinoin safety data.

Conclusion:

This case adds to the growing evidence of retinoid-triggered autoimmunity and is among the first to document dermatomyositis in this context. Isotretinoin, though immunomodulatory, may unmask or trigger autoimmunity in predisposed individuals. While musculoskeletal aches are common during therapy, persistent symptoms accompanied by systemic signs—such as fatigue, rash, or weakness—warrant further evaluation. Interface dermatitis with mucin, ANA positivity, and classic skin features support the diagnosis. Clinicians should remain vigilant in patients who develop new-onset systemic symptoms on isotretinoin, especially when labs suggest immune activation. Early recognition allows for prompt cessation and intervention, preventing disease progression. This case emphasizes the need for thoughtful patient selection and close monitoring during retinoid therapy. It also opens avenues for future research into isotretinoin's immunologic effects and autoimmune risks.

Abstract N°: 7879

Unmasking the Mimic: Bullous SLE Presenting as Chronic Bullous Disease of Childhood in a Teenage

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Title: Unmasking the Mimic: Bullous SLE Presenting as Chronic Bullous Disease of Childhood in a Teenage

Introduction & Objectives: Bullous systemic lupus erythematosus (BSLE) is a rare vesiculobullous manifestation of systemic lupus erythematosus (SLE) that may closely resemble chronic bullous disease of childhood (CBDC), particularly in pediatric patients. Accurate differentiation is critical due to the systemic implications and therapeutic requirements of BSLE, including potential lupus nephritis.

Materials & Methods: A 13-year-old female presented with pruritic, tense bullae arranged in a “string of pearls” configuration, predominantly over the trunk and flexural surfaces, morphologically characteristic of CBDC. Histopathology and direct immunofluorescence initially supported this diagnosis. However, persistent microscopic hematuria, proteinuria, and elevated serum autoantibodies like anti-dsDNA suggested systemic disease. Further evaluation revealed class II lupus nephritis, confirming a final diagnosis of BSLE.

Conclusion: This case underscores the diagnostic challenge posed by BSLE with a clinical phenotype mimicking CBDC. The presence of systemic signs, even in morphologically typical bullous lesions, warrants a thorough autoimmune and renal workup. Prompt recognition enables appropriate immunosuppressive therapy and improves prognosis.

Abstract N°: 7881

Periungual Dermoscopy in Systemic Autoimmune Diseases

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Introduction & Objectives:

Periungual dermoscopy is a simple, rapid, non-invasive, and reproducible examination. It is an increasingly common and valuable tool for evaluating digital microcirculation. Its primary indication lies in the assessment of vascular acrosyndromes, particularly Raynaud's phenomenon, and it also plays an important role in systemic sclerosis. The aim of our study is to describe the various periungual dermoscopic patterns observed in systemic autoimmune diseases managed in our department.

Materials & Methods:

We conducted a retrospective, descriptive study including all patients managed in the dermatology department . The objective was to describe the different periungual dermoscopic abnormalities in systemic autoimmune diseases and to monitor their evolution after treatment.

Results:

We included 56 patients, of whom 44 were women and 12 were men, with a marked female predominance (female-to-male ratio of 3.66). The mean age of the patients was 49.55 years, ranging from 9 to 87 years. Seven patients had systemic sclerosis, 20 had dermatomyositis (including 7 with a paraneoplastic form), 10 had systemic lupus erythematosus (SLE), and 19 had cutaneous lupus erythematosus (CLE).Analysis of periungual capillaroscopy images in systemic sclerosis revealed the presence of megacapillaries in 71.5% of cases, avascular areas, exudates, and microhemorrhages in 28.5% of cases each, and tortuous vessels in 14% of cases, with a normal appearance in 14% of cases. In dermatomyositis, capillaroscopic abnormalities were mainly represented by megacapillaries in 65% of cases, followed by microhemorrhages in 50%, bushy tortuous vessels in 35%, architectural disorganization in 30%, capillary rarefaction and exudates in 20% of cases each, avascular areas in 15%, and "stacked coin" hemorrhages in 10% of cases.In cutaneous and systemic lupus, periungual capillaroscopy appeared normal in 58.6% of patients. Megacapillaries were observed in 31%, avascular areas in 24%, microhemorrhages in 17%, and bushy tortuous vessels in 6%.Regarding disease progression, 50% of the patients were lost to follow-up, 30% showed a stationary capillaroscopic appearance, 12.5% showed improvement after treatment, and 4% died.

The usefulness of periungual capillaroscopy has been demonstrated in the diagnosis and screening of systemic autoimmune diseases. The capillaroscopic pattern in systemic sclerosis is known as the "scleroderma pattern," which is highly specific for the disease and is characterized by capillary rarefaction and avascular zones, megacapillaries on at least two fingers, branched capillaries, and major architectural disorganization. In dermatomyositis, capillaroscopy often reveals abnormalities similar to those seen in systemic sclerosis. In cutaneous and systemic lupus, capillaroscopy is nonspecific. In lupus, a normal capillaroscopic appearance was observed in most cases, while the other abnormalities—such as avascular areas—were consistent with the literature, although avascular zones have rarely been reported in previous studies.

Conclusion:

Periungual dermoscopy is increasingly recommended in the etiological work-up of systemic diseases, as well as for monitoring their progression under treatment.

Abstract N°: 7884

Recalcitrant IgA pemphigus with successful response to adalimumab: case series from a tertiary hospital.

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Introduction & Objectives:

IgA pemphigus is an autoimmune blistering disease presenting as vesicles and pustules mainly on flexures. It is characterized by a neutrophilic infiltrate with intercellular IgA deposition in the epidermis. (1)

Two subtypes have been described, according to the histopathological level of cleavage: subcorneal pustular dermatosis (SPD) and intraepidermal neutrophilic dermatosis (IEN). The diagnosis should be confirmed with immunofluorescence studies. (1)

Systemic treatments include dapsone, colchicine, prednisone, acitretin, isotretinoin, tetracycline, sulfamethoxazole/trimethoprim, methotrexate, cyclosporine, adalimumab, sulfamethoxypyridazine, azathioprine, and PUVA. (1, 2)

We present a case series of three patients with IgA pemphigus in whom disease control was only achieved with adalimumab. Our aim is to demonstrate that this may be an effective treatment.

Materials & Methods:

This is an observational, descriptive and retrospective study. We reviewed records of three patients with confirmed diagnosis of IgA pemphigus under follow-up at our tertiary hospital from 2002 to 2025.

Results:

1- A 24-year-old female with SPD has been followed-up for over 20 years with frequent relapses, including an ICU admission, despite use of prednisone, azathioprine, colchicine, dapsone, isotretinoin, acitretin and cyclosporine. In 2023 adalimumab was prescribed with complete remission. It was discontinued in 2025 due to a diagnosis of cervical cancer, triggering a relapse.

2- A 21-year-old female with SPD showed partial response to prednisone, isotretinoin, NB-UVB and PUVA and adverse effects to other drugs (dyslipidemia with acitretin, nausea with methotrexate; renal dysfunction with cyclosporine). Adalimumab was prescribed in 2011 with rapid response, but was withheld in two episodes of suspected infections. We achieved complete control in 2021, even after increasing the dosing interval.

3- A 10-year-old female with SPD presented recalcitrant lesions with doxycycline, colchicine, dapsone and isotretinoin. Corticosteroids were contraindicated due to congenital dilated cardiomyopathy. In 2024 adalimumab was prescribed after cardiology clearance with complete remission and the patient only relapsed when the drug was unavailable.

Conclusion:

We report three cases of refractory IgA pemphigus with complete sustained response to adalimumab monotherapy (Table 1).

Adalimumab - a recombinant monoclonal antibody that binds to TNF-α - decreases the neutrophilic epidermal infiltration, thus enabling improvement of IgA pemphigus. (3)

Previously, at least four cases of Iga pemphigus successfully treated with adalimumab were reported (2, 3, 4, 5). We conclude that adalimumab may be an effective and safe treatment for IgA pemphigus.

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Table 1 - Three cases of IgA pemphigus (Subcorneal Pustular Dermatosis variant) with successful response to adalimumab

	Case 1	Case 2	Case 3
Sex	Female	Female	Female
Age of onset (years)	24	21	10
Medical history	Non-significant	Non-significant	Congenital dilated cardiomyopathy
Previous treatments	Acitretin, Azathioprine, Colchicine, Cyclosporine, Dapsone, Isotretinoin, Prednisone	Acitretin, Colchicine, Cyclosporine, Dapsone, Isotretinoin, Methotrexate, Prednisone, PUVA, UVB-NB	Colchicine, Dapsone, Doxycycline, Isotretinoin
Dosing (subcutaneous)	80 mg on day zero, followed by 40 mg every 14 days	80 mg on day zero, followed by 40 mg every 14 days Dosing interval increased to 40 mg every 28 days with unsatisfactory response Current dosing interval 40 mg every 21 days with sustained response	80 mg on day zero, followed by 40 mg every 14 days
Significant events after treatment initiation	Cervical cancer (probably not related to adalimumab) Relapse after adalimumab discontinuation following cervical cancer	Blepharitis and conjunctivitis with no need to stop the medication Respiratory syndrome (Tuberculosis and fungal infection ruled out), skin abscess Relapse when adalimumab was withheld while infection investigation and treatment	Relapse when adalimumab was unavailable
Current status	Adalimumab reintroduced in March 2025 after oncology clearance with sustained response	Adalimumab reintroduced in December 2021 with sustained response on a 21 day dosing interval	Adalimumab reintroduced in February 2025 with sustained response

Abstract N°: 7965

Cicatricial alopecia as the key manifestation of pemphigus erythematosus

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Introduction & Objectives:

Pemphigus erythematosus (PE), also known as Senear-Usher syndrome, is a rare variant within the pemphigus group of autoimmune blistering diseases. It represents an overlap between pemphigus foliaceus (PF) and lupus erythematosus (LE). Patients exhibit clinical, histopathological, and immunological features of both conditions, including a reticular pattern of intercellular IgG and C3 upon direct immunofluorescence (IF) microscopy, and serum anti-desmoglein (Dsg) antibodies and/or antinuclear antibodies (ANA). The autoantibodies recognize Dsg1, as well as Ro, La, and double-stranded DNA antigens. Clinically, PE typically presents with erythematous, scaly plaques and shows a female predominance. Progression to systemic LE is rare. We present an unusual case of cicatricial alopecia in which a detailed investigation revealed previously unrecognized PE.

Materials & Methods:

An 18-year-old woman presented with scalp patches of scarring alopecia. The patient had a history of "small ulcers" appearing on her head and body since the age of 9 but her condition had progressively worsened over the past year. On physical examination, the parietal zone was almost entirely occupied by several atrophic patches of varying sizes, coalescing to form a large bald plaque with a few band-like islands of preserved hair. A large dry erosion, partly covered by a thick crust was present in the center of the alopetic area along with a few smaller erosions in its periphery. Careful examination of the entire skin revealed a few discrete hyperpigmented macules on the trunk. There was no history of other associated diseases.

Results:

Histological examination demonstrated superficial epidermal acantholysis, dermal fibrosis, and lack of hair follicles. Direct IF on perilesional skin showed intercellular IgG throughout the epidermis and granular deposits of IgM and C3 along the dermo-epidermal junction. Indirect IF on monkey esophagus substrate was negative for anti-epithelial cell surface antibodies and ANA. On ELISA-Dsg1 and Dsg3, the patient's serum contained antibodies only against Dsg1. The diagnosis of PE was retained based on the immunopathologic findings. The patient was treated with dapsone, topical corticosteroids, and antiseptics, which led to healing of the erosions.

Conclusion:

Scarring and non-scarring alopecia are well-documented in LE, often correlating with a prolonged disease course. In contrast, scalp involvement is common in pemphigus vulgaris (PV) and PF but rarely results in permanent hair loss. Previous studies have assessed alopecia in various autoimmune blistering diseases, with mechanisms potentially involving antibody-mediated damage to the hair follicle structures. Although scalp involvement in PE has been reported, no previous cases have specifically focused on the development of scarring alopecia. Our case adds to the limited literature by expanding the known spectrum of PE-associated manifestations.

Abstract N°: 7980

Chronic Bullous Disease of Childhood Induced byPulmonary Tuberculosis in a Malnourished 12-Year-Old Girl

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Introduction & Objectives:

In endemic countries such as Indonesia, tuberculosis (TB) infection is a disease that is difficult to eradicate and cure. TB can also trigger rare diseases such as an autoimmune disease i.e. chronic bullous disease of childhood (CBDC). Autoantibodies in TB can increase due to imbalanced immune response. The mechanism of AAB formation in TB includes excessive cell death and insufficient clearance of dead cells.

TB therapy must be completed to prevent the worsening of the disease, which can trigger the emergence of autoimmune diseases. However, multidrug treatment (MDT) and corticosteroids in autoimmune disease can worsen TB in immunocompromised patients, which increases the risk of adverse drug reactions, such as acute drug-induced hepatitis (ADIH). Therefore, second-line therapy such as sulfasalazine (SS) should be administered in this condition.

Materials & Methods:

We report an interesting case about a 12-year-old girl with stunting who presented with widespread itchy reddish patches covering almost her entire body, accompanied by blisters on various areas since one month before admission. She also had a history of fluctuating fever and productive cough over the past few years. At the age of 5 months old, she was diagnosed with pulmonary TB and treated for one month, but the treatment was discontinued. Dermatological findings showed annular urticated erythematous patches along with multiple tense bullae and vesicles clustered at the periphery, resembling a “string of pearls” on the trunk and lower limb. Laboratory investigations revealed an elevated erythrocyte sedimentation rate (ESR). Chest X-ray showed cardiomegaly without pulmonary congestion and findings suggestive of active pulmonary TB. Histopathological examination (HE) of the blister on the back demonstrated a vesicobullous and acantholytic reaction that supported CBDC.

Initial therapy was methylprednisolone 25 mg every 12 hours, but the dose was reduced after the diagnosis of TB was confirmed. The patient then received MDT for TB, and no new blisters developed after the initiation of MDT therapy. However, five days after MP therapy was reduced to 8 mg twice daily, new blisters appeared. Simultaneously, there was an increase in transaminase enzymes indicating the occurrence of ADIH, so MDT was stopped and oral SS was given.

Results:

In this patient’s complex situation, malnourished with pulmonary TB and CBDC followed by drug adverse conditions such as ADIH, second-line treatment for CBDC, i.e., sulfasalazine, should be given. After 1.5 months of SS therapy, the patient’s condition improved rapidly with no new blister formation.

Conclusion:

In the pathogenesis of autoimmune disease, systemic infection could act as an autoantigen that triggers or worsens the disease. This comorbidity creates a dilemma in therapy since systemic steroids are the gold standard

for autoimmune diseases, but they should be limited to treating infection, including pulmonary TB. On the other hand, malnutrition caused by TB infection can create an immunocompromised condition, and it increases susceptibility to hepatotoxicity and reduces hepatic resilience, which can lead to ADIH.

This case highlights pulmonary TB as a trigger factor of CBDC, and autoimmune conditions could increase the ADIH of TB treatment, and vice versa. Thus, sulfasalazine could replace steroid treatment and make a good result.

Abstract N°: 8020

Pemphigus vulgaris and nephrotic syndrome in a pediatric patient

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Introduction & Objectives:

Pemphigus vulgaris (PV) is a rare autoimmune blistering disease (AIBD), characterized by the occurrence of tissue-bound and circulating IgG4 and IgG1 autoantibodies directed against desmosomal structural proteins. PV is classified as skin-specific AIBD but there is growing evidence that systemic manifestations may also occur. PV has rarely been described in association with renal diseases, namely minimal change nephropathy, membranous nephropathy, renal amyloidosis, or compromising end-stage renal disease patients. We report a case of PV in a pediatric patient with nephrotic syndrome and suggest a possible relationship between the AIBD and the renal manifestations.

Materials & Methods:

A 14-year-old girl diagnosed with PV a year before, was readmitted to the dermatology department for another severe PV flare while being on oral methylprednisolone and azathioprine. Multiple blisters and erosions, covered with thick and adherent serohaemorrhagic crusts due to profuse exudation, spread over the face, scalp, trunk, and extremities. The mucosal surfaces were significantly involved with numerous oral and nasal erosions, and bilateral conjunctival injection. Nail changes included onychomadesis, Beau lines, and periungual vesicles. The child was in a slightly deteriorated general condition with moderate bilateral edema of the lower limbs.

Results:

Over the course of the hospitalization, systemic corticosteroids, antibiotics and antimycotics along with topical antiseptics and potent corticosteroids, remained ineffective and the patient gradually developed the signs of nephrotic syndrome with oliguria, proteinuria, hypoproteinemia, hypoalbuminemia, and pronounced edema of the face and extremities. The child was transferred to the department of pediatric nephrology and hemodialysis for further tests and treatment. Kidney biopsy was compatible with focal segmental glomerulosclerosis (FSGS) or tubulointerstitial nephritis. Kidney direct immunofluorescence failed to reveal immune deposits due to the absence of glomeruli in the respective specimen. Therapy with rituximab 500 mg/weekly for four weeks in addition to the systemic corticosteroids achieved significant general improvement and parallel control of both the skin and renal diseases.

Conclusion:

To our knowledge this is the second reported case of PV and FSGS and further adds to the short list of associations between PV and kidney diseases, despite the kidney being a frequent target of immune-mediated injury. The occurrence of nephrotic syndrome and PV in our case, as well as their parallel control by the B-cell depleting agent rituximab may suggest a common pathogenic mechanism. PV is the best known IgG4 mediated autoimmune disease but similar immunopathological mechanisms are considered also in myasthenia gravis, thrombotic thrombocytopenic purpura, autoimmune encephalitis, and glomerulonephritis. Further investigations

are needed to prove these relations.

Abstract N°: 8057

Managing a Paradox: Exacerbation of Pemphigus Vulgaris Following Rituximab Infusion

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Introduction & Objectives:

Pemphigus vulgaris is a rare, chronic, and potentially life-threatening autoimmune blistering disorder. Rituximab, a CD20+ B-cell-depleting monoclonal antibody, is considered first-line therapy for moderate-to-severe cases, achieving disease control in the majority of patients. Although generally well-tolerated, rare cases of paradoxical worsening following rituximab infusion have been reported.

Materials & Methods:

We report the case of a 43-year-old Caucasian male with a 4-year history of pemphigus vulgaris who experienced a paradoxical disease exacerbation 20 days after his fourth rituximab infusion. He had previously completed three cycles of rituximab over the past year without complications and had discontinued oral prednisolone shortly after the third cycle. His Pemphigus Disease Area Index (PDAI) score rose from 5 prior to the fourth infusion to 68 post-infusion, prompting hospitalization. Clinical examination revealed multiple painful erosions with hemorrhagic crusts and several flaccid blisters affecting the skin and oral mucosa. The patient was obese (BMI 32.7) and had a history of corticosteroid-induced osteoporosis. Prior treatments with systemic corticosteroids, mycophenolate sodium, and azathioprine had failed to achieve adequate disease control before the initiation of rituximab.

Results:

High-dose prednisolone (1 mg/kg/day) was initiated upon admission and tapered according to clinical response. Mycophenolate sodium was reintroduced two weeks later. Marked clinical improvement was observed within 10 days, with substantial resolution of lesions by one month. Notably, a fifth rituximab infusion was administered post-recovery without recurrence or adverse effects.

Conclusion: This case highlights a rare but clinically significant complication—paradoxical exacerbation of pemphigus vulgaris following rituximab infusion. Although the underlying mechanism remains unclear, such flares have been observed with both the original rituximab drug and biosimilars. The absence of previous adverse reactions does not eliminate the risk of future flares, nor does a prior exacerbation necessarily predict recurrence. Prophylactic corticosteroid administration prior to rituximab infusion may be beneficial in selected cases. Clinicians should remain vigilant for this potential complication and manage it promptly with high-dose corticosteroids and/or adjunctive immunosuppressive therapy. Re-challenging with rituximab may remain a viable option under close monitoring.

Abstract N°: 8081

Periorbital Edema and Erythema as a Singular Manifestation of Cutaneous Lupus Erythematosus – A Diagnostic Challenge

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Introduction & Objectives:

Cutaneous lupus erythematosus (CLE) is an autoimmune skin disease with diverse clinical manifestations. Periorbital erythema and edema represent an uncommon variant of CLE that poses significant diagnostic and therapeutic challenges. This case aims to highlight the need for a high index of suspicion and a multidisciplinary approach when dealing with persistent periorbital inflammation of unclear etiology.

Materials & Methods:

We present the clinical case of a 34-year-old female patient with unilateral, non-painful, periorbital edema and erythema, persisting for the past six months, without associated systemic symptoms or visual impairment. Her medical history was notable for three early pregnancy losses, all occurring between 8 and 12 weeks of gestation. Initial symptom relief was achieved with high-dose systemic corticosteroid therapy; however, recurrence occurred upon dose tapering and discontinuation. A comprehensive laboratory workup for tumoral and autoimmune panels was within normal limits. Serum ACE, IgG4 levels, and serologies for Toxocara canis, subcutaneous dirofilariasis and nematode screening blot were also normal. Genetic testing identified a homozygous mutation in the PAI-1 gene. Neck MRI and ultrasound of parotid and submandibular glands revealed no abnormalities. Imaging studies included contrast-enhanced cranial and orbital MRI scans, which ruled out orbital masses or infiltrative lesions, but showed findings consistent with right-sided periorbital cellulitis.

Results:

We performed skin biopsy due to clinical suspicion of sarcoidosis, granulomatosis with polyangiitis, cutaneous lymphoma or IgG4-related disease. Histological findings were consistent with cutaneous lupus erythematosus. The patient was initiated on hydroxychloroquine, which resulted in a gradual but sustained clinical improvement.

Conclusion:

This case underscores the importance of including CLE in the differential diagnosis of chronic periorbital inflammation, particularly when initial evaluations are inconclusive. The absence of classic cutaneous or systemic lupus features may delay diagnosis. Skin biopsy remains a cornerstone for establishing the etiology of atypical cutaneous presentations, and early recognition is essential for optimal management.

Abstract N°: 8101

Real-World Effectiveness of Janus Kinase Inhibitors in Alopecia Areata and Vitiligo: A Case Series

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Title: Real-World Effectiveness of Janus Kinase Inhibitors in Alopecia Areata and Vitiligo: A Case Series

Authors: V.Lazarou, N.Lolou, C.Daniil, E.Papadavid

Introduction & Objectives: Janus kinase inhibitors have shown promise for severe alopecia areata (AA) and non-segmental vitiligo. Oral baricitinib resulted in significant hair regrowth in AA (BRAVE-AA1, BRAVE-AA2), while topical ruxolitinib was approved for vitiligo based on repigmentation observed in TRuE-V1 and TRuE-V2 trials. This case series presents real-world outcomes of AA patients treated with oral JAK inhibitors and vitiligo patients treated with topical ruxolitinib.

Materials & Methods: We conducted a retrospective chart review of 11 patients treated with JAK inhibitors at our institution. The cohort included six patients with severe AA treated with oral baricitinib (n=4) or oral upadacitinib (n=2), and five patients with vitiligo treated with topical ruxolitinib. Treatment selection was guided by clinical presentation, comorbidities, and national health system access criteria. All AA patients concurrently received oral minoxidil (2.5–5 mg/day). Follow-up for the AA cohort ranged from 3 to 30 months (median: 16.5 months); the vitiligo cohort had a minimum follow-up of 9 months.

Results: All AA patients presented with severe disease at baseline (SALT:55–100). Treatment with oral JAK inhibitors, combined with oral minoxidil, led to significant and clinically meaningful hair regrowth. Improvements in SALT scores, ClinRO for eyebrows and eyelashes, and DLQI were observed in responding patients, with initial responses noted as early as 3 months and sustained in some individuals for up to 30 months. At baseline, most patients exhibited significant loss of eyebrows and eyelashes. After 3 months of treatment, SALT scores showed a spectrum of responses, from non-response to substantial reductions. Further reductions in SALT scores were evident at 6 months in multiple patients, with values as low as 22. Longitudinal follow-up data extending up to 30 months for a subset of patients demonstrated sustained or continued improvements, with one patient achieving near-complete hair regrowth at 16 months from a baseline of 100.

The impact on quality of life, as reflected by DLQI scores, consistently improved in the AA cohort, with baseline scores ranging from 4 to 18 reducing to values as low as 1 at later time points. The median follow-up duration for the AA cohort was 16.5 months (3–30 months). Adverse events observed with baricitinib included transient lymphopenia, thrombocytosis, and mild acne.

In the vitiligo cohort, treatment with topical ruxolitinib resulted in reductions in mean VASI scores and corresponding improvements in DLQI scores at the 9-month assessment. Mild to moderate repigmentation was observed in treated areas in the majority of patients. No significant adverse events were reported in either treatment group during the follow-up period.

Clinical response is shown in Table 1 for Alopecia Areata and in Table 2 for Vitiligo

Table 1.

PATIENT	Age of onset	JAK inhibitor initiation	DLQI baseline	Additional therapy	SALT score baseline	clinRO for eyebrows and eyelashes baseline	SALT score 3 months	clinRO 3 months	DLQI 3 months	SALT score 6 months	clinRO 6 months	DLQI 6 months
1	4 years old	JAN-2024 minoxidil 2.5mg baricitinib 4mg	4	topical corticosteroids, topical minoxidil	100	3 eyebrows and eyelashes	100	3 eyebrows and eyelashes	4	100	2 eyebrows 3 eyelashes	4
2	4 years old	NOV-2023 minoxidil 2.5mg upadacitinib 15mg	15	topical corticosteroids	100	3 eyebrows and eyelashes	75	2 eyebrows 3 eyelashes	12	55	2 eyebrows 3 eyelashes	8
3	18 years old	SEP-2022 minoxidil 2.5mg baricitinib 4mg	15	topical corticosteroids, topical minoxidil	100	3 eyebrows and eyelashes	43.8	3 eyebrows and eyelashes	11	33.4	2 eyebrows and eyelashes	7
4	3 years old	OCT/2024 minoxidil 2.5mg upadacitinib 15mg	15	topical corticosteroids, topical minoxidil	61.2	1 eyebrows and 2 eyelashes	54	1 eyebrows and 2 eyelashes	11	45	0 eyebrows and eyelashes	9
5	17 years old	APR-2024 minoxidil 2.5mg baricitinib 4mg	18	topical corticosteroids, topical minoxidil	100	3 eyebrows and eyelashes	32	2 eyebrows and 3 eyelashes	5	22	0 eyebrows and eyelashes	3
6	46 years old	FEB-2025 minoxidil 2.5mg baricitinib 4mg	18	topical corticosteroids, topical minoxidil	55	2 eyebrows and 0 eyelashes	45	0 eyebrows and eyelashes	15	-	-	-

PATIENT	SALT score 12 months	clinRO 12months	DLQI 12months	SALT score 16months	clinRO 16months	DLQI 16months	SALT score 30 months	clinRO 30months	DLQI 30 months
1	95	1 eyebrows and eyelashes	4	95	0	4	-	-	-
2	33	1 eyebrows 3 eyelashes	8	25	0 eyebrows and 3 eyelashes	8	-	-	-
3	9	1 eyebrows and eyelashes	4	2	0 eyebrows and eyelashes	2	25	0 eyebrows and eyelashes	2
4	-	-	-	-	-	-	-	-	-
5	16	0 eyebrows and eyelashes	1	-	-	-	-	-	-
6	-	-	-	-	-	-	-	-	-

Table2.

PATIENT	Age of onset	JAK Inhibitor initiation	Additional therapy	VASI score baseline	DLQI baseline	VASI score 3 months	DLQI score 3 months	VASI score 6 months	DLQI score 6 months	VASI score 9 months	DLQI score 9 months
1	14 years old	AUG-2024 Ruxolitinib	topical corticosteroids, tacrolimus, pimecrolimus	8.2	16	7.6	12	6.9	11	6.5	8
2	55 years old	SEP-2024 Ruxolitinib	topical corticosteroids, tacrolimus, pimecrolimus	5.1	12	4.9	10	4.2	8	-	-
3	42 years old	NOV-2024 Ruxolitinib	topical corticosteroids, tacrolimus, pimecrolimus	4.2	14	3.8	6	3.5	6	-	-
4	25 years old	JUL-2024 Ruxolitinib	topical corticosteroids, tacrolimus, pimecrolimus	6.8	11	6.2	9	5.5	7	5.2	6
5	37 years old	MAY-2024 Ruxolitinib	topical corticosteroids, tacrolimus, pimecrolimus	4.5	9	4.1	7	3.9	7	3.5	5

Conclusion: This real-world case series provides compelling evidence that oral JAK inhibitors, particularly when combined with oral minoxidil, result in significant hair regrowth and improved quality of life in patients with severe AA, with responses demonstrating durability over extended follow-up. Topical ruxolitinib shows promising preliminary effectiveness in promoting repigmentation and enhancing quality of life in patients with vitiligo.

Abstract N°: 8104

Upregulated glucose metabolism may correlate with CLE clinical activity

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Introduction & Objectives:

Cutaneous lupus erythematosus (CLE) is an autoimmune skin disease that may present as a localized cutaneous condition or as a manifestation of systemic lupus erythematosus, potentially indicating severe, multi-organ involvement. Higher glucose availability and a rapid shift in metabolism to glycolysis and oxidative phosphorylation are necessary for T cell and keratinocyte activation and proliferation, both of which play a major role in CLE pathomechanism. However, glucose metabolism in CLE is yet unknown. Here, we seek to test the hypothesis that elevated GLUT1 transporter expression and, consequently, increased intracellular glucose availability correlates with CLE disease activity.

Materials & Methods:

Skin biopsy samples were collected from 51 patients with active, untreated CLE lesions at the Department of Dermatology, Venereology, and Dermatooncology, Semmelweis University, Budapest, Hungary. GLUT1 and MxA (Mx1) expression were evaluated using immunohistochemistry, while gene set enrichment analysis (GSEA) was performed to identify immunometabolism-related transcriptional changes. The CLASI score was used to determine disease activity.

Results:

Significantly increased GLUT1 expression was observed in keratinocytes in CLE compared to healthy control. A significant positive correlation was observed between keratinocyte GLUT1 expression and both keratinocyte and lymphocyte MxA expression in CLE samples ($p=0.004$, $r=0,394$; $p= <0.001$, 0.466). Furthermore, a significant positive correlation was observed between the CLASI score and keratinocyte GLUT1 expression (<0.001, $r=0.47$). Consistently, GSEA revealed that inflammatory pathways were upregulated in CLE skin samples with high GLUT1 expression.

Conclusion:

To our knowledge, this is the first study to assess the impact of glucose metabolism on CLE disease activity. The observed upregulation of GLUT1 expression in CLE skin and its correlation with MxA expression and the CLASI score may suggest a potential role of glucose metabolism in the immunopathogenesis and clinical activity of CLE.

Abstract N°: 8119

Localized Scleroderma (Morphea) Associated with Borrelia burgdorferi infection: A Case Report

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Introduction & Objectives: Localized scleroderma, or morphea, is a rare, chronic, inflammatory skin disorder characterized by dermal fibrosis. While the precise etiology remains unclear, infectious triggers—particularly *Borrelia burgdorferi*—have been implicated in some cases. This case report aims to present a clinically and histologically confirmed case of morphea in a young adult patient with positive serology for *B. burgdorferi*, highlighting the diagnostic process and therapeutic outcomes.

Materials & Methods: A 45-year-old male patient was referred to our Department with hyperpigmented lesions and significant pruritus. Dermatological examination revealed hyperpigmented plaques with centrally palpable induration on the trunk and left inguinal region, along with palmar and plantar hyperkeratosis. Skin biopsy was performed and histopathological sample (orcein histochemical staining was performed to evaluate elastic fibers) confirmed dermal fibrosis consistent with morphea. Serological testing for Borrelia burgdorferi antibodies (IgG, IgM) was repeatedly positive over a 12-month follow-up period. Therapeutic interventions included systemic antibiotic therapy, topical corticosteroids and PUVA cream photochemotherapy.

Results: Histopathology revealed dermal fibrosis consistent with localized scleroderma. Persistent positive B. burgdorferi serology supported a potential infectious etiology. Significant clinical improvement was achieved with the multimodal treatment approach, as demonstrated by a notable reduction in pruritus and partial regression of skin lesions.

Conclusion: This case supports the hypothesis that *Borrelia burgdorferi* may play a role in the pathogenesis of morphea in certain individuals. The positive clinical response to antibiotic therapy further emphasizes the potential benefit of addressing underlying infections in affected patients. Continued serological monitoring and multidisciplinary treatment approaches are recommended. Further prospective studies are needed to clarify the relationship between *B. burgdorferi* infection and localized scleroderma and to develop standardized treatment protocols.

Abstract N°: 8163

Refractory mucocutaneous lesions in SLE: rapid and sustained response to anifrolumab

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Introduction & Objectives:

Mucocutaneous lesions are a major unmet need and therapeutic challenge in systemic lupus erythematosus (SLE), often leading to visible disfigurement and poor quality of life, even in well-controlled systemic disease. Anifrolumab is a monoclonal antibody targeting the type I interferons (IFN) receptor subunit 1 (IFNAR1), thereby blocking the downstream type I IFN signalling cascade, which is critically overactivated in SLE. Phase III trials demonstrated anifrolumab efficacy in reducing SLE activity, however, real-world data remain limited, particularly in refractory and severe cutaneous LE (CLE).

Materials & Methods:

We present a 41-year-old female with SLE initially diagnosed at age 12, with mucocutaneous lesions, arthritis, pancytopenia, and class IV lupus nephritis, complicated with TEN-like SLE. She was treated with intravenous pulse and continuous oral glucocorticoids, hydroxychloroquine, cyclophosphamide, azathioprine, and more recently mycophenolate mofetil (MMF), achieving good control of systemic manifestations, but with persistent mucocutaneous involvement, encompassing malar rash, discoid and vasculopathic skin lesions, and oral ulcerations. Intravenous 300 mg of anifrolumab every four weeks was started alongside MMF and prednisone, while hydroxychloroquine was discontinued due to macular degeneration.

Results:

At baseline, Cutaneous Lupus Erythematosus Disease Area and Severity Index activity score (CLASI-A) was 22, damage score (CLASI-D) 2 and SLEDAI-2K score 8 (inflammatory type rash, oral ulcerations, low-positive anti-dsDNA and low C4). Rapid clinical improvement was demonstrated, starting from week 4 of anifrolumab initiation, with CLASI-A reduced to 11. By week 16, almost complete resolution of lesions was achieved, with CLASI-A reduced to 2 and SLEDAI-2K to 0. Currently, at week 48, remission is sustained (CLASI-A=2, CLASI-D=2, SLEDAI-2K=4 due to low-positive anti-dsDNA and low C4), with tapering of prednisone from 30 mg to 5 mg on alternate days, and MMF from 2 g to 1 g/day. The only notable adverse event was an episode of herpes labialis.

Conclusion:

Our case supports the findings of phase III clinical trial TULIP-2, where 49% of patients receiving anifrolumab achieved ≥50% CLASI reduction by week 12, compared to 25% in the standard care group. The mucocutaneous efficacy of anifrolumab may be attributed to the more pronounced type I IFN response in the skin compared to other tissues. In LE, this response is further amplified by defective clearance of cell debris and accumulation of plasmacytoid dendritic cells, primary IFN-I producers, in lupus-affected skin. Consistent with larger studies, we did not observe significant variations in SLE serological parameters. Despite demonstrated efficacy and safety, with mild upper respiratory tract infections and herpes zoster as the most common adverse events, current approval excludes CLE without systemic involvement. Rapid clinical improvement that prevents permanent damage of CLE, and the reduction of glucocorticoids and immunosuppressants doses, minimizing their side-effects, are key benefits of anifrolumab. To maximize these benefits, our case and emerging evidence from other cases underscore

the importance of early initiation of anifrolumab, and the need for expanded indications to include CLE without systemic disease.

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Abstract N°: 8176

Serum Chemokine Profiling Reveals Elevated CXCL9 and CXCL10 in Lichen Planus

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Introduction & Objectives:

Lichen planus (LP) is an idiopathic chronic inflammatory dermatological condition characterized by violaceous papules and plaques, often accompanied by pruritus, affecting skin, mucous membranes, hair, and nails. LPs pathophysiology is poorly understood but involves T-cell-mediated cytotoxicity, driven by Th1 and IL-23/Th17 pathways. It is theorized that exposure to agents such as viruses, drugs, or allergens may trigger cross-reactions with self-antigens in basal keratinocytes, leading to immune activation. This process is marked by damage signals, such as S100A8/A9, which initiate interferon-α release from dendritic cells and cytokine production, amplifying immune cell infiltration.

Materials & Methods:

To elucidate the systemic immunological mechanisms underlying LP pathophysiology, a case-control study profiled serum chemokines in 8 LP patients and 41 age- and sex-matched non-itch controls. Plasma samples were collected to assess circulating cytokine levels, offering a systemic perspective beyond localized tissue analysis. Cytokine expression was analyzed, and differences in serum levels between LP patients and controls were evaluated using the Wilcoxon rank-sum test. False Discovery Rate (FDR) correction, applied via the Benjamini-Hochberg procedure, identified significant cytokines at FDR < 0.05. Welch's t-test assessed age differences, while chi-squared tests evaluated variations in sex and race distributions to ensure comparable baseline demographics.

Results:

The average age of LP patients was 53.5 ± 20.82 years, with 6 out of 8 patients being female. No significant demographic differences were found compared to controls. CXCL9 (MIG) (mean ± SD [pg/mL]: 11745.7 ± 9717.4 LP vs. 2830.7 ± 1534.7 HC, $p < 0.01$) and CXCL10 (IP-10) (mean ± SD [pg/mL]: 662.1 ± 706.7 LP vs. 132.9 ± 90.1 HC, $p < 0.01$) were significantly elevated in LP patients' serum compared to controls.

Conclusion:

Interferon-γ-induced chemokines, CXCL9 and CXCL10, are critical for T-cell recruitment, aligning with LP's T-cell-driven pathogenesis. Previous studies identified CXCL9 as a key marker in LP gene expression profiles and CXCL10 as a mediator of T-cell migration to LP lesions, secreted by fibroblasts and basal keratinocytes. The elevation of CXCL9 and CXCL10 suggests their pivotal role in sustaining the inflammatory cascade in LP, making them promising targets for novel therapies. Inhibiting these chemokines or their receptors, such as CXCR3, could modulate T-cell infiltration and reduce inflammation, offering potential treatment avenues. These findings highlight CXCL9 and CXCL10 as key players in LP pathophysiology and as viable biomarkers for developing targeted immunomodulatory therapies, warranting further research into their therapeutic applications in immune-mediated dermatological diseases.

Abstract N°: 8273

Dermatomyositis with positive anti-TIF1 antibody: Severe form About a case

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Introduction & Objectives: Dermatomyositis is a chronic autoimmune disease that manifests with cutaneous and muscle involvement. Its strong association with neoplasms and lung involvement constitute the severity of the disease. We report the case of a patient with severe dermatomyositis with a positive anti-TIF1 antibody without neoplasia and with a fatal outcome.

Materials & Methods: This is a 60-year-old patient with no particular past medical history, hospitalized for the management of dermatomyositis. He had significant muscle weakness. On examination, he presented with bilateral periorbital erythema and edema, a V-sign rash on the décolletage, erythema of the upper back and the extensor surfaces of the arms (shawl sign). He also presented with Gottron's papules and erythema, notably at the metacarpophalangeal joints. The Holster sign was positive, characterized by erythema of the extensor surfaces of the thighs. He also had mechanic's hands. Paraclinical investigations revealed an inflammatory syndrome, elevated muscle enzymes, and a positive anti-TIF1 antibody. Electromyography showed myopathic involvement of all four limbs, predominantly in the proximal muscles of the upper limbs, with active features. Muscle biopsy showed inflammatory myositis. Paraneoplastic workup was negative with a normal CT scan, showing no evidence of progressive malignant lesions. The patient was started on corticosteroid therapy at 1mg/kg/day, with adjuvant treatment of methotrexate 15mg/week. The initial course was marked by clinical improvement and normalization of muscle enzymes. The patient experienced a worsening of his symptoms after 5 months, leading to his death.

Results: Dermatomyositis (DM) is an inflammatory disease with constant cutaneous involvement and inconstant muscle involvement, predominantly affecting the limb girdles. DM affects all age groups, with a clear female predominance (male-to-female ratio of 1.5 to 2/1). The age of onset is often later in men compared to women, who have a higher risk of association with neoplasms. The form associated with cancer is more frequent in older individuals. Age over 45 years, male sex, dysphagia, skin necrosis, the presence of vasculitis on histology, elevated erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP), and the extent of muscle involvement are criteria associated with the risk of cancer. Anti-transcriptional intermediary factor 1 (anti-TIF1) antibodies are associated with an increased risk of cancer. The choice of treatment in DM is related to the severity of the disease. Corticosteroids have revolutionized the prognosis, with mortality reduced from 50 to 14%.

Conclusion: Dermatomyositis can precede a neoplasm, accompany it, or be revealed by it. Predictive factors for an association with cancer are: age over 50 years, male sex, presence of skin necrosis, presence of erythroderma, severity of muscle involvement, and positive anti-TIF1 antibody. The presence of anti-TIF1 antibodies in a patient with DM warrants a systematic and extensive screening for cancer, which should be repeated if the initial workup is negative. Advanced age, malignancy, dysphagia, and muscle involvement are associated with an increased risk of mortality.

Abstract N°: 8275

Clinical, Dermoscopic, and Therapeutic Insights into Pemphigus Vegetans: A rare case report

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Introduction & Objectives:

Pemphigus vegetans is a rare variant of pemphigus, characterized by predominant flexural involvement and vegetating lesions. This study reports a new case, highlighting its clinical, dermoscopic, and therapeutic specificities.

Case report:

A 43-year-old woman, with a history of cholecystectomy 11 years prior, was hospitalized for flaccid bullous lesions evolving into erosive and vegetating plaques, primarily in flexural areas over three months. Initial painful oral involvement impaired nutrition, accompanied by moderate pruritus, chronic constipation, rectal bleeding, and general deterioration.

Clinical examination revealed post-bullous erosions with an erythematous and bleeding base, overlying healthy skin, and topped by coalescing, malodorous, brownish, papillomatous vegetations in axillary, submammary, inguinal, intergluteal, suprapubic, and vulvar folds, as well as the neck, occipital scalp, mid-back, and posterior thighs.

Pustules were present in some areas, with flaccid serous bullae on the upper back and perioral region. Nikolsky's sign was positive. Pigmented macular scars were noted on the posterior thighs, along with a diagonal brownish hypochondrial scar. Oral examination revealed erosive gingivitis, fissured crusted cheilitis, painful buccal and lingual ulcerations without pseudomembranes or synechiae. Anal pseudo-marqués and a large anal fissure were confirmed via rectoscopy. Nails were normal.

Dermoscopy of the vegetative lesions revealed a brown cobblestone pattern with pink-white lacunae, some scattered dotted and linear vessels, whitish scales and darkish browner areas.

Histopathology revealed suprabasal acantholysis, hyperparakeratotic acanthosis, and intraepidermal IgG/C3 deposition on direct immunofluorescence. Indirect immunofluorescence was strongly positive. Paraneoplastic screening was negative. Bacterial culture identified Pseudomonas aeruginosa and Staphylococcus aureus both ciprofloxacin-sensitive.

Treatment included corticosteroids (1.5 mg/kg/day), azathioprine (150 mg/day), local wound care, and antibiotic/antiviral therapy. Evolution showed no new lesions, healing of existing erosions, regression of vegetations, and residual hyperpigmented macules.

Conclusion:

Pemphigus is an autoimmune blistering dermatosis; the vegetative form represents 1 to 2% of cases. It manifests as flexural bullae progressing to vegetating plaques. Diagnosis relies on histology and immunofluorescence. Although rarely described, dermoscopy may help correlate vascular structures with disease activity and the presence of scales with the vegetative form. Treatment is based on corticosteroids, immunosuppressants, and potentially rituximab.

Abstract N°: 8353

A Case of Refractory Bullous Pemphigoid: Good Response to Omalizumab

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Introduction & Objectives:

Bullous pemphigoid (BP) is the most common autoimmune blistering dermatosis, accounting for 70% of all subepidermal autoimmune blistering diseases. The pathogenic role of IgE in BP has been demonstrated. Elevated IgE levels are frequently observed in the serum of patients or at the dermoepidermal junction. Several reports have described the effectiveness of omalizumab, an anti-IgE monoclonal antibody, in the treatment of BP. We report a case of corticosteroid-resistant BP that was successfully treated with omalizumab, leading to significant clinical improvement.

Materials & Methods:

A 72-year-old woman with a history of type 2 diabetes, hypertension, atrial fibrillation, and thyroidectomy was hospitalized for a blistering dermatosis evolving over one month. Dermatological examination revealed multiple tense, pruritic bullous lesions with a negative Nikolsky sign, mostly on an erythematous and urticarial-like base. Lesions were located on the trunk, face, and limbs. There was also involvement of the oral mucosa, with erosions affecting the palate and buccal mucosa. A skin biopsy and ELISA confirmed the diagnosis of BP (subepidermal split and positive anti-BP180 and BP230 antibodies). Laboratory investigations showed normal eosinophil count and elevated total IgE levels at 442.7 ng/ml (normal < 240 ng/ml). Oral corticosteroid monotherapy 1 mg/kg/day was ineffective. The addition of methotrexate (12.5 mg/week) led to hematologic side effects and digestive intolerance, requiring its discontinuation. Due to persistent disease activity, worsening severity scores (ABSIS and BPDAI), and the onset of bacterial pneumonia, treatment with omalizumab (300 mg every 15 days subcutaneously) was initiated, along with a gradual tapering of corticosteroids. The clinical course rapidly improved, with clear signs of efficacy after the second injection (no new lesions or pruritus, and early healing of erosive lesions). Complete clinical remission was achieved after 4 months of treatment, with excellent clinical and biological tolerance.

Results:

BP typically affects elderly patients with multiple comorbidities. This frail context often necessitates the use of immunosuppressive therapies. However, their delayed onset of action and iatrogenic effects make them ineffective or even dangerous in urgent situations. In such cases, immunomodulatory therapies like omalizumab should be considered as a preferred option. Omalizumab is a humanized monoclonal antibody that selectively binds to IgE, thereby decreasing free IgE levels. IgE may play a central role in the pathophysiology of BP, either by recognizing—like IgG—the extracellular domain of collagen XVII, or by promoting eosinophil recruitment via mast cell degranulation. By binding to IgE, omalizumab prevents their attachment to high-affinity class I receptors and inhibits the inflammatory cascade. Omalizumab appears to induce a rapid clinical response in BP, especially in forms with intense pruritus, pseudo-urticarial plaques, peripheral eosinophilia, and elevated IgE levels. Furthermore, unlike immunosuppressants, it has an excellent safety profile, making it an attractive treatment option in BP, as demonstrated by our patient’s case.

Conclusion:

IgE seems to play a key role in the pathophysiology of bullous pemphigoid, which explains the effectiveness of omalizumab, particularly in patients with severe forms, significant eosinophilia, and high IgE levels.

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Abstract N°: 8384

A rare presentation of keloidal morphea in a 7- year old girl

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Introduction & Objectives:

Morphea is a localized autoimmune, inflammatory skin disorder characterized by hardening of the skin due to sclerotic changes of the skin and subcutaneous tissue.

Keloidal morphea, a seldom encountered variant of morphea is characterized by spontaneous keloid like scars predominantly found on neck, trunk and extremities. It is more common in females than in males. Here in, we report a case of keloidal morphea in a 7-year-old girl.

Materials & Methods:

A -7- year- old girl was brought to outpatient department of dermatology with complaints of dark colored swellings over her back which were first noticed by the girl's mother when the girl was 6 months old. Initially the swellings presented as pea sized nodules which gradually enlarged in size and coalesced to form linear band like scars. The swellings however were asymptomatic except for mild itching. Progressive increase in their size over a period of 6 months was reported by the girl's mother. There is no history of preceding trauma, familial tendency and irradiation at the site. Patient's attender denied any symptoms of morning stiffness, Raynaud's phenomenon, difficulty in breathing, dryness and difficulty in opening of mouth. On cutaneous examination two hyperpigmented, parallelly arranged linear scars which were firm to hard in consistency on palpation, measuring roughly about 3 to 4 cm in length were present over right mid scapular region with induration of the underlying skin. Few irregularly shaped hyperpigmented nodules were found at the periphery of the lesions. A provisional diagnosis of keloidal morphea was made and serum anti-nuclear antibody test with profile was advised and a punch biopsy was performed to confirm the diagnosis.

Results:

The serum anti- nuclear antibodies test was negative however skin biopsy showed evidence of keloidal morphea with papillary dermis and subcutis showing thickened collagen bundles with a haphazard arrangement. perivascular mononuclear cell infiltrates and loss of peri-appendageal fat were the remarkable findings. Epidermis showed mild acanthosis and elongation of rete ridges. The patient is currently undergoing treatment with monthly intralesional injections of Triamcinolone acetonide.

Conclusion:

Keloidal morphea or nodular Morphea is a rare variant of Morphea with less than 50 cases reported in English literature. The salient features in this case are the lack of systemic involvement and the presentation in childhood, which is relatively rare. This could raise a possibility that keloidal or nodular Scleroderma could be potentially underdiagnosed.

Abstract N°: 8386

Bullous Pemphigoid in Focus: A Case Series Review of Diagnostic and Therapeutic Insights

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Introduction & Objectives:

Bullous pemphigoid is a subepidermal autoimmune bullous dermatosis. It is the most common acquired autoimmune bullous disease. Its incidence has increased in recent decades due to the aging of the population. Treatment remains difficult due to the advanced age of patients and frequent comorbidities. The aim of our work is to study the epidemiological, clinical, therapeutic and evolutionary profile of bullous pemphigoid in our department.

Materials & Methods:

This is a retrospective descriptive study including all patients hospitalized in our dermatology department for confirmed bullous pemphigoide, over a 10-year period from January 2013 to January 2023.

Results:

We enrolled 87 patients with bullous pemphigoide, with a clear female predominance. The sex ratio was M/F = 0.4. The mean age of our patients was 63.8 years, with extremes ranging from 6 to 96 years. Phototype ranged from III to IV. 62% of patients were of urban origin. Socioeconomic status was considered low in 73% of cases. Association with neuro-psychiatric pathologies was found in 10 cases (11%), with 5 cases of stroke (5.74%), 3 cases of psychic disorders (3.44%), 1 case each of dementia and epilepsy (1.14%). Other comorbidities included arterial hypertension in 27 cases (31%), diabetes in 15 cases (17%) and chronic renal failure in 2 cases (2%). Pruritus was almost constant in over 96.5% of cases. Mucosal involvement was present in 40% of cases. The BDPAI activity score was moderate in 51% of cases, mild in 31% and severe in 18%. Clinical forms in our series were predominantly bullous pemphigoide (76%), followed by pemphigoide gestationis (13%), then cicatricial pemphigoide (8%), and 3 cases of pemphigoide of children (3%). Hypereosinophilia was noted in 56 patients (64%). Complications related to the disease included skin superinfection (56%) and urinary tract infections (27.58%). Septic shock was noted in only one patient. Only one death was recorded. In terms of treatment, local corticosteroids of the strong class alone (20 to 30 g/d) were prescribed in 47 cases (54.02%). Oral corticosteroid therapy (0.5 to 1.5mg/kg/d) was administered in 74 patients (85%). The addition of an immunosuppressant was indicated in 12 cases (13.8%), with 2 cases receiving methotrexate (5 to 20mg/week) and 9 cases cyclophosphamide (1g bolus with an average of 3 boluses). Dapsone (50 to 100 mg/d) was prescribed in 7 patients. The average length of hospital stay was 20 days. The average cost of each hospitalization was 5480 DH.

Conclusion:

Bullous pemphigoid is typically a chronic disease with unpredictable relapses. Its mortality rate is higher because of the elderly population. The faster and better the treatment, the better the prognosis. Hence the importance of establishing national recommendations to codify management.

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Abstract N°: 8434

Population-based Mortality in Dermatomyositis: A Systematic Review and Meta-Analysis

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Introduction & Objectives:

Dermatomyositis (DM) is a rare inflammatory myopathy with hallmark skin findings and muscle weakness. Mortality in DM varies with comorbidities and disease subsets, influenced by factors such as older age at onset, delayed therapy after onset of myopathy, pulmonary disease, malignancy, and infections. Despite this, no prior meta-analysis has focused solely on population-based mortality rates. This systematic review and meta-analysis aimed to quantify the standardized mortality ratio (SMR) in patients with DM.

Materials & Methods:

Following PRISMA guidelines, we conducted a systematic review and meta-analysis registered on PROSPERO (CRD420250633407). We searched MEDLINE, EMBASE, and Cochrane databases from inception to January 2025 for population-based studies reporting mortality in DM. Two reviewers independently screened articles, extracted data, and assessed study quality using the Joanna Briggs Institute checklist. We conducted a random-effects meta-analysis to generate pooled SMRs and assessed heterogeneity using I^2 statistics. Publication bias was evaluated with funnel plots and statistical tests. The GRADE framework was used to rate the certainty of evidence.

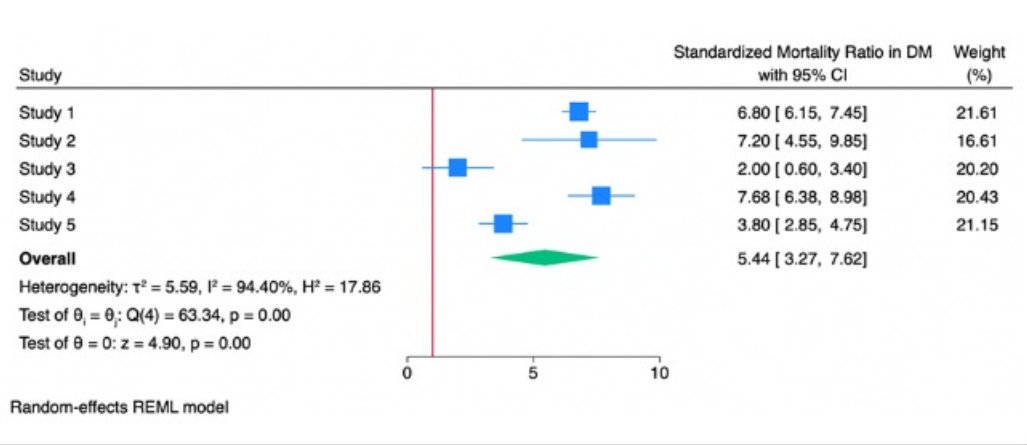
Results:

Of 304 records screened, 12 studies (14,823 DM patients, 42,000,124 controls) were included in the systematic review, and 5 in the meta-analysis. Studies were of high methodological quality. Pooled analysis revealed a significantly elevated mortality risk in DM (SMR 5.44; 95% CI 3.27–7.62; I^2 = 94.4%, Figure 1). Funnel plots showed no significant publication bias (Begg's p=0.46; Egger's p=0.74). Narrative synthesis confirmed an association between DM and mortality (twelve studies, high certainty) (Table 1). There is likely an association between increased mortality in females compared to males with DM (three studies, moderate certainty). There may be an association between increased mortality in Black patients compared to controls (one study, low certainty). Patients with cardiovascular comorbidities and DM had higher mortality than controls with similar comorbidities, suggesting an additive risk. One study also reported declining mortality over time, likely reflecting earlier diagnosis and improved therapeutic strategies.

Conclusion:

Overall, these findings reinforce the well-established link between DM and mortality, while highlighting the influence of sex and racial disparities. This is the first meta-analysis to quantify population-based mortality in DM, reporting a fivefold increase in mortality risk. Future research should explore underlying mechanisms and develop targeted interventions to improve outcomes in high-risk populations with DM.

Figure 1: Meta-analysis of pooled standardized mortality ratio in dermatomyositis


Study
Standardized Mortality Ratio in DM with 95% CI
Weight (%)
Study 1
6.80 [6.15, 7.45]
21.61
Study 2
7.20 [4.55, 9.85]
16.61
Study 3
2.00 [0.60, 3.40]
20.20
Study 4
7.68 [6.38, 8.98]
20.43
Study 5
3.80 [2.85, 4.75]
21.15
Overall
5.44 [3.27, 7.62]
Heterogeneity: τ² = 5.59, I² = 94.40%, H² = 17.86
Test of θi = θj: Q(4) = 63.34, p = 0.00
Test of θ = 0: z = 4.90, p = 0.00
0
5
10
Random-effects REML model

Abstract N°: 8483

an unusual presentation of pemphigoid gestationis: a case report

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Introduction & Objectives:

Pemphigoid gestationis (PG), formerly herpes gestationis, is a rare, self-limiting, autoimmune subepidermal bullous dermatosis of pregnancy that shares some clinical and pathogenic features with bullous pemphigoid. The incidence of PG is variably estimated, ranging from 1 out of 2,000 to 60,000 pregnancies, with most cases occurring in the second and third trimesters. The condition is more prevalent in multiparous than primiparous women, commonly recurring in subsequent pregnancies. PG typically presents during the third trimester, though it can present during any trimester or the postpartum period with inflammatory skin lesions and severe pruritus. The skin lesions commonly begin in the periumbilical area (characteristic eruption site) and spread across the abdomen to the extremities. Treatment of PG depends on the severity and the stage of the skin lesions. The main goal is to relieve itching and prevent the formation of new blisters. The mainstay of treatment is topical high-potency corticosteroids and antihistamines. However, additional therapies may be needed for patients unresponsive to initial therapy. Fetal risks have also been observed in pregnancies with PG, including fetal growth restriction, preterm labor, and temporary neonatal skin lesions, which usually resolve several weeks after birth. This risk to pregnancy appears to be correlated with disease severity; however, no increased risk of stillbirth or miscarriage has been observed.

Materials & Methods:

Case report

Results:

We describe a case of a pregnant patient with severe PG, including an unusual initial palmoplantar presentation. A 34-year-old woman developed pruritus and palmoplantar exanthema at week 30 of her first pregnancy. After starting topical corticosteroids daily, the patient reported complete resolution of the lesions. Generalized pruritus was persistent. Within hours after her first delivery (preterm labor at week 37), pruritic reddish raised urticarial lesions and large fluid-filled blisters appeared over the abdomen and limbs. The diagnosis of PG was confirmed by skin biopsy and direct immunofluorescence. Her baby boy was healthy and did not show any skin lesions. The patient was started on oral prednison 60 mg daily along with supportive measures. New blistering appeared in the following weeks, so we decided to add another immunosuppressive agent. Azathioprine was chosen as an additive treatment. This treatment regimen resulted in complete resolution of skin lesions within a 4 months.

Conclusion:

PG is a rare disease requiring fast and accurate diagnosis and therapy. Due to the rarity of the disease both treatment and diagnosis can be challenging.

Abstract N°: 8532

Calcinosis Cutis: A Case Report.

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Introduction & Objectives:

Calcinosis cutis is characterized by calcium salt deposition in the skin and subcutaneous tissues, forming firm nodules that vary in location depending on the underlying condition. These nodules may ulcerate and discharge chalky material. Histologically, they show basophilic, granular or amorphous calcium deposits with foreign body reactions, including histiocytes and multinucleated giant cells.

There are five subtypes: dystrophic, metastatic, idiopathic, iatrogenic, and calciphylaxis. Contributing conditions include autoimmune connective tissue diseases, chronic kidney failure, malignancies, genodermatoses, trauma, infection, and calciphylaxis.

Materials & Methods:

A 57-year-old woman presented with unilateral firm nodules in the femoral region and lymphadenopathy, which had developed over several months. Examination showed scar tissue and hyperpigmentation, suggesting prior ulceration and chronicity.

A skin biopsy was performed on a nodule near the right sacroiliac region. Blood tests were ANA-positive but negative for systemic lupus erythematosus, dermatomyositis, and systemic sclerosis. Thyroid and PTH levels were normal; serum calcium was elevated. CMV IgG was positive, and urine calcium was normal.

Ultrasound showed enlarged lymph nodes in the right inguinal area. CT imaging of the chest and abdomen identified lymphadenopathy in the axillary and inguinal regions, along with hepatosplenomegaly. PET-CT demonstrated increased metabolic activity in diaphragmatic lymph nodes, prompting further investigation. Mammography and Pap smear were negative. Histological analysis confirmed calcinosis cutis in the skin lesion. A subsequent biopsy of an inguinal lymph node established a diagnosis of Hodgkin lymphoma.

Results:

Multiple potential causes for calcinosis cutis were considered. Extensive hematologic and imaging evaluations were conducted, but definitive diagnosis was achieved through biopsy—especially of the enlarged lymph node.

Conclusion:

This case emphasizes how a dermatological finding can be the first sign of an underlying systemic disease. A multifactorial condition like calcinosis cutis requires focused diagnostic steps to rule out causes, with biopsy often being the decisive tool. Prompt recognition and interdisciplinary cooperation enable earlier diagnosis and more effective management, particularly when malignancy is involved.

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Abstract N°: 8536

Sleep, IL-10, and vitamin D: exploring the interplay between these components in the immunoregulation of vitiligo

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Introduction & Objectives: Vitiligo is an autoimmune skin disorder closely related to psychological distress. Sleep disturbance, as a stressful condition, may be a risk factor for vitiligo worsening, potentially through mechanisms involving immune dysregulation. Vitamin D levels and inflammatory cytokines can interact and influence both immune pathways implicated in vitiligo and sleep regulation. The interplay between circadian-influenced cytokines, vitamin D, sleep, and vitiligo remains unclear. This study aimed to investigate the potential immunological connections between sleep disturbances and vitiligo, with a particular focus on vitamin D and inflammatory cytokines.

Materials & Methods: Individuals were attended at the Dermatology Service of Federal University of São Paulo (UNIFESP). A total of 28 adults with clinically diagnosed vitiligo was included in this study. Serum components were measured, including 25-hydroxy-vitamin D [25(OH)D] and immune cytokines – interleukin (IL)-4, IL-6, IL-10, IL-17A, IFN-γ and TNF-α. Two sleep questionnaires were accessed: Pittsburgh Sleep Quality Index (PSQI) and Insomnia Severity Index (ISI). PSQI score>5 means poor sleep, and ISI>7 suggests clinical insomnia. Clinical variables considered for vitiligo were previous phototherapy, signs of skin activity, mucosal and acral lesions. For statistical analysis, T-test and Mann-Whitney test were used.

Results: Data from 28 patients with vitiligo were analyzed. The mean age was 46.92 years-old. Most of the sample were women (n=20). Regarding sleep, 24 individuals were classified as having poor sleep (PSQI), whereas only 13 patients reported insomnia symptoms (ISI). In the analysis of numerical variables (serum components, age, and disease duration in months) according to PSQI classification, IL-10 levels were significantly increased in poor sleepers (2.31 pg/mL) compared to normal sleepers (0.98 pg/mL). The 25(OH)D levels were higher in poor sleepers (24.63 ng/mL) than in normal sleepers (17.87 ng/mL). Considering ISI, none of the components were statistically significant. In the analysis of the variables with skin activity (17 individuals with vitiligo activity), IL-10 levels were significantly elevated in patients without active lesions (2.66 pg/mL) compared to those with skin activity (1.77 pg/mL). Vitiligo duration was higher and statistically significant in the sample with skin activity (198.35 vs. 88.09 months). The analysis of these variables with previous phototherapy, and presence of mucosal or acral lesions were not significant.

Conclusion: Higher levels of IL-10 and vitamin D were found in vitiligo patients with poor sleep quality, which may be influenced by the larger representation of this subgroup in the overall sample. This suggests a possible interaction between sleep disturbances and immunomodulatory mechanisms in vitiligo. Increased IL-10 levels were detected in individuals without active lesions, which may reflect a compensatory anti-inflammatory response associated with sleep-related immune imbalance. As an anti-inflammatory cytokine with circadian influence, IL-10 may initially increase to counteract inflammatory processes induced by poor sleep. These findings support further investigation of IL-10 and vitamin D as potential biomarkers for both sleep quality and vitiligo activity, and underscore the importance of sleep as a modulator of immune function in autoimmune skin disorders.

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Abstract N°: 8544

Linear IgA Bullous Dermatosis Associated with Malignancy: A Case Report

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Introduction:

Linear IgA bullous dermatosis (LABD) is an autoimmune subepidermal vesiculobullous eruption that can affect children and adult1. It occurs due to the linear deposition of IgA autoantibodies against antigens located in the basement membrane zone 2.

The disease is predominantly idiopathic, although several associations have been reported, including drugs, autoimmune disorders, inflammatory bowel diseases, and malignancies 2.

We present a case of paraneoplastic linear IgA bullous dermatosis in an adult patient.

Case report:

A 65-year-old male presented with a one-year history of erythematous papules and plaques, some exhibiting peripheral vesiculation, predominantly distributed over the extensor surfaces of elbows, knees, the gluteal region, and the medial thoracic back. He had a medical history of systemic arterial hypertension and diabetes mellitus and was undergoing investigation for a right renal nodule. A cutaneous biopsy showed a subepidermal bullous dermatosis with a predominance of neutrophils. Direct immunofluorescence demonstrated linear IgA deposition along the basement membrane zone, confirming the diagnosis of linear IgA bullous dermatosis.

A few months after the diagnosis, the patient underwent a right nephrectomy, and histopathological examination confirmed the diagnosis of clear cell renal cell carcinoma. Following tumor excision, significant improvement of the cutaneous lesions was observed.

Discussion:

LABD is a rare subepidermal bullous disorder characterized by the deposition of antibodies against basement membrane antigens, such as LABD97, laminin 332, and type VII collagen 1,3. It shows two incidence peaks: in children between 2 and 6 years old, and in adults in their sixth decade of life [1]. In the present case, the patient was within the most common age group, at 65 years old.

The disease has been associated with immunological triggers, including drugs, intestinal inflammation, infections, and vaccines [1]. More rarely, it is associated with malignancies, with hematological disorders being more commonly than solid organ neoplasms [4]. In the described case, the patient had clear cell renal cell carcinoma, with no other evident immunological triggers.

The clinical presentation is variable, ranging from erythematous and urticarial plaques, tense blisters, to annular lesions with peripheral vesiculation, presenting a ‘pearl necklace’ or rosette-like appearance, which, although characteristic, is not pathognomonic[5]. In adults, lesions typically affect the trunk, extremities, face, often involving extensor surfaces, as seen in dermatitis herpetiformis 6. The clinical presentation in this case is consistent with descriptions in the literature, particularly the predilection for extensor areas.

Complementary exams are essential for diagnosis. Histopathology shows subepidermal vesicles with neutrophils, while direct immunofluorescence reveals linear IgA along the basement membrane[5]. In the present case, the complementary exams were consistent with these findings and were crucial for the diagnosis.

Conclusion:

LABD is a rare autoimmune disorder, usually idiopathic, but it can be associated with malignancies, as demonstrated in this case of clear cell renal cell carcinoma. This case highlights the importance of considering solid organ tumors as a potential underlying cause in adult patients with LABD. Early recognition of this association is essential for timely diagnosis and optimal management.

Abstract N°: 8552

Case report: Therapeutic Challenges of adult-onset refractory Discoid Lupus Erythematosus with comorbid rosacea

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Introduction & Objectives:

Discoid lupus erythematosus (DLE) is a predominant subtype of cutaneous lupus erythematosus (CLE), presenting as a chronic inflammatory cutaneous pathology with discoid lesions. Originally DLE lesions present as violaceous macules or papules, later evolving into scaly plaques concurrent with peripheral hyperpigmentation. The treatment course is gradual: originally treated with topical corticosteroids, if unresponsive, hydroxychloroquine is introduced, followed by immunosuppressants in refractory cases. The case report aims to outline a therapeutically challenging case of DLE, clinically marked by resistance to conventional treatments due to comorbidly occurring rosacea and demodicosis. This case report highlights the multidisciplinary therapeutic challenges in refractory DLE and acknowledges the negative impact of comorbid inflammatory conditions to successful treatment.

Materials & Methods:

Case report.

Results:

The 54-year-old female presented with persistent facial erythema and pustules in the central facial region. Initial topical treatment with metronidazole was ineffective, oral doxycycline yielded only a modest response. Punch biopsy was performed and DLE was histologically confirmed, observing superficial and deep perivascular and periadnexal dermatitis and thickened PAS+ basement membrane. Direct immunofluorescence revealed IgG, IgM, and C3 linear deposition in the basal membrane. Following diagnosis, the patient alternated between topical treatments, including the calcineurin inhibitors tacrolimus and pimecrolimus, as well as clobetasol, with no significant clinical improvement. Hence, hydroxychloroquine was initiated at 200 mg/d for 1 month, but the skin condition worsened. Therefore, a secondary punch biopsy was performed, disclosing granulomatous dermatitis and establishing the diagnosis of DLE concomitant with granulomatous rosacea. Microscopic evaluation of the skin scrape revealed a positive result for Demodex mite. Combined treatment with doxycycline 40mg/day and prednisolone 40 mg/day was initiated with limited response. Then, treatment with methotrexate 10 mg/week and topical Ivermectin was initiated, resulting in the gradual remission of DLE.

Conclusion:

This case urges the importance of recognising treatment-resistant DLE due to comorbid granulomatous rosacea and demodicosis: the disease can present with refractory behaviour and not respond to immunomodulators and topical corticosteroids. The case highlights how coexisting rosacea exacerbates and prolongs DLE symptoms, emphasizing the need for a multifaceted treatment approach and a more escalated treatment strategy. This instance underscores how refractory DLE can be effectively managed with immunosuppressants (methotrexate) and anti-parasitic Ivermectin for comorbid rosacea, when conventional therapies fail.

Abstract N°: 8655

Successful management of obesity and metabolic implications in patients with hidradenitis suppurativa

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Introduction & Objectives:

Hidradenitis suppurativa is a chronic inflammatory skin disorder. The disease tends to recur and may leave permanent scarring. The disease is known to be associated with obesity and metabolic syndrome. The aim was to describe successful management of obesity with the administration of GLP-1 receptor agonists in a cohort of patients with hidradenitis suppurativa, obesity and metabolic syndrome and its effect on lesions of hidradenitis suppurativa.

Materials & Methods:

A cohort of 10 female patients with hidradenitis suppurativa and obesity is presented. Laboratory evaluation revealed mild diabetes mellitus type 2 in 6 of the patients. Metformin was administered to all patients. However, as weight loss was not achieved, GLP-1 receptor agonists were administered to 9 of the patients.

Results:

The administration of GLP-1 receptor agonists led to significant weight loss in 7 of the patients, whereas in 2 of the patients it led to weight stabilization. Diabetes mellitus type 2 control improved in all patients with diabetes. Hidradenitis suppurativa lesions improved. Active lesions were easier to manage. No new active lesions appeared.

Conclusion:

Hidradenitis suppurativa is an inflammatory disorder associated with obesity and metabolic syndrome. In some cases frank diabetes mellitus may be observed. GLP-1 receptor agonist administration led to weight loss or weight stabilization in this cohort. Diabetes mellitus management improved. Hidradenitis suppurativa lesions improved. Thus, it appears that GLP-1 receptor agonists may lead to weight loss and to improved management of hidradenitis suppurativa. Thus, GLP-1 receptor agonists may be an ideal treatment for the management of obesity and metabolic syndrome in patients with hidradenitis suppurativa.

Abstract N°: 8669

When Skin Meets Eye: Ocular Involvement in Pemphigoid Disorders

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Introduction & Objectives:

Ocular involvement in bullous pemphigoid is a severe form of the disease, threatening the visual prognosis of affected patients and even leading to blindness.

The aim of our work is to illustrate the role of multidisciplinary consultation between dermatologists and ophthalmologists in the detection of ocular manifestations in bullous pemphigoid, and the importance of early management.

Materials & Methods:

This is a retrospective descriptive study including patients followed in our dermatology department for bullous pemphigoid, conducted in collaboration with the ophthalmology department, over a period of 10 years and 6 months from January 2013 to June 2023.

Results:

We identified 21 patients with ocular involvement out of 87 with bullous pemphigoid, with a clear female predominance. The sex ratio was M/F = 0.4. The mean age of our patients was 63.8 years. Phototype ranged from III to IV. 62% of patients were of urban origin. Ophthalmological involvement was present in 24.13% of patients, predominantly cicatricial pemphigoid in 13 cases (61.91%), followed by bullous pemphigoid in 8 cases (38.09%). Ocular involvement was bilateral in 81% of cases. Symptoms were mainly chronic conjunctivitis (41%), scarring conjunctivitis (7%), dry eyes (53%), corneal abrasions (13%) and microbial keratitis (6%). The evolution was marked by the development of symblépharons (5%), ankyloblépharons (3%), entropion (3%), ectropion (1%), one case of corneal perforation complicated by blindness and 3 cases of corneal neovascularization. Therapeutic management was carried out in consultation with ophthalmologists, and included a predominantly medical component in the acute phase (42%), while the sequelae phase was mainly surgical, consisting of a cure of symblepharon in 5 cases, with scleral lenses in 6 cases, cure of entropion in 4 cases, corneal grafting in 2 cases, amniotic membrane grafting in 6 cases, buccal membrane grafting in 3 cases, as well as recourse to autologous serum in severe dryness in 13 cases.

Conclusion:

Early diagnosis of ocular involvement in bullous pemphigoid is crucial, hence the importance of good multidisciplinary consultation between dermatologists and ophthalmologists. Early treatment can prevent non-reversible palpebral abnormalities and limit secondary corneal complications.

Abstract N°: 8689

Pemphigus Vegetans Limited to the Axilla: Diagnostic Insight from a Unique Case

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Introduction & Objectives:

Pemphigus vegetans is a rare clinical variant of pemphigus characterized by vegetative plaques, often occurring in intertriginous areas. Due to its unusual appearance, it can mimic various other dermatological conditions, making diagnosis particularly challenging, especially in elderly or immunocompromised individuals. We report the case of a 71-year-old male patient with type 2 diabetes who presented with a persistent axillary lesion. The aim of this report is to detail the diagnostic approach and therapeutic aspects in this uncommon presentation.

Materials & Methods:

The patient, a 71-year-old man with known diabetes, was evaluated for a unilateral axillary lesion that had been slowly evolving since August 2024. Clinical examination revealed a hypertrophic, vegetating plaque in the left axillary region. There was no mucosal involvement or presence of similar lesions elsewhere. A skin biopsy was performed. Histological analysis revealed suprabasal acantholysis and marked eosinophilic infiltration, consistent with pemphigus vegetans. Direct immunofluorescence testing showed intercellular deposition of IgG and C3.

Results:

The clinical presentation, along with histological and immunofluorescence findings, supported the diagnosis of pemphigus vegetans. The patient was started on systemic corticosteroids. Close monitoring was implemented to manage both therapeutic response and glycemic control due to the underlying diabetes.

Conclusion:

While pemphigus vegetans is rare, its unilateral presentation in older patients may complicate diagnosis due to overlapping symptoms with other dermatological conditions. Early recognition and treatment are vital to prevent morbidity associated with the disease.

Abstract N°: 8691

Diffuse Extragenital Lichen Sclerosus: Clinical Case and Therapeutic Response to PUVA Phototherapy

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Introduction & Objectives: Lichen sclerosus (LS) is a rare, chronic inflammatory dermatosis of unknown etiology that predominantly affects postmenopausal women. While it typically involves the anogenital region, extragenital manifestations can also occur. The exclusively extragenital form is uncommon, accounting for approximately 2.5% of cases. This report presents a rare case of diffuse extragenital LS with successful therapeutic response to PUVA phototherapy.

Materials & Methods: A comprehensive review of the literature was conducted, highlighting the rarity of exclusively extragenital LS, with only a limited number of cases reported to date. Moreover, data on the use of PUVA phototherapy as a therapeutic option for this clinical form remain scarce.

Results: A 50-year-old female patient reported the appearance of shiny, hypopigmented, pearly plaques with mild induration following vascular surgery. The lesions began on the lower extremities and gradually spread to the trunk and upper limbs, while sparing the genital region. Pruritus was present. An incisional skin biopsy revealed basal vacuolar

degeneration, subepidermal hyalinization, and dilated capillaries—findings consistent with the diagnosis of extragenital LS. Due to the extent of skin involvement, PUVA phototherapy was initiated. The patient underwent 24 treatment sessions, resulting in significant improvement in pruritus, skin induration, and overall surface texture.

Conclusion: Exclusively extragenital lichen sclerosus is an uncommon presentation that requires a high index of clinical suspicion for accurate and timely diagnosis. The characteristic lesions—hypopigmented, shiny, atrophic plaques—must be promptly recognized to ensure early intervention. In extensive cases, phototherapy has demonstrated efficacy through cytokine release and immunosuppressive mechanisms, leading to clinical improvement and symptom control. For more severe or refractory cases, systemic therapies such as methotrexate, hydroxychloroquine, cyclosporine, acitretin, and mycophenolate mofetil may be considered. This case reinforces the importance of early recognition and individualized therapeutic strategies in the management of extragenital LS.

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Abstract N°: 8714

GLP-1 Receptor Agonists as Potential Pemphigus Risk Modifiers: A Propensity-Score-Matched, Population-Level, Retrospective Cohort Analysis

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Introduction & Objectives:

Obesity has long been conceived of as a simple problem of imbalance between energy intake and expenditure. However, recent advances have led to an increased appreciation of its complex pathogenesis, which encompasses environmental exposures acting on a background of genetic predisposition, chronic low-grade inflammation, and aberrant immune activity. These features are similarly hallmarks of human autoimmune diseases, including the pemphigus group of blistering disorders.

In recent years, the class of Glucagon Like Peptide-1-Receptor Agonists (GLP-1RAs) have received attention not only for their ability to induce weight-loss but, more interestingly, for their pleiotropic effects on immune function and, subsequently, on autoimmune disease. For example, emerging evidence in psoriasis has consistently shown that psoriatic individuals with type 2 diabetes mellitus (T2DM) treated with GLP-1RAs experienced significant reductions in disease activity independent of, and prior to, changes in weight or glycemic status, indicating that exposure to these drugs is independently associated with decreased local and/or systemic inflammation.

Materials & Methods:

Despite the significant corpus of evidence highlighting the bidirectional relationship between metabolic and immunologic function, the capacity of GLP-1RAs to improve both, as well as to directly ameliorate autoimmune disease activity, a population level assessment of the potential for these drugs to modify the risk for the development of pemphigus remains absent from the literature. To address this gap, we conducted a retrospective cohort study using the Global Collaborative Network of TriNetX. We constructed two cohorts: Cohort 1 included individuals exposed to any GLP-1RA (semaglutide, liraglutide, tirzepatide, and dulaglutide) and never exposed to metformin. Cohort 2, our comparator, comprised individuals exposed to metformin, but never exposed to any GLP-1RAs. To reduce potential confounding, the two cohorts (each n=737,166). were propensity-score-matched for age, sex, race, ethnicity, diagnoses of T2DM, exposure to a broad swath of drugs previously reported to trigger/exacerbate pemphigus, and weight status by BMI.

We then evaluated the odds among the two matched cohorts for the**subsequent** development of: *any* subtype of pemphigus, pemphigus vulgaris (PV) or pemphigus foliaceous (PF) specifically, and neurofibromatosis-type-II (NF2) as a genetically determined negative control, each identified via ICD-10 codes.

Results:

Among the >1.4 million individuals assessed herein, those exposed to GLP-1RAs had significantly decreased odds for the subsequent development of any subtype of pemphigus as well as PV/PF specifically, compared to the GLP-1RA-non-exposed cohort; there was no significant difference in the odds of subsequent diagnosis of NF2 (**Table 1**).

Conclusion:

Given that cardiovascular (CV) disease is a leading cause of mortality among pemphigus patients, the well-

established efficacy of GLP-1RAs in reducing rates of major adverse CV events and the promising early results in psoriasis, together with the epidemiologic data presented here demonstrating a protective effect of GLP-1RA exposure against the development of pemphigus, future investigations of these agents in pemphigus patients are warranted. These insights highlight avenues for the development of safer treatment regimens which transcend the present, immunosuppressive paradigm.

Table 1.

Outcome	Cohort(s)	Number of Eligible Individuals	Number with Outcome	Odds Ratio (95% CI)
***Any* subtype of pemphigus** (ICD-10: L10)	Cohort 1, GLP-1RA exposed	736,995	50	**0.391** (0.282, 0.542)
	Cohort 2, GLP-1RA non-exposed	736,997	128	
Pemphigus vulgaris (PV) or **Pemphigus foliaceous (PF)** (ICD-10: L10.0, L10.2)	Cohort 1, GLP-1RA exposed	737,041	30	**0.417** (0.272, 0.638)
	Cohort 2, GLP-1RA non-exposed	737,031	72	
Neurofibromatosis Type II (ICD-10: Q85.02)	Cohort 1, GLP-1RA exposed	737,141	11	0.687 (0.319, **1.481**)
	Cohort 2, GLP-1RA non-exposed	737,120	16	

Abstract N°: 8773

Case of vegetative pemphigus vulgaris following rituximab infusion

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Case of vegetative pemphigus vulgaris following rituximab infusion

Introduction & Objectives:

Pemphigus vulgaris (PV) is a chronic autoimmune blistering disease caused by autoantibodies against desmoglein 1 and 3, leading to intraepidermal acantholysis and flaccid blisters on skin and mucous membranes. Pemphigus vegetans is a rare PV variant (<5%), marked by vegetating, verrucous plaques in intertriginous and mucosal areas. Rituximab, an anti-CD20 monoclonal antibody, transformed PV treatment, proving effective in refractory cases, reducing steroid use, and improving outcomes. Despite its efficacy, rituximab carries infection and skin complication risks, posing diagnostic challenges in immunosuppressed patients. Verrucous lesions in PV patients under rituximab need evaluation, as they may indicate infections, drug-induced lesions, or atypical disease. No prior reports describe pemphigus vegetans or progressing from PV to vegetans variant after rituximab. We report a rare case of pemphigus vegetans post-rituximab in a PV patient, highlighting diagnostic, histopathological, and therapeutic challenges.

Materials & Methods:

A 64-year-old male from São Paulo, hypertensive and ex-smoker, presented with throat and tongue discomfort and dysphagia. Initially treated symptomatically, he developed pruritic, non-painful blisters with serous discharge, oral and genital erosions, worsening dysphagia. Skin biopsy confirmed PV; he received methylprednisolone pulses and rituximab, achieving remission. After stopping azathioprine and losing follow-up, he returned with a forearm blister (Nikolsky negative), oral ulcers, and a verrucous lesion on the lip commissures. Biopsies confirmed PV on the forearm and pemphigus vegetans on the lips. Treatment included prednisone 80 mg/day, oral triamcinolone, and planned rituximab re-infusion.

Results:

PV management relies on immunosuppression to prevent lesions and promote healing. Corticosteroids remain first-line, alone or with azathioprine or mycophenolate. Rituximab is pivotal for refractory and initial treatment. Adverse effects include infusion reactions (headache, chills, hypertension, nausea), infections (warts, herpes, bronchitis, herpes zoster, UTIs, fungal infections, conjunctivitis), and rare severe reactions. Disease exacerbation post-rituximab is rare but manageable. No prior reports describe pemphigus vegetans post-rituximab, making this case notable. Differential diagnosis with viral warts is challenging, as both show verrucous lesions, especially in immunosuppressed patients. Histopathology is key: warts show hyperplasia with koilocytosis; pemphigus vegetans shows suprabasal acantholysis with eosinophils. Immunofluorescence shows intercellular IgG and C3. Misdiagnosis risks poor treatment; warts respond to topical therapy, pemphigus vegetans requires systemic immunosuppression.

Conclusion:

This case documents pemphigus vegetans post-rituximab in PV. It underscores diagnostic complexity, histopathological confirmation, and need for further research.

Abstract N°: 8779

The Role of IL-36 Family Cytokines in Pyoderma Gangrenosum: Evidence for an Imbalanced Inflammatory Response

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Introduction & Objectives:

Pyoderma gangrenosum (PG) is a rare dermatological condition characterized by the rapid development of painful ulcers. The pathogenesis of PG remains largely elusive, complicating the development of effective therapeutic strategies. The IL-36 family includes IL-36α, IL-36β, and IL-36γ, which interact with the IL-36 receptor (IL-36R). IL-36Ra, IL-37, and IL-38 function as antagonists of IL-36R, modulating inflammatory processes by inhibiting the actions of the aforementioned cytokines. Despite the documented involvement of IL-36 group members in diseases associated with PG, their specific role in PG pathogenesis remains unexplored. This study aims to evaluate the serum levels of IL-36R agonists and antagonists in PG patients and their potential clinical correlations.

Materials & Methods:

This study included 44 adult patients with PG and 40 healthy volunteers. Stringent exclusion criteria were applied to eliminate individuals with active comorbidities, malignancies, recent treatments, or ongoing infections. At admission, a thorough assessment of skin lesions was performed, evaluating lesion count, dimensions, and surface area. Concurrently, peripheral venous blood samples were collected to measure various clinical parameters, including levels of IL-36α, IL-36β, IL-36γ, IL-36Ra, IL-37, IL-38.

Results:

The study group consisted of 29 female (65.9%) and 15 male (34.1%) patients with PG, while the control group comprised 21 female (52.5%) and 19 male (47.5%) individuals. The mean age of PG patients was 50.2 ± 16.4 years, whereas the mean age of the control group was 48.2 ± 13.0 years. PG patients demonstrated significantly lower levels of IL-36α ($p = 0.0003$) and IL-36γ ($p = 0.02$) compared to the controls. In contrast, PG patients had markedly higher levels of IL-36Ra ($p = 0.000$), IL-37 ($p = 0.000$), and IL-38 ($p = 0.000$) relative to controls. Moreover, a positive correlation was observed between IL-36β and IL-36α ($r = 0.4$, $p = 0.001$) and between IL-36β and IL-36γ ($r = 0.2$, $p = 0.02$). On the other hand, negative correlations were found between IL-36γ and white blood cell count ($r = -0.33$, $p = 0.028$), as well as between IL-36γ and IL-38 ($r = -0.2$, $p = 0.03$). Additionally, a negative correlation was found between IL-36α and total ulcer surface area ($r=-0.4$, $p=0.003$) and between IL-36α and IL-36Ra ($r = -0.4$, $p = 0.001$).

Conclusion:

Patients with PG exhibit significantly lower serum levels of IL-36α and IL-36γ compared to healthy controls. Conversely, PG patients have markedly higher levels of IL-36Ra, IL-37, and IL-38. Moreover, this study highlights a correlation between larger ulcer surface areas and reduced IL-36α levels, as well as higher white blood cell count and decreased IL-36γ levels. The findings suggest that the dysregulation of IL-36 cytokines plays a role in the

pathogenesis of PG. This emphasizes the necessity for further investigation into the role of the IL-36 group in the pathogenesis of PG, as it holds potential implications for developing and planning innovative therapeutic approaches in addressing this challenging to treat dermatosis.

Abstract N°: 8793

A rare case of pemphigus herpetiformis evolving into hypertrophic discoid lupus erythematous

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Introduction & Objectives:

Pemphigus and lupus erythematosus (LE) are classified as B-cell-mediated autoimmune diseases, both depending on autoreactive CD4+ T lymphocytes. Many forms of pemphigus have been anecdotally reported to occur in association with LE as well as other autoimmune diseases but the pathogenic mechanism remains unclear. (1) We report a rare case of pemphigus herpetiformis (PH) evolving into hypertrophic discoid lupus erythematous (HDLE) treated with hydroxychloroquine and isotretinoin.

Materials & Methods:

A 56-year-old male with no underlying medical condition visited our department with a one-year history of erythematous, papular and vesicular lesions of the trunk, upper and lower limbs distributed in a herpetiform pattern and with severe pruritus. A skin biopsy was taken from the trunk showing eosinophilic spongiosis and a neutrophilic infiltrate compatible with PH. Laboratory tests and radiological explorations were within normal limits. Patient was treated with Disulone 100mg/day. He responded well and was lost to follow up. One year later he came back with erythematous and violaceous plaques showing atrophy and hyperpigmentation at the margins located on the nose, scalp and upper trunk. Sun exposed upper and lower limbs showed verrucous plaques. On the buccal mucosa there was a white reticular pattern. Histology of a skin biopsy on the forearm demonstrated acanthosis and hyperkeratosis of the epidermis with plugged follicles and an interface inflammatory pattern with peri-vascular and peri-adnexal lymphocytic infiltrate. The patient was negative for ANA. He was diagnosed as HDLE and started on hydroxychloroquine 400 mg/day and clobetasol; however, he did not respond. Isotretinoin 10mg/day was added. After 2 months the plaques resolved with residual pigmentation.

Results:

PH is a rare subtype of intraepidermal autoimmune bullous diseases that shares clinical characteristics with dermatitis herpetiformis and immunologic features with pemphigus. (5) Many forms of pemphigus have been reported to occur in association with Cutaneous LE (1), but to our knowledge there have not been case reports of pemphigus evolving into HDLE, the case of our patient.

HDLE is a rare subtype of DLE, characterized by verrucous and hyperkeratotic nodulo-plaques with tendency of affecting the sun exposed skin (2) like our patient. It is characterized histologically by irregular epidermal hyperplasia associated with features of classic chronic cutaneous lupus erythematous, including interface changes. (3) Chronicity, absence of regression, therapeutic failures and potential to develop malignancy are causes of concern. (4) In our case, combination of isotretinoin with hydroxychloroquine proved highly effective and the hyperkeratotic plaques disappeared promptly within the first 2 months of treatment.

This can be explained by the hypothesis that the presence of any autoimmune diseases increases the probability of additional ones occurring during the disease's course because they can share common immunopathogenic mechanisms and risk factors (1) which can explain why PH evolved into HDLE in our case.

Conclusion:

We report an uncommon clinical presentation of PH evolving into HDLE that may highlight an underlying pathophysiologic link between the two conditions. This case also shows a good response to treatment with isotretinoin and hydroxychloroquine in HDLE which is known to be resistant to most of the therapies. (4)

Abstract N°: 8830

JAK/STAT signalling pathway is involved in the immune mechanism ofBullous pemphigoid

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Introduction & Objectives: Bullous Pemphigoid (BP) is a common immune bullous disease, mainly affecting the elderly. However, the molecular pathogenesis of the JAK/STAT pathway in BP is not fully understood. We aim to characterize immune profiles and the key JAK/STAT pathway in BP patients. The clinical efficacy of the JAKi for BP patients was also assessed.

Materials & Methods: Skin transcriptome profile, plasma cytokine/chemokine levels, and *in vitro* T cell activation and JAKi blocking assay for BP patients were performed. Clinical improvement for steroid-resistant BP patients treated with JAK inhibitors was evaluated.

Results: A total of fifty BP patients as well as thirty-one healthy individuals were enrolled.** The mRNA expression levels of JAK3 and STAT3 were increased in skin lesions from the BP patients. The BP-related inflammatory-mediated cytokines/chemokines such as IL-5, CCL22, TARC/CCL17, PARC/CCL18, MMP9, and Granzyme B (*P* values <10-3-10-5) were elevated in the BP patients compared with those in the healthy control. *In vitro* T cell activation and JAKi blocking assay revealed that tofacitinib (JAK1/3i) and ritlecitinib (JAK3i) had better inhibitory effects than upadacitinib for granzyme B and TARC secretions in BP patients. Eight steroid-resistant BP patients were treated with oral tofacitinib. Among these patients, five patients had a rapid reduction in their Bullous Pemphigoid Disease Area Index (BPDAI) from 104.2 to 34.8 within five weeks..

Conclusion: JAK3 inhibitors can attenuate JAK3/STAT3-mediated inflammatory factors, providing an alternative treatment strategy for refractory BP patients in combination with low-dose steroids.

Abstract N°: 8845

Skin Manifestations of Adult-Onset Immunodeficiencies

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Introduction & Objectives:

Immunodeficiencies represent a group of diseases characterized by dysregulated immune function. Although these conditions are frequently diagnosed in childhood, some forms manifest later in life, complicating diagnosis. Adult immunodeficiencies generally stem from genetic mutations which result in immune dysfunction: namely autoimmunity and increased infection risks. They commonly affect the skin, as it defends against many pathogens and is populated by resident immune cells. Typical presentations of immunodeficiencies include recurrent and refractory cutaneous infections, unexplained weight loss and inflammatory dermatoses. Often, they can be mistakenly attributed to hematological disorders or interpreted as adverse events related to therapeutic measures. However, a more thorough clinical examination may reveal that immunodeficiency itself may be responsible for the physiopathologic cascade. This review examines the spectrum of skin involvement in adult immunodeficiencies and highlights its importance in the clinical assessment and management of affected patients.

Materials & Methods:

A comprehensive review of adult immunodeficiencies was carried out, focusing on dermatological presentations and associated hematological conditions.

Results:

The results reveal that several autoimmune pathologies can cause cutaneous symptoms, with some reaching over 50% of cases. Pathologies such as common variable immunodeficiency, selective IgA deficiency and idiopathic CD4+ lymphocytopenia can manifest as bacterial skin infections and resistant warts. Autoimmune manifestations such as vitiligo, lichen planus, atopic dermatitis and other skin infections are associated with Good’s syndrome, APECED, VEXAS and LRBA deficiency. These findings often coincide with systemic hematological complications, including lymphomas, thymomas and myelodysplastic syndromes. The attached table provides a summary of the associations between skin manifestations and corresponding hematological disorders.

Table 1. Adult-onset immunodeficiencies with skin involvement

Disease	Typical skin manifestations	Associated hematological diseases
Antibody defects		
Common variable immunodeficiency	Bacterial skin infections	CLL
	Severe rosacea	Risk of lymphoma
	Seborrheic dermatitis	
Selective IgA deficiency (SIgAD)	Skin abscesses	Transfusion reactions
Lymphocyte defects		
Idiopathic $CD4^+$ lymphocytopenia	Warts	Risk of lymphoma
Immune dysregulation syndromes		
Hyper IgE syndrome (HIES)	Eczema	Risk of lymphoma
	Staphylococcal infections	
Autoimmune polyendocrinopathy with candidiasis and ectodermal dystrophy (APECED)	Mucocutaneous candidiasis	Pernicious anemia
	Nail dystrophy	
Good syndrome	Lichen planus	Thymoma
VEXAS (vacuoles, E1 enzyme, X-linked autoinflammatory, somatic syndrome)	Herpes reactivation	Myelodysplastic syndrome
	Chondritis	Hemophagocytic lymphohistiocytosis (HLH)
	Vasculitis	
LRBA deficiency	Atopic dermatitis-like eczema	Lymphadenopathy
	Vitiligo	
Innate immune deficiencies		
GATA2 deficiency	Warts	Myelodysplastic syndrome
WHIM syndrome	Warts	Leukopenia
Chronic granulomatous disease (CGD)	Severe bacterial and fungal infections	Hemophagocytic lymphohistiocytosis (HLH)

Conclusion:

Skin manifestations are important diagnostic indicators of immunodeficiency in adulthood. Early recognition of these signs is key to prompt detection and targeted intervention. Addressing gaps in our knowledge of these pathologies is essential to refining diagnostic approaches and tailoring management to the unique challenges of adult phenotypes.

Abstract N°: 8931

Association between anti-Ku autoantibodies and organ involvement in patients with autoimmune connective tissue diseases

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Introduction & Objectives:

Anti-Ku antibodies are a type of autoantibody found in patients with autoimmune connective tissue diseases (ACTDs). They target the Ku protein, which is essential for DNA repair. The presence of anti-Ku antibodies is often linked to organ damage, particularly in the lungs, muscles, and kidneys. Their detection can help identify patients at higher risk of organ involvement and guide treatment strategies. Therefore, we aimed to examine the expression of anti-Ku antibodies among ACTDs and the association with organ involvement

Materials & Methods:

A cross-sectional cohort study recruited patients diagnosed with one of ACTDs, including systemic lupus erythematosus (SLE), systemic sclerosis (SSc), dermatomyositis (DM), and overlap syndromes (OVLS), whose serum samples were screened for anti-Ku antibodies using the ANA 23 profile test kit.

Results:

The study was comprised of 215 eligible patients, with the mean age of 43.6 ± 16.4 years and women (80%) being predominant among the cohort. Overall, 27.9% (60/215) of ACTD patients were anti-Ku positive. Sub-analyses revealed the anti-Ku positive rate of 43.3%(26/60) within SLE patients; SSc, 10.3%(6/58); DM, 18.3%(11/18.3); OVLS, 29.8%(17/57). Regarding organ involvement, we observed a statistically higher risk of increased serum creatine kinase (53.3%) and pulmonary arterial hypertension (61.7%) among the anti-Ku positive group (compared to 29.7% and 31.0%), respectively, for the anti-Ku negative group (all $P<0.05$). There were other differences in the risk between the positive and negative group, such as interstitial lung disease (28.3% vs 36.6%), presence of proteinuria (15.5% vs 13.2%). Yet these findings did not reach statistical significance (all $P>0.05$).

Conclusion:

These findings reaffirm the favorable prevalence of anti-Ku antibodies in SLE among ACTDs and underscore the prognostic role in critical organ involvement including pulmonary arterial hypertension and preclinical myositis

Abstract N°: 8936

Bullous systemic lupus erythematosus: experience from a single-center retrospective study

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Introduction & Objectives:

Bullous systemic lupus erythematosus (BSLE) is a rare presentation of systemic lupus erythematosus (SLE) that occurs in less than 5% of SLE patients, which represents a challenging entity due to clinicopathological similarities to other primary bullous dermatoses. This study aimed to investigate clinical and laboratory findings, treatment and outcome of patients with BSLE through a single-center retrospective study

Materials & Methods:

This retrospective study recruited patients who met the criteria for BSLE confirmed by board-certified dermatologists at National Hospital of Dermatology and Venereology from 2019 to 2025. Eligible cases were reviewed for the clinical presentations, histopathology, immunofluorescence, treatment response, and follow-up for flare-ups

Results:

16 eligible patients were included in this study. Females predominated the cohort, 12/16 (75%). The mean age was 27.4±15.7 years, with 50% of patients aged 20-40 years; particularly, there were 5 pediatric patients (31.3%). 10/16 (62.5%) patients had been diagnosed with SLE prior to bullae manifestation, whereas blisters preceded the diagnosis of SLE in the remaining 6 patients (37.5%). 100% of patients presented with vesicles and tense bullae that appeared spontaneously over a background of erythema or healthy skin, primarily concentrated on the face, neck, trunk, and upper limbs. 68.8% of patients featured urticarial wheal-like lesions, and 56.3% experienced mucosal erosions in the mouth. In addition, 50% of patients were identified renal involvement. On histopathology, 100% of patients revealed the typical findings: subepithelial bullae with significant infiltration of neutrophils. Upon direct immunofluorescence (DIF), 81.3% of biopsy specimens demonstrated linear positivity for IgG at the basement membrane zone (BMZ) and vascular walls, while 25% exhibited coarse granular IgG along the BMZ. Furthermore, 68.8% of patients showed positivity for at least one among other antibodies (IgA, IgM, C3, and Fibrinogen) on DIF studies. On indirect immunofluorescence and salt-split skin, all patients (100%) showed positivity for IgG in nuclei and a linear pattern at the base of the blisters. Dapsone provided a rapid and sustained resolution of bullae in all administered patients (7/7, 100%), while systemic steroids showed a slower improvement among 9/9 patients with recurrence of blisters occurred in 3/9 (33.3%) patients. Lupus-related organ involvement was managed with systemic steroids and immunosuppressants (6 patients)

Conclusion:

BSLE is a rare autoimmune subepithelial blistering disease, with the definitive diagnosis based on clinical and histopathological findings. Dapsone is considered as the first-line treatment offering rapid and prolonged clearance of blisters, while corticosteroids and other immunosuppressants are selected for cases with organ involvement. Concomitant management of SLE is essential for disease control

Abstract N°: 8938

Clinical Characteristics, Management and Outcomes of Pemphigoid Nodularis: A Systematic Review

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Introduction & Objectives:

Pemphigoid nodularis (PN) is a rare variant of bullous pemphigoid (BP), characterized by nodular lesions and persistent itching. PN remains underreported in the medical literature due to uncertainties surrounding its clinical features and management. This systematic review summarizes the clinical characteristics, management strategies, and outcomes of PN.

Materials & Methods:

Following PRISMA guidelines, Embase and MEDLINE databases were searched using specific keywords. Study quality was appraised using the Oxford Centre for Evidence-Based Medicine 2011 Levels of Evidence. After independent screening by two reviewers, 35 articles (published between 1981-2023) were included, encompassing 52 patients. Evidence included cohort studies (2.9%, 1/35), case series (11.4%, 4/35), and case reports (85.7%, 30/35).

Results:

The mean patient age was 65 years (range: 11-82), with 26.9% (14/52) males and 73.1% (38/52) females. The mean age of onset was 65.1 years, with a mean disease duration of 39.2 months. Fifty-one cases (98.1%) had biopsy-confirmed diagnoses. Frequent morphological features included excoriated nodules (38.5%; 20/52), hyperkeratotic nodules (23.1%; 12/52), and blisters (19.2%; 10/52). Typical locations were the extremities (84.6%; 44/52), trunk (71.2%; 37/52), and scalp (15.4%; 8/52). Common histopathological features included perivascular mononuclear inflammatory infiltrates (46.2%; 24/52), acanthosis (32.7%; 17/52), and direct immunofluorescence (DIF)showing linear IgG (32.7%; 17/52) and C3 deposition (26.9%; 14/52).

Topical corticosteroid monotherapy was used in 15.4% (8/52) of cases. Clobetasol was the most common successful topical treatment, achieving complete resolution in 33.3% (2/6) of cases and partial resolution in 66.6% (4/6). Systemic therapies were used in 69.2% (36/52) of cases. Oral corticosteroid monotherapy was the most common successful systemic treatment, achieving complete resolution in 25% (3/12) of cases and partial resolution in 75% (9/12). Biologics were used in 15.4% (8/52) of cases. Rituximab was most frequently used, with complete resolution in 33.3% (2/6) of cases and partial resolution in 33.3% (2/6). Recurrence occurred in 25% (2/8) of cases treated with topicals and 13.9% (5/36) treated with systemic therapies. Nausea was the only adverse event reported, occurring with the combination of oral azathioprine and prednisolone (2.8%; 1/36).

Conclusion:

This review offers a comprehensive overview of PN, addressing uncertainties related to its clinical presentation and management. Our findings reveal that PN often mimics prurigo nodularis, presenting with intensely pruritic, excoriated, hyperkeratotic nodules more commonly than the blisters typical of BP. PN predominantly involves the extremities and trunk, unlike the flexural and intertriginous distribution frequently seen in BP. Despite these clinical differences, PN shares BP's immunopathological features, including BP180 and BP230 autoantibodies and linear

IgG and C3 deposition along the basement membrane zone. While topical corticosteroids may suffice for mild BP, PN often requires systemic therapy, such as oral corticosteroids, which generally led to resolution. However, recurrence in some cases suggests persistent immune dysregulation. Systemic treatments were well-tolerated, with few serious adverse events.