Abstract N°: 34

Targeted and whole-body narrowband UVB phototherapy stabilizes acral vitiligo with minimal repigmentation outside of the wrists and ankles.

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Introduction & Objectives: Vitiligo is a determined melanocytopenia characterized by depigmentation, in which skinpatches lose brightness due to a lack of melanin. In cases of vitiligo, phototherapy with narrowband ultraviolet B (NB-UVB) stimulates stability and repigmentation. Few studies have examined whole-body and localized NB-UVB for the treatment of acral vitiligo.This randomized split-body trial examined the effects of targeted and whole-body NB-UVB on repigmentation and stability in acral vitiligo.

Materials & Methods: Twenty individuals with bilaterally symmetrical lesions of acral vitiligo (distal to the knees and elbows) were included. Patients underwent targeted NB-UVB treatment on the side that was shielded after receiving whole-body NB-UVB treatment. One hand and one foot were shielded up until the elbow and knee. Using the Vitiligo Disease Activity (VIDA) score, Vitiligo Skin Activity Score (VSAS), Vitiligo Area Scoring Index (ascertained by fingertip method using the method to compute facial-VASI), and degree of repigmentation, patients were evaluated at one-month intervals for three months.

Results: There were six (30%) females and fourteen (70%) males aged 18 to 60.At 12 weeks, 87.5% of patients had a VIDA score of 3, and at 24 weeks, none of them still had active illness. In the whole-body and targeted groups, above 50% repigmentation was seen in 45.2% and 36.7% of limbs, respectively (p =.95). Over the course of the 24-week period, neither modality showed any improvement in the hands' or feet's (distal to the wrist and ankles) F-VASI scores.

Conclusion: Comparable rates of repigmentation were observed in our study between the targeted and whole-body NB-UVB groups. Phototherapy limited success in repigmenting the hands and feet highlights a significant therapeutic gap.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 36

Effectiveness of Skin Microneedling Combined with Narrow Band UVB Phototherapy for Vitiligo

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Introduction & Objectives: The depigmented, milky white patches that are the hallmark of the chronic cutaneous condition vitiligo have a psychological effect on the patient's quality of life. In order to treat vitiligo more quickly and with fewer adverse effects, several therapeutic techniques have been created. To assess the effectiveness and safety of using micro needling and narrow band UVB together to treat vitiligo.

Materials & Methods: Thirty patients with stable vitiligo were included in this investigation. For three months, they received treatment with micro needling (one session every two weeks) and narrow band-UVB (three sessions per week). Repigmentation was assessed using the Vitiligo Area Scoring Index (VASI).

Results: Following three months of therapy, 10% of the patients in the study reported good improvement, 20% showed moderate improvement, 40% showed mild improvement, and 30% showed no improvement. These results represent statistically significant clinical improvements. The mild and temporary adverse effects that were recorded included erythema at the micro needling site, burning sensation, and mild pain that went away on its own in a few hours.

Conclusion: For the treatment of vitiligo, micro needling combined with narrow band UVB phototherapy may be useful. Microneedling is a manageable method that has very little negative side effects and is safe.

Abstract N°: 53

The effectiveness of the treatment of psychosomatic disorders in patients with vitiligo

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Introduction & Objectives The autoimmune disease vitiligo is a long-term condition that causes skin depigmentation. It impacts on the patient's psychological profile in addition to his appearance. Psychosomatic illnesses can lessen the efficacy of therapy and patients' commitment to long-term treatment. They are particularly prevalent in patients with vitiligo, with a focus on secondary (nosogenic) anxiety and depressive disorders. Examining the connection between results and psychosomatic diseases. The creation of all-encompassing methods for treating this illness depends on the management of vitiligo. This study aims to assess the impact of psychological problems on the efficacy of different vitiligo treatment approaches. The purpose of the research is to determine whether a patient's psychological profile and treatment outcomes are related. It also intends to create guidelines for the incorporation of psycho-corrective therapy.

Materials & Methods :100 vitiligo patients were included in the study. The subjects were split into two groups: 50 patients in the control group and 50 patients in the experimental group, which started with a dosage of 2.5 mg of alimemazine, the first-line psychiatric medication. Patients in this group will get combined therapy with alimemazine in accordance with routine medical care for patients with vitiligo. One of the most widely used first-line psychotropic medications, mecamyezine has demonstrated its efficacy and safety in a variety of patient populations. Patients exhibiting symptoms of central nervous system depression, such as alcohol intoxication, barbiturates, or other drugs that depress the central nervous system, pathological blood pattern changes, myelodepression, pregnancy, lactation, or hypersensitivity to mecamyezine, are not permitted. The Hospital Anxiety and Depression Scale (HADS), a Dermatological Quality of Life Index (DLQI), VitiQoL, and the Vitiligo Impact Scale (VIS)-22 were among the tests and scales used to evaluate the patients' psychological profiles. After 6 months, changes in the afflicted skin area and degree of repigmentation were used to assess the efficacy of vitiligo therapy.

Results: The study's findings indicated that, in contrast to individuals with a normal psychological profile, those with severe psychosomatic illnesses exhibited less efficacy from vitiligo therapy. 40% of the patients in the experimental group showed improvement in their skin condition, compared to 70% in the control group. These findings suggest that psychosomatic variables have a major impact. Further counseling in psychotherapy, such as cognitive behavioral therapy. In addition to enhancing patient psychological profiles, relaxation techniques have been shown to boost vitiligo therapeutic efficacy. More noticeable changes in the dynamics of the skin process were connected with lower levels of social anxiety and depression in the experimental group.

Conclusion: The research verified the noteworthy influence of psychological problems on the efficacy of vitiligo treatment. Patients showed poorer response to treatment when their levels of stress, sadness, and social anxiety were higher. The psychological profiles of patients and treatment outcomes improve when psycho-corrective therapy is incorporated into traditional vitiligo therapy. This highlights the significance of an integrated approach to vitiligo therapy, which can greatly enhance patient outcomes and quality of life.

Abstract N°: 54

Children and adolescents with vitiligo have reduced quality of life due to disease-related behavioral patterns and experiences

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Introduction & Objectives :An acquired, non-contagious depigmentation condition called vitiligo causes a patchy loss of skin color. Adult patients frequently experience stigmatization, shame, and a decline in their quality of life (QoL). The responses of children are not well understood. Goals This study looked at a global group of children and adolescents' experiences and quality of life in relation to sickness.

Materials & Methods: Quality of life, disease-related experiences and behavior, and sociodemographic data were examined in 25 boys and 15 girls (age range: 7–17 years) using the Children's Dermatology Life Quality Index (CDLQI) and additional questions. Eighteen children without skin disorders served as age-, sex- and skin color-matched controls.

Results: The average length of the illness was 3.5 years. The head and neck, legs, and trunk were the most often affected areas. In total, 85% of the 40 participants had seen a doctor, 35% had a positive family history, and 75% had undergone treatment. Sixty-two percent were upset about their vitiligo, and ninety-three percent reported low-key stigmatization, forty-four percent hurtful remarks, and twenty-seven percent bullying. Of those with vitiligo, 25% had hidden the condition from others, and 30% had avoided certain circumstances as a result. Stigmatization frequency affected avoidant conduct. Friends and parents, especially moms, were valuable resources of support. The number of friends and leisure activities was similar between the patient and control groups. The group's average DLQI score was a low 2.8. Stigmatization, hiding white spots, facial depigmentation, avoiding situations, and a family background free of vitiligo were all associated with higher DLQI ratings.

Conclusion: QoL is impacted by avoidant behavior, bad experiences, and stigmatization associated with disease. Therefore, in order to screen for disease burden, the CDLQI should be used in conjunction with other tools. These findings demand that young individuals with vitiligo be carefully evaluated.

Abstract N°: 55

Assessment of Behavioral and Psychological Symptoms in Vitiligo Patients

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Introduction & Objectives: One common acquired depigmenting condition is vitiligo. Although the exact etiology is unknown, neuropsychological issues might be at play. A person with vitiligo may experience varying degrees of psychological behavior issues as a result of its effects on both their mental and physical health. Identification and assessment of behavioral reactions and psychosomatic disorders in patients with vitiligo

Materials & Methods: Adult vitiligo patients were drawn from the dermatological clinic between March and November of 2024 using a straightforward sample technique. The Dermatology Life Quality Index (DLQI), Symptom Checklist-90 (SCL-90), Social Avoidance and Distress Scale (SADS), and Medical Coping Modes Questionnaire (MCMQ) were used to assess them.

Results: At 7.46 ± 6.13, the DLQI score fell into the third category. Patients with vitiligo scored 126.84 ± 40.18 on the SCL-90, with the major symptoms being interpersonal sensitivity, depression, anxiety, and phobia. The patient's gender, marital status, disease severity, stage of the condition, and location of skin lesions may all have an impact. The patients' overall SADS score was 10.20 ± 6.56. The overall score as well as scores in each of the SADS aspects that were correlated with the patient's gender, level of education, and the kind, stage, and severity of skin lesions. The fucking score, as well as the yielding and avoiding scores, are for MCM.

Conclusion: Patients with vitiligo experience varied degrees of quality of life impairment, which can lead to a range of behavioral and psychiatric issues. We should actively care and improve the psychological health state and behavior of patients, and multidisciplinary treatment options and information regarding vitiligo should be delivered to the patients.

Abstract N°: 86

rheumatodermatologic mysteries - uncommon presentations and insight: a case series

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Introduction:

Rheumatodermatologic disorders are complex conditions with both dermatological and rheumatological manifestations often posing diagnostic challenge.Shared clinical insights, combined with diagnostic tests improve diagnostic accuracy.Early recognition of skin manifestations can prevent systemic complications.I hereby present a series of six cases of rheumatodermatological disorders with diagnostic dilemmas.

Observations:

24 year old female,multiple alopecic patches over scalp for 4 years.On examination multiple scarring and non-scarring alopecic patches over scalp,hyperpigmented atrophic plaques over face. HPE scalp:Lupus erythematosus profundus; ANA: 3+ nuclear homogenous;anti-ds DNA positive. Diagnosis:Lupus erythematosus profundus in a case of alopecia.

42 year old female,known case of Obsessive compulsive disorder,hypothyroidism with multiple raised lesions over extremities for 3 years.On examination multiple excoriated papules,nodules over extremities.HPE: Papulonodular mucinosis.ANA-3+,anti-U1RNP positive.C/O papulonodular mucinosis presented as prurigo nodularis in a case of mixed connective tissue disorder.

52 year old female,raised lesions over palms and soles,generalised fatigue, proximal myopathy for 2 years.Examination revealed multiple violaceous plaques over palmar aspect of MCP and PIP joints (inverse Gottron sign).Muscles biopsy:Inflammatory myositis ENA panel: MDA 5, Jo1, Ro 52,U1RNP,PM-Scl positivity.Diagnosis:Dermatomyositis with inverse Gottron’s sign and rapidly progressing interstitial lung disease.

20 year old female,multiple red scaly lesions over face and extremities for 15 days.H/o chicken pox 2 weeks prior to onset.O/e: multiple scaly plaques,few targetoid over extremities.Ana: 3+, ds Dna+.C/o acute cutaneous lupus erythematosus post viral exanthem

48 year old female,red,scaly skin lesions over face for 03 years.H/o photosensitivity,oral ulcers present.Examination revealed erythematous plaques with adherent scales over face, neck and posterior trunk.HPE:DLE. ANA- 4+, ds DNA positivity.Case of disseminated DLE with SLE

54 years,known case of Rheumatoid arthritis presented with multiple scaly lesions over upper back for 2 months. Examination revealed erythematous scaly plaques, erosions of the posterior trunk, petechiae on the legs. Ana 3+, ds DNA+. HPE: vasculitis.C/o SCLE. Rapidly progressed in skin lesions with acute cutaneous lupus erythematosus type.A case of SCLE with rapid progression to ACLE lesions in a case of RHUPUS.

Conclusion:

Rheumatologic diseases present with specific dermatologic manifestationsEarly recognition crucial for proper management.Skin biopsies and immunological workups are key in rheumatodermatologic diagnosis.However overlap in findings across different diseases can complicate interpretation making clinical correlation critical.Hence collaboration between dermatologists and rheumatologists is crucial.Presenting specific cases from the series that illustrate unique or atypical presentations of rheumatodermatologic diseases,emphasising diagnostic challenges and how these cases were eventually resolved.

Abstract N°: 171

Three Faces of a Line: Three Rare Cases With Facial Atrophic Linear Lesions

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Introduction & Objectives:

Embryological migration lines, also known as Blaschko lines, represent developmental growth patterns on the human body and mucous membranes. In this study, we present three patients with similar linear atrophic plaques on the right side of the face. Despite the resemblance in presentation, each patient was diagnosed with a distinct condition based on histopathological and clinical findings. These cases emphasize the importance of differential diagnosis and aim to raise awareness of the diverse etiologies of linear atrophic lesions of the face.

Materials & Methods:

This study describes three patients with linear atrophic lesions on the face, providing a detailed analysis of their phenotypic, histopathological, and clinical characteristics. Therapeutic approaches and the critical aspects of differential diagnosis are thoroughly examined to offer a comprehensive understanding of these cases.

Results:

Case 1: A 40-year-old female presented with a linear sclerotic, atrophic, hyperpigmented plaque on the right side of the scalp, extending from the frontal region. Additionally, she had cicatricial alopecia in three distinct areas of the scalp and erythematous, scaly, itchy annular plaques behind the ears. Dermoscopic examination revealed follicular plugs, white structureless areas, and superficial telangiectasias. Serological testing showed a positive antinuclear antibody (ANA) with a titer of 1/80 and a granular pattern, along with positive anti-SSA. A biopsy from the frontal hairline showed epidermal atrophy, basal cell vacuolar degeneration, and granular IgM deposits at the dermoepidermal junction on direct immunofluorescence (DIF). The patient was diagnosed with **discoid lupus erythematosus (DLE) mimicking en coup de sabre.**

Case 2: A 36-year-old female presented with a linear atrophic sclerotic plaque on the right forehead, nasal area, and chin, which first appeared at age 9. ANA was positive with a titer of 1/80, exhibiting a granular pattern. The patient also reported severe headaches, but cranial imaging showed no abnormalities. Histopathological examination revealed loss of follicular structures and diffuse subcutaneous fibrosis within the dermis. Based on clinical and histopathological findings, the patient was diagnosed with **en coup de sabre morphea.**

Case 3: A 35-year-old female presented with progressive concave deformity originating in the right frontotemporal region, gradually extending towards the forehead and nose over the past 12 years. A linear area of atrophy and regional indentation was noted, affecting the right third of the forehead and nasal region. ANA was positive with a titer of 1/320, showing a granular pattern, along with a strongly positive (+3) anti-DFS70. Cranial imaging revealed minimal atrophy of subcutaneous fat and soft tissues in the forehead and nasal areas. Based on clinical, imaging, and histopathological findings, the patient was diagnosed with **Parry Romberg syndrome.**

Conclusion:

These cases highlight the critical importance of differential diagnosis in patients presenting with linear Blaschkoid scleroatrophic plaques on the face. Early and accurate diagnosis and appropriate treatment are essential to preventing

complications. It is important to differentiate between diseases with similar clinical appearances, which may even mimic one another, and to consider these possibilities in clinical practice.

Abstract N°: 208

Generalized Sweet syndrome

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Introduction & Objectives:

Sweet syndrome (SS), also known as acute febrile neutrophilic dermatosis, is characterized by fever, elevated white blood cell count predominantly composed of neutrophils, painful red plaques on the skin, and dermal neutrophilic infiltration without vasculitis. SS can be triggered by various factors and is diagnosed based on clinical, laboratory, and histopathological criteria. SS is classified into three types: classical (or idiopathic), malignancy-associated, and drug-induced. Classical Sweet syndrome is the most common form, often linked to infectious diseases, inflammatory disorders, or pregnancy. Our main objective is to describe a patient with an aggravated and generalized form of Sweet syndrome who had a favorable outcome.

Materials & Methods:

A 59-year-old female presented to the emergency department with a three-week history of high fever, cough, and painful red lesions on her face, trunk, and extremities. Her medical history included long-term use of Clozapine for schizoaffective disorder. Initial psychiatric consultation raised suspicion of Clozapine-induced SJS/TEN, leading to the initiation of Risperidone.

Results:

The dermatologist noted erythematous, edematous, and painful plaques of varying sizes and target-like lesions. Laboratory tests revealed elevated white blood cell count and neutrophilia. Skin biopsy showed subcorneal pustules, eosinophils and neutrophils, spongiosis in the epidermis, and dense papillary edema with mixed inflammatory infiltrates. These findings confirmed Sweet syndrome. The patient did not meet the criteria for drug-induced SS, and malignancy was excluded, leading to a diagnosis of classic Sweet syndrome. Treatment with intravenous Prednisolone 100 mg was initiated and gradually tapered, resulting in significant clinical improvement without relapse or complications.

Conclusion:

Sweet syndrome is a rare condition requiring precise diagnosis based on clinical, laboratory, and histopathological evidence. Early and accurate diagnosis, combined with appropriate corticosteroid therapy, can lead to rapid symptom resolution and favorable outcomes, even in cases with extensive lesion involvement.

Abstract N°: 323

Plasma exchange for dermatomyositis

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Introduction & Objectives:

Dermatomyositis (DM) is a life-threatening idiopathic inflammatory myopathy that requires lifesaving treatment in severe forms. Plasma exchange (PE) involves removing the patient's plasma and replacing it with fresh frozen plasma. The aim of our study is to evaluate the efficacy of therapeutic plasma exchange as a first-line treatment for DM.

Materials & Methods:

This is a descriptive retrospective study. We included all patients with definite DM according to Peter and Bohan criteria who received first-line plasma exchange associated with corticosteroid therapy and/or immunosuppressants.

Results:

Six patients received PE. The mean age of onset was 62+ /-12 years, with a female predominance. All patients presented with a highly characteristic skin involvement consisting of periorbital erythema, Gottron's sign and Gottron's papules. All patients had muscular involvement, with bilateral, symmetrical proximal muscle weakness involving the scapular and pelvic girdles with an estimated average upper right and left muscle rating of 2.8+/-0.4 and an estimated lower right and left muscle rating of 2.6+ /- 0.5 according to MRC score. Three patients (50%) had severe pharyngoesophageal muscle weakness with dysphagia. All patients had a creatine phospho-kinase (CPK) assay prior to PE sessions.

For the treatment administered, oral corticosteroid therapy was prescribed for all patients, at a dose of 1.5mg/kg/d in 5 patients and 2mg/kg/d in one patient. Bolus corticosteroids at a dose of 1g/d for 3 days were administered in 2 patients presenting dysphagia and dyspnea. Immunosuppressive treatment was administered in 4 patients: azathioprine at a dose of 150mg/d in one patient and methotrexate at a dose of 25mg/week in 3 patients. For skin involvement, dermocorticoids and photoprotection were administered in all patients.

All patients in our series received plasma exchange, the average number of sessions being 3+/1. The mean skin surface area fell from 59.8 +/-19.9 to 37.8+ /- 15.5, and the CDASI activity score rose from 28.6+ /- 11.6 to 18.3 +/- 8.8.

Muscle strength and CPK values improved. The average right and left upper muscle score after the end of the plasmapheresis sessions was estimated at 3.8+/-0.4, and an estimated right and left lower muscle score of 3.5+ /- 0 ,5, with an average CPK that rose to 76 +/- 96 from 196 + /- 164.

For the 3 patients who presented dysphagia, we noted the disappearance of dysphagia with the possibilitý of oral feeding without a nasogastric tube after the 2nd plasma exchange session in one patient. A disappearance of dysphagia on D3 of the 3rd plasma exchange session in another patient, a stationary state in one patient after plasma exchange.

Conclusion:

Plasma exchange is a lifesaving treatment in severe forms of DM characterized by life-threatening pharyngoesophageal and respiratory muscle weakness. They are also more effective in acute forms of myositis.

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Abstract N°: 325

Factors associated with the severity of bullous pemphigoid

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Introduction & Objectives:

Bullous pemphigoid (BP) is the most common autoimmune subepidermal bullous blister disease. It is associated with tissue-bound and circulating autoantibodies directed against BP 180 and BP 230 antigens. The BPDAI score is approved by international guidelines to assess the severity of BP.

The aim of our study is to evaluate the biological factors correlated with the severity of BP.

Materials & Methods:

This is a retrospective descriptive and analytical study over an 8-year period from June 2015 to June 2022, including all patients with bullous pemphigoid (BP) selected on the basis of clinical, histological and immunological criteria. PB severity was calculated using the BPDAI score, with a BPDAI score >57 corresponding to severe involvement. A correlation study was performed using the spearman test. A $p<0.05$ threshold was considered statistically significant.

Results:

A total of 35 patients were enrolled, with an average age at diagnosis of 79.11+/-12.20 years, and a slight female predominance with a sex ratio of 1.05/ 1. Neurological disease was present in 17 patients (48.57%), including dementia in 8 patients (22.9%), Parkinson's disease in 2 patients (5.7%), stroke in 4 patients (11.4%), epilepsy in 2 patients (5.9%) and myxopapillary ependymoma in one patient.

The severity of bullous pemphigoid was assessed by BPDAI score: 28.6% had a low BPDAI score, 34.3% a moderate BPDAI score and 37.1% a severe BPDAI score. Anti-BP180 antibodies were positive in 71.4% of patients, with a mean of 7+/-5.6, and anti-BP230 antibodies were positive in 32.3%, with a mean of 1.2+/- 2. There was a highly significant ($p<0.001$) positive correlation ($R=0.746$) between BP severity and anti-BP180 antibody levels. However, no correlation was found between BP230 antibody levels, the presence of mucosal involvement, eosinophil levels and the severity of PB.

In our study, we observed a significant positive correlation between the severity of bullous pemphigoid (BP), as assessed by the BPDAI score, and the level of anti-BP180 antibodies ($R = 0.746$; $p < 0.001$). Our results corroborate those of the literature, where anti-BP180 antibodies are known to be associated with the most severe forms of PB.

On the other hand, it is interesting to note that in our study, anti-BP230 antibodies, although found in 32.3% of patients, showed no significant correlation with the severity of PB. This result suggests that anti-BP230 antibodies play a less predominant role in disease worsening. Furthermore, the lack of correlation between the severity of PB and other parameters such as eosinophil count and mucosal involvement reinforces the idea that anti-BP180 antibodies are the major predictive factor to be considered.

Indeed, anti-BP180 ELISA values > 27 U/mL have been reported to be predictive of relapse after treatment discontinuation. These tests may be considered before discontinuing treatment.

Conclusion:

This study highlights the central role of anti-BP180 antibodies as the main predictors of PB severity, reinforcing their

importance in the management and follow-up of this autoimmune disease. However, further studies are needed to identify other factors predictive of PB severity.

Abstract N°: 423

B-cell and T-cell cytofluorimetric repertoire characterize clinical response to Rituximab in Pemphigus vulgaris patients: long-term remission vs relapse

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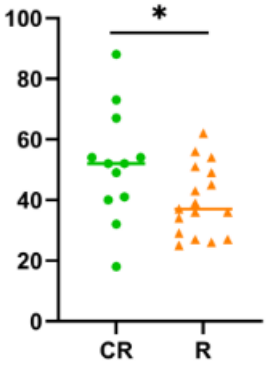
Introduction & Objectives: The immunological mechanism leading to Pemphigus vulgaris (PV) prolonged remission after Rituximab has not been elucidated yet. This study investigated the role of humoral and cellular immunity in mediating clinical response to B-cell depletion comparing a wide repertoire of B and T lymphocyte subpopulations between Rituximab-treated patients who achieved long term remission and those who relapsed.

Materials & Methods: The clinical course of** 29 PV patients treated with Rituximab was evaluated after a median 35-month follow-up. At the end of follow-up B and T cells were phenotyped by flow cytometric assay as follows: naïve B cells (CD19+ IgD+ IgM+ CD27-), transitional B cells (CD19+ IgM++ CD38++), pre-switch memory B cells (CD19+ IgD+ IgM+ CD27+), post-switch memory B cells (CD19+ IgD- IgM- CD27+), naïve helper T cells (CD3+CD4+ CD45Ra+) and memory helper T cells (CD3+CD4+ CD45RO+). CD24, CD25 and CD127 were additionally used to assess the following regulatory lymphocyte subsets: B reg naïve cells (CD19+ CD24hi CD38hi), B reg memory cells (CD19+ CD24hi CD27+) and T reg cells (CD4++CD127- CD25+). Any correlation was finally evaluated between B and T cell immunophenotyping and patients' clinical response to B cell depletion.

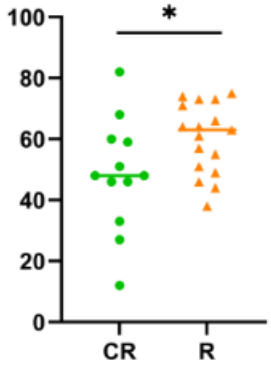
Results: After Rituximab 12* patients experienced long term remission while 17* relapsed at a median time of 33 months from the last infusion of the treatment cycle. At the end of follow-up relapsing patients reported a three-fold higher count and a four-fold higher frequency of post-switch memory B cells compared to patients in remission ($P<0.05$) while no difference was described in pre-switch memory and naïve B cell concentration. When assessing T cell subsets, a significant correlation was found between both naïve and memory helper T cells and therapeutic response with patients in remission reporting significantly higher frequency of naïve subset and significantly lower frequency of memory subset ($P<0.05$). No difference, instead, was described, according to clinical response, in peripherical immune regulation and similar values of B reg naïve cells, B reg memory cells and T reg cells were reported in relapse and remission groups.

Conclusion: The shift of B and T cells towards a memory phenotype is correlated to post-Rituximab relapse. The evaluation of a novel cytofluorimetric panel including both B and T cell phenotypes could provide a better understanding of the cellular mechanism of post-Rituximab long-term remission and improve the prognostic monitoring of PV.

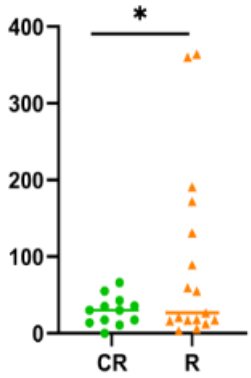
naïve helper T cells %



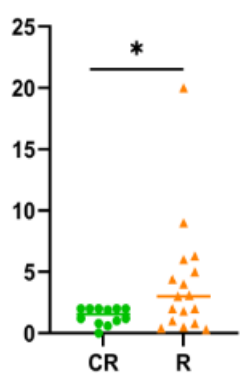
memory helper T cells %



post-switch memory B cells



post-switch memory B cells %



Abstract N°: 424

Remission maintenance with conventional immunosuppressants in Pemphigus vulgaris treated with Rituximab: a multicenter retrospective study

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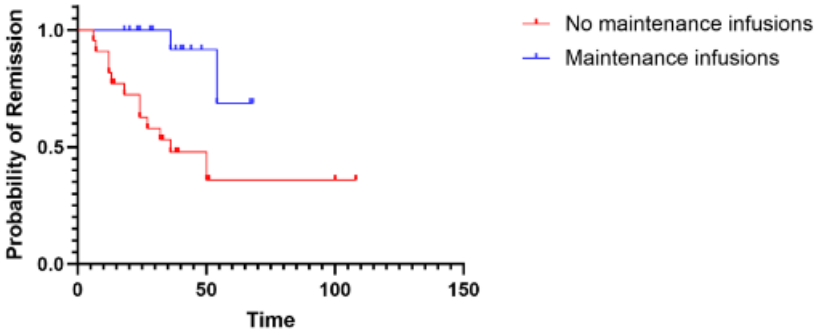
Introduction & Objectives: In Pemphigus vulgaris (PV) patients the impact of conventional immunosuppressants in post-Rituximab maintenance has been scarcely investigated and different treatment strategies are currently followed by clinicians to induce prolonged remission after B-cell repopulation. This study aimed to assess patients' clinical response to B cell depletion according to the type and duration of maintenance in order to evaluate the role of systemic steroids and adjuvant immunosuppressants in inducing Rituximab long-term effectiveness.

Materials & Methods: Forty recalcitrant PV patients treated with two doses of 1g Rituximab at two weeks apart were recruited in this retrospective study.** Patients' clinical course was evaluated after a minimum 18-month follow-up from the treatment cycle and data about the conventional immunosuppressive therapy administered after Rituximab infusions were collected. Different therapeutic strategies were eventually compared assessing the correlation between the type and duration of maintenance treatment and patients' response to B-cell depletion.

Results: After Rituximab all the patients received minimal doses of conventional immunosuppressants as maintenance: 27 (67,5%) patients were administered prednisone or prednisolone at a dose of less than or equal to 10mg/day while for the others 13 (32,5%) the same dosage of systemic steroids was used in association with mycophenolate mofetil at a dose of less than or equal to 1g/day. Eighteen (45%) patients were additionally treated with two maintenance infusions of Rituximab at 12 and 18 months from the first treatment cycle. During the study 26 (65%) patients achieved clinical remission off therapy while 14 (35%) relapsed at a median time of 24 months from the first infusion. Patients receiving maintenance Rituximab infusions reported significantly lower relapse risk (p-value<0,05*) and interestingly the only two patients who relapsed after 18-month maintenance dose were successfully treated with a short-lasting (less than 8 weeks) prednisone course without the need of additional Rituximab cycles. No correlation was described between the length and the type of maintenance treatment with conventional immunosuppressants and clinical response to B cell depletion.

Conclusion: Conventional immunosuppressants have a minor role in post-Rituximab management. Maintenance infusions are crucial for increasing Rituximab success rate and prolonging the duration of clinical remission.


Probability of Remission
1.0
0.5
0.0
0
50
100
150
Time
No maintenance infusions
Maintenance infusions

EA DV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 493

Gestational Pemphigoid and Plasmapheresis

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Introduction & Objectives:

Gestational pemphigoid (GP) is a rare autoimmune blistering dermatosis. It most commonly affects multiparous women and can occur at any stage of pregnancy, but especially during the 2nd or 3rd trimester and in the immediate postpartum period as well. The aim of this study is to describe the epidemiological, clinical, and therapeutic profile of gestational pemphigoid, while highlighting the key role of plasmapheresis in the management of this condition.

Materials & Methods:

We conducted a retrospective descriptive study of patients followed in our department, presenting with gestational pemphigoid confirmed by histological analysis and the presence of positive anti-BP180 antibodies.

Results:

Seven cases of gestational pemphigoid were studied, with a mean age of 27 years. Most patients were multiparous, and the disease typically occurred in the 2nd and 3rd trimesters. All patients experienced localized pruritus before lesions appeared. Initial lesions were mostly periumbilical, and patients exhibited tense vesicular and bullous lesions. The severity of lesions ranged from mild to severe, with 28.6% showing facial involvement.

The average skin surface affected was 26.29% ± 16.12, with values ranging from 7% to 45%. The median BPDAI score was 31 [14 - 75]. The severity of lesions was classified as mild in 1 case (14.28%), moderate in 4 cases (57.14%), and severe in 2 cases (28.60%).

The therapeutic management of our patients included:

Topical corticosteroids (clobetasol propionate: 30g/day) were used alone as the first-line treatment in 4 patients (57.14%). The median time for complete lesion resolution was 8 weeks [8; 8.45].

Oral corticosteroids (prednisone: 0.5 mg/kg/day) were used alone in 1 patient with extensive postpartum involvement. The time to complete lesion resolution was 5 months.

Two patients (28.6%) underwent a single plasmapheresis session each in the postpartum period:

In one case, plasmapheresis was performed as second-line treatment after failure of corticosteroids alone. The time to complete lesion resolution was 33 days after the plasmapheresis session.

In the other case, plasmapheresis was performed in combination with corticosteroids and topical corticosteroids as first-line treatment in a patient who refused further care and hospitalization. The time to complete lesion resolution was 16 days after the plasmapheresis session.

Tetracyclines (doxycycline: 200mg/day) were used in 1 patient postpartum. It was combined with clobetasol propionate

during the first recurrence, with a total resolution time of 4 months, and during the second recurrence in the same patient, in combination with a second-generation antihistamine, with a total resolution time of 3 months.

Regarding complications and adverse treatment effects:

Cataract, striae, and dyslipidemia were revealed in 1 patient treated with oral corticosteroids alone.

Fetal hypotrophy was revealed in 2 patients treated with topical corticosteroids alone (28.60%).

Conclusion:

Gestational pemphigoid (GP) is a rare dermatosis typically occurring in the 3rd trimester, with treatment mainly involving topical corticosteroids, and systemic corticosteroids for severe cases. Our study highlighted the significant efficacy of plasmapheresis, showing short remission times, suggesting its potential as a primary treatment. Further studies are needed to confirm its role in initial management.

Abstract N°: 524

Effectiveness and safety of Bimekizumab in Psoriasis, Psoriatic Arthritis and Hidradenitis Suppurativa diseases

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Introduction & Objectives:

Bimekizumab is the first and only selective dual inhibitor of IL-17 A and IL-17 F approved for the treatment of moderate-to-severe Psoriasis (P), Psoriatic Arthritis (PA) and Hidradenitis suppurativa (HS).

The objetive of this study is to evaluate the effectiveness and safety of Bimekizumab in patients with moderate-to-severe Psoriasis (P), Psoriatic Arthritis (AP) and Hidradenitis suppurativa (HS) diseases of Dermatology Service in a third-tier hospital from March 2023 to January 2025.

Materials & Methods:

Observational and retrospective study of patients treated for moderate-to-severe Psoriasis (P), Psoriatic Arthritis (PA) and Hidradenitis suppurativa (HS) with Bimekizumab of Dermatology Service in a third-tier hospital from March 2023 to January 2025.

The information has been obtained from the Electronic Clinical History (DRAGO-AE®) and the Pharmacy Service Managing Software (FARMATOOLS®).

Clinical variables collected were: sex, age, diagnose, dose and frequency of Bimekizumab, total skin clearance at weeks 4 and 16, adverse events, PASI (P and PA) and ISH4 (HS) before and after treatment, suspension of Bimekizumab.

Results:

31 patients were included in the study, mean age 49 yeaars old (17 women, 13 men) with diagnosis of P (23 patients), PA (3 patients) and HS (5 patients).

The doses of Bimekizumab were 320 mg every 8 weeks (with the induction dose every 4 weeks during the first 16 weeks) in patients with P, 160 mg every 4 weeks (without induction) in patient with PA and 320 mg every 4 weeks (with the induction dose every 2 weeks during the first 16 weeks) in patients with HS.

The average baseline PASI was 11,3 (±7.6) while the average PASI at the time of analysis was 0,5 (±0,5) in patients with P and PA. The average ISH4 before treatment was 20 (±11) and after was 10 (±5) in patients with HS.

100% of the patients had failed at least one previous biological therapy while 42% had received ≥2 previous biological therapies.

Of the 26 patients (P and PA) with follow-up at week 61% had achieved total clearance that increased at week 16 to 81% of patients achieving complete clearance.

No new safety alerts were identified, with the most frequently reported adverse effect being oral candidiasis in 3 patients, which did not lead to discontinuation of treatment. Therapy with Bimekizumab was discontinued in 2 patients (P).

Conclusion:

The effectiveness of Bimekizumab observed in our patients was similar or even higher than reported in pivotal clinical trials, with no new safety alerts.

In view of these results, Bimekizumab is an effective and safe alternative for patients with moderate-to-severe Psoriasis, Psoriatic Arthritis and Hidradenitis suppurativa.

Abstract N°: 600

Urticarial Vasculitis and Systemic Lupus Erythematosus association and treatment with Omalizumab: Case report

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Introduction & Objectives:

Urticarial vasculitis (UV) is a challenging condition that presents a reddish, itchy rash with histopathological findings of leukocytoclastic vasculitis. It is most commonly seen in connective tissue diseases such as systemic lupus erythematosus (SLE) or in Sjögren's syndrome [Alharbi S., et al 2019]. We present a patient with SLE who developed UV that was resistant to treatment.

Materials & Methods:

A case report.

Results:

A 52-year-old woman complained of a rash affecting her whole body. She also had swelling of her fingers and wrists, joint pain in her hands and muscle aches. The patient said the rash first appeared in August 2023. At that time, the patient was diagnosed with virus-induced erythema multiforme, and a treatment plan with prednisolone 30 mg was initiated. However, this treatment did not yield the anticipated results, and the skin lesions gradually subsided by December 2023. In May 2024, the lesions reappeared, this time on the patient's chest and thighs. Skin examination of the patient's trunk, upper and lower extremities revealed the presence of several reddish plaques, each measuring 2-4 cm in diameter. A skin biopsy and subsequent histological examination with direct immunofluorescence (DIF) showed perivascular inflammatory infiltration with lymphocytes and isolated neutrophils in the dermis, as well as perivascular fibrin extravasations. DIF revealed perivascular enhancement of fibrinogen, which allowed for the diagnosis of urticarial vasculitis. ANA was positive 3+, ACNA 1:320, C3 and C4 low at 0.67 and 0.10 g/l, and IgE was normal. The patient had several comorbidities, including hypertension, thyroiditis, Sjögren's syndrome with symptoms of xerostomia, xerophthalmia, Raynaud's syndrome, cryoglobulinemia, chronic glomerulonephritis, and monoclonal gammopathy of unknown significance. The patient also had polyneuropathy. Her renal failure was diagnosed as stage 3a and was thought to be a consequence of the glomerulonephritis. The diagnosis of SLE was made using a combination of history, skin examination and histological findings. The patient was discussed in a multidisciplinary team meeting, where methotrexate treatment had previously been administered and a positive response had been observed. However, subsequent deterioration in renal function led to discontinuation of treatment. The patient was intolerant to cyclophosphamide, and when rituximab 1000 mg was administered, minimal efficacy was observed. All treatment options were followed by high-dose corticosteroids. However, the use of methotrexate is dependent on renal function and was not initialized, so we started treatment with omalizumab 300 mg/monthly. After a few weeks on the new treatment, the lesions disappeared and the patient has no symptoms or complaints. However, the patient still requires close management and collaboration between several healthcare professionals in order to achieve clear skin and joint pain relief.

Conclusion:

IgE antibodies have been implicated in the pathogenesis of SLE in the literature, and omalizumab, an anti-IgE monoclonal

antibody, has been shown to improve SLE by reducing the production of theses molecules [Hasni S, et al. 2020]. This proves why our patient responded well to omalizumab, although poor effects of high-dose corticosteroids and other treatments were seen in her UV and SLE.

Abstract N°: 610

Linear IgA bullous dermatosis of multifactorial etiology

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Linear IgA bullous dermatosis of multifactorial etiology

Introduction & Objectives:

Linear IgA bullous dermatosis (LABD) is an immune-mediated blistering dermatosis characterized with linear deposition of IgA at the dermo-epidermal junctions. So far, the pathogenesis of the disease remains unknown. According to the literature drugs (e.g. vancomycin, trimethoprim-sulfamethoxazole) or neoplastic process (e.g. lymphomas, leukemias, lung cancers) could be potential triggering factors for LABD. The course of the disease (clinical manifestation, treatment response, relapse rate) is usually more severe in drug-induced LABD.

Materials & Methods:

69-year-old male patient was consulted due to scattered, annular vesiculobullous eruptions arising on an inflammatory base located on the trunk and limbs accompanied with targetoid, erythema-edematous lesions on thighs and back. Vesicles were also located on vermilion zone, but the mucosa was not affected. Skin lesions appeared three months after the patient had performed transplantation of both lungs due to chronic obstructive pulmonary disease. Pathomorphological result of the patient's left lung revealed presence of squamous cell carcinoma (pT2b, N0, R0). As a part of post-transplant lung infection prevention, the patient received trimethoprim-sulfamethoxazole as well as allopurinol due to increased level of uric acid.

Results:

Histopathological examination suggested LABD. Direct immunofluorescence (DIF) result showed abundant linear deposition of IgA at the dermo-epidermal junction what confirmed the diagnosis of LABD. Treatment with prednisone 35 mg orally per day was initiated together with topical treatment of clobetasol ointment – a gradual clinical improvement was observed. Trimethoprim-sulfamethoxazole was not withdrawn as the post-transplant prophylaxis at that time was of utmost importance. In a long-term perspective treatment with dapsone was planned but the patient did not consent. In the literature several cases of LABD in course of a lung cancer as well as LABD induced by trimethoprim-sulfamethoxazole or allupurinol are reported.

Conclusion:

In case of our patient, both the lung cancer and the intruduction of new drugs could potentially elicit the eruption of LABD. The patient is currently under oncological supervision with further oncological diagnostics being in progress.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 633

Retrospective Analysis of Patients with Pemphigus from 2018 to 2023: Clinical and Social Characteristics, Factors of Disability, and Disease Onset Patterns

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Introduction & Objectives:

Pemphigus is a rare autoimmune disease with a chronic, relapsing course that significantly impacts patients' quality of life. Studying its clinical manifestations, disability factors, and social aspects can help optimize therapeutic strategies and improve patient rehabilitation.

To analyze the clinical and socio-demographic characteristics, frequency of relapses, and factors contributing to disability in patients with pemphigus treated between 2018 and 2023.

Materials & Methods:

A total of 412 medical records were reviewed (198 men and 214 women), with patient ages ranging from 20 to 77 years, all diagnosed with pemphigus.

Results:

- **Gender and Age Characteristics**: The mean onset age was 45 years in men and 41 years in women.
- **Clinical Course and Onset Patterns**:
  - Of the 198 men, 36 (18.2%) initially presented with lesions on the oral mucosa.
  - Among the 214 women, 94 (43.9%) developed their first symptoms as trunk skin lesions.
- **Disability**:
  - Seventy-two patients (17.5%) were classified with second-degree disability due to severe chronic relapses, widespread mucocutaneous involvement, and limited response to standard therapies.
- **Frequency of Rehospitalizations**:
  - Twenty-one patients (5.1%) underwent multiple (5–8) inpatient treatment cycles because of frequent relapses and resistance to basic therapy.
- **Social Aspects**:
  - Two hundred and forty-two individuals (58.7%) were unemployed, reflecting decreased work capacity, societal stigma, and the need for prolonged or repeated medical interventions.

Conclusion:

These findings indicate considerable variation in clinical onset between men and women, a high rate of relapse, and notable disability. Timely identification of mucosal lesions, alongside individualized therapeutic and rehabilitation approaches, is essential for effective disease management. Addressing social and employment barriers remains a critical component in improving overall treatment outcomes and enhancing quality of life for individuals with pemphigus.

Abstract N°: 635

Exploring the Uncommon: A report of Two Rare Pemphigus Cases

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Exploring the Uncommon: A report of Two Rare Pemphigus Cases

Introduction & Objectives:

Pemphigus herpetiformis (PH) and IgA pemphigus are rare entities of the bullous autoimmune diseases group characterized by immunologic findings consistent with pemphigus but with unique clinical and pathologic presentations. We herein report two recent cases from our clinic to raise awareness of rarer forms of the bullous pathologies that can pose diagnostic and therapeutic difficulties.

Materials & Methods:

Case 1. An 57-year-old male presented with a 3 weeks history of pruritic pustules, with a tendency to confluence, initially located on the extremities, with subsequent extension. The clinical exam also reveals fragile bullae and isolated erythematous plaques with annular disposition, with pustules at the periphery and central hematic crust. Biopsy reveals subcorneal pustulosis, neutrophils and marked spongiosis. The identification of IgA and C3 deposits at direct immunofluorescence (DIF) confirms the diagnosis of IgA pemphigus. The appearance of new lesions is now controlled with dapsone and low, decreasing doses of systemic corticosteroids.

Case 2.** An 46-year old woman presented with a 10-month history of erythematous lesions, covered with fine scales and crusts, located bilaterally on the legs. The first biopsy taken revealed acute dermatitis. The patient was treated for 8 months with antihistamines and corticoid cream without improvement. The clinical exam reveals numerous vesicles and pustules isolated and grouped in clusters on erythematous, itchy plaques, located on the shins and dorsum of the hands. Rebiopsy of lesions was considered. The diagnosis of pemphigus herpetiform was confirmed histopathologically and by DIF. A therapeutic regimen with dapsone was initiated with satisfactory clinical results, but with discontinuation after 3 months because of abnormal liver samples, with initiation of systemic corticosteroid therapy.

Results:

PH and** IgA pemphigus stands out as two of the most uncommon types of pemphigus, with a limited understanding of their epidemiology. Diagnosis and subtype determination rely on histopathology and immunofluorescence findings. IgA pemphigus manifests as painful, itchy blisters that fill with neutrophils and evolve into pustules. PH shares clinical characteristics with dermatitis herpetiformis and immunologic features with pemphigus It is characterized by an intense inflammation that may not be associated with acantholysis. There is no universal treatment for this disorders, but many patients do respond to dapsone and steroids.

Conclusion:

This cases underscore the importance of considering PH and Pemphigus with Ig A in the differential diagnosis of vesicobullous diseases and the need for further research to elucidate their pathogenesis and optimal management. The clinical presentation of these conditions is remarkably similar, and careful investigation using histology and DIF is necessary to differentiate them.

Abstract N°: 637

Unveiling Chilblain Lupus Erythematosus: A Rare Skin Manifestation of Autoimmune Disease

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Introduction & Objectives:

Chilblain lupus erythematosus (CHLE), also known as 'Hutchinson lupus", is a rare skin disorder, with only a limited number of cases reported to date.

This condition is a subtype of chronic cutaneous lupus erythematosus, an autoimmune inflammatory disease affecting the skin. It can occur in individuals with or without systemic lupus erythematosus (SLE). Approximately 20% of patients with chilblain lupus erythematosus eventually develop SLE.

The condition typically manifests as a rash on acral surfaces most exposed to cold, such as the toes, fingers, ears, and nose. The rash is characterized by tender, purple plaques, nodules, sometimes with central erosions or ulcerations, and oedematous skin. The exact pathophysiology remains unclear, but it is believed to be autoimmune in nature, involving altered microcirculation, stasis, and vascular occlusion triggered by exposure to low temperatures. Chilblain lupus erythematosus may be diagnosed based on clinical presentation and histopathological examination or direct immunofluorescence study.

This case report aims to highlight the potential role of CHLE as an early manifestation of SLE, emphasizing the importance of timely recognition and appropriate management of this condition.

Materials & Methods:

We present the case of a 56-year-old female patient with a history of Hepatitis C, who presented to the Dermatovenerology Department in Iași for the first time with erythematous-violaceous plaques covered by serohematic crusts, located on her central face, mainly the nose, and dorsal hands. The lesions were painless and exacerbated by cold exposure, having first appeared about four months prior to her hospital admission in late February. The patient reported a similar episode the previous winter, which had resolved with the arrival of warmer weather in late spring. To assess the patient's condition, a skin biopsy from the dorsal side of one hand was performed.

Results:

Laboratory results revealed elevated levels of C-reactive protein, erythrocyte sedimentation rate, SS-A, SS-B, and ANA antibodies, with normal rheumatoid factor levels. Histopathological examination of the skin biopsy confirmed lupus chilblain, characterized by keratinized stratified squamous epithelium with hyperkeratosis and parakeratosis, an area of ulceration, and underlying lymphoplasmacytic inflammatory infiltrates in the dermis and hypodermis. Edema, vascular ectasia, and congestion were also observed. Van Gieson staining showed collagen and elastic fibers without pathological changes. Therefore, lupus pernio was confirmed as a manifestation of an ongoing SLE.

Conclusion:

Chilblain lupus erythematosus is a rare chronic condition that predominantly affects women. While it is less severe than SLE, it can serve as an early indicator of various underlying autoimmune diseases. Physicians should approach CHLE with caution, as its symptoms can be subtle and often resemble those of other similar conditions.

EA DV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 711

Eosinophilic fasciitis presenting as unilateral edema in a 40-year-old female

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Eosinophilic fasciitis presenting as unilateral edema in a 40-year-old female

A 40-year-old female presented with a five-year history of progressive enlargement, induration, erythema, and hyperpigmentation of the right arm and leg, accompanied by occasional burning pain and pruritus. She denied Raynaud phenomenon, travel, medication or supplement intake, and exposure to toxic agents. She had occasional rhinorrhea and unexplained 6-kilogram weight loss over 6 months. She had controlled hyperthyroidism, a history of smoking (1.5 pack years), and no illicit drug use. She worked as a vendor and laundrywoman.

Physical examination revealed diffuse, non-pitting edema, erythema, and induration of the right arm up to the shoulder, as well as the right leg up to the hip, extending to the left trunk and buttocks**.** The right hand was affected but the face and feet were spared. She had no arthritis nor peripheral neuropathy. Systemic examination was unremarkable save for non-tender, right cervical and axillary lymphadenopathy.

Data Review *(see Figure 1)*

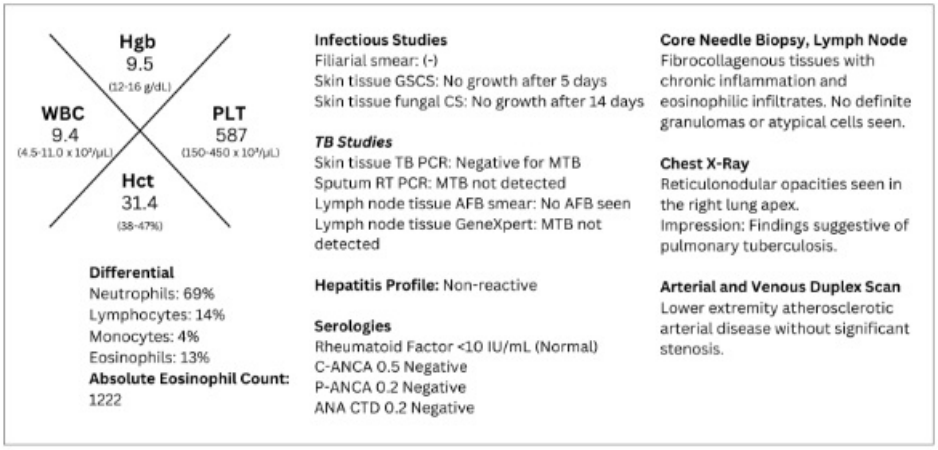
The patient had anemia and peripheral eosinophilia. Anti-neutrophil cytoplasm antibody (ANCA) serologies and tuberculosis work-up were negative. Chest computed tomography (CT) scan with contrast showed homogenous non-enhancing subcutaneous thickening of affected tissue and several apical nodular densities, some with spiculated borders. Lymph node biopsies showed no granulomas nor atypical cells. A wedge biopsy of the chest revealed eosinophilic cellulitis with granulomatous vasculitis**.**

Conclusion

Albeit atypical in presentation, the overall picture was consistent with eosinophilic fasciitis. Concomitant malignancy could not be ruled out given the pulmonary nodules, progressive anemia, weight loss, and lymphadenopathy. Bone marrow aspiration, total protein and albumin/globulin ratio test, eosinophilia FISH panel, and abdominal CT scan were planned. She was started on low-dose oral prednisone (0.2 mg/kg daily), with plans to increase dose after further work-up; however, she was unwilling to pursue further invasive tests and was lost to follow-up.

The rarity of eosinophilic fasciitis demands a higher index of suspicion in similarly-appearing patients. Multispecialty collaboration is essential, especially in complex cases with concomitant pathologies.

Figure 1. Laboratory results



Abstract N°: 809

Refractory bullous pemphigoid with mucosal involvement in young patients: Overcoming resistance with rituximab

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[2]2nd Department of Pathology, National and Kapodistrian University of Athens, "Attikon" General University Hospital, Medical School, Athens, Greece

Introduction & Objectives: Bullous pemphigoid (BP) is an autoimmune blistering disease primarily affecting elderly patients. Its occurrence in young individuals is rare and presents a significant therapeutic challenge. Our objective is to present two cases of refractory BP in young female patients, aged 30 and 40, who failed multiple standard treatments but achieved disease control with rituximab.Τέλος φόρμας

Materials & Methods: We retrospectively analyzed the two BP cases, extracting diagnostic findings, disease course, treatment history, and therapeutic response from medical records. Diagnosis was confirmed by histopathology, direct immunofluorescence of perilesional skin, and indirect immunofluorescence.Τέλος φόρμας

Results: Both patients presented with extensive bullae and erosions affecting the skin and oral mucosa. Initial treatment with high-dose systemic corticosteroids and various immunosuppressive agents, including azathioprine, mycophenolate mofetil, cyclosporine, methotrexate, and doxycycline/nicotinamide, failed to achieve disease control due to either lack of efficacy or adverse effects. Persistent disease activity, along with steroid-induced complications, necessitated treatment with rituximab, following the dosing regimen used for pemphigus vulgaris (1 g per infusion at weeks 0 and 15).

Following rituximab therapy, both patients exhibited significant clinical improvement with progressive lesion resolution. Corticosteroids were successfully tapered without relapse, and no rituximab-related adverse events were reported. Both achieved sustained remission, which remains ongoing.

Conclusion: The present study highlights an uncommon presentation of BP in young patients and emphasizes the therapeutic potential of rituximab in refractory cases. In patients unresponsive to conventional immunosuppressive therapy, early consideration of B-cell–depleting agents may be beneficial for achieving sustained disease remission while minimizing corticosteroid-related complications.

EA DV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 828

Bullous Pemphigoid and Gliptin: A Case Report.

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Introduction:

Bullous Pemphigoid (BP) is an autoimmune blistering dermatosis, primarily observed in elderly individuals. Several risk factors contribute to its onset, including medications such as dipeptidyl peptidase-4 inhibitors (DPP-4), or gliptins, which have been recently implicated in the development of BP.

Case report:

We report the case of a 74-year-old patient with acral vitiligo for 20 years and type 2 diabetes for 10 years, initially treated with metformin and sulfonylureas. Three years ago, vildagliptin, a DPP-4 inhibitor, was introduced. The patient presented to dermatology with bullous lesions that appeared a month prior.

Dermatological examination revealed:

- Multiple tense blisters containing serohematic fluid on an erythematous base, evolving into erosions and crusts, localized on the trunk, axillae, and proximal lower limbs.

- Negative Nikolsky's sign.

- Moderate pruritus.

- No mucosal involvement.

The diagnosis of BP was confirmed by histology, showing a subepidermal blister with eosinophilic infiltrate and linear deposits of immunoglobulin G and C3 at the basement membrane on direct immunofluorescence. Anti-basement membrane antibodies were negative on indirect immunofluorescence. No hypereosinophilia was observed.

Treatment with topical steroids class IV (30g/day) and systemic corticosteroids (0.5 mg/kg/day) was initiated, resulting in rapid clinical improvement.

Discussion:

Bullous pemphigoid is an autoimmune blistering dermatosis characterized by tense blisters on erythematous or urticarial skin, often with pruritus. Lesions predominantly affect the trunk and the proximal limbs, sparing the face and mucous membranes. It results from an immune response to BP180 and BP230 antigens, crucial for epidermal-dermal adhesion.

Oral antidiabetic DPP-4 inhibitors, particularly vildagliptin and linagliptin, are risk factors for BP, with an incidence of 0.42 cases per 1000 person-years. The exact mechanism remains unclear, but some studies suggest a link with the HLA DQB1 haplotype. BP induced by gliptins typically appears months to years after treatment.

Induced forms differ clinically, histologically, and biologically from spontaneous forms. They are characterized by lesions predominantly on the trunk and proximal limbs, with a less inflammatory phenotype, moderate skin involvement (< 10 new blisters per day), and mild pruritus. Furthermore, eosinophilic infiltrates in the dermis and blood, as well as serum levels of BP180 and BP230 autoantibodies, are significantly lower, which is consistent with our case.

Recommendations on stopping DPP-4 inhibitors are conflicting. Some studies show quick remission after discontinuation, while others see no impact on disease progression or relapse. Generally, stopping DPP-4 inhibitors, along with topical steroids class IV (10-30g/day) or low-dose systemic corticosteroids (0.5 mg/kg/day), leads to remission.

Conclusion:

This case highlights the potential association between DPP-4 inhibitors and the development of bullous pemphigoid in an elderly patient. It underscores the importance of vigilance when prescribing gliptins and considering the diagnosis of BP in diabetic patients.

Abstract N°: 855

Comparing the effectiveness of oral methotrexate and phototherapy vs phototherapy alone for the treatment of vitiligo

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Comparing the effectiveness of oral methotrexate and phototherapy vs phototherapy alone for the treatment of vitiligo

Introduction & Objectives: Vitiligo is an acquired skin condition that affects 0.5% to 2% of people worldwide and tends to affect both sexes. There is some indication that autoimmunity plays a part in this condition, even though the precise pathogenic mechanism is unknown. Numerous immunosuppressive medications, including systemic or topical corticosteroids and phototherapy (particularly narrowband UVB), are employed in accordance with this notion. Another immunosuppressant that has gained popularity recently as a stand-alone treatment for vitiligo is methotrexate; nevertheless, two important aspects that are still unclear are its superiority over alternative treatments and its synergistic effect. The purpose of this study was to assess the effectiveness of methotrexate+NB-UVB with placebo+NB-UVB in patients with vitiligo.

Materials & Methods: 60 patients were randomly assigned to one of two groups in this double-blind, randomized controlled trial: the first group received NB-UVB three times a week along with a placebo, while the second group received NB-UVB three times a week together with a weekly dose of 12.5 mg MTX. Patients were monitored every two months for the length of the 6-month treatment, and VASI (repigmentation indicator) and VIDA (disease activity indicator) scores served as the evaluation instruments.

Results: During the 6-month follow-up, both treatment groups' VASI and VIDA scores improved, but there was no statistically significant difference between the two approaches.

Conclusion: This study showed that all treatment approaches were equally successful; however, more research with a bigger sample size and longer follow-up is needed to assess the effectiveness of MTX in combination with other drugs.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 864

Labial Adhesions as a Rare Mucosal Manifestation of Bullous Lupus Erythematosus in a 15-year-old Female

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Introduction:

Bullous lupus erythematosus is a rare cutaneous manifestation of lupus erythematosus with a variable presentation. Mucosal manifestations in lupus usually include oral or nasopharyngeal ulcerations, ocular manifestations, serositis, among many others; however, genital lupus is only reported in a few case reports.

Case Report:

The patient, a 15-year-old female, presented with a 5-year history of multiple, intensely pruritic, erythematous vesicles and bullae that erode and form erythematous erosions and milia formation. She also developed vaginal ulcerations with erythematous borders that caused her labia to fuse causing difficulty in urinating. A biopsy of blister edge on the left shoulder showed a subepidermal split with vacuolar interface change and dense diffuse neutrophilic infiltrates. Immunohistopathologic findings of perilesional skin showed basement membrane zone deposition of IgG, IgM, IgA. She had positive ANA 1:80, but normal anti-dsDNA and C3 levels. Hormonal work-up showed hypogonadotropic, hypogonadism, and transperineal/transabdominal/transrectal ultrasound showed infantile uterus, thin endometrium, and silent ovaries. Since she also has developmental delays, Turner Syndrome was considered. It was later ruled out after karyotyping results showed normal female karyotype. Complete blood count showed anemia for age with direct Coomb's test positive (+1). Urinalysis showed albuminuria, hematuria, and positive leukocytes which may indicate possible lupus nephritis or urinary tract infection. Urine protein-creatinine ratio was 0.23 indicating microalbuminuria. The final assessment was bullous lupus erythematosus and early systemic lupus erythematosus (SLE). Epidermolysis bullosa acquisita (EBA) was a very close differential, due to formation tense blisters in areas of trauma, the characteristic scarring, and the histopathologic and DIF findings similar with bullous LE. However, vacuolar interface changes favor more bullous LE than EBA. Initial management was mid-potent topical steroids, photoprotection with sunscreen, and wound care. She was started on hydroxychloroquine and underwent labiaplasty using Z-plasty technique under spinal anesthesia. Colchicine was added after and there was no noted recurrence of the strictures.

Conclusion:

Labial adhesions and strictures are very unusual presentations in the setting of bullous LE. Lack of appropriate treatment could lead to grave consequences like repeated urinary tract infection and kidney failure. Multidisciplinary approach using combined oral and topical immunomodulators, together with surgical intervention, may be considered in patients already presenting with bothersome genitourinary symptoms.

Abstract N°: 925

Pemphigoid Nodularis : An Uncommon Presentation of Bullous Pemphigoid: A Case Report

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Pemphigoid Nodularis : An Uncommon Presentation of Bullous Pemphigoid: A Case Report

Introduction & Objectives:

Bullous pemphigoid (BP) is an autoimmune subepidermal blistering disorder primarily affecting the elderly. While the classical form presents with tense bullae, atypical variants, such as nodular prurigo-like BP (pemphigoid nodularis PN), can mimic chronic prurigo, delaying diagnosis and treatment. We report a case of PN in a Moroccan patient with metabolic comorbidities, underscoring the need for histopathological and immunological confirmation.

Materials & Methods:

A 72-year-old Moroccan male with type 2 diabetes mellitus and hypertension presented with intensely pruritic nodules on the trunk, back, and limbs, evolving for one month. Examination revealed multiple excoriated, violaceous nodules with atrophic changes, sparing the face and mucosa. Given the clinical presentation, prurigo nodularis was initially suspected, but the acute onset in an elderly patient raised suspicion of an underlying autoimmune disorder.

Laboratory tests, including autoimmune markers, were unremarkable. A skin biopsy revealed an acanthotic epidermis with a subepidermal blister and eosinophilic infiltrates. Direct immunofluorescence revealed linear C3 deposits along the basement membrane. Enzyme-linked immunosorbent assay (ELISA) detected BP230 antibodies (79.0 U/ml), while BP180 antibodies were negative, confirming PN.

The patient was treated with oral prednisone (0.5 mg/kg/day) tapered over six weeks and topical clobetasol. Pruritus improved significantly, and nodular lesions regressed. After three months, the patient showed marked improvement with no recurrence.

Results:

Nodular BP, or pemphigoid nodularis, is a rare clinical variant of BP that closely mimics prurigo nodularis, leading to frequent misdiagnoses and treatment delays. Autoantibodies targeting BP230 disrupt keratinocyte adhesion, distinguishing it from classical BP, which is typically associated with BP180. The absence of bullous lesions complicates diagnosis, requiring histopathological and immunological confirmation.

Systemic corticosteroids are the mainstay treatment, but long-term use poses risks, particularly in patients with diabetes and hypertension. Steroid-sparing agents such as antihistamines, leukotriene receptor antagonists, doxycycline, or dapsone can be useful. Recent studies have suggested that biologics, including rituximab and omalizumab, may be beneficial for refractory cases. In our patient, a careful balance was maintained between disease control and minimizing corticosteroid-related complications, leading to successful remission.

Conclusion:

This case highlights the importance of considering BP in elderly patients presenting with chronic pruritic nodules, particularly those unresponsive to conventional prurigo treatments. Early recognition, histopathological confirmation, and targeted immunological studies are essential for appropriate diagnosis and management.

EA DV SYMPOSIUM
PRAGUE
22-24 MAY 2025

Abstract N°: 963

The role of neutrophil-to-lymphocyte ratio in bullous pemphigoid: systematic review

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Introduction & Objectives:

Bullous pemphigoid (BP) is a chronic autoimmune blistering disorder characterized by subepidermal blister formation and inflammatory processes affecting the skin and mucous membranes. Recent studies have explored the ratio between the number of neutrophils and lymphocytes (NLR) in the peripheral blood as a potential biomarker in various autoimmune and inflammatory disorders. However the role of NLR has not been clearly defined and remains poorly studied in BP. The aim of this study was to evaluate the role of NLR as a potential biomarker in BP.

Materials & Methods:

A systematic search was conducted in MEDLINE, EMBASE, and Google Scholar databases according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. Studies assessing the role of NLR in BP were included in the analysis. The methodological quality of each study was assessed using the Newcastle-Ottawa Scale (NOS).

Results:

A total of 21 articles were identified and screened, 8 full texts were reviewed and 7 studies were included in the analysis. Only one study was performed prospectively and the remaining studies were retrospective. A total of 428 patients were included. The mean age of the patients was 72,6 years (SD 9.02). There was wide heterogeneity in the use of NLR in patients with BP, however 5 of the seven (71%) studies showed that NLR had a positive predictive value in BP. NLR's diagnostic role was evaluated in four studies, with three (75%) showing statistical significance. Therapeutic role of NLR was further assessed in one study and prognostic role in two studies. Only one of these studies compared NLR between patients with BP and pemphigus disease (PD) at the time of initial presentation, showing higher values in BP patients. The methodological quality of the studies according to NOS was moderate to high.

Conclusion:

Despite limited literature, NLR holds promise as a non-invasive and cost-effective biomarker in BP. The results indicate that NLR is applicable across multiple settings in BP, particularly as a diagnostic and prognostic tool. Retrospective nature of majority of the studies and small sample of patients was noteworthy. Future well-designed prospective studies are necessary to validate its role as a BP biomarker.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 987

Beyond the Rash: A Rare Early-Onset Case of Juvenile Dermatomyositis

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Introduction:

Juvenile Dermatomyositis (JDM) is a rare autoimmune condition marked by muscle weakness, skin lesions, and joint inflammation, typically affecting children aged 5-10, with a female predominance. Early symptoms include fever, dysphagia, and myalgias. This case report presents an early onset of DM emphasizing dermatological clues for diagnosis and importance of timely intervention.

Case Presentation:

A 5-year-old girl presented with a 1.5-year history of erythematous papules and plaques spreading across her body, accompanied by moderate to severe pruritus, increase in pigmentation and photosensitivity. Initial treatments for presumed atopic dermatitis were ineffective. Dermatological findings include Heliotrope rash, Gottron papules, Gottron sign, V-neck sign, Shawl sign, Holster sign, ovoid palatal patch, poikiloderma, and generalized xerosis. This was supported by elevated creatine kinase and other enzyme levels, and a confirmatory diagnosis with electromyography and a skin biopsy consistent with the disease. The patient responded well to topical steroids and immunosuppressants, showing marked improvement of lesions.

Conclusion:

This case report underscores the crucial role of dermatologists in recognizing dermatological clues, even in the absence of muscle weakness, for an accurate diagnosis. Timely diagnostic testing, including skin biopsy and electromyography, was essential in confirming the patient's condition. Although more common in adults, early-onset juvenile dermatomyositis may occur, as in this case. Early detection is vital to prevent severe complications, emphasizing the importance of heightened clinical awareness of the distinctive features of Juvenile Dermatomyositis. This awareness ensures not only an early diagnosis but also the prompt initiation of appropriate management, ultimately improving patient's quality of life.

Abstract N°: 1084

The Lupus Hand

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Introduction & Objectives:

Cutaneous manifestations of lupus affect approximately 80% of patients during the course of the disease, and hand involvement is common. The lesions can be lupus-related or non-lupus-related and may mimic vascular lesions, which increases the risk of diagnostic error. The aim of this work is to describe hand involvement in lupus patients managed in our department.

Materials & Methods:

This is a retrospective descriptive study conducted in the Dermatology Department of Mohammed VI University Hospital in Oujda over a 10-year period, from December 2014 to July 2024, including patients with lupus managed in our department.

Results:

Our study included 29 lupus patients hospitalized during the studied period, with 10 patients having systemic lupus and 19 having chronic cutaneous lupus. There was a clear female predominance with a female-to-male sex ratio of 2.6. The average age of our patients was 46 years, with ages ranging from 28 to 63 years.

Hand involvement was present in 17 patients, accounting for 58.6% of the cases. The most common lesions were erythematous-squamous and atrophic lesions, seen in 47% (8) of the cases, followed by papulo-nodular lesions with a keratotic center in 35% (6) of the patients, and atrophic hypochromic lesions in 11.7% (2) of the patients. These lesions were located on the dorsal surfaces of the hands and fingers, sparing the interphalangeal and metacarpophalangeal joints.

Only one patient had lupus pulpitis, and erosions were noted in 11.7% of the cases. Chilblains were observed in one patient, and Raynaud's phenomenon was noted in 17.6%.

Regarding nail involvement, cuticular hypertrophy was found in 17.6% (3) of the patients, distal onycholysis was present in 23.5% (4), longitudinal striations in 47% (8) of the cases, and multi-digit melanonychia in one patient on hydroxychloroquine. Periungual capillaroscopy revealed mega-capillaries in 29.4% (5) of the patients, flame hemorrhages in 5.9% of cases, and avascular areas in 17.6% (3) of the patients.

Conclusion:

Cutaneous lesions on the hands in lupus can be lupus-specific, with polymorphic lesions, or non-lupus-related. They are sometimes difficult to define and pose a diagnostic challenge, which is why biopsy is often necessary for a more accurate diagnosis.

Abstract N°: 1093

IMACS Guidelines: An External Validation in Dermatomyositis Patients at 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 at a metropolitan academic center.

Materials & Methods:

Charts for DM patients were reviewed retrospectively for patients with a known DM diagnosis and 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 autoantibodies, 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 137 patients with a diagnosis of DM. 15% (n=21) patients had a diagnosis of cancer at any time period, with 6.5% (n=9) classified as paraneoplastic DM. Most patients were female, comprising 79% of non-paraneoplastic and 89% of paraneoplastic patients. The mean age at disease onset for paraneoplastic patients was 61.3 ± 13.4, and for non-paraneoplastic patients was 47.6 ± 18.2. Patients were stratified by the risk factors outlined in the IMACS guidelines into high-, intermediate-, and low-risk factors. 67% of paraneoplastic DM patients were categorized as high risk, compared to 53.1% of non-paraneoplastic patients. In our sample, TIF1y antibodies were statistically significant for an increased risk of paraneoplastic DM (p=0.02). Of the 9 paraneoplastic DM cases, all 4 breast cancer patients were detected using age-appropriate screenings via mammograms, both lung cancer patients were detected via chest x-rays with subsequent chest computed tomography (CT), and both uterine cancer patients were detected via abdominal CT. The one patient with cervical cancer presented with vaginal bleeding at age 78 following the initiation of blood thinners and not via post-diagnostic screening.

Conclusion:

Our results provide a validation for the guidelines set forth by the IMACS, with nearly all paraneoplastic DM patients (n=8) being found via the screening guidelines set forth by the IMACS guidelines. These findings suggest that the screening guidelines are appropriate for the identification of paraneoplastic DM, leading to more targeted and risk-based screening. Future large-sample analyses in diverse populations are needed to further validate these screening guidelines.

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

	Non-Paraneoplastic (n=128) N(%)	Paraneoplastic (n=9) N(%)	P-Value	Odds Ratio	95% CI
High Risk Factors					
Dermatomyositis	60 (46.9)	5 (55.6)	0.74	1.41	0.29–7.46
Anti-TIF1y antibodies	7 (5.5)	3 (33.3)	0.02*	8.37	1.12–51.0
Anti-NXP2 antibodies	6 (4.7)	0 (0)	1.00	0.00	0–13.4
Age ≥40 at IIM onset	86 (67.2)	8 (88.9)	0.27	3.88	0.49–177
Persistent high disease activity despite therapy	3 (2.3)	0 (0)	1.00	0	0–37.1
Dysphagia (moderate to severe)	31 (24.2)	4 (44.4)	0.23	2.48	0.46–12.4
Cutaneous necrosis	7 (5.5)	1 (11.1)	0.43	2.14	0.04–20.6
≥ 2 'High risk' factors	68 (53.1)	6 (66.7)	0.51	1.76	0.36–11.3
Intermediate Risk Factors					
Amyopathic Dermatomyositis, Polymyositis, IMNM	46 (35.9)	4 (44.4)	0.72	1.42	0.27–7.0
Anti-SAE1 antibodies	4 (3.1)	0 (0)	1.00	0	0–23.6
Anti- HMGCR antibodies	1 (0.8)	0 (0)	1.00	0	0–550
Anti-Mi2 antibodies	7 (5.5)	0 (0)	1.00	0	0–11.0
Anti-MDA5 antibodies	12 (9.4)	0 (0)	1.00	0	0–5.61
Male Sex	27 (21.1)	1 (11.1)	0.69	0.47	0.01–3.77
≥ 2 'Intermediate risk' factors	39 (30.5)	2 (2.22)	0.72	0.65	0.06–3.6
Low Risk Factors					
Antisynthase syndrome	9 (7.0)	0 (0)	1.00	0	0–8.0
Anti-SRP antibodies	1 (0.8)	0 (0)	1.00	0	0–550
Anti-Jo1 antibodies	7 (5.5)	0 (0)	1.00	0	0–11.0
Non-Jo1 ASSD antibodies	1 (0.8)	0 (0)	1.00	0	0–550
MAA (PM-Scl, Ku, RNP, SSA/Ro, or SSB/La)	33 (25.8)	4 (44.4)	0.25	2.29	0.43–11.3
≥ 2 'Low risk' factors	21 (16.4)	1 (11.1)	1.00	0.64	0.01–5.2

Table 2. Paraneoplastic DM Diagnostic Criteria

Cancer Type	Number of patients N(%)	Imaging Modality for Diagnosis	Clinical Cancer Symptoms	Mean Time between DM Diagnosis and Cancer Diagnosis
Breast	4 (44.4)	Mammogram	None	23.1 months
Lung	2 (22.2)	Chest CT, Chest X-ray	Cough, dysphagia	22.9 months
Uterus/Endometrial	2 (22.2)	Abdominal CT	None	20.0 months
Cervix	1 (11.1)	N/A	Bleeding after starting blood thinner prescription	0.2 months

Abstract N°: 1129

Lupus-psoriasis overlap syndrome: a dermatological conundrum

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Introduction & Objectives:

We present 3 cases of lupus-psoriasis overlap syndrome, a known but rare entity; treatment is challenging and complex as the treatment for one can trigger the other.

Materials & Methods:

A 58-year-old male with severe psoriasis developed erythroderma following phototherapy, which was partially treated with Ciclosporin. Bloods showed positive ANA and anti-Ro60 antibody; co-existing SCLE was diagnosed. Treatment with Hydroxychloroquine caused psoriasiform erythroderma, managed with Prednisolone. Further failed therapies: Methotrexate (ineffective), Adalimumab (neutropenia), Acitretin (weakness), Bimekizumab (secondary failure). He was commenced on Risankizumab and his erythrodermic psoriasiform rash improved but he developed a lupus-type rash with anaemia, acute kidney injury, proteinuria and complement depletion. Skin biopsy showed psoriasiform features with positive IgG, IgM and C3, suggestive of lupus-psoriasis overlap syndrome. Renal biopsy showed proliferative crescentic glomerulonephritis, initially felt to be sepsis driven and he was treated with antibiotics. Unfortunately, his renal failure rapidly progressed, and the renal impression changed to immune complex driven end-stage renal failure from connective tissue disease, and he is now dialysis-dependent. Decruvacitinib has been started.

A 39-year-old female had a history of biopsy proven DLE, arthralgia, Raynaud's, previous miscarriages and nephrotic syndrome, with positive ANA, anti-cardiolipin and anti-Ro antibodies. The DLE improved with MMF and Hydroxychloroquine but four years later she developed widespread biopsy proven psoriasis with nail changes and plantar hyperkeratosis. She was treated successfully with Acitretin and Ustekinumab.

A 65-year-old female presented with a generalised SCLE rash, inflammatory arthritis, photosensitivity, sicca symptoms and lymphadenopathy. She was on Etanercept for psoriatic arthritis and had recently discontinued Methotrexate due to deteriorating renal function. Bloods showed positive ANA, anti-Ro52 and anti-Ro60 antibodies. Biopsy confirmed SCLE, Prednisolone and Hydroxychloroquine were commenced, and Etanercept was discontinued. The lupus symptoms resolved but the psoriasis recurred. She restarted Methotrexate with improvement in her skin.

Results:

Historically psoriasis and SLE were considered to have distinct immune processes: psoriasis related to activation of Th1 cells and SLE resulting from abnormal Th2 responses. The rare co-existence of lupus and psoriasis with a probability of <1.2% and an incidence of around 0.69% suggests a common immune pathway. This may be due to shared genetic risk loci (PTPN22), IL-23/Th17 axis and involvement of IL-17 cytokines in the pathogenesis of both conditions. Methotrexate and JAK inhibitors can be used effectively in both psoriasis and lupus and are useful options in the long-term management of lupus-psoriasis overlap syndrome.

Conclusion:

We have highlighted the difficulties faced when managing patients with the rare but reported lupus-psoriasis overlap syndrome through these 3 contrasting cases.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1285

Evaluating the Role of Ciclosporin as an Adjunctive Therapy in Severe Morphoea: Experience from a Tertiary Centre

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Introduction & Objectives:

Morphoea is a rare autoimmune disorder that affects the skin and underlying tissues, presenting in subtypes such as plaque, linear, generalized, and pansclerotic morphoea. Treatment options are limited, and data on the efficacy of ciclosporin in managing the condition remain sparse. This observational study aimed to assess the efficacy and tolerability of ciclosporin in adult patients with morphoea.

Materials & Methods:

In this cross-sectional study, 21 adult patients with clinically confirmed morphoea were reviewed over a 12-year period (2011–2023) from a tertiary Morphoea Service. Data collected included patient demographics, disease severity measured using the Localized Scleroderma Assessment Tool (LoSCAT), physician global assessment, immune profiles, treatment regimens, and adverse effects. Clinical outcomes were classified as improved, stable, or progressive, with treatment responses systematically evaluated at regular intervals for up to one year.

Results:

The cohort comprised 21 patients, with a female predominance (81.0%). The median age at presentation was 50 years (IQR: 31–63), with a median age of disease onset of 49 years (IQR: 21.5–60). Ciclosporin was initiated at a median dose of 1.9 mg/kg (IQR: 1.4–2.3 mg/kg). 33.3% (n=7) patients responded to treatment as early as six weeks, with 47.6% of patients demonstrating clinical improvement at one year. Ciclosporin was introduced as an adjunct to systemic therapy in patients with progressive disease despite or intolerant of standard treatments. Disease stability or improvement was achieved in 81.0% of patients following ciclosporin initiation. Adverse effects were predominantly mild, with hypertension and myalgia being the most frequently reported. Remission was documented in 52.4% of patients.

Conclusion:

Ciclosporin demonstrates efficacy as an adjunctive therapy in morphoea, facilitating rapid clinical improvement, with responses observed as early as six weeks in some patients. The treatment is generally well-tolerated, with predominantly mild adverse effects; however, most patients remained on standard systemic therapies for long-term disease management. This study provides evidence supporting ciclosporin as a viable therapeutic option for recalcitrant morphoea, particularly in cases refractory to conventional systemic treatments. Further research with larger cohorts and extended follow-up is warranted to better delineate ciclosporin's role within treatment algorithms for this complex condition.

Abstract N°: 1313

Characterizing Unique Clinical and Serologic Phenotypes of Classic and Immune Checkpoint Inhibitor Induced Bullous Pemphigoid

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Introduction & Objectives:

Bullous Pemphigoid (BP) is a subepidermal autoimmune blistering disease which most commonly occurs in elderly patients. It can also occur as an immune-related adverse event associated with immune checkpoint inhibitor (ICI) therapy. Management of BP is challenging due to disease heterogeneity and limited treatment options. We aim to improve understanding of the variety of presentations of BP and ICI-BP by characterizing unique clinical and serologic phenotypes.

Materials & Methods:

We included 446 cases of BP (BP=417, ICI-BP = 29) across the Mayo Clinic Enterprise between 1998-2024 based on diagnosis, clinical features, pathology, and serology. ICI-BP required onset of BP during or within 12 weeks following ICI treatment. Cases without cutaneous disease involvement were excluded. Unsupervised clustering analysis was conducted using hierarchical clustering on principal components (HCPC) and multiple correspondence analysis (MCA).

Results:

Compared to classic BP, patients with ICI-BP were more likely to be male (BP: 50.1% vs ICI-BP: 72.4%, p=0.020) and have a comorbid diagnosis of malignancy (BP: 29.7% vs ICI-BP: 86.2%, p<0.0001). Patients with treatment refractory BP requiring 2+ systemic medications were significantly younger (n=272, mean 73.8 years (SD 12.1), p=0.037) than those requiring fewer treatments (n=174, mean 76.3 years (SD 10.2)). Four phenotypic clusters were identified, with no significant differences in gender, age, race, ethnicity, or ICI-BP occurrence amongst clusters. Cluster 1 (C1) was the largest (N=281; ICI-BP=20) and was characterized by high levels of upper extremity (UE) (92.5%) and trunk (100%) involvement, with minimal oral involvement (3.6%). C1 had the highest percentage of BP180+/BP230+ serotype (29.5%) of all clusters. Cluster 2 (C2) (N=65; ICI-BP=3) was characterized by absence of truncal disease (0%), frequent lower extremity (LE) (87.7%) involvement, minimal oral mucosal involvement (7.7%), and lowest head/neck (H/N) (10.8%) involvement of all clusters. Cluster 3 (C3) (N=27; ICI-BP=2) was characterized by mucosal disease (100%) and anogenital (81.5%) involvement with high levels of UE (88.9%), LE (81.5%), trunk (92.6%), and H/N (48.1%) involvement. Patients in C3 had the highest percentage of BP180+/230- serotype (55.6%) and were significantly more likely to have treatment refractory disease (81.5%, p=0.040). Cluster 4 (C4) (N=73; ICI-BP=4) was the only cluster with eye involvement (12.3%). C4 was also notable for low involvement in UE (9.6%) and LE (16.4%), high H/N (42.5%) involvement, and the highest percentage of BP180-/230- serotype (52.1%).

Conclusion:

We characterized demographics, treatment response, and identified four unique phenotypes of BP based on clinical

presentation and antibody serotype. There were no significant differences in ICI-BP occurrence between clusters, suggesting that ICI-BP and classic BP cannot be differentiated on cluster phenotype alone. Patients presenting at a younger age with BP or ICI-BP and patients with the C3 phenotype may be more likely to have disease refractory to treatment, requiring multiple systemic medications. Future investigation into the cellular mechanisms within each cluster is warranted.

Figure 1: Cluster Plot

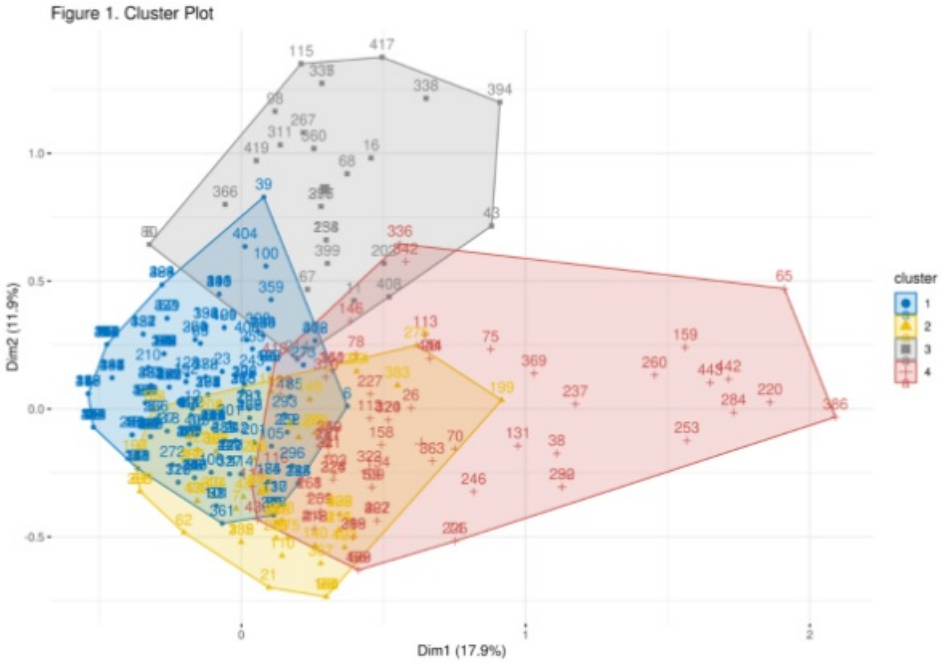

Table 1: Demographics BP vs ICI-BP

	BP (N=417)	ICI-BP (N=29)	P-Value
Age at Biopsy			0.1490[1]
Mean (SD)	74.9 (11.5)	72.4 (10.0)	
Gender, (%)			**0.0202[2]**
Female	208 (49.9%)	8 (27.6%)	
Male	209 (50.1%)	21 (72.4%)	
Race			0.9994[2]
White	391 (93.8%)	28 (96.6%)	
Ethnicity			0.9732[2]
Not Hispanic or Latino	405 (97.1%)	29 (100.0%)	
Comorbidities			
Malignancy	124 (29.7%)	25 (86.2%)	**<.0001[2]**
Hypertension	249 (59.7%)	14 (48.3%)	0.2260[2]
Diabetes	92 (22.1%)	7 (24.1%)	0.7948[2]
IIF			**0.0008[2]**
Inconsistent	173 (50.4%)	12 (48.0%)	
Consistent	47 (13.7%)	10 (40.0%)	
Definitive	123 (35.9%)	3 (12.0%)	
DIF			0.0890[2]
Inconsistent	73 (17.5%)	1 (4.2%)	
Consistent	344 (82.5%)	23 (95.8%)	
BM180			0.5535[1]
Mean (SD)	59.5 (67.7)	73.8 (80.5)	
BM230			**0.0008[1]**
Mean (SD)	29.0 (43.5)	8.9 (25.6)	

Table 2: Demographics non-refractory vs treatment refractory

	Non-Refractory treatment group* (N=174)	Refractory treatment group* (N=272)	P-Value
ICI Cohort	10 (5.7%)	19 (7.0%)	0.6049[2]
Age at Biopsy			**0.0371**[1]
Mean (SD)	76.3 (10.2)	73.8 (12.1)	
Gender, (%)			0.7367[2]
Female	86 (49.4%)	130 (47.8%)	
Male	88 (50.6%)	142 (52.2%)	
Race			0.6640[2]
White	162 (93.1%)	257 (94.5%)	
Ethnicity			0.6280[2]
Not Hispanic or Latino	171 (98.3%)	263 (96.7%)	
Comorbidities			
Malignancy	52 (29.9%)	97 (35.7%)	0.2071[2]
Hypertension	102 (58.6%)	161 (59.2%)	0.9049[2]
Diabetes	32 (18.4%)	67 (24.6%)	0.1218[2]
IIF			0.5229[2]
Inconsistent	69 (47.3%)	116 (52.3%)	
Consistent	22 (15.1%)	35 (15.8%)	
Definitive	55 (37.7%)	71 (32.0%)	
DIF			0.8219[2]
Inconsistent	28 (16.3%)	46 (17.1%)	
Consistent	144 (83.7%)	223 (82.9%)	
BM180			0.6107[1]
Mean (SD)	60.0 (66.9)	60.7 (69.7)	
BP230			0.6300[1]
Mean (SD)	27.5 (42.6)	27.9 (43.1)	

Table 3: Cluster data

	Cluster 1 (N=281)	Cluster 2 (N=65)	Cluster 3 (N=27)	Cluster 4 (N=73)	P-Value
Gender					0.621[1]
Female	141 (50.2%)	27 (41.5%)	12 (44.4%)	36 (49.3%)	
Age					0.137[2]
Mean (SD)	75.9 (10.7)	75.4 (12.9)	68.5 (17.0)	75.2 (9.9)	
Race					0.910[1]
White	262 (93.9%)	61 (93.8%)	25 (92.6%)	71 (97.3%)	
Ethnicity					0.940[1]
Not Hispanic or Latino	272 (98.6%)	64 (98.5%)	27 (100.0%)	71 (98.6%)	
Mucosal Involvement					
Mucosal (All)	10 (3.6%)	5 (7.7%)	27 (100.0%)	32 (43.8%)	**< 0.001[1]**
Eyes	0 (0.0%)	0 (0.0%)	0 (0.0%)	9 (12.3%)	**< 0.001[1]**
Mouth	10 (3.6%)	5 (7.7%)	16 (59.3%)	30 (41.1%)	**< 0.001[1]**
Anogenital	0 (0.0%)	0 (0.0%)	22 (81.5%)	1 (1.4%)	**< 0.001[1]**
Skin Involvement					
Upper Extremity	260 (92.5%)	44 (67.7%)	24 (88.9%)	7 (9.6%)	**< 0.001[1]**
Lower Extremity	245 (87.2%)	57 (87.7%)	22 (81.5%)	12 (16.4%)	**< 0.001[1]**
Head and Neck	53 (18.9%)	7 (10.8%)	13 (48.1%)	31 (42.5%)	**< 0.001[1]**
Trunk	281 (100.0%)	0 (0.0%)	25 (92.6%)	60 (82.2%)	**< 0.001[1]**
Serotype					**< 0.001[1]**
BP180-BP230-	77 (27.4%)	29 (44.6%)	7 (25.9%)	38 (52.1%)	
BP180-BP230+	40 (14.2%)	5 (7.7%)	2 (7.4%)	3 (4.1%)	
BP180+BP230-	81 (28.8%)	20 (30.8%)	15 (55.6%)	24 (32.9%)	
BP180+230+	83 (29.5%)	11 (16.9%)	3 (11.1%)	8 (11.0%)	
IIF					**< 0.001[1]**
Inconsistent	90 (38.8%)	31 (59.6%)	16 (66.7%)	48 (80.0%)	
Consistent	40 (17.2%)	9 (17.3%)	4 (16.7%)	4 (6.7%)	
Definitive	102 (44.0%)	12 (23.1%)	4 (16.7%)	8 (13.3%)	
DIF					0.328[1]

Table 4: Treatment refractory data by cluster

	Cluster 1 (N=281)	Cluster 2 (N=65)	Cluster 3 (N=27)	Cluster 4 (N=73)	P-Value
Treatment Refractory					**0.040[1]**
No	105 (37.4%)	31 (47.7%)	5 (18.5%)	33 (45.2%)	
Yes	176 (62.6%)	34 (52.3%)	22 (81.5%)	40 (54.8%)	
ICI-BP					0.869[1]
No	261 (92.9%)	62 (95.4%)	25 (92.6%)	69 (94.5%)	
Yes	20 (7.1%)	3 (4.6%)	2 (7.4%)	4 (5.5%)	

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1330

The Sclerotic Pan-Morphea: A Complex and Rare Medical Challenge (case report)

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Introduction & Objectives:

Pan-sclerotic morphea is a rare and severe form of localized cutaneous scleroderma, characterized by rapid progression. It leads to deep skin fibrosis, severe joint contractures, and ulcerations. This disease typically manifests in childhood and has an unfavorable prognosis. Currently, no effective treatment exists, though some therapies, such as vasodilators and photochemotherapy, have shown promising results. The objective is to explore the clinical features, management, and treatment outcomes of this condition.

Materials & Methods:

A 6-year-old girl, with no prior medical history, presented with progressive sclerosis of the left lower limb for one year, starting with a hypopigmented plaque. Clinical examination revealed extensive sclerotic plaques, ulcerations, and joint contractures at the left knee and ankle. A biopsy confirmed localized scleroderma, and pan-sclerotic morphea was diagnosed. The treatment involved topical corticosteroids, bolus corticosteroid therapy, methotrexate, and local wound care.

Results:

The patient exhibited classic symptoms of pan-sclerotic morphea, including widespread sclerotic lesions and joint contractures. Despite aggressive treatment, the prognosis remains poor, as current therapies have limited effectiveness. Recent advancements, such as photochemotherapy and vasodilators, have shown some positive effects.

Conclusion:

Pan-sclerotic morphea is a rare and severe disease that significantly impacts the patient's quality of life. Ongoing research is crucial to better understand the pathology and to develop more effective treatments. A multidisciplinary approach, involving dermatologists, rheumatologists, and other specialists, is essential for managing this complex condition.

Abstract N°: 1350

phenotypic change from pemphigus vulgars to pemphigus vegetans

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Introduction & Objectives:

Pemphigus vegetans (PV) is an infrequent variant of pemphigus vulgaris. We present the case of a patient who, after years of being diagnosed with pemphigus vulgaris, developed vegetative lesions.

Materials & Methods:

Clinical case.

Results:

A 49-year-old male patient had been followed in dermatology consultations for eight years due to oral erosions consistent with pemphigus vulgaris. He had been treated with azathioprine, systemic corticosteroids, and two doses of rituximab. Two months after the last administration of rituximab, he presented to the emergency department with painful cutaneous plaques of two weeks evolution, progressively growing and localized in skin folds. Physical examination revealed mamillated plaques with pustules and blisters on the surface that eroded upon friction, located in the axillae and intergluteal fold. Additionally, pustules and erosions coalescing in the pubic region, lower abdomen, and lateral costal region were observed, along with oral mucosal erosions with a cerebriform tongue appearance. Two skin biopsies were performed for histopathological analysis, revealing epidermal acanthosis with suprabasal intraepidermal blister formation, acantholytic cells, and an inflammatory component composed of eosinophils and neutrophils. Direct immunofluorescence (DIF) showed granular basal deposits of C3 and IgG, confirming the diagnosis of pemphigus vegetans. Treatment with oral corticosteroids at a dose of 0.75 mg/kg/day was initiated, following a descending taper over two months, along with topical corticosteroids and antibiotics. The patient is currently on 10 mg of prednisone with significant improvement in vegetative cutaneous lesions and, to a lesser extent, in oral lesions.

Conclusion:

PV is an autoimmune disease with two clinical variants (Neumann and Hallopeau), characterized by pustules and blisters that easily erode, forming vegetative plaques predominantly in skin folds. These variants, along with pemphigus vulgaris, form a clinical spectrum, with the Hallopeau variant being the mildest, as observed in our patient. Histopathologically, besides suprabasal acantholysis, PV also presents acanthosis with papillomatosis and epidermal hyperkeratosis. Regarding treatment, no specific protocols exist, with systemic corticosteroids, azathioprine, mycophenolate mofetil, and rituximab being the most commonly used therapies. In conclusion, our case represents a rare phenotypic shift from pemphigus vulgaris to pemphigus vegetans, despite prior treatment with rituximab.

Abstract N°: 1372

Artificial intelligence-based quantification to assess the Automatic Vitiligo Area Scoring Index

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Introduction & Objectives:

Vitiligo is a chronic skin disorder characterized by the progressive loss of pigmentation affecting 0.5% to 2% of the global population. While clinically identifiable, accurately assessing its severity remains challenging due to the subjective nature of manual scoring systems, such as the Vitiligo Area Scoring Index (VASI). VASI, though widely used, is prone to interobserver variability and time-intensive calculations, limiting its consistency and efficiency in clinical and research settings. To address these limitations, this study introduces an automated deep learning-based approach, the Automated Vitiligo Area Scoring Index (AVASI), designed to objectively quantify vitiligo severity by automating the segmentation of depigmented areas and computing VASI scores. The primary objectives were to develop a robust, reproducible, and time-efficient system for vitiligo assessment and to evaluate its performance against manual annotations.

Materials & Methods:

A retrospective, non-interventional study was conducted using a dataset of 791 images sourced from publicly available domains, capturing diverse clinical scenarios, including variations in skin tones, disease severities, imaging conditions, and anatomical regions. Each image was independently segmented by three annotators, and the ground truth masks were generated by averaging their annotations. A deep learning model was trained to automate the segmentation of depigmented areas and calculate the regional automatic VASI scores. Performance was assessed using eight-fold cross-validation, with Intersection over Union (IoU) and F1-score as evaluation metrics.

Results:

An analysis of the annotations using a one-vs-rest methodology, where one annotator served as the reference while the others contributed to the ground truth, revealed high interobserver agreement. The mean Intersection over Union (IoU) was 0.76, and the F1-score was 0.86, demonstrating the reliability of the dataset annotations. The deep learning model achieved comparable performance, with a mean IoU of 0.73 ± 0.01 and an F1-score of 0.83 ± 0.01 across all cross-validation folds, indicating minimal variability and strong alignment with manual segmentations.

Conclusion:

AVASI effectively addresses key limitations of traditional VASI scoring, including interobserver variability, subjectivity, and time inefficiency. Its strong agreement with manual annotations, coupled with its ability to streamline assessment, makes it a valuable tool for clinical practice and research. AVASI has the potential to enhance the accuracy and reproducibility of vitiligo assessments, particularly in pharmaceutical trials, where precise and consistent metrics are critical. Ultimately, it provides a scalable, cost-effective solution for improving vitiligo management and patient outcomes.

Abstract N°: 1375

Localized bullous pemphigoide in a young patient with cerebral stroke: Case report

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Introduction & Objective:

Bullous pemphigoid (BP) is an autoimmune subepidermal blistering disease, which typically presents in the elderly patients with neurologic conditions. Localized bullous pemphigoid (LBP) is an infrequent BP variant restricted to a body region. In this report, we describe a 42-year-old patient with cerebral stroke with localized bullous pemphigoide restricted to the hemiplegic side.

Materials & Methods:

Case report

Results:

A 42-year-old female patient with a history of untreated hypertension and a left –sided hemiplegia secondary to ischemic stroke four days prior, presented with blisters limited to the hemiplegic side and appearing after the cerebral vascular accident. Clinical examination revealed tense bullae with clear contents on slightly erythematous skin associated with post-bullous erosions located on the left side in the left buttock region, the left lower limb and the back. There was no mucosal involvement and Nikolsy's sign was negative. Skin biopsy showed dermal-epidermal cleavage with lymphoplasmacytic inflammatory infiltrate in the dermis. Direct immunofluorescence (DIF) showed linear IgG deposition along the dermo-epidermal junction. Indirect immunofluorescence (IFI) antibody assay showed negative anti-BP180, negative anti BP230, and positive anti-intercellular space antibodies at 40. The diagnosis of localized pemphigoide was based on clinical and histological criteria. The course of the disease was marked by spontaneous regression of the erosions after local treatment.

Discussion:

LBP is a rare variant of bullous pemphigoid, with only about 100 cases reported up to date. LBP is a variant of BP with similar clinical and immunopathologic features to generalized forms, yet with some singularities: a high prevalence of localized triggers, a better prognosis, higher chances of therapeutic success with topical corticosteroids, and a risk of generalization. A literature review showed a clear female preponderance in LBP which is consistent with our case. In fact, association between neurologic disorders and BP is well known but the exact mechanisms linking BP to neurodegenerative diseases is not entirely elucidated .It has been proposed that local neuromuscular and vascular changes in the paretic leg, together with immobility and scratching can contribute to the onset of LBP lesions.

Authors proposed diagnostic criteria for LBP which will be confirmed by fulfillment of three of the following four criteria, with the clinical criteria being mandatory :1)Clinical criteria: Presence of a bullous eruption confined to a single anatomic region, sometimes preceded by a predisposing factor, without a previous history of generalized BP;2) DIF criteria: Positive DIF with linear deposits of IgG and/or C3 along the BMZ ;3)Serological criteria: Positive IgG antibodies against the epidermal side of BMZ by IIF and/or Positive IgG antibodies reacting with BP180 and/or BP230 by ELISA, IIF, immunoblot, or immunoprecipitation;4)Histological criteria: Subepidermal blister with the presence of eosinophils.

The particularities to be underlined in our case are: The limitation of bullae to the hemiplegic side, the young age of the patient, and the spontaneously evolution without specific treatment.

Conclusion:

LBP should be considered in patients presenting local bullous eruptions, especially in patients with a history of exposure to any known trigger or associated to neurologic disorders .

Abstract N°: 1381

Coexistence of generalized morphea and vitiligo: a rare case report

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Introduction :

The simultaneous appearance of two autoimmune skin diseases is rarely observed. Generalized morphea is a rare subtype of morphea characterized by more than four lesions occurring at two or more body sites. Although not yet fully elucidated, it is becoming increasingly clear that autoimmunity plays a central role in the pathogenesis of morphea and vitiligo. Several case reports describe the association of morphea with autoimmune diseases, but coexistence with vitiligo has rarely been reported.

Observation:

A 49 year old female, with a history of vitiligo since the age of 9 with no follow-up, and no other personal or family history, presented with cutaneous sclerosis of the hands, forearms and feet 3 months prior to admission, associated with burning pain in all 4 limbs, with no Reynaud's phenomenon or other associated signs, such as respiratory or digestive, all evolving in a context of apyrexia and a good general condition.

Physical examination revealed symmetrically distributed achromic macules on the face, trunk and limbs, suggestive of vitiligo, associated with multiple lilac-pink sclerotic plaques (N>4) on the trunk and limbs, suggestive of generalized morphea. Skin biopsy confirmed the diagnosis of morphea. Immunological tests such as antinuclear antibodies, anti native DNA antibodies were negative. Thyroid tests and glycated hemoglobin were normal, as were the rest of the investigations. On the basis of clinical and laboratory findings, the diagnosis of generalized morphea associated with vitiligo was made.

The patient was put on general corticosteroid therapy and methotrexate with very good evolution, with an indication for UVB phototherapy for the management of vitiligo.

Discussion:

Morphea is a rare disease of the skin and underlying tissues, characterized by initially erythematous lesions that develop into hypo-pigmented sclerotic plaques with a lilac-pink border. Generalized morphea is an uncommon subtype.

Vitiligo is an acquired pigment disorder characterized by well-demarcated achromic spots. The relationship between vitiligo and autoimmune diseases, and the detection of organ-specific autoantibodies in vitiligo patients, suggest that vitiligo is an autoimmune pathology.

And in the literature, several cases report the involvement of autoimmunity in the development of morphea, indicating its association with various autoimmune diseases such as Hashimoto's thyroiditis.

But although morphea and vitiligo are both autoimmune diseases, the simultaneous appearance of these entities has rarely been studied. In our patient, the appearance of generalized morphea on a background of vitiligo supports the hypotheses concerning the autoimmune nature of these entities.

Conclusion:

Although we can't exclude the possibility of a simple coincidence between the development of vitiligo and morphea, our case suggests the existence of an autoimmune phenomenon in both conditions.

Further case studies would provide a better understanding of the mechanisms of these diseases, as well as the role of autoimmunity in the pathogenesis of morphea and vitiligo.

Abstract N°: 1437

Cabergoline-Induced Pemphigus Foliaceus: A Unique Case Report

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Introduction & Objectives:

Pemphigus foliaceus (PF) is an autoimmune blistering disorder characterized by the loss of adhesion between keratinocytes due to autoantibodies targeting desmoglein-1. Drug-induced PF is a recognized phenomenon, yet its association with **cabergoline**, a dopamine agonist used for hyperprolactinemia, has not been previously reported. Interestingly, **elevated prolactin levels have been implicated in autoimmune diseases, including pemphigus**, suggesting a possible immunomodulatory role. However, in this case, **the strong temporal relationship with cabergoline initiation suggests a drug-induced etiology rather than hyperprolactinemia itself**. This study presents a **novel case of cabergoline-associated PF**, aiming to highlight its potential role as a trigger and raise awareness among dermatologists regarding this possible adverse reaction.

Materials & Methods:

A **32-year-old female** with a history of hyperprolactinemia, treated with cabergoline for one year, developed **pruritic papules and small vesicles** over her back, neck, and posterior scalp, healing with **post-inflammatory hyperpigmentation (PIH)**. She had no other chronic illnesses, medications, or family history of blistering disorders. A **skin biopsy and direct immunofluorescence (DIF) confirmed PF**, showing subcorneal acantholysis, IgG and C3 deposits, with negative IgM and IgA. Given the temporal relationship between cabergoline initiation and symptom onset, **drug-induced PF was strongly suspected**

Results:

Following **dermatological and endocrinological consultation**, the patient independently **discontinued cabergoline**, leading to symptom resolution. She was managed with **topical corticosteroids** and remained **disease-free over two years** without recurrence. A comprehensive **literature search revealed no previously documented cases** of cabergoline-induced PF, emphasizing the uniqueness of this case

Conclusion:

This case **presents the first documented instance of cabergoline-associated PF**, suggesting that dopamine agonists may contribute to **autoimmune blistering diseases**. While **hyperprolactinemia itself has been linked to pemphigus**, this case supports the hypothesis that **cabergoline may act as a direct trigger for PF rather than prolactin dysregulation being the cause**. Recognizing this **potential drug association is crucial for early diagnosis and optimal management**. Future research is necessary to elucidate the underlying mechanisms and establish definitive causality. Clinicians should maintain a **high index of suspicion for drug-induced PF** in patients presenting with blistering dermatoses while on cabergoline therapy.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1441

Familial bullous lichen planus: the first cases from outside Asia

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Introduction & Objectives: ## Lichen planus (LP) is an idiopathic inflammatory disease affecting the skin, hair, nails, and mucous membranes. Bullous lichen planus (BLP) is a rare LP variant characterized by blisters and bullae that usually develop on pre-existing LP lesions. BLP is usually sporadic; familial BLP (FBLP) cases are highly scarce and mainly reported from China. We present the first cases, mother and son, of FBLP from outside Asia and discuss the outcomes of methotrexate treatment.

Materials & Methods:

A 41-year-old woman was presented with violaceous plaques on the distal parts of both lower legs, accompanied by numerous milia and vesicles on the surface of the plaques. Violaceous papules and some bullae were also observed on the right elbow and forearm. She had no mucosal involvement. The patient has had cutaneous eruptions since her twenties. She stated that her sister, daughter, and son had similar lesions. The patient's son, age 18, accepted the examination, and pink-purple papules with bullae on his hands, fingers, knees, and elbows were observed.

Results:

Histopathological examination of both cases showed orthokeratosis, hypergranulosis, focal dermo-epidermal separation, and perivascular interstitial dermatitis in the upper dermis. The immunofluorescence examination of the skin samples did not show autoantibody deposits. Circulating antibodies against desmoglein 1, desmoglein 3, BP180, BP230, envoplakin, periplakin, and type 7 collagen were all negative. Depending on clinical, pathological, and laboratory findings and the presence of familial cases, the patients were diagnosed with FBLP. The index case accepted systemic therapy and she was put on weekly 20 mg methotrexate and daily topical clobetasol propionate ointment. After about 3-month of treatment, the skin lesions remarkably regressed.

Conclusion:

Familial BLP should be considered in the differential diagnosis of bullous eruptions over lichenoid papules and plaques in patients with a family history. Methotrexate seems to be a promising treatment for FBLP.

Abstract N°: 1442

Rare clinical presentations of immunobullous diseases

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Rare clinical presentations of immunobullous diseases

Introduction & Objectives:

Immunobullous diseases are autoimmune blistering conditions like pemphigus vulgaris, pemphigus foliaceous and bullous pemphigoid. Even though they commonly present with blisters, there are rare occasions that they may present with other clinical manifestations initially.

Materials & Methods:

case1

75 year old male patient with ischemic heart disease admitted with intensely itchy erythematous papules and plaques over the trunk, upper limbs and lower limbs for three months. The rash showed sparing of skin folds over the abdomen and flexures producing the deck chair sign. He didn't have fever, loss of appetite or loss of weight. While being investigated, he gradually developed tense blisters over trunk and extremities.

Routine investigations were normal except high eosinophil count in full blood count. Initial biopsy over erythematous plaque showed acanthosis and mild perivascular inflammation. The second biopsy from a blister showed sub epidermal blister filled with eosinophils. Immunofluorescence study was negative for IgA, IgG and C3. Clinically he was diagnosed as bullous pemphigoid and he resolved completely following treatment with steroid pulses and doxycycline.

case2

57 year old previously healthy male, presented with generalized erythema and scaling for 2 weeks. He didn't have mucosal involvement. On examination there was generalized erythema and scaling and it was more prominent over upper chest and arms. There were erythematous thin annular plaques with concentric ring like appearance over arms and chest mimicking characteristic erythema gyratum repens appearance. Clinically there were no intact blisters. On histophathological examination a blister was present below the corneal layer and immunofluorescence showed intercellular deposition of IgG and C3 in epidermis. The definitive diagnosis of pemphigus foliaceus was confirmed. Patient was treated with steroids and complete resolution of lesions occurred.

Results:

Both pemphigus foliaceus and bullous pemphigoid commonly present with flaccid and tense blisters respectively. Deck chair sign indicates the sparing of major skin folds and it characteristically occurs in papuloerythroderma of ofuji. However it is also reported in acute contact dermatitis, angioimmunoblastic lymphoma,leprosy and bullous pemphigoid. Our first patient had encountered this rare presentation during the course of bullous pemphigiod as he developed tense blisters later and biopsy confirmed the diagnosis of bullous pemphigoid. This is an unusual finding in bullous pemphigoid.

Erythema gyratum repens is a figurate disorder with wood-grain appearance, typically associated with malignancy and rarely occuring with mycobacterial infections, connective tissue diseases and pregnancy. Our second patient had presented with erythema gyratum repens like appearance in pemphigus foliaceus which was confirmed with biopsy and immunofluorescence.

Conclusion:

The above cases show case the rare clinical presentations of common autoimmune blistering disorders. This highlights the importance of knowing the rare presentations, continuous follow up of the patients who may develop typical lesions during the progression the disease and histopathological correlation of the cutaneous lesions in diagnosing blistering disorders.

Abstract N°: 1453

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 is most common in adults between the third and sixth decade of life with no gender predominance, but it affects females more commonly. The classical lichen planus lesion 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 the upper limbs, lower limbs, abdomen, and back. Dermoscopic examination revealed a red-brown background, a starburst pattern of Wickham's striae, and brown dots. Histopathology of the lesion showed a moderately dense lichenoid infiltrate of lymphocytes at the dermo-epidermal junction. The epidermis showed saw-tooth acanthosis, wedge-shaped hypergranulosis, and orthohyperkeratosis. Based on the clinical, dermoscopy, and histopathological findings, a diagnosis of lichen planus with corymbiform lesions was made. After routine investigations, the patient was started on oral prednisolone (1mg/kg/day) daily for three weeks. The patient was shifted to tablet methotrexate 7.5 mg weekly with regular monitoring. The patient was followed up at two weekly intervals and she noticed improvement in her skin lesions

Conclusion:

Lichen planus is an autoimmune disorder affecting the skin, mucosa and nails. There are several clinical variants of LP such as hypertrophic, eruptive, linear, annular, atrophic, actinic, nail, oral, vulvovaginal, inverse, lichen planus pemphigoides, bullous, lichen planus- lupus erythematosus overlap syndrome. In Botany, the word corymb refers to racemose inflorescence in which flowers are located at the same level due to the gradual shortening of the length of the pedicels along the axis. Corymbiform arrangement of skin lesions in dermatology is characterized by the presence of a central larger papule which is surrounded by smaller satellite papules. Corymbose lesions have been described in the literature in various dermatological disorders such as secondary syphilis, Lichen sclerosus et atrophicus, nodular amyloidosis, and sarcoidosis. In syphilis, corymbose lesions are associated with recurrences, and relapses and have evidence of increased severity. In Lichen planus, papules arranged in a corymbiform pattern are not yet reported in the literature. In our patient, the lesions had a 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.

EA DV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1533

Linear Pigmented Discoid Lupus Erythematosus On The Scalp Of A 52-Year-Old, Male, Filipino Treated With Pico Laser: A Case Report

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Introduction & Objectives:

Lupus erythematosus (LE) is a multisystem disorder affecting the skin through complex interactions of immunologic, genetic, and environmental factors. Linear cutaneous lupus erythematosus (LCLE) is a rare variant characterized by linear erythematous plaques along Blaschko lines.

Due to its limited data, further researches are needed to understand its pathogenesis, clinical features, and management.

Materials & Methods:

A 52-year-old Filipino male, presented with a 3-month history of a solitary, hyperpigmented linear plaque on the left frontal area extending to the forehead. The lesion had progressively increased in size and pigmentation, developing fine, whitish adherent scales. Examination revealed a well-demarcated, linear violaceous-to-hyperpigmented plaque measuring 8x2 cm on the left frontal area, along with a smaller plaque (3x2 cm) with hair loss on the left parietal area. A biopsy confirmed the diagnosis of linear pigmented discoid lupus erythematosus (DLE). Laboratory tests, including CRP and ANA, were negative.

Results:

Patient was treated with super high potency topical corticosteroid twice a day for two weeks then once a day for the next two weeks. Medication was shifted to topical calcineurin inhibitor, applied twice a day to lesions for 20 weeks showing further decrease in the thickness, hyperpigmentation, scaling of plaques. Dermoscopy showed disappearance of the follicular plugs, perifollicular white halos, rosettes, yellow dots, and telangiectatic vessels from the baseline.

Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) decreased from six to two.

Pico laser treatment was administered using an Nd:YAG picosecond laser (1,064 nm, 3-mm spot size, fluence 0.8 J/cm^2, one pass, with whitish gray spot as the endpoint). Follow-up every four weeks showed reduced hyperpigmentation after two sessions.

Discussion and Conclusion:

From 1978 to 2022, 72 cases of linear DLE were reported in 45 global journals. The patients had an average age of 24.47 years, and there was nearly an equal distribution between genders, with 35 males and 37 females. Between 1978 and 2022, 72 cases of linear DLE were reported globally, with an average patient age of 24.47 years and nearly equal gender distribution. The most commonly affected sites included the face (72%), upper extremities (21%), and neck (19%) with an average duration from onset to presentation was approximately 4.5 years. The initial diagnoses included linear cutaneous lupus erythematosus (24%), lichen planus (18%), and discoid lupus erythematosus (12%), morphea (7%), and lichen striatus (6%). Among the patients, 22% had positive ANA tests, and 4.6% experienced photosensitivity symptoms. Treatment strategies included corticosteroids, calcineurin inhibitors, hydroxychloroquine, and photoprotection, although post-hyperpigmentation was common.

This is the first reported case of linear pigmented DLE on the scalp in the Philippines and first linear DLE to use pico laser

as treatment, based on previous literature.

This case emphasizes considering CLE as a potential diagnosis in patients with linear hyperpigmented plaques and sun exposure history, even without systemic symptoms. Treatment included a super high potency topical corticosteroid, topical calcineurin inhibito, and strict photoprotection led to decreased thickness, hyperpigmentation, scaling, and CLASI. Pico laser treatment was administered resulting decreased hyperpigmentation.

Abstract N°: 1543

blisters and sight : unveiling the ocular challenges of pemphigoid

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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°: 1548

bullous pemphigoid: insights from a case series on diagnosis and management

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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.

Abstract N°: 1561

Parry-Romberg-syndrome Associated with En Coup de Sabre: Case Report

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Introduction & Objectives: Parry-Romberg syndrome (PRS), also known as progressive facial hemiatrophy, is a rare neurocutaneous disorder characterized by the progressive, unilateral atrophy of the facial soft and hard tissues. Typically present in childhood or early adulthood, late-onset cases are becoming more recognized. PRS can lead to significant aesthetic and functional issues, including facial asymmetry, neuralgia, migraines, and ocular involvement. Although its exact cause remains unknown, the disorder is thought to have a multifactorial etiology involving genetic, autoimmune, vascular, and neurogenic factors. PRS is frequently associated with localized scleroderma, particularly en coup de sabre (ECDS), a linear form of scleroderma. This case report discusses an adult-onset PRS case, initially diagnosed as localized scleroderma, which progressed into PRS over several decades. The objective is to emphasize the importance of diagnosing PRS in adults and to explore treatment options, particularly immunosuppressive therapies.

Materials & Methods: We present the case of a 52-year-old female diagnosed with adult-onset PRS associated with ECDS.

Results: The patient, a 52-year-old female, experienced progressive facial atrophy initially diagnosed as localized scleroderma in 1999. She had no history of trauma or preceding illnesses. Over the next two decades, she developed gradual atrophic changes on the right side of her face, beginning with the chin and progressing to the cheek. By 2023, significant facial asymmetry prompted a re-evaluation of her diagnosis. Physical examination revealed prominent right hemifacial atrophy, with a distinctive “sword stroke” mark on her forehead, consistent with ECDS. Additional findings included alopecia, hypopigmented atrophic macules, and sunken periorbital fat. Laboratory tests for antinuclear antibodies, Scl-70, and SSA/SSB antibodies were all negative, ruling out systemic involvement. Two skin biopsies confirmed the diagnosis of localized scleroderma. Imaging studies, including CT scans, showed no significant brain changes, although air inclusions were found in the right eye socket. Ophthalmic examination revealed a 4 mm difference in eye socket size and enophthalmos of the right eye. Based on patient history, clinical examination, histopathological results, and imaging studies, a diagnosis of PRS with associated linear scleroderma (en coup de sabre) was confirmed. Systemic treatment with methotrexate 10 mg/ weekly was initiated to manage disease progression, with regular follow-ups scheduled for monitoring.

Conclusion: This case highlights an uncommon form of adult-onset PRS, initially presenting as localized scleroderma and later progressing to PRS. It underscores the need for recognizing PRS in adults, particularly when it overlaps with localized scleroderma-like en coup de sabre. Early diagnosis and intervention are crucial to prevent further progression, with immunosuppressive therapies such as methotrexate playing a key role in halting the disease. A multidisciplinary approach is essential for managing both the aesthetic and functional aspects of PRS and monitoring for potential neurologic and other complications.

Abstract N°: 1652

Beyond the Blisters: The Untold Challenges of Diagnosing and Treating Pemphigus Vulgaris: A Case Report

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Introduction & Objectives:

Chronic, non-healing oral lesions are frequently misdiagnosed as infectious or inflammatory conditions, leading to delayed and often inappropriate treatment. Pemphigus vulgaris (PV), a rare autoimmune blistering disorder, commonly presents with mucosal involvement, complicating early recognition. Diagnostic challenges increase when PV exhibits paraneoplastic features. This case explores the complexities in diagnosing and managing PV with suspected paraneoplastic involvement, emphasizing the importance of personalized treatment strategies.

Materials & Methods:

A 65-year-old male with diabetes and hypertension developed chronic oral erosions covered by white-yellowish pseudomembranes on the soft palate, jugal and labial mucosa, and tongue. Following multiple antibiotic courses prescribed by infectious disease and ENT specialists, the lesions had an oscilating evolution. Histopathology and ELISA testing of anti-desmoglein 3 antibodies confirmed PV, while direct immunofluorescence findings suggested paraneoplastic pemphigus (PNP), raising concerns about an occult malignancy, though imaging results were negative. At first, the patient refused systemic corticosteroid therapy, determined to avoid it because of potential side effects. Initial treatment with mycophenolate mofetil (MMF) at 2 g/day resulted in worsening of the lesions, which led to its discontinuation, and rituximab was introduced while receiving a low dose of prednisone (0.5 mg/kg/day).

Results:

Although malignancy screening was negative, the presence of PNP-like features complicated disease management. The paradoxical worsening of lesions under MMF suggested either an inadequate response, or potential disease flare. Between the 2 doses of rituximab, the patient showed significant clinical improvement, highlighting its efficacy in refractory PV.

Conclusion:

This case underscores the importance of early recognition of autoimmune mucosal diseases to prevent diagnostic delays and unnecessary treatments. It illustrates the diagnostic complexity of PV with paraneoplastic features and the challenges associated with immunosuppressive therapy selection, particularly when systemic corticosteroids are avoided. Further research is needed to understand paradoxical responses to immunosuppressants like MMF in PV and to explore the role of biologics as viable alternatives.

Abstract N°: 1659

Migration of Hyaluronic Acid Dermal Fillers During Pregnancy: A Case Study

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Introduction & Objectives:

Background

Hyaluronic acid (HA) is a widely used dermal filler. Dermal fillers, especially those based on HA, have gained popularity as non-invasive solutions for restoring facial volume and minimizing wrinkles. Their safety profile, reversibility, and longevity make them preferred options. However, HA fillers are not without complications. Here, we present a case study of a pregnant woman presenting with chin asymmetry due to HA filler migration 11 years post-injection.

Materials & Methods:

A 36-year-old pregnant woman (24 HBD) presented with asymmetry and swelling in the chin region. The patient reported undergoing HA filler injection for lip augmentation 11 years earlier. Symptoms began four weeks prior to presentation and progressively worsened. Ultrasound imaging revealed hyperechoic structures in the soft tissues of the chin, consistent with migrated filler material. The absence of signs of inflammation or infection suggested a non-infectious aetiology.

Results:

Pregnancy-specific factors, such as hormonal changes and tissue remodelling, were hypothesized to have reactivated and displaced encapsulated HA filler deposits. Management included conservative monitoring, given the patient's asymptomatic status, and follow-up imaging.

Conclusion:

HA fillers can persist beyond the marketed 6–12 months. Pregnancy introduces physiological changes which may destabilize previously injected fillers. Ultrasound is useful to visualize hyaluronic acid fillers as hypoechoic or anechoic structures within the dermis or subcutaneous tissues. This case emphasizes the need for careful consideration in pre-procedure counselling. Patients should understand the potential for long-term complications, especially if planning pregnancy.

Abstract N°: 1686

Reticular Erythematous Mucinosis: A Variant of Systemic Lupus Erythematosus or a Distinct Entity? A Case Report

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Introduction & Objectives: Reticular erythematous mucinosis (REM) is a rare dermatologic condition characterized by reticulated erythematous patches, commonly associated with mucin deposition in the dermis. Its relationship with systemic lupus erythematosus (SLE) remains uncertain, with some studies suggesting it may represent a subtype of lupus erythematosus. The objective of this study is to investigate the potential link between REM and SLE by presenting a clinical case with concurrent positive autoantibodies, low complement levels, and lymphocytic infiltration of the skin, examining whether REM could be a manifestation of systemic lupus erythematosus or a distinct dermatologic entity.

Materials & Methods: A 45-year-old male patient with a history of skin changes was evaluated for suspected REM. Data were collected from the patient's clinical history, neck, abdominal, axillary and inguinal ultrasound to identify lymph node abnormalities, a skin biopsy for histopathological examination and immunohistochemistry, laboratory blood tests including ANA screen, complement levels (C3 and C4), and D-dimer.

Results: The patient presented with reticular erythematous lesions localized to the chest and upper back, with islands of normal skin. Urticarial lesions were also observed on the neck. The skin biopsy showed perivascular and periadnexal lymphocytic infiltrates along with increased mucin deposition in the dermis, consistent with REM. Immunohistochemistry also showed features suggestive of lupus erythematosus: Immunohistochemical analysis revealed myeloperoxidase expression in the infiltrating cells, with CD3 and CD20 positive for T and B lymphocytes, respectively. Glycophorin A and CH61 were negative, while CD34 highlighted microvascular structures. Additionally, CD117 staining identified a limited number of mast cells within the dermis. Laboratory results revealed a positive ANA screen (4.9), low complement levels (C3 = 0.31g/L and C4 = 0.07g/L), and an elevated D-dimer (6.96ug/ml), suggesting an autoimmune process. Additionally, the neck ultrasound showed reactive lymph nodes, which raised the suspicion of systemic involvement. These findings support the hypothesis that the patient may have an underlying autoimmune condition, possibly systemic lupus erythematosus (SLE).

Conclusion: This case highlights the overlapping features between reticular erythematous mucinosis (REM) and systemic lupus erythematosus (SLE). Literature includes cases that aim to clarify the relationship between REM and SLE, with some indicating a potential connection. Our case further supports this hypothesis, but additional studies are needed to determine whether REM is a variant of SLE or a distinct entity.

Abstract N°: 1711

Impact of the Coronovirus area on the incidence of autoimmune diseases

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Introduction & Objectives:

The hypothesis of the involvement of environmental factors in the pathogenesis of autoimmune diseases is well established; various infections and vaccines result in hyper-stimulation of the immune system. Similarly, SARS-CoV-2 may give rise to similar manifestations, with numerous papers demonstrating that patients with COVID-19 are likely to develop several types of autoantibodies and autoimmune diseases.The aim of our work is to compare the incidence of different autoimmune diseases before and after the SARS-COV-2 area in Morocco.

Materials & Methods:

This is a retroprospective descriptive study over a period of 8 year. We included all adult patients with Pemphigus , Bullous Pemphigoide and Dermatomyositis . We calculated the annual hospital incidence of these different pathologies in order to investigate a possible peak in incidence after the SARS-COV2 pandemic.

Results:

We collected 27 cases of DM . The average annual incidence was estimated at 3.4 new cases per year, with extremes ranging from 0 to 7. We noted a peak in incidence during the year 2020, coinciding with the onset of the corona virus (COVID-19) pandemic in Morocco. The total number of cases recorded during this year was 7, corresponding to the highest incidence described during our entire study period extending from 2015 to 2022. We included 35 cases of bullous. The average annual incidence of BP was 4.3 new cases per year, and we noted an increase in the incidence of bullous pemphigoid from the year 2020 onwards, where the number of new cases varied between 7 and 11 per year, compared with the area before the corona virus pandemic, where the number of new cases varied between 1 and 3 per year. We also included 63 cases of pemphigus .The mean annual incidence was 8 new cases per year, with extremes ranging from 3 to 10 new cases. We noted a peak in incidence during the year 2020, when the number of new cases was estimated at 8, corresponding to the highest incidence during our study period.

Autoimmunity can be secondary to a variety of factors leading to a state of hyper-stimulation of the immune system. It has long been established that viruses are an important component of the environmental factors that contribute to the production of autoimmune antibodies. SARS–CoV-2 is responsible for hyper-stimulation of the immune system, with increased concentrations of pro-inflammatory cytokines. In addition, a molecular mimicry between SARS-CoV-2 and humans has been established, contributing to the production of autoantibodies.

Conclusion:

We noted an increase in the incidence of various autoimmune diseases after the coronavirus area in Morocco. The role of SARS-COV2 and its vaccination is strongly suspected. Further studies in vaccinated patients or infected patients with SARS-COV2 are needed to corroborate our results.

Abstract N°: 1744

Trauma-Induced Bullous Pemphigoid Following Coronary Bypass Surgery in a Gliptin-Treated Patient: A Case Report

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Introduction & Objectives: Bullous pemphigoid (BP) is the most common subepidermal autoimmune blistering disorder which can be triggered by various factors, including trauma and certain medications such as gliptins. This case report presents a rare instance of trauma-induced BP in a patient on gliptin therapy following coronary artery bypass graft surgery.

Materials & Methods: A 63-year-old male patient with a history of diabetes, hypertension and coronary artery disease presented with pruritic, intact and ruptured bullae along the saphenous vein donor site and a solitary vesicle on the sternal surgical scar for about two months. The patient had undergone coronary artery bypass graft surgery with saphenous vein harvesting from the left lower limb one month prior the eruption. The mucous membranes were unaffected. The patient has been using vildagliptin and metformin for approximately 5 years for the treatment of diabetes mellitus.

Results: Histopathology of biopsy revealed dermo-epidermal separation and inflammatory cell infiltration including eosinophilic leukocytes in the upper dermis. Direct immunofluorescence examination revealed linear deposition of C3 (+) and IgG (+) in the basal layer of the epidermis. Genetic predisposition along with gliptin usage were considered as predisposing factors. Trauma causing basal membrane damage was speculated as the last hit to the presentation of BP clinic. The patient was put on daily application of high-potency topical corticosteroid. Regarding the potential association between gliptins and BP, the patient's gliptin therapy was switched to an alternative antidiabetic medication as a precautionary measure. After a month of treatment, the patient was totally lesion free. No recurrence was observed in the subsequent follow up visits.

Conclusion: Bullous pemphigoid should be considered in the differential diagnosis of blistering disorders along surgical scars. One may speculate that gliptin usage may facilitate trauma induced BP in genetically predisposed patients.

Abstract N°: 1748

Regulatory γδ T cells Protect Human Scalp Hair Follicles from Alopecia Areata In Vivo and Represent Potential Therapeutic Target

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Introduction & Objectives:

Recently, we showed that IFN-γ-secreting, NKG2D+/Vδ1+ γδT cells can induce alopecia areata (AA). However, the role of immunosuppressive Foxp3+ γδTregs in AA remains unexplored.

To clarify whetherFoxp3+ γδTregs can prevent or treat human AA.

Materials & Methods:

Autologous Foxp3+γδTregs were generated by pre-stimulating PBMCs with IL-2, TGF-β1, IL-15, and zoledronate. Recognized γδTreg markers and secretory activities were confirmed by FACSAria analysis amd ELISA (see below) . These γδTregs were either co-cultured with stressed human scalp hair follicles (HFs) *ex vivo* or were injected intradermally into experimentally induced AA lesions in human skin xenotransplants on SCID/beige mice *in vivo*.

Results:

The number of Foxp3+ γδTregs was increased around lesional AA HFs. When these γδTregs (CD3+, TCRVδ2+, FOXP3+, ICOS+, CTLA4+; TGFb1- and IL-10-secreting) were co-cultured with stressed, MICA/B-overexpressing human scalp HFs *ex vivo* in the presence of AA-pathogenic NKG2D+/CD8+ T cells, γδTregs mitigated CD8+T cell-induced HF immune privilege collapse and hair growth inhibition *ex vivo* by secreting IL-10 and TGF-β1. *In vivo*, intradermal injection of peripheral blood-derived human autologous γδTregs significantly reduced the development of experimentally induced AA lesions in human scalp skin xenotransplants on SCID/beige mice, reduced lymphocytic HF infiltration, and restored HF immune privilege.

Conclusion:

We provide the first evidence that γδTregs are both preventive and therapeutic in human AAand thus as as potent autoimmunity-protective immune cells. This invites the use of autologous γδTregs as novel cell-based therapeutics in future AA management.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1779

Botulinum Toxin Therapy in Raynaud's Phenomenon: A Systematic Review

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Introduction & Objectives:

Raynaud's Phenomenon (RP) is characterized by episodic vasospasms** in the peripheral arteries, affecting the** fingers and toes**.** Botulinum toxin therapy has emerged as a promising treatment due to its ability to inhibit neurotransmitter release and modulate vascular tone**.** This review evaluates the effectiveness and safety of botulinum toxin in RP management by synthesizing current evidence. By altering vascular reactivity and improving blood flow, botulinum toxin holds potential for addressing underlying vascular dysfunction and developing targeted treatment approaches for RP.

Materials & Methods:

A thorough search of electronic databases including PubMed, Embase, and Scopus was conducted using relevant keywords such as "Botulinum Toxin", "Raynaud's Phenomenon". The search identified a total of 50 studies that met the inclusion criteria. We followed the Preferred Reporting Items for Systematic Reviews and Meta-Analysis Statements (PRISMA) for writing this review. These studies evaluated the use of Botulinum Toxin therapy as a therapeutic intervention for Raynaud's Phenomenon.

Results:

The studies included in this systematic review demonstrated that Botulinum Toxin therapy appears to be a promising treatment option for Raynaud's Phenomenon. Several studies reported significant improvements in symptoms such as frequency and duration of vasospastic attacks, pain severity, and overall quality of life. One study found that Botulinum Toxin injections led to a reduction in Raynaud's attacks and improved vascular perfusion in the affected areas.

Conclusion:

Botulinum toxin therapy shows promise as a treatment for Raynaud's phenomenon** by modulating** neurotransmitter release and vascular tone**,** potentially improving** blood flow and symptom relief**.** However,** further clinical studies** are needed to establish its** efficacy, safety, optimal dosing, and injection sites**.** Continued research is essential to define** long-term outcomes** and refine its** therapeutic role in RP management**.**

Abstract N°: 1790

'Inverse Gottron's Sign In A Patient With Dermatomyositis And Rapidly Progressing Interstitial Lung Disease - A Case Report'

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Inverse Gottron's Sign In A Patient With Dermatomyositis And Rapidly Progressing Interstitial Lung Disease - A Case Report

Introduction:

Dermatomyositis (DM) is a rare autoimmune disorder characterized by inflammation of skin and muscles,leading to symptoms such as muscle weakness,skin rashes and joint pain.A notable feature of DM is Gottron's papules which are lichenoid papules typically found over joints of dorsal hand.Inverse Gottron's papules appear on palms are less common and can be particularly significant as they may indicate underlying interstitial lung disease.

Case Description: We present a case of 52-year-old female with history of hypothyroidism who experienced multiple red violaceous skin lesions on her face,hands and feet over 9 months.She reported swelling around her eyes,difficulty rising from a squatting position for 3 months and breathlessness on exertion for past 20 days.She noticed red purple discoloration around her eyes which gradually spread to malar areas,hands and feet.Accompanied by mild itching and burning,exacerbated by sun exposure.She also experienced painless swelling around her eyes,weakness in standing from a squatting position and joint pain in her ankles/knees.History of photosensitivity,recurrent oral ulcers,intermittent non-productive cough.On examination pallor noted.Vitals stable,respiratory examination revealed bilateral fine inspiratory crackles.Proximal muscle weakness was observed in both hip and shoulder joints(power rated 3/5)

Dermatological examination revealed a heliotrope rash with swelling and telangiectasias around eyes along with multiple dusky erythematous and violaceous papules on palmar aspect of the distal and proximal interphalangeal and metacarpophalangeal joints,indicative of inverse Gottron's sign.Mechanic's hands.Healed atrophic digital vasculitic scars.Musculoskeletal examination showed swollen and tender joints.

NFC: dilated tortuous capillaries with capillary dropout and ragged cuticles. Routine investigations indicated elevated muscle-specific enzymes,deranged hepatic enzymes, elevated serum ferritin, and anti-TPO levels. Immunological workup showed positive ANA and a strongly positive anti-Ro 52. Histopathological evaluation of skin lesions consistent with dermatomyositis.Muscle biopsy revealed inflammatory myositis.Pulmonary function tests indicated a restrictive pattern consistent with interstitial lung disease.HRCT chest imaging showed features suggestive of interstitial lung disease,MRI thighs inflammatory myositis.Malignancy screen-negative.

A clinicopathological diagnosis of dermatomyositis was established,she was initiated on systemic corticosteroids,methotrexate,hydroxychloroquine along with topical emollients and photoprotection.The patient showed symptomatic improvement.Two months post-discharge,she returned with worsening breathlessness,proximal muscle weakness,hoarseness of voice,dysphagia and generalized fatigue.She was treated with intravenous methylprednisolone pulse therapy, followed by intravenous immunoglobulin and rituximab,to which she responded positively.

Conclusion:

This case highlights a rare presentation of dermatomyositis with inverse Gottron's sign and rapidly progressing interstitial lung disease.The recognition of such atypical manifestations is crucial for timely diagnosis and management of this complex autoimmune condition.

Abstract N°: 1797

Pretibial Myxedema as the First Indicator of Hidden Graves' Disease

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Introduction & Objectives:

Pretibial myxedema (PTM) is a dermatological manifestation of different autoimmune disorders, particularly in autoimmune thyroiditis, most commonly Graves' disease, due to intradermal deposition of mucin.Graves' disease is caused by autoantibodies to the thyroid-stimulating hormone receptor (TSHR) that act as agonists and induce excessive thyroid hormone secretion, releasing the thyroid gland from pituitary control. Antibodies to TSHR also underlie Graves' orbitopathy (GO) and pretibial myxedema. The anterior tibia is most commonly affected but lesions may be presented also on thighs, feet and upper extremities.

Materials & Methods:

A 64-year-old woman presented with nearly 10-year-old swelling of the legs, characterized by nodular and verrucous lesions on both tibias. In 2014, the lesions were initially diagnosed as lymphoedema, and excision followed by skin transplantation was performed. However, recurrence occurred six months post-surgery. The patient underwent a total thyroidectomy in 2012 due to nodular goiter associated with hyperthyroidism and was subsequently treated with levothyroxine.Further clinical examination also revealed exophthalmos.

Results:

A biopsy performed in the Dermatology Department revealed focal hyperkeratotic, parakeratotic and slightly acanthotic epidermis and edematous dermis with abundant mucinous infiltrates. A diagnosis of pretibial myxedema was suspected. Laboratory tests showed the following results: TSH 0.33 ng/mL (reference range 0.27–4.2), T3 2.27 ng/mL (reference range 2–4.4), and T4 1.69 ng/mL (reference range 0.93–1.7). Anti-TPO antibodies were <0.21, while TSH receptor antibodies (TRAb) were markedly elevated at 412.10 IU/L. MRI of the ankles further confirmed the diagnosis. Following the endocrinologist's recommendation, the diagnosis of a thyroid disorder was established. Treatment with Lanreotide was initiated at a dosage of 120 mg via subcutaneous injections every 4 weeks.

Conclusion:

We presented a case of pretibial myxedema in a patient with Graves' disease, initially misdiagnosed as lymphoedema. This case underscores the critical importance of accurately diagnosing dermatological conditions to uncover underlying systemic diseases. Comprehensive diagnostic testing is essential in identifying the systemic causes of cutaneous manifestations in order to start effective therapy to increase a life quality of the patient.

Abstract N°: 1798

Interstitial Granulomatous Dermatitis as a Rare Paraneoplastic Manifestation of a PET-Avid Lung Nodule: A Multidisciplinary Diagnostic Challenge

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Introduction & Objectives:

Interstitial Granulomatous Dermatitis (IGD) is a rare inflammatory condition typically linked to autoimmune diseases or drug reactions. Its association with malignancy remains poorly understood, with limited documented cases in literature . This case highlights IGD as a potential paraneoplastic manifestation of an undiagnosed lung nodule, emphasising the role of dermatological findings in systemic disease recognition.

Materials & Methods:

A 69-year-old male presented with a two-year history of progressive erythematous maculopapular eruptions on the trunk and extremities, associated with fatigue, myalgia, and weight loss. Initial histopathology suggested granuloma annulare, but the persistent and refractory nature of the rash prompted further investigation. A deep skin biopsy revealed mid-dermal non-caseating granulomas with multinucleate giant cells, confirming IGD.

Systemic evaluation identified hypercalcaemia, elevated inflammatory markers, autoimmune and vasculitis screen were negative. PET scan revealed an avid cavitating lung nodule. A CT-guided biopsy was inconclusive due to insufficient tissue. Given the clinical and radiological findings, IGD was suspected to be part of a paraneoplastic process. A multidisciplinary team recommended surgical wedge resection for definitive diagnosis.

Results:

Despite symptomatic management with topical corticosteroids and emollients, the skin lesions persisted, further supporting an underlying systemic driver. The presence of hypercalcaemia and inflammatory markers suggested a malignancy-associated inflammatory response. The diagnostic complexity of IGD was compounded by histopathological overlap with other granulomatous dermatoses, necessitating a high index of suspicion for malignancy.

Conclusion:

This case underscores IGD as a potential paraneoplastic phenomenon requiring comprehensive systemic evaluation. Recognition of persistent granulomatous dermatitis in conjunction with systemic symptoms is crucial for early malignancy detection. A multidisciplinary approach remains key to optimising patient outcomes. This case further highlights the need for continued research into the pathophysiological mechanisms linking IGD with malignancy, as a comprehensive understanding will facilitate early diagnosis and enhance patient prognosis.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1909

Development of generalized morphea after COVID-19 vaccination

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Localized scleroderma (LS) is part of the broader group of sclerosing dermatoses. The most common form of LS is morphea, with an incidence ranging from 0.4 to 2.7 cases per 100.000 inhabitants per year. LS predominantly affects females, usually between 40 and 50 years. The pathogenesis of LS is still unclear, although several factors have been reported as possible triggers, including genetic predisposition, repeated microtrauma, and environmental factors. One variant of LS is generalized morphea (GM), defined by the presence of at least four sclerotic plaques larger than 3 cm in diameter, affecting at least two of seven anatomical regions, namely head-neck district, chest and back, and the four limbs. We describe a 58-year-old Caucasian male with multiple sharp, hard, whitish plaques on the abdomen, back, upper and lower limbs. The patient denied any previous systemic or dermatological diseases, drugs intake or tick bites. He had developed the plaques after the administration of the second dose of the Vaxzevria® (AstraZeneca) COVID-19 vaccine. Histological examination of a plaque showed widespread proliferation of dermal connective tissue, with homogenized and enlarged collagen fiber bundles oriented parallel to the skin surface. Based on the clinical and histopathological findings, the diagnosis of GM induced by COVID-19 vaccination was made. The patient was treated with methotrexate (12.5 mg weekly s.c.) and potent topical corticosteroids, leading to a massive clinical improvement after three months. Potential mechanisms underlying the onset of GM after COVID-19 vaccination include molecular mimicry and the stimulation of autoreactive lymphocytes. To the best of our knowledge, only nine cases of GM after COVID-19 vaccination have been reported in the literature, two of them induced by Vaxzevria®.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1916

Cutaneous Lymphocytic Infiltrate in a Patient with Non-Hodgkin's Lymphoma: Case report

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Title: Cutaneous Lymphocytic Infiltrate in a Patient with Non-Hodgkin's Lymphoma: Case report

Introduction: Jessner-Kanof disease is a rare, benign, and self-limited condition characterized by a cutaneous lymphocytic infiltrate. Although it is not typically associated with underlying systemic diseases, it can occur in patients with lymphoproliferative disorders. The histopathologic features of Jessner-Kanof disease include a superficial dermal lymphocytic infiltrate with a "mantle" arrangement around blood vessels and the dermo-epidermal junction.

Case report: I will present a case of Jessner-Kanof infiltrate in a patient with stage IV-A non-Hodgkin lymphoma (small B-cell marginal zone type), currently in remission. On dermatological examination, pruritic, erythematous-violaceous lesions were observed on the forearms and lower legs, more pronounced in the extensor regions of the knees. Histopathological analysis of the lesions confirmed the diagnosis of Jessner-Kanof infiltrate. The histopathological features included epidermal acanthosis, hyperorthokeratosis, and a dense perivascular lymphocytic infiltrate. Treatment with systemic antihistamines and topical corticosteroids was initiated, resulting in favorable progress and complete resolution of the lesions. The patient has not experienced any recurrences and remains under long-term dermatological and hematological monitoring.

Discussion: This case highlights the importance of considering Jessner-Kanof disease as a differential diagnosis in patients with cutaneous lymphocytic infiltrates, particularly in those with underlying systemic conditions like Non-Hodgkin's lymphoma. The disease can mimic other cutaneous lymphocytic conditions, and its diagnosis is primarily based on histopathologic findings. The response to treatment with antihistamines and corticosteroids in this case supports the benign and self-limited nature of the disease.

Conclusion: Jessner-Kanof disease is a rare, self-limited skin condition characterized by a lymphocytic infiltrate. Clinicians should be aware of this diagnosis, particularly in patients with lymphoproliferative disorders, as it can present with distinctive clinical and histopathological features. Effective management with antihistamines and corticosteroids resulted in complete resolution of the lesions in our patient.

Abstract N°: 1935

Linear IgA dermatosis in adults and children : distinctive clinical and immunopathological features

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Introduction & Objectives:

Linear IgA dermatosis (LAD) is a rare autoimmune bullous dermatosis (ABD) that affects both adults and children.

Our objective was to Evaluate clinical and immunopathological differences between adults and children.

Materials & Methods:

Retrospective, descriptive, and analytic study including patients with LAD from January 2014 to June 2024.

Results:

15 patients were included: 8 children and 7 adults. The mean age was 6.25 years for children vs 37.4 for adults. Five adults (71.4%) were male vs five children (62.5%). A rosette pattern was observed in 4 adults (57.1%) and 5 children (62.5%). Pseudo-urticarial plaques were noted in 2 adults (28.5%). Mucosal involvement was noted in 2 adults (28.5%) and 3 children (37.5%). Facial involvement was significantly more frequent in children: 7 cases (87.5%) vs 1 adult (14.2%) (**p=0.009**); particularly peri-oral involvement (**p=0.019**). Trunk involvement was significantly more frequent in adults: 100% vs 37.5% (**p=0.019**). External genitalia involvement was observed in 4 children (p= 0.051). No significant histological differences were observed. Adults were treated with dapsone (85,7%) and systemic corticosteroids (57,1%), while children received dapsone (75%), systemic corticosteroids (37,5%), and colchicine (12,5%). All treatments were combined with topical corticosteroids. Treatment-related complications were observed in children: one developed adrenal insufficiency due to corticosteroid, and another developed anemia under dapsone. Complete remission was achieved in 6 adults (85.7%) and 6 children (75%). One adult and one child required higher corticosteroid doses, and rituximab was effective in one child.

Conclusion:

LAD is the most common form of ABD in children. In adults, it may be less typical. Our study showed a higher prevalence of trunk involvement in adults and a higher prevalence of facial, particularly peri-oral, involvement in children. However, immunopathological features were identical in both populations. These clinical features can serve as diagnostic tools. Nevertheless, larger studies are necessary to better investigate these distinctive features.

Abstract N°: 1954

Relapse of Systemic Lupus Erythematosus with Chilblain and Oral Lesions: Case Presentation

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Introduction & Objectives: Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease of unknown origin that can affect almost any human body organ, including the skin and the oral mucosa. Chilblain lupus, also known as "Hutchinson lupus" or perniotic lupus, is a rare chronic form of systemic LE on the extremities, usually triggered by exposure to cold. Oral lesions in SLE are more common but rarely co-exist with chilblain cutaneous LE.

Materials & Methods: A 27-year-old Caucasian male presented with a two-week history of painful erythematous-violaceous macules and papules bilaterally on his palms. The patient had an 8-year history of SLE in remission on oral hydroxychloroquine at a dose of 200 mg once daily. Clinical examination also revealed scattered, painless, polymorphous erosions, ulcers on the gingiva and buccal mucosa, and a well-demarcated erythematous plaque with a central whitish papule on the hard palate. The oral lesions caused mild discomfort to the patient when consuming spicy foods.

Results: Laboratory examination of the peripheral blood revealed leukopenia (2940/μL), neutropenia (1600/μL), and lymphopenia (1000/μL). Anti-nuclear antibodies (ANA) were positive (1:640), and there were elevated anti-double-stranded deoxyribonucleic acid (anti-dsDNA) titers, low C3 and C4 levels, and a positive Coombs test without hemolysis. SLE Disease Area Index (SLEDAI) Score was 8, indicating moderate disease activity.

Hydroxychloroquine was increased to 200 mg twice daily, and oral methylprednisolone was initiated at a dose of 16 mg daily with a 25-day tapering. In addition, topical mometasone 0.1% cream was applied twice daily for 20 days on the cutaneous lesions. Significant improvement in cutaneous and mucosal lesions along with partial normalization of laboratory findings was noted within four weeks.

Conclusion: Chilblain lupus is an uncommon clinical manifestation of SLE, with fewer than 70 cases reported in the literature according to a 2007 report. The aetiology is not clear, but it has been classified as a type III hypersensitivity reaction, mediated by immune complex deposition in skin vessels.

During an SLE flare, oral lesions are unusual and may signal heightened immune activation.

Given SLE's female predominance, this case report of mucocutaneous manifestations in this young male emphasizes the importance of skin and mucosal examination in all patients with SLE.

This case highlights the importance of early diagnosis of rare mucocutaneous manifestations as a marker of SLE relapses.

EA DV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1965

six-month clinical outcomes of omalizumab treatment in bullous pemphigoid:a retrospective study

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Introduction & Objectives: Bullous pemphigoid (BP) is the most prevalent autoimmune blistering disorder, predominantly affecting elderly individuals. Systemic and topical corticosteroids are considered the first-line treatment for BP. In this study, we aimed to assess the clinical efficacy of omalizumab (omalizumab) in the treatment of BP.

Materials & Methods: We retrospectively analyzed the medical records of 46 patients with a BP diagnosis who underwent omalizumab treatment for at least six months.

To evaluate treatment response, the patients' initial presentation, pre-omalizumab condition, and their status at the 1st and 6th months after omalizumab treatment were assessed. The following parameters were used to evaluate treatment efficacy: Bullous Pemphigoid Disease Area Index (BPDAI) score, Pruritus Visual Analog Scale (PVAS), treatment response scale (complete response (CR), partial response (PR), no significant clinical improvement (inefficiency), recurrence).

The study protocol is detailed in the flowchart (Figure 1).

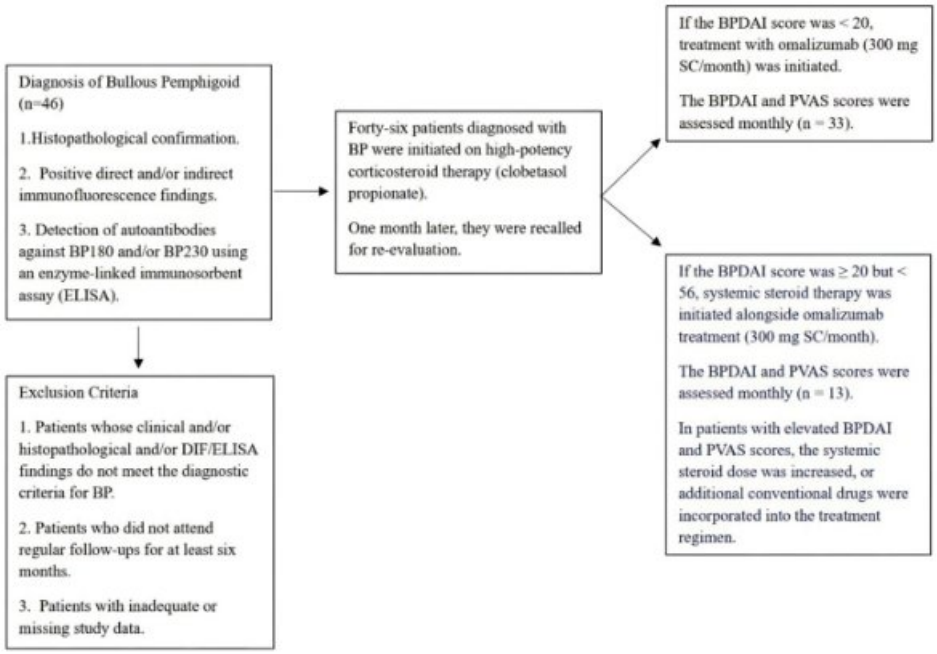

Figure 1. Flowchart illustrating the selection of the study sample

Results: Of the 46 patients included in the study, 45.7% were male, and the mean age at diagnosis was 79.6 years. The upper and lower extremities were the most commonly affected areas, observed in 93.5% of cases (n=43) (Table 1).

Table 1. Sociodemographic and clinical characteristics of the patients (n=46)

Gender (n,%)	N=46
Male	21 (45.7)
Female	25 (54.3)
Age (years), mean (range)	76 (54-94)
Lesion Localization (n,%)	
Trunck	36 (78.3)
Upper/Lower extremities	43 (93.5)
Oral mucosa	16 (34.8)
Scalp	6 (13)
Clinical presentation (n,%)	
Severe urticarial lesions	19 (41.3)
Eczematous lesions	18 (39.1)
Bullous lesion	42 (91.3)

Among the patients, 71.7% (n=33) did not require any treatment other than omalizumab. Systemic steroids were the most commonly used treatment in patients in addition to omalizumab (% 21, n=12)

In patients who did not respond completely to treatment, the mean age was higher (p=0.030), the BPDAI score was higher in the 1st month of treatment (p=0.000), the PVAS score was higher in the 1st month of treatment (p=0.016), and the IgE level was higher (p=0.040) (Table 2).

Table 2. Analyzing the characteristics of patients who complete responders versus non-complete responders

	Absense of CR	CR	X2 / Z	p*
Gender				
Male	7 (46.7)	14 (45.2)	0.009	0.923
Female	8 (53.3)	17 (54.8)		
Age (years)	81 (66-94)	75 (54-91)	-2.170	**0.030**
BPDAI (In the 1st month)	12 (2-30)	0 (0-6)	-5.603	**0.000**
BPDAI (In the 6th month)	0 (0-8)	0 (0-9)	-1.625	0.104
PVAS (In the 1st month)	2 (0-8)	0 (0-8)	-2.414	**0.016**
PVAS (In the 6th month)	0 (0-7)	0 (0-3)	-0.140	0.889
Individuals With Eosinophilia	7 (46.7)	15 (48.4)	0.012	0.913
Elevated IgE Level	15 (100)	23 (74.2)	4.686	**0.040**

*Pearson Chi-Square, Fisher's Exact test, Mann Whitney U analysis.

Conclusion: In a review the CR rate in patients with BP treated with omalizumab was reported as 68%. In our study, although this rate was found to be similar in the first month of treatment, it was higher in the sixth month. This suggests that the effectiveness of omalizumab treatment may be higher over a longer period.

There are not many studies comparing changes in BPDAI scores during omalizumab treatment. In the study by Alexandre et al., the average BPDAI score was reported as 56. In our study, the average BPDAI score was lower, and the CR rate was found to be lower in patients with high BPDAI scores. It can be concluded that omalizumab treatment is beneficial in patients with mild to moderate BP.

Abstract N°: 1982

Blister Fluid as a Source of Biomarkers for Steroid Resistance in Patients with Life-Threatening Bullous Dermatoses: A Case-Control Study

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Introduction & Objectives:

Bullous dermatoses (BD) are life-threatening skin conditions characterized by blistering and erosions on the skin and mucous membranes. These conditions are genetically predisposed and antibody-induced. Systemic glucocorticosteroids (CSs) remain the cornerstone of treatment for BD. However, a small yet significant proportion of patients with pemphigus and bullous pemphigoid (BP) exhibit resistance to standard CS therapy. While the mechanisms underlying steroid resistance (SR) have been explored at the receptor and gene levels, investigations at the level of blister fluid remain lacking.

The aim of this study was to evaluate the levels of IL-10, IL-15, IL-4, TNF-α, CCL11, CXCL8, and granulysin in the serum and blister fluid of patients with pemphigus and BP, as well as to assess their correlation with steroid resistance.

Materials & Methods:

A case-control study was conducted involving 39 patients with pemphigus and 10 patients with BP. The pemphigus group comprised 9 men and 30 women, aged between 30 and 80 years. The BP group included three men and seven women, aged from 37 to 85 years. The control group consisted of 40 healthy volunteers, including 18 men and 22 women, with an average age of 51 years. Steroid resistance was determined based on the Murrell consensus (2008). Among the pemphigus patients, 45% (n=18) were identified as steroid-resistant, while only 7% (n=3) of BP patients did not adequately respond to CS treatment. Cytokine and chemokine levels were measured using the ELISA method.

Results:

In steroid-resistant patients, there was a statistically significant increase in serum levels of CXCL8 compared to those who were steroid-sensitive. Interestingly, CXCL8 levels in blister fluid were highest in patients who responded well to CS therapy ($p<0,001$). In steroid-resistant BP patients, serum TNF-α levels were significantly elevated compared to the steroid-sensitive group ($p<0,0485$). Conversely, in blister fluid, levels of IL-15, IL-4, CXCL8, and granulysin were significantly higher in patients with a poor response to CSs ($p<0,0317$; 0,0241; 0,0121; and 0,0485, respectively).

Conclusion:

This study identified potential biomarkers of CS therapy in the blister fluid of patients with pemphigus and BP. Analysis of blister fluid can serve as a complementary tool to histological methods, enhancing diagnostics and differential diagnosis of severe BD. Furthermore, it may facilitate real-time monitoring of therapy response and prognosis for these diseases. This approach could enable timely initiation of adjuvant therapies for patients who are refractory to CSs, thereby minimizing the risk of complications and potential side effects.

Abstract N°: 2016

Histopathological Surprise in Achromic Lesions: Diagnosing Cutaneous Lupus Erythematosus Without Classic Clinical Features

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Introduction & Objectives: Cutaneous lupus erythematosus (CLE) encompasses a heterogeneous spectrum of dermatological manifestations, often associated with erythematous lesions, photosensitivity, and, in some cases, systemic involvement. However, its diagnosis may be challenging in the absence of these hallmark clinical features. The variability in presentation requires a comprehensive approach to clinical and histopathological evaluation to ensure accurate diagnosis and timely intervention.

Materials & Methods: We present a case of CLE diagnosed histopathologically in a patient with asymptomatic achromic lesions, without prior erythematous lesions, photosensitivity, or systemic symptoms, highlighting the pivotal role of histopathological evaluation in atypical presentations.

Results: A 70-year-old male, with recurrent ulcers on the left shin in the context of chronic venous insufficiency, reported the gradual development of asymptomatic, achromic, and slightly atrophic patches over the past nine months that appeared on the dorsal aspect of the hands but progressively extended to the face, trunk, limbs, and buttocks. The patient denied any history of erythematous lesions prior to the achromic ones, photosensitivity, or systemic symptoms associated with lupus. Upon clinical examination, the lesions were achromic with a slight degree of atrophy. Given the appearance of the lesions, a differential diagnosis was considered, including morphea, vitiligo, and pityriasis versicolor. Wood's lamp examination was negative. Histopathological analysis of the biopsy specimen revealed epidermal atrophy with follicular plugging and interface dermatitis. In the superficial dermis, there was a dense lymphocytic infiltrate, both perivascular and perifollicular, along with interstitial mucin deposition. These histopathological findings were pivotal in confirming the diagnosis of CLE, despite the absence of classical clinical signs and a negative antinuclear antibody test. Throughout the follow-up period, the lesions remained stable without progression, and there was no evidence of systemic involvement. The final diagnosis, based on the clinical presentation and histopathological features, was cutaneous vitiligoid lupus erythematosus.

Conclusion: This case highlights the critical role of histopathological evaluation in diagnosing CLE, particularly in patients who do not present with classical clinical markers. The absence of erythematous lesions and photosensitivity posed a diagnostic challenge, emphasizing the need for heightened clinical suspicion and reliance on histopathology for accurate identification. Recognizing atypical presentations of CLE is essential to ensuring timely diagnosis and appropriate management, thereby preventing potential complications associated with delayed recognition of the disease.

Abstract N°: 2020

Pemphigus induced alopecia : a rare presentation

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Introduction:

Pemphigus vulgaris (PV) is a rare autoimmune bullous disease characterized by autoantibodies targeting desmogleins, leading to intraepidermal acantholysis. While scalp involvement is frequent, hair loss remains an uncommon manifestation, potentially delaying diagnosis. We report a rare case of PV initially presenting as erosive alopecia.

Case report:

An 80-year-old female presented with a three-month history of a progressive alopecic plaque affecting the right frontal, vertex, and parietal scalp. Despite multiple antibiotic treatments, no improvement was noted. Two weeks prior to admission, the patient developed widespread flaccid bullae and painful oral erosions. Histopathology confirmed suprabasal acantholysis, and direct immunofluorescence revealed intercellular IgG and C3 deposits, establishing the diagnosis of severe PV with a Pemphigus Disease Area Index (PDAI) score of 55. Unfortunately, within 24 hours of admission, the patient developed a massive pulmonary embolism and succumbed to the complication, preventing further assessment of scalp lesion evolution under treatment.

Discussion:

Although scalp erosions are frequently observed in PV, associated alopecia is rarely reported. In a study by Veraitch et al., only 5.4% of PV patients exhibited alopecic scalp lesions, making it an uncommon but distinct manifestation. The pathophysiology of PV-related alopecia is thought to involve direct autoimmune-mediated acantholysis within the hair follicle unit, specifically targeting keratinocytes of the outer root sheath. Immunopathological studies have demonstrated intercellular IgG deposition in hair follicle structures, reinforcing the hypothesis of an autoimmune-mediated hair loss mechanism.

Scalp involvement in PV is associated with increased disease severity and prolonged remission times. A prospective study demonstrated that patients with scalp lesions had significantly higher PDAI scores and longer times to achieve remission compared to those without scalp involvement. This suggests that the presence of alopecia may indicate a more severe disease course, necessitating a more aggressive therapeutic approach.

Differential diagnoses of erosive alopecia include lichen planopilaris, discoid lupus erythematosus, cicatricial alopecia, and bullous impetigo. Histopathology and direct immunofluorescence remain essential in differentiating PV from other inflammatory scalp disorders. Additionally, trichoscopic evaluation and the hair pull test may serve as valuable non-invasive diagnostic tools, with the latter demonstrating anagen effluvium, a potential scalp equivalent to the Nikolsky sign.

Conclusion:

This case underscores the importance of recognizing alopecia as a rare but significant manifestation of PV. Early identification of scalp involvement may facilitate prompt diagnosis, preventing unnecessary treatments and disease progression. Given its potential association with more severe disease, clinicians should consider PV in patients presenting with progressive scalp erosions and alopecia, even in the absence of classical mucosal or cutaneous lesions. Timely intervention and appropriate immunosuppressive therapy are crucial for improving patient outcomes and preventing further complications.

Abstract N°: 2027

Effective Use of Tofacitinib in Refractory Pansclerotic Morphoea: a 2 case series from a tertiary centre

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Introduction & Objectives:

Pansclerotic morphoea is a rare, progressive, and debilitating form of localized scleroderma, often unresponsive to conventional treatments. Despite maximal immunosuppressive therapy, the prognosis for severe cases remains poor, with substantial impacts on quality of life. Emerging evidence supports the use of Janus kinase inhibitors (JAKi) such as tofacitinib in modulating fibrotic pathways and immune dysregulation, offering hope in refractory disease.

Materials & Methods:

Two patients with severe pansclerotic morphoea demonstrate the challenges in management and the potential benefit of tofacitinib. Both had progressive disease despite therapies, including methotrexate, mycophenolate mofetil, hydroxychloroquine, abatacept, systemic corticosteroids, and repeated courses of intravenous methylprednisolone.

Results:

The first patient experienced worsening skin thickening, blistering with secondary infections, and abdominal pain caused by restrictive fibrotic bands. Disease progression severely impaired mobility and daily functioning. Despite extensive treatment, including the addition of abatacept, significant clinical control was unattainable. Following initiation of tofacitinib at 10 mg daily, the patient demonstrated marked clinical improvements by three months, including enhanced skin pliability, reduced pain, and cessation of new infections. Skin thickness improved within six months, and functional mobility was restored without adverse effects.

The second patient faced severe skin sclerosis and immobility, leaving her reliant on a wheelchair and unable to perform basic daily tasks. This had significant personal and social implications, including losing her independence. Despite escalating treatments, her disease continued to worsen. Three months after starting tofacitinib, her skin texture has slowly improved, pain levels decreased, and she regained some functional ability and mobility.

Conclusion:

These cases highlight the significant burden of refractory pansclerotic morphoea and the limitations of current NHS-approved treatments. Tofacitinib, by targeting Janus kinase-STAT signaling pathways implicated in fibrosis, provided meaningful clinical improvement in two patients otherwise unresponsive to conventional therapy. Further research is needed to establish the long-term efficacy and safety of JAK inhibitors in morphoea and expand therapeutic options for this challenging condition.

EA DV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 2048

Rosacea-Like Lupus Erythematosus: A Challenging Diagnostic Case

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Rosacea-Like Lupus Erythematosus: A Challenging Diagnostic Case

Introduction & Objectives:

Lupus erythematosus (LE) is a chronic autoimmune disease with various cutaneous manifestations, often mimicking other dermatologic conditions such as rosacea. Misdiagnosis can lead to therapeutic delay and disease progression. We report the case of a 35-year-old male patient with persistent facial erythema initially treated as rosacea without improvement, ultimately diagnosed as lupus erythematosus.

Materials & Methods:

A 35-year-old male patient, with no relevant medical history, presented with persistent centrofacial erythema, papules, and telangiectasia. Based on clinical and dermoscopic findings : linear vessels and follicular pustules; rosacea was initially suspected. He was treated with topical metronidazole and oral doxycycline for two months without improvement. Dermoscopic reevaluation revealed follicular plugs and fine white scales, raising suspicion of lupus erythematosus.

Laboratory tests showed positive ANA and anti-Ro/SSA antibodies. A skin biopsy confirmed interface dermatitis with perifollicular lymphocytic infiltration and dermal mucin deposition, consistent with cutaneous lupus erythematosus (CLE).Treatment with hydroxychloroquine 200 mg daily, topical calcineurin inhibitors, and strict photoprotection led to significant improvement within three months.

Results:

Rosacea-like lupus erythematosus remains an uncommon and diagnostically challenging entity due to overlapping features with rosacea. Clinically, both conditions can present with centrofacial erythema, papules, pustules, and telangiectasia. However, lupus erythematosus typically exhibits follicular plugs, white scales, and arborizing vessels on dermoscopy features absent in classic rosacea.

The pathophysiology of rosacea is primarily linked to dysregulation of the innate immune response, increased Demodex mite density, and abnormal vascular reactivity. In lupus erythematosus, immune complex deposition, complement activation, and a predominantly Th1/Th17-driven immune response lead to characteristic skin lesions. Dermoscopic differentiation is critical: while rosacea typically presents with superficial linear vessels, lupus erythematosus exhibits deeper arborizing vessels and follicular plugs due to immune-mediated folliculotropism.

Another key differentiator is the response to initial treatment. Rosacea typically improves with topical metronidazole, or oral doxycycline, whereas rosacea-like lupus erythematosus remains refractory. In such cases, laboratory evaluation and histopathological analysis are essential.The delayed diagnosis in our case can be attributed to the misleading rosacea-like appearance, leading to initial empirical rosacea treatment. Early recognition and differentiation are crucial, as lupus erythematosus requires immunomodulatory treatment to prevent progression to systemic involvement, unlike rosacea, which is primarily managed with anti-inflammatory and antiparasitic agents.

Conclusion:

Rosacea-like lupus erythematosus is a rare yet clinically significant presentation of lupus erythematosus. This case

highlights the importance of considering lupus erythematosus in patients with refractory rosacea-like symptoms.

Abstract N°: 2051

Cutaneous Gluten Sensitivity Beyond Celiac Disease: A Case of Dermatitis Herpetiformis Controlled by Gluten-Free Diet Alone

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Introduction & Objectives:

Dermatitis herpetiformis (DH) is an autoimmune blistering disorder strongly associated with gluten sensitivity, classically linked to celiac disease (CD). However, rare cases of DH occur without intestinal involvement, raising questions about the underlying mechanisms. While dapsone is often the first-line symptomatic treatment, a strict gluten-free diet (GFD) is known to provide long-term disease control. We present a case of DH without CD, where the patient experienced a significant and sustained improvement with GFD alone, emphasizing the potential therapeutic role of dietary intervention even in the absence of enteropathy.

Materials & Methods:

A 26-year-old male with no significant medical history presented with a 4-month history of a polymorphic, pruritic eruption characterized by symmetrical papulovesicular, crusted, and urticarial lesions on the back and limbs. The lesions were intensely pruritic and resistant to topical corticosteroids.

Clinical suspicion of DH was confirmed by direct immunofluorescence (DIF) of a perilesional skin biopsy, revealing granular IgA deposits in the dermal papillae. Serologic tests for anti-tissue transglutaminase (tTG) IgA, anti-endomysial antibodies, and deamidated gliadin peptides (DGP) were all negative. Duodenal biopsy showed normal villous architecture.

Despite the absence of celiac disease, the patient was advised to initiate a strict gluten-free diet without dapsone. After three months, pruritus and lesions resolved completely, with sustained remission for over one year.

Results:

The observed improvement with GFD alone in a DH case without serologic or histopathologic evidence of celiac disease raises important considerations regarding the pathophysiology and management of DH.

DH and CD share a genetic predisposition (HLA-DQ2/8) and gluten-driven immune activation. However, DH can occasionally present as a skin-limited disorder, where epidermal transglutaminase (eTG) serves as the primary autoantigen without concomitant intestinal damage. In such cases, IgA deposits in the skin are gluten-dependent but do not necessarily reflect ongoing enteropathy.

Our patient’s rapid and sustained response to GFD supports the hypothesis that gluten elimination can significantly reduce the immunological trigger for DH, even in the absence of CD. This aligns with reports suggesting that DH represents the most specific cutaneous manifestation of gluten sensitivity, independent of gastrointestinal involvement.

From a clinical perspective, this case underscores the importance of considering a gluten-free diet in DH patients without CD markers, especially when dapsone is contraindicated or poorly tolerated. While seronegative DH remains a diagnostic and therapeutic challenge, this case suggests that dietary intervention alone can be both diagnostic and therapeutic.

Conclusion:

This case demonstrates that a strict gluten-free diet can lead to significant clinical improvement in dermatitis herpetiformis. The findings support the role of gluten as a central driver of DH and emphasize the importance of considering dietary intervention as a primary therapeutic strategy in selected cases. Further research is needed to better understand the immunological mechanisms underlying skin-limited gluten sensitivity.

Abstract N°: 2080

Coexistence of Pustular Psoriasis and IgA Pemphigus: A Rare Duet

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Introduction & Objectives:

The coexistence of psoriasis and bullous pemphigoid is well-documented, but its association with pemphigus, particularly IgA pemphigus, is exceptionally rare. We report a case of pustular psoriasis associated with IgA pemphigus, highlighting the diagnostic and therapeutic challenges of this unusual presentation.

Materials & Methods:

A 75-year-old man with a history of pustular psoriasis presented with new-onset flaccid bullous lesions following treatment discontinuation. Clinical examination revealed erythematous, squamous annular plaques with peripheral pustules (consistent with pustular psoriasis) and half-half blisters on the forearms and back. Histopathology confirmed pustular psoriasis (subcorneal pustules, psoriasiform hyperplasia) and IgA pemphigus (intraepidermal bullae, subcorneal cleavage, and IgA deposits on direct immunofluorescence). The patient was treated with dapsone (100 mg/day), resulting in resolution of bullous lesions and no flare-ups at two-month follow-up.

Results:

This case represents a rare association between pustular psoriasis and IgA pemphigus, with only one similar case previously reported. Proposed mechanisms include shared inflammatory pathways, genetic predisposition (e.g., HLA DRB1 alleles), and UV therapy triggering pemphigus. The temporal relationship between the two conditions varies, with psoriasis often preceding pemphigus. Distinguishing between the two can be challenging due to overlapping clinical features, but histopathology and immunofluorescence are critical for accurate diagnosis. This case raises the possibility of an overlap syndrome and underscores the need for awareness of this rare association, especially in cases of atypical presentations of both diseases.

Conclusion:

The confluence of pustular psoriasis and IgA pemphigus is a dermatological rarity that demands vigilance and a nuanced diagnostic approach. This case not only underscores the importance of histopathology and immunofluorescence in unraveling complex clinical presentations but also calls for deeper exploration into the mechanistic interplay between these two entities to optimize management strategies.

Abstract N°: 2134

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.

Abstract N°: 2162

A Rare Case: Pustular Pyoderma Gangrenosum With Atipical Localisation Triggered By Flare Of Ulcerative Colitis, Using Tzanck Smear To Support Early Diagnosis And A Multidisciplinary Treatment Approach – A Case Report

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Introduction & Objectives: Pyoderma gangrenosum(PG) is a rare neutrophilic dermatosis. Its diagnosis can be difficult due to its variable presentation, clinical similarities with other conditions, association with several systemic diseases, and lack of specific histopathologic or laboratory findings. Early diagnosis and treatment can minimize morbidity,prevent unnecessary aggressive surgery, improve aesthetic outcomes and help minimize complications from long term use of systemic treatment. In this report, we present a rare case of pustular PG with atipical localization that was triggered by flare of ulcerative colitis (UC) and evaluate its multidisciplinary treatment approach. This case also highlights the importance of the using Tzanck smear for early and quick diagnosis.

Materials & Methods: A 56-year-old female patient presented to our clinic with pustules on her face, neck, gluteal and inguinal region. 3 weeks prior to her presentation she started methylprednisolone 48 mg/day for UC. She was not under follow-up for UC and had initiated flare therapy on her own. As the dose tapered to 4 mg/day, pustules began to appear. Dermatological examination revealed multiple pustules, the largest measuring 1,5 cm in diameter, located at the malar region, dorsum of the nose, left auricula, frontal region, right lateral of the neck and intergluteal cleft. Dark erythematous bordered ulcerated plaques were observed below the right areola and in the left inguinal region. Tzanck smear ruled out herpetic ulcer as no acantholytic cells and multinucleated giant cells were observed. The presence of abundant neutrophils suggested PG. (Figure 1) No growth was observed in bacterial and fungal culture. Rectosigmoidoscopy confirmed active UC. Diagnosis of PG was made based on pustules extending into the dermis on histopathology (Figure.2), the presence of neutrophils on Tzanck smear, exclusion of the infection, presence of ulcerated pustules within the 4 days, rapid regression of pustules with methylprednisolone.

Results: Methylprednisolone 32 mg/day, azathioprine 2x50 mg were started. After 2 weeks, all lesions regressed completely. At the two-month follow-up no new lesion was observed. Her follow-up is being continued with azathioprine 50 mg twice daily, methylprednisolone 12 mg/day and mesalazine 2x2gr.

Conclusion: This case demonstrates that dermatological and systemic diseases can occur together and that such conditions require a multidisciplinary approach. PG, triggered by flare of UC, completely regressed with appropriate systemic treatment. This case emphasizes the efficacy of immunosuppressant treatment options in addition to steroids. After early appropriate treatment, ulcers and pustules healed completely. That case also shows the importance of early treatment.

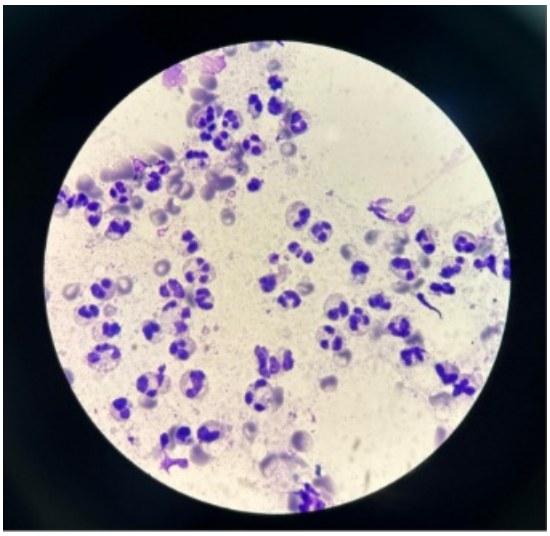

Figure.1: Abundant neutrophils on Tzanck smear.

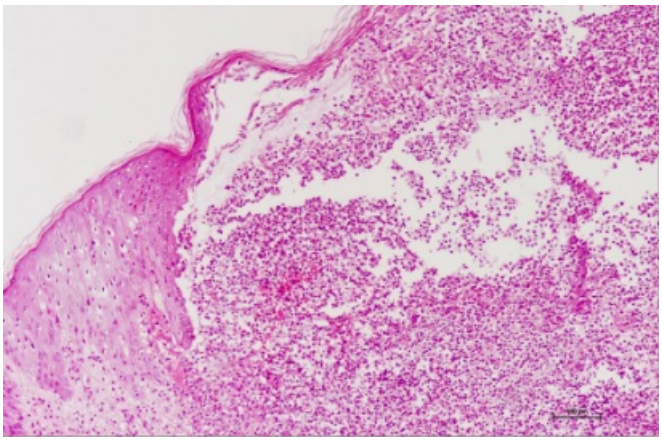

Figure.2: Epidermal mild orthokeratosis, acanthosis and subepidermal pustul extending into dermis.(H&E stain)

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Abstract N°: 2196

Bullous pemphigoid in association with vitiligo

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Introduction & Objectives: Bullous pemphigoid (BP) and vitiligo both are autoimmune disorders of the skin and mucous membranes. Each of them may be associated with other diseases of autoimmune nature, but BP is most frequently preceded by degenerative neurological disorders while vitiligo is mainly related to thyroid abnormalities. On most occasions the treatment for vitiligo, namely phototherapy, may trigger BP and only few cases in the literature describe their fortuitous concurrence.

Materials & Methods: We report an 87-year-old woman with 6-month history of pruritic vesiculobullous eruption on the trunk, upper and lower extremities. Her past medical history included stroke at the age of 83 and vitiligo since the age of 84. The clinical examination revealed generalized polymorphic rash consisting of tense bullae with clear contents on an erythematous-edematous background, and erosions covered with haemorrhagic crusts. Additionally, well-demarcated depigmented macules of various size were present on the trunk and on the scalp, particularly in the frontal and occipital regions along with poliosis of the right eyebrow. Some of the blisters were within the borders of the vitiliginous patches. Besides the thorough medical and drug history, routine laboratory, skin biopsies for histology, direct immunofluorescence (DIF), and immunoserologic tests were performed.

Results: : Routine laboratory investigations revealed peripheral eosinophilia, and elevated blood glucose and inflammation markers. Histopathologic and immunofluorescence investigations were compatible with BP. ELISAs for BP180 and BP230 were both strongly positive. Treatment with methylprednisolone 30mg/24h, doxycycline 100mg/24h, chloropyramine hydrochloride 1amp/24h, and topical corticosteroids resulted in marked clinical response.

Conclusion: The reported association of BP and vitiligo is extremely rare, with only few cases reported in the literature mainly in the setting of treatment regimens with PD-1 and PD-L1 inhibitors for metastatic melanoma or carcinoma. In our case, the appearance of both diseases is spontaneous. Besides, vitiligo occurred at an older age (84 years) whereas in general, it is usually found in younger individuals. As for BP, it is considered an autoimmune blistering disease of the elderly. While the pathogenesis of BP and vitiligo is not unveiled and is likely to involve a combination of genetic susceptibility and multiple environmental factors, their association may be regarded as a mere coincidence, or one could suspect the potential role of the pre-existing neurologic disease as a triggering factor in our patient.

Key words: bullous pemphigoid, vitiligo, neurological disorders

Abstract N°: 2228

AI Precision Imaging and Analysis for Epidermolysis Bullosa: Advancing Digital Clinimetry and Photometry

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Introduction & Objectives:

Accurate assessment of Epidermolysis Bullosa (EB) lesions is essential for monitoring disease progression and optimizing treatment strategies. However conventional lesion evaluation methods necessitate in-person visits, posing a significant burden for patients. Our proof-of-concept study aimed to improve EB lesion assessment using single-image analysis, facilitating remote evaluation and reducing the need for in-person clinical visits.

Materials & Methods:

Our research utilized 157 images from 29 patients diagnosed with EB. Images were captured using the Scarletred®Vision mobile app, a clinically validated Software as a Medical Device (SaMD) platform. This imaging tool enabled high-quality image capture in a real-world setting, providing consistency and precision across various environments.

The images were analyzed using a combination of advanced image processing techniques and deep learning models. A deep learning model pre-trained to assess psoriasis images was fine-tuned and used on 157 EB images to obtain an in-depth segmentation of EB lesions, delineating the affected tissue. In addition to lesion segmentation, we applied a second model specifically trained to detect open wounds. This model differentiated between intact and broken skin, enabling the identification of wounds that may require further medical attention or monitoring.

Additional image processing techniques were employed to detect and count the number of blisters within the segmented lesion areas. By leveraging augmented signal maps, the system identified the boundaries of the blisters by enhancing the contrast between the lesions and surrounding tissue, allowing quantification of blisters.

Results:

We developed a robust system capable of automatically detecting and analyzing EB lesions. The deep learning-based convolutional neural network (CNN) model effectively segmented EB lesions, accurately isolating affected areas from healthy tissue. This segmentation enabled more targeted analysis of the images, facilitating precise blister quantification through the image processing pipeline. Furthermore, the open wound detection model demonstrated its capability to identify regions of broken skin, providing valuable insights into lesion severity which may support wound care decisions. The combination of lesion segmentation, blister counting, and open wound detection resulted in a comprehensive system providing detailed insights into EB progression, offering a powerful tool for remote disease monitoring.

Conclusion:

By leveraging smart imaging technologies and deep learning algorithms, we developed an automated system for analyzing EB lesions, aiding clinicians in remote monitoring and personalized treatment planning. The findings of our research lay the foundation for integrating image processing and AI tools into remote diagnostic practices, enhancing accessibility, efficiency, and improving patient outcomes for rare and chronic skin conditions like EB.

Abstract N°: 2240

The combination of generalized morphea and eosinophilic fasciitis: a rare case report

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Introduction & Objectives:

Generalized morphea (GM) is presented by many morphea patches (greater than four plaques on the trunk, arms, head, or neck). Eosinophilic fasciitis (EF), also called Shulman syndrome, is a rare variant of scleroderma or deep form of morphea involving the subcutis and fascia of the skin characterized by fascial thickening with an eosinophilic tissue infiltrate and peripheral eosinophilia. A potential link between epoxy resin, GM and EF overlap has been reported previously. However, the causative mechanism remains unclear. [Chan. H. W, et. al. 2018].

Materials & Methods:

We present a rare case of combination GM and EF.

Results:

A 62-year-old woman since March 2024 felt pain and tightness in her left arm. After 4 months she noticed painful sclerotic patches on her back, which spread to the neck and abdomen areas. Her medical history included wrist trauma 3 years ago. Occupational history involved furniture restoration job, which suggested possible exposure to epoxy resin.

Physical examination revealed painful, indurated, longitudinally arranged sclerotic patches with the involvement of deeper layers on the left arm and abdomen. Sclerotic whitish plaques with central atrophy were observed on the upper back and neck. The “scleroderma pattern” on capillaroscopy was not found. Laboratory tests showed eosinophilia (1.1x109/L), elevated erythrocyte sedimentation rate (27 mm/h) and C-reactive protein (11.7 mg/L). Antinuclear antibodies were not detected, but a positive rheumatoid factor (111.2 kU/l) and cryoglobulinemia were present. These findings led to a rheumatologist consultation for suspected systemic connective tissue disease, which was not confirmed. Histology of a deep excisional skin biopsy from abdomen patch revealed subatrophy of the overlying epidermis, parakeratosis, significant collagen deposition in the subepithelial stroma, dermal fibrosis, and sparse chronic inflammatory infiltration around small blood vessels. Wrist ultrasound showed mild compression of the right median nerve in the carpal tunnel. Left elbow magnetic resonance imaging identified thickening of the ulnar nerve and fibrosis-like structures around Osborne’s fascia.

Based on clinical presentation, histological findings, laboratory test results, and radiological investigations, the diagnosis of GM with EF was confirmed. Regarding the progression and severity of the disease, systemic treatment with methylprednisolone 250 mg IV pulse therapy for 3 days was initiated, then followed by oral prednisolone 0,5 mg/kg daily for 3 weeks, with a subsequent dose tapering. Additionally, methotrexate 15 mg SC once weekly was prescribed, along with folic acid 5 mg on the next day. A 12-month course of systemic treatment was planned to achieve remission. Local treatment included clobetasol cream once daily for one month.

Conclusion:

The patient’s history of possible epoxy resin exposure and preceding trauma raises interesting considerations regarding potential environmental triggers in this case. EF remains a disease with potential for further research.

Abstract N°: 2241

Delayed diagnosis of lupus erythematosus panniculitis: a case report

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Introduction & Objectives: Lupus erythematosus profundus (LEP) is a rare form of chronic cutaneous lupus erythematous (LE), characterized by inflammation of the subcutaneous fat, presenting in 1%–3% of patients with cutaneous LE. The disease manifests as persistent, subcutaneous nodules with or without surface changes such as erythema and discoid lupus erythematosus features. Skin lesions are usually described as tender and deep nodules that appear on the buttocks, arms, thighs, or face. Healing of lesions is associated with scarring, lipoatrophy and rarely ulceration. In this report, we present a case of widespread LEP with delayed diagnosis and treatment.

A 47-year-old, otherwise healthy woman presented with a 2-year history of recurrent erythematous nodular and infiltrative lesions, on the face, followed by atrophic depressions. One and a half years later, she noted erythematous plaques on the left forearm and bilateral gluteal regions, accompanied by pain and burning sensations. The patient was initially treated for lumboischialgia due to persistent gluteal pain.

Skin examination revealed multiple painful, slightly indurated and depressed round nodules, as well as large depressed areas on superior aspects of her left and right buttocks. The differential diagnoses at this time included pancreatic panniculitis, lipoatrophy, morphea profunda, cold-panniculitis, LEP, and subcutaneous panniculitis-like T-cell lymphoma. .

Laboratory tests revealed positive values for CENP-B and ANA. Deep skin biopsy revealed a lobular panniculitis and a patchy superficial and deep dermal perivascular, perifollicular, and periadnexal lymphocytic inflammatory infiltrate without accompanying interface or epidermal changes.

Conclusion: Our case highlights the complexity of diagnosing autoimmune and inflammatory disorders affecting the subcutaneous tissue, emphasizing the need for a comprehensive clinical, serological, and histopathological assessment to establish an accurate diagnosis. The inclusion of LEP in the differential diagnosis is necessary in patients with disorders affecting the subcutaneous tissue.

Abstract N°: 2294

Neuro-Behçet Following COVID-19 Vaccination: A Case Report

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Introduction & Objectives:

Behçet's disease is a rare autoimmune disorder characterized by systemic vasculitis, the pathophysiology of which remains poorly understood. Neurological involvement is not uncommon in this condition, defining what is known as Neuro-Behçet. We report a case of Neuro-Behçet in a woman following her COVID-19 vaccination.

Materials & Methods:

We present the case of a 49-year-old woman with a history of acute rheumatic fever. She received a dose of an mRNA-based COVID-19 vaccine in September 2020. A few days later, she developed severe headaches, followed by a deviation of the right labial commissure and dysarthria, which she initially ignored. Due to the persistence of her symptoms, she visited the emergency department, where a stroke was diagnosed. Further etiological investigations revealed a thrombotic origin.

Four months later, the patient presented with bipolar oral aphthosis, prompting her to consult us. During the interview, she reported blurred vision with bilateral visual field loss, predominantly on the left side, and complained of gait disturbances. She was referred to ophthalmology and neurology consultations. Given these thrombotic, ophthalmological, and neurological manifestations, an etiological workup was conducted. Physical examination revealed aphthous ulcers on the inner side of the lower lip and cheeks, a bilateral positive Babinski sign, and absent patellar reflexes. A pathergy test was performed and returned positive. Ophthalmological examination revealed sequelae of retrobulbar optic neuritis. Brain MRI showed signal abnormalities in the periventricular and deep white matter, consistent with vasculitis lesions. A diagnosis of Neuro-Behçet was established, and corticosteroid therapy was initiated.

Results:

The etiopathogenesis of Neuro-Behçet remains poorly understood, with emerging evidence suggesting a role for infectious or environmental triggers in genetically predisposed individuals. During the COVID-19 pandemic, the virus and its derivatives have demonstrated close interactions with the immune system, suggesting their role in triggering various autoimmune diseases. The SARS-CoV-2 vaccination is not without risks and is associated with a wide range of complications, including dysimmune disorders, such as Neuro-Behçet in this case. While rare, such cases underscore the importance of vigilance in recognizing and managing post-vaccination autoimmune complications.

Conclusion:

This case highlights the potential for Neuro-Behçet to manifest following COVID-19 vaccination, emphasizing the need for heightened awareness of dysimmune complications in vaccinated individuals.

Abstract N°: 2311

Analysis of skin microbiome in patients with autoimmune pemphigus depending on the therapy received

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Introduction & Objectives: Autoimmune pemphigus is a group of life-threatening diseases. The basic treatment is high doses of systemic glucocorticosteroids followed by slow decreases. However, pemphigus relapses occur in 50% of patients, and severe side effects associated with glucocorticosteroid administration - in 65%. It is important to note that the leading causes of death are pneumonia and sepsis. The origin of infection can be the skin. To study the composition of the skin microbiome of pemphigus patients and of the control group by culture method, to analyze the composition correlation with the therapy received.

Materials & Methods: 40 patients with pemphigus: 34 (85%) - with pemphigus vulgaris and 6 (15%) - pemphigus foliaceus (15 males (37.5%), 25 females (62.5%), age - 53.05±12.62 years).

In patients in the active stage of pemphigus samples were taken before the start of therapy, in patients in remission - at the background of glucocorticosteroids in the maintenance dose (12.7±8.2 mg). The control group included 10 people (3 males (30%), 7 females (70%), age - 40±12.5 years).

Results: Samples from patients before treatment were characterized by the presence of a large number of S.aureus on the background of a decrease in total microbial diversity. In turn, samples from patients treated with glucocorticosteroids contained a greater diversity of species, with S.epidermidis and S.hominis species predominating in the samples. In addition, the composition of the microbiome from apparently unaffected areas in pemphigus patients was not different from the skin microbiome of the control group.

Conclusion: Our correlation analysis of the microbial composition of the skin of patients with pemphigus depending on the therapy with glucocorticosteroids demonstrated a significant difference. The importance of studying the microbiome is to prevent the development of severe infection complications and further progression of the disease and to improve the effectiveness of therapy.

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Abstract N°: 2327

When chilblain lupus mimics microbial eczema

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Introduction & Objectives:

Systemic lupus erythematosus is a complex autoimmune disease that can manifest itself in many different ways. Among its manifestations is Chilblain lupus, a rare but classic form.

This case report aims to examine the atypical presentation of lupus with chilblains resembling eczema, emphasizing the diagnostic difficulties.

Case report:

74-year-old women presented to dermatology department with a month history of an erythematous, infiltrated lesion occupying the pinna and tragus of the right ear, initially treated by an otorhinolaryngologist as otitis externa by antibiotic therapy without improvement, and referred to our department on suspicion of microbial eczema.

Dermoscopic examination revealed an erythematous background with white rosettes, dotted and linear congested vessels and white scales.

A skin biopsy was performed and demonstrated a discreetly acanthotic epidermis surmounted by orthokeratosis hyperkeratosis, the dermis was fibrous, with a dense interstitial and perivascular inflammatory infiltrate consisting mainly of lymphocytes and rare neutrophils arranged around vascular structures and the direct immunofluorescence was negative.

The patient was positive for antinuclear antibodies as well as anti-SSA antibodies without signs of dry syndrome on salivary gland biopsy. No other systemic signs or symptoms were present. A diagnostic of Chilblain lupus was made.

Treatment with synthetic antimalarials was initiated. The skin lesion started to subside 8 days after the start of treatment and completely disappeared after 6 months.

Discussion:

Chilblain lupus derives its name from its distinctive clinical presentation, characterized by papular or nodular, keratotic, purplish lesions, often occurring on the hands and feet and rarely on the nose and ears after exposure to cold or a drop in temperature (like classic frostbite), but persisting outside the cold season.

Diagnosis is based on clinical examination, skin biopsy results, the presence of anti-nuclear antibodies and even anti-SSA antibodies, which are present in most patients with this form of cutaneous lupus.

Treatment options may include local application of topical corticosteroids to reduce inflammation, antimalarials such as hydroxychloroquine to control skin lesions and systemic symptoms, and oral corticosteroids in more severe disease. Patients with lupus chilblain should also take steps to protect their skin from the cold by wearing warm clothing and using moisturizers

Conclusion:

The Chilblain lupus typically follows a chronic, recurrent, and unpredictable course. Remissions can last for several years. If the acute initial phase is managed effectively, even when severe (cerebral thrombosis or severe nephritis), the long-term prognosis is generally favorable.

Abstract N°: 2339

Cutaneous Lupus Erythematosus in Kyrgyzstan and LMICs: Disparities in Diagnosis, Treatment, and Patient Outcomes

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Introduction & Objectives: Lupus erythematosus (CLE), a chronic autoimmune disease with significant morbidity, disproportionately affects populations in low- and middle-income countries (LMICs), including Kyrgyzstan, where systemic healthcare disparities exacerbate diagnosis delays, limit treatment access, and lower patient outcomes. In this study, with an emphasis on Kyrgyz population, we examines differences in CLE management among LMICs.

Materials & Methods: MeSH terms (Lupus Erythematosus, Cutaneous, Healthcare Disparities, Delayed Diagnosis) and free-text keywords “cutaneous lupus,” “diagnostic delay,” “treatment access,” “LMICs,” and “Kyrgyzstan” were used in a comprehensive search across PubMed, Embase, Scopus, Web of Science, Cochrane Library, and regional databases (inception-2023). The findings were refined using Boolean operators (AND/OR), and filters for English/Russian and full-text availability. Inclusion criteria encompassed observational studies, clinical trials, or qualitative research reporting CLE epidemiology, diagnostic practices, treatment patterns, or patient outcomes in LMICs. Exclusion criteria removed case reports (<5 patients), non-peer-reviewed articles, and studies lacking primary data on disparities. From 897 screened records, 16 articles met eligibility after dual independent review (PRISMA 2020). Quality assessment employed the Newcastle-Ottawa Scale for observational studies.

Results: Key results showed a median diagnosis delay of 18 months (IQR: 10–26) in Kyrgyzstan and comparable LMICs caused by low availability to dermatologists (0.8 per 100,000 population) and dependence on clinical evaluation over histology (<25% of cases). While biologics and immunosuppressants were available to just 5% of patients, mostly in urban settings, corticosteroids (94% of studies) and antimalarials (68%) dominated regimens in treatment discrepancies. Travel restrictions and cost—mean out-of-pocket expense: 22% of monthly income—cause 3.2-fold greater risks of treatment stoppage (95% CI: 1.9–5.4) for rural patients. Patient outcomes, assessed via DLQI in 9 studies (n=632), showed severe quality-of-life impairment (mean score: 16.8 ± 6.4), with psychosocial stigma and mental health comorbidities reported in 74% of qualitative studies.

Conclusion: The study highlights systematic disparities in CLE treatment across LMICs, including Kyrgyzstan, and demands immediate action: increasing dermatological training, including telemedicine for remote diagnosis, subsidizing antimalarials, and giving patient-centered research first priority to help with psychological burdens. Achieving fair CLE management in underprivileged areas depends critically on strengthening healthcare infrastructure and promoting cross-sector alliances.

Abstract N°: 2342

An exceptional cutaneous presentation of systemic lupus erythematosus

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Introduction & Objectives:

Lupus erythematosus (LE) is a multi-system autoimmune disease in which skin involvement is prevalent and well-documented. However, ocular presentation with erythema and edema of the bilateral eyelids is exceptional. We present a case of recrudescence, during pregnancy, of quiescent systemic lupus erythematosus (SLE) with periorbital involvement.

Materials & Methods:

NA

Results:

A 32-year-old female patient, with a history of systemic lupus erythematosus (SLE) diagnosed 4 years ago without skin involvment, five months pregnant, presented to our department with an asymptomatic rash which gradually expanded to the face over the past five days. Physical examination revealed erythematous papular lesions located on the trunk, the roots of the upper limbs, and the face with erosions of the oral mucosa. In addition, bilateral erythematous-violaceous ecchymotic plaques were noted on both eyelids. A skin biopsy with immunofluorescence study was performed. Histopathological examination revealed epidermal interface dermatitis with vacuolation of basal keratinocytes. Direct immunofluorescence was negative. Laboratory tests showed leukopenia, lymphopenia, normocytic normochromic anemia, an accelerated sedimentation rate, hypoalbuminemia, hypoprotidemia, and proteinuria accompanied by positive antinuclear, anti-Sm, anti-SSA/SSB, and anti-RNP antibodies. In light of the clinical, biological, and histological findings, the rash was deemed consistent with cutaneous lupus. The patient was treated with daily doses of prednisone and hydroxychloroquine, resulting in the regression of the lesions within a period of four weeks.

Conclusion:

Our patient was diagnosed with systemic lupus erythematosus (SLE), without cutaneous involvement. During pregnancy, the patient presented a severe relapse of the disease, associated with renal involvement and atypical skin involvement of the eyelids. Periorbital erythema and edema are uncommon in acute LE, with usually unilateral involvement of the left orbit, and the upper eyelid more frequently affected. A few cases reported in the literature describe bilateral involvement, as in our patient’s case. The periorbital manifestation of the disease is often refractory to topical treatments, thus necessitating systemic medication. The management of LE during pregnancy poses a significant therapeutic challenge due to the contraindication of several treatment options. Synthetic antimalarials are a safe and effective therapeutic option. In resistant cases, azathioprine may be used. It is noteworthy that periorbital LE may represent the sole manifestation of acute cutaneous lupus, and diagnosis can be delayed due to clinical mimicry and the frequent negativity of the immunological work-up.

Abstract N°: 2411

Lupus Lyell-Like Syndrome : a case report

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Introduction:

Toxic epidermal necrolysis (TEN), also known as Lyell's syndrome, is a life-threatening dermatological condition typically induced by drug hypersensitivity. However, epidermal necrolysis can also occur in the context of autoimmune diseases, particularly systemic lupus erythematosus (SLE). Differentiating between TEN and lupus Lyell-like syndrome is crucial, as their management differs significantly. Lupus Lyell-like syndrome presents as an acute severe cutaneous flare of SLE, mimicking drug-induced TEN but with distinct clinical and immunopathological characteristics.

Case report:

We report the case of a 30-year-old female with a 10-year history of SLE involving polyarthritis, lupus nephritis, malar rash, anemia, and lymphopenia, managed with corticosteroids and synthetic antimalarials. She presented to the emergency department with multiple post-bullous erosions on the upper limbs and trunk, associated with cheilitis. The lesions were negative for Nikolsky's sign, and she was afebrile with no recent drug exposure. The initial differential diagnosis included TEN; however, the absence of a drug trigger, the progressive onset, and the distribution of lesions in photo-exposed areas raised suspicion of a lupus-related flare rather than drug-induced TEN. Histopathological examination revealed extensive epidermal necrosis, a feature common to both TEN and lupus-related necrolysis. However, direct immunofluorescence (DIF) demonstrated a characteristic lupus band, supporting the diagnosis of lupus Lyell-like syndrome.

Discussion:

Epidermal necrolysis is most commonly associated with drug reactions. However, in rare cases, lupus can present with severe bullous eruptions. The clinical presentation of lupus Lyell-like syndrome includes a gradual onset of cutaneous lesions, photo-distributed involvement, and limited mucosal damage, often without systemic deterioration. Importantly, lupus Lyell-like syndrome lacks the recent drug exposure. Key diagnostic features that differentiate the two entities include: progressive onset rather than the abrupt presentation seen in TEN, photo-exposed distribution of lesions, as opposed to widespread involvement in TEN, negative Nikolsky sign, which is often positive in TEN, limited mucosal involvement, whereas TEN typically presents with extensive mucosal damage, histopathological findings, showing epidermal necrosis in both conditions but often with features of connective tissue disease in lupus, DIF positivity for a lupus band, supporting an autoimmune etiology. Misdiagnosing lupus-related epidermal necrolysis as TEN can lead to inappropriate treatment, as systemic corticosteroids are generally contraindicated in drug-induced TEN but are essential for controlling lupus flares. In this case, early administration of systemic corticosteroids led to rapid improvement, confirming the autoimmune nature of the condition.

Conclusion:

Lupus Lyell-like syndrome is a rare but critical differential diagnosis in patients presenting with epidermal necrolysis. Recognizing its distinguishing features—progressive onset, photo-distributed lesions, absence of drug exposure, and lupus-specific histopathology—is essential to avoid misdiagnosis and ensure appropriate management. Clinicians should maintain a high index of suspicion in SLE patients with severe cutaneous flares mimicking TEN, as early immunosuppressive therapy leads to favorable outcomes.