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Abstract N°: 51

When a blister is the first clue

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Introduction:

Bullous pemphigoid (BP) is an autoimmune skin disorder characterized by the formation of tense subepidermal blisters triggered by antibodies against hemidesmosomal proteins BP180 and BP23011. BP typically has an incidence of 10 to 43 per million people per year2,3, with up to 18% of cases showing concurrent or subsequent malignancy4. However, a direct link with a specific cancer type remains elusive5. Notably, the literature documents a correlation between BP, neurological comorbidities, and oncological pathologies6. We present the case of a man who began with a nonbullous pemphigoid (NBP) and later evolved to BP in the context of a stroke and a prostatic adenocarcinoma.

Results:

An 81-year-old male patient with a history of hypertension, anxiety, and insomnia presented to the emergency department with a 2-year history of nonblistering skin lesions accompanied by scaling and itching that a month ago turned into multiple generalized tense bullae with serous content over the erythematous base. BP was suspected, which led to a skin biopsy that showed a subepidermal bullae with eosinophils and neutrophils and direct immunofluorescence with a lineal deposit of IgG and C3 on the basal membrane, both studies confirming the diagnosis. Given the association of BP with malignancies, extension studies were performed. Paraclinical investigations showed a marked inflammatory response, with the culture of the bullous lesions revealing methicillin-resistant Staphylococcus aureus. Additionally, a brain MRI was executed and revealed a subacute ischemic stroke and a suspicious lesion of metastatic involvement in the right parietal bone. On further investigations, total prostate-specific antigen was high, and CT revealed findings suggestive of prostatic neoplasia and inguinal lymphadenopathies. A lymph node biopsy was made and disclosed a florid dermatopathic lymphadenopathy. A prostate biopsy finally reported an acinar adenocarcinoma. Confirmation of BP diagnosis by dermatology led to systemic steroid and azathioprine therapy, resulting in significant re-epithelialization of the lesions and symptom resolution.

Conclusion:

The presented case provides a comprehensive insight into BP. The patient debuted with clinical symptoms consistent with NBP, a variant that comprises 20% of BP cases3. Then, two years later, it evolved to BP, and in this context, it led to the diagnosis of a solid organ neoplasm and stroke, both with association with this subepidermal dermatosis6. Highlighting that while not as common as lymphoproliferative disorders, the association with prostatic neoplasms is striking and has been infrequently reported in the literature3,6. This case underscores the critical relevance of cutaneous manifestations as indicators of underlying systemic diseases. Therefore, early diagnosis and multidisciplinary management are necessary to enhance patient prognosis.

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Abstract N°: 60

The role of UV-induced DNA damage, matrix metalloproteases and mi-RNAs in the pathogenesis of cutaneous and systemic lupus erythematosus

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Introduction & Objectives:

Cutaneous (CLE) and Systemic Lupus erythematosus (SLE) are autoimmune diseases with multifactorial pathogenesis that includes genetic predisposition, environmental triggers, and abnormalities of the innate and adaptive immune response. Clinical manifestations of LE range from mild effects limited to the skin in CLE, to serious and possibly life-threatening manifestations found in SLE. CLE can be further subdivided into acute cutaneous lupus erythematosus (ACLE), subacute cutaneous lupus erythematosus (SCLE), and chronic cutaneous lupus erythematosus (including discoid erythematosus (DLE)), LE tumidus (LET), and Chilblain lupus. UV radiation, the most important environmental trigger of LE, is known to cause DNA damage and cell apoptosis, especially in the upper layers of the epidermis. In a healthy skin, damaged keratinocytes are cleared effectively without triggering an immune reaction. However, in LE patients, the disposal of damaged cells is impaired, leading to an autoimmune response. Matrix metalloproteases (MMPs) are involved in the apoptotic clearance and skin homeostasis by rearranging extracellular matrix and thus enabling cell migration. Many MMPs are also UV regulated, making them potential players in the development and progression of LE. Besides MMPs, micro RNAs have also been shown to play important role in LE pathogenesis. Mir-31 and mir-150 are especially interesting since irregularities in their expression has been observed in LE samples compared to healthy skin. The UV-regulated mir-31 is involved in a variety of cellular processes, including the mediation of inflammatory cytokines and glucose metabolism involving GLUT1. As for mir-150, its downregulation has been shown to promote keratinocyte proliferation under hypoxic conditions.

Materials & Methods:

In our current work, we hypothesized that CLE subtypes and SLE react differently to sun exposure due to clinically observed difference in photosensitivity and manifesting skin lesions. We investigated the amount of DNA damage in lesional skin samples from patients with CLE and SLE, and compared them to healthy skin and samples from patients with polymorphous light eruption (PLE). Furthermore, we correlated the expression of mir-31 and mir-150 with the expression of the GLUT1 receptor, as well as MMP1 and MMP28. In addition, we have investigated the influence of UV radiation and pyruvate (as an intermediate product of glucose metabolism) on the expression of MMP28 in healthy skin cells.

Results:

We found out that MMP28 expression is up-regulated by UVA. Furthermore, high concentrations of pyruvate, in combination with UVA, have different effects on MMP28 expression in human fibroblasts compared to keratinocytes. MMP28 also showed distinct vertical distribution in Chilblain samples compared to all other LE subtypes. This expression pattern also correlated with decreased GLUT1 expression in the epidermis of the Chilblain patients, which in turn overlapped with high expression of both mir-31 and mir-150. The remaining LE subtypes showed no differences in mir-31 and mir-150 expression compared to normal skin and PLE. As for the GLUT1 expression, all other LE subtypes, except Chilblain LE had epidermal GLUT1 levels similar to normal skin. On

the other hand, the dermal expression of GLUT1 was uniformly higher in LE samples than in normal skin.

Conclusion:

This data present evidence for possible metabolic dysregulation playing a role in the etiology of LE.

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Abstract N°: 162

Chronic cutaneous lupus erythematosus in a patient with a history of Kikuchi–Fujimoto disease

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Introduction & Objectives:

Kikuchi–Fujimoto disease (KFD), histiocytic necrotizing lymphadenitis, is a benign and self-limiting disease, characterized by lymphadenopathy, fever, fatigue, and leukopenia. The symptoms of KFD can also mimic more sinister conditions such as non-Hodgkin lymphoma, tuberculous lymphadenitis, or reactive lymphadenopathy. The pathogenesis is unknown, but viral or autoimmune aetiologies. Several reports have emphasized the importance of KFD and SLE associations.

Materials & Methods:

A 26-year-old Caucasian male was admitted to the Outpatient Clinic with erythematous and infiltrative lesions on the right cheek and forehead in the left retro auricular area of irregular shape behind peeling on the periphery, without subjective symptoms since February 2023.

Eight years earlier, he was diagnosed with lymphadenopathy of the cervical and submandibular nodes (up to 2 cm) with leukopenia 2.21 G/l, elevated transaminases (ALAT exceeded the upper limit of normal by 6.6 times, AST by 4.3 times), moderately elevated LDH values and antibodies. against EBV in the IgG class. Based on hist-path examination of the lymph node, trepanobiopsy, and additional tests excluding the lymphoproliferative process, KFD disease was diagnosed as histiocytic, necrotizing lymphadenitis, and the liver damage was considered to be the result of EBV infection. The symptoms of KFD lasted for about 1.5 years and resolved spontaneously.

Laboratory tests showed no significant deviations from the norm, ANA 1: 160, immunoblot negative. The hist-pat examination of the lesion on the forehead revealed the features occurring in the course of lupus erythematosus. The patient did not report any other symptoms typical of SLE. A diagnosis of CCLE was made and the patient received hydroxychloroquine 200 mg/d, methylprednisolone aceponate topically, with good tolerance and improvement within a month.

Results:

KFD can occur in patients with pre-existing SLE, coexist with SLE, or evolve into SLE. Histologic and immunohistochemical features of SLE lymphadenopathy are, in some cases, indistinguishable from KFD. Therefore, it is recommended antinuclear antibody (ANA) screening at diagnosis and close follow-up, especially in patients with cutaneous lesions for the early detection of an autoimmune disease.

My patient had skin lesions typical of chronic lupus, without general symptoms and the patient didn’t fulfill SLE diagnosis criteria according to both ACR and EULAR classification. However, early initiation of hydroxychloroquine may prevent the development of SLE in the future. Unfortunately, there is no current literature advising how best to follow KFD patients regarding long-term complications and disease associations such as SLE.

Conclusion:

I present this case to highlight the rare clinical entity of KFD in the Caucasian population, the need for a long-term

follow-up, and the high risk of associated autoimmune diseases such as SLE. Our patient only developed symptoms of chronic lupus, approximately 7 years after the symptoms of KFD disappeared. Therefore, long-term follow-up of patients with this syndrome seems necessary.

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Abstract N°: 189

Consensus-based guidelines for the provision of pallia5ve and end-of-life care for people living with epidermolysis bullosa

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Introduction & Objectives:

Inherited epidermolysis bullosa (EB) is a cluster of rare, genetic skin and mucosal fragility disorders with multisystem and secondary effects, in which blistering and erosions occur in response to friction/mechanical trauma. Considering the incurable and potentially life-limiting nature of the condition and the challenges posed by its symptoms, a palliative approach to EB-related care is necessary. However, knowledge and experience related to the provision of EB palliative care is minimal. Evidence-based, best care guidelines are needed to establish a base of knowledge for practitioners to prevent or ease suffering while improving comfort at all stages of the illness, not just the end of life.

Materials & Methods:

This consensus guideline (CG) was begun at the request of DEBRA International, an international organization dedicated to improvement of care, research, and dissemination of knowledge for EB patients, and represents the work of an international panel of medical experts in palliative care and EB, people living with EB, and people who provide care for individuals living with EB. Following a rigorous, evidence-based guideline development process, the author panel identified six clinical outcomes based on the results of a survey of people living with EB, carers, and medical experts in the field, as well as an exhaustive and systematic evaluation of literature. Recommendations for the best clinical provision of palliative care for people living with EB for each of the outcomes were reached through panel consensus of the available literature.

Results:

This article presents evidence-based recommendations for the provision of palliative healthcare services that establishes a base of knowledge and practice for an interdisciplinary team approach to ease suffering and improve the quality of life for all people living with EB. Any specific differences in the provision of care between EB subtypes are noted.

Conclusion:

Because there is yet no cure for EB, this evidence-based CG is a means of optimizing and standardizing the IDT care needed to reduce suffering while improving comfort and overall quality of life for people living with this rare and often devastating condition.

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Abstract N°: 272

Generalized lichen planus pigmentosus and a new management approach

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Introduction & Objectives:

Lichen Planus Pigmentosus (LPP) is an uncommon form of lichen planus,in which lesions are mainly distributed over the sun-exposed areas, including the face and neck. Only a limited number of reports have indicated that the palms and soles could be affected in LPP. We are reporting the first case of generalized LPP accompanied by palmar involvement that experienced notable improvement with tofacitinib (Janus Kinase inhibitor) after two months.

Materials & Methods:

A 50-year-old female patient was referred to our dermatology clinic with widespread skin lesions that had been gradually developing over the past 14 years.

In her examination, the lesions were distributed over 90% of her body particularly her face, and other sun-exposed areas. The lesions were mostly violaceous and brown with fine scale, especially on the borders. Exfoliation in the violaceous lesions was detected on the palmar regions of her both hands. Furthermore, telangiectasia was observed at the center of the lesions predominantly the ones on the face.

Results:

A skin biopsy was conducted with differential diagnoses such as Subacute Cutaneous Lupus (SCLE), mixed connective tissue disease, dermatomyositis, systemic lupus erythematosus, Lichen planus (LP) pigmentosus, lichen planus atrophicus, ashy dermatosis, and Mycosis Fungoides (MF).

The microscopic examination indicated acanthosis and mild superficial perivascular inflammation. The inflammation produces a lichenoid interface reaction with the presence of some Civatte bodies in the basal layer. Upper dermal melanin incontinence and vascular ectasia are also noted. The result of Direct immunofluorescence (DIF) showed multiple globular deposits of immunoglobulin (Ig) G and M at the epidermis and dermoepidermal junction.

Collagen vascular tests such as anti-SCL-70, anti-SSA Ab, anti-SSB, anti-Jo-1 Ab, anti-smith Ab, and anti-RNP/Sm IgG were all negative.

Assuming that the lichen planus pigmentosus with telangiectasia due to the clinical and histopathological features, Tofacitinib (15 mg daily) was started for the patient. As a result, the medication was continued until the disease was completely resolved.

Conclusion:

Diffuse form of the LPP is a rare variant in which nails and palmoplantar area are often spared; however, palmar involvement has been observed in a few cases similar to our patient.

In 2018, Dabas et al. conducted a study of the 10 LPP cases focused on palmoplantar involvement. Like our report, most of the cases experienced rapidly advancing type of LPP, that necessitated a systemic treatment approach. In

addition, the palmar involvement was mostly accompanied by the atypical form of the LPP like our patient.

The inflammation and vulnerability of keratinocytes to destruction are mainly influenced by the signaling of interferon-g through the Janus kinase (JAK) - signal transducer and activator of the transcription pathway. In this process, chemokines induced by interferon-g (such as CXCL9, CXCL10, and CXC11) play a significant role in attracting inflammatory cells to infiltrate the interface dermatitis. Hence, promising therapeutic candidates for addressing LPP are JAK inhibitors. To our knowledge, this is the first report in which tofacitinib dramatically improves the generalized LPP with palmar involvement following 4 months.

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Abstract N°: 293

Relationships between systemic sclerosis and atherosclerosis: screening for mitochondrial-related biomarkers

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Introduction & Objectives:

Patients with systemic sclerosis (SSc) have a higher incidence of atherosclerosis (AS), and SSc can accelerate the process of AS. Mitochondrial injury in SSc can cause endothelial dysfunction, which leads to AS. Mitochondria seems like a hub that links SSc to AS. The aim of this study is to identify mitochondria-related biomarkers between SSc and AS.

Materials & Methods:

We downloaded SSc (GSE58095) and AS (GSE100927) datasets from the Gene Expression Omnibus (GEO) database. Then, we identified common differentially expressed genes (DEGs). After taking the intersection between genesthat have identical expression trends and mitochondrial genes, we used least absolute shrinkage and selection operator (LASSO) and random forest (RF) algorithms identified four mitochondria-related hub genes. Diagnostic nomograms were constructed to predict the likelihood of SSc and AS. We used CIBERSORT algorithms to evaluate immune infiltration in both disorders. We predicted transcription factors for hub genes and further validated in two datasets.

Results:

A total of 112 genes and 13 genes mitochondria-related genes were identified. These genes were significantly enriched in macrophage differentiation, collage containing extracellular matrix, collagen binding, antigen processing and presentation, leukocyte transendothelial migration and apoptosis. Four mitochondria-related hub DEGs (IFI6, FSCN1, GAL and SGCA) were identified using two machine learning algorithms. Nomograms showed good diagnostic value in GSE58095 (AUC = 0.903) and GSE100927 (AUC = 0.904). Memory B cells, γδT cells, Mo Macrophages and activated mast cells were significantly increased in AS. T-cell CD4 memory resting was decreased and M1 macrophages were increased in SSc. Four hub genes were closely linked with multiple immune cells. Gene set enrichment analysis (GSEA) showed IFI6 and FSCN1 were both related to immune-related pathways in AS and SSc. GAL and SGCA were both related to the mitochondrial metabolism pathway in SSc and AS. We predicted 20 transcription factors for 4 hub DEGs. Two transcription factors BRCA1 and PPARγ were highly expressed in SSc and AS.**Conclusion:**

Four mitochondria-related biomarkers were identified in SSc and AS. They had high diagnostic values and were associated with immune cell infiltration in both disorders. This study provides new insights into shared pathomechanisms between SSc and AS.

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Abstract N°: 474

B Cell Function in Systemic lupus erythematosus Patients is Regulated by the Upregulation of JUND

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Introduction & Objectives: Systemic lupus erythematosus (SLE) is driven in large part by B cells. JUND is an activator protein 1 (AP-1) family protein that has been linked to the regulation of apoptotic and cell proliferation, but the precise mechanisms whereby it functions remain to be fully elucidated. As such, this study was developed to clarify the functional importance of JUND gene expression in SLE, with further analyses of the functional role that JUND plays as a regulator of B cell proliferation and immune function.

Materials & Methods: RT-qPCR was employed to analyze JUND expression in the B cells of SLE patients and healthy subjects. CCK-8 and flow cytometry assays were used to characterize the proliferative activity, cell cycle progression, and apoptosis of B cells in which JUND was knocked down or overexpressed, while the immune status and autophagic activity in these cells was assessed through Western blotting and ELISAs. A Mouse model in which JUND was knocked down was also established, and the functional role of B cell JUND expression in the pathogenesis of SLE was assessed through Western blotting, ELISAs, and H&E staining.

Results: SLE patient B cells exhibited the upregulation of JUND, with such overexpression facilitating in vitro cellular proliferation and modulating the immune and autophagic status of these B cells. JUND knockdown was also sufficient to modulate the in vivo immune functionality and autophagic status of B cells.

Conclusion: JUND is upregulated in the B cells of patients with SLE wherein it functions to regulate proliferation, autophagy, and immunity.**

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Abstract N°: 562

Linear Morphea unilateral: “en coup de sabre” and extremities. Complete spectrum of both variants in a mexican girl case.

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Introduction & Objectives:

Morphea is a rare autoimmune disease of connective tissue characterized by localized sclerosis of the skin, with inflammatory, fibrotic, and vascular components and extracutaneous manifestations. The etiology is not fully understood, but it could be an immunologic predisposition, genetic factors, or epigenetic modification.

This disease encompasses a wide variety of clinical phenotypes. Laxer and Zulian (2006) described the most used classification, which includes circumscribed morphea, lineal morphea, generalized morphea, pansclerotic morphea, and mixed subtypes.

Linear Morphea (LM) is the most frequent form of scleroderma in childhood. It is usually a single unilateral lesion of linear distribution that presents two variants: one involves the extremities/trunk and the other face and scalp (LMcs).

The first is generally unilateral and progressive in extension and may affect muscle and bone tissue. The second, “en coup de sabre” (LMcs), is the most common, affecting 90% of pediatric patients diagnosed between 2 and 14 years of age. It is more prevalent in females than males (3:1).

We presented a Mexican girl case with unilateral LM, including both subtypes that affect extremities with linear lesions involving dermis, subcutaneous or deep tissue, and “en coup the sabre.” These subtypes of LM have not been reported simultaneously in the same patient.

Materials & Methods:

CASE REPORT: Mexican 12-year-old girl. She started at 3 years old with hyperchromic lesions on the face and forearm unilaterally on the right side, which progressed to hypochromic lesions. A decrease in the thickness of the skin and muscle mass reduction were also noticed. Later, the same injuries occurred in the right leg and distal extremities.

Physical examination. Plagiocephaly, asymmetric right unilateral affection shown facial hemiatrophy and atrophic extremities, with shortening and extension limitation.

Face strikingly linear and demarcated medial depression extending from the frontal bone, with greater involvement towards the part of the lower jaw, characteristics “en coup de sabre” with right malar deviation. Bad dental occlusion. The medial tongue groove deviated to the right and is retractable. Chest: A recent hyperchromic spot was seen in the right posterior thorax.

Right Extremities: Upper limb: brachydacty, hypo and hyperpigmented areas. More evident in the hand and forearm, were limited movements. Lower limb: shortening and muscle atrophy. The examined skin showed thickening, scleroderma changes, and linear cutaneous induration streaks, which also involved muscle.

Results:

There was no Neurological or Ophthalmological finding. Radiographically, hemiatrophy was shown on the right side, on the face, and in the long bones, with radiolucent images in the soft tissue area. The diagnosis of LM is mainly clinical, with a careful physical examination of the skin. Therefore, histopathological studies are not necessary.

Conclusion:

Linear Morphea is a type of connective tissue disease that is usually unilateral. The diagnosis is often based on characteristics of clínical findings and may affect muscles and underlying bones, causing growth disturbance, ankylosis, aesthetic damage, and nutritional imbalance. In most cases of LM. The subtypes of ML have been reported independently.

The case’s importance is to report that the two LM variants present unilateral simultaneously in the same patient, showing the complete clinical spectrum of the disease.

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Abstract N°: 710

Efficacy and Safety of Low-dose Corticosteroids as Maintenance Therapy in Pemphigus Vulgaris, A Randomized Trial

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Introduction & Objectives:

Pemphigus Vulgaris (PV) is an autoimmune blistering disease characterized by circulating antibodies against Desmoglein 1 (DSG-1) and 3 (DSG-3). Despite the promising effect of rituximab in treating pemphigus, many patients still need a minimal systemic corticosteroid (CS) dose to control the disease. There is still controversy about the risk/benefit ratio of using the minimal dose of CS in managing patients with pemphigus. Thus, in this study, we try to identify the efficacy and safety of low-dose CS maintenance therapy in preventing disease relapse and factors associated with relapse-free survival time.

Materials & Methods:

We studied pemphigus patients in complete remission on minimal therapy with positive DSG1 and/or DSG3 values. We split them into tapering and maintenance groups and followed them for 29 months. In the tapering group, the prednisolone dose was decreased to zero in 3-4 months, while the maintenance group had a minimal prednisone dose (<10mg/day).

Results:

Out of 57 patients enrolled (35 in the maintenance group and 22 in the tapering group), thirteen (52.63%) relapsed. The mean and median time to first relapse were 18.61 (±11.00) and 22 months, respectively. We found the maintenance group (hazard ratio, HR: 0.20, P=0.005) and the disease duration (HR: 0.98, P=0.01) to be associated with a longer time to relapse, while positive Anti-DSG1 results (HR: 11.61, P<0.001) and cumulative prednisolone dosage before study onset (HR: 1.07, P<0.001) were related to a shortened time to relapse. There were no significant differences in adverse effects between the study groups.

Conclusion:

Overall, maintaining a minimal dosage of CS may offer more prolonged remission with minimal side effects.

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Abstract N°: 851

Treatment in bullous pemphigoid: analyzing the morbimortality of systemic steroids

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Introduction & Objectives:

Bullous pemphigoid (BP) is the most prevalent autoimmune bullous disease. High-potency topical corticosteroids have been accepted as first-line treatment in BP. However, it is a chronic disease, characterized by multiple relapses and topical therapies may not be sufficient. Systemic steroids are usually prescribed as second line; however, we must try to limit side-effects and minimize interventions when controlling the disease, particularly in the elderly. The aim of this study was to analyze the management of BP in our setting, with particular interest in the outcomes of patients treated with systemic steroids.

Materials & Methods:

A retrospective observational study was carried out; all patients with a diagnosis of BP, attended in our department between the year 2000 and the first semester of 2020 were included. The epidemiological, clinical, immunological, histopathological and therapeutic characteristics of all patients, including dosage, duration and lines of treatment, efficacy, need for hospitalization and adverse events, if present.

Results:

Of the total of 257 patients with BP studied, in which high/very high-potency topical corticosteroids were used as first-line treatment, 41 (16%) achieved remission. Systemic steroids were prescribed in 209 cases; however, 91 patients needed further intervention to control the disease. However, in 64.6% of BP cases included in the study cohort, topical and/or systemic corticosteroids were the definitive treatment line (**Table 1**). When analyzing steroid dosage, 86.4% received doses < 0.5 mg/kg/day (0.3-0.4 mg/kg/day) and mean steroid treatment duration was 3.33 (±1.77 SD) months. 155 BP cases were deceased at the end of the study period; no differences in steroid doses employed in relation to hospitalization or death were found (**Table 2**). In terms of survival time, doses > 0.5 mg/kg/day were not related to poor prognosis. No adverse events directly related to systemic steroids was observed.

Discussion/Conclusion:

Despite being the second line treatment accepted for most BP guidelines (0.5-1 mg/kg/day prednisone), systemic corticosteroids have been classically associated to higher mortality and increased side-effects. However, mortality has also been associated to higher titles of BP180 NC16A antibodies, higher disease activity and, therefore, higher systemic inflammation. Advance age in BP patients does not facilitate to achieve the goal of improving the quality of life, preventing/reducing the risk of recurrence, when immunosuppressive drugs are needed. However, 0.3-0.4 mg/kg/day doses of prednisone can be sufficient to control the skin eruption, minimizing the morbimortality in BP patients.

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Abstract N°: 853

In Focus: Pemphigus Vegetans - A Rare Dermatological Puzzle Explored

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Introduction & Objectives:

Pemphigus is a group of autoimmune blistering diseases characterized by intraepidermal blister formation due to autoantibodies targeting desmosomal proteins. Pemphigus vegetans, a rare subtype of pemphigus, presents with verrucous, vegetating, and pustular lesions, predominantly involving skin folds and mucosal surfaces. This case report aims to illustrate the clinical presentation, diagnostic evaluation, and therapeutic management of pemphigus vegetans through the presentation of a patient case.

Materials & Methods:

A 62-year-old woman with no significant medical history, was admitted for vegetating lesions involving skin folds. Clinical examination revealed erosive plaques with vegetating centers surrounded by an epithelial collar, oozing and malodorous, located in the mammary, inguinal, umbilical, and anal folds. Non-follicular pustules, resting on erythematous skin, were observed in the right axillary, mammary, and leg extension folds, with a negative Nikolsky sign. Painful mucosal erosions, some covered with hemorrhagic crusts, were present on the lips, along with variable-sized erosions on the vaginal and anal mucosa.

Histological examination showed normo-acanthosic epidermis with a thin layer of orthokeratosis on top. Presence at multiple levels of deep sections of a dermo-epidermal cleavage containing rare fibrinous elements. Absence of acantholytic cells. Presence of images of neutrophilic granulocyte exocytosis marked elsewhere.

The dermis contains numerous congestive vascular elements, surrounded by a discreet infiltrate of regular mononuclear inflammatory cells.

Direct immunofluorescence demonstrates IgG and C3 deposits in a “net-like” pattern at the epidermal level.

The diagnosis of pemphigus vegetans was established based on clinical and histological features of the cutaneous and mucosal lesions.

The patient initially received intravenous corticosteroid bolus 15mg/kg daily for 3 days, followed by oral therapy (Prednisone) alongside with dermocorticoids with a significant improvement but with a relapse after a few months.

We decided to initiate rituximab as a second-line treatment, resulting in a spectacular improvement without complications

Results:

Pemphigus vegetans is a rare variant of pemphigus, characterized by distinct clinical and histopathological features. Diagnosis relies on clinical evaluation, histopathological examination, and direct immunofluorescence studies of lesional skin or mucosa. Treatment typically involves immunosuppressive therapy with corticosteroids and immunosuppressants, aimed at suppressing autoimmune activity and preventing disease progression. Close monitoring for treatment response and adverse effects is essential to optimize patient outcomes.

Conclusion:

Pemphigus vegetans is a rare autoimmune blistering disorder with unique clinical and histological characteristics. Early recognition and prompt initiation of appropriate immunosuppressive therapy are crucial for achieving remission and preventing disease complications. Further research is warranted to elucidate the underlying pathophysiological mechanisms and explore novel therapeutic approaches for pemphigus vegetans.

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Abstract N°: 892

mild cognitive impairment in pemphigus

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Introduction & Objectives: Pemphigus refers to a spectrum of severe autoimmune blistering disorders that has been associated with dementia in previous studies. Mild cognitive impairment (MCI) can be the first stage of progression into dementia. The objective of the present study was to evaluate the frequency of MCI in pemphigus patients compared to a control group.

Materials & Methods: This case-control study included 80 patients with pemphigus referred to our dermatology clinics in 2021. A group of 80 individuals without pemphigus who visited the same clinics for cosmetic consultation or interventions were regarded as controls. Age, sex, marital status, and education were recorded for all participants. For pemphigus patients, disease duration, medications, and disease severity (based on pemphigus disease area index [PDAI]) were also noted. The Persian version of Montreal cognitive assessment (MoCA) test was used to assess cognitive function.

Results: Pemphigus patients and controls were comparable in terms of age, sex, marital status, and education. Mild cognitive impairment was significantly more frequent in pemphigus patients than in controls (55% vs. 37.5%, P=0.026). Furthermore, the total MoCA score was significantly lower in pemphigus patients compared to controls (23.98 ± 3.77 vs. 25.21 ± 3.45, P=0.032); however, among MoCA's different domains, only the score pertaining to executive functions was significantly lower in pemphigus patients (P=0.010). After adjusting for age, marital status, and disease severity, multivariable logistic regression analysis revealed that with every one-year higher education in pemphigus patients the odds of MCI decreased by 52% (adjusted odds ratio=0.483, 95% confidence interval 0.326; 0.715, P<0.001).

Conclusion: The frequency of MCI was found to be significantly higher among individuals with pemphigus in this study, as compared to the control group. Similar results were noted for the overall scores of the MoCA test as well as its executive function domain. Additionally, higher level of education was associated with a decrease in the odds of MCI in pemphigus patients. Identifying pemphigus patients with mild cognitive impairment through the use of the MoCA test can facilitate early intervention, enabling them to seek help and support sooner.

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Abstract N°: 1070

New onset or flare-up BullousPemphigoid associated with COVID-19vaccines: A systematic review of casereport and case series studies

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Introduction & Objectives:

Numerous cutaneous manifestations have been associated with the COVID-19 outbreak and vaccination, but new-onset bullous Pemphigoid (BP) or flaring up the pre-existing BP is a rare side effect of COVID-19 vaccines that mentioned in a lesser extent in the literature. Therefore, we aimed to conduct a systematic review focused on the association of new onset or flare-up

BP and COVID-19 vaccination.

Materials & Methods:

A comprehensive literature search was conducted using PubMed (MEDLINE), Scopus, and Web of Science databases up to 11 March 2023. The search aimed to identify English-language studies reporting new-onset or flare-ups of BP as a potential side effect of COVID-19 vaccination. The search terms included bullous Pemphigoid and COVID-19 vaccination related mesh terms.

Results:

The systematic review of 40 included articles investigating the incidence of BP in individuals who received various COVID-19 vaccines revealed pertinent findings. Among the 54 patients with new-onset BP, the median age was 72.42 years, and most were male (64%). Conversely, the median age of the 17 patients experiencing a flare-up of BP was 73.35 years, with a higher proportion of females (53%). Regarding vaccination types, a significant number of patients (56%) developed new-onset BP after receiving the BNT162b2 vaccine (Pfizer-BioNTech).

Conclusion:

This study indicates a potential association between COVID-19 vaccinations, particularly mRNA vaccines, and the occurrence of BP. It suggests that this rare autoimmune disorder may be triggered as an adverse event following COVID-19 vaccination. However, it is important to note that the majority of BP patients in our study were unaffected by the COVID-19 vaccine, and even those who experienced worsening of their conditions were managed without significant consequences. These findings provide additional evidence supporting the safety of COVID-19 vaccines. Physicians should be mindful of this uncommon adverse event and encourage patients to complete their planned vaccination schedules.

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Abstract N°: 1078

A rare case of Acute Generalized Exanthematous Pustulosis in a Filipino Female Pediatric Patient with Systemic Lupus Erythematosus

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Introduction & Objectives:

Acute Generalized Exanthematous Pustulosis (AGEP) is a rare, febrile, drug-eruption which clinically presents with non-follicular pustules. Majority of AGEP cases are associated with medications like antibiotics, anti-hypertensive, immunosuppressors which are also used in Systemic Lupus Erythematosus (SLE) and its complications. To the extent of our knowledge, this is the first Filipino pediatric case of AGEP in SLE.

Materials & Methods:

Results:

An 18-year-old Filipino, female patient, diagnosed with SLE in remission, initially presented with multiple erythematous annular plaques on admission with subsequent antibiotic use. On Day 2 of Azithromycin, the patient presented with multiple non-follicular pustules, fever, leukocytosis, and subcorneal pustule on histopathology (AGEP Score: 11 – Definitive AGEP). Resolution was noted ten (10) days from the withdrawal of Azithromycin. IV Intravenous Hydrocortisone (7 mg/kg/day), topical potent steroids, moist dressing, and antihistamine were also utilized in this case.

Conclusion:

While there are still limited data on AGEP in SLE, this case has noted the intuitive increased risk of SLE patients to develop AGEP. Prompt drug withdrawal is still the standard of treatment in AGEP in SLE. Good prognosis is also observed in this case of AGEP in pediatric patient with SLE.

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Abstract N°: 1128

Hypogonadotropic hypogonadism, cutaneous horns, face mutilation with discoid lupus rash in young female with systemic lupus erythematosus

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Introduction & Objective:

systemic lupus erythematosus (SLE) in children and adolescent is rare, more common in females and is a great mimicker with heterogenous manifestation. Systemic involvement, cosmetic disfigurement caused by skin lesions and long term disease management add to the morbidity. Untreated and misdiagnosed disease can be fatal. We describe a case of young female with SLE, having severe cosmetic facial disfigurement and endocrinopathies

Materials & Methods: a young 20 year old female presented with multiple horny hyperkeratotic papules and plaques all over the face with surrounding erythema and scarring for 2 years. She had history of arthralgia, myalgia since childhood and was stunted with low body weight. On investigation she was found to be hypothyroid, had autoimmune hepatitis, high Parathyroid hormone (PTH), anemia and hypogonadotropic hypogonadism. Further investigations showed ANA 4 plus, positive anti-ds DNA, positive anticentromere B antibody and low C3 levels. Skin biopsy and Direct immunofluorescence (DIF) done were compatible with the diagnosis of Discoid lupus erythematosus.

Results: The patient was diagnosed as SLE as she fulfilled American College of Rheumatology classification criteria and was started on broad spectrum sunscreens, along with oral steroids and is currently under follow up with multidisciplinary approach .

Conclusion: five to ten percent of patients with DLE has coexistent SLE and when present specially in children and adolescent may have multi-system autoimmune involvement with wide diversity of disease manifestations. In adolescent and young females dealing with the disease is a special challenge with remitting relapsing nature of the disease, morbidity associated with it and its treatment. Our patient had severe facial disfigurement along with endocrinopathy, autoimmune hepatitis making it more challenging. This case also highlights the importance of awareness, early diagnosis and treatment of childhood and adolescent LE.

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Abstract N°: 1308

Expanding the clinical experience in vitiligo: quick response in 2 months treatment using topical ruxolitinib

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Introduction & Objectives: Vitiligo is an autoimmune disease that results in the loss of pigment in the skin due to immune-inflammatory pathogenesis. Though the estimated global prevalence of the disease is 0.5-2%, it has a profoundly deleterious effect on the patient's quality of life. Various off-label treatment options have been attempted in the past; however, their efficacy has been limited. In 2022 in the USA and 2023 in Europe, ruxolitinib cream was the only medication approved, to restore lost skin color in patients aged 12 years and above with non-segmental vitiligo. In our case study, we utilized ruxolitinib cream to treat a 53-year-old woman who had not responded to previous treatment modalities. The patient responded excellently to the therapy with visible improvements observed from the initial month of treatment.

Materials & Methods:

A female patient, aged 53 years and of Brazilian descent, presented with nonsegmental focal vitiligo on her face, which had been causing significant psychological distress, for the last three years that was suffering. The severity of the condition was assessed using the Dermatology Quality Index (DLQI), with a baseline score of 21. Despite previous attempts to alleviate the disease through traditional treatments such as topical corticosteroids, topical calcineurin inhibitors, and narrow-band ultraviolet UVB phototherapy, the patient reported no improvement in her condition. The patient's medical history revealed a comorbidity of autoimmune thyroid disease, as well as the last five years. After clinical diagnosis and photographic assessment of vitiligo activity, the patient was prescribed ruxolitinib cream as a treatment.

Results:

The use of ruxolitinib cream in a twice-daily application resulted in a swift and significant re-pigmentation of the skin in the patient within the first month of treatment. A two-month follow-up revealed a further improvement in the clinical presentation, with a reduction in the DLQI score to 8 and 3, respectively.

Conclusion:

Ruxolitinib cream, a selective Janus Kinase (JAK) 1/2 inhibitor, is the first medication that has been approved for repigmentation in patients with vitiligo, supporting the implication of the IFN-γ-chemokine signaling axis in the pathogenesis of the disease. In phase III clinical trials, treatment for 24 weeks resulted in a 60.7% improvement in 75% of the facial vitiligo area score. Of this, 29.8% was observed in true- V1 study and 30.9% in true -V2, one. In real-world scenarios, such as our case report, the response time was even quicker. A 75% improvement was observed in clinical, photographic, and DLQI evaluation in only two months of follow-up. Future studies with a larger number of patients will elucidate the efficacy, rapidity of action, and maintenance of response in vitiligo patients treated with ruxolitinib cream.

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Abstract N°: 1318

Gestationis Pemphigoid triggred by a DRESS Syndrome

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Introduction & Objectives:

DRESS syndrome is a severe drug hypersensitivity reaction characterized by rash, eosinophilia, and organ involvement, typically occurring weeks after exposure to the culprit drug. Diagnosis relies on clinical and laboratory criteria, with various scoring systems used for confirmation. The severity of DRESS is linked to systemic involvement, with its pathogenesis involving viral reactivation and ensuing inflammatory response. Serious complications and mortality in DRESS are mainly associated with systemic involvement and immunosuppressive treatments.

The main objectives are to assess the link between DRESS syndrome and the later onset of autoimmune conditions, particularly in young individuals, Identify possible risk factors for the emergence of autoimmune diseases following an episode of DRESS syndrome and emphasize the significance of ongoing monitoring for patients with a history of DRESS syndrome to promptly detect early indications of potential autoimmune disorders.

Here, we present a rare case of gestational pemphigoid following an episode of DRESS in a multiparous woman.

Case report:

A 34-year-old multiparous woman was admitted with a rash and fever 10 days after taking paracetamol for flu-like symptoms. Physical examination revealed a morbilliform rash with lymphadenopathy and facial edema. Blood tests showed elevated transaminases and positive HHV6 serology. Symptoms resolved within 10 days without treatment.

Two years later, during her fourth pregnancy, she developed gestationis pemphigoid, confirmed by histopathological and immunofluorescence tests, successfully treated with oral corticosteroids.

Discussion:

DRESS syndrome, a severe drug-induced cutaneous reaction with a 10% mortality rate, shares similarities with Stevens-Johnson syndrome and toxic epidermal necrolysis. It often occurs in older patients, associated with renal impairment, but in younger individuals, it appears to predict subsequent development of autoimmune diseases such as autoimmune thyroiditis, diabetes, systemic lupus erythematosus, and autoimmune hemolytic anemia.

Autoimmune diseases are more prevalent in younger and middle-aged women, suggesting an increased response to antigenic stimuli.

The mainstay of treatment involves discontinuation of the offending drug, with systemic corticosteroids typically improving symptoms.

Studies have documented the long-term development of autoimmune diseases, including autoantibody production, underscoring the importance of ongoing surveillance in patients with a history of DRESS.

Conclusion:

DRESS syndrome is associated with the risk of developing autoimmune diseases, particularly in young patients. Our case and other studies suggest that the pathophysiology of DRESS may explain this predisposition to autoimmunity.

Patients with DRESS should undergo long-term monitoring to detect potential autoimmune diseases.

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Abstract N°: 1413

Ultrasound and Shear-wave elastography as a tool for evaluating response to treatment of localized scleroderma: a prospective study

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Ultrasound and Shear-wave elastography as a tool for evaluating response to treatment of localized scleroderma: a prospective study

Introduction & Objectives:

Morphea is an autoimmune connective tissue disorder characterized by skin inflammation and fibrosis. Due to its insidious nature, measuring disease activity is hard, and clinical scoring tools have limitations. This study aims to investigate the accuracy and usefulness of ultrasound and shear-wave elastography (SWE), in determining morphea lesions activity in relation to clinical scores during a 6-month follow-up.

Materials & Methods:

This prospective study was conducted in a tertiary dermatology hospital. Eighteen newly biopsy-confirmed patients with at least one active plaque and normal skin on the contralateral side were examined.

Results:

Eighteen patients with an average age of 39.78±14.68 were examined. Of them, 15(83.3%) were female, 13(72.2%) had plaque-type morphea and 5(27.8%) had linear morphea. The most frequent lesions sites were upper limb (39.9%) and trunk (39.9%). After six months, patient's average dermis thickness (DT) (2.19±0.78 mm vs. 1.88±0.88 mm) as well as mean LoSCAT, LoSAI, LoSDI, PGA-A, and PGA-D scores decreased significantly ($p<0.05$). In contrast, hypodermis thickness and elasticity changes at the lesion site weren't significant ($p>0.05$). Although, both dermis elasticity and hypodermis elasticity in control sites were significantly lower than lesion sites after follow-up($p<0.05$). The decrease in PGA-D was correlated with DT decrease ($p > 0.05$, R: 0.514). Also, hypodermis thickness decrease was correlated with decrease in LoSCAT, LoSDI and LoSAI ($p>0.05$, R:0.610, 0.518, and 0.524 respectively).

Conclusion:

DT shown to be the most promising ultrasound measure for evaluating morphea activity. Although SWE isn't accurate for measuring disease activity, it is feasible to detect morphea lesions and damage in comparison to normal skin.

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Abstract N°: 1575

Therapeutic challenges in the treatmnet of calcinosis in autoimmune diseases.

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Introduction & Objectives: Calcinosis is a common complication of systemic autoimmune diseases. It can affect up to 20% of adult patients with dermatomyositis (DM) and may occur up to twice as often in juvenile dermatomyositis (JDM). Calcinosis is usually present in the form of superficial nodules or plaques which tend to accumulate in the skin, subcutaneous tissue, fascia, muscles or blood vessels. Clinical effects include contractures, ischemia, muscle atrophy, itching and ulcers, what significantly can restrict patients' quality of life and is considered as a therapeutic challenge requiring a multidisciplinary approach. Here we aim to add to the knowledge of treating calcinosis using STS injections.

Materials & Methods: We present a case of 71-year-old, male patient with dermatomyositis and a past medical history of primary cutaneous T-cell lymphoma (CTCL, *Mycosis Fungoides* form). First skin lesions appeared at age of 47, a diagnosis of CTCL was made 13 years later. Throughout that time the patient received PUVA, UVB phototherapy, INF-a and Targretin treatment. One year later, patient started to report periodic muscle weakness, swelling of the face and numbness with tingling in the fingers. Subsequently, Gottron's papules and calcinosis appeared. After correlating the laboratory and histopathological results with the patient's clinical features and meeting Bohdan and Peter classification criteria, in 2014 DM with Wonga subtype was diagnosed. Applied treatment included intravenous immunoglobulins, methotrexate with prednisone and mycophenolate mofetil. Due to mediocre clinical improvement, therapy with rituximab was performed. However, implemented treatment did not result in complete remission.

Results: One year after rituximab treatment, the patient was admitted to our clinic to undergo therapy with intralesional (IL) sodium thiosulfate (STS) and platelet-rich plasma (PRP) injections due to the presence of ulceration and calcinosis. Performed injections included affected areas of the fingertips, lesions on the abdomen and armpits. Treatments were scheduled once per month. After seven months, therapy was temporarily suspended due to surgery of the ectropion, which was caused by calcium deposits as well. Following therapy, a reduction of calcified lesions located on the fingers and armpits, a decrease in pain, and the healing of fingertip ulcers were reported.

Conclusion: Despite the numerous treatment strategies, calcinosis still remains a therapeutic challenge. Here, we present a case of successful use of a method involving PRP and STS injections. Due to the fact that the beneficial effect of described above treatment among patients with dermatomyositis remains poorly documented in the literature, further research and standardization of the procedure are needed.

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Abstract N°: 1716

Sphingosine-1-Phosphate Receptor 1 (S1P1) Agonist, Vibozilimod (BS6.890C), Effect on Bradycardia, Preclinical to First-in-human Clinical Translation

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Introduction & Objectives:

Sphingosine-1-Phosphate receptor 1 (S1P1R) modulators have been approved for the treatment of inflammatory and autoimmune disorders. S1P1R stimulation sequesters lymphocyte subsets in peripheral lymphoid organs, preventing inflammation. Due to effects on other subtypes of S1P receptors, these modulators may be associated with first dose effects such as bradycardia and atrioventricular (AV) conduction delays, along with macular edema and bronchoconstriction. Vibozilimod (VBZ, BS6.890C) is a novel S1PR agonist that acts on S1P1R and S1P5R while antagonizing S1P4R with significant lack of activity towards S1P3R. The effects of VBZ on cardiovascular parameters in rats along with results from the first-in-human study are reported here.

Materials & Methods:

VBZ, etrasimod, ozanimod and siponimod were administered intravenously at a dose of 3 mg/kg in anesthetized male Sprague-Dawley rats and their effects on heart rate (HR) and mean arterial blood pressure (MAP) were studied. All agents were infused slowly as solutions in distilled water or PEG-400 after a stabilization period of 20 minutes. In a single ascending dose (SAD) Phase 1 study (n = 6 + 2 placebo), the effects of VBZ on cardiac function in healthy adult males and post-menopausal females were evaluated by measuring heart rate, recording 12-lead ECGs and conducting 24-hour Holter recordings.

Results:

Bradycardia was observed after infusion of all the S1P1R agonists in rats. VBZ infusion was associated with a decrease in heart rate to 130 bpm, which returned to baseline (BL) of 400 bpm within 2 minutes. Etrasimod infusion reduced the HR to 100 bpm and took about 3 minutes to return to the BL of 370 bpm. Ozanimod infusion decreased the HR to 50 bpm, and the effect lasted for 6 minutes before returning to the BL of 360 bpm. However, the reduction in HR with siponimod, which was around 100 bpm, did not return to pre-dose values during the 10-minute recording period (see Fig. 1).

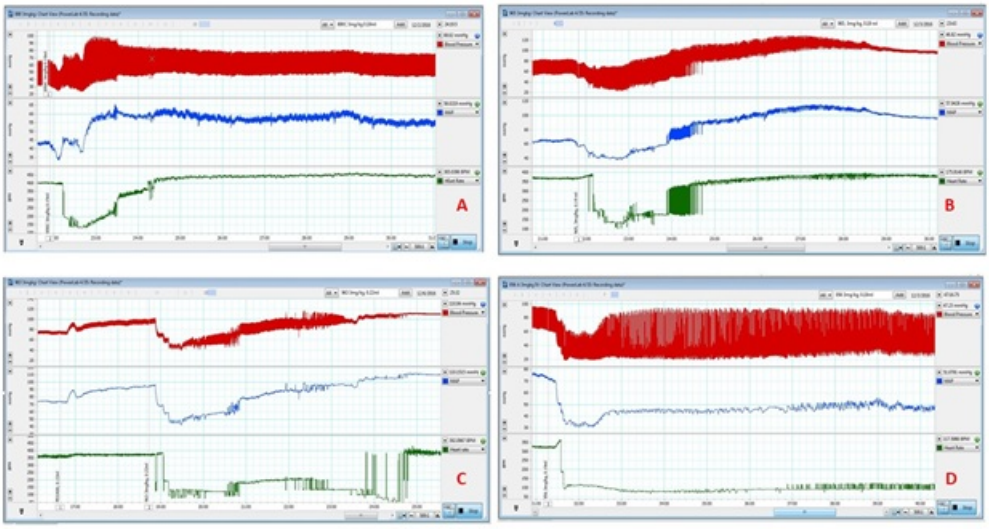
Following administration of VBZ to humans, effects mimicked those observed in the rat model with a dose response observed after doses of 0.1 to 6.0 mg. Mean bradycardia of magnitude greater than 10 bpm was observed following a dose of 1.0 mg or higher. The maximum change in mean (± SD) heart rate from baseline following the administration of single doses of 0.1 mg, 0.3 mg, 1 mg, 2 mg, 4 mg, and 6 mg in males was -2.2 (±3.1), -6.0 (±6), -15.3 (±6), -16.8 (±6.3), -17.2 (±5.9), and -16.5 (±3.9), respectively and for females, it was -15.0 (± 3.5) at 2 mg. The maximum change in HR was transient and observed between 2 to 3 hours following administration of VBZ. None of the treatment groups had a mean HR decrease of more than 10.5 bpm more than

5 hours after administration. Recovery from bradycardia for doses ≥4.0 mg in males and at 2.0 mg in females was faster, after a transient reduction at 2 to 4 hours, suggesting that subjects adapt and develop more rapid tolerance with higher exposure to VBZ. The mean change in HR from baseline is presented in Table 1. Thus, the observed bradycardia in humans closely resembled observed in the rat model.

Conclusion:

VBZ, a differentiated agonist at S1P1 and S1P5 receptors produced only a transient and well manageable effect on heart rate in humans, with faster recovery as observed in the preclinical model. Thus, the effects of VBZ observed in animal studies were successfully translated in humans in terms of reduction in magnitude of heart rate in Phase I study, thus providing improved safety on cardiac function.

Figure 1: Effects of different compounds on heart rate, blood pressure, and mean arterial pressure in anesthetized Sprague-Dawley rats



Changes in blood pressure (BP-Red line in mm of Hg), mean arterial BP (MAP-Blue line in mm of Hg) and heart rate (HR-green line in beats per minutes, bpm) were monitored continuously using a PowerLab® data acquisition system with LabChart 8 software (AD Instruments). X-axis represents time in minutes.

HR decrease (Bradycardia) : Vibozilimod (A)< Etrasimod (B)< Ozanimod (C)<Siponimod (D)
MABP change : Vibozilimod (A)<Siponimod (D)<Ozanimod (C)<Etrasimod (B)
The observed changes with Vibozilimod (VBZ) being transient and minimal in all the parameters

Table 1: Summary of Mean Change in Heart Rate (bpm) from Baseline (Δ) by dose level in Phase 1 Single Ascending Dose Study of Vibozilimod

Time (h)	0.1 mg	0.3 mg	1 mg	2 mg (M)	2mg (F)	4 mg	6 mg	Placebo
N	6	6	6	6	6	6	6	14
BL	56.8 ± 7.4	55.0 ± 6.1	60.8 ± 11.7	61.5 ± 7.5	61.2 ± 6.3	59.5 ± 5.6	56.2 ± 2.1	60.0 ± 10.8
0.5	0.8 (3.1)	1.2 (2.0)	0.8 (3.2)	0.2 (4.0)	-0.3 (5.2)	0.3 (2.1)	1.8 (4.5)	-0.9 (3.5)
1	1.7 (2.6)	0.7 (2.8)	-4.7 (4.4)	-6.3 (6.5)	-9.7 (4.3)	-8.3 (8.2)	-8.3 (4.9)	-1.1 (3.4)
2	-2.2 (3.1)	-4.7 (4.7)	-13.2*** (4.4)	-16.8*** (6.3)	-15.0*** (3.5)	-17.2*** (5.9)	-16.5*** (3.9)	0.2 (5.1)
3	-1.3 (4.1)	-6.0 (5.3)	-15.3*** (6.0)	-16.3*** (7.1)	-13.7*** (5.1)	-15.0*** (5.4)	-15.5*** (3.8)	-0.4 (4.0)
4	-0.8 (3.1)	-6.0 (6.0)	-14.3*** (6.2)	-13.8*** (7.4)	-11.0* (4.6)	-13.2*** (8.6)	-14.3*** (3.1)	0.6 (4.7)
5	5.7 (3.2)	5.0 (5.3)	-4.2 (5.3)	-6.5 (6.1)	-4.7 (4.5)	1.8 (8.6)	-1.5 (5.0)	15.9*** (9.3)
24	0.0 (3.2)	0.8 (4.5)	-5.3 (3.7)	-7.8 (6.7)	-3.8 (4.5)	-1.5 (4.8)	-6.3 (5.5)	1.4 (4.1)

Values are expressed as mean (±SD); BL – Baseline
For statistical analysis heart rate, entire data from pre-dose to EOS (Day 14) h was used; only data at BL, 0.5h, 1h, 2h, 3h, 4h, 5h, and 24 h represented in the table.
*p<0.05, **p<0.01, ***p<0.001 vs respective group BL heart rate. Two-way ANOVA followed by Bonferroni's test

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Abstract N°: 1719

tofacitinib in alopecia totalis

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Introduction & Objectives: Alopecia totalis (AT) is a subtype of alopecia areata which is an autoimmune condition characterized by the complete loss of hair on the scalp. emerging therapies such as tofacitinib, a Janus kinase (JAK) inhibitor, have shown promising results and gives potential treatment resulting in promoting hair regrowth n some patients with alopecia totalis.

The objective was to determine the efficacy and superiority of using the tofacitinib drug over other conventional treatment protocols in alopecia totalis.

Materials & Methods: A 17 year old male patient was presented with near complete hair loss on scalp since 6 months in our clinic. Few patches were noticed even when he was 9 years old which recovered by its own. There was no history of similar illness in family members and also no history of any drug. At the age of 17 he started to notice few patches again which was in alopecia ophiasis pattern in the beginning but then increased and involved the full scalp within 2 months.

After thorough investigations which included CBC, HbAIC, LFT, CREATININE, TB GOLD TEST, TSH we started the treatment with tofacitanib 5mg twice a day with monthly follow up.

The treatment was continued for 6 months

Results: Within 2 months hair started growing back and by the end of 6 month patient had recovered almost 90% of the hair back. During this time patient was asked to continue local application of fluticasone cream and tacrolimus ointment.

Conclusion: Treatment options for alopecia totalis are limited, and the prognosis for spontaneous hair regrowth is generally poor. However, emerging therapies such as tofacitinib, a Janus kinase (JAK) inhibitor, have shown promising results and gives potential treatment resulting in promoting hair regrowth n some patients with alopecia totalis. Tofacitinib is an off-label treatment option that requires careful consideration of the potential risks and benefits, as well as the challenges associated with insurance coverage and monitoring.

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Abstract N°: 1798

Epidermolysis bullosa acquisita pseudo-cicatricial pemphigoid in a child

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Introduction:

Epidermolysis bullosa acquisita (EBA) belongs to the group of subepidermal autoimmune blistering dermatoses. It is a rare pathology linked to the production of autoantibodies against type VII collagen. Pediatric cases are rarely reported in the literature. We present an atypical case with severe mucosal involvement.

Case presentation:

A 9-year-old girl presented with a pruritic mucocutaneous bullous eruption. Questioning revealed recurrent oral erosions for more than 6 months. Clinically, there were tense bullae resting on erythematous skin, located on the trunk, the extension areas of the limbs and the palmoplantar surfaces. Nikolsky's sign was negative. Oral (jugal and palatal erosions) and ocular (bilateral symblepharon and Meibomian gland dysfunction) mucusal damage was severe. The genital mucosa was unaffected. The biology work-up was unremarkable. Skin histology showed a bullous detachment with linear deposits of IgG and C3 along the dermal-epidermal junction in direct immunofluorescence (DIF). Indirect immunofluorescence (IIF) on cleaved skin showed the presence of anti-basal membrane antibodies on the dermal side. Systemic treatment with prednisolone 1 mg/kg/day combined with dapsone 2 mg/kg/day was started, resulting in rapid skin remission with the appearance of grains of melium. Mucosal involvement improved more slowly.

Discussion:

EBA is rarely seen in children. Two distinct clinical presentations have been described: the classic or mechanobullous form, characterized by cutaneous fragility and bullae at sites of trauma, and the inflammatory form, mimicking any other subepidermal autoimmune blistering dermatoses. The diagnosis of EBA in its pseudo-cicatricial pemphigoid variant was made in our patient on clinical grounds (severe pleural-mucosal involvement, moderate cutaneous involvement with development of milium grains), histological grounds (bullous detachment) and immunological grounds (linear deposits of IgG and C3 in DIF, positivity of anti-basal membrane antibodies on the dermal side of the dermo-epidermal junction in IIF of cleaved skin). The Elisa anti-collagen VII test could not be performed. Meibomian gland dysfunction has been described in hereditary epidermolysis bullosa, more rarely in EBA. The treatment of EBA is not codified. A number of immunosuppressive, anti-inflammatory and anti-neutrophilic treatments are proposed. Dapsone, alone or in combination with systemic corticosteroids, is often the first-line treatment. In cases where the ocular prognosis is compromised, ciclosporin is recommended. forms of this condition.

Conclusion:

We report a rare case of pediatric EBA with pseudo-cicatricial pemphigoid presentation, illustrating the great heterogeneity of clinical forms of this condition.

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Abstract N°: 1841

Can we use the asas criteria for axial spondyloarthritis and the modified new york criteria for ankylosing spondylitis to identify axial psoriatic arthritis patients

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Introduction & Objectives: there isn't any unified definition and diagnostic criteria for axial psoriatic arthritis (PsA). To analyze whether axial PsA (axPsA) patients (pts) meet the Assessment of SpondyloArthritis International Society (ASAS) classification criteria for axial spondyloarthritis (axSpA) and modified New York (mNY) criteria for ankylosing spondylitis (AS).

Materials & Methods: 104 pts (M/F–66/38), a hospital cohort, with PsA according to CASPAR criteria were observed. All pts had chronic back pain (CBP), which rheumatologist suspected to be axial involvement. All pts were evaluated for presence of inflammatory back pain (IBP) by ASAS criteria. 97 pts underwent pelvic radiographs, cervical and lumbar spine, hands and feet X-ray. Radiographic sacroiliitis (rSI) was defined as bilateral grade ≥ 2 or unilateral grade ≥ 3. Pts without rSI underwent sacroiliac joints MRI on Philips Multiva 1.5 T scanner. Active MRI sacroiliitis (MRI-SI) was categorized using ASAS 2016 criteria. Radiographic spondylitis (rSp) was defined as ≥1 syndesmophyte(s) of the cervical and/or lumbar spine. All results were interpreted by two experienced musculoskeletal radiologists. 92 patients underwent HLA B27 examination. The pts meeting ASAS classification criteria for axSpA and mNY criteria for AS were identified.

Results: IBP was found in 67 (64.4%) pts, CBP in 37 (35.6%) pts. 31 (29.8%) pts were at the age of above 40 at the IBP/CBP onset, 18 (17.3%) pts – at the age of above 45. RSI was found in 57 (58.8%) pts, syndesmophytes – in 57 (58.8%) cases; 6 pts of the 19 examined (31.6%) had MRI-SI. Among the 40 pts without rSI, 19 (47.5%) had syndesmophytes. Among the 97 examined pts, 38 (39.2%) pts had rSI along with syndesmophytes, while 19 (19.6%) pts had rSI without spondylitis, and 19 (19.6%) pts had isolated spine involvement (syndesmophytes) without rSI. HLA B27 was present in 28 (30.1%) cases. Among the 92 pts who underwent pelvic radiographs, sacroiliac joints MRI and HLA B27 examination, 51 (55.4%) pts met ASAS criteria for axSpA as follows: 33 pts met the imaging stream criterion (rSI or MRI-SI + ≥ 1 SpA feature), 6 pts met the clinical stream (HLA B27+ ≥ 2 other SpA features) and 12 pts met both of those. 41 (44.6%) pts didn't meet ASAS criteria for axSpA: they had neither rSI/ MRI-SI nor HLA B27; however, 27 (65.9%) of them had syndesmophytes. 48 (48.5%) PsA pts met mNY criteria for AS. However, specific PsA features were revealed among these pts: 18 (37.5%) pts had no IBP, 18 (37.5%) pts had older age (>40 years) at back pain onset, 34 (70.8%) pts had dactylitis, 38 (79.2%) – erosive polyarthritis, 23 (48.8%) – juxta-articular new bone formation, 14 (30.2%) pts had osteolysis, 23 (48,9%) pts – "chunky" non-marginal syndesmophytes, 40 (82,6%) pts had nail psoriasis, 28 (66,6%) pts were HLA-B27 negative.

Conclusion: in our cohort of axPsA pts 45% have not met ASAS classification criteria for axSpA. These criteria are therefore not useful for identifying axial involvement in PsA pts. A group of clinical features, more typical for axPsA pts has been identified, which will help to differentiate axPsA from AS in pts meeting both CASPAR and mNY criteria.

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Abstract N°: 1844

Gender-related differences in patients with axial psoriatic arthritis

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Introduction & Objectives: gender-related differences in axial psoriatic arthritis (axPsA) patients haven't been sufficiently studied. To analyze the effect of gender on clinical characteristics of axPsA patients.

Materials & Methods: 104 patients [M/F–66 (63.5%)/38 (36.5%)] with PsA according to CASPAR criteria were observed. All patients had chronic back pain, which rheumatologist suspected to be axial involvement. Patients' age 44.0±11.4 years, disease duration 9.8±8.8 years. Patients underwent standard clinical examination of PsA activity. BASDAI 5.9±3.6; ASDAS-СРБ 3.5 [2.8; 4.2]; DAPSA 38.0±21.7. Enthesitis was found in 56.7%, dactylitis in 41.8% of patients. All patients had psoriasis: BSA 4 [1; 11]%, PASI 10.5 [5.6; 18.0]. Nail psoriasis was found in 77.2% of patients. All patients were evaluated for presence of inflammatory back pain (IBP) by ASAS criteria. Patients underwent pelvic radiographs, cervical and lumbar spine, hands and feet X-ray. Radiographic sacroiliitis (rSI) was defined as bilateral grade ≥2 or unilateral grade ≥3. Examination included HLA-B27 status. Radiographs were interpreted by two musculoskeletal radiologists. All clinical and imaging features and patient reported outcomes were compared between male and female patients. Me [Q25; Q75], Pierson-$\chi 2$ tests were performed. All $p<0.05$ were considered to indicate statistical significance.

Results: the following differences were found between males and females with axPsA. Females had longer duration of spinal morning stiffness: 60 [60; 120] vs 60 [40; 90] minutes. More women than men had high activity by BASDAI: 34 (89.5%) vs 46 (69.7%; p=0.028) patients. Arthritis mutilans was found only in females, in 3 (8.1%) cases (p=0.02). Ankylosis of the hands and feet joints was more often found in women: 8 (25.8%) vs 2 (3.0%; p=0.04) cases. No differences between men and women were found in the frequency of IBP, cervical spine involvement, rSI, syndesmophytes in the lumbar and/or cervical spine, HLA-B27 positivity. Females more often had anxiety and depression according to the hospital anxiety and depression scale (HADS): 5 (13.5%) vs 2 (3.0%; p=0.04) cases. Women were more likely than men to receive TNF inhibitors: 14 (37. 2%) vs 16 (25.1%; p=0.043) patients; no differences were found in the frequency of interleukin inhibitor therapy.**

Conclusion: gender-related differences are observed in the clinical expression of axPsA. Though males are more likely to develop axial involvement, females have significantly worse disease status as measured by spinal morning stiffness, activity of spondylitis by BASDAI, more severe radiographic damage in the peripheral joints, and the frequency of anxiety and depression. Women more often received TNF inhibitor therapy. This information highlights the importance of considering gender differences in clinical practice.

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Abstract N°: 1845

Axial disease in psoriatic arthritis

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Introduction & Objectives: the latest data show that axial psoriatic arthritis (PsA) is associated with distinct radiographic features. To analyze characteristics of PsA patients (pts) with axial involvement.

Materials & Methods: 104 pts (M/F–66/38) with PsA according to CASPAR criteria were included. Pts had either a present or past history of back pain, which rheumatologist suspected to be axial involvement. Pts' age 44.0±11.4, disease duration 9.8±8.8 years. Pts underwent standard clinical examination of PsA activity: BASDAI 5.9±3.6, ASDAS-CRP 3.5 [2.8; 4.2], DAPSA 38.0±21.7, CRP 9.9 [3.5; 28.2] mg/L. Pts were evaluated for presence of inflammatory back pain (IBP) by ASAS criteria. HLA-B27 antigen status was observed. Physical examination included Bath Ankylosing Spondylitis Metrology Index (BASMI). Pts underwent X-ray of sacroiliac joints (pelvic radiographs), cervical and lumbar spine, and hands and feet. Radiographic sacroiliitis (rSI) was defined as bilateral grade ≥ 2 or unilateral grade ≥ 3. Hands and feet X-rays were evaluated using PsA-modified Sharp/van der Heijde score (SHS). Radiographs were interpreted by 2 musculoskeletal radiologists. Me [Q25; Q75], Pierson-$\chi 2$ tests were performed. All $p<0.05$ were considered to indicate statistical significance.

Results: IBP was identified in 67 (64.4%) pts, while 37 (35.6%) pts didn't meet IBP criteria – they had chronic back pain. In 31 (29.8%) pts back pain started at age of above 40. RSI was found in 57 (58.8%) cases; 26/57 (44.8%) pts developed rSI without IBP. A correlation was revealed between the presence of rSI and limited spine mobility by BASMI (r=0.347). 79 (76%) pts had erosive arthritis, 37 (36%) pts had numerous erosions. The average value of SHS was 82.79±64.77. Association was found between rSI and numerous erosions (p=0.003), as well as between rSI and juxtaarticular new bone formation (p=0.02). A correlation was revealed between the presence of rSI and the value of SHS (r=0,46; p<0,05). Syndesmophytes were detected in 57 (58.8%) cases. In 38/97 (39.2%) pts rSI was detected along with syndesmophytes, while 19/97 (19.6%) pts had isolated rSI without spondylitis, and 19/97 (19.6%) pts developed syndesmophytes without rSI. Restricted neck rotation was found in 85% of cases. 29 (28.0%) pts had isolated involvement of the cervical spine. Syndesmophytes in the cervical spine were found twice as often (in 52 pts) as in the lumbar spine (in 26 pts). Asymmetrical syndesmophytes in the lumbar spine were found in 53.8% of pts. An association was detected between asymmetrical syndesmophytes and radiographic changes on hands and feet X-rays such as osteolysis (p=0.005) and juxtaarticular new bone formation (p=0.044). Pts having osteolysis developed asymmetrical syndesmophytes 10 times more often than pts without osteolysis (OR=10; 95% CI: 1.63–61.327). HLA-B27 antigen was positive in 28 (30.1%) cases.

Conclusion: axial PsA is often asymptomatic, one third of pts develop back pain at age above 40. HLA-B27 positivity was found in only 30% of pts. Pts with rSI are more likely to have severe peripheral arthritis with multiple joint erosions. RSI is associated with juxtaarticular new bone formation. Syndesmophytes may occur in absence of sacroiliitis (in 20% of pts). Cervical spine involvement is more frequent in axial PsA pts. Asymmetrical syndesmophytes characteristic of PsA, are associated with typical, for PsA, radiographic changes on hands and feet X-rays, such as osteolysis and juxtaarticular new bone formation.

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Abstract N°: 1915

"The Great Imitator": A Case of Rowell Syndrome in the Setting of Preexisting Systemic Lupus Erythematosus

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Introduction

Rowell syndrome, characterized by the triad of lupus erythematosus, erythema multiforme-like lesions, and positive lupus immunology, remains a diagnostic challenge due to its rarity and varied clinical presentation. Named after Dr. Barbara M. Rowell who first described it in 1963, it remains a diagnostic challenge and underscores the importance of heightened awareness among dermatologists and rheumatologists. The hallmark features of Rowell Syndrome include cutaneous manifestations reminiscent of both lupus erythematosus and EM, leading to frequent misdiagnoses or delays in proper identification. Characterized by the abrupt onset of erythematous, targetoid lesions, often accompanied by mucosal involvement, it can mimic a spectrum of dermatologic disorders, necessitating a comprehensive diagnostic approach. Despite advances in understanding autoimmune diseases, the precise etiology of Rowell Syndrome remains elusive, highlighting the need for further research elucidating its immunopathogenic mechanisms. Treatment strategies typically include systemic corticosteroids, immunosuppressive agents, and adjunctive therapies targeting specific autoimmune pathways. However, optimal therapeutic regimens remain speculative, necessitating individualized management guided by disease severity and patient response. Here, we present a case of Rowell syndrome occurring in a patient with preexisting systemic lupus erythematosus (SLE), highlighting the complexities in diagnosis and management when two autoimmune conditions converge.

Results

A 70-year-old female with a 5-year history of SLE and Sjogren syndrome presented with a 2-week history of pruritic and painful skin rash. Despite being on long-term hydroxychloroquine and corticosteroid therapy for SLE management, her symptoms persisted and worsened over time. On examination, she exhibited targetoid lesions on the limbs and trunk, oral and eye mucosa involvement, accompanied by systemic symptoms including fever and malaise. Laboratory investigations revealed leukopenia, anemia, and elevated inflammatory markers. Autoantibody testing showed positive antinuclear antibodies, anti-Ro and anti-La antibodies. Given the characteristic cutaneous and systemic manifestations, a differential diagnosis including Lyell and Rowell syndrome superimposed on preexisting SLE was suspected. Hydroxychloroquine was discontinued, and systemic corticosteroids were initiated along with supportive therapy for skin lesions and ocular involvement. Improvement in skin lesions and systemic symptoms was noted within days of treatment initiation. Skin biopsies confirmed the diagnosis of Rowell syndrome, further supporting the clinical impression. The patient was discharged and referred for further treatment of SLE with belimumab.

Conclusion

This case underscores the importance of considering Rowell syndrome in the differential diagnosis of cutaneous manifestations in patients with preexisting autoimmune conditions like SLE. Clinicians should maintain a high index of suspicion for Rowell syndrome in patients with SLE presenting with atypical cutaneous lesions, even in the absence of recent changes in therapy. Through diligent clinical evaluation and multidisciplinary collaboration,

timely recognition and management of Rowell syndrome can be achieved, leading to improved outcomes and quality of life for affected individuals.

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Abstract N°: 1940

Ulcerated papuloplaques in middle-aged women, an exclusion diagnosis

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Introduction

Recurrent cutaneous eosinophilic vasculitis (RECV) is a rare necrotizing vasculitis described in 1994 (Chen et al), with less than twenty cases reported in the literature.

Case Report:

A 46-year-old female patient with no previous medical history was referred to our outpatient dermatological clinic for violaceous papules-plaques, some of them ulcerated, on the lower limbs and trunk of two months of evolution. They resolved leaving an hyperpigmented scar. No systemic symptoms were referred. Complementary tests including bacterial and mycobacterial culture of the lesions, and leishmania Polymerase Chain Reaction (PCR) ruled out infectious pathology. Blood tests including hemogram, ions, liver and renal function, autoimmunity, and serologies for hepatotropic viruses, HIV and syphilis did not show significant results. Histologic study revealed epidermal ulceration, intra- and subepithelial edema with microvesicle formation, perivascular and interstitial lymphohistiocytic infiltrate with many eosinophils, and signs of lymphocytic vasculitis. Clonal rearrangement study ruled out lymphomatoid papulosis. Given the clinical and histological findings, the diagnosis of recurrent cutaneous eosinophilic vasculitis (RCEV) was stablished. Treatment with dapsone 50mg/day was initiated, with resolution of lesions at 3-month follow-up.

Discussion: **

RCEV is an uncommon entity characterized by recurrent outbreaks of violaceous papules-plaques with ulceration tendency, which predominantly affects lower limbs in middle-aged or elderly Asian women. Systemic involvement is infrequent, although angioedema or mild general symptoms can be present. Laboratory common alterations include peripheral blood eosinophilia, leukocytosis, elevated acute phase reactants and IgE. Histology is characterized by a perivascular dermal infiltrate rich in eosinophils and small vessel necrotizing vasculitis, with no evidence of leukocytoclasia. Quijano et al proposed diagnosis criteria, although exclusion of other diseases is crucial for its diagnosis. Differential diagnosis includes different entities: those with an special role of eosinophils (urticarial vasculitis, Wells syndrome, eosinophilic granulomatosis with polyangiitis, hyperosinophilic syndrome), cutaneous leishmaniasis, lymphangitis nodularis or lymphomatoid papulosis . Complementary tests and distinctive clinical findings are essential in the distinction. First line treatment consists on oral corticosteroids at medium dose with gradual tapering, with frequent recurrences after discontinuation. Other therapeutic lines include dapsone, oral tacrolimus, indomethacin or mepolizumab.

Conclusion:

CRPV is characterized by recurrent outbreaks of typical lesions with no systemic involvement. Its peculiar histological findings (with eosinophils as the main cellularity), a rapid response to corticosteroids and the absence of systemic involvement are keys to its diagnosis.

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Abstract N°: 1948

Panniculitis as the predominant cutaneous finding in a patient with dermatomyositis

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Introduction & Objectives:

Materials & Methods:

Results:

A 58-year-old woman was referred to our Dermatology clinic for pruritic disseminated skin lesions with 18 months of evolution. The patient reported erythematous cutaneous lesions on the extensor surfaces of both arms, thighs and buttocks, that progressively became firmer. In the last month, there was a sudden spread of lesions to the face, neck and chest. Concurrently, she experienced asthenia, anorexia and myalgia. Upon observation, erythematous-violaceous plaques were symmetrically disseminated to the mandibular region, neck, chest and arms. In the sacral region, buttocks and thighs, these plaques were annular, with a central brownish area and an erythematous-violaceous border, displaying a stony consistency and marked areas of lipoatrophy. The proximal muscular strength of the upper and lower limbs was markedly reduced. A skin biopsy revealed histopathological findings consistent with interface dermatitis, and, in depth, a lymphoplasmacytic infiltrate with extensive lobular hyalinization and necrosis, indicating panniculitis associated with dermatomyositis. Analytical study revealed a significant increase in muscle and liver enzymes, antinuclear antibodies at a titer of 1: 640, and a positivity for the anti-Mi-2 antibody. The electromyogram supported the diagnosis of dermatomyositis. Subsequent complementary studies yielded negative results. The patient was advised to strictly adhere to photoprotection measures and was medicated with prednisolone 1mg/kg/day, hydroxychloroquine 400mg/day, methotrexate 15mg/week and high-potency topical corticosteroids. After three months, we verified a substantial improvement in muscle strength and an excellent cutaneous response.

Conclusion:

Dermatomyositis is an idiopathic inflammatory myopathy primarily affecting skeletal muscle and skin. Panniculitis is a rare manifestation of dermatomyositis, with less than 40 cases described in the literature. It most often affects adult women, presenting with hardened plaques that typically affect the upper and lower limbs, which may lead to secondary lipoatrophy. Generally, patients exhibit high levels of muscle and liver enzymes, with a variable autoantibody profile. Histopathology reveals a predominantly lobular lymphoplasmacytic panniculitis, and, in advanced stages, lobular hyalinization and necrosis. Typically, these patients present less aggressive forms of dermatomyositis, with a good response to immunosuppressive therapy and a favorable prognosis.

The authors present an atypical case of dermatomyositis with panniculitis as the predominant finding, highlighting the importance of a thorough clinical history and observation for recognition of less common findings of this entity.

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Abstract N°: 1962

Immunobullous diseases among the geriatric population in Lagos Nigeria: Diagnostic challenges and management.

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Introduction & Objectives: Studies from Nigeria revealed that a low percentage (2%) of geriatric individuals have immunobullous diseases probably on account of a low index of suspicion, and lack of access to quality health care. Geriatric individuals are prone to immunobullous diseases that may be linked to high morbidity and mortality, thus prompt diagnosis is required for specific drug treatment. Although immunofluorescence is the gold standard, it is expensive, requires a medium, and is done in a sophisticated laboratory. Histopathological diagnosis can serve as a useful substitute. The objective is to document the disease patterns and the frequency of occurrence among the geriatric population.

Materials & Methods: A retrospective study of all patients aged 65 and above seen at Lagos University Teaching Hospital, Lagos Nigeria between January 2020 and July 2023. Case notes of all the patients were retrieved. Data was obtained and analyzed using SPSS version 25.

Results: A total of 236 individuals were seen during the study period;110 males and 126 females, with a ratio of 1.1:1, the age range was 65-95 years, median age of 80. The five most common skin disorders were papulosquamous disorders in 62 patients (26%), Infections in 48 patients (20.3%), Pigmentary disorders in 32 patients (13.6%), Xerosis in 20 patients (8.5%), and Scar in 15 patients (6.4%). Immunobullous diseases were noted in 14 patients (5.9%); 6 males and 8 females, the age range was 65 years to 86 years, median age of 76 years, and bullous pemphigoid was the most common seen in 9 patients (3.8%). Patients were diagnosed with skin biopsy and histology due to the unavailability of immunofluorescence in our center, placed on corticosteroids and immunosuppressive agents, and comorbidities were also managed. Patients showed remarkable improvement after management and were discharged home with subsequent follow-up in the clinic.

Conclusion: Immunobullous diseases can be diagnosed clinically with a high index of suspicion with the aid of skin biopsy and histology, and can be managed successfully even in resource-poor countries where immunofluorescence facilities are lacking. Hence, improve the quality of life of the geriatric population.

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Abstract N°: 2173

Infantile Bullous Pemphigoid: A Case Report

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Abstract:

Background:

Infantile bullous pemphigoid (IBP) is an exceptionally rare acquired autoimmune subepidermal bullous disorder characterized by vesicles, bullae, and additional manifestations such as urticarial and infiltrated papules, plaques, or eczematous lesions. These skin lesions can lead to eroded and crusted regions upon healing, and in some cases, rapid blister rupturing causes extensively eroded areas. Reporting these rare cases is essential to improve our understanding, diagnosis, and treatment strategies for IBP.

Case Presentation:

We present the case of a 4-month-old Saudi male infant who presented with generalized multiple tense bullae on an erythematous base, initially appearing on the feet and soles and progressing to involve the trunk and extremities over 2 weeks. The bullae caused discomfort, irritability, and difficulty sleeping. Perinatal history was unremarkable, with regular antenatal care and exclusive breastfeeding. Clinical examination revealed tense bullae, crusts, and erythematous urticarial plaques without mucosal involvement. Laboratory findings showed leukocytosis, eosinophilia, and thrombocytosis. Histopathological examination revealed subepidermal bullae with inflammatory infiltrate and linear deposition of complement factor C3 and immunoglobulin G (IgG) along the dermo-epidermal junction. Diagnosis of infantile bullous pemphigoid (IBP) was confirmed. Treatment with oral prednisolone and later addition of topical steroids and dapsone led to resolution of lesions over 1 year, with no recurrence thereafter.

Conclusion:

This case highlights the importance of early recognition and appropriate management of IBP. Our comprehensive evaluation, which includes pathological confirmation, and therapeutic interventions, enhances our understanding of IBP. The case underscores the necessity for prompt diagnosis and individualized treatment strategies in affected infants.

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Abstract N°: 2265

Comparative Study Assessing Multiple Switches Between Biosimilar ABP 501 and Adalimumab Reference Product in Patients with Plaque Psoriasis

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Introduction & Objectives:

ABP 501 is an approved biosimilar to adalimumab reference product (RP) in the US and EU. ABP 501 is used to treat chronic inflammatory conditions that include rheumatoid arthritis, juvenile idiopathic arthritis, psoriatic arthritis, ankylosing spondylitis, Crohn's disease, ulcerative colitis, plaque psoriasis (Ps), hidradenitis suppurativa, and uveitis. This study aims to demonstrate that switching between adalimumab RP and ABP 501 does not result in differences in pharmacokinetic (PK), efficacy, safety, and immunogenicity, when compared to continued use of adalimumab RP.

Materials & Methods:

This switching study was carried out with adults with moderate to severe Ps, see Figure 1. The study had 2 periods: a lead-in period where patients received adalimumab RP 100 mg/mL open-label, followed by a randomized, double-blind 2-parallel arm period. In the 2-parallel arm period, patients were randomized to either the continued use group or the switching group. The continued use group continued on adalimumab RP 100 mg/mL every 2 weeks (Q2W) through end of study (EOS). The switching group switched to ABP 501 100 mg/mL Q2W at week 12, back to adalimumab RP 100 mg/mL Q2W at week 16, and again to ABP 501 100 mg/mL Q2W at week 20 through EOS. The primary endpoint was a demonstration of PK similarity based on area under the serum concentration-time curve from time 0 over the dosing interval (AUCtau) and the maximum observed serum concentration (Cmax) between week 28 and week 30. Secondary endpoints included psoriasis area and severity index (PASI) percent improvement from baseline and PASI 75, 90 and 100 scores, immunogenicity, and safety.

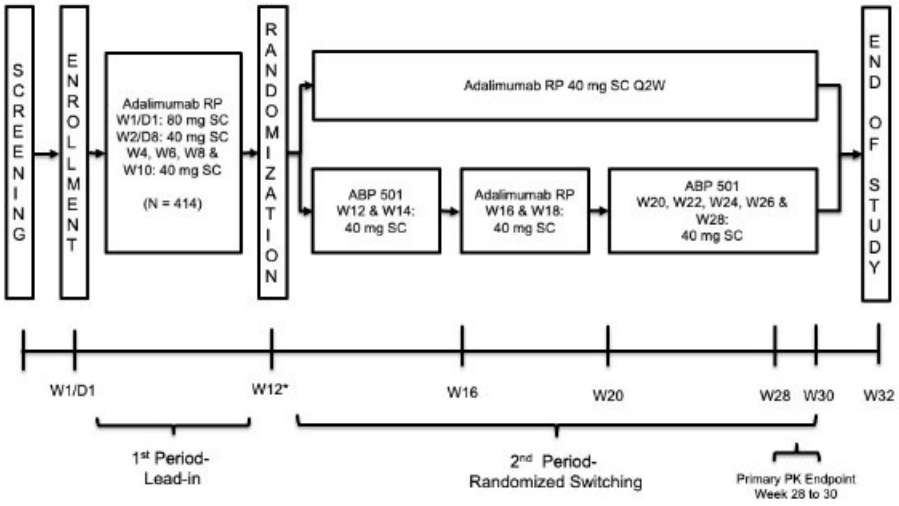
Results:

A total of 425 patients were enrolled at 83 centers across 6 countries. Overall, 347 (91.3%) of the 380 randomized patients completed the study and 33 (8.7%) patients discontinued the study, mainly due to "consent withdrawn." Occurance of investigational product dosing, study completion/discontinuation, demographic, baseline physical, and baseline disease characteristics were similar between the 2 treatment groups. The ratio of geometric least squares means (90% CI) between the switching group and the continued use group for AUCtau was 1.0516 (0.9010, 1.2273) and for Cmax was 1.0044 (0.8717, 1.1574); both were within the prespecified similarity margin of 0.8 to 1.25 (Table 1). Secondary outcomes, including PASI percent improvement, PASI scores, and immunogenicity were overall similar between the 2 groups. Post randomization, the frequency, type, and severity of adverse events, serious adverse events, and events of interest were also similar between the 2 groups.

Conclusion:

Results of this study demonstrated similarity for PK, efficacy, immunogenicity, and safety in patients with Ps switching 3 times between adalimumab RP and ABP 501 as compared to patients receiving continued use of adalimumab RP. These results support a demonstration of interchangeability between ABP 501 and adalimumab RP.

Figure 1. Study Design



*Subjects with less than PASI 50 response at W12 will be discontinued.

Table 1: Assessment of PK Parameters between Week 28 and Week 30 – Post-randomization Period (PK Parameter Analysis Set)

Treatment and Comparison	AUC_{tau} (hr*µg/mL)	C_{max} (µg/mL)
	Geometric LS Mean [n]	Geometric LS Mean [n]
Switching Group	1616.68 [146]	5.49 [153]
Continued Use Group	1537.42 [145]	5.47 [153]
Ratio of Geometric LS Means (90% CI)		
Switching Group vs Continued Use Group	1.0516 (0.9010, 1.2273)	1.0044 (0.8717, 1.1574)

AUC_{tau} = area under the curve from time 0 over the dosing interval; C_{max} = maximum concentration; LS = least squares; PK = pharmacokinetic

Note: Geometric LS mean, ratio of geometric LS means, and 90% CI were estimated based on the ANCOVA model adjusted for baseline weight, PK trough concentration at the end of the Lead-in Period (week 12), and actual stratification factors of prior biologic use for psoriasis and geographic region.

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Abstract N°: 2315

Nail unit involvement in pemphigus vulgaris: a manifestation to be considered

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Introduction & Objectives: Background: Involvement of the nail unit in patients with pemphigus vulgaris (PV) appears to be uncommon, with great variability among case series. Moreover, the scarce evidence regarding this manifestation of PV leads to a lack of specific treatment approaches. The aim of this study was to report 2 new cases of pemphigus vulgaris with nail involvement, as well as to describe the clinical response to treatment.

Materials & Methods: Case report study of 2 cases of patients with PV with nail unit involvement. Clinical and sociodemographic characteristics were collected, with a 2-year follow-up after treatment.

Results: We present the case of a 16-year-old male and a 40-year-old female with PV. Both presented cutaneous and mucosal erosions in the form of flaccid blisters, as well as lesions on the fingers of both hands, in the form of paronychia and detachment of the nail plate (onychomadesis, onycholysis). In both cases, nail involvement was concurrent with the oral lesions and resolved without sequelae after treatment with systemic corticosteroids and Rituximab.

Conclusion: Nail involvement in patients with pemphigus vulgaris is a poorly understood entity that could be useful in predicting oral mucosal involvement in these patients. They seem to appear at any time during the clinical course and usually have a good response to systemic treatment, including corticosteroids and Rituximab.

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Abstract N°: 2316

Experience in the use of Rituximab for the treatment of pemphigus vulgaris: case series

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Introduction & Objectives: Pemphigus is a cluster of autoimmune blistering diseases affecting the outermost layers of the skin and mucous membranes. Accurate diagnosis and therapeutic management is often complex, with the consequent impact on the quality of life of these patients. The aim of this study was to evaluate the effectiveness of Rituximab (RTX) in the treatment of patients with pemphigus, as well as to identify possible predictors of good response and the occurrence of adverse events.

Materials & Methods: case series of patients with blistering diseases treated with RTX. Sociodemographic characteristics, previous treatments and response to treatment were evaluated with a subsequent follow-up up to 3 years.

Results: 12 patients with pemphigus vulgaris were included. Women accounted for 66.7% and the mean age was of 52.3 (SD 17.3) years. The mean time to initiation of RTX was 11.25 (SD 14.6) months, which was effective in more than half of the cases. All patients had received oral corticosteroids previously and together with RTX. The median time to complete response was 4 months and only 30% had mild adverse effects that did not require hospital attention. 1 patient relapsed after discontinuation of treatment.

Conclusion: RTX appears to be an effective and safe treatment for most patients with pemphigus vulgaris. Patients usually respond within 4 months of the first administration of the drug. This reduces the adverse effects associated with oral corticosteroids and rapidly improves the quality of life of these patients.

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Abstract N°: 2353

Severe and recalcitrant bullous pemphigoid in an infant successfully treated with intravenous human immunoglobulin

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Introduction & Objectives:

Bullous pemphigoid (BP) is an autoimmune dermatosis presenting with subepidermal blistering, urticarial plaques, eosinophilia, and pruritus. BP usually develops in the elderly and is rare in infants. We report a severe BP case in an infant with partial response with systemic corticosteroids who achieved complete remission after intravenous immunoglobulin (IVIg) therapy.

Materials & Methods:

We reviewed the medical records and laboratory results of a BP patient under follow-up since February 2024 at the Autoimmune Blistering Disease Clinic in a tertiary hospital.

Results:

A 6-month-old male patient presented with a 2-month history of diffuse tense blisters and urticarial plaques on the trunk and face, with vesicles on the hands and feet and pruritus. Due to the cutaneous lesions, he had been hospitalized in three different occasions and was receiving prednisolone 3 mg/kg/day for 2 months without improvement. We considered the hypothesis of BP and performed a biopsy on the abdomen. Histopathology revealed a subepidermal blister with superficial perivascular lymphohistiocytic and eosinophilic infiltrate. Direct immunofluorescence (IF) from perilesional skin showed linear C3 and IgG deposition at the basement membrane zone (BMZ), thus confirming BP. Additional evaluation revealed eosinophilia of 11,538/mm3 (20-700) and elevated IgE levels of 1,629 units/mL (<15). A normal echocardiogram and abdominal ultrasound ruled out eosinophilic infiltration. As BP remained active despite high corticosteroid dose, prednisolone was replaced with betamethasone (equivalent dosage of prednisolone 2 mg/kg/day). Despite absence of new blisters after three weeks, urticarial plaques and eosinophilia worsened after meningococcal and pentavalent vaccines and furunculosis. We started a 5-day course of IVIg 400 mg/kg/day, after considering the partial response to systemic corticosteroid, a previous history of blood transfusion due to dapsone-induced hemolytic anemia and a recent pyodermitis. Complete remission and decrease of eosinophil count (1,600/mm3) were achieved after 3 days of IVIg therapy, thus enabling betamethasone tapering (equivalent dosage of prednisolone 1,3 mg/kg/day) with no recurrence.

Conclusion:

BP occurs mainly in the elderly, with the second peak of incidence during childhood. It has been speculated the correlation between BP onset and vaccination without a definitive conclusion. BP pathogenesis is similar in both age groups and involves the production of antibodies against BP180 and BP230, which triggers subepidermal blistering with eosinophils. Diagnostic confirmation requires the demonstration of C3 and/or IgG deposits at the BMZ with direct IF or circulating IgG antibodies binding to the epidermal side using indirect IF with the salt-split skin technique. Case reports of BP in infants generally describe successful response to systemic corticosteroid with sustained remission. Adjuvant therapy with dapsone, mycophenolate mofetil, IVIg and/or rituximab has rarely

been reported. In our case, as conventional treatment with high-dose corticosteroid failed to control BP lesions and eosinophilia, and dapsone was contraindicated due to previous hemolytic anemia, IVIg therapy was introduced with a remarkable improvement. We highlight that although BP in infants usually has a good prognosis, exceptional severe and recalcitrant cases may require IVIg treatment to achieve systemic and cutaneous disease control.

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Abstract N°: 2448

Design of a phase 2a, double-blind, placebo-controlled, global trial of MK-6194, a modified form of IL-2, in participants with non-segmental vitiligo.

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Introduction & Objectives:

Current systemic treatments for vitiligo offer limited efficacy, have potential safety concerns with extended use and can be burdensome or undesirable in application. In vitro and in vivo data suggests regulatory T cells (Tregs) are involved in the pathogenesis of Vitiligo. Treg expansion using low dose IL-2 has shown preliminary evidence of clinical efficacy in a variety of autoimmune diseases. However, its use is limited by a narrow therapeutic window of dose for minimizing activation of the cytotoxic immune cells and frequent injections due to its short half-life.

MK-6194 is a modified form of IL-2 that selectively expands Tregs without significant effects on other immune cell types. MK-6194 administered subcutaneously is generally well tolerated in the completed and ongoing Phase 1 clinical studies at single doses up to 10 mg. In healthy adults, the half-life of MK-6194 is ~20–28 hours after single-dose administration, with dose-related increases in total Tregs.

This study will evaluate the efficacy and safety of MK-6194 at two dose regimens in patients with nonsegmental vitiligo (NCT06113328).

Materials & Methods:

This phase 2a, multicentre, randomized, double-blind, placebo-controlled trial is enrolling adults (aged 18-75 years) with a clinical diagnosis of non-segmental vitiligo of ≥6 months (**Figure 1**). Eligible participants (**Table 1**) will be randomized to receive subcutaneous administration of either placebo or one of the dose regimens of MK-6194 for 24 weeks (double-blind placebo controlled treatment period) and followed-up in a further 24 weeks double-blinded extension. In the double-blinded extension period, participants will continue their original treatment regimen if treated with MK6194 in the double-blind treatment period and participants will be randomized to receive one of the two MK-6194 regimens if previously treated with placebo.

The study will also be stratified by history of previous JAKi use (yes vs no) and stable vs active non-segmental vitiligo at randomization. The primary efficacy endpoint is percent change from baseline in Facial Vitiligo Area Scoring Index (F-VASI) and the primary safety endpoint is number of adverse events (AE) and discontinuation due to AEs at Week 24. Percentage change from baseline in total Vitiligo Area Scoring Index (T-VASI, including the face) will be assessed as a secondary endpoint at Week 24, along with other exploratory endpoints, including patient-reported outcomes, pharmacokinetics and pharmacodynamics.

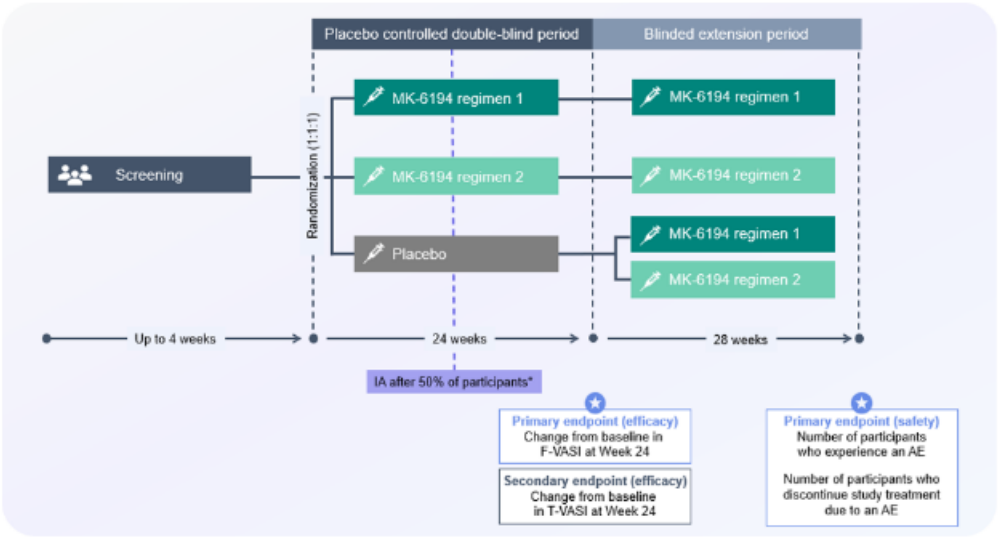
Results:

Approximately 165 participants (55 per treatment group) will be randomized in Europe, USA, Canada and the Asia Pacific region. The study is actively recruiting; no data are yet available.

Conclusion:

This phase 2a trial will outline the efficacy, safety, and tolerability of MK-6194 compared with placebo in patients with non-segmental vitiligo.

Figure 1. Study design and key endpoints



*An interim futility analysis will be performed when the first 50% randomized participants complete the Week 24 evaluation or prematurely discontinue.
AE, adverse event; F-VASI, Facial Vitiligo Area Scoring Index; IA, interim analysis; T-VASI, Total Vitiligo Area Scoring Index (including the face).

Table 1. Inclusion/exclusion criteria

Key inclusion criteria	Key exclusion criteria

• Adults (18–75 years) of any sex/gender
• Clinical diagnosis of non-segmental vitiligo for a duration of ≥6 months*
• At screening and baseline:
  • Depigmentation contributing to F-VASI ≥0.3
  • Depigmented facial BSA ≥0.3%
  • T-VASI ≥4
  • Total body vitiligo area ≥4%†
  • Candidate for systemic therapy based on investigator judgment
	• Segmental vitiligo or ≥50% leukotrichia on face or body
	• History of (or current) inflammatory condition that could interfere with vitiligo evaluation‡
	• Known systemic hypersensitivity to IL-2, or modified IL-2 including MK-6194, or its inactive ingredients
	• Known history of hypereosinophilic syndrome or an eosinophil-related condition§
	• Active clinically significant infection,¶ any infection requiring treatment with IV anti-infectives ≤4 weeks prior to randomization, or oral/intramuscular anti-infectives ≤2 weeks prior to randomization
	• Symptomatic heart failure (NYHA class III or IV) or MI or unstable angina pectoris within 6 months prior to screening
	• Severe chronic pulmonary disease requiring oxygen therapy
	• Has a transplanted organ which requires continued immunosuppression, had major surgery within 3 months prior to screening or has a major surgery planned during the study
	• History of any malignancy, except for successfully treated non-melanoma skin cancer or localized carcinoma *in situ* of the cervix

For full details on the inclusion and exclusion criteria please refer to NCT06113328.

*Vitiligo diagnosis must be made by a trained physician who is a board-certified dermatologist (or local equivalent). Disease duration is defined as the length of time since onset of symptoms; †Excluding hand and foot involvement; ‡In the opinion of the investigator; §For example, eosinophilic pulmonary disease including eosinophilic asthma, eosinophilic esophagitis, eosinophilic nephritis, eosinophilic granulomatosis with polyangiitis; ¶Including COVID-19, HBV, HCV, HIV and TB.

BSA, body surface area; COVID-19, coronavirus disease 19; F-VASI, Facial Vitiligo Area Scoring Index; HBV, hepatitis B virus; HCV, hepatitis C virus; HIV, human immunodeficiency virus; IL, interleukin; IV, intravenous; MI, myocardial infarction; NYHA, New York Heart Association; TB, tuberculosis; T-VASI, Total Vitiligo Area Scoring Index (including the face).

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Abstract N°: 2496

Bullous Pemphigoid induced by local radiotherapy or by immunotherapy?

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Introduction & Objectives:

Bullous Pemphigoid (BP) is the most prevalent autoimmune blistering disease, characterized by circulating and tissue-bound autoantibodies (BP180/BP230) targeting the dermal-epidermal junction (DEJ). While typically idiopathic, BP has been increasingly associated with immune checkpoint blockade (ICB) in oncologic patients as well as with local radiotherapy (RT).

Materials & Methods:

A 51-year-old patient with known end-stage renal failure undergoing dialysis and diagnosed with breast cancer T4N1M0, triple-negative, non-mutated, underwent mastectomy (left breast) and axillary lymph node dissection. Subsequently, the patient received 6 injections of pembrolizumab in conjunction with local radiotherapy. She was addressed to the dermatological department with a polymorphic, pruritic eruption consisting of multiple tense bullae and erosions localized around the mastectomy scar, appearing 5 days after the final radiotherapy session.

Radiation-induced skin reactions (RISRs), infection, and PB were retained as potential differential diagnoses. Histological examination revealed junctional blistering disease with a superficial lymphocytic infiltrate and occasional eosinophils, suggestive of PB, along with possible acute radiation-induced skin reactions showing minimal features of discrete vacuolar interface dermatitis, without apoptotic changes. Direct immunofluorescence microscopy (DIF) demonstrated linear deposits of IgG and C3 along the DEJ. Positive enzyme-linked immunosorbent assay (ELISA) for antibodies antiMB and antiPB180 confirmed the diagnosis of BP.

Treatment with a topical high-potency corticosteroid (clobetasol propionate 0.05%), initially resulted in local regression of the lesions. However, 10 days post-treatment initiation, the eruption disseminated to the trunk, abdomen, upper limbs, and thighs, characterized by well-delimited vesicles, small bullae, and multiple erosions on unaffected skin, without mucosal involvement. Immunotherapy was discontinued, and topical treatment was escalated to three tubes of clobetasol propionate per day, gradually tapered over three months, achieving disease control.

Results:

While most bullous pemphigoid cases are due to autoantibodies against proteins arranged at the dermal-epidermal junction, some cases of bullous pemphigoid are caused by systemic medications. Several drugs, including PD-1/PD-L1 inhibitors, have been reported in the literature to cause BP. Drug-induced BP typically manifests within three months of medication initiation, often in younger patients. Additionally, several case reports suggest a potential role of radiotherapy as a trigger for BP. Literature review suggests that while BP lesions may initially appear at irradiated sites, they often extend beyond these areas, potentially implicating radiotherapy in disease pathogenesis. The etiology of BP in this case remains debated, with triggers including immunotherapy and local radiotherapy.

Conclusion:

Clinicians encountering pruritic skin lesions or blistering in patients receiving immunotherapy and radiotherapy should consider skin biopsy for histopathology and direct immunofluorescence. Treatment for radiotherapy-induced BP or ICB-induced BP parallels that one for idiopathic BP, involving topical and systemic corticosteroids and immunosuppressive agents depending on disease severity.

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Abstract N°: 2508

Case Report: Evaluating a Possible SITRAME Diagnosis in a Patient with Recurrent Sharply Demarcated Erythematous Patches

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Introduction & Objectives: Undifferentiated systemic autoinflammatory disorders (SAIDs) are associated with innate immunity and do not align with any established autoinflammatory diseases, nor do they have confirmed molecular diagnoses (1,2). These disorders are characterized by recurrent, generalized inflammation that manifests without infection or autoimmune disease, with skin manifestations often observed as the primary clinical feature (3). A recent report suggested a new clinical entity represented by 16 adult patients, characterized by systematic inflammatory responses and a stereotyped pattern of trunk macular eruptions recurring with consistent topography. This pattern displays criteria suggestive of an undetermined SAID and has led to the proposed acronym SITRAME, standing for Systemic Inflammatory Trunk Recurrent Acute Macular Eruption (4). This report presents a male patient with clinical presentations and findings closely resembling those described in SITRAME, with additional prurigo, potentially shedding extra light on this proposed clinical entity.

Materials & Methods: An otherwise healthy 54-year-old male presented to our department with a two-decade history of recurrent sharply demarcated erythematous patches. These patches initially affected only the trunk and upper arms on areas of skin unexposed to the sun. Over the years, the eruptions progressively extended to cover the entire body, except for the face, palms, and soles. The affected skin areas became pruritic a few days following each eruption. No desquamation was observed post-resolution. The initial outbreak occurred in his early thirties, with episodes initially lasting approximately 2-3 days, but extending to about a week in recent years. He reported over 50 similar episodes, typically after high fever or extreme physical exertion (e.g., running a marathon), but not after regular physical activities. Concurrent with these skin eruptions, he experienced significant fatigue. Multiple antihistamine treatments failed to alleviate his symptoms. He takes no regular medications, and his family history was non-contributory. Diagnostic tests for physical triggers of urticaria and cholinergic urticaria were negative.

Results: Our patient's clinical presentation aligned with a stereotyped, exact topography, as determined from iterative photographs, consistent with the description of SITRAME. The diagnosis was not solely based on photographic evidence but also on the behaviour of the lesions, fitting the descriptions provided by the original authors. Interestingly, our patient developed prurigo a few days post-eruption, a symptom absent in the original cohort of 16 patients. Furthermore, the eruptions have notably expanded in recent years to cover most of the body, sparing only the face, palms, and soles, indicating a potentially unreported progression pattern of the disease.

Conclusion: In conclusion, we have documented a potential case of a patient with SITRAME, detailing the disease's progression over two decades and presenting previously unreported prurigo. Iterative photographs, coupled with a detailed clinical description, significantly aided in diagnosing this patient. Consistent with the views of the disease's initial proposers, we agree that further research is necessary to determine the aetiology and pathophysiology of SITRAME and thus establishing the most effective treatment protocols.

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Abstract N°: 2523

Bilateral Axillary Deformity Caused by Dystrophic Calcification Followed by a Surprising Presentation: A Case of Dermatomyositis in an Adult

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Introduction & Objectives: Dermatomyositis (DM) is a common autoimmune inflammatory connective tissue disease that occurs in two age groups, juvenile and adult types. It manifests with proximal muscle involvement. Particularly in sun-exposed areas, such as the scalp, periocular areas, and extensor surfaces, it is characterized by characteristic pink-purple poikiloderma, prominent nail bed changes, and pink-purple papules on the finger joints. Calcinosis cutis was described as an abnormal deposition of calcium phosphate. The etiology of calcium deposition can be divided into four subtypes: dystrophic, metastatic, idiopathic, and iatrogenic. The type of calcinosis in dermatomyositis is dystrophic in nature. The extremities and pressure areas are the most commonly affected .

We intend to report an unusual case of bilateral axillary deformity with dystrophic calcification in adult dermatomyositis.

Materials & Methods: : A 43-year-old female patient had been experiencing increasing difficulty in movement due to hardness on both axillae for the past 3 years. Therefore, an incisional biopsy was performed, revealing widespread calcifications and collagen degeneration consistent with autoimmune connective tissue diseases. The patient was advised to undergo further investigation for autoimmune connective tissue diseases. However, she did’t attend follow-up appointments. She presented with weakness and episodes of falls in the legs, along with redness and swelling on the face and arms, and erythematous nodules on the fingers for the past year. Severe calcified plaques causing significant sequelae in both axillae had been progressing. The patient’s muscle biopsy revealed findings consistent with myositis and dermatomyositis both clinically and pathologically. The degree of calcification and sclerosis in the axillae had increased. ANA was positive at a titer of 1:3, and creatine kinase levels were elevated. Along with rheumatology, systemic methotrexate 15 mg/week and deltacortril 8 mg/day were initiated. Physical therapy and sodium thiosulfate were started for calcifications.

Results: Dermatomyositis is a idiopathic chronic acquired autoimmune inflammatory myopathy with skin manifestations that vary in severity. In childhood dermatomyositis, the incidence of calcinosis, ulceration, and vasculopathy is higher. Dystrophic calcinosis is observed in 45-75% of juvenile cases while this rate is limited to 10-20% in adult-onset DM cases. Calcinosis appears within a period of 4 months to 12 years from the onset of the disease. Calsinosis is more common in cases with severe skin involvement. Surgical treatment is the primary approach in extensive lesions of dystrophic calcinosis, while local injection of sodium thiosulfate is another treatment option for superficial calcifications in cases with a limited number of lesions.* Sodium thiosulfate acts as a calcium chelating agent by producing calcium thiosulfate, increasing the solubility, and hastening the clearance of the calcium deposit.

Conclusion: The case is considered significant due to dystrophic calcification being a precursor lesion of dermatomyositis, leading to disfigurement, significantly impairing quality of life and accompanying dermatomyositis in adulthood.

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Abstract N°: 2530

Safety, Tolerability, Pharmacokinetics, and Pharmacodynamics of Vibozilimod: A Randomized, Double-blind, First-in-human, Study in Healthy Male and Female Subjects

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Introduction & Objectives:

Vibozilimod (VBZ) is a potent, oral, selective agonist of the sphingosine-1-phosphate receptors (S1PR) 1 and 5, an inverse agonist at S1P4R, and devoid of activity on S1P3R. S1P1 receptor modulators sequester circulating lymphocytes within lymph nodes, causing reduction of pathogenic autoimmune cells in the blood stream and inflamed tissues, making S1P1R agonists an effective therapeutic agent for autoimmune disorders. VBZ is in clinical development for the treatment of chronic immune-mediated, inflammatory disorders.

Materials & Methods:

In this randomized, double-blind, phase 1, single ascending dose study, healthy male and post-menopausal female volunteers received single doses of either VBZ 0.1, 0.3, 1, 2, 4, or 6 mg in males and 2 mg in post-menopausal females, or placebo in a 3:1 ratio. Safety, tolerability, pharmacokinetics (PK), and pharmacodynamics (PD) were evaluated.

Results:

Fifty-six healthy subjects were randomly assigned to VBZ or placebo in seven groups (groups 1 to 6; males: n=48, and group 7; females: n=8). Vibozilimod was well tolerated and adverse events (AE) were mild to moderate. Transient bradycardia was noted between 2 and 4 h post-dose, the maximum drop in heart rate was 17.2 beats/min, which plateaued at 2 mg and higher doses (Table 1). A dose-dependent percent reduction of absolute lymphocyte count (ALC) compared to baseline was observed, with onset at approximately 4 h post-dose, 30.9 ($p<0.05$), 50.0 ($p<0.001$), 43.6 ($p<0.01$), 58.6 ($p<0.001$), 60.5 ($p<0.001$) and 68.1 ($p<0.001$), vs. placebo (PBO) 22.4 and at 24 h with 21.0 (non-significant; ns), 32.1 ($p<0.05$), 36.3 (ns), 51.8 ($p<0.001$), 57.0 ($p<0.001$) and 74.0 ($p<0.001$), vs. PBO 17.7 at doses of 0.3, 1.0 , 2.0, 2.0 (F), 4.0 and 6.0 mg respectively. Nadir ALC observed between 4-12 h and was similar to 4 h effect, whereas at highest dose of 6 mg the percent reduction was 77.0 (<0.001). However, at 96 h the ALC changes were non-significant other than at the 6 mg dose. The ALC effects were rapid and resolved within 96h (Table 2).

Vibozilimod showed dose proportional pharmacokinetic parameters of maximum concentration (Cmax) and area under the curve (AUC). The mean time to maximum concentration (tmax) was between 2 h and 2.5 h, and the median t1/2 was between 33 h and 37 h. Exposure was higher in females than in male subjects. The Vd/F was in the range of 232 L to 299 L in males and 165 L in females, however with comparable t1/2 in both (Table 2).

Conclusion:

Vibozilimod was well-tolerated with only mild to moderate adverse events (AEs) up to a dose of 6 mg. Transient bradycardia, a known class effect, was observed with a mean heart rate reduction of less than 10 beats/min up to a dose of 0.3 mg. Bradycardia effects plateaued at doses of 2 mg and above. Dose dependent reductions in ALC were significant at doses of 0.3 mg and above. Demonstrated a dose proportional and favorable PK profile. Vibozilimod is a potent sphingosine agonist with potential promise for the treatment of immune disorders such as inflammatory bowel disease, multiple sclerosis, lupus, psoriasis, atopic dermatitis, and alopecia areata.

Table 1: Summary of Mean Change in Heart Rate (bpm) from Baseline (Δ) by dose level in phase 1 Single Ascending Dose Study of Vibozilimod

Time (h)	0.1 mg	0.3 mg	1 mg	2 mg (M)	2mg (F)	4 mg	6 mg	Placebo
N	6	6	6	6	6	6	6	14
BL	56.8 ± 7.4	55.0 ± 6.1	60.8 ± 11.7	61.5 ± 7.5	61.2 ± 6.3	59.5 ± 5.6	56.2 ± 2.1	60.0 ± 10.8
1	1.7 (2.6)	0.7 (2.8)	-4.7 (4.4)	-6.3 (6.5)	-9.7 (4.3)	-8.3 (8.2)	-8.3 (4.9)	-1.1 (3.4)
2	-2.2 (3.1)	-4.7 (4.7)	-13.2*** (4.4)	-16.8*** (6.3)	-15.0*** (3.5)	-17.2*** (5.9)	-16.5*** (3.9)	0.2 (5.1)
3	-1.3 (4.1)	-6.0 (5.3)	-15.3*** (6.0)	-16.3*** (7.1)	-13.7*** (5.1)	-15.0*** (5.4)	-15.5*** (3.8)	-0.4 (4.0)
4	-0.8 (3.1)	-6.0 (6.0)	-14.3*** (6.2)	-13.8*** (7.4)	-11.0* (4.6)	-13.2*** (8.6)	-14.3*** (3.1)	0.6 (4.7)
5	5.7 (3.2)	5.0 (5.3)	-4.2 (5.3)	-6.5 (6.1)	-4.7 (4.5)	1.8 (8.6)	-1.5 (5.0)	15.9*** (9.3)

Values are expressed as mean (±SD); BL – Baseline; bpm-beats per minute; F-Female
For statistical analysis of heart rate, entire data from pre-dose to EOS (Day 14) h was used; only data at BL, 1h, 2h, 3h, 4h, and 5h, represented in the table.
*p<0.05, **p<0.01, ***p<0.001 vs respective group BL heart rate. Two-way ANOVA followed by Bonferroni's test

Table 2: Summary of Mean Change in Absolute Lymphocyte Count (ALC) from Baseline (Δ) and PK parameters by dose level in phase 1 Single Ascending Dose Study of Vibozilimod

Dose	Sex	ALC (Lymphocytes/µL blood) [Mean ± SD]					Mean PK Parameters			
(N)		BL	4h	Nadir	24h	96 h	C_{max} (ng/mL)	AUC_{0-t} (h*ng/mL)	$t_{1/2}$ (h)	Vd /F (L)
Placebo (14)	Male, Female	2226.4 ± 379.1	1727.1 ± 423.7 (-22.4)	1727.1 ± 423.7 (-22.4)	1839.3 ± 451.8 (-17.7)	1958.6 ± 386.6 (-11.6)	----	----	----	----
0.1 mg (6)	Male	2078.3 ± 596.6	1666.7 ± 337.9 (-17.8)	1666.7 ± 337.9 (-17.8)	1873.3 ± 433.8 (-7.4)	1933.3 ± 337.4 (-1.5)	0.67	17.83	35.47	200.28
0.3 mg (6)	Male	2851.7 ± 525.2	1986.7 ± 589.5* (-30.9)	1971.7 ± 565.2* (-31.1)	2268.3 ± 507.7 (-21.0)	2383.3 ± 460.4 (-16.3)	1.85	55.76	34.45	235.86
1 mg (6)	Male	2576.7 ± 557.9	1291.7 ± 300.7*** (-50.0)	1160.0 ± 278.6*** (-55.0)	1721.7 ± 255.3* (-32.1)	1968.3 ± 457.1 (-23.6)	6.03	185.83	32.67	249.55
2 mg (6)	Male	2078.3 ± 355.4	1130.0 ± 315.4** (-43.6)	1020.0 ± 394.3*** (-49.0)	1281.7 ± 432.5 (-36.3)	1671.7 ± 485.4 (-20.1)	9.74	324.71	34.31	305.09
2 mg (6)	Female	2441.7 ± 464.2	981.7 ± 107.2*** (-58.6)	851.7 ± 178.8*** (-64.5)	1181.7 ± 399.0*** (-51.8)	1716.7 ± 213.8 (-28.8)	19.94	551.47	32.54	166.77
4 mg (6)	Male	1908.3 ± 477.9	743.3 ± 265.7*** (-60.5)	713.3 ± 102.5*** (-60.4)	775.0 ± 103.3*** (-57.0)	1375.0 ± 343.1 (-26.3)	22.61	720.03	37.18	284.76
6 mg (6)	Male	1995.0 ± 306.9	636.7 ± 208.9*** (-68.1)	440.0 ± 122.1*** (-77.0)	505.0 ± 115.4*** (-74.0)	1156.7 ± 108.4* (-40.6)	33.66	1207.29	36.2	295.68

AUC- Area under the curve; BL – Baseline; C_{max} – maximum concentration; PK – Pharmacokinetic; $t_{1/2}$- terminal half-life; V_d/F- apparent volume of distribution. Values in parasthesiae indicate percent changes from BL.
For PK parameters, C_{max} and AUC are geometric means, $t_{1/2}$ is median; Vd/F is arithmetic mean.
For statistical analysis of absolute lymphocyte count, entire data from pre-dose to EOS (Day 14) h was used; only data at BL, 4h, 24 h and 96 h represented in the table.
*P<0.05, **P<0.01, ***P<0.001 vs. respective group BL values, Two-Way ANOVA followed by Bonferroni's test.

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Abstract N°: 2637

Deep Learning-Based Histopathologic Assessment of Bullous Pemphigoid Heterogeneity

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Introduction & Objectives: The main histopathological characteristics of bullous pemphigoid (BP) may be heterogeneous and related with disease severity or treatment response. This study aimed to investigate association between BP clinical phenotypes and histopathological characteristics based on a deep learning system.

Materials & Methods: We reviewed records of BP patients diagnosed from January 1, 2008 to December 31, 2022 in Xijing Hospital and trained an AI-based deep learning model using whole-slide histopathological imagings to accurately calculate and classify infiltrated inflammatory cells including eosinophils, neutrophils, and lymphocytes in BP lesions.

Results: The study cohort composed of 280 BP patients. Based on the deep learning model, 59 (21.1%), 179 (63.5%) and 43 (15.4%) specimens were found to include eosinophil-predominant, lymphocyte-predominant and neutrophil-predominant inflammatory infiltrates, respectively. BPDAI scores, anti-BP180 NC16A IgG antibody ELISA values and prevalence of skin infection conditions were positively correlated with numbers of eosinophil infiltrate ($P < 0.05$). There is a negative correlation between the degree of neutrophil infiltration and the serum total IgE level ($P < 0.05$). Relapse after glucocorticosteroid reduction within 3 months were negatively correlated with lymphocyte infiltrate ($P < 0.05$). Numbers of inflammatory cells, linear deposits of C3 by direct immunofluorescence and comorbidity with hypertension were statistically significant in the relapse prediction model.

Conclusion: BP patients demonstrated heterogeneity in clinic and histopathology. An AI-based deep learning model may supply a more straightforward and reliable prognosis for BP patients in treatment and management.

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Abstract N°: 2726

One diagnosis, a plethora of presentations: a series of cases of lupus erythematosus

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Introduction & Objectives:

Lupus erythematosus(LE)is a autoimmune, multisystem, chronic inflammatory diseases. It is antibody or t-cell response directed against a self-antigen leads to tissue damage and organ failure. Subtypes:

- Acute cutaneous LE (ACLE)
- Subacute cutaneous LE (SCLE)
- Chronic cutaneous LE (CCLE)

CASE DESCRIPTION:

CASE 1:

A 30 years old , female with complaints of multiple joint pains since 1 and ½ years , multiple crusted lesions over lips, face, and chest since 4 months. Scalp showed few erythematous crusted plaques. Multiple hyperpigmented crusted papules and plaque present over face and targetoid lesions present over neck, extremities and trunk. Oral mucosa and genital mucosa showing erosions and hemorrhagic crusted plaques. Investigations showed ANA – positive, RA factor – positive, AntidsDNA – positive, ESR – 20 mmhr, 24 hour urine protein – 1693 mg. Diagnosed as Rowell's syndrome and treated.

CASE 2:

A 17 years, female presented with red lesions all over body since 1 year. Severe crusting of both lips since 5 months. On examination symmetrical erythema, edema and scaling present all over the face with relative sparing of chin area and nasolabial folds, V area of neck. Periungual erythema, nail fold telangiectasia present. ANA profile showed Anti ds DNA, Anti sm, AntiRNP/Sm, Antihistone antibody positive. Diagnosed as SLE and treated.

CASE 3:

A 9years old , male with reddish scaly lesions all over body since 2 months. Fever and Joint pain since 1 month. On examination multiple hyperpigmented ill defined scaly plaque present over face, neck, trunk , extremities, palms and soles. Oral cavity showed erosions with crust. Periungual erythema present. ANA - RNP/Sm, anti sm, RO/SS-A, ribosomal p protein, anti ds DNA, antihistone antibody positive. Diagnosed as Childhood onset SLE and treated.

CASE 4:

A 60 years, female with pink scaly lesions over face, neck, arms and lower back since 8 years and Joint pains since 3 months. On examination multiple well defined erythematous plaques with adherent scales and hyperpigmented borders, over the scalp, face, trunk, back, and forearms. Confirmed by biopsy, diagnosed as disseminated DLE and treated.

CASE 5 :

A 35 years old, female with blackish discoloration over face since 6 months, Multiple joint pains since 6 months. Symmetrical hyperpigmentation and scaling present all over the face, V area of neck, lower back. Periungual erythema and erosions present. 24 hr urine protein – 1000mg/dl, ANA profile – Anti RNP/Sm, Anti ribosomal P antibody positive. Diagnosed as SLE and treated.

CASE 6 :

A 13 yr old female, with multiple erosions over face, neck, chest, abdomen, back since 15days . Oral erosions, fever and joint pains since 7 days. On examination multiple symmetrical erosions, edema present all over the face. Petechiae present over tips of fingers and toes. Multiple ulcers present over chest and back. Periungal erythema present. ANA profile showed ANA - RNP/Sm, anti sm, RO/SS-A, ribosomal p protein, anti ds DNA, antihistone antibody positive. Diagnosed as Acute syndrome of apoptotic pan-epidermolysis(ASAP) and treated.

Discussion:

LE is a common autoimmune disease that manifests in systemic and cutaneous forms. Cutaneous features seen 75% to 80% of all patients with LE. It might still be an underdiagnosed and underestimated disease.We report this case series to highlight the necessity to suspect Lupus in all patients.

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Abstract N°: 2735

Lichen sclerosus mimicking vitiligo in a young boy

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Lichen sclerosus mimicking vitiligo in a young boy

Introduction & Objectives:

Vitiligo and lichen sclerosus (LS) are two inflammatory skin diseases characterised by depigmented lesions. They both frequently affect the anogenital area.

Materials & Methods:

We report the case of a 13-year-old boy, with a history of circumcision due to phimosis and suspicion of LS 5 years ago, who was referred to our department for asymptomatic depigmented macules on his penis. These lesions were stable for 4 years. He experienced no difficulties with urination. Physical examination revealed, depigmented macules on the penis without porcelain-like sclerotic aspect or other textural changes. There were no lesions on the perianal region and the rest of the body. The diagnosis of vitiligo was suspected.

Results:

The patient was initially treated with topical tacrolimus twice daily during 3 months without improvement. At follow up a sclerotic aspect of urethral orifice was noticed compatible with LS. Urology evaluation confirmed urethral stricture. These observations led to the diagnosis of vitiligoid LS.

A treatment with clobetasol propionate cream 10g per months for 3 moths was introduced with good evolution.

Conclusion:

LS and vitiligo are both chronic inflammatory autoimmune dermatoses, that can present with depigmentation. LS may result in skin depigmentation without textural changes making hard to distinguish it from vitiligo. However timely diagnosis of LS is essential as the treatment with topical corticosteroids prevent long-term outcomes, including architectural changes and squamous cell carcinoma. The two entities seem to have some similarities in their pathophysiology. Multiples cases involving co-localization of LS and vitiligo are reported.

Histological examination can help to distinguish these entities. We report this case to highlight the presence of vitiligoid-like lichen sclerosus, a condition frequently misdiagnosed.

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Abstract N°: 2751

concomitant presence of frontal fibrosing alopecia (ffa) and linear scleroderma (morphea): a report of two cases

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Introduction & Objectives:

Frontal fibrosing alopecia (FFA) is a primary lymphocytic cicatricial alopecia and is thought to be a variant of lichen planopilaris (LPP). Morphea (localized scleroderma) is an autoimmune connective tissue disease and En coup de sabre is a localized linear morphea. Herein, we report the concomitant appearance of both morphea and FFA in two patients.

Materials & Methods:

Two patients with morphea and FFA were enrolled.

Results:

Case 1

A 49-year-old woman with a history of linear morphea of face and scalp since she was 15 years old and a six-year history of frontal hair loss presented to our clinic. She had multiple erythematous atrophic plaques on the right side of her forehead, right cheek, and also the left side of her neck. Also, she had regression of frontal hairline on both sides of her face with atrophy of the skin and perifollicular scale. Scattered alopetic patches were seen on her frontal scalp and a sclerotic plaque of morphea was detected on her lower back. Biopsies from the scalp showed FFA and also the biopsy from the facial plaque showed morphea.

Based on clinical and histological findings, the patient was diagnosed with concomitant FFA and morphea.

She had not received any systemic therapy for her morphea until six years ago when hair loss started. She had taken methotrexate (MTX) on and off during the last six years. We prescribed MTX with the dosage of 15 mg/week and dutasteride for her FFA.

Case 2

A 70-year-old woman with a previous medical history of diabetes mellitus, and hypertension presented to our clinic. She also had hairline regression during the past five years which was diagnosed with FFA. Moreover, she had been diagnosed with morphea three years ago.

Hairline regression with mild cutaneous atrophy on the frontal hairline and multiple skin-colored facial papules were observed. A biopsy taken from the frontal hairline showed FFA. Furthermore, multiple erythematous shiny sclerotic plaques were observed on her breasts and inframammary skin, stomach, and lower extremities. Histologic examination taken from her trunk lesions showed morphea.

Based on clinical and histological findings had both FFA and morphea. She has been taking mycophenolate mophetil 2 g/day since one year ago for morphea.

Moreover, she was taking finasteride 5 mg/daily, topical tacrolimus, and isotretinoin 20mg twice/week for FFA since diagnosis. Due to the erythema of morphea lesions, we added MTX for her 15 mg/week.

Conclusion:

We presented a rare association of FFA and morphea in two patients. One patient had severe linear morphea on the face and scalp that developed FFA more than 28 years later after the morphea onset. Both morphea and FFA were progressive in this case.

The other patient was an old woman who developed morphea two years after the onset of FFA.

There exists only one reported case of concomitant morphea and FFA reported by Abdalla et. al. They reported a 51-year-old Caucasian female with a 2-year history of frontal hair loss and noticed a linear atrophic plaque on the frontal region. These three cases (the one reported by Abdalla et. al and the two cases reported here) are all middle-aged to old female patients. Two of them first demonstrated FFA before the presentation of morphea and one developed morphea years before developing FFA.

LPP, FFA, and morphea en coup de sabre are cutaneous conditions including inflammation autoimmune response, and fibrotic changes. Physicians should be aware of the possible associations in these patients and their managements.

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Abstract N°: 2752

Lupus Panniculitis Masquerading as Mastitis: A Diagnostic Challenge in a 17-Year-Old Female Patient

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Introduction & Objectives:

Lupus mastitis is a rare presentation of lupus panniculitis marked by inflammation of the subcutaneous fat tissue. It affects about 2-3% of people with systemic lupus erythematosus (SLE) and may be the first symptom of the disease. It can present as masses in the breast, axillary lymphadenopathy, fibrosis, and calcifications. It should be differentiated from infection and malignancy. We describe a case of lupus panniculitis presenting as lupus mastitis highlighting the importance of a thorough diagnostic approach in such challenging cases.

Materials & Methods:

We report a case of a 17-year-old female patient who presented with lupus mastitis. History and physical examination were reviewed. Histologic sections were assessed with hematoxylin and eosin (H&E), and alcian blue stain.

Results:

A 17-year-old female, presented with a 4-month history of multiple tender pruritic erythematous nodules with erosions and ulcerations on the right breast which subsequently increased in size and number affecting the right lateral neck, associated with periorbital swelling. She was admitted under pediatric service and was initially diagnosed with facial cellulitis and breast abscess. Blood chemistries, gram stain and culture of blood, wound, and tissue, chest radiographs, ultrasound of the bilateral breast and whole abdomen, CT scans of the cranial, orbit, and chest, and breast and submandibular tissue biopsies were done. Breast ultrasound revealed a large mass with a small pocket of fluid on the right breast, while chest CT scan revealed swelling of the right breast parenchyma and areas of subcutaneous edema, both of which were consistent with a right breast abscess. Despite receiving a variety of oral antibiotics, pain relievers, antifungal medications, and intravenous corticosteroids, no improvement was observed, prompting the referral to our service. On physical examination, multiple well-demarcated erythematous to hyperpigmented indurated plaques on the left eye, submandibular area, neck, trunk, and bilateral arms, and multiple firm tender nodules on the right breast were noted. A 4 mm skin punch biopsy was done on the left arm and trunk, and sections revealed vacuolar interface changes, a mild inflammatory infiltrate composed of lymphocytes surrounding the upper vessels, adnexal structures, and fat lobules consistent with superficial and deep perivascular dermatitis with panniculitis on H&E stain, and an increase in dermal mucin on alcian blue stain. After further laboratory investigations revealing leukopenia, anemia, protenuria and granular cast on urinalysis, elevated erythrocyte sedimentation rate and C-reactive protein, positive for antinuclear antibody (ANA) with a value of 447, and negative for anti-double-stranded-DNA (dsDNA) antibody, she was diagnosed with lupus panniculitis and was given hydroxychloroquine 200mg/day and prednisone 20mg/day. After 1 month of treatment, there was a decrease in swelling and erythema of previously noted lesions.

Conclusion:

We presented a rare case of lupus mastitis. This case highlights the diagnostic challenges posed by lupus panniculitis masquerading as mastitis and emphasizes the need for a comprehensive evaluation in complex dermatologic cases. Early recognition and appropriate treatment of lupus panniculitis can improve patient

outcomes and prevent unnecessary interventions.

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Abstract N°: 2758

Pulmonary interstitial lesions in pemphigus mouse model: verifying pemphigus may not be only limited to skin and mucosa

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Introduction & Objectives:

In previous studies, pemphigus mouse models were mostly used to study the clinical manifestations and pathogenesis of the disease itself, and less used for pemphigus comorbidities. Based on the high incidence and poor prognosis of interstitial lung disease (ILD) in patients with pemphigus observed in clinical practice, we here consider establishing a pemphigus mouse model by subcutaneous injection of pemphigus patient 's serum IgG to evaluate its pulmonary lesions and further reveal the correlation between pemphigus and ILD.

Materials & Methods:

The serum of pemphigus patients and healthy people was collected, and the IgG in the serum of the two groups was extracted and injected subcutaneously into 6-week-old male Balb/c mice. The skin tissue was extracted for section staining to confirm whether the pemphigus model was successfully established. Subsequently, the lung tissues of mice in each group were extracted for section staining, qRT-PCR and Sircol assay collagen detection to evaluate lung lesions.

Results:

HE staining showed that the early lung tissue of pemphigus mice showed alveolar septal thickening, local lung tissue destruction with a large number of inflammatory cell infiltration, and the late inflammation gradually subsided, but followed by fibroblast proliferation, partial alveolar septal thickening, fibrosis nodules, and finally showed significant fibrosis occlusion. Masson staining showed that there was obvious fibroblast proliferation in the alveolar space of pemphigus mice, and the Ashcroft pulmonary fibrosis score was significantly higher than that of the control group. With the increase of subcutaneous injection of serum IgG dose, the collagen content of lung tissue increased and the degree of fibrosis increased. The expression of collagen genes (Col1a1, Col1a2) in lung tissue of pemphigus mice was significantly increased. DIF staining showed serum IgG deposition in the lung tissue of pemphigus mice, suggesting that subcutaneous injection of human IgG may be deposited in the lungs of mice to cause damage.

Conclusion:

The pemphigus mouse model has interstitial lung disease, which breaks the traditional definition of pemphigus skin mucosal targeted organ-specific autoimmune disease and classifies it as a systemic autoimmune disease that can be accompanied by other organs (especially the lungs).

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Abstract N°: 2820

Linear IgA dermatosis of childhood: a series of 6 cases

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Introduction:

Linear IgA dermatosis (LAD) is an acquired autoimmune bullous disease (AIBD) characterized by linear deposition of IgA along the dermal-epidermal junction. Although rare, it is the most common AIBD in children. We report a paediatric series of 6 cases.

Materials & Methods:

A single-centre, retrospective study conducted between 2021 and 2023, including all children (under 15 years old) hospitalised for confirmed LAD.

Results:

6 cases were identified (male = 5 and female = 1). The mean age was 6.6 years with extremes ranging from 3 to 12 years. The median interval from onset to diagnosis was 3.5 months. No cases of drug-induced triggering were noted. In 66% of cases, cutaneous involvement was disseminated and multi-bullous. The vesiculo-bullae exhibiting rosette-like pattern in 4 cases. Mucosal involvement occurred in 16.6% of children. Cutaneous superinfection was observed in 2 cases. Immunohistologically, junctional cleavage and linear IgA deposition were present in all cases. Treatment consisted of Dapsone alone at a dose of 1 to 2 mg/kg/d in 5 cases (83%). One child received a combination of dapsone and systemic corticosteroid therapy. All cases showed favorable evolution.

Conclusion:

Our series is notable for the absence of drug-induced forms and the marked predominance of males. Furthermore, our results reproduce the main characterized of pediatric LAD, including the peak incidence in second childhood, the rosette-like bullae presentation, the high prevalence of pruritus, the efficacy of dapsone and the good prognosis.

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Abstract N°: 2831

Bullous pemphigoid: a study of 23 cases

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Introduction & Objectives:

Bullous pemphigoid (BP) is a rare autoimmune subepidermal blistering dermatosis. It most commonly affects elderly patients. The aim of our study was to describe the epidemiological epidemio-clinical, histopathological, therapeutic and evolutionary aspects of patients with BP in a hospital series of 23 cases.

Materials & Methods:

A monocentric retro-prospective study, including all patients hospitalized, between January 2020 and December 2023, for BP. Diagnosis was based on clinical, histological and immunological criteria.

Results:

Twenty-three cases were identified. The mean age was 54.2 years (1-90). A female predominance was noted with a sex ratio of 0.35. A history of neurodegenerative disease was found in 3 cases (13%), with two cases of Alzheimer's disease and one case of stroke. Skin involvement was disseminated and multi-bullous in more than 85% of cases. Mucosal involvement was present in 17.4% of cases and nail involvement in 4.4% of cases. Pruritus was constant. Eosinophilia was observed in 17 patients (73.9%). Cutaneous superinfection was found in 6 cases (26.1%). Immunohistologically, junctional cleavage was observed in all our cases. linear labeling along the epidermal-dermal junction in direct immunofluorescence was present in 91.3% of cases. Indirect immunofluorescence was positive in 10/11 (90.9%). Therapeutically, topical corticosteroid alone was prescribed in 5 cases (21.7%). Systemic corticosteroid (0.5 to 1 mg/kg/d) was administered in 18 patients (78.3%). The addition of an immunosuppressant was indicated in 2 cases (8.7%), with one case receiving cyclosporine and one azathioprine. A good initial clinical response (after an average of 2-3 weeks) was achieved in more than 85% of cases. Progression was marked by 2 cases (8.7%) of relapse and one case of recurrence.

Discussion:

The results of our series of PB concur with those reported in the literature concerning the predominance of females and the rarity of mucosal involvement. In addition, our study is distinguished by a relatively young population (54 years, vs. over 67 years in most series), a low association with neurological disorders, and an overall favorable evolution.

The treatment most commonly used in our patients was general corticosteroid. Current international recommendations favor topical corticosteroid as first-line treatment because of their similar efficacy and fewer side effects.

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Abstract N°: 2846

Ulcerative skin manifestations in Wegener granulomatosis

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Introduction & Objectives: Wegener granulomatosis or granulomatosis with polyangiitis is a small-medium vessel necrotizing vasculitis, which is assosieted with the anti-neutrophil-cytoplasmic-antibody (ANCA). The disease can affect any part in the body, but it has a predisposition for certain organs, the upper respiratory tract (sinuses, nose, ears, and trachea), the lungs, and the kidneys but sometimes can start with the skin. According to a large multicenter study, skin affection in GPA is common and was found in 34% of patients.

Materials & Methods: A 46-year-old male patient with lesions on the body that appeared 20 days ago, resembling an erythematous/hyperpigmented macula on the right upper leg, which gradually expanded with progression to necrotizing ulcers. During the examination, changes in the lower limbs are observed in the form of erythematous patches, hemorrhagic crusts and numerous necroses. On the left leg, dorsal to the lower leg, there is a necrotic ulcer covered with an eschar the size of a child's palm, while on the dorsum of the upper leg to the subgluteal, three smaller necroses are observed surrounded by erythemalivid beaches. On the right lower leg on the flexor side, a more accentuated erythematous plaque with a necrotic focus can be observed.Changes of the same kind are observed on the upper limbs, but they are less pronounced. There are numerous point necrosis on the palms. On the forearms and upper arms, disseminated isolated hyperpigmentation. On the left foot, the left knee and the dorsum of the right hand, individual sero-hemorrhagic bullae are observed. Extensive erosions with white adherent deposits 1x1 cm are present in the oral mucosa and on the glans penis.

Laboratory analyzes indicate increased erythrocyte sedimentation, leukocytosis, increased c-reactive protein (crp), increased transaminases, increased values of d-dimers. Microbiologically, Citrobacter freundi ESBL+, Candida albicans/species, MRSA, Pseudomonas aeruginosa were isolated from the wound swab. Immunological analyses-antidsDNA, c-ANCA, and ACL-IgG are negative. From diagnostic procedures, an X-ray of the lungs was made with an accentuated bronchovascular pattern, with fibrotic changes on the right in the middle parts. A skin biopsy was also taken, which supports Wegener's granulomatosis in corelation with skin changes.

The patient was treated with parenteral corticosteroids (prednisolone equivalents initial dose of 75mg, with gradual reduction). Anticoagulation therapy, antibiotic therapy according to antibiogram (Imipenem, Vancomycin). Oral antibiotic (Ciprofloxacin), gastroprotective, antihistamine and local antiseptic and antibiotic therapy. After starting the therapy, there was an improvement in the patient's condition and the laboratory parameters, as well as an improvement in the skin changes.

Results: In the following months, the patient continued with oral immunosuppressive (Azathioprine) and corticosteroid therapy with dose reduction, as well as local antiseptic and antibiotic therapy, during which complete epithelialization of the changes occurred. Unfortunately, the patient was diagnosed during the covid-19 pandemic, who later tested positive for sars-covid-19 and unfortunately lost the battle as a result of numerous complications.

Conclusion: There are not many studies that GPA develops and starts with cutaneous manifestations and later develops symptoms from other involved systems.

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Abstract N°: 2857

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

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

Case report: A 59-year-old female presented with intermittent low-grade fever and multiple painful crusted erosions over lips/oral/nasal/genital mucosa for the last 4-months. She also complained of mild redness and burning in both the eyes. During hospitalization, she developed similar lesions, predominantly over the venepuncture sites and a few over the trunk (pathergy phenomenon). She had anemia, raised 24-hour urine protein and ESR. Anti-Dsg 1/3 antibody titres were 1:300/1:400 respectively and HLA-B*51 was positive.Histopathology of the oral mucosa showed ulceration of the epidermis with dense mixed inflammatory infiltrate extending till the subcutaneous tissue and muscle along with perivascular inflammation with features of leukocytoclastic vasculitis and intraepidermal collection of neutrophils. Direct immunoflourescence (DIF) revealed IgG and C3 deposits in the intercellular spaces of the epidermis in a fish-net pattern. Histopathology of the skin could not be performed due to tissue loss during processing caused by extreme fragility; however, DIF revealed similar findings suggestive of PV. She did not respond to oral prednisolone (1mg/kg/day), injection dexamethasone pulse, and azathioprine. The therapy was shifted to infliximab infusion at 5mg/kg (300mg) at 0, 2, and 4-week without any significant healing of the lesions. Subsequently, injection Adalimumab was started with a loading dose of 80mg followed by 40mg subcutaneous every 2-weeks which resulted in significant healing of the lesions starting within the first two weeks. The oral steroids could be tapered rapidly over next four weeks and she is maintained on injection Adalimumab. The last follow-up at 6-month revealed no new lesions and the existing lesions had healed significantly.

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

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Abstract N°: 2895

Anti-p200 pemphigoid: a clinicopathological challenge

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Introduction & Objectives:

Anti-p200 pemphigoid is an uncommon autoimmune subepidermal bullous disease characterized by the presence of IgG-type antibodies against a 200kDa protein located in the lamina lucida of the basement membrane called laminin γ1.

Materials & Methods:

We present the case of a 47-year-old female patient from Venezuela who arrived at the emergency department with a three month story of pruritic blistering lesions that appeared with friction or with minor trauma. On a first visit, she had erosions, post-inflammatory hyperpigmentation and milium cysts in rubbing areas and nail dystrophy in her toes. During her evaluation, she presented with an outbreak of disseminated erythematoedematous lesions, accompanied by tense blisters, erosions and involvement of the oral and pharyngeal mucosa that conditioned dysphagia to liquids and solids. She didn't refer to worsen with the sun. At the same time as the appearance of the cutaneous clinic, she was evaluated by an ophthalmologist and she was diagnosed of a left corneal ulcer.

Results:

Histopathology revealed a subepidermal blister with neutrophilic infiltration, with linear deposits of IgG, IgM, C3 and C1q. No anti-BP180, anti-BP230 antibodies or anti-collagen VII were detected, ruling out bullous pemphigoid (BP) and epidermolysis bullosa acquisita (EBA), respectively. Elevated ANAs and anti-Ro/SSA were highlighted with no other clinical-analytical criteria for systemic lupus erythematosus, which ruled out the diagnosis of blistering systemic lupus erythematosus.

In view of the suspicion of anti-p200 pemphigoid, immunohistochemistry revealed the presence of collagen IV at the base of the blister, reinforcing the diagnosis of this entity. The patient was then treated with sulfone with remission of almost every lesion.

Conclusion:

Anti-p200 pemphigoid preferentially affects young people and is characterized by a variable clinical presentation, with lesions often on palms of hands and feet and in 20% of cases with mucosal involvement. Identification of collagen IV at the base of the blister by immunohistochemistry is a practical and economical strategy to differentiate this disease from EBA. Likewise, more specific confirmation methods such as ELISA or immunoblot are sometimes necessary to detect the p200 antigen, however, they are not always available at every centre.

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Abstract N°: 2938

Amyopathic dermatomyositis presenting as a unilateral heliotrope erythema and palpebral edema mimicking Romaña's sign

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Introduction & Objectives:

A 44-year-old Colombian woman of mestizo ethnicity presented to our clinic with a history of non-painful unilateral palpebral edema and erythema and an apparent bug sting when the symptoms started. She had no remarkable medical history at the moment. The patient sought medical attention and initial diagnosis was periorbital cellulitis and was treated with multiple antibiotic schemes with no improvement. She received metamizole for general symptoms and subsequently developed a pruriginous and extensive erythematous rash with plaques and papules that persisted.

Materials & Methods:

Patient lived in a tropical area, had a history of fibromyalgia and had contact with domestic animals, but reported no previous history of traveling, and neither remembered being bit by a tick or a triatominae. She reported having no allergies. Skin findings included unilateral edematous heliotrope erythema predominantly of the upper eyelid, Gottron's sign and V-sign.

Initial diagnostic workup included Trypanosoma cruzii serology, Coxsackie virus, antinuclear antibodies (ANA), extractable nuclear antigens (ENA), rheumatoid factor and complement levels but they all turned normal. The probability of Drug reaction with eosinophilia and systemic symptoms (DRESS) was calculated by the RegiSCAR score but was unlikely due to absence of symptoms. Physical exam was otherwise normal, with preserved strength and sensibility of the limbs. CPK and electromyography showed no alterations and no samples of muscle biopsy were collected. Skin biopsy of abdomen and limbs revealed findings of toxicoderma, which is a common finding in autoimmune diseases. Furthermore, myositis-specific antibodies reported positive NXP-2 and given that she had heliotrope erythema, Gottron's sign and V sign with no muscle involvement amyopathic dermatomyositis was diagnosed according to both EULAR and JAAD criteria. Patient was treated with azathioprine, prednisone and topical corticosteroids with improvement of edema but persistence of violaceous erythema.

Results:

Amyopathic dermatomyositis is a rare clinical diagnosis. It has been previously described that presentation can be unnoticed in some patients, specially in dark-colored skin groups. This patient presented with edematous heliotrope erythema sparing the contralateral eye, which is a very uncommon finding. Gottron's sign and papules were less noticeable in her and rashes presented in a subtle way. Characteristic findings are of foremost importance to diagnosis, and it should be taken into account that skin involvement can precede muscle disease for months.

Conclusion:

Anti-NXP2 dermatomyositis is usually associated with muscle involvement and classical skin findings, with a higher

malignancy risk. Occult cancer risk can persist for years and individual factors should be taken into account. Patient is at high risk and active screening is recommended.

Table 1

Clinical findings: Heliotrope erythema, Gottron's sign, V sign.	**Electromyography: no electrodiagnostic evidence of a neuropathic or a myopathic process.**
Infectious diseases studies: T. cruzii negative, VDRL negative, Coxsackie negative,HIV negative. Autoimmunity: ANA negative, Anti La/SSa, Anti-Ro/Ssb, Anti-RNP negative. Rheumatoid factor < 15 C3: 170.8 C4: 12.8	CPK and routine testing: CPK in normal ranges (37), AST and ALT in normal ranges.
Myositis-specific antibodies: NXP2++, PM-Scl 75+ and OJ +positive.	MRI and muscle biopsy were not performed.

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Abstract N°: 2980

Clinical significance of myositis-specific and myositis-associated antibody profiles in dermatomyositis

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Introduction & Objectives: Myositis-specific autoantibodies (MSA) and myositis-associated autoantibodies (MAA) are clinically useful biomarkers to help the diagnosis of dermatomyositis (DM). Most of them are associated with a specific clinical subset of DM, making them useful in predicting certain clinical manifestations. Prognosis also differ depending on the MSA positivity and hence could help in determining the therapeutic strategy as well.

Objective: To estimate the prevalence of MSA as well as MAA and analyze possible clinical correlations of these autoantibodies in patients diagnosed with DM.

Materials & Methods: We conducted a cross-sectional study of 30 patients who attended dermatology and rheumatology departments of a quaternary care center in South India during the period of April 2016 to March 2021and were diagnosed with DM who met at least four of the criteria defined by Bohan and Peter. Data regarding demographic features, clinical manifestations, laboratory and radiographic investigations, as well as the presence of internal malignancies and interstitial lung disease in these patients diagnosed with dermatomyositis during this period was retrieved from the electronic medical records in hospital information system.

Results: Mi 2 was positive in 8 (27%) patients and this was the most frequently found MSA. MSA was positive in 19 patients (63%). AntiPM/Scl 75 and anti- Ro 52 were positive in 5 (16.7%) each and these were the most commonly found MAA. Anti- La was absent in all our patients. A total of 11(36.7%) patients showed positive MAA. There were 8 (27%) patients in whom both MSA and MAA were positive. Either MSA and/or MAA were positive in 22 (73%) patients. On a bivariate analysis the patients who were positive for anti-PM/Scl 75 showed a significant difference in manifesting cutaneous ulcers (p-value 0.023). It was also found that anti-SAE positive patients showed a significant difference with malignancy (p-value 0.014). Anti-Ro 52 positive patients were less likely to have symmetrical proximal muscle weakness (p-value 0.006).

Conclusion: Anti-MDA5 is predominantly seen in clinically amyopathic dermatomyositis (CADM) / amyopathic dermatomyositis (ADM), but our study revealed that all patients who were anti-MDA 5 positive had myositis. None of the anti-MDA 5 positive patients had rapidly progressive interstitial lung disease (RPILD). More than one MSA in the same patient was noted in this study. All patients with of anti-Jo1 antibodies had anti- synthetase syndrome. Anti-TIF1γ/α positivity showed associated malignancy. Anti-SRP positive patients had significant myopathy. The small sample size was a limitation in our study which made it difficult to arrive at significant differences.

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Abstract N°: 3032

Dramatic response to anifrolumab in refractory cutaneous systemic lupus

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Introduction & Objectives: The wide range of clinical manifestations in systemic lupus erythematosus (SLE) is due to the heterogeneity of the disease in which different etiopathogenic factors and immunological abnormalities intervene. Patients can present any combination of characteristics, and these can change throughout the disease and treatment, with cutaneous lesions in a high percentage of cases.

Materials & Methods: We present the case of a 54-year-old woman in follow-up in dermatology and rheumatology department for SLE with cutaneous, articular, and renal affection (class IV, in remission, received cyclophosphamide and mycophenolate mofetil) with different lines of treatment required. As for cutaneous lesions, subcutaneous lupus lesions on the face, ears, and hands along with no scarring alopecia was present, with a severe Raynaud phenomenon. She had thrombopenia, low levels of complement and high levels of anti-dsDNA antibodies. She was treated with methotrexate, prednisone, hydroxychloroquine and bosentan with a bad control of inflammatory arthralgias and skin manifestations. Other treatments added for a better cutaneous control was dapsone with no improvement and mepacrine with partial control but abandoned for a marked yellow pigmentation. She showed flares of erythematous non-scarring scaling skin lesions, on the face where she also had oedema and ears, arms, neckline, and dorsum of hand along with Raynaud and arthralgias, and due to this maintained bad control anifrolumab was added. The response was fast with excellent control after one infusion and maintained 9 months until now with a great improvement in quality of life.

Results: Knowledge gained over many years has implicated type I interferon (IFN) in the pathogenesis of SLE. Anifrolumab, a human monoclonal antibody against type 1 IFNα receptor subunit 1 (IFNAR1) inhibitor, has been recently approved for moderate to severe SLE while on standard therapy

Conclusion: Early use of anifrolumab in moderate to severe cutaneous manifestations of systemic lupus may result in significant improvement in patients as in our case.

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Abstract N°: 3039

Evaluation of the efficacy and safety of dapsone associated with topical corticosteroids for pemphigus foliaceus: a retrospective multicenter study.

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Introduction & Objectives:

European guidelines recommend using dapsone in addition to topical corticosteroids (TCS) as first-line-treatment for mild pemphigus foliaceous (PF). However, very few studies have evaluated dapsone's efficacy as a single agent in PF. The aim of this study was to evaluate the efficacy and safety of dapsone with TCS for PF.

Materials & Methods:

This is a descriptive, retrospective, multicenter, national French study, including PF patients, followed between May 1990-May 2023, treated with dapsone and TCS. PF severity was evaluated by affected body surface or PDAI-score. The primary end point was to evaluate the objective response rate (ORR) defined as the percentage of patients presenting partial response (PR) or complete response (CR) on the treatment regimen at 12 (M12), 24 (M24), 36 months (M36) and at the end of follow-up (global follow-up). The secondary end points aimed to identify the

factors predicting response and influencing* the drug survival on the treatment regimen, to estimate the duration* of response and to specify its safety profile.

Results:

77 cases were included (men n=42, 56%; median age=58 yo). The patients' baseline characteristics are available in Table 1. At some point during follow-up, 50 patients (64,9%) demonstrated an objective response (PR or CR), but only 21 patients (27,3%) achieved CR. CR was achieved respectively in 9 (29%), 11 (27%) and 1 (14%) patient with mild, moderate, and severe PF. Global follow-up durations varied amongst patients (median: 3.3 years, range: 0.25-32.0 years). Best objective response rate (ORR) was achieved at M12 (40.9%) whereas ORR dropped for later timepoints at M24 (21%) and M36 (22%). Events that led to dapsone discontinuation included treatment inefficacy for 38 cases (49%), adverse events for 18 cases (23%), and obtention of CR/PR for 6 cases (8%). 55 patients (72%) required dapsone replacement or association with an immunosuppressant due to flares (69%) and adverse events (AEs) (31%). AEs occurred in 39 patients (50%), 9 of whom (12%) had grade 3-4 CTCAE AEs, including death (n=2, 3%).

Multivariate analysis did not find any predictive factor for objective response at M12. In later timepoints, having a mild PF was associated with an objective response at M24 (p=0,0119, OR: 5.99), while having a PF treated after 2017 was associated with a lack of response at M24 (p=0,0363, OR= 0.223) and M36 (p=0,0020, OR=0.071) (Table 2). 2017 was the year of publication of the ritux3 trial demonstrating the value of using rituximab as first-line treatment. Survival analysis found a median survival of 18 months on dapsone and TCS. The results of the multivariate cox regression analysis are available in Table 3. When considering inefficacy and AEs as treatment cessation causes (Table 3a), the survival on dapsone and TCS was significantly longer for mild PF (P = 0.0462, HR= 0,559), and shorter for cases treated after 2017 (P = 0.0007, HR= 2.891). When considering treatment cessation for inefficacy only (Table 3b), the survival was longer for mild PF (P = 0.0077, HR = 0.3451) and shorter for cases treated after 2017 (P = 0.0062, HR= 3.041) and for pemphigus herpetiformis cases (P = 0.0039, HR = 5.799).

Conclusion:

The only significant predictors of good long-term-response were low PF severity and decade of inclusion (<2017). Otherwise, dapsone's long-term efficacy and safety were modest. Nevertheless, its place in mild disease, especially in severely immunosuppressed patients remains to be determined.

Table 1: Baseline characteristics and dapsone monotherapy efficacy through overall follow-up of 77 patients with PF

	N	%
Age (yo), median, (IQR)	58	(7-88)
Female sex, n (%)	35	(44%)
Diagnostic delay (years), median, (IQR)	0.25	(0.08-17.00)
Compatible histopathology (n, %)	77	(100%)
Positive DIF (n, %)	74	(96%)
Positive IIF (n, %)	38	(50%)
ELISA anti-Dsg1		
Positivity (n, %)	59	(77%)
Dosage in IU/mL, mean (IQR)*	151	3-209
Drug-induced PF, n (%)	8	(10%)
P. herpetiformis, n (%)	5	(7%)
PF severity		
Mild (n, %)	31	(40%)
Moderate (n, %)	41	(53%)
Severe (n, %)	5	(7%)
Comorbidities (n, %)	38	(49%)
Cancer (n, %)	4	(5%)
Auto-immune disorders (n, %) **	6	(7%)
Cardiovascular diseases (n, %)	12	(16%)
Thyroid dysfunction (n, %)	4	(5%)
Depression (n, %)	6	(7%)
Osteoporosis (n, %)	8	(10%)
Maximal dapsone dosage (mg/day), mean (IQR)	250	(0.4-4.3)
TCS (n, %)	75	(97.5%)
Best response to dapsone monotherapy		
Objective response (PR + CR), N (%)	50	(64.9)
Partial response, N (%)	29	(27.3)
Complete response, N (%)	21	(37.6)
Median survival time on dapsone (years), median (IQR)	1.5	(0.08-16.30)
Reasons for dapsone monotherapy discontinuation		
Global follow-up (years), median (IQR)	3.3	(0.25-32.00)

*Median and range of anti-Dsg1 titers were calculated for 52/59 patients (quantitative values available for 52 patients)
**Hashimoto thyroiditis, Biermer's disease, rheumatoid arthritis, type 1 diabetes mellitus and systemic lupus erythematosus

Table 2: Results of backward stepwise multivariate linear regression analysis: factors associated with objective response at 12, 24 and 36 months in PF patients treated with dapsone monotherapy.

	Model 1*		Model 2**	
	OR (95% CI)	**P value**	**OR (95% CI)**	**P value**
M12				
Pemphigus herpetiformis	2.780E-6 (2.667E-283 – 2.897E271)	0.9686	-	-
Treatment after 2017	0.440 (0.163-1.191)	0.1061	-	-
M24				
ELISA Ab titres	0.911 (0.979-1.003)	0.1331	-	-
Mild PF	3.964 (0.885-18.378)	0.0784	5.990 (1.484-24.174)	0.0119
Treatment after 2017	0.162 (0.033-0.0803)	0.0258	0.223 (0.055-0.909)	0.0363
M36				
Mild PF	0.644 (0.071-2.313)	0.1283	-	-
Treatment after 2017	0.085 (0.010-2.551)	0.0258	0.071 (0.013-0.380)	0.0020

* Model 1 includes variables with P < 0.2 in univariate analysis.
** Model 2 includes variables with significant differences selected by backward elimination

Table 3: Results of multivariate cox regression analyses.

	HR (95% CI)	*P value*	*VIF*
A. Cox regression model considering inefficacy and adverse events*			
Herpetiformis PF	2,485 (0,839-5,952)	0,0628	1,385
History of other auto-immune disease	2,651 (0,884-6,479)	0,0501	1,083
Treatment before / after 2017	**2,891 (1,581-5,436)**	**0,0007**	1,450
Mild PF	**0,559 (0,308-0,976)**	**0,0462**	1,027
B. Cox regression model considering only inefficacy**			
Treatment before / after 2017	**3,041 (1,384-6,863)**	**0,0062**	1,372
Herpetiformis PF	**5,799 (1,549-18,01)**	**0,0039**	1,081
Mild PF	**0,3451 (0,149-0,725)**	**0,0077**	1,444

* Cessation for response in 6 patients were considered as censures. The first part of the table reports the results of multivariate cox regression analysis when considering cessation for adverse events and inefficiency as an event.
** The second part of the table report the results when only considering cessation for inefficacy.

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Abstract N°: 3166

Rituximab alone is more cost effective and efficacious than combined rituximab with Intravenous Immunoglobulin in treatment of pemphigus, a prospective study

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Rituximab alone is more cost effective and efficacious than combined Rituximab and Intravenous Immunoglobulin in treatment of pemphigus, a prospective study

Introduction & Objectives:

Pemphigus is a rare but life-threatening autoimmune blistering disease. Rituximab, is anti-CD 20 monoclonal antibodies which showed to be effective treatment in pemphigus, but there is potential risk of severe infection. IVIg has been used in patients with acquired immunodeficiency. We aimed to investigate the efficacy and safety of combined intravenous immunoglobulin (IVIg) and rituximab compared to rituximab alone in disease control and infection risk reduction in pemphigus.

Materials & Methods:

This is a prospective open-labelled randomized controlled study. Patients with moderate-to-severe pemphigus (PDAI>15) and BSA>5% are randomly assigned in a 1:1 ratio to receive rituximab (RX arm: 375mg/cm2 weekly, total of 12 injections, every 6months) or combined rituximab-IVIg regime (combined RI arm: rituximab 375mg/cm2 weekly, total of 12 injections, together with monthly IVIg (2g/kg/infusion) of 12). Disease activity score (PDAI), BSA, serum Desmoglein (DG) 1,3 autoantibodies titre and CD19 counts were recorded at baseline and follow-up.

Results:

20 subjects are recruited with a mean age of 54.8±9.84 years old (range 28-68), 11(55%) are men, a median of disease activity (PDAI) 31 (range 23.7-41.8) and BSA 7% (range 5-13), and DLQI 18 (8.8-28). Patients in RX arm are able to wean off systemic corticosteroid (Cs) faster than patients in combined arm (median week 16 (6-24) vs week 19 (16-40) and more likely to remain in clinical remission without Cs up to week 72 (0% vs 60%) ($p<0.003$). At week 24 & 72, patients in RX arm achieved greater PDAI improvement than those in combined arm (82.2% vs 70.5%, p=0.024*, 96% vs 69% p=0.034* respectively. One patient in combined arm suffered from Pneumocystis cariini pneumonia and required hospitalized during treatment while none suffered from severe infection or required hospitalization in RX arm. The cost of combined treatment arm is much higher than RX alone arm. Due to the cost of IvIg, limitation of this study is small cases number with potential bias.

Conclusion:

Our prelimary result showed that Rituximab only arm is more efficacious and cost-effective treatment than combined arm for pemphigus. A study of larger samples size is warranted.

(342 words)

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Abstract N°: 3247

Unusual presentation of pemphigus vegetans

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Introduction & Objectives:

Pemphigus is a group of autoimmune blistering disorders mediated by circulating and in vivo bound autoantibodies against desmosomal proteins. Pemphigus vegetans (P Veg) is a rare variant of pemphigus vulgaris, comprising up to 1-2% of all pemphigus cases. Based on its clinical presentation and course, P Veg has been divided into the Neumann and Hallopeau type. The Hallopeau type of P Veg appears with polycyclic pustules that gradually evolve into verrucous and vegetating plaques usually located in the intertriginous areas. In rare cases, Hallopeau type of P Veg can start as solitary lesions at unusual sites which makes the diagnosis challenging. We report a case of Hallopeau type of P Veg with an uncommon manifestation that caused delay in the diagnosis and treatment.

Materials & Methods:

A 68-year-old man presented with a solitary, large, well-defined verrucous and vegetating plaque involving his right foot and toes with no history of preceding trauma. The lesion, which had gradually appeared 2 months before, was initially interpreted as pyoderma for which the patient was admitted to the septic surgery ward and underwent ineffective treatment with broad-spectrum antibiotics and topical antiseptics. The patient was further directed to dermatologic care. On physical examination, apart from the hypertrophic verrucous vegetating plaque, there was verrucous paronychia and toenail dystrophy with first toenail loss. Pustular lesions were present at the periphery. Careful examination of the mucous membranes revealed a cerebriform tongue and bilateral eroded vegetations at the mouth angles. Additionally, the patient suffered from thrombophlebitis and gouty arthritis. The diagnostic procedures included routine laboratory tests, histology, direct immunofluorescence (DIF) microscopy and immunoserologic investigation.

Results:

Routine laboratory tests revealed mild normocytic anaemia, mild leukocytosis, and elevated erythrocyte sedimentation rate. Histologic examination demonstrated suprabasal acantholytic cleft formation and spongiosis. DIF showed intercellular deposition of immunoglobulin G (IgG) and complement C3 in the epidermis. Indirect IF on monkey oesophagus substrate detected circulating anti-epithelial cell surface IgG antibodies at a titer of 1:160. ELISA anti-desmoglein (Dsg) tested strongly positive for Dsg1 and Dsg3. The patient was treated with oral prednisolone 0.5 mg/kg, azathioprine 100 mg daily, and topical clobetasol propionate ointment. This led to progressive improvement of the vegetating lesions and to a clinical remission on minimal therapy for the next two years of follow-up.

Conclusion:

Unlike "classic" pemphigus vulgaris, P Veg, especially the Hallopeau type, has no clinical resemblance to a vesiculobullous disorder, which is why it may remain unrecognized for a long time. It presents as accumulated vegetating and verrucous plaques mainly in the flexures. Oral lesions are common. Other areas are rarely involved, though reports of lesions limited to the foot or toes have been described, as in our case. The differential diagnosis

may include Hailey-Hailey disease, acne inversa, pyoderma gangrenosum, condylomata lata in secondary syphilis, acanthosis nigricans, and pyodermatitis-pyostomatitis vegetans. Further studies should be conducted to elaborate on the atypical nature of this unusual case.

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Abstract N°: 3305

Dose Selection of Upadacitinib in Non-Segmental Vitiligo (NSV): Pharmacokinetic and Exposure-Response Analyses of a Phase 2 Study in Subjects with NSV

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Introduction & Objectives:

Upadacitinib (UPA) is an oral selective Janus kinase inhibitor approved for several autoimmune diseases, including Atopic Dermatitis. The safety and efficacy of UPA was evaluated in adult subjects with extensive non-segmental vitiligo (NSV) in a single Phase 2 dose-ranging study (NCT04927975). The study evaluated dosing regimens of UPA 6 mg QD, 11 mg QD, and 22 mg QD in a placebo-controlled manner through Week 24, followed by a blinded extension through Week 52.

Herein, we present the pharmacokinetics of UPA as well as exposure-response relationships for efficacy and safety in subjects with NSV.

Materials & Methods:

A two-compartment population pharmacokinetic model with combined first- and zero-order absorption for the extended-release formulation was used to describe the pharmacokinetics of UPA in subjects with NSV.

Exposure-response analyses for efficacy and safety were conducted by relating the UPA average plasma concentration to select efficacy and safety variables. These analyses included graphical assessments and regression modeling at Week 24 (placebo-controlled period) and Week 52 (blinded extension). The models were used to additionally evaluate the predicted efficacy for 15 mg QD and 30 mg QD regimens that were not evaluated in the clinical trial.

Results:

UPA pharmacokinetics in subjects with NSV were similar to the pharmacokinetics previously observed in adult subjects with rheumatoid arthritis, atopic dermatitis, ulcerative colitis, and Crohn's disease.

Exposure-response analyses for efficacy during the 24-week placebo-controlled period showed that increasing UPA average plasma concentration was associated with increasing response for percent change from baseline (CFB) in facial vitiligo area scoring index (F-VASI), percent CFB in total vitiligo area scoring index (T-VASI), achievement of ≥ 75% improvement in F-VASI (F-VASI 75) and achievement of ≥ 50% improvement in T-VASI (T-VASI 50). In the Week 24 analysis a linear exposure-response relationship best described the relationship between UPA average plasma concentration and percent CFB T-VASI and percent CFB F-VASI, whereas a logarithmic model best described the relationships for F-VASI 75 and T-VASI 50. Simulations based on the models for Week 24 estimated that there was limited additional benefit across all the evaluated endpoints for a dose higher than 15 mg QD.

Exposure response relationships for percent CFB F-VASI and percent CFB T-VASI at Week 52 showed that the response increased with increasing UPA average plasma concentration, with the response approaching a plateau

at plasma exposures associated with the 15 mg QD regimen.

No exposure-response trends were observed for lymphopenia (≥ Grade 3), neutropenia (≥ Grade 3), hemoglobin < 8 g/dL, a > 2 g/dL decrease in hemoglobin from Baseline, serious infections, herpes zoster and pneumonia at or through Week 24 or Week 52, given that there were limited (< 10) events observed for each safety variable.

Conclusion:

These analyses demonstrated that UPA plasma exposures associated with a 15 mg QD regimen are predicted to provide a favorable benefit-risk profile, thereby supporting the evaluation of UPA 15 mg QD in the Phase 3 program for NSV (NCT06118411).

Figure 1. Observed and Model-Predicted Exposure-Response Relationships for Efficacy Endpoints at Week 24

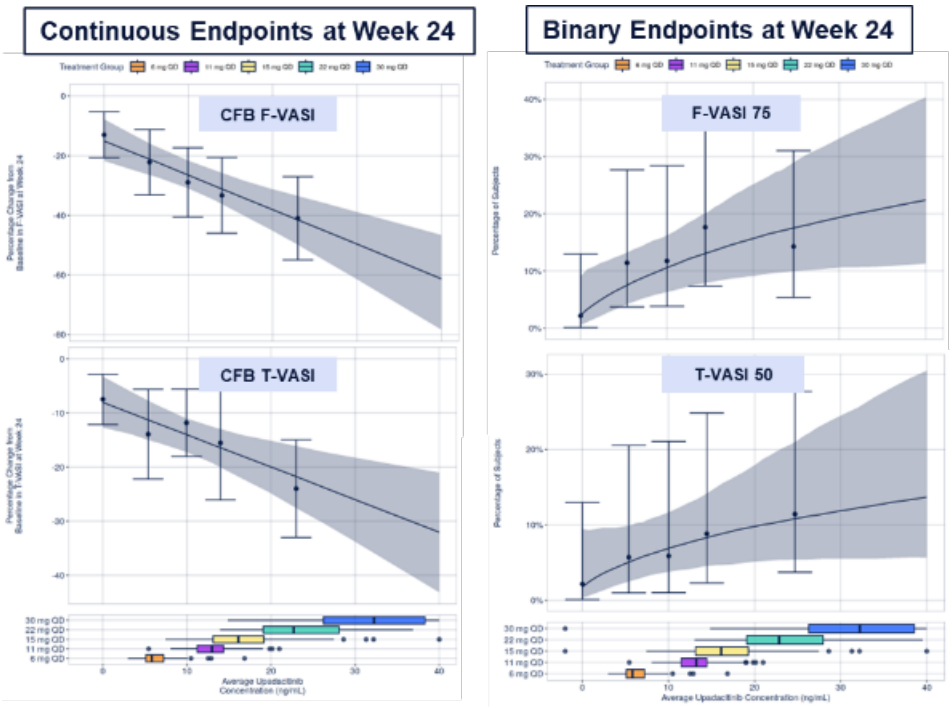
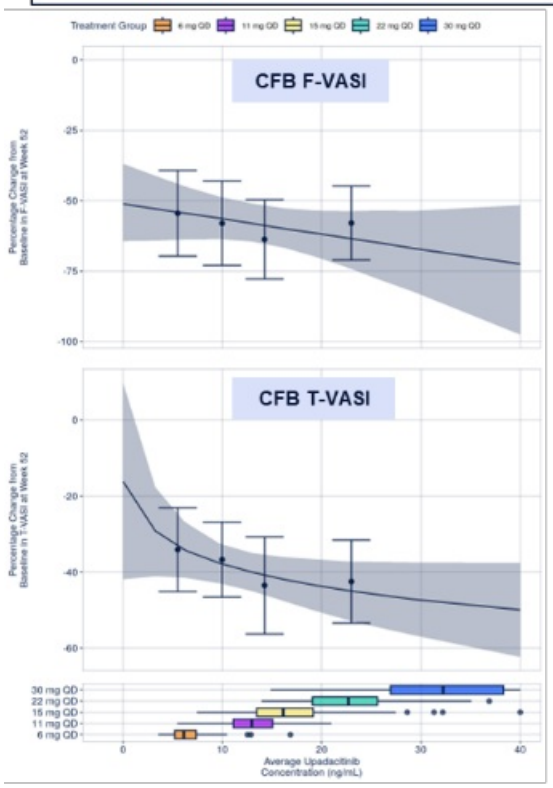


Figure 2. Observed and Model-Predicted Exposure-Response Relationships for Efficacy Endpoints at Week 52

Continuous Endpoints at Week 52



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Abstract N°: 3405

A case of immunoglobulin G4-related disease that pruritic skin lesions preceded 9 years to the diagnosis and that lymphadenopathy became apparent leading to the diagnosis while receiving dupilumab

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Introduction:

Immunoglobulin G4-related disease (IgG4-RD) is a systemic fibro-inflammatory condition characterized by tissue infiltration of IgG-4 expressing plasma cells and an elevation of serum IgG4 level affecting multiple organs. Skin manifestations are considered to be divided into primary lesions with direct infiltration of IgG4+ plasma cells and secondary non-specific inflammatory lesions, and can be the first overt clinical symptom. Dupilumab (DUP) is a monoclonal antibody that act on the interleukin (IL)-4 receptor alpha and inhibits IL-4/13 signaling resulting anti-type 2 inflammation. Several cases of IgG4-RD complicated with type-2 inflammation diseases treated with DUP have been reported and its efficacy to IgG4-RD is under discussion.

Results (case report):

A 55-year-old man suffered from pruritic skin lesions for 6 months and visited our department. He revealed erythematous plaques and multiple prurigo-like nodules almost over the body especially on his scalp, face, back and abdomen. He had diabetes mellitus and was well controlled by treating with linagliptin and metformin hydrochloride. Since his father also had itchy skin lesions and his blood test showed IgE 790ml/IU and high levels of IgE radioallergosorbent test for cider pollen, *Dermatophagoides pteronyssius and farina*, we considered him as late-onset atopic dermatitis at that moment. We treated him with oral antihistamines, topical corticosteroids, narrow band UVB phototherapy but he did not improve. Skin biopsy was done and histological findings revealed non-specific. 7 years later from his initial visit, he suffered from idiopathic thrombocytopenia, and was treated with oral prednisolone initially at 20mg daily and eltrombopag olamine. During oral corticosteroid treatment, his skin manifestations had been improved but recurred after stopping it. Since it was very refractory and his quality of life was poor, we started the administration of DUP. After 2 months, his skin manifestations improved, but cervical lymph nodes got swollen elastic hard sized to 3cm. CT showed also enlargement of mediastinal, supraclavicular, axial and inguinal lymph nodes and slight interstitial infiltration in lung. Cervical lymph node biopsy showed interfollicular expansion and infiltration of IgG4+ plasma cells (IgG4/Ig4 >80%). Serum IgG4 level was 651mg/dl. Therefore, diagnosis was changed to IgG4-RD. DUP was stopped and serum IgG4 level decreased to 369mg/dl and lymphadenopathy improved gradually but incompletely. Then, oral prednisolone was added at 20mg daily.

Conclusion:

A number of underlying diseases for refractory chronic generalized eczema in the middle-aged and elderly are known. IgG4-RD should be considered as one of them. In addition, the majority of the reported cases of IgG4-RD administered DUP showed favorable outcomes, but our case showed opposite response.

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Abstract N°: 3450

Difficulties in diagnosing the erosive-ulcerative form of vulvar lichen sclerosis.

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Introduction & Objectives: Vulvar lichen sclerosis (LS) is a chronic T-cell-mediated inflammatory dermatosis. The erosive and ulcerative form of the disease is the rarest. It is characterized by spontaneous formation of bleeding painful erosions or ulcerative defects on the background of hyperemia and atrophy of the mucous membrane without previous formation of blisters.

Materials & Methods: Patient's outpatient record, evaluation of anamnestic and laboratory data.

Results: Woman, 49 years old, visited a dermatologist in December 2023 with complaints of rashes, itching, pain in the anogenital area, discharge accompanied by an unpleasant odor. These symptoms had been bothering the patient for about 1 year. The patient was consulted by gynecologist and was diagnosed with acute vaginitis, prescribed suppositories neomycin + nystiatine + prednisalone + ternidazole intravaginally once a day -10 days, without effect. Also prescribed suppositories with clotrimazole intravaginally 1 once a day -8 days without effect. A smear was taken for cytology: cytogram of inflammation. Cytogram corresponds to cervicitis. Intraepithelial lesions and malignant processes are absent. Associated chronic diseases: Intramural uterine myoma. Cystocele. Iron deficiency anemia of 1 degree. Bronchial asthma. Allergoananmesis is not aggravated.

On examination the skin process had a localized chronic character. On the skin of the external genitalia in the vulva area with transition to the perianal zone, it was represented by white plaques with marked infiltration, scales on the surface, erosions and ulcers 0.5-0.7 cm in diameter with an irregular edge. The labia minora and clitoral complex are partially reduced. The skin in the foci is thickened. Diagnostic biopsy of vulvar skin was performed: epidermis is somewhat thickened, hyperkeratosis, formation of subepidermal slits is noted. Edema and thickening of collagen fibers in the upper third of the dermis. There is moderate histiolymphocytic infiltration around the vessels localized at the border between the papillary and reticular layers of derma. Conclusion: pathologic changes correspond to sclrosus lichen. Examination for sexually transmitted infections: HPV type 51 was detected. Therapy was recommended: mometasone furoate cream with gentamicin, econazole and dexpanthenol 2 times a day for 10 days, then clobetasol propionate ointment 0.05%, 1 time a day for 1 month, then every other day for 1 month, then 2 times a week for a month. From the first day emollients, 40 minutes after topical glucocorticosteroids 1 time a day. With positive dynamics. On the background of treatment in a month the patient noted a significant reduction of subjective symptoms. Also, significant clinical positive dynamics was noted.

Conclusion: at present there are still difficulties in establishing the diagnosis of lichen sclerosis. In most cases, the average time from symptom onset to diagnosis is 3-5 years, during which time patients receive inappropriate, ineffective treatment and symptoms progress. There are rare descriptions of the erosive-ulcerative form of lichen sclerosis in the literature. With this clinical case, we wanted to remind about the diversity of clinical manifestations of LS and the importance of timely diagnosis.

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Abstract N°: 3472

Off-label Treatment of Chronic Cutaneous Lupus with tacrolimus 0.1% ointment and roflumilast 0.3% cream: A Split Face Comparison Case Study

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Introduction & Objectives:

Cutaneous cutaneous lupus (CCLE) is a type of cutaneous lupus that causes pruritis, pain and can lead to scarring. It can be refractory to standard and off-label treatment, including topicals (TCS and TCI) and traditional systemics. It does impact quality of life, especially if in an area that is easily visible. Topical roflumilast cream 0.3% is a topical PE4 inhibitor which is thought to reduce inflammation by accumulating cAMP in skin cells, leading to increased signaling1; it is indicated for psoriasis. There has been one case report detailing success for CCLE, and anecdotal evidence. Here I report a successful split-face case study for the treatment of cutaneous lupus using tacrolimus 0.1% on the left side and roflumilast 0.3% cream on the ride side, leading to clinical and subjective improvements with both agents.

Materials & Methods:

A 65 year-old female with history for more than ten years of CCLE on the face was referred for a second opinion on topical treatment. Previous therapies included topical, intralesional and oral steroids, and systemics such as hydroxychloroquine, IVIG, and tofacitinib. She was also involved in several clinical trials. She was interested in further topical treatments of agents she had not tried. She was prescribed topical tacrolimus 0.1% ointment to use on the left side of the face and topical roflumilast to use on the right side of the face, both daily. She was reassessed at two months and five months with photos taken with her consent at baseline and these intervals.

Results:

Improvement of CCLE was achieved with both topical tacrolimus 0.1% and roflumilast 0.3% daily. The roflumilast was better tolerated.

Conclusion:

Topical roflumilast 0.3% cream appears to be as effective as TCIs for the off-label treatment of CCLE. It is a safe option with better tolerability compared to TCIs and should be considered more often in patients with recurrent or refractory CCLE.

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Abstract N°: 3477

pustular variant of lichen planopilaris: a diagnostic challenge

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Introduction & Objectives: lichen planopilaris (LPP) is a primary lymphocytic cicatricial alopecia that mainly involves the vertex of the scalp. It presents clinically as patches of alopecia with perifollicular erythema and scales. The presence of pustules in LPP is a rare clinical finding that was only reported in few cases in literature. This pustular variant of LPP can be easily misdiagnosed as folliculitis decalvans, bacterial folliculitis or erosive pustular dermatosis. Delayed diagnosis and treatment can lead to more extensive progression of the fibrosing alopecia. In this study, we aim to spot the light on this variant through a case series of 6 patients of LPP presenting with perifollicular pustules on the scalp.

Materials & Methods: Six patients of LPP, presenting with pustules on the scalp, were evaluated regarding the demographic, clinical, dermoscopic, and histological data. Laboratory investigations were done including blood picture, renal and liver function tests, hepatitis C virus markers (HCV), and thyroid profile.

Results: The case series included 4 female and 2 male patients; age ranged from 47-65 years old. The duration of hair loss ranged from 4-20 years. The most presenting symptom was scalp itching in all patients 6/6 while burning and pain was reported in 3/6 patients. The entire scalp was involved in 4 cases while 2 patients had LPP in the vertex and frontal scalp only. Laboratory investigations demonstrated HCV infection in a single patient and hypothyroidism in two patients. Dermoscopic examination showed erythema, mottled pigmentation, perifollicular tubular scale, perifollicular pustules, and crusts in all patients. Histological examination displayed perifollicular fibrosis and lymphocytic infiltrate in all subjects while 2 cases had perifollicular neutrophilic infiltrate.

Conclusion: The pustular variant of LPP represents a diagnostic challenge. Further studies are required to investigate the pathogenesis of this presentation. Dermatologists should be aware of this unique variant for proper and early management of the disease.

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Abstract N°: 3521

A Case of Grover's Disease in an 85-year old Caucasian responding to Dapsone

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Introduction & Objectives:

Transient acantholytic dermatosis, or Grover's disease (GD), is a rare pruritic papulovesicular eruption that occurs on the trunk and proximal extremities of elderly patients. In some patients, disease course may resolve spontaneously. However, in other cases, this condition may be chronic, relapsing, and difficult to treat. The main etiologic factor is unknown but certain trigger factors have been identified, such as heat and sweat, certain immunomodulators, end-stage renal disease, and even hematologic malignancy. Treatment includes emollients, topical corticosteroids, topical vitamin D analogies, oral antihistamines, oral retinoids, and oral corticosteroids.

Materials & Methods:

We report a case of an 85-year Caucasian who had a 2-month history of erythematous papules and flaccid vesicles on the anterior trunk. Lesions were moderately pruritic and are aggravated by heat & sweat. He initially self-medicated with various anti-itch creams with minimal improvement. Punch biopsy was done and revealed parakeratosis with collection of rounded keratinocytes with pyknotic nuclei and elongated keratinocytes in the stratum corneum. Suprabasilar and intraepidermal clefting are noted. There is acantholysis of the keratinocytes resembling a dilapidated brick wall. DIF was negative. Transient acantholytic dermatosis was highly considered. The patient was screened for all other comorbidities and was noted to be unremarkable.

Results:

Patient was initially given topical and oral corticosteroids. This treatment however failed to give good control of his condition. As steroids were tapered off, lesions would recur. G6PD level was screened and showed to have normal results. Dapsone was then started at 100mg/day and had good control his condition within 2 weeks. The patient maintained on it for 2 months and has been asymptomatic since then.

Conclusion:

This case highlights dapsone as an effective alternative treatment of choice of patients diagnosed with transient acantholytic dermatosis, or Grover's disease who are not responding to the first line treatment of choice. There are very limited treatment options for GD, and Dapsone has been reported to be an effective alternative treatment of choice.

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Abstract N°: 3569

Unraveling drug-associated bullous pemphigoid pathophysiology with lymphocyte transformation tests

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Introduction & Objectives:

Although the etiology of bullous pemphigoid (BP) remains unclear, it has been associated with the exposure to nearly one hundred different medications. However, this association is based on epidemiological studies relying on temporal relationships, with limited evidence of the underlying immunological mechanisms. The objective of this study is to explore the value of in vitro tests in identifying drugs related to BP development.

Materials & Methods:

We have conducted a prospective study including patients with a newly diagnosis of bullous pemphigoid. Three months after the diagnosis, lymphocyte transformation tests were performed, with a clinical follow-up extending to at least six months.

Lymphocyte transformation tests (LTT) are the standard in vitro test for diagnosing delayed hypersensitivity reactions to drugs. Patient's mononuclear cells are incubated with the suspected drug for six days, followed by H3-thymidine addition and quantification of the proliferation of drug-exposed T lymphocytes compared to unexposed T cells. A stimulation index equal to or above 3 was considered positive.

Results:

Eighteen patients have been included in the study, with a mean age of 78.5 years and a female-to-male ratio of 1:2. Prior to BP onset, eight patients (44.4%) had received treatment with dipeptidyl peptidase-4 inhibitors (DPP4i). An average of 2.1 drugs per patient were tested in the LTT, including DPP4i, other antidiabetics, diuretics, antihypertensives, lipid lowering agents, anticonvulsants, and immunotherapy. LTT was positive in 10 patients (55.6%): 3 for DPP4i, 2 for thiazides, and one each for a loop diuretic, nivolumab, levetiracetam, enalapril and amlodipine.

Among the 10 patients with positive LTT results, in four cases the culprit drug was discontinued, achieving clinical remission in 75% of them at the 6-month follow-up. Conversely, among the six cases where the positive drug identified in LTT was not discontinued, only a 33% remission rate was observed at 6-month-time. Nevertheless, the small sample size of our study limits the ability to identify significant associations.

Conclusion:

Our findings provide objective evidence confirming the existence of an immune reaction following exposure to certain drugs in BP patients. Conducting lymphocyte transformation tests could help identify drugs potentially associated with BP development, contributing to better understanding its etiopathogenesis and subsequently

improving the management of these patients.

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Abstract N°: 3574

A challenging patient with Inflammatory bowel disease associated pyoderma gangrenosum in pregnancy, with good response to infliximab and maintenance hydroxychloroquine, and review of the literature

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A challenging patient with Inflammatory bowel disease associated pyoderma gangrenosum in pregnancy, with good response to infliximab and maintenance hydroxychloroquine, and review of the literature

Introduction & Objectives:

Materials & Methods:

Results:

Conclusion:

Pyoderma gangrenosum is a type of neutrophilic dermatosis that is a diagnosis of exclusion. Pyoderma gangrenosum presenting during pregnancy remains rare, and treatment modalities can be challenging in gravid patients.

We present a rare case of pyoderma gangrenosum in a pregnant patient in her third trimester with concurrent newly diagnosed severe ulcerative colitis, that was successfully treated with infliximab, systemic steroids, with subsequent uncomplicated delivery and in remission on maintenance hydroxychloroquine therapy. We highlight the importance of multidisciplinary care in the management of such patients.

A 33-year-old female with a strong family history of autoimmune disease and personal history of quiescent seropositive rheumatoid arthritis on baseline hydroxychloroquine was first referred to the dermatology department for suppurative ulcerative nodules and papulopustules on her scalp, arms and legs for the past 1-2 weeks. (Figure 1,2) This was associated with systemic symptoms of fever, severe abdominal pain, nausea and vomiting and bloody diarrhea. She had otherwise no conjunctivitis or active synovitis. On examination, she was also noted to have pathergy over her intravenous cannula sites.

A diagnostic skin biopsy revealed an erosion with a predominantly neutrophilic infiltrate, with no evidence of vasculitis. (Figure 3) Infective stains for gram stain, giemsa, periodic acid-schiff, fite, ziehl nelson stain were all negative. Direct immunofluorescence was negative. Tissue cultures for bacteria, fungal and atypical mycobacteria were also unyielding. In view of initial possible differentials of IgA pemphigus, blood tests for desmoglein 1 and 3 were also performed, which returned negative.

She was seen urgently by our gastroenterology colleagues and underwent an emergency flexible sigmoidoscopy as tolerated, which revealed extensive circumferential colitis with exudates, with 2 deep ulcers seen in the rectum. A diagnosis of acute severe ulcerative colitis was made, and she was immediately started on intravenous hydrocortisone and infliximab infusion, which was maintained at 400mg per week. Histology from the ulcers subsequently revealed chronic moderately active colitis with cryptitis and crypt distortion. No granulomas, viral inclusion bodies, parasites or neoplasia were seen.

With the initiation of induction immunosuppressant therapy, her ulcerative colitis and suppurative skin ulcers all resolved rapidly with good clearance of the initial cutaneous eruption. She was continued on mesalazine therapy

by gastroenterology with remission of her ulcerative colitis. After discussion with the patient and rheumatology, she was continued on oral hydroxychloroquine, with her arthritis and pyoderma gangrenosum remaining quiescent. The patient also proceeded to deliver her fetus uneventfully by normal vaginal delivery, with close monitoring.

Here, we evaluate the literature with regards to the management and treatment of inflammatory bowel disease associated pyoderma gangrenosum, and summarize the options available in special cohorts, such as pregnancy as in our patient, and review the evidence.

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Abstract N°: 3587

Improvement of Severe Scleroderma Cases with Lung and Heart Involvement Treated Using Methotrexat

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Introduction & Objectives: Scleroderma or also known as systemic sclerosis (SSc) is a systemic autoimmune rheumatic disease that is quite rare, characterized by dysregulation of the immune system, vasculopathy, skin fibrosis and involvement of visceral organs such as peripheral blood vessels, digestive system, cardiopulmonary and kidney. The cause of scleroderma is not fully known but is thought to occur due to environmental triggers in individuals with genetic susceptibility who then experience activation of the inflammatory cascade, auto-amplification of autoantibody production, vascular changes (vasculopathy), and fibrosis tissue formation.

Materials & Methods: Female patient 54 years old, complained that both hands, feet and facial areas were said to have stiffness. Inspection of the skin found hypopigmentation and hyperpigmentation with a generalized "salt & pepper" appearance. From palpation, the skin are hard resembling wood or boards in the upper and lower extremities, truncus and facial areas. Complaints were also accompanied by shortness of breath and nausea since 1 month ago and aggravated the last 2 days, causing the patient to be taken to the emergency unit and received oxygen therapy. X-ray examination of the thorax shows a picture of the infiltrate in the upper-middle field of the right and left lung. The results of echocardiography examination obtained mild degree mitral regurgitation and mild degree pulmonary regurgitation. Patients diagnosed with scleroderma with accompanying interstitial lung disease, chronic obstructive pulmonary disease, mitral regurgitation and congestive heart failure. The therapy given is methotrexate 15 mg/week, folic acid 10 mg/week, mometasone furoate cream on the face, and clobetasol propionate ointment on other skin areas. Other treatments are salbutamol nebules, methylprednisolone 125 mg/day with tapering down, lansoprazole, erdosteine, furosemide, spironolactone and bisoprolol.

Results: According to the classification of American College of Rheumatology (ACR)/European League Against Rheumatism (EULAR), patients have a total score of 16. Immunosuppressant therapy options that can be given to scleroderma patients include methotrexate (MTX). There are two types of MTX mechanisms of action, namely as an anti-proliferation mediated by folate-influenced pathways and an anti-inflammatory caused by elevated levels of aminoimidazole carboxamide ribonucleoside (AICAR). MTX will only be active in the S-phase of the cell cycle. MTX binds to dihydrofolate reductase (DHFR) with high affinity. MTX binds to DHFR and has a fairly high affinity for enzymes requiring folate cofactor, including thymidylate synthetase (TS) and 5-aminoimidazole-4-carboxamide ribonucleotide (AICAR) mylase transphore. TS inhibition, induced by MTX, interferes with DNA synthesis in actively dividing cells, and an increase in the AICAR enzyme system, which plays an important role in cell purine metabolism, leads to increased release of adenosine into the blood.

Conclusion: The patient is diagnosed with diffuse type scleroderma where there is involvement of major organs so that the patient has a poor prognosis. However, in the observations on patients for 1 month after therapy showed improvement of the condition of skin stiffness and reduced shortness of breath. MTX accompanied by systemic corticosteroids can be an inexpensive treatment option even in severe scleroderma conditions.

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Abstract N°: 3741

Facial hyperpigmentation: Consider melanotic lupus!

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Introduction & Objectives:

Chronic cutaneous lupus erythematosus (CLE) can present in multiple clinical variants. The most common form is discoid lupus. Melanotic lupus (ML) is a rare form of CLE recently described.

Materials & Methods:

Since 2019, the diagnosis of ML has been made in 3 patients in our dermatology department.

Results:

Our patients were aged 25, 49, and 53, respectively. They all had skin type IV. The median duration of evolution was 7 months (45 days – 9 years). The latter patient only sought consultation after developing a squamous cell carcinoma of the lower lip. Patients presented with poorly defined slate-gray hyperpigmentation, oval in 2 cases and diffuse in one case. It affected the face (3 cases), neck (1 case), and forearms (1 case). Pruritus was present in one case. One patient described preceding erythema before hyperpigmentation. Two patients reported photosensitivity and polyarthralgia. Two patients had frontal alopecia with thinning of eyebrows in one case. One patient had cheilitis complicated by carcinoma of the lower lip. Dermoscopy in 2 patients showed brown globules and perifollicular peppering, with erythema observed in one case. Anti-nuclear antibody (ANA) levels were positive and speckled in all cases, associated with positive native anti-DNA and anti-SSA antibodies in one case. Histology showed interface dermatitis in all cases with presence of cornified tunnels, vacuolated basal layer, and peri-vascular and peri-adnexal lymphoplasmacytic inflammatory infiltrate with melanophages. Treatment with sunscreen, corticosteroids, and hydroxychloroquine (HCQ) only led to moderate improvement.

Conclusion:

ML is a newly described variant of CLE with only about thirty published cases. It occurs in subjects with medium to dark skin types and relatively advanced age. It can present as poorly defined hyperpigmented patches or diffuse hyperpigmentation usually affecting the face without scales, cutaneous atrophy, or telangiectasias. ML may be associated with photosensitivity in 50% of cases. It rarely associates with systemic involvement. The alopecic lesions and cheilitis complicated by squamous cell carcinoma observed in our patients are exceptional. Dermoscopic features are dominated by follicular signs and pigmentary structures. The pathogenesis of ML could be explained by pigmentary incontinence caused by interface dermatitis, especially in individuals with dark skin types. Management of ML is not well codified. Encouraging results have been obtained with corticosteroids and HCQ.

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Abstract N°: 3760

Alopecia universalis in children and adolescents – treatment challenges

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Introduction: Alopecia areata is a chronic autoimmune disease mediated by autoreactive T lymphocytes which affects hair follicles and nails. The disease occurs on a predisposing genetic background and represents a type of non-scarring, non-inflammatory and asymptomatic alopecia, encountered both in children (10-20% of cases) and adults. Alopecia universalis is the most severe form of the disease and clinically presents as a complete loss of terminal hair on the scalp as well as on the eyebrows, eyelashes and body.

Case report: We report the case of a female 14-year old patient, first diagnosed with Alopecia areata at the age of 1-year old, who have had multiple periods of remission and exacerbations since the diagnose. The patient presents in our Dermatology department for an almost complete loss of scalp hair, only presenting a few strands of hair in some areas of the scalp and total absence of eyelashes, eyebrows and body hair. Family history reveals that the patient's maternal uncle was diagnosed with Alopecia areata in adolescence The patient does not have any other significant medical history. Previous episodes of the disease have responded well to topical treatments and local immunotherapy with Diphenylcyclopropenone. The current episode started 8 moths ago and has been the most severe since now, having rapidly evolved from AA in patches to the universalis form. The patient underwent treatment with 3 courses of systemic corticotherapy (Methylprednisolone) at monthly intervals and local treatment with dermatocorticoids, vitamin D analogs, Minoxidil solution and vasodilatory agents with no favorable evolution until present. New treatments with JAK inhibitors could not be administered in Romania because it is not approved for the pediatric population until now.

Conclusion: The early onset in childhood and the family history of Alopecia areata provide a reserved prognosis of the case. Therapeutic options are limited to topical, intralesional or systemic corticosteroids, immunosuppressive agents or induction of allergic contact dermatitis, all of which has not showed favorable results in this patient. Currently, innovative therapies with Janus kinase (JAK) inhibitors such as Baricitinib have been approved in Romania for the adult population with promising results, which could be applicable for the presented case in the future.

Key words: alopecia areata, alopecia universalis, adolescents, JAK inhibitors

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Abstract N°: 3762

Linear IgA Bullous Dermatosis : A series of 11 cases

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Introduction & Objectives:

Linear IgA bullous dermatosis (LABD) is a rare subepidermal autoimmune bullous disease that affects both children and adults. Our objective was to determine the clinical characteristics, treatments, and outcomes of this disease.

Materials & Methods:

This is a retrospective study including all cases of LABD confirmed by histopathology and direct immunofluorescence over a period of 17 years (2005-2022).

Results:

We included 11 patients: 5 adults and 6 children. The median age was 13 years with a sex ratio (M/F) of 1. Clinically, tense vesicles or bullae on a background of erythematous or non-inflamed skin with the classical “string of pearls” arrangement was seen in all cases. The limbs and trunk were the most commonly involved sites (90% of cases), particularly in adults. Genital and perioral involvement was observed in children. Mucosal involvement was seen in 60% of cases. Nine cases (82%) were idiopathic while 2 cases were triggered by amoxicillin with a latency time from the intake of 4 and 7 days. Dapsone was used in 60% of cases. In the other cases, the treatment was systemic corticosteroid therapy and stopping treatment in drug-induced forms. The evolution was favorable in all cases.

Conclusion:

LABD is an autoimmune bullous disease, with a bimodal age of onset, characterized immunopathologically by linear deposition of IgA at the basement membrane zone. Clinically, LABD is characterized by an eruption of tense vesiculo-bullae grouped in clusters, giving an appearance of a ‘‘string of pearls”. The anogenital and perioral regions are the favorite sites in children against the trunk and the limbs in adults. Several associated pathologies have been described such as autoimmune and inflammatory bowel diseases. Several drugs have been implicated, especially in adults, in particular vancomycin, amoxicillin and cephalosporin. The standard treatment is dapsone. However systemic corticosteroid therapy has given good results.

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Abstract N°: 3828

The quality of life in patients with alopecia areata in Romania

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Introduction & Objectives: Alopecia areata is a skin disease that impairs the quality of life in patients across different severity levels, sexes, and age groups. In addition to assessing the effects of alopecia areata on quality of life, the EQ-5D-5L data can be used to compute utilities which are further used in economic evaluations (necessary to assess the most cost-effective option among two or multiple concurring therapeutic alternatives). To the best of our knowledge, this is the first study evaluating the quality of life in patients with alopecia areata in Romania.

Materials & Methods: We analyzed quality of life data collected using the EQ-5D-5L instrument from 38 patients with complete EQ-5D-5L data (out of 41). In addition, we also analyzed other variables, such as age, sex, time since diagnosis, and severity levels. We computed descriptive statistics and ANOVA.

Results: Most patients were female (n = 30), had an S1 (characterized by between 1% and 24% hair loss) level (n = 9), were aged over 36 years (n = 22). Utilities ranged between 0.205 and 1 (female), and 0.705 and 0.909 (male). Level frequency scores and other descriptive statistics are presented in tables 1 and 2. No statistically significant differences in utilities were found between severity levels, sex, age, and time since diagnosis.

Conclusion: Most patients that exhibit pain/discomfort and anxiety/depression reported scores over one. This points out to the need for future studies that, in addition to measuring quality of life, are also including instruments that focus on mental health problems.

Table 1. Level frequency scores

Scores	*Mobility*	*Self-care*	*Usual activities*	*Pain/Discomfort*	*Anxiety/Depression*
1	26	25	19	6	7
2	5	9	6	16	10
3	3	3	9	13	13
4	4	1	4	2	6
5	0	0	0	1	2

Table 2. Descriptive statistics

Variable	*Min*	*Max*	*Mean*	*Median*
Level sum scores	5	20	10.03	9.5
Utilities	0.205	1	0.8037	0.8327
VAS	3	90	55.55	65

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Abstract N°: 3832

Assess Disease End Point In Pemphigus Vulgaris Patients Treated With High Dose (1000mg) And Low Dose (500mg) Of Rituximab

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Introduction & Objectives:

Pemphigus Vulgaris (PV) is the commonest one in a group of chronic autoimmune blistering diseases characterised by the presence of pathogenic antibodies directed against desmosomal adhesion proteins, desmogleins 1 and 3 (Dsg1&3).

Rituximab a chimeric human-mouse monoclonal IgG1 antibody against CD20 B lymphocytes cell surface antigen has been shown to be effective in the management of pemphigus vulgaris.

To assess the late endpoints of disease activity in pemphigus vulgaris treatment with low dose of Rituximab 500mg and high dose of Rituximab 1000mg as (1) complete remission off therapy (2) complete remission on therapy (3) partial remission (4) relapse or flare

Materials & Methods:

Case history and thorough clinical examination with Pemphigus Area and Activity Score (PAAS) was done in all 68 patients as per case record form. Direct Immunofluorescence (DIF) and Enzyme-linked Immunosorbent assay (ELISA) of antibodies to Dsg 1 & 3 were evaluated at baseline and subsequent follow up at 3 ,6, and 12 months. Rituximab 500 mg or 1000 mg in 500ml normal saline was given at two weeks apart. All patients were reassessed clinically and serologically (Dsg 1&3) at regular intervals at 3,6 and 12 months after completion of second infusion. Late endpoints of disease activities at 1 year were assessed as complete remission (off or on therapy), partial remission (off or on therapy), treatment failure and relapse. The analysis of the data was done by paired T Test.

Results:

Patients treated with 1000 mg of Rituximab, 7 (33.3%)of the patients were in complete remission and off therapy, 9 (42.8%) of the patients were in complete remission on minimal therapy (2.5-5mg prednisolone, 5 (23.8%) of the patients were in partial remission , no patients had relapse or treatment failure. Patients treated with 500 mg of Rituximab, 9 (19.1%) of the patients were in complete remission and off therapy whereas 21 (44.6%) of the patients were in complete remission on minimal therapy (2.5-5mg prednisolone), 6 (12.7%) of the patients were in partial remission, 5 (10.6%) patients relapsed and 6 (12.7%) patients had treatment failure. The mean time to relapse was 6.6 months

Difference in the fall of PAAS was statistically significant at 3, 6 and 12 months ($P<0.05$, Confidence Interval of 95%) for patients who received 1000mg and 500 mg of Rituximab in complete remission and complete remission with minimum treatment group of patients. PAAS was statistically not significant at 3,6 and 12 months ($P>0.05$) for patients who received 1000mg and 500 mg of Rituximab in partial remission group of patients.

Difference in the fall of Dsg1&3 was statistically significant at 3,6 and 12 months ($P<0.05$ Confidence Interval of 95%) for patients who received 1000mg and 500 mg of Rituximab in complete remission and complete remission

with minimum treatment group of patients. Dsg1&3 was statistically not significant at 3,6 and 12 months (P >0.05) for patients who received 1000mg and 500 mg of rituximab in partial remission group of patients.

Dsg1&3 was not significantly reduced in treatment failure and relapse group of patients treated with 500mg of Rituximab at 12 months. (P>0.05)

Conclusion:

Decline in Dsg 1&3 titres and PAAS with high doses 1000mg of Rituximab in initial 3 months of treatment implies that early control of disease activity with beginning and end of the consolidation phase of disease, which indicate dose of steroid can be reduced early. Quality of life of patients with life threating comorbidities can be upgraded.

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Abstract N°: 3861

Hypertrophic discoid lupus erythematosus successfully treated with photodynamic therapy, a case report.

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Introduction & Objectives: Lupus erythematosus (LE) is a multisystemic disorder that predominantly affects the skin. There are several types of cutaneous lupus and discoid erythematosus lupus (DLE) is one of the most common subsets. This type of lesions are difficult to treat and sometimes refractory to different therapies.

Materials & Methods:

We present the case of a 63-year-old woman diagnosed with DLE at the age of 36, under follow-up by dermatology and rheumatology. She presented a large, atrophic, erythematous-crusty plaque in the right pretibial region and other in the upper lip, which had been treated with multiple therapies over the years. In 2018, despite treatment with clovate, hydroxychloroquine 200mg daily and oral methotrexate, the lesions persisted without improvement. After reviewing the literature, it was decided to start with red light photodynamic therapy with methyl aminolevulinate. 15 sessions were performed throughout 2018 and 2019 with very good response and resolution of the lesions. Currently the patient remains without lesions.

Results: DLE is the most common form of chronic cutaneous erythematosus lupus. It is unusual for discoid lesions to present below the neck. Sometimes they exhibit a photodistribution, so sun exposure seems to play a role in its development.

The first morphological sign is a well-defined, annular erythematous plaque, followed by follicular hyperkeratosis, which is adherent to the skin. The lesions slowly expand at the periphery, leaving central atrophy and scarring.** Hyperkeratotic plaques are noted in hypertrophic DLE, a rare entity, whose clinical course is generally marked by chronicity and resistance to therapy.

Early treatment may lead to the total clearing of skin lesions. Current first-line therapy consists of topical or intralesional corticosteroids and topical calcineurin inhibitors. Lesions which do not respond to topical therapy are candidates for systemic treatment. Antimalarials (hydroxychloroquine, chloroquine, quinacrine) are considered first-line systemic therapy. Immunosuppressive agents such as methotrexate, dapsone, mycophenolate mofetil, azathioprine, cyclophosphamide, and cyclosporine, have been trialed as second-line. Topical photodynamic therapy (PDT) has been applied to the treatment of an extensive range of skin diseases, however, reviewing pubMed, there are only 2 published cases of discoid lupus successfully treated this way. In animals PDT exhibit the ability of decreasing the number of lymphocytes and inactivate the activited ones. In psoriasis, the capacity of PDT to incude apoptosis in T cells has been shown. Infiltrating T lymphocytes are considered to play a major pathological role in DLE.

DLE may be induced or exacerbated by exposure to ultraviolet radiation, but not to other light length. Thus, there are previous case reports of DLE improvement with pulsed dye laser (595nm).

Conclusion:

The ability of PDT to alter the course of inmunologic and inflamtory diseases such as DLE is not well known, and there are just a few cases published with successful results, so this report may be useful, as dermatologists, for

cases that do not respond to conventional treatment.

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Abstract N°: 3937

Basophils: The Overlooked Players in Bullous Pemphigoid Inflammation

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Title: Basophils: The Overlooked Players in Bullous Pemphigoid Inflammation

Introduction & Objectives: Bullous pemphigoid (BP) is an autoimmune disease characterized by the formation of blisters and the infiltration of inflammatory cells. Most of previous studies suggested that the formation of a subepidermal cleft, accompanied by a significant eosinophil infiltration, is a prominent characteristic of BP. In addition to eosinophil and neutrophil infiltration, various other inflammatory cells and factors, including basophils, play a role in the pathophysiology of BP. Now few studies have demonstrated a significant increase in basophil numbers in skin inflammation, particularly in the bullous phase, suggesting their involvement in the pathogenesis and resolution of BP. However, the specific pathological role of basophils in BP remains largely unexplored. This study aimed to investigate the relative contributions of inflammatory cells in serum and blister fluid of BP.

Materials & Methods: Serum and blister fluid samples were obtained from patients diagnosed with bullous pemphigoid (BP) and toxic epidermal necrolysis (TEN). Cell classification was conducted using an automated hematology analyzer within a body fluid analysis framework. Inflammatory cells were phenotyped through the measurement of surface expression of CD45, while lymphocyte subsets were identified via flow cytometry. Cytokine profiles were analyzed through the examination of soluble mediators, and statistical analysis was performed using GraphPad Prism 7.0.

Results: Using flow cytometry, we found eosinophil and neutrophil, basophil infiltration in BP blister fluid. We found the expression of activation marker of CD4+ and CD8+ T cell in BP and TEN patients of blister fluid. Cytokine profile analysis across peripheral blood and blister fluid displayed increase IL-4, IL-5, IL-10, IL-6, IL-8 and CCL2 expression in BP patients. By contrast, in TEN patients, the highest increased cytokine were IL-6, IL-8 and especial higher IFN-γ. Detected the secretion of intracellular cytokines, we found basophil-derived IL-4, but not CD4+T lymphocytes, are the main IL-4 secreting cell types under no stimulation.

Conclusion: These findings highlighted basophils are involved in the development of BP. Besides eosinophil and neutrophil, basophil infiltration is a prominence feature during the bullous phase of BP. And type 2 inflammation responses was the most important immune characters in BP. Our above results showed an increased numbers of basophil and predominance of activated CD4+T cells in blister fluid. Basophil-derived IL-4 may play a more important role in naive T cell activation and type 2 inflammation responses.

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Abstract N°: 3986

Janus kinase inhibitor, tofacitinib, in the treatment of clinically amyopathic juvenile dermatomyositis

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Introduction & Objectives:

Juvenile dermatomyositis (JDM) is a rare systemic connective tissue disorder in the pediatric age group.

Materials & Methods:

We present an 18-year-old patient with no family history of note, who came to our clinic at the age of 13 with facial skin lesions, on the back of the hands, elbows, and knees of years of evolution. She did not present systemic symptoms.

Physical examination revealed malar and palpebral erythema, and papular lesions on the dorsum of the interphalangeal joints of the hands. Laboratory tests showed no abnormalities, with negative results for ANA, Anti SS-A, Anti SS-B, Anti RNP, Anti SCL-70, Anti SM… transaminases and muscle enzymes were normal. Given the pathognomonic cutaneous findings and a skin biopsy compatible with a connective tissue disorder, a diagnosis of JDM was made.

Magnetic resonance imaging (MRI) did not show muscle edema. In the absence of muscle symptoms, normal muscle enzymes, and EMG findings, it was classified as clinically amyopathic juvenile dermatomyositis (JDM) in its hypomyopathic variant. Specific myositis antibody testing (Anti-Jo1, Anti-MI2, MDA5, NXP-2, TIF-1) revealed no positivity.

Treatment with hydroxychloroquine was initiated, but was discontinued due to the appearance of grayish facial pigmentation and was replaced with subcutaneous methotrexate. After 6 months of methotrexate treatment, subcutaneous indurated lesions began to appear on the thighs. With suspicion of calcinosis, a bone series was performed, confirming the diagnosis, showing numerous subcutaneous calcifications in the pelvis, elbows, and lower limbs.

A new study of muscle involvement was requested, with both muscle enzymes and MRI being normal, except for the described calcifications. It was decided to initiate treatment with tofacitinib on compassionate use.

Results:

After 12 months, the cutaneous symptoms resolved, and the calcinosis lesions improved both clinically and radiologically.

Conclusion:

During the course of JDM, complications such as calcinosis may arise, hence the importance of early recognition for appropriate treatment initiation.Although more studies are needed, there are several publications supporting the use of JAK inhibitors to stabilize the lesions.

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Abstract N°: 4020

Painful plaques, pustules in folds and photosensitivity in a patient on infliximab: piecing the puzzle together

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Introduction & Objectives:

Neutrophilic lupus erythematosus has emerged as a proposed term to encapsulate the spectrum of neutrophilic dermatoses linked to systemic lupus erythematosus (SLE). Despite ongoing research, the precise pathogenesis remains unclear. However, there is mounting evidence implicating dysregulation of key cytokines within specific inflammatory pathways, including interleukin (IL)-1, tumor necrosis factor and type I interferon. Herein we report a patient with SLE who developed lesions of Sweet syndrome and aseptic flexural pustulosis.

Materials & Methods:

Case report and literature review.

Results:

A 23-year-old patient with a history of Crohn's disease treated with infliximab presented with abdominal symptoms resistant to prednisone, accompanied by fever and painful cutaneous lesions. Clinical examination revealed photodistributed erythema and scaling on the face, in addition to erythematous and edematous plaques on the extremities and pustules with exudative crusts in groin, axillae and scalp. Histopathological examination of skin lesions suggested features consistent with histiocytoid Sweet syndrome and plasmacytoid dendritic cell infiltration. Laboratory analysis revealed elevated inflammatory markers, diminished complement C3 levels and positive autoantibodies, including antinuclear antibodies (ANA) at 1/640 titre with a nucleolar pattern, anti-Ro antibodies and lupus anticoagulant. Furthermore, the patient was diagnosed with SLE and aseptic flexural pustulosis in addition to exacerbation of colonic Crohn's disease. Treatment necessitated a multidisciplinary approach, incorporating intravenous methylprednisolone, ciprofloxacin and metronidazole. Hydroxychloroquine was initiated and infliximab was discontinued due to potential exacerbation. Ustekinumab initiation led to improved Crohn's disease control, albeit with required dose adjustments for recurrent symptoms.

Conclusion:

This case underscores the unique challenge posed by the coexistence of Crohn's disease, SLE, histiocytoid Sweet syndrome and aseptic flexural pustulosis. It emphasizes the imperative of a holistic and multidisciplinary management approach. Additionally, our findings underscore the necessity for tailored treatment strategies, considering the individual autoimmune-inflammatory profile of each patient. Furthermore, previous infliximab exposure may have triggered disease exacerbation, while the observed positive response to ustekinumab suggests potential involvement of IL-12 and IL-23 pathways in these dermatoses.

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Abstract N°: 4021

Association of genito-urinary calculus disease and lichen planus: a study of 21 patients

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Introduction & Objectives: Lichen Planus (LP) is a common papulosquamous skin disorder whose underlying etiology is unknown but genetic and exogenous influences have been implicated. Infectious agents (Hepatitis C virus), chemicals (dental amalgams), and drugs are associated with some variants of LP. Here we have discussed a rare and interesting association between urolithiasis and LP observed in 10 patients.

Materials & Methods: Twenty one patients (16 male and 5 female; age range 20 to 62 yrs) were included in the prospective study from 2022 to 2023 in a single Dermatology setup with a single consultant. Clinical diagnosis included eruptive LP in 11, hypertrophic LP in 6, annular LP in 2, and oral LP in 2 patients. These patients had either a preceding or concomitant history of urolithiasis. All patients of LP were histologically proven and detection of urolithiasis was done by ultrasonography of the abdomen and X-ray of kidney, ureter, and urinary bladder.
All patients were seronegative for hepatitis C, VDRL and HIV. Patients did not have diabetes or any other systemic disorders.

Results: Interestingly, after successful management of urolithiasis, nineteen patients had complete resolution. In the other two, the disease frequency and intensity were significantly diminished. Thus removal of calculi resulted in either complete resolution in most cases or led to a significant reduction in morbidity of LP. Immunoreactive substances in stones, bacterial antigens over stones, low-grade UTI, or calculus could be the cause of LP.

Conclusion: Our study indicates urolithiasis may be an underlying inciting event for some patients of LP. Careful attention to systemic complaints of such patients is important. A dermatologist may be the first physician to suspect urolithiasis in certain patients of chronic and recurrent LP. Treatment of urolithiasis alone might be sufficient for resolution of LP in some patients.

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Abstract N°: 4047

Oropharyngeal involvement in patients with pemphigus vulgaris: experience in a tertiary hospital.

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Introduction & Objectives:

Oropharyngeal involvement in pemphigus vulgaris (PV) is often underdiagnosed due to its difficult access by physical examination. In previous studies, it seems that the existence of oropharyngeal involvement is associated with worse prognosis of the disease.

Therefore, the aim of our work is to describe the cases in our center with oropharyngeal disease, as well as, establish possible differences compared to subjects without oropharyngeal PV.

Materials & Methods:

Patients diagnosed with PV (according to the latest diagnostic criteria) in current or previous follow-up in our center between 2010 and 2023 were collected. Subjects with a diagnosis of active malignancy (except non-melanoma skin cancer) were excluded.

From the initial cohort of individuals, we selected those subjects who presented oropharyngeal involvement (defined as the presence of erosions on or beyond the tonsillar pillars).

Results:

Fifty-two individuals with PV were collected, of whom 17 (32.7%) had oropharyngeal involvement (13 females and 4 males). Of the 17 patients, all of them had oral mucosal involvement; 52.9% of the nasal mucosa; 41.2% of the genitourinary mucosa. Twelve subjects had concomitant skin involvement. The mean PDAI in the mucosa was 21.76 +/- 11.63, and the mean PDAI in the skin was 5.65 +/- 8.18. Eleven patients presented odynophagia over the course of the disease and seven of them dysphagia. Six patients were asymptomatic. Eight patients (47.1%) required hospitalization, mostly due to inability to eat. With regard to treatment, the mean maximum dose of prednisone required was 53.23 mg/day. Thirteen patients (76.5%) required rituximab, with an average of 1.87 cycles.

In comparison with patients without oropharyngeal involvement, collected in the same period, oropharyngeal involvement was associated with a statistically higher mean PDAI in mucosa and a lower mean PDAI in skin. Oropharyngeal involvement was more frequent in female subjects. Nasal involvement and involvement of the hypopharynx and larynx were also more frequent in subjects with oropharyngeal PV, but without statistical significance. There were no significant differences in the proportion of hospital admissions due to PV. Patients with oropharyngeal involvement received on average more cycles of rituximab but without statistical significance.

Conclusion:

In patients with PV, oropharyngeal involvement is frequent and seems to be associated with greater mucosal disease. Its utility as a predictor of response to treatment is not clear, and future studies with a larger sample size

are needed.

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Abstract N°: 4053

Localized Juvenile Scleroderma, Pansclerotic Subtype: A Case Report

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Introduction:

Localized juvenile scleroderma consists of a functional disorder of fibroblasts and autoimmunity resulting in collagen deposit, fibrosis, atrophy, and deformities. It presents in 4 subclassifications: linear morphea - the most prevalent; superficial circumscribed morphea; generalized morphea; and pansclerotic morphea. The latter is characterized by rapid progression of cutaneous fibrosis, involving subcutaneous adipose tissue and, eventually, fascia, muscles, and bones. Initially, it predominantly affects the extremities, and as the disease progresses, it involves the trunk, face, and scalp. The etiology is still undefined and under investigation; however, epidemiologically, it is known to primarily affect female children around the age of 8.

Case Report:

In this case report the patient was a 4-year-old boy, without known previous comorbidities. Family members reported that at the age of 1 he developed pruritic violaceous macules on the left lower limb, which later spread throughout the limb forming crusts. One year later they also reported that he began experiencing ankle arthralgia, associated with gait alteration, secondary to valgus flat foot and claw toe. At the time of the medical consultation, at 4 years of age, he presented diffuse cutaneous thickening, with areas of retraction of fibrotic components, including on the face and scalp, with the en coup de sabre. At this moment, the clinical diagnosis of pansclerotic localized juvenile scleroderma was made.

Discussion:

After diagnosis, he was hospitalized for pulse therapy with methylprednisolone 0.5 mg/kg/day and started on methotrexate 15mg/m^2/week, with relative improvement of the lesions. Chest and head CT scans were performed, which did not show systemic involvement, and he received clinical follow-up from the orthopedic, dermatology, and rheumatology services. Scleroderma can present as localized scleroderma (morphea) or systemic scleroderma. Juvenile localized scleroderma has an incidence of 0.34 to 2.7 cases per 100,000 children per year. Pansclerotic morphea is even rarer, and the delay in diagnosing its atypical form can worsen the morbidity and mortality of the condition. There is a possibility of neuropsychomotor development impairment with significant psychological and social repercussions due to reduced body mobility and gait impairment due to musculoskeletal system involvement. The relevance of studies on rare and debilitating conditions such as juvenile localized scleroderma aiming at early diagnosis and management of complications is emphasized. There are also reports that the disease progression may lead to the development of squamous cell carcinoma in affected patients. This reinforces the importance of periodic dermatological consultations for evaluation and follow-up of lesions suspected of malignancy.

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The Feverish Quest: Unmasking Kikuchi-Fujimoto's Peculiar Patterns

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Introduction & Objectives:

Kikuchi-Fujimoto disease is a rare cause of lymphadenopathy, mainly affecting young women, usually self-limiting, which can also be associated with fever, asthenia and weight loss, but also with cutaneous rash. We present a clinical case and brief literature review.

Materials & Methods:

We examined a 23-year-old man with a history of congenital hearing loss and cognitive delay presents with a one-month history of fever, associated with decreased C3 and positive antinuclear antibodies (ANA >1/1280), as well as visualized lymphadenopathy on computed tomography and a palpable left laterocervical lymph node measuring 2 cm in diameter, which persists despite medical antibiotic treatment, leading to a decision to perform a core needle biopsy, confirming a typical histological finding of Kikuchi-Fujimoto disease. One month later, he is evaluated by the dermatology service for the appearance of excoriated papules, some with central ulceration, on the forehead and auricular pavilions, as well as some less significant ones on the trunk and thigh. A skin biopsy was performed showing Civatte bodies and vacuolar interface dermatitis, findings compatible with cutaneous involvement of Kikuchi-Fujimoto disease. No medical treatment was necessary, as the fever and cutaneous manifestations had a self-resolving nature.

Results:

Regarding cutaneous involvement, although the rash is more frequent in children than in adults, other manifestations such as erythematous macules or papules, plaques, nodules, or ulcers have been reported

Cases of sweet-type cutaneous involvement or paraqueratosis have also been described, and it has frequently been associated with systemic lupus erythematosus, although the relationship between the two remains a matter of debate. For diagnosis, a biopsy of the affected lymph node is necessary, especially to perform a correct differential diagnosis with lymphomas and other infectious pathologies. Nodal histology shows a deficiency of neutrophils and eosinophils, with myeloperoxidase and CD68 +, CD8 + histiocytes, and minimal presence of B cells. Generally, it does not require treatment, and the prognosis is good. However, in severe cases, systemic corticosteroids or immunosuppressants may be applied.

Conclusion:

We present a case of a young man with confirmed Kikuchi-Fujimoto disease with cutaneous involvement.

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Abstract N°: 4110

Palatal ovoid patch in dermatomyositis with Anti-Mi 2b antibodies

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Introduction & Objectives: Dermatomyositis (DM) is a disease with a broad spectrum of cutaneous manifestations. The palatal ovoid patch has gained relevance in recent years due to its association in the literature with the anti-TIF1γ (transcription intermediary factor 1-gamma) antibody and consequently with the presence of underlying malignancy. We present a case of DM with the presence of a palatal ovoid patch associated with another myositis antibody, anti-Mi2b.

Materials & Methods: A 43-year-old woman with no medical history reported two weeks of asthenia, myalgias, and generalized arthralgias. Physical exam revealed erythemato-violaceous papules on hand joints, elbows, and knees, along with dilated capillaries around nails and a hyperpigmented plaque on the neckline. Oral exam showed an erythematous oval macule on the palate. Skin biopsies confirmed DM. Positive anti-Mi-2b antibody was detected. Elevated CA 15-3 (32.6 U/ml) and SCC antigen (2.4 ng/mL) were found. Imaging showed no abnormalities. Treatment with high-dose corticosteroids and weekly methotrexate 15 mg led to improvement. No malignancy was detected after 6 months of follow-up.

Results: The ovoid palatal patch presents as a well-defined erythematous spot, non-ulcerative, on the posterior hard palate. This finding was first documented in 2016 by Bernet et al., who reported in their cohort of 45 patients that 18 of them had the ovoid palatal patch (40%), and that it was significantly associated with the presence of anti-TIF1γ antibodies as well as highly associated with internal malignancy. Three subsequent articles discuss similar cases that are summarized in Table 1.**

The studies suggest the ovoid palatal patch is a specific finding of DM with positive anti-TIF1γ antibodies, often indicating malignancy. However, in our case, it wasn't linked to neoplasia or TIF-γ immunophenotype, but to anti-Mi2b, not previously reported. This antibody is associated with classic DM, responsive to treatment with low neoplasia or lung disease risk.

Conclusion: We want to emphasize the importance of oral examination in patients with suspected DM, even though, according to the case presented, the ovoid patch does not seem to be an exclusive finding in patients with anti-TIF1γ antibodies.

Reference	Number of cases (n)	Sex (n)	Age	Antibody Subtype (n)	Underlying neoplasia (n)	Type of neoplasia
Bernet et al. (2016)	18	W (17) M (1)	59 (median)	anti TIF1γ (15) No antibody (3)	Yes (7) No (11)	Unspecified
Bhattacharjee et al. (2020)	1	W	48	Not carried out	Not carried out	
Franciosi et al. (2020)	1	W	80	anti TIF1γ	Not carried out	-
Liu et al. (2023)	1	W	58	anti TIF1γ	Yes	Right ovarian tumor
Our case	1	W	43	anti Mi2b	No	-

Table 1. Summary of the cases published in the literature of dermatomyositis with palatal ovoid patch.W: woman M: male, TIF1γ: transcription intermediary factor 1-gamma

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Refractory Dermatomyositis Treated with Anifrolumab

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Introduction: Dermatomyositis (DM) remains challenging to treat. As more is learned about the inflammatory pathways associated with DM, new targeted therapies are being developed. A strong type 1 interferon (IFN) signature has been demonstrated in DM patients, and there are both available and developing drugs targeting IFN pathways. Anifrolumab, a monoclonal antibody that binds with high specificity and affinity to the type I IFN receptor subunit 1, has been developed more recently.

Clinical Case: We present the case of a 42-year-old woman who consulted for the appearance of pruritic skin lesions on her forehead and cheeks, which had been evolving for 1 month with progressive extension to the neck, anterior thorax, back, upper limbs, and thighs. She also reported asthenia and mild muscular weakness in the scapular and pelvic girdle without other systemic symptoms associated. Physical examination showed a typical cutaneous picture of dermatomyositis, confirmed by a skin biopsy. At that time, screening for neoplasia was negative, diagnosing classic adult dermatomyositis. Initially, the patient was treated with systemic corticosteroids, hydroxychloroquine, azathioprine, and methotrexate, with persistent intense cutaneous activity for years until a ductal carcinoma of the breast was diagnosed and treated. Throughout her evolution, she developed multiple foci of calcinosis in limbs and face, as well as several outbreaks of DM skin lesions, treated with different cycles of intravenous immunoglobulins. Since cutaneous activity was not controlled, compassionate use of anifrolumab was requested. The patient received 4 doses of intravenous anifrolumab every 4 weeks. Notably, three days after starting treatment, the patient reported significant improvement in pruritus, and in less than a month, the lesions greatly improved, becoming hyperpigmented. Dystrophic calcinosis has also been stable since then.

Discusion: A strong type 1 interferon signature is present in patients with DM, similar to lupus. Anifrolumab is a monoclonal antibody that blocks the type 1 IFN receptor. It is already approved for the treatment of systemic lupus erythematosus and has shown a good safety profile. Given that lupus and DM share dysregulation of the type I IFN pathway in their pathogenesis, anifrolumab could be a potential treatment for DM. There are only 2 published cases in the literature of dermatomyositis treated with anifrolumab with good efficacy and safety results.

Conclusion: We present a patient with paraneoplastic dermatomyositis showing severe dystrophic calcinosis and cutaneous activity despite numerous lines of treatment presenting good response to treatment with anifrolumab. Anifrolumab is a monoclonal antibody directed against the type 1 IFN receptor and represents a new therapeutic option for patients with refractory DM.

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Abstract N°: 4312

Successful Treatment of Refractory Oral Discoid Lupus Lesions with Anifrolumab Patient with Systemic Lupus Erythematosus: A Case Report

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Introduction:

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that can affect various organs, including the skin. Discoid lupus erythematosus (DLE) is a form of chronic cutaneous lupus erythematosus, which typically affects the upper body areas, including facial area, scalp and oral mucosae affecting mostly the lips. Anifrolumab, a type I interferon receptor antagonist, has shown promising results in the treatment of SLE. Here, we report a case of a 38-year-old female patient with SLE who had shown a good response to anifrolumab for oral discoid lupus lesions that were refractory.

Case Presentation:

The patient, diagnosed with SLE with anti-double-stranded DNA antibodies for six years, presented with arthralgia and cutaneous lesions during this time, initially diagnosed as subacute, later developing erythemateous scaly plaques and crusts on the lips. These lesions were biopsied showing perivascular and periadnexal lymphohistiocytic infiltrate under an interface dermatitis and the presence of mucin plus the presence of Plasmacytoid dendritic cells. Despite treatment with mycophenolate mofetil, belimumab, and colchicine, the skin condition did not improve. After initiating anifrolumab therapy by the Reumatology department, a significant improvement was observed in the lesions, leaving a small cicatricial area in the upper perioral region.

Conclusion:

This case contributes to the growing evidence that anifrolumab is useful for refractory CLE, including special areas such as lips and oral mucosa. Further studies are necessary to explore the efficacy of anifrolumab in treating chronic mucocutaenous lupus lesions and its impact on treatment of cutaneous lupus. Furthermore, it underscores the importance of a thorough differential diagnosis for cutaneous lupus involving the lips. The early recognition and appropriate management of these lesions are essential to prevent disfigurement and improve the patient's quality of life.

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Abstract N°: 4350

Bullous Pemphigoid: A Case Report of a 15-Month-Old Female Infant

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Introduction & Objectives: Bullous pemphigoid (BP) is a rare autoimmune disease that typically occurs at an advanced age but is unusually observed in children and infants. Herein, we present a 15-month-old female infant who developed BP, possibly triggered by vaccinations and ultraviolet radiation.

Materials & Methods: A 15-month-old female infant was presented to our outpatient clinic with widespread, pruritic, erythematous patches and fluid-filled blisters for approximately three days. Personal and familial medical history was unremarkable. Dermatological examination revealed erythematous macules, tense bullae, and erosions on the face, hands, feet, anterior and posterior trunk, and extremities. Nikolsky's sign was negative, and mucosal regions were not involved. The patient had been initially examined in a healthcare center and suspected of having an allergic reaction or a herpes virus infection. One month before the onset of the bullous eruption, the patient received varicella, measles-mumps-rubella, and a 13-valent pneumococcal conjugate vaccine. Subsequently, she was exposed to extensive sunlight on a vacation.

Results: The Tzanck smear did not show acantholytic cells. Skin biopsy and direct immunofluorescence examinations were performed from a bullous lesion and peri-lesional area. Histopathology revealed predominantly lymphocytic superficial perivascular dermatitis, focal spongiosis-exocytosis, focal basal fibrin deposition, and linear C3, IgM, and fibrinogen deposits in the basal layer. The patient was treated with ketotifen suspension and 0.1 % methylprednisolone acetate cream twice daily. Two weeks later, all bullae were regressed entirely. There were no relapses during a 6-month follow-up period.

Conclusion: Bullous pemphigoid should be considered in the differential diagnosis of bullous lesions observed during infancy.

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Abstract N°: 4365

A case of Pyoderma gangrenosum unmasking Systemic lupus erythematosus

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Introduction: Pyoderma gangrenosum (PG) is a rare non-infectious ulcerative neutrophilic dermatosis that is clinically characterized by a well-defined erythematous to violaceous margin commonly associated with underlying systemic disease like inflammatory bowel disease, arthritis, malignant disorders and monoclonal IgA gammopathy. Although pyoderma gangrenosum is associated with Systemic lupus erythematosus (SLE), it's been rarely reported as initial manifestation of lupus. Here we present a case of Pyoderma gangrenosum later diagnosed with SLE.

CASE report: A 19-year-old female patient presented with complaints of large wound over the left thigh of 5 days duration. Started as a small raised lesion over the inner aspect of left thigh which rapidly progressed into a wound approximately of one palm size. Associated with intense pain and mild discharge. History of joint pain was present. No other positive history was elicited. Patient gives history of similar complaint 1 year ago over the right leg which healed spontaneously with scarring. On examination: a solitary, tender, oval shaped ulcer, measuring 5x6cm in size located over the inner aspect of left thigh with well-defined borders, violaceous margin and undermined edges. Floor consists of red healthy granulation tissue with serous discharge, the base was formed by underlying muscles, and surrounding skin was warm and erythematous.

On investigation: complete hemogram showed low haemoglobin (10.3g/dl) and urine routine showed mild proteinuria. Other Routine blood investigations like liver function test, renal function tests, chest x-ray, ECHO was unremarkable. Pathergy test was positive. Anti-Nuclear Antibody was strongly positive for anti dsDNA .Biopsy from the ulcer edge showed focal ulceration, irregular acanthosis, spongiosis and basal cell vacuolar degeneration, dermis showed RBC exocytosis, multiple foci of dermal abscesses, perivascular and peri adnexal mixed inflammatory infiltrate composed of lymphocytes, neutrophils and eosinophils extending till deep dermis. Patient was diagnosed with Pyoderma gangrenosum secondary to SLE after fulfilling the criteria of PG and SLE, and was started on oral steroids and colchicine with supportive wound care and regular dressing. Patient showed complete resolution of lesion, which healed with cribriform scarring. Patient was advised for regular follow up for monitoring of systemic involvement of lupus.

Conclusion: Ulcer development in systemic lupus erythematosus is rare but well-documented with reported rates of around 5 and 8% of cases, most commonly secondary to vasculitis of medium-sized arteries. But pyoderma gangrenosum has been rarely reported as sole cutaneous manifestation of SLE. Hence here we report a rare case of PG that unmasked the systemic lupus erythematosus in a young female patient.

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Abstract N°: 4380

The impact of clinical phenotypes of psoriatic arthritis on patient-reported outcomes

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Introduction & Objectives: The variety of clinical manifestations of psoriatic arthritis (PsA) leads to a significant deterioration in the health status of patients, including health-related quality of life (HRQoL). In 2016, according to the recommendations of the Group for Research and Assessment of Psoriasis and Psoriatic Arthritis (GRAPPA), in addition to the standard rheumatological assessment of the patient, must conduct an assessment of patient reported outcomes (PROs) for timely correction of therapy. The aim of our study To assess the PROs according to patients with PsA and determine the relationship with clinical phenotypes (peripheral arthritis, enthesitis, dactylitis, psoriasis severity, axial lesions, psoriatic onychodystrophy).

Materials & Methods: the study included 172 patients with PsA (male/female (n/%) - (52.3)/ 82(47.7)), meeting the CASPAR (2006). Mean age was 45.1±11.8 years, duration of PsA was 113.1±80.4 months (mo), duration of psoriasis was 246.1±147.7 mo, DAPSA was 28±22.2. A standard rheumatological examination and PROs were performed. At the time of inclusion in the study, enthesitis was observed in 41.7%, dactylitis - 41.1%, severe psoriasis in 36.6% of cases. PROs included: VAS global assessments (mm), VAS global pain (mm), BASDAI, PsAID-12, FACIT and HAQ). Me [Q25; Q75], Pierson-χ2 M±SD, %, t-test, Pierson-χ2, Manna-Whitney tests were performed. All $p<0.05$ were considered to indicate statistical significance.

Results: patients with peripheral arthritis, axial lesions, enthesitis and severe psoriasis were significant in all measured scales ($p<0.0001$). Patients with dactylitis had a significantly worse pain assessment ($p<0.0001$), functional impairments (HAQ) were detected more often ($p=0.005$), a higher assessment of the activity of their disease according to the BASDAI index ($p=0.002$), other indicators were comparable to patients without dactylitis. In the group with psoriatic onychodystrophy, higher disease activity according to BASDAI ($p=0.0002$) and severe fatigue (FACIT) ($p<0.0001$) were detected.

Conclusion: Thus, some clinical phenotypes of PsA (peripheral arthritis, enthesitis, axial lesions, severe psoriasis) have a significant impact on PROs, which, along with a physician's assessment of disease activity, clinical manifestations, can make it possible to assess the effectiveness of therapy and timely adjust it.

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Abstract N°: 4455

Pemphigus and asphalt: a possible relation

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Pemphigus and asphalt: a possible relation

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Introduction & Objectives:

Pemphigus is an autoimmune blistering skin disease mediated by autoantibodies directed against epidermal desmosomal proteins, namely desmogleins (Dsg) 1 and 3. Depending on the clinical presentation, the level of epidermal clefting and antibody profile, two main types of pemphigus exist, pemphigus vulgaris (PV) and foliaceus (PF). As in most of the autoimmune diseases the etiology of pemphigus remains unknown but multiple environmental factors may be related with its onset, including physical agents (UV radiation, high temperature), chemicals (pesticides, drugs) or biological reasons (infectious diseases or vaccines). We report a case of pemphigus occurring in a patient who is professionally exposed to asphalt.

Materials & Methods:

A 58-year-old man was admitted to the Department of dermatology with a history of progressive skin blistering over the past 6 months. The initial cutaneous lesions have appeared during the last summer when the patient was hired by a road construction company as an asphalt worker. He was occupationally exposed to newly-laid asphalt and bitumen fumes in combination with high temperatures and UV-radiation. For his complaints he was previously treated with systemic and topical antibiotics without improvement. Upon admission the clinical examination revealed vesiculo-bullous eruption and painful erosions covered with crusts on his scalp, face, chest and back in the absence of mucosal involvement. Our diagnostic algorithm included routine laboratory tests, histological examination, and immunological investigations.

Results:

Routine laboratory demonstrated mild lymphocytopenia and elevated inflammatory markers. Histological examination revealed an inflammatory infiltrate, eosinophilic spongiosis, and superficial epidermal blister formation. Direct immunofluorescence from perilesional skin showed intercellular deposition of IgG and C3 complement in the epidermis. ELISA tested highly positive for Dsg1. Based on the clinico-laboratory findings the presumptive diagnosis of PF was confirmed. Therapy with moderate doses of systemic corticosteroids and antibiotics in combination with topical corticosteroids resulted in rapid disease control. Additionally, the patient was advised to change his professional environment.

Conclusion:

Occupational asphalt exposure might appear a new possible trigger for pemphigus as in the case presented here. Pemphigus is frequently occurring following exposure to chemicals, UV-rays, or heat. Asphalt, used for road paving, consists of bitumen which is a by-product of crude oil refining, binders and fillers, fibres, and crushed stones. Exposure of road pavers to asphalt mixtures is related to numerous health effects including headache, skin

rash, sensitization, fatigue, reduced appetite, throat and eye irritation, cough, and skin cancer. Further experience would be necessary to elucidate the possible relationship between this culprit and pemphigus.

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Abstract N°: 4482

Azathioprine hypersensitivity syndrome in patient with pemphigus foliaceus

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Introduction & Objectives: Azathioprine hypersensitivity syndrome is a rare but potentially serious adverse drug reaction. This syndrome occurs in about 2% of cases. This adverse reaction is not dependent on the dose of the drug and is not dependent on the activity of thiopurine S-methyltransferase. The presentation most closely resembles sepsis or the recurrence of the disease, usually occurring about four weeks after taking the drug, and leads to recovery typically about 7 days after exclusion of the drug.

Materials & Methods: We report a 69 years old patient with pemphigus foliaceus treated with azathioprine. Four weeks later he developed severe unrecognized azathioprine hypersensitivity syndrome. It was during the pandemic, and the patient was vaccinated against COVID-19. However, until receiving negative results of antigen test and hemocultures, fever and other signs and symptoms were initially considered to be a possible sepsis or covid 19 disease.

Results: Before azathioprine introduction our patient developed severe side effects from methotrexate and later Dapsone. Since oral corticosteroids are not a treatment of choice for chronic disease, we introduced azathioprine. After four weeks, he was readmitted to our department for a regular checkup of blood analyses and skin. He was feeling good, erosions were in the end stage of epithelization, and routine blood analyses were within normal limits. But within 72 hours he developed fever, nausea, and vomiting. In routine analyses we found leukocytosis with neutrophilia, increased liver and pancreatic enzymes, decreased parameters of renal function. CRP was 202 mg/L, procalcitonin was 3.85 ng/ml, and D-dimer was 4.52 mg/L. Hemocultures remained sterile. We discontinued azathioprine treatment, but we were not aware that his condition was caused by azathioprine hypersensitivity syndrome. Within 3-4 days, all symptoms disappeared, and blood analyses were within normal limits after 10-14 days. We did not have an explanation for his condition. After two weeks of good general condition and treatment with topical corticosteroid treatment, we reintroduced azathioprine in dose of 50 mg/day. Within 48 hours he developed the same severe azathioprine hypersensitivity syndrome, which resolved about 7 days after stopping the drug.

Conclusion: This syndrome should be suspected when there is an absence of infection and serological evidence of relapse of the disease. Therefore, high clinical suspicion and early recognition of this syndrome are important to minimize morbidity and mortality.

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Abstract N°: 4563

Detection of herpes simplex viruses in the oral lesions of patients with pemphigus vulgaris: Is it diagnostic or predictive of disease severity?

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Introduction & Objectives: Some studies emphasize the relationship between the herpes simplex virus (HSV) and pemphigus. Although the possible role of HSV in the pathogenesis of pemphigus and the severity of the disease is obscure, we aimed to evaluate the presence of herpes simplex viruses (HSV 1/2) in the oral lesions of patients with pemphigus vulgaris and also assess its association with disease severity and types of lesions.

Materials & Methods: A retrospective study was conducted on collected data in the form of collecting paraffin blocks, slides, and relevant pathology reports and referring to patients' medical records. A questionnaire containing details on the degree of skin, scalp, and mucosal involvement (Pemphigus Disease Area Index (PDAI)) was fulfilled. The immunoassay result was also collected to check the anti-desmoglein 3 and 1 antibodies (using ELISA technique).

Results: In this study, 52 patients of pemphigus vulgaris with oral lesions (case) and 52 patients with oral lesions not related to the disease (control) were evaluated. HSV1 was detected in 13.5% of oral pemphigus lesions and 1.9% of the control group ($p = 0.0598$). There were no positive cases of HSV2 in either group. There was no significant association between the positivity of HSV1 and the site of lesions ($p = 1.00$) or disease severity ($p = 0.28$). However, we found a strong correlation between the PDAI disease severity score with the titer of the AntiDsg3 antibody ($r = 0.487$, $p = 0.001$) and AntiDsg1 antibody ($r = 0.309$, $p = 0.026$).

Conclusion: This study demonstrated a significant prevalence of HSV1 in oral pemphigus lesions, and acyclovir therapy may play a significant role in managing these patients. However, HSV's role in the *pathogenesis* of pemphigus vulgaris cannot be clearly determined.

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Abstract N°: 4638

Efficacy and safety of Tofacitinib in refractory pemphigus : a retrospective cohort study

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Title:

Efficacy and safety of Tofacitinib in refractory pemphigus : a retrospective cohort study

Introduction & Objectives:

High-dose corticosteroids are considered the standard treatment for pemphigus. However, due to the serious and potentially life-threatening side effects of long-term corticosteroid therapy for this disease, refractory pemphigus may use rituximab as an adjuvant therapy. But there are disadvantages such as high risk of immunodeficiency due to B-cell depletion and high cost. JAKi Tofacitinib as the potent treatment for refractory pemphigus.Therefore, we evaluated whether using Tofacitinib as adjuvant therapy during the active phase of the disease could accelerate the rate of steroid reduction in patients and achieve rapid disease remission.

Materials & Methods:

We retrospectively reviewed the use of Tofacitinib as an adjuvant treatment for pemphigus patients in our department (retrospective, single-center) over a period of 2 years (n=21), and compared them with a matched group of pemphigus patients who did not receive Tofacitinib during the same period (n=21). We assessed the baseline severity of the disease in both groups, calculated the average time to achieve disease remission, and compared the proportion of patients achieving complete remission at 3 and 6 months, as well as the average amount of steroids used.

Results:

The proportion of patients with pemphigus achieving complete remission at 3 and 6 months with adjunctive Tofacitinib therapy was 61.9% and 90.5%, respectively, compared to 52.4% and 80.9% with steroids alone

Conclusion:

Our data shows that using Tofacitinib in combination with prednisone is more effective in treating pemphigus patients during the active phase of the disease compared to using prednisone alone, but it is necessary to screen for the patient’s tumor and thrombotic risk before using JAKi.

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Abstract N°: 4641

IMG-004, a selective noncovalent reversible Bruton's tyrosine kinase inhibitor, demonstrated potent pharmacodynamic effects

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Introduction & Objectives:

Bruton's tyrosine kinase (BTK) is a critical kinase mediating signaling downstream of various receptors, including B cell receptor (BCR), Fc receptor (FcR), Toll-like receptor (TLR) and some G protein-coupled receptors (GPCR). Several BTK inhibitors have shown clinical efficacy in various immunological and inflammatory (I&I) disorders such as chronic spontaneous urticaria (CSU), hidradenitis suppurativa (HS) and systemic lupus erythematous (SLE). IMG-004 is a novel noncovalent reversible oral BTK inhibitor designed for chronic use in I&I diseases. Here we report the preclinical pharmacodynamic effects of IMG-004 in various in vitro and in vivo studies.

Materials & Methods:

In vitro studies were conducted to determine the inhibitory effects of IMG-004 on BTK enzymatic activities using Z'-LYTE™ kinase assay kit-Tyr 1 peptide system (Invitrogen). BTK Tyr223 phosphorylation was measured by using HTRF kit (Cisbio) with human B cell line Ramos, B cell activation was measured by CD69 expression in human whole blood, and basophil activation was measured by CD63 expression in human whole blood. IMG-004's residence time on wild-type BTK was also investigated with fluorescence competition assay. In vivo efficacy was evaluated in animal disease models, including a prophylactic major histocompatibility complex (MHC)-matched but minor histocompatibility antigen (miHA)-mismatched mouse model of chronic graft-versus-host disease (cGvHD) in BALB/c mice.

Results:

IMG-004 showed high potency in BTK inhibition, with low IC90s for inhibiting BTK enzymatic activities, BTK phosphorylation, B cell activation in human whole blood, and basophil activation in human whole blood in vitro (table 1). IMG-004 bound to BTK with a prolonged residence time of 34.75 h, compared with 11.01 h shown by an in-house made fenebrutinib analog. Prophylactic treatment of oral IMG-004 at 10 mg/kg twice daily significantly prolonged animal survival time, reduced proteinuria incidence and severity, and completedly suppressed ascites in the cGvHD animals.

Conclusion:

IMG-004, a unique noncovalent reversible BTK inihibitor, demonstrated potent BTK inhibition in vitro and protective efficacy in a cGvHD model in vivo. IMG-004 is a promising potential therapeutic agent for chronic autoimmune diseases.

Table 1. Summary of inhibitory potency of IMG-004 in various in vitro studies

Inhibitory Potency (nM)	IC50	IC90
BTK enzyme activity	4	25
pBTKY223 in Ramos	4	32
Human whole blood B-cell activation (CD69)	4	15
Human whole blood basophil activation (CD63)	10	22

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Abstract N°: 4651

A systematic review of case series and clinical trials investigating systemic oral or injectable therapies for the treatment of vitiligo

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Introduction & Objectives:

The purpose of this study is to investigate the effectiveness and safety of oral and injectable systemic treatments, such as methotrexate, azathioprine, cyclosporine, tofacitinib, baricitinib, corticosteroids, statins, zinc, apremilast, etc., for treating vitiligo lesions.

Materials & Methods:

Databases including PubMed, Scopus, and Web of Science were meticulously searched for studies spanning from 2010 to August 2023, focusing on systemic oral and injectable therapies for vitiligo, using comprehensive keywords and search syntaxes tailored to each database. Key data extracted included study design, treatment efficacy, patient outcomes, patient satisfaction, and safety profiles.

Results:

In a total of 42 included studies, oral mini-pulse corticosteroid therapy (OMP) was the subject of six studies (14.2%). Minocycline was the focus of five studies (11.9%), while methotrexate, apremilast, and tofacitinib each were examined in four studies (9.5%). Antioxidants and Afamelanotide were the subjects of three studies each (7.1%). Cyclosporine, simvastatin, oral zinc, oral corticosteroids (excluding OMP) and injections, and baricitinib were each explored in two studies (4.8%). Azathioprine, mycophenolate mofetil, and Alefacept were the subjects of one study each (2.4%).

Conclusion:

Systemic treatments for vitiligo have been successful in controlling lesions without notable side effects. OMP, Methotrexate, Azathioprine, Cyclosporine, Mycophenolate mofetil, Simvastatin, Apremilast, Minocycline, Afamelanotide, Tofacitinib, Baricitinib, Antioxidants, and oral/injectable corticosteroids are effective treatment methods. However, oral zinc and alefacept did not show effectiveness.

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Abstract N°: 4660

Onset of bullous pemphigoid during treatment with anti-interleukin-5 drugs in two asthmatic patients

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Introduction & Objectives: Bullous pemphigoid (BP) is the most prevalent subepidermal autoimmune disease worldwide. Improved knowledge about the pathogenesis of BP has motivated various clinical trials of new treatments, including benralizumab and mepolizumab (monoclonal antibodies that target interleukin (IL)-5). We report two cases of BP onset during treatment with anti-IL-5 drugs for severe asthma.

Materials & Methods: We report two cases of BP onset during treatment with anti-IL-5 drugs for severe asthma.

Results: The first case was a 71-year-old woman with multirefractory allergic asthma treated with benralizumab every 8 weeks for 7 months before she developed BP. The symptoms persisted despite treatment with oral corticosteroids (1mg/kg/day), so she was started on methotrexate (MTX) 20mg/week and intravenous immunoglobulins monthly. Benralizumab was discontinued we decided to start dupilumab 300mg every 2 weeks to control asthma and BP together, with good control persisting after two years of treatment.

The second case was an 85-year-old male with non-allergic eosinophilic bronchial asthma treated with mepolizumab for one year before he developed BP. During the evolution the patient was treated with systemic corticosteroids (1mg/kg/day), dapsone 200mg/day for 11 months and MTX 12.5mg/week for 6 months, with poor response. Subsequently, we switched mepolizumab to dupilumab, without achieving an adequate response. The patient has been under good control with omalizumab for the past year.

Conclusion: IL-5 is expressed abundantly in the lesional skin and serum of people with BP, and is associated with peripheral eosinophilia and cutaneous eosinophilic infiltration. This evidence has motivated various clinical trials that aim to determine the efficacy in BP of anti-IL5 drugs currently approved for the treatment of severe eosinophilic asthma, such as mepolizumab and benralizumab.

An ongoing randomised, double-blind, placebo-controlled phase III trial (NCT04612790) is assessing the efficacy and safety of benralizumab in people with BP. On the other hand, a recent phase II clinical trial investigated the efficacy and safety of mepolizumab in people with BP, with disappointing results: compared with placebo, the intervention failed to significantly reduce BP relapse rates, pruritus intensity, or titres of anti-BP180 and anti-BP230 antibodies.

Although various drugs may be associated with BP development, to date there is only one case of BP has been reported in a patient receiving anti-IL-5 drugs. This leads us to hypothesize that these drugs may be ineffective at controlling or preventing BP, or may produce a paradoxical effect, analogous to the paradoxical psoriasis observed in people treated with tumour necrosis factor-alpha inhibitors.

The increasing use of new biological drugs may lead to unknown cutaneous adverse effects in the future. This possible association merits further investigation to establish causality and the best treatment approach.

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Abstract N°: 4714

Effectiveness of treatment with intravenous immunoglobulins in refractory skin manifestations of dermatomyositis

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Introduction & Objectives: Dermatomyositis (DM) is a heterogeneous idiopathic inflammatory myopathy with varied skin and muscle involvement. Treatment is not standardized, with systemic corticosteroids being the first-line therapy followed by immunosuppressants. The use of intravenous immunoglobulins (IVIG) presents numerous scientific evidence as a second-line adjuvant treatment improving muscle and skin symptoms, although there are no differences regarding the improvement of the different skin manifestations.

Materials & Methods: We conducted a retrospective observational study from 2017 to 2022 in patients with refractory DM who received at least one dose of IVIG, to determine the therapeutic effect on each of the skin manifestations during the follow-up period.

Results: 17 patients were collected and 9 typical lesions were analyzed (psoriasiform dermatitis of the scalp, heliotrope erythema, shawl sign, neckline erythema, mechanic's hands, Gottron papules, elbow erythema, Gottron inverse and Holster sign) in addition to lesions secondary to vasculopathy (periungual erythema and ulcers), thrush and pruritus. We define improvement as the disappearance of the lesions after the IVIG cycle and subsequent reduction/suspension of corticosteroids. The most prevalent skin involvement was on the back of the hands (94%), followed by heliotrope and malar erythema (88%), pruritus (69%), and shawl sign (65%). An improvement of more than 75% has been observed in all lesions, except for scalp involvement and lesions secondary to vasculopathy. Pruritus has shown the fastest response (83% from the first dose). We have not found differences in the response between classic DM and those associated with neoplasia.

Conclusion: We present to our know the first observational study with the specific skin improvements of patients with refractory DM treated with IVIG. The limitations of the study were the retrospective nature and the small sample size, so more studies would be needed to confirm the results.

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Abstract N°: 4725

High frequency ultrasound and lipofilling in facial sclerosing dermatoses: a multicenter retrospective case series.

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Introduction & Objectives:

The aesthetic sequelae of facial sclerosing inflammatory dermatoses have a significant impact on quality of life. Autologous fat grafting, or lipofilling (LF), provides volume and immunomodulatory factors to atrophic areas. However, multiple LF sessions are often required due to high reabsorption rates. We explored the use of high-frequency cutaneous ultrasound (HFU) as a potential tool for guiding patient management in facial sclerosing dermatoses.

Materials & Methods:

We present a retrospective series of 23 patients from 3 different hospitals in Spain with facial atrophy due to progressive hemifacial atrophy, lineal morphea, dermatomyositis and lupus panniculitis. HFU was performed with a 10-22 MHz probe. Measurements of thickness from the epidermis to the underlying bone on both affected and unaffected sides, along with Doppler activity, were collected.

Results:

Twenty-three patients were included (mean age 41 ± 13 years, 91.3% female)**(Table 1)**. Of these, 11 had undergone LF treatment and 12 had not. HFU was performed pre-LF (n=20), post-1st LF (n=9), post-2nd LF (n=3), and post-last LF (n=11). Following the 1st LF, the mean thickness on the affected side compared to the unaffected side increased from 52 ± 19% to 83 ± 20%. The 2nd LF was performed at a mean of 10 ± 5 months after the 1st LF, with thickness increasing to 87 ± 16%. After a mean of 2.6 ± 1.4 LF sessions, the mean thickness on the affected side reached 84 ± 18%, representing an absolute thickness increase of 32% compared to the initial defect. HFU was performed on average 5 ± 4 months after LF. Conversely, in patients without LF, the thickness of the affected side compared to the unaffected side was of 67 ± 15% **(Figure 1)**. Increased vascularization was not observed in any patient by HFU neither before nor after the LP.

Conclusion:

To our knowledge, this study provides the first objective description of LF effectiveness in correcting facial defects from sclerosing dermatoses. We observed an absolut thickness increase of 32% in atrophic area after an average of 2.4 LF sessions. HFU is an accessible, inexpensive, and non-invasive method for assessing dermal and subcutaneous thickness. HFU could measure LF reabsortion rate and potentially guide the timing of further LF sessions, which are currently decided subjectively. Limitations include the small sample size and retrospective nature of the study. HFU emerges as a valuable tool for evaluating LF outcomes and optimizing patient

management in facial sclerosing dermatoses. Nonetheless, further prospective studies with larger samples are required to characterize the temporal dynamics of LF reabsorption in such cases and to assess the impact of this treatment on the disease's course.

	Lipofilling	No Lipofilling
Patients (n)	11	12
Age in years (mean, SD)	42 (14)	41 (13)
Women (n,%)	9 (81)	12 (100)
Lineal morphea (n,%)	3 (27)*	9 (75)*
Hemifacial progressive atrophy (n,%)	6 (54)*	3 (25)*
Dermatomyositis (n,%)	1 (9)	0 (0)
Lupus panniculitis (n,%)	2 (18)	1 (9)

Table 1: Demographic characteristics of our case series. *In both groups 1 patient was affected by both hemifacial progressive atrophy and lineal morphea.

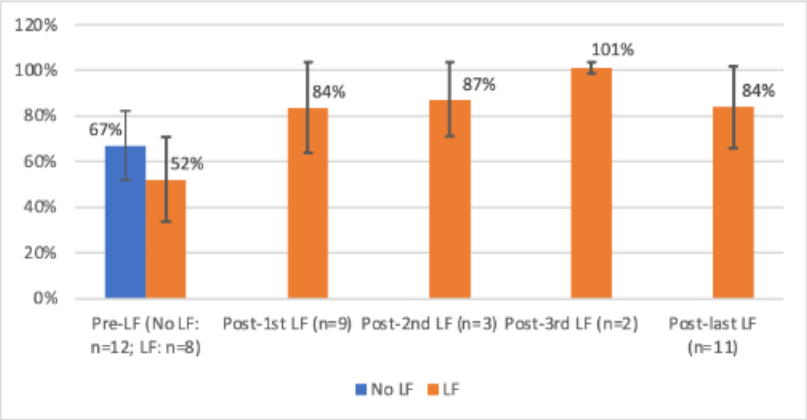

Figure 1: Thickness of the atrophic side compared to the healthy side (100%), measured from the epidermis to the underlying bone by high frequency ultrasound.

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Abstract N°: 4726

Parry Romberg's Rapid Development in Early Childhood

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Introduction: Parry-Romberg syndrome, also known as progressive hemifacial atrophy, is a rare degenerative disorder with numerous distinctive clinical presentations. **Case Report:** A 4-year-old boy was brought by his parents to a dermatological consultation due to a facial lesion. The lesion appeared when he was around 2 years old. On dermatological examination, a linear area of atrophy was observed on the right frontal region, extending from the eyebrow to the scalp. Adjacent to this linear area, there was marbled-colored skin extending on the right side of the face, with a slightly sclerotic aspect. A skin biopsy was performed, confirming the diagnosis of segmental facial scleroderma, better known as Parry Romberg syndrome. General exams were within normal limits, and the patient was under azathioprine treatment for disease control. **Conclusion:** The Parry-Romberg syndrome is a very rare entity that has devastating consequences due to its various systemic involvements; Early recognition and prompt management of Parry Romberg syndrome, including the use of immunosuppressive agents like azathioprine, can help mitigate disease progression and improve patient quality of life. Long-term follow-up and further studies are warranted to better understand the etiology and optimal treatment strategies for this rare condition.

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Abstract N°: 4826

Lupus erythematosus tumidus

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Introduction & Objectives:

Lupus erythematosus tumidus (LET) is an uncommon inflammatory skin disease. It is a form of intermittent cutaneous lupus erythematosus. LET is characterised by the high photosensivity and it manifests itself as various skin lesions that mimics other diseases. Therefore, this condition is rarely diagnosed at the initial appointment and is a challenge to distinguish it from Jessner lymphocytic infiltration of the skin (JLIS), reticular erythematous mucinosis, polymorphous light eruption urticarial vasculitis and urticaria. There is little literature on this problem. Even today LET causes a lot of debate regarding its clinical and histological features.

Materials & Methods:

Medical history, clinical picture and results of histology and immunofluorescence tests of 2 patients are presented in Table 1.

Table 1. Description of patients

	Patient 1	Patient 2
Sex / Age	Male / 30 yo	Male / 53 yo
Medical history of interest	Hepatomegaly	Hypertension, dyslipidemia, smoking
Time from outbreak to diagnosis	6 months	2 years
Geographical location	Central Asia	North-Western region of Russia
Season	N/A	Spring-Summer
Trigger factors	Sun exposure, doxycycline (2 months)	Sun exposure, alcohol, ACE inhibitor (3 years)
Previous diagnoses	**Dermatitis, JLIS, Rosacea, cutaneous sarcoidosis**	Dermatitis, Urticaria, Rosacea
Clinical manifestation	3 round shape urticarial-like erythematous-violaceous plaques (oedematous erythema) with clearly defined boundaries	Multiple annular and semi annular urticarial-like erythematous-violaceus plaques (oedematous erythema) and succulent papules
Localisation	Face	Face, neck, chest, back
Itch	No	Yes
Indirect immunofluorescence	ANA < 160 dsDNA – 1,3 IU/ml	ANA < 160 dsDNA 0,02 IU/ml
Histology	Focal perivascular and pronounced perifollicular lympocytic infiltration penetrating the follicular epithelium; in the deep parts of the dermis perivascular basophilic staining was detected. Deposition of mucin was noted by additional stains (PAS, Van Gieson, Masson's trichrome)	Dense perivascular and perifollicular lymphocytic infiltration, PAS-positive inclusions; no obvious damage to connective tissue was noted when staining according to Van Gieson and Weigert.
Direct immunofluorescence (DIF)	Small granular deposits IgM (3+), IgA (+), C3 (1+) along the epidermal basement membrane zone.	Small granular deposits IgA (+), IgG (2+), IgM (+) along the epidermal basement membrane zone

Results:

Based on the clinical picture, medical history, as well as the results of DIF and histology, both patients were diagnosed with LET. UV protection and smoking cessation were recommended. Hydroxychloroquine 400 mg/day and topical methylprednisolone aceponate 0,1% were prescribed with favourable response. Both patients were under supervision for 2 years. During this time, long-term remission was noted.

Conclusion:

LET is a very rare condition, which makes it difficult to diagnose. When collecting the anamnesis, it is important to pay particular attention to the presence of increased photosensitivity. Clinical picture of this dermatosis has various manifestations on the skin such as oedematous erythema, succulent papules and plaques without signs of scaling, ulceration and scarring. The diagnosis of LET should be confirmed by pathomorphological examination and DIF. Provided treatment with Hydroxychloroquine 400 mg/day, topical methylprednisolone aceponate 0,1% and SPF 50+ cream showed good results in patients with LET.

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Abstract N°: 4851

Clinical presentation and characteristics of dermatomyositis in women: Experience of a female department

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Introduction & Objectives: Dermatomyositis (DM) is a rare inflammatory disease that can present with skin involvement alone, muscle disease, or even be associated with malignancy. The main variants of DM include classic DM, clinically amyopathic DM (CADM), paraneoplastic DM, and juvenile DM. Diagnosing DM requires a comprehensive evaluation of clinical signs, laboratory tests, imaging studies, muscle and skin biopsies, and myositis-specific autoantibodies (MSAs). In patients with CADM, there is a need for malignancy screening and, as well as paraneoplastic DM, it often requires early and aggressive treatment.

Materials & Methods: The aim was to evaluate the clinical presentation and characteristics of dermatomyositis in females. We encompassed 16 patients in a cross-sectional study over a five-year period. For each patient, we analyzed skin manifestations, muscle weakness, biochemistry analyses (creatine kinase and lactate dehydrogenase), and the presence of MSAs. If performed, electromyoneurography (EMNG), MRI findings, and histopathology confirmation of dermatomyositis were also considered.

Results: Sixteen women were included. The median age was 61 years (range 39-77). In all patients, photoexposed areas (face, neck, upper chest, and dorsal aspect of hands) were affected. The most common skin manifestations were heliotrope rash and Gottron papules, each observed in 10 patients. Periungual erythema and/or telangiectasia were noted in eight patients, while the “shawl” sign was present in four patients. Eight patients had clinically manifested proximal muscle weakness, and CADM was noted in the same number of subjects. Seven patients had shown findings consistent with myopathy on EMNG. In seven patients, MRI findings suggested soft-tissue edema and inflammation, which can be seen in dermatomyositis. Histopathology diagnosis confirmation was performed on 12 patients. CK and/or LDH levels were elevated in 11 patients. Immunology analysis was performed for all patients; ANA Hep-2 was positive in 13, while 10 patients had positive MSAs at the moment of diagnosis. Four patients had paraneoplastic dermatomyositis, which was triggered by breast, endometrial, and ovarian cancer.

Conclusion: In our female patients, among typical but heterogeneous skin manifestations, their localization was in photoexposed areas without any exception. Most commonly, heliotrope rash and Gottron papules were observed. Biochemical and immunology abnormalities were registered in more than 2/3 of the subjects, but clinical or electrophysiological muscle involvement was noted only in half of the sample. The underlying malignancy was detected in 25% of patients. Due to heterogeneous manifestations, the suspected case warrants thorough and systematic examination. Prompt recognition is extremely important, especially in patients with underlying malignant disease.

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Abstract N°: 4948

Spectrum of Cutaneous Manifestations of Connective Tissue Diseases in North East India

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Introduction & Objectives: Connective tissue diseases (CTDs) are chronic, multisystem inflammatory disorders, with the skin as a primary target. Cutaneous involvement in CTDs typically occurs early, and understanding its regional manifestations can lead to earlier diagnosis and treatment. Therefore, this study aimed to observe, document, and highlight the various patterns of cutaneous involvement in CTDs prevalent in the North East Indian population.

Materials & Methods: A year-long cross-sectional study was conducted at a tertiary care hospital. Cases of CTDs with cutaneous manifestations were included and clinically assessed. The findings were analysed using descriptive statistics. Continuous variables were presented as mean ± SD or median (range), and categorical variables as absolute numbers and percentages.

Results: The prevalence of CTDs was 1.35%. Of the 147 patients, 84% were female, with a mean age of 36.26 ± 12.90 years. Eight patients belonged to the paediatric age group. Lupus erythematosus was the most common CTD (67%), followed by rheumatoid arthritis (12%), mixed connective tissue disease (9%), overlap syndrome (4%), systemic sclerosis (4%), dermatomyositis (3%) and Sjogren's syndrome (1%). Acute cutaneous lupus erythematosus (53.06%) was the most common subtype in the lupus group. Limited cutaneous subtype (67%) was the most common among the systemic sclerosis patients with one case of CREST syndrome. All patients with dermatomyositis exhibited Gottron's papules and poikiloderma. Systemic and nail involvement was seen in 32% and 30% of cases, respectively. Antinuclear antibodies (ANA) were positive in 85.71% of patients.

Conclusion: CTDs, despite their diversity, often present early skin symptoms. Clinical features vary among populations due to genetic and environmental factors. This study underscores the vital importance of dermatological assessment in the early identification and management of CTDs.

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Abstract N°: 4984

Pemphigus Vulgaris localized to the Penis

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Introduction & Objectives:

Pemphigus vulgaris is a chronic, autoimmune vesiculobullous disease affecting the mucosae and skin. Mucosal involvement precedes cutaneous involvement. Oral mucosa is most affected. Involvement of penile mucosa is rare as a part of generalized cutaneous disease or as an isolated involvement.

Case Report:

A 48-year-old married male presented with non-healing raw areas on glans penis for 6 months. The patient was previously treated for candidial balanoposthitis, herpes genitalis, and syphilis with oral antifungals, antivirals, and oral antibiotics multiple times without significant improvement. There was no history of associated comorbidities, presence of similar lesions in the spouse, or history of high-risk sexual behavior. Cutaneous examination revealed moist crusted erosions on the glans and shaft of the penis with nonsignificant left inguinal lymphadenopathy. A provisional diagnosis of genital lichen planus was considered. His routine investigations including HIV and VDRL did not reveal any abnormality. Histopathology showed the presence of intra-epidermal acantholysis with multiple acantholytic cells and inflammatory infiltrate in the upper dermis. Desmoglein 3 antibodies were raised suggesting the diagnosis of Pemphigus vulgaris localized to the penis was made. Screening for paraneoplastic pemphigus did not reveal any abnormality. The patient was treated with oral steroids and azathioprine with complete resolution of the lesions. The disease is in remission for almost 1 year.

Conclusion:

Pemphigus vulgaris of penile mucosa may present as erosive lesions. Isolated localized involvement to penile mucosa without generalized disease, may pose a diagnostic challenge as it can be easily mistaken for sexually transmitted diseases. Physicians should be aware of this rare presentation and skin biopsy should be considered in patients not responding to standard care.

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Abstract N°: 5091

"Red on white", an unspecific feature in autoimmune connective tissue diseases?

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Introduction & Objectives:

"Red on white" patches were initially described in 2014 by Fiorentino et al. as a distinctive cutaneous feature in dermatomyositis patients positive for anti-transcriptional intermediary factor (TIF)-1γ antibodies. These patches manifest as hypopigmented areas accompanied by follicular erythema.

Materials & Methods:

Case Report: We present the case of a 41-year-old female with a history of celiac disease, presenting with a one-year history of facial and cervical erythema. Upon examination, characteristic "red on white" plaques were noted in the neckline, retroauricular and frontal regions. Suspecting dermatomyositis, a skin biopsy was performed showing atrophic epidermis, a vacuolar-interface pattern at the dermoepidermal junction and a superficial perivascular and perianexial lymphocytic infiltrate with increased dermal mucin. Additionaly, a myositis-specific antibody panel was conducted, revealing a positive result for small ubiquitin-like modifier-1-Activating Enzyme (anti-SAE-1). Muscle involvement was ruled out, and a diagnosis of amyopathic dermatomyositis was established. Despite treatment with hydroxychloroquine, methotrexate, and immunoglobulins yielding no improvement, off-label use of baricitinib resulted in progressive resolution of pruritus and lesions, with gradual repigmentation of the "red on white" patches on the chest, neck, and face.

Results:

While "red on white" patches have been primarily associated with anti-TIF1γ dermatomyositis, our case suggests their presence in dermatomyositis patients with alternative antibody profiles. Additionally, we propose a potential correlation between these patches and the "salt and pepper" lesions observed in scleroderma patients with low skin phototypes. However, comprehensive reports are needed to better understand this phenomenon.

Conclusion:

This Dermatomyositis SAE-1 case treated with baricitinib emphasizes the significance of "red on white patches" as an unspecific indicator in autoimmune skin diseases, urging enhanced clinical attention and research into their diagnostic and therapeutic implications.

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Abstract N°: 5153

Induction of Pustular Psoriasis Following Abrupt Withdrawal of Immunosuppressants in a Patient with Pemphigus Foliaceus: A Case Report

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Introduction & Objectives: Numerous reports have documented the complex interplay between pemphigus and various forms of psoriasis, with bullous pemphigoid frequently co-occurring in patients with psoriasis vulgaris. This suggests shared pathogenetic mechanisms. However, instances of pustular psoriasis emerging in patients with pemphigus foliaceus remain undocumented. We present a unique case of pustular psoriasis triggered by the cessation of immunosuppressants in a patient with pemphigus foliaceus, exploring the potential pathophysiological connections between these conditions.

Materials & Methods: We present the case of a 62-year-old female patient, managed at our dermatology department, diagnosed with pemphigus foliaceus and concurrently suffering from type 2 diabetes mellitus. The patient was undergoing treatment with azathioprine for her pemphigus foliaceus for a period of three years. During her treatment, she began experiencing migraines, which led to her growing dissatisfaction with the medication regimen. Subsequently, the patient made the decision to discontinue her azathioprine therapy. Within three days of stopping the medication, there was a marked exacerbation of her pemphigus foliaceus symptoms. This deterioration was characterized by the emergence of diffuse erosive, scaling, and crusted lesions across her scalp, body, and extremities. Notably, during this episode, she developed new pustular lesions specifically on the palms of her hands and the soles of her feet.

Results: To investigate these new symptoms, a histopathological examination was performed. The biopsy from the pustular lesions showed features typical of pustular psoriasis, including neutrophilic exocytosis with intraepidermal or subcorneal pustule formation. There was also noted edema in the papillary dermis and a mixed neutrophilic and eosinophilic infiltrate within the dermis. These histopathological findings confirmed the diagnosis of pustular psoriasis, developing concurrently with an exacerbation of pemphigus foliaceus post-immunosuppressant withdrawal. Following the diagnosis of pustular psoriasis in addition to the exacerbation of pemphigus foliaceus, a decision was made to administer an immunomodulatory treatment with rituximab. The patient received a total of four doses, each consisting of 1 gram of rituximab. This treatment was aimed at controlling the autoimmune activity underlying her conditions. After the completion of rituximab therapy and observing initial improvements, the patient was transitioned to a maintenance regimen with methotrexate to manage both pemphigus foliaceus and pustular psoriasis. She was prescribed a weekly dose of 25 mg of methotrexate. This strategy was intended to sustain the remission of her symptoms and prevent further relapses. The effectiveness of this approach was monitored through regular clinical assessments and periodic re-evaluation of her skin condition.

Conclusion: This case highlights the diagnostic challenges and management complexities when dealing with concurrent autoimmune dermatoses, especially following the abrupt cessation of immunotherapy.

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Abstract N°: 5219

Pemphigoid nodularis induced by gliptins

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Introduction and Objectives:

Bullous pemphigoid (BP) is a chronic subepidermal autoimmune blistering disease affecting elderly individuals. It is clinically characterized by intensive itch and tense blisters on erythematous background but some rare variants such as vesicular, erythrodermic, erythema multiforme-like, dyshidrosiform, and nodular BP, may also be observed. Any of the above-mentioned clinical forms of BP may be drug-induced and the list of culprit drugs is constantly increasing. Recently, dipeptidyl peptidase 4 inhibitors (DPP-4i) also known as gliptins, have been identified as potential triggers for BP, as highlighted in numerous studies. We present a patient with newly developed pemphigoid nodularis while undergoing gliptin therapy for underlying diabetes mellitus.

Materials and methods:

A 67-year-old woman with a history of diabetes mellitus treated with linagliptin for the last 2 years, presented with complaints of a severely pruritic rash of four-months duration affecting the scalp, trunk and extremities. This was accompanied by episodic blistering that progressed to ulceration upon scratching. The patient was initially treated with emollients and topical corticosteroids without improvement. The dermatological examination revealed a widespread polymorphic eruption on the trunk, and extremities consistent with both prurigo nodularis and BP, namely excoriated and umbilicated papules and nodules, single vesicles on erythematous urticaria-like background, and multiple small and large erosions covered with haemorrhagic crusts.

Results:

Routine laboratory findings indicated elevated blood sugar levels warranting attention. Histopathology examination was compatible with prurigo nodularis, showing epidermal hyperplasia and an excoriation with overlying crust, and mild perivascular lymphocytic infiltrate. Direct immunofluorescence analysis revealed linear deposits of IgG and C3 at the dermo-epidermal junction. ELISA BP180 and BP230 were strongly positive. Based on the clinico-laboratory findings the diagnosis of nodular BP was confirmed. After linagliptin was replaced with gliklazide, moderate dose of methylprednisolone, doxycycline, and high potency topical corticosteroids were administered resulting in a marked improvement.

Conclusion:

Pemphigoid nodularis is a rare atypical form of BP combining the clinical features of pemphigoid nodularis with the immunopathologic profile of BP. More than 50 cases of pemphigoid nodularis have been reported so far, but only in few of them a particular trigger could be identified in contrast to BP in general which can be precipitated by various physical or chemical agents, and medications, DPP-4i among them. A few cases of pemphigoid nodularis under gliptins were found in the literature, which may support linagliptin as a possible "culprit" in the present case. The favorable outcome after gliptin discontinuation is a further argument towards DPP-4i induced pemphigoid nodularis in those cases including ours.

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Abstract N°: 5221

Glutathione induced relapse of pemphigus vulgaris

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Introduction & Objectives:

Pemphigus vulgaris (PV) is a rare autoimmune blistering disease with chronic relapsing course which is characterized by the formation of intraepidermal blisters and erosions on the skin and mucous membranes. It is mediated by autoantibodies targeting desmosomal proteins, specifically desmoglein (Dsg) 1 and 3, leading to loss of cell-cell adhesion within the epidermis. The etiology of pemphigus remains unknown but multiple environmental factors or drugs may trigger its debut or relapses. The three main groups of drugs which have been reported to be associated with new onset or exacerbation of pemphigus are the thiol drugs, phenol drugs and non-thiol/non-phenol drugs. We report a patient with relapse of PV following infusions of glutathione, another thiol containing substance

Materials & Methods: #### A 49-year-old man with a 1-year history of mucosal-dominant PV presented with newly appeared bullae and erosions on the scalp, face and trunk as well as soft palate enanthema while being on long-term maintenance therapy with methylprednisolone 16mg/24h. The patient attributes the occurrence of the relapse to intravenous infusions with glutathione and vitamin C. Additionally, he has undergone physiotherapy using a device with varying radiofrequencies and was taking multiple herbal supplements.

Results: #### Routine laboratory investigations were within the reference ranges, except for slightly elevated D-dimers. Mycological examination from the oral cavity was positive for Candida albicans. The diagnosis PV was reconfirmed and systemic therapy with dexamethasone 8mg/24h with subsequent tapering in combination with systemic antibiotics, antimycotics and topical high potency corticosteroids resulted in progressive improvement.

Conclusion: #### The case presented underscores the need for vigilance regarding the potential triggers of PV or its relapses, especially in patients undergoing adjunctive therapies. Thiol drugs have sulfhydryl (-SH) group in their chemical structure and are probably the better studied group of drugs in the pathogenesis of drug-induced pemphigus. Glutathione and vitamin C infusions are currently widely administered as a cocktail with anti-oxidative and reparative potential applied both in healthy individuals and patients suffering from different diseases. Although they are perceived as innocuous, glutathione is a biologically active thiol substance and has been shown to induce acantholysis in human skin fragments under certain experimental conditions. While such interventions may offer perceived benefits, their impact on disease activity and treatment outcomes must be carefully evaluated.

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Abstract N°: 5317

Interest of Rituximab in pemphigus (Evaluation at Month 18)

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Introduction & Objectives:

Rituximab (RTX), a monoclonal anti-CD20 antibody, represents a major advancement in the treatment of pemphigus, ensuring both efficacy and tolerability. Currently,

its use as first-line therapy is recommended for moderate to severe cases. The aim of our study is

to evaluate and compare the effectiveness of repetitive RTX treatments up to the M18 regimen

in pemphigus treatment, aiming to optimize therapeutic strategies for this autoimmune disease.

Materials & Methods:

A retrospective cross-sectional descriptive and analytical single-center

study conducted at the Dermatology-Venereology Department of Ibn Sina University Hospital in Rabat over a 20-year period from 2003 to 2023.

Results:

69 patients were included with a female-to-male sex ratio of 1.65 and a mean age of 49 years, with an average consultation delay of 13.64 weeks. 43.5% of patients had pemphigus

vulgaris and 76.8% had severe pemphigus disease activity index (PDAI). Rituximab was

indicated in 50.7% of cases in “naive” patients (29% had severe, 14.5% moderate, and 7.2%

mild forms) and in 49.3% of cases in “non-naive” patients (37.7% were in relapse, 11.6% had

refractory pemphigus). The average duration of the consolidation phase was 5.6 weeks. At

Month 6, 46.4% were good responders. Complete remission was achieved on average at 6.5

months in 82.6% of patients at Month 6, 89.9% at Month 12, and 97.1% at Month 18,

demonstrating the efficacy of rituximab as first-line therapy in achieving complete remission at

Month 18 ($p<0.05$). The average prednisone dose at Day 1 was 80mg and 22mg at Month 6,

14.6mg at Month 12, and 4.8mg at Month 18. 15.9% of our patients relapsed over an average

duration of 19.9 months. 81.8% of relapse cases occurred in patients who skipped a rituximab

infusion, either at Month 6 or Month 12. Over time, 30 of our patients received maintenance

therapy at Month 24. Side effects were noted at doses <2g of rituximab, primarily infusion-

related reactions in the majority of cases.

Conclusion:

Our results highlight a significant efficacy of rituximab in the management of pemphigus, particularly as first-line therapy to achieve complete remission at Month 18 through the administration of successive infusions at Day 1/Day 15, Month 6, Month 12, and Month 18. Given the significant therapeutic advancements with rituximab in pemphigus treatment, our study serves as a reference for the development of multicenter studies aimed at establishing a national protocol for pemphigus management tailored to the Moroccan population.

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Abstract N°: 5336

The profile of autoimmune markers (antinuclear factor and thyroid antibodies) in a vitiligo sample and the possible link of autoimmunity with subjective sleep pattern

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Introduction & Objectives: Changes in circadian rhythm and the stress caused by poor sleep can lead to immune repercussions, with consequent role in the autoimmune mechanism of vitiligo. Signs of skin activity may be associated with dysregulation of etiopathogenic pathways, and concomitant autoimmune diseases, including those of thyroid, and positivity of antithyroid markers. The antinuclear factor (ANA) profile in vitiligo, especially related to phototherapy, is not totally clear. As ANA is not usually requested to vitiligo, this study aimed to identify the frequency of ANA and antithyroid markers in a sample with vitiligo; relate ANA with disease activity and phototherapy; and measure subjective sleep in the sample.

Materials & Methods: Study approved by the Ethics Committee. It was a cross-sectional study, including 30 patients with vitiligo, treated at a Dermatology Service, and 26 controls. Signs of vitiligo activity and previous phototherapy were registered as clinical variables. Clinical-epidemiological protocol and self-reported sleep questionnaire (Pittsburgh Sleep Quality Index) have been accessed. Serum autoimmune markers were measured in both groups: ANA and autoantibodies antithyroid peroxidase (anti-TPO) and antithyroglobulin (anti-TG). In the statistical analysis, the chi-square test was used to correlate ANA with the variables “phototherapy” and “disease activity”.

Results: Regarding clinical data, the average age of the vitiligo group was 47 years-old, with predominance of females (66.7%) and phototype III/IV (60%). Signs of lesions activity comprehended 63.3% of the sample. Previous UVB narrow band phototherapy was recorded in 43.3%. Most individuals (85% in vitiligo group and 62.5% of controls) have PSQI score>5 (classification as “poor sleeper” individual), indicating predominance of poor sleep quality. Reactive ANA occurred in 33.3% of the vitiligo sample, and 23.1% of the controls. Anti-TPO and anti-TGB positivity were, respectively, 26.7% and 20% in the vitiligo group (0% and 3.8% in the controls). The positivity of both these markers, in addition to reactive ANA, was 6.7% (vitiligo group) and 0% (controls). In vitiligo group, reactive ANA was analyzed with previous phototherapy (p=0.25) and vitiligo activity (p=0.24) through qui-square test, without statistical difference. Reactive ANA was detected in 46.2% of patients treated with phototherapy, and in 23.5% of those who not performed phototherapy. A total of 36.8% of individuals with active lesions presented reactive ANA; and 18.2% of those without vitiligo activity had reactive ANA. Regarding sleep, all patients with reactive ANA were “poor sleepers”.

Conclusions: Few studies have reported ANA analysis before phototherapy in individuals with vitiligo. This should be useful especially when associated autoimmune diseases and photosensitivity are suspected, due to the risks of their exacerbation due to phototherapy. Poor sleep, as a stressful condition, can be a potential trigger to vitiligo worsening. The inflammatory state can induce the activity of autoimmune diseases and positivity of autoimmune markers, such as ANA and those from thyroid, in addition to other pathways involved in the pathophysiology of vitiligo. We raise the importance of investigating the interaction of ANA and antithyroid antibodies with vitiligo activity, that may be triggered by sleep disturbance, and the behavior of ANA in the management of vitiligo with phototherapy.

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Abstract N°: 5350

Immunomodulatory therapy of patients with lichen planus

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Introduction & Objectives: Immunomodulatory therapy of patients with red squamous lichen planus

Lichen planus (LP) is characterized by polymorphism of clinical manifestations, especially lesions of the oral mucosa, which may be complicated by malignization. To date, not all aspects of the pathogenesis of LP have been disclosed, and there are difficulties in the treatment of this dermatosis.

Materials & Methods: 120 patients with LP were under observation, 80 (66.7%) of them were diagnosed with the classical form, 40 (33.3%) - with atypical forms (verrucous, atrophic, etc.) of dermatosis. All patients with atypical forms of LP and 50% of patients with the classical form of dermatosis had lesions of the oral mucosa. Inflammatory indices in the form of C-reactive protein (CRP, hs-CRP) and pro- and anti-inflammatory cytokines (IL-4, IL-8, IL-17, TNF-α) were studied by IFA. Injectable methotrexate (1.5 ml) weekly was used in the treatment of LP patients, 8-10 injections were used per course of treatment.

Results: the conducted treatment allowed to obtain positive clinical effect in 62 out of 80 (77,5%) with classical form and in 25 out of 40 (62,5%) with atypical forms of LP. CRP values were significantly decreased in both classic (hs-CRP: from 6.7±0.8 mg/mL to 1.3±0.05 mg/mL at $p<0.001$) and atypical (hs-CRP: from 9.1±1.3 mg/mL to 4.2±0.5 mg/mL at $p<0.05$) forms of LP. The values of anti-inflammatory cytokine increased both in classical (from 1.2±0.2 pg/mL to 4.5±0.3 pg/mL at $p<0.05$) and atypical (from 1.9±0.2 pg/mL to 3.3±0.7 pg/mL at $p<0.05$) forms of LP. The most informative of proinflammatory cytokines was TNF-α, which values significantly decreased during methotrexate treatment both in classical (from 16.1±2.5 pg/mL to 6.3±0.9 pg/mL at $p<0.001$) and atypical (from 20.5±3.3 pg/mL to 15.9±3.6 pg/mL at $p<0.05$) forms of LP.

Conclusion: Methotrexate injections should be used in the treatment of LP patients, especially with manifestations on the oral mucosa, which can have immunomodulatory and anti-inflammatory effects, contributing to the regression of rashes on the skin and oral mucosa.

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Abstract N°: 5368

Immunomodulatory therapy of patients with lichen planus

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Introduction & Objectives: Red squamous lichen planus (LP) is characterized by polymorphism of clinical manifestations, especially lesions of the oral mucosa, which may be complicated by malignization. To date, not all aspects of the pathogenesis of LP have been disclosed, and there are difficulties in the treatment of this dermatosis.

Materials & Methods: 120 patients with LP were under observation, 80 (66.7%) of them were diagnosed with the classical form, 40 (33.3%) - with atypical forms (verrucous, atrophic, etc.) of dermatosis. All patients with atypical forms of LP and 50% of patients with the classical form of dermatosis had lesions of the oral mucosa. Inflammatory indices in the form of C-reactive protein (CRP, hs-CRP) and pro- and anti-inflammatory cytokines (IL-4, IL-8, IL-17, TNF-α) were studied by IFA. Injectable methotrexate (1.5 ml) weekly was used in the treatment of LP patients, 8-10 injections were used per course of treatment.

Results: the conducted treatment allowed to obtain positive clinical effect in 62 out of 80 (77,5%) with classical form and in 25 out of 40 (62,5%) with atypical forms of LP. CRP values were significantly decreased in both classic (hs-CRP: from 6.7±0.8 mg/mL to 1.3±0.05 mg/mL at $p<0.001$) and atypical (hs-CRP: from 9.1±1.3 mg/mL to 4.2±0.5 mg/mL at $p<0.05$) forms of LP. The values of anti-inflammatory cytokine increased both in classical (from 1.2±0.2 pg/mL to 4.5±0.3 pg/mL at $p<0.05$) and atypical (from 1.9±0.2 pg/mL to 3.3±0.7 pg/mL at $p<0.05$) forms of LP. The most informative of pro inflammatory cytokines was TNF-α, which values significantly decreased during methotrexate treatment both in classical (from 16.1±2.5 pg/mL to 6.3±0.9 pg/mL at $p<0.001$) and atypical (from 20.5±3.3 pg/mL to 15.9±3.6 pg/mL at $p<0.05$) forms of LP.

Conclusion: Methotrexate injections should be used in the treatment of LP patients, especially with manifestations on the oral mucosa, which can have immunomodulatory and anti-inflammatory effects, contributing to the regression of rashes on the skin and oral mucosa.

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Abstract N°: 5456

Hair regrowth in patients with severe alopecia areata receiving long-term continuous treatment with baricitinib 4 mg up to Week 76

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Introduction & Objectives: Therapeutic efficacy has been demonstrated with continuous baricitinib treatment through Weeks 36 and 52 in severe alopecia areata. Pooled longer-term outcomes for all randomized patients eligible to continue the same dose have not been previously reported. We report Week 76 outcomes from all patients eligible to continue baricitinib 4-mg.

Materials & Methods: Data were pooled from the BRAVE-AA1/AA2 studies. At Week 52, non-responders (Severity of Alopecia Tool [SALT] >20 without significant eyebrow/eyelash improvement) were discontinued per protocol. This analysis integrates three populations: (1) responders (achieving SALT ≤20 at Week 52) who continued on baricitinib 4-mg; (2) responders rerandomized into down-titration or withdrawal arms (their data were censored and imputed based on the overall responder rate); (3) mixed responders (having significant eyebrow or eyelash regrowth by Week 52, or an unsustained SALT score ≤20 before Week 52) who continued on baricitinib 4-mg. Outcomes were achievement of SALT ≤20 and Clinician-Reported Outcome (ClinRO) score 0/1 (full hair or minimal loss) for eyebrows and eyelashes.

Results: At Week 52, 44.5% (as observed) of all baricitinib-treated patients from the BRAVE-AA trials achieved SALT ≤20. Among responders and mixed responders, the proportion of patients achieving SALT ≤20 increased to 75.8% at Week 76. The proportion of patients with ClinRO (0/1) increased from 49.5% (Week 52) to 80.0% (Week 76) for eyebrows and 49.6% to 77.0% for eyelashes.

Conclusion: Among patients receiving continuous baricitinib treatment, improvement in hair regrowth outcomes continued with higher proportions of patients achieving meaningful scalp hair, eyebrow, and eyelash regrowth over 76 weeks.

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Abstract N°: 5465

Among scalp non-responder patients with eyebrow/eyelash regrowth in the first year, continued treatment with baricitinib resulted in meaningful scalp responses for patients with severe alopecia areata

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Introduction & Objectives: Baricitinib is approved for the treatment of severe alopecia areata (AA). The timing and extent of hair regrowth with treatment can differ across body sites.

Materials & Methods: We evaluated scalp hair regrowth among baricitinib 4mg-treated patients with severe AA from two phase 3 trials (BRAVE-AA1/2) through 152 weeks, who achieved an eyebrow (EB) and/or eyelash (EL) response (score of 0 or 1 with ≥2-point improvement from baseline in Clinician Reported Outcome Measures for EB© and/or EL©) but no scalp response (Severity of Alopecia Tool [SALT] score >20) during the first year of treatment. The proportion of responders (SALT score ≤20) and SALT percent changes from baseline from Week-52 to Week-152, SALT score distribution at Week-52, and baseline disease characteristics are reported. Last observation carried forward was used to impute missing or censored data.

Results: Of 78 patients in the analysis population, 58 completed Week-152. At baseline, 74% had alopecia universalis compared to 46% in the intent-to-treat population. By Week-104, 39% had achieved SALT score ≤20, which persisted overall through Week-152. From Week-52 to Week-152, the mean percent improvement from baseline in SALT score increased from 35% to 55%.

Conclusion: This analysis, in a subgroup comprised predominantly of patients with alopecia universalis, suggests that continuous treatment with baricitinib beyond Week-52 in patients who have already achieved regrowth of EB or EL can allow a large proportion of them to ultimately achieve meaningful scalp hair regrowth.

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Abstract N°: 5470

Tofacitinib- a safe and effective agent in 3 cases of bullous pemphigoid

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Introduction & Objectives: Bullous pemphigoid (BP) is the most common subepidermal autoimmune blistering disease affecting elderly patients. BP is thought to occur due to autoantibody response toward hemidesmosomal proteins (BP180 and BP230. There is no cure for this condition. Topical, systemic corticosteroids & other agents have been used, but there are limitations due their inefficacy & side effects. There is growing data on the positive role of oral tofacitinib, a Janus kinase inhibitor (JAKi), for the treatment of autoimmune cutaneous diseases. This drug is now available at a relatively cheaper price in India. We report 3 cases of BP seen at a solo private Dermatology clinic in Mumbai, India. All 3 cases were treated with 5 milligrams (mg) of off-label oral tofacitinib twice daily. Baseline and follow-up laboratory evaluations were done (complete blood count, complete metabolic panel, glucose, lipid panel, and tuberculosis screening). ### Materials & Methods: Case 1

A 63-year-old woman presented for itchy lesions on her body of 3 weeks duration. There were tense bullae, erosions & erythematous plaques spread all over the skin. A clinical diagnosis of BP was confirmed by biopsy and elevated serum BP180 immunoglobulin G autoantibodies. This patient initially healed on prednisolne 40mg and dapsone 100mg combination over 2 months. But, developed severe anemia due to dapsone at 2 months, so it was stopped. She also developed steroid induced hyperglycemia. On tapering of prednisone, she developed new lesions. Mycophenolate mofetil 2 grams per day was ineffective. Thus, Tofacitinib was started after omitting other drugs. All lesions healed fully at 3 weeks. No side effects were seen.

Case 2

A 70-year-old woman presented with 1-month history of severe itching & bullous lesions on her body. Physical examination showed erythematous patches, tense bullae and erosions all over body. The diagnosis was confirmed by histopathology & positive antibody to BP-180 antigen. She was steroid dependent & had developed steroid induced diabetes as well as other side effects. She was not responding to other agents. It was then decided to start tofacitinib. After starting oral tofacitinib, she had complete clearance of lesions at 5 weeks. She tolerated the tofacitinib well.

Case 3

A 70 year old woman presented with severely itchy bullous lesions all over the body and multiple erosions on the skin. She was a known diabetic, hypertensive & was on anti-epileptics too. The skin biosy & positive autoantibody to BP-230 antigen confirmed the diagnosis of BP. She did not respond to doxycycline-niacinamide combination. Systemic steroids were not considered in view of high blood sugars.Thus, a decision was made to start Tofacitinib. All the lesions healed at 12 weeks. She continues to be on follow up monitoring.

Results: BP is a chronic and highly debilitating disease. Our first 2 patients had failed topical and systemic commonly used drugs. In case 3, steroids were not considered in view of high blood sugars. In case 1, blisters and pruritus subsided fully at 3 weeks showing a fast onset of action of this drug. Case 2 healed at week 5 while case 3 did so at week 12.

Conclusion: Tofacitinib is a good agent in managing bullous pemphigoid with complete healing in all the 3 cases. At the dose of 5mg twice daily, there were no significant side effects. It is a good option to use in resource poor

setting like India.

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Abstract N°: 5471

Long-term efficacy of baricitinib in alopecia areata: 3-year results from BRAVE-AA1 and BRAVE-AA2

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Introduction & Objectives: This analysis evaluates long-term efficacy over three years of continuous therapy in baricitinib 4mg and 2mg Week 52 (W52) responders (SALT score ≤20 (≤20% scalp hair loss)) with severe alopecia areata (AA).

Materials & Methods: Integrated data from the BRAVE-AA1/BRAVE-AA2 phase 3 trials included patients with Severity of Alopecia Tool (SALT) scores ≥50 (≥50% scalp hair loss) randomized to baricitinib, who had achieved SALT score ≤20 at W52 and remained on the same dosage through Week 152 (W152). W152 outcomes included the proportions of patients maintaining SALT score ≤20 and those achieving or maintaining eyebrow and eyelash regrowth measured by Clinician-Reported Outcomes (ClinRO) for Eyebrow and Eyelash Hair LossTM scores 0/1 (full coverage/minimal gaps) and ≥2-point improvements from baseline (among those with baseline scores ≥2). Data after permanent treatment discontinuation or rescue were censored; missing data is handled with modified last observation carried forward imputation.

Results: Among baricitinib 4mg-treated and 2mg-treated W52 responders, respectively, 115/129 (89.1%) and 56/67 (83.6%) maintained SALT score ≤20 at W152; among W52 responders with baseline ClinRO scores ≥2, 64/80 (80%) and 26/39 (66.7%) had ClinRO Eyebrow (0,1) with ≥2-point improvements from baseline regrowth, and 55/68 (80.9%) and 25/32 (78.1%) had ClinRO Eyelash (0,1) with ≥2-point improvements from baseline regrowth at W152, comparable with results reported at W104.

Conclusion: Considering the known safety profile of baricitinib, the high proportion of W52 responders maintaining efficacy over 3 years as demonstrated by scalp hair, eyebrow, and eyelash regrowth, supports the long-term continuous use of baricitinib in severe AA.

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Abstract N°: 5475

Safety analysis of baricitinib in adult patients with severe Alopecia areata from 2 randomized clinical trials over a median of 2.3 years and up to 4 years of exposure

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Introduction & Objectives: We report pooled safety data for baricitinib in patients with severe alopecia areata (AA) from BRAVE-AA1 (NCT03570749; phase 2/3) and BRAVE-AA2 (NCT03899259; phase 3), including the long-term extension periods.

Materials & Methods: Data are reported from the all-BARI dataset, combining all patients receiving ≥1 dose of baricitinib (1 mg, 2 mg or 4 mg) at any time during the trials. Safety outcomes include treatment-emergent adverse events (TEAEs), adverse events (AEs) of special interest, and abnormal laboratory changes. Incidence rates (IRs) per 100 patient-years were calculated based on time at risk. Data cut-offs were June/22/2023 for BRAVE-AA1 and May/08/2023 for BRAVE-AA2.

Results: In all-BARI, 1303 patients received ≥1 dose of baricitinib, reflecting 2789.7 patient-years of exposure (median 825 days, maximum 1460 days). Most TEAEs (93.2%) were mild to moderate in severity. IRs of serious AEs (IR=2.6) and treatment discontinuations due to AEs (IR=1.7) were generally low and remained similar to previous analysis (May 2022). Since the previous analysis, there were no new cases of serious infections, opportunistic infections, major adverse cardiovascular events, deep vein thromboses, or pulmonary embolisms. The IRs for non-melanoma skin cancer (IR=0.1) and other malignancies (IR=0.2) remained stable over time. The IR of herpes zoster was comparable to prior reports (IR=1.9). Laboratory changes were generally consistent over time. No deaths have been reported in either study.

Conclusion: This safety analysis in patients with severe AA is consistent with previously reported data from the baricitinib AA clinical trial program.

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Abstract N°: 5480

Outcomes of down-titration in patients with severe alopecia areata treated with baricitinib: An update through Week 152 from BRAVE-AA2

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Introduction & Objectives: Baricitinib is approved for the treatment of patients with severe alopecia areata (AA). The efficacy results from the down-titration portion of the phase 3 trial BRAVE-AA2 have been reported up to Week-104. Here we report both long-term efficacy outcomes from the down-titration portion of BRAVE-AA2, and recapture data after retreatment.

Materials & Methods: BRAVE-AA2 enrolled 546 adults with severe AA (Severity of Alopecia Tool [SALT] score ≥50). At Week-52, patients initially randomized to 4mg baricitinib who had scalp hair response (Responders: SALT score ≤20) were eligible to be rerandomized 1:1 to remain on 4mg or down-titrate to 2mg. Retreatment with baricitinib 4mg for those patients who were down-titrated was triggered by a loss of treatment benefit defined as a 20 points or greater worsening in SALT score from Week-52. Last observation carried forward was used to impute missing or censored data.

Results: Among 86 responders at Week-52, 42 were down-titrated from 4mg to 2mg baricitinib. At Week-104 and 152 respectively, 66% and 59% of down-titrated patients had maintained a SALT score ≤20, compared to 91% and 89% of responders who remained on baricitinib 4mg. Among down-titrated patients, at Weeks 104 and 152, 29% and 37% experienced a loss of treatment benefit, respectively. For patients that remained on 4mg baricitinib, these proportions were 5% and 7%. Among patients who had achieved a SALT score ≤20 by Week-36 and had maintained it up to Week-52 before down-titration (sustained response), loss of treatment benefit by Week-152 occurred in 33% (11/33), compared to 50% (4/8) among patients who had not achieved sustained response. Among patients experiencing loss of benefit and retreated with baricitinib 4mg, the proportion of patients recapturing SALT score ≤20 increased over time, reaching 67% (10/15) for patients with ≥48 weeks of retreatment.

Conclusion: These data show that more than half of patients down-titrated to 2mg maintained clinical response up to 2 years after the down-titration. While more work is needed to define timing and conditions for a successful down-titration, data suggests that dosing of baricitinib can be modulated between 2mg and 4mg to adapt to the clinical response.

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Abstract N°: 5494

Assessing the quality of life and anxiety-depressive disorders in patients with pemphigus

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Introduction & Objectives: Pemphigus is characterised by its chronic course, often associated with disability and the threat of serious complications, which have a considerable impact on daily life. Today, there is no doubt about the impact of pemphigus on the quality of life (QoL) of those affected, or about its involvement in anxiety-depressive disorders.

Our objective is to determine the quality of life and anxiety-depressive disorders and the factors associated with them in patients with pemphigus.

Materials & Methods: A cross-sectional study of patients with pemphigus recruited from a tertiary care hospital from January 2022 to June 2023. The DLQI and HAD-S questionnaires were completed.

Results: fifty-four patients were recruited (27 PV and 27 PF). The mean age was 46.83 years. The sex ratio Female/male was 3, 15/1. Rural origin was noted in 57.4% of cases, and low socio-economic status in 72%. Severe disease was observed in 39% of patients, and pruritus was reported in half of them. The median time to clinical remission was 13 months. The median follow-up was 24 months, with 44.4% of patients having 3 or more relapses during follow-up and a median hospital stay of 13 days. Corticosteroid-induced diabetes was noted in 37% of patients and an infectious complication in 50%. The mean DLQI was 11.04±7.23. The mean HAD for Anxiety score was 10.20±4.67, with anxiety symptoms present in 63% of patients, and the mean HAD for Depression score was 8.22±5.07, with depressive symptoms present in 37% of patients.

In the multivariate analysis, the severity of the disease and the occurrence of Corticosteroid-induced diabetes were retained as independent factors predicting poor QoL. Similarly, disease severity and high-dose corticosteroid therapy were factors independently associated with depression and anxiety.

Conclusion: We recommend assessing the psychological impact and QoL of patients with pemphigus. This dimension should be integrated into the multidisciplinary management of patients in order to diagnose psychosocial problems quickly and to request the necessary therapeutic consultation.

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Abstract N°: 5499

Atypical pemphigus vulgaris with isolated nail involvement

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Introduction & Objectives:

Pemphigus vulgaris (PV) is an autoimmune blistering dermatosis mediated by circulating and in vivo bound anti-desmoglein (Dsg) autoantibodies. The clinical hallmark of PV are the flaccid blisters and painful erosions primarily affecting the skin and mucous membranes. Nail involvement is relatively uncommon and underreported. When available it is regarded as closely correlated with disease severity. The most common nail symptoms observed are paronychia and Beau lines but onychomadesis, onycholysis, nail discoloration, pitting or even complete nail loss are uncommon. We report a rare and difficult for recognition case of PV with isolated nail involvement.

Materials & Methods:

A 65-year-old woman presented with nail abnormalities affecting the nails on her hands and feet. The clinical observation revealed periungual erythema, paronychia, Beau lines, subungual hyperkeratosis, onychodystrophy and nail brittleness. These lesions were initially interpreted as onychomycosis but the respective antimycotic treatment was ineffective. Upon admission routine laboratory, mycological investigation from the nails, histology, direct immunofluorescence (DIF) microscopy from perilesional periungual area, indirect IF (IIF), and enzyme-linked immunosorbent assay (ELISA) tests for anti-Dsg autoantibodies were performed.

Results:

Routine laboratory parameters indicated mild leukocytosis, eosinophilia, and elevated gamma- glutamyl-transferase. Histological examination demonstrated spongiosis and intraepidermal vesicles. DIF on perilesional skin showed intercellular deposition of IgG (+) in the lower third of the epidermis. ELISA anti-Dsg1 and Dsg3 tested negative. Based on the clinico-laboratory findings we confined to the diagnosis of PV with isolated nail involvement. Treatment with moderate dose systemic and topical corticosteroids resulted in clinical improvement.

Conclusion:

Nail involvement in PV is rare and is usually seen in severe cases but it may also be an isolated primary manifestation of the disease. The most frequently reported presentations were paronychia and Beau lines, but almost all types of nail pathology may be observed. The mechanisms underlying nail involvement in PV remain unclear, but it is hypothesized to involve differences in the expression of certain proteins in the nail tissue compared to skin and mucous membranes. An intriguing finding in our case of isolated nail PV is the negative ELISA testing, which may be due to such low quantity of antibodies that are deposited only *in vivo* in the most sensitive nail matrix. Treatment typically involves systemic and topical corticosteroids, which can lead to improvement in both nail and muco-cutaneous lesions.

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Abstract N°: 5589

Anifrolumab in cutaneous lupus erythematosus: real-life data from a tertiary care hospital in Spain

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Introduction & Objectives:

Cutaneous lupus erythematosus (CLE) is a chronic autoimmune skin pathology characterized by various morphological subtypes, all sharing a cytokine profile where type I interferon (IFN-I) plays a key role. To date, there are no drugs specifically indicated for CLE, with off-label use of medications approved for systemic lupus erythematosus (SLE). Anifrolumab, a monoclonal antibody IFN-I receptor subunit 1 (IFNAR1), was approved by the EMA in February 2022 for SLE. Secondary objectives of the TULIP-2 clinical trial showed significant benefits in CLE. In June 2023, the Spanish Ministry of Health approved anifrolumab funding for moderate-to-severe active SLE with positive antinuclear antibodies (ANA). We present our experience with anifrolumab in patients with severe and refractory CLE in SLE.

Materials & Methods:

A descriptive retrospective study was conducted on all patients with severe and refractory CLE in SLE undergoing monthly treatment with 300 mg anifrolumab from July 2023 to April 2024 at the Dermatology Department of a tertiary referral hospital in Spain. Study variables included gender, age, CLE subtype, systemic involvement of SLE, autoantibodies, prior treatments, pre- and post-treatment CLASI-A (CLE Area and Severity Index Activity), response and follow-up time, treatment optimization, and adverse effects.

Results:

Six patients were included (Table 1), all female, with a median age of 50 years (range 19-65). The median duration of SLE progression was 15 years (range 2-42). The most prevalent systemic involvements were lymphopenia, polyarthritis, and lupus nephropathy. All patients expressed ANA, with anti-Ro and anti-dsDNA specificities being the most predominant. The most observed subtypes of CLE were discoid lupus erythematosus (DLE) and acute lupus erythematosus (ACLE). The median number of previous treatments used was 4 (range 4-12). All patients showed a rapid remission of CLE lesions after a median of 2 months (range 1-3), corresponding with two anifrolumab infusions. The median CLASI-A decreased from 35 (range 17-47) to 2 (range 0-5). In three patients, treatment was optimized every 2 months without recurrence. No improvements were observed in non-cutaneous SLE manifestations. There were no serious adverse effects.

Conclusion:

Anifrolumab presents as a promising therapy for CLE. Its efficacy in managing cutaneous symptoms surpasses that of belimumab and rituximab, previously the only approved biologic agents for SLE. Furthermore, its efficacy and safety profiles, as observed in clinical trials and real-world studies, support its use in refractory cases of CLE, with notable reductions in disease activity and minimal adverse effects. The potential for complete remission and the option for dose optimization further enhance its clinical utility in CLE.

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Abstract N°: 5665

Proteomic analysis reveals different top canonical pathways and top upstream regulators in dermal infiltrates of cutaneous lupus erythematosus and dermatomyositis

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Introduction & Objectives:

Systemic lupus erythematosus (SLE) and dermatomyositis (DM) are systemic autoimmune diseases often affecting the skin. The histologic features of skin biopsies from patients with cutaneous lupus (CLE) and DM are indistinguishable despite the differential clinical manifestations with respect to distribution of the rash and organ system involvement. The presence of inflammatory infiltrate under an interface dermatitis is a well-known histopathological feature in CLE and DM and in a previous study we found CD66b+ neutrophils to be increased in both symptomatic and non-symptomatic skin of SLE and DM patients when compared to healthy controls. By proteomic techniques we previously also identified IL-16 as a key cytokine in CLE but not in DM. It is not clear what molecular pathways play a role contributing to the clinical differences and histopathological similarities between CLE and DM. To explore this further in the same proteomic database, with the aim to reveal the top canonical pathways and top upstream regulators in CLE and DM compared to controls, we performed pathway analyses. Our aim was to systematically identify and explore the top canonical pathways and top upstream regulators in CLE and DM compared to controls for better understanding the pathogenesis of these two different autoimmune diseases.

Materials & Methods:

Skin biopsies from six patients with CLE, five patients with DM and six controls were investigated. Biopsy sections were examined by a pathologist and inflammatory dermal infiltrates and control dermis were micro-dissected by laser capture and protein content measured by nano-LC tandem mass spectrometry. Data was analyzed by String and Ingenuity Pathway analysis (IPA). P-values<0.05 were considered significant, adjustment for multiple testing was performed.

Results:

The top canonical pathways in CLE identified by IPA were the glucocorticoid receptor signalling, neutrophil extracellular trap signalling, interferon signalling, EIF2 signalling and antigen presentation pathways (p-value range 2.59E-06 to 4.88E-08). The identified top canonical pathways in DM were the protein ubiquitination, antigen presentation, microRNA Biogenesis signalling, microautophagy signalling and BAG2 signalling pathways (p-value range 4.49E-05 to 3.91E-10).

The top upstream regulators identified by IPA in lupus were IL4, interferon alpha, CHROMR, TREX1 and dexamethasone (p-value range 1.21E-24 to 7.20E-28). In DM, the top upstream regulators were tumor protein P53 (TP53), interferon alpha, NKX2-3, oncostatin M (OSM) and SN-011 (p-value range 1.69E-11 to 2.77E-15).

Conclusion:

Our comparative proteomic analysis revealed different key canonical pathways and different upstream regulators in CLE and DM. This novel information may contribute to a better understanding of the molecular pathogenesis of respective disease, and what pathways might be of interest for pharmaceutical targeting.

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Abstract N°: 5818

Dapsone and Surgery Combination in the Pemphigus Vegetans Management: A Case Report

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Introduction & Objectives:

Pemphigus vegetans is a rarely seen subtype of pemphigus vulgaris and characterized by vesicles, bullae, and pustules that extend and coalesce; then evolve into vegetating plaques on intertriginous areas. Mucosal involvement, and the cerebriform tongue can be one of the features of the disease. While systemic corticosteroids and immunosuppressants are the mainstay of treatment of pemphigus vegetans, surgery can be an alternative on the resistant vegetative plaque management.

Materials & Methods:

Here, we report a patient with pemphigus vegetans diagnosis who treated with dapsone and surgery combination.

Results:

A 30-year-old male patient with a known pemphigus vegetans diagnosis was evaluated by us. The patient had comorbidities of type-2 diabetes mellitus and obesity. In the anamnesis taken from the patient, he defined that his complaints started as discharged lesions located in the inguinal region three years ago, and over time, new lesions developed in the axilla, abdomen and nape, and the lesions grew and turned into swollen masses. During this period, the patient, who had used systemic corticosteroid, azathioprine, and mycophenolate but did not benefit and was diagnosed with pemphigus vegetans by clinical examination, histopathological evaluation and serological analyses performed by us a year ago. At the first-year follow-up of the patient, who treated with dapsone (200 mg/day), no complications or recurrence were observed, it was determined that the lesions had largely regressed and healed with post-inflammatory hyperpigmentation, and the discharge had completely disappeared. On dermatological examination, there was post-inflammatory hyperpigmentation in the abdomen, inguinal region, axilla, and vegetative plaque on the nape. The patient, whose disease activity was under control and whose quality of life had increased significantly, stated that the lesion on the back of the neck made it difficult for him to perform daily tasks. Upon treatment resistance to intralesional corticosteroid treatments, it was decided to excise the patient's lesion and to excise the lesion in two stages to evaluate the wound healing process after excision. After sterile conditions were achieved, half of the lesion was excised with a scalpel down to the subcutaneous fat tissue under local anesthesia. The patient was followed up with regular dressings following the procedure. At the first month follow-up, it was observed that the procedure area was completely healed without recurrence, so the other half of the lesion was excised in the same way. During the patient's follow-up, an improvement was observed in the procedure area. The patient continues dapsone treatment (100 mg/day) and is being followed without any recurrence.

Conclusion:

Although pemphigus treatment is basically a treatment based on immunosuppression, excision of vegetative plaques after disease activity in pemphigus vegetans is suppressed may be important, especially to improve the quality of life of patients. There is limited data in the literature regarding pemphigus vegetans surgery. In our case, surgical excision of the vegetative plaque was performed after the disease activity was suppressed with dapsone.

With this case report, we would like to emphasize that the dapsone and surgery combination may be an effective combination in the management of pemphigus vegetans and want to point out that surgical excision may be a treatment method in resistant vegetative plaques.

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Abstract N°: 5843

Facial srcoidosis as an important diagnosis for clinical outcome

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INTRODUCTION: Sarcoidosis is a rare disease of unknown etiology, resulting from the interaction between environmental triggers and genetic predisposition. The clinical presentation is variable, with the most common nonspecific symptoms being fatigue, reduced concentration, weight loss, fever, and night sweats. However, the presentation depends on the degree of organ involvement and may vary over time. The disease can affect any organ, but its most common form involves the lungs, causing cough, dyspnea, and chest pain. Diagnosis involves clinical and anatomopathological evaluation, as well as exclusion of other diseases. Treatment is reserved for symptomatic patients and may include corticosteroids and immunosuppressive agents. **CASE REPORT:** A 50-year-old female patient presents with complaints of lesions on the face for approximately 3 months, after being scratched by a cat. The patient also reported a weight loss of 10kg in one month, dizziness, and asthenia. On physical examination, she presented erythematous-infiltrated papules and plaques, ranging from 0.5 to 1.2cm in diameter, in the region of the right auricular pavilion, right cervical, right pre-auricular, left mandibular, and left frontal areas. Based on the clinical picture and personal history, the diagnostic hypotheses of Cat Scratch Disease and sarcoidosis were raised. Local biopsy showed: diffuse dermal granulomatous infiltration, with numerous epithelioid granulomas and numerous multinucleated giant cells. Serology for Bartonella Hanselae and quintana were non-reactive (negative IgM and IgG). Chest computed tomography revealed a micronodular pulmonary infiltrate, characterized by perilymphatic and peribronchovascular nodules, mild lymphadenopathy, mediastinal lymphadenomegaly + bilateral mammary nodules. Transbronchial biopsy showed exuberant chronic granulomatous inflammatory process in bronchial wall and lung parenchyma, rich in Langhans-type giant cells and epithelioid cells, surrounded by fibrosis. PAS and Grocott fungal stains were negative, as well as the ziehl-neelsen acid-fast bacilli (AFB) stain. **CONCLUSION:** Sarcoidosis is a disease with variable clinical presentation, thus adequate history-taking and clinical evaluation are crucial in diagnosis. It is essential for the physician to be attentive to the clinical signs of disease progression in order to prevent morbidity and mortality and improve the patient's quality of life.

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Abstract N°: 5860

Autoimmune Blistering Diseases: Characterizing Remission Status, Quality of Life, and Disability

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Introduction & Objectives: Modern treatments for autoimmune blistering diseases (AIBDs) have considerably improved mortality rates, with greater numbers of patients experiencing remission. Despite these advances, patients with AIBDs may have limited access to treatments, may require systemic steroids for more prompt response, may not achieve remission, and may experience significant disability and impairment in quality of life (QOL). The objective of this study was to quantify and describe the daily impact of living with an AIBD, in terms of symptom expression, physical functioning, disability, and quality of life. The influence of remission status on current functioning was also examined.

Materials & Methods: An anonymous survey was distributed to all participants of the International Pemphigus & Pemphigoid Foundation Externally-Led Patient-Focused Drug Development Meeting (January 2023). The questionnaire battery included the Autoimmune Bullous Disease Quality of Life Questionnaire and key items evaluating treatment and remission history and work-related disability.

A total of 437 patients with AIBDs completed the survey. Of these, 45% (n=196) had pemphigus vulgaris, 24% (n=103) had bullous pemphigoid, 22% (n=98) had mucous membrane pemphigoid, 7% (n=32) had pemphigus foliaceus, and 2% (n=8) reported “other” disease. The sample was 73% (n=320) female.

Results: Over one-half (51%) of the sample had never achieved remission; 19% had remitted with multiple relapses, 16% had remitted with one relapse, and only 14% had remitted with no relapse. Of those who had experienced remission, 45% had a relapse within one year. Participants reported that pain (51%), difficulty eating or swallowing (39%), mental health (38%), and fatigue (34%) exerted the most significant impact on their lives. The majority of the sample (59%) reported work-related disability, with 48% missing work due to initial disease activity and 30.4% missing work due to relapse/flare activity. Remission status was significantly related to disease-specific quality of life ($p<.001$), with those who had never remitted reporting significantly worse QOL than those who had remitted in full. Participants who had relapsed reported intermediate QOL scores. Mean QOL scores for all remission groups are presented in the figure.

Conclusion: Patients with AIBDs experience high rates of disability and significant impairments in quality of life, which are magnified for those who have not experienced full remission. These data suggest that despite significant treatment advancements, there is a need to address many clinically significant symptoms that affect the wellbeing of patients with AIBDs.

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Abstract N°: 5879

Tackling the Complexity: A Case of Bullous Cutaneous Lupus Erythematosus

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Introduction: Bullous cutaneous lupus erythematosus is a rare form of lupus that mainly affects the skin, resulting in painful blisters and skin lesions. **Case Report:** A 47-year-old female patient, presented with an intense outbreak of blisters all over her body. These blisters initially appeared on the scalp and face, spreading from cranial to caudal with a sensation of pain and burning in the lesions. She had areas with ulcerated lesions on her face, cheek regions, dorsum of the nose, and auricular pavilions, with areas of atrophy. She also had lesions on the oral mucosa, ulcerated, and presented lymphadenopathy, mainly cervical and axillary. She reported episodes of chills and overall compromised health. Her treatment was started with prednisone 80mg/day, with little response in 15 days, and she opted for hospitalization for corticosteroid pulse therapy. Subsequently, hydroxychloroquine 400mg/day was added, with a good therapeutic response. **Conclusion:** The case illustrates the complexity and importance of early diagnosis and proper management of rare dermatological conditions, such as bullous cutaneous lupus erythematosus. The varied clinical presentation and specific characteristics of the skin lesions required a multidisciplinary approach and careful consideration of available therapeutic options.

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Abstract N°: 5925

Real-life experience of anifrolumab to treat lupus erythematosus patients at a third- level University Hospital

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Introduction & Objectives:

Anifrolumab is a human IgG1κ monoclonal antibody recently approved for adults with moderate to severe systemic lupus erythematosus (SLE). The approved dose is 300 mg administered intravenously every 4 weeks. Anifrolumab acts by binding to type I interferon (IFN) receptor, inhibiting its signaling pathway. The objective of the present work was to collect the experience of lupus patients treated with Anifrolumab at a third-level University Hospital in Spain. The remission was defined as follows: complete remission (CR) was considered as achieve totally inactive disease; partial remission (PR) as clinical improvement but not CR.

Materials & Methods and Results:

Our series consisted of 11 women (8 Caucasian, 1 Mestiza, 1 Arabian and one Asian) with a median of 43 years (range 36-58). The median time since lupus diagnosis was 16 years (range: 9-34). All patients had received at least 2 doses of the drug (until March 2024) at our center to be included. One patient had SLE without cutaneous lupus erythematosus (CLE); 3 presented SLE+CLE, with SLE symptoms predominating; 5 had SLE+CLE with cutaneous symptoms predominating; and 2 presented exclusively CLE. All patients had previously received prednisone and antimalarials, in addition to an average of 2.8 immunosuppressant/biologic drugs (azathioprine, methotrexate, mycophenolate, rituximab or belimumab). All patients have responded to anifrolumab (response rate: 100%), with more than half have achieving CR after the first infusions of the drug (CR rate: 55%; PR rate: 45%). No differences in effectiveness were observed among the different types of lupus nor according to the previous treatments received. The median of anifrolumab infusions was 5 (range 3-8), so patients have received the drug for a median of 5 months. Peripheral blood type I IFN gene signature of 28 genes was performed in 4 patients. It showed high interferon signature values before starting anifrolumab, that normalized after a few months with the drug. Only 3 patients presented mild and short-term adverse events: gastrointestinal symptoms (abdominal pain, nausea and diarrhea); mild gluteal hidradenitis; and infectious bronchitis that required antibiotics. No herpes zoster infections were seen during follow-up.

Conclusion:

In our series of 11 patients, anifrolumab was effective in all of them. responded and 55% achieved a complete response with few infusions. No differences among the different lupus types were observed. Furthermore, the expression of IFN-related genes normalized after the initiation of anifrolumab in some patients. Only three patients have presented mild adverse events that did not require drug discontinuation.

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Abstract N°: 5929

Reticular erythematous mucinosis developed in an atypical body part associated with thyroid disease and menstrual cycle: a rare case report

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Introduction

Reticular erythematous mucinosis (REM) is a rare form of diffuse cutaneous mucinosis.It often presents as erythematous macules or papules that coalesce into a reticulated pattern in the midline of the upper chest or back of middle-aged women1-3. Although the pathophysiology has been poorly identified, correlations have been observed with autoimmune disorders and various types of cancers1-5.We report a rare case of REM that affected the left upper forearm and was triggered by menstruation, sunlight and thyroid disease.

Case

This is a case of a 48-year-old female patient, presenting with longstanding unilateral reticulated erythro-violaceous papules in the anterior part of the left forearm. They were itchy, photosensitive and deteriorating before menstruation. The exanthema was previously managed unsuccessfully with topical corticosteroids. The medical background included iron-deficiency anaemia and orally treated hypothyroidism, following thyroidectomy.

Results

Thorough investigations including autoimmune screen were unremarkable. The histology with Hematoxylin-Eosin stain revealed predominantly tense perivascular infiltration by small lymphocytes at the level of the reticular dermis, focal mucus deposition in dermis (Alcian Blue) and a normal epidermis.** Immunohistochemistry revealed T lymphocytes with CD2+, CD3+, CD4+, CD5+, CD7+ more than CD8+, TCRδ- and a Ki67/MiB1 cell proliferation index around 5% revealing the diagnosis of REM1-5. Therefore, we treated with photoprotection, topical clobetasol proprionate 0,05% cream for a month followed by Tacrolimus 0,1% ointment and Hydroxychloroquine 200mg twice daily6. We noticed complete clearance within two months. A trial to reduce the Hydroxychloroquine was unsuccesful, so we maintained the initial dosage. Six months later she remained exanthema-free.

Conclusion

To summarise, we presented a rare case of REM in an atypical body part treated succesfully with Hydroxychloroquine. This case highlights the need for biopsy in cases of longstanding recalicitrant exanthemas.Additionally, we noted an association with hypothyroidism and we urge the scientific community to investigate the conjunction to thyroid disorders further. A correlation with hormonal changes during menstrual cycle was likewise observed. The pathophysiology behind this is yet to be clarified. Finally, the success of antimalarials as a first line therapy of REM has been attested in our case while only limited response to topical corticosteroids was observed6

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Abstract N°: 5972

scRNA-seq of human vulva lichen sclerosus reveals complex networks of fibroblasts involvement and a role for ASPN in extracellular matrix synthesis and CD8+ T cell chemotaxis

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Introduction & Objectives:

To explore the potential role of Asporin (ASPN) in the pathogenesis of vulva lichen sclerosus (VLS).

Materials & Methods:

scRNA-seq was used to compare the skin lesions of patients with VLS and normal vulvar skin tissues to analyze the core role of fibroblasts in VLS and the function of communication with related cells. Immunofluorescence, RT-qPCR, Western Blot and Transwell techniques were used to explore the role of ASPN in extracellular matrix synthesis and CD8+ T cell chemotaxis in fibroblasts.

Results:

The skin tissue was divided into 12 cell subsets by scRNA-aeq. The differential genes of fibroblasts composed of VLS are enriched in the pathways of collagen synthesis and extracellular matrix. Fibroblasts were divided into four subtypes and only the proportion of Mesenchymal fibroblasts subsets increased significantly in VLS. The differential genes of the four fibroblast subsets were enriched in the collagen synthesis pathway, and the differential genes of Pro-inflammatory, Mesenchymal and Secretory-reticular fibroblasts were enriched in the MHC protein complex recognition pathway. In the disease group, the intercellular communication between fibroblast subsets was enhanced, and the communication between CD8+ T cells and fibroblast subsets in COLLAGE and other pathways was enhanced. The expression of ASPN was increased in fibroblasts of VLS tissue. ASPN promoted the synthesis of collagen Ⅰ, Ⅲ, Ⅳ and FGF2 in fibroblasts, inhibited the expression of elastin. It promoted the migration of CD8+ T cells by secreting chemokines such as CXCL9 and CXCL11.

Conclusion:

Fibroblasts in VLS can be divided into four subtypes, all of which promote the formation of extracellular matrix and enhance communication with CD8+ T cells. ASPN in VLS can promote fibroblasts to synthesize extracellular matrix and promote the chemotaxis of CD8+T cells by secreting chemokines.

Keywords: vulvar lichen sclerosus, single-cell RNA sequencing, fibroblasts, ASPN, extracellular matrix

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Abstract N°: 6013

IgA pemphigus manifested as acrodermatitis continua of Hallopeau: a case report

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Introduction & Objectives:

IgA pemphigus is a rare autoimmune blistering disease characterized by acantholysis and tissue-bound and circulating IgA antibodies targeting components in the epidermis.

Materials & Methods:

A 41-year-old female with a 17-year history of refractory IgA pemphigus was under remission with acitretin 30mg/day when periungueal pustules, erythema and edema and nail plate loss suddenly developed on the 4th toe. Erythematous scaling plaques with pustules were also observed on the palms and soles. The initial clinical hypothesis was acrodermatitis continua of Hallopeau.

Results:

A new biopsy revealed acantholysis and intercellular intraepidermal IgA fluorescence without evidence of infection confirming the diagnosis of IgA pemphigus. Complete improvement occurred 3 months after the association of colchicine 1.0mg/day to acitretin.

Conclusion:

As the clinical and histopathological features of IgA pemphigus may resemble pustular psoriasis, Sneddon-Wilkinson disease, impetigo, and pemphigus foliaceus, definitive diagnosis mostly relies on IF studies. The concomitant occurrence of psoriasis and IgA pemphigus in the same patient has been seldom reported. Some authors advocate that both belong to the same spectrum of neutrophilic dermatoses. Patients with long-standing pustular psoriasis with negative IF studies may later develop detectable serum IgA autoantibodies against cell surface antigens. Our patient had the diagnosis of IgA pemphigus since the beginning of the cutaneous disease with c-DNA transfection test revealing positivity to desmocollin 1. To the best of our knowledge, this is the first report of IgA pemphigus manifested as acrodermatitis continua of Hallopeau.

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Abstract N°: 6059

Psychosocial Impacts of Autoimmune Blistering Diseases: Results from a Large Cross-sectional Survey

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Introduction & Objectives: Autoimmune blistering diseases (AIBDs) are severe dermatologic disorders known for their debilitating physical impact and disfiguring nature. Recent research has reported mental health comorbidities in AIBDs, including depression and anxiety. Missing from the literature is an examination of the impact of AIBDs on body image and related psychological factors. This study examined the interrelationships among disease severity, body image dissatisfaction, and mental health comorbidities in a large sample of individuals with AIBDs.

Materials & Methods: We conducted a large survey study of 451 adults living with AIBDs. Participants were recruited through the International Pemphigus and Pemphigoid Foundation email distribution lists and social media. Validated self-report questionnaires assessed disease course and severity, body image disturbance, depressive symptomatology, quality of life, and psychiatric history. Participants reported their gender, age, race, ethnicity, country of residence, educational attainment, and employment/disability status. No personally identifying information was collected.

Results: By diagnosis, the sample composition was: 49.0% pemphigus vulgaris, 18.7% bullous pemphigoid, 19.6% mucous membrane pemphigus, 4.5% pemphigus foliaceous, 2.9% ocular cicatrical pemphigoid, 0.7% pemphigoid gestationis, 3.7% other/unclassified or still unknown, and 0.9% with multiple conditions.

Participants reported increased incidence of psychiatric disorders following AIBD diagnosis, especially depression, post-traumatic stress disorder, and eating disorders. Participants reported high levels of depressive symptomatology and impairments in quality of life. Patients with AIBDs reported extremely high levels of body image disturbance compared to patients with other disfiguring diseases or injury. Correlation analyses revealed significant relationships between body image variables and quality of life, even after controlling for depression.

Conclusion: Treatment guidelines for AIBDs focus primarily on the management of disease flares and the consequences of immunosuppression, without consideration of the psychosocial consequences of the diseases and treatments. The current study highlights the need for mental health support for patients with AIBDs.

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Abstract N°: 6124

Systemic lupus erythematosus exacerbation presenting as Stevens-Johnson syndrome: a case report

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Introduction & Objectives:

Stevens–Johnson syndrome (SJS) is a severe life-threatening, necrotic mucocutaneous reaction. In SJS, drugs are the overwhelming majority cause, while other triggers like autoimmune disease, infections or vaccinations are uncommon. One of the autoimmune disease, which can present primaly as SJS is systemic lupus erythematosus (SLE), which is a systemic disease that affects many organs. The relationship between SLE and SJS is unknown.

Here, we present a case report of a patient with systemic lupus erythematosus who presented with clinical manifestation of SJS without drug causality.

Materials & Methods:

A 36-year-old male patient with febrile illness and erythematous lesions was admitted to the clinic due to an acute exacerbation of his skin changes. The patient developed erythematous and maculopapular lesions with erosions and hemorrhagic crusts on his scalp, trunk, upper extremities as well as mucous membranes erosions following sun exposure. The patient has a 25-year history of systemic lupus erythematosus and has been taking oral hydroxychloroquine and prednisone for several years with good therapeutic effect. The patient had no history of new medication use or previous infections.

Results:

Our dermatological examination revealed positive Nikolsky sign. Additionally, laboratory tests revealed renal injury and hematological changes.

In order to confirm the diagnosis, we performed a biopsy from the trunk. The histopathology analysis revealed skin fragment covered with atrophic epidermis, without signs of necrosis. Hyperkeratosis and parakeratosis were visible on the surface. Features of severe inflammatory changes of the interface dermatitis type (vacuolization and destruction of the basal layer by lymphocytes) were present. In addition, numerous dyskeratocytes were visible. The microscopic picture may correspond to changes in the course of systemic lupus erythematosus.

Even though the histiologial findings corresponded to the SLE lesions, based on the typical clinical presentation and histopathological re-evaluation with the pathomorphologist we diagnosed SJS in the course of SLE. The patient received intravenous deksamethasone, antibiotics and acyclovir. The lesions resolved within 3 weeks after treatment and he was discharged in good condition.

Conclusion:

The relationship between SLE and SJS is unknown. There are only few causes in the literature of SJS or recurrent SJS in patients suffering from SLE without previous notable cause. The diagnosis of SJS can be made based on clinical picture and typical histological changes which involve basal keratinocyte vacuolation, epidermal necrosis

with mild dermal lymphocytic infiltration. Most patients with SJS and SLE demonstrate clinical and histopathological characteristics allowing for clear differentiation, however in some cases acute cutaneous manifestations of those diseases can be phenotypically similar, what can disrupt the implementation of proper treatment.

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Abstract N°: 6125

Herpesviridae Infections and Pemphigus Risk: Insights from a Population-Level, Propensity-Matched, Retrospective Cohort Study

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Introduction & Objectives:

The Pemphigus group of intra-epidermal autoimmune blistering diseases are characterized by complex, multifactorial etiologies, including potentially pathogenic contributions from environmental triggers such as viral infections with Herpes Simplex Virus (HSV) and/or Epstein Barr Virus (EBV). HSV induced autoimmunity has been documented in both autoimmune encephalitis and stromal keratitis and, recently, longitudinal data has implicated EBV as the primary environmental factor in the pathogenesis of multiple sclerosis. However, the current literature on the role of viral infections in the pathogenesis of pemphigus is marked by conflicting evidence and study limitations.

Materials & Methods:

To address these gaps, namely reliance on case reports and small sample sizes, we conducted a large-scale, propensity-matched, global retrospective cohort study to elucidate the relationship between objectively confirmed HSV or EBV infection and the subsequent development of pemphigus. Utilizing the Global Collaborative Network of TriNetX, a federated health research network, we analyzed electronic health records (EHRs) from 113 healthcare organizations. We compared the frequency of pemphigus development among a cohort of individuals age >18 following objectively confirmed HSV1/2 infection, as evidenced by positive anti-HSV 1/2 IgG or IgM (n=88,902), against a matched cohort (n=88,902) without either laboratory (IgG/IgM) or clinical (ICD-10) evidence of HSV infection. We conducted a similar analysis among a cohort age >18 following confirmed EBV infection as evidenced by positive anti-EBV IgG or IgM (nuclear, capsid, or early antigens) (n=197,944), against a matched cohort (n=197,944) without either laboratory or clinical evidence of EBV infection. We employed 1:1 propensity score matching based on age, gender, and healthcare interactions relating to sexually transmitted infection care (ICD-10 A50-A64) in the case of HSV, and healthcare interactions for viral infections (ICD-10 B25-B34) in the case of EBV, to ensure robust comparisons.

Results:

Our findings reveal that previous HSV infection is associated with significantly higher odds of subsequently developing pemphigus (OR, 2.274; 95% CI, 1.119-4.261) compared to a matched cohort without HSV. There was no significant difference in the development of pemphigus following infection with EBV (OR, 1.3; 95% CI, 0.726-2.323).

Conclusion:

Limitations include our retrospective study design, which precludes causal attribution of the observed differences in pemphigus development to HSV or EBV exposure; additionally, the use of EHR data introduces the possibility of misdiagnosis and/or coding errors. In conclusion, our study, distinguished by its scale, adds crucial evidence supporting the role of environmental triggers, specifically HSV, in the etiopathogenesis of pemphigus. These findings have implications for the management of pemphigus and other autoimmune diseases influenced by environmental factors.

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Abstract N°: 6154

A case report of MDA5-type seronegative dermatomyositis

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Introduction & Objectives: Dermatomyositis (DM) is an autoimmune idiopathic inflammatory myopathy with characteristic skin manifestations. Myositis-specific autoantibodies (MSA) help distinguish between DM subtypes and their prognoses. We describe one of the few cases of seronegative, amyopathic MDA5-DM in a 70-year-old female. This case underscores the importance of clinical judgment in diagnosing seronegative, amyopathic DM, and provides hypotheses for this phenomenon.

Materials & Methods: With the patient's consent, we reviewed the records from 2021-2024. Our institution uses the MitogenDx Myositis Panel, which includes Jo-1, Mi2-α, Mi2-β, MDA5, NXP2, TIF1γ, PL7, PL12, PM/Scl75, PM/Scl100, Ku, SRP, EJ, OJ, Ro52/TRIM21, SAE1, HMGCR, and NT5C1A/Mup44.

Results: A 70-year-old female presented with a six-month history of eruptive, pruritic, erythematous, and scaly ill-defined plaques that initially appeared on the face, eyelids, arms, and trunk. Accompanying symptoms included facial erythema, generalized myalgias, subjective leg muscle weakness, and persistent dyspnea for over a year. Her condition advanced to involve erosions and ulcerations of the digits, worsening facial erythema and edema of the eyelids bilaterally, and persistence of photodistributed skin findings extending across the lateral aspects of the upper arms, chest, and upper back. Her hands exhibited bilateral Gottron's sign, scaling, xerosis, fissuring, leukonychia, dystrophic cuticles, and periungual erythema, with dilated capillaries and capillary dropout noted on nailfold examination.

Despite the clinical picture of DM, all initial autoimmune serology, including ANA and myositis panels were negative. Creatine kinase, CRP and other biochemical markers remained within normal range. Within one year, her repeat autoimmune serology returned positive for speckled ANA and weak positive dsDNA but remained negative for ENA, myositis, and scleroderma panels. Skin biopsy showed interface dermatitis. She had non-contributory CT chest, MRI pelvis, and electromyography; therefore, muscle biopsy was not pursued. As clinical indicators strongly suggested DM, we eventually diagnosed her with an early, amyopathic DM, MDA5 subtype, as supported by other case reports in the literature. Initial treatments with methotrexate and azathioprine were not tolerated and the patient worsened with hydroxychloroquine; however, the patient achieved remission with tapering prednisone and mycophenolate mofetil.

Conclusion: Uncommonly, the diagnosis of DM can be complicated by incongruencies in clinical presentation and antibody testing, emphasizing the need for astute clinical judgment. Possible reasons for seronegativity include interference from corticosteroid treatment as seen with historical seronegative lupus, and limitations of current antibody testing assays. This case also suggests that non-MSA immune pathways may contribute to the pathogenesis and progression of DM, which has previously been suggested.

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Abstract N°: 6159

Parry Romberg syndrome: a series of five cases

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Introduction & Objectives:

Parry-Romberg syndrome (PRS) is progressive hemifacial atrophy. We present a study of five cases of PRS over three years, analyzing clinical features and therapeutic approaches.

Materials & Methods:

A retrospective study over a 3-year period, from 2021 to 2024, was performed on patients admitted to our department diagnosed with PRS on the basis of clinical features of progressive hemifacial atrophy.

Results:

We encountered five patients with a diagnosis of PRS. The average age was 18 years old, with a clear female prevalence; only one patient was male. The median age of onset was 11, 4 years old. The right side was involved in 80%. Four patients had mild PRS (Type 2), and one patient had severe PRS (Type 3).One patient had PRS associated with ipsilateral segmental vitiligo. A significant psychological impact was noted in all patients. Cutaneous induration was observed in 40% (2 cases) on the chin and neck, respectively, with histology of morphea. One patient had amputation of the right nasal wing. A facial CT was performed for one patient and a facial MRI for another, confirming fat atrophy with no bone involvement. There were no neurologic symptoms except for one patient who had a migraine. A cerebral MRI was performed on one patient and showed punctiform microbleeds in the right cerebral hemisphere with peri-lesional and right parafalcoral edema as part of his syndrome. A cerebral CT scan was performed for another patient and was without anomalies. The ophthalmological examination was without anomalies, except for one patient who had enophthalmos. Panoramic dental X-rays were performed on two patients with no abnormalities. All patients were treated with oral corticosteroids at a dose of 0.5 mg/kg/day combined with methotrexate at a dose of 12.5 mg/week, with an average treatment duration of 14 months.** Stabilization of the disease by medical treatment was achieved in all patients. Three patients underwent lipofilling under general anesthesia in plastic surgery, with subjective satisfaction of 70%.

Discussion:

PRS is a rare disease of unknown etiology; it had an estimated prevalence of 1 out of 700.000 people. PRS occurs in late adolescence and early adulthood. It affects three times more women than men. In our cases, there was a feminine prevalence. Fifty percent of all patients present are around the age of 10 years. The mean age of onset in our patients was 11, 4 years old.

Guerrerosantos et al. divided patients with PRS into four groups according to the severity of the defect to plan treatment. Type 1 patients had a mild form of the disease, which can be easily noticed. There is no bony involvement. In Type 2, the defect is more obvious but does not involve hard tissue. Type 3 patients show both soft tissue and hard tissue involvement with severe deformity. Type 4 patients have more severe deformity with the skin almost attached to the bone.

Treatment options include anti-inflammatory drug therapy, lipofilling, and reconstructive orthognathic surgery. PRS is a self-limiting disease, and the need for medical treatment arises from the coexistence of other autoimmune disorders like scleroderma. Immunosuppressants like methotrexate, corticosteroids, cyclophosphamide have been used in progressive cases of PRS. In our cases, patients were treated with corticosteroids in combination with methotrexate.

Conclusion:

PRS is a rare disorder that leads to severe disfigurement with possible functional impairment after years of progressive hemifacial atrophy.

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Abstract N°: 6321

Exploring the Complexities of Dermatitis Herpetiformis Diagnosis: Integrating Psychosocial Stress and Celiac Disease

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Introduction & Objectives:

Diagnosing dermatitis herpetiformis (DH) poses challenges due to its varied clinical manifestations and potential overlap with other dermatological conditions. Psychological stress has been implicated in exacerbating autoimmune diseases, including celiac disease (CD), but its role in DH remains underexplored. We present a case study highlighting the interplay between DH, CD, and psychosocial stress, emphasizing the importance of comprehensive evaluation in challenging diagnostic scenarios.

Materials & Methods:

We conducted a retrospective analysis of the medical records of a 69-year-old female patient who presented with a pruritic erythematous-papular-vesicular rash correlating with psychotraumatic stressors following her husband's death in 2021. Despite recurrent flare-ups and conventional therapies, including Dexamethasone and topical corticosteroids, lesions persisted. The patient's initial presentation to our department was in February 2023. Skin biopsies initially suggested bullous pemphigoid, later confirmed as DH through direct immunofluorescence examination in April 2023. Gastroenterological consultations confirmed celiac disease in November 2023 through positive serological markers, corroborated by duodenal biopsy findings.

Results:

Initiation of treatment with Dapsone and a gluten-free diet in April 2023 led to initial improvement until August 2023 when therapy discontinuation due to adverse effects prompted alternative management. Notably, the patient reported remission of cutaneous lesions with a gluten-free diet but experienced relapses during periods of heightened psychosocial stress. Psychiatric evaluations revealed mixed anxiety-depressive disorder with panic attacks post her husband's death, requiring pharmacological and psychological interventions.

Conclusion:

This case underscores the intricate interplay between DH, CD, and psychosocial stressors, indicating a mutual relationship. Stressful life events may exacerbate pre-existing conditions or even contribute to their onset. The diagnostic complexity of DH is highlighted, emphasizing the importance of considering celiac disease in patients presenting with DH-like lesions. Furthermore, it highlights the bidirectional relationship between psychosocial stress and autoimmune conditions, suggesting a potential role in disease exacerbation. Comprehensive multidisciplinary approaches integrating dermatology, gastroenterology, psychiatry, and immunology are essential for optimal management and improved patient outcomes in DH.

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Abstract N°: 6418

Amyopathic dermatomysositis presenting as severe scalp alopecia and prominent eyelid edema: a case report

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Introduction: Dermatomyositis is an autoimmune connective tissue disease of uncertain etiology. It most often affects both the muscles and the skin, but may occur as amyopathic dermatomyositis (AD). Multiple studies suggest an association between malignancies and AD, the reported frequency varies from 15–25%. Moreover, some also report fulminant lung disease in patients with amyopathic dermatomyositis. Therefore, both malignancy screening and evaluation for pulmonary involvement are indicated.

Results:

A 62-year-old female patient with a medical history of hypertension and chronic gastritis presented to our Dermatovenerology department with a severe symmetrical livid edema of upper and lower eyelid that obscured her vision, diffuse scaly erythema on the scalp and face, diffuse severe alopecia, livid erythematous plaques on the neck and upper chest, symmetrical erythematous livid plaques on elbows and clinical picture of mechanic hands with the presence of Gottron papules. She denied muscle weakness.

The patient first reported the onset of the symptoms three years prior to the visit with a scaly rash on the scalp and hair loss, which deteriorated within one year, presenting with face and scalp erythema, diffuse frontal alopecia and mild eyelid edema. The results of histopathology, direct immunofluorescence and clinical picture coincided with the diagnosis of discoid lupus erythematosus (LE), for which she was treated with local corticosteroid and immunomodulatory therapy, later on treatment with hydroxychloroquine was introduced. Due to the continuous deterioration she was referred to our tertiary centre where the suspicion of AD was made. Results of laboratory tests revealed positive anti-MDA-5 antibodies. Based on the neurological assessment, electromyography and laboratory testing (creatine kinase, myoglobulin, aldolase, lactate dehydrogenase, aspartate aminotransferase, alanine aminotransferase) muscle involvement was excluded. Paraneoplastic etiology and lung involvement was excluded with an extensive laboratory and radiologic investigations. Due to insufficient regression with hydroxychloroquine and intensive local corticosteroids concurrent therapy with methylprednisolone and methotrexate was initiated.

Conclusion:

Diagnosis of AD may be at times tricky. The clinician must remain vigilant in the key differential diagnosis of cutaneous LE as they can both present with similar clinical features, clinical course, histopathology and without myopathy.

Severe alopecia of the scalp with prominent eyelid edema can be one of the key presentations in AD, that aid in the vast differential diagnosis and may encourage the clinician to search for other key features of dermatomyositis.

When the diagnosis is established, extensive tests must be performed to exclude possible paraneoplastic etiology and lung involvement. Achieving regression in AD is difficult and both intensive topical and concurrent three track systemic therapy may be indicated in refractory cases in order to achieve remission.

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Abstract N°: 6434

Efficacy and safety of oral tofacitinib in non-segmental vitiligo

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Introduction & Objectives:

Vitiligo affects both adults and children and can be a source of significant psychosocial distress. Immune mediated destruction of melanocytes in vitiligo has been shown to be driven by IFN-γ and CD8+ T cells. Re-pigmentation of lesions with current traditional treatment options tends to be variable and treatment response is often unsatisfactory. There are hardly any systemic agents approved for the treatment of this distressing condition. Tofacitinib, a Janus kinase (JAK) 1/3 inhibitor is a promising treatment for vitiligo due to its inhibition of IFN-γ signalling.

Our objective was to to study the efficacy and safety profile of oral tofacitinib in non-segmental vitiligo.

Materials & Methods:

We performed a record-based review of patients with non-segmental vitiligo who were treated with oral tofacitinib. Following institutional ethical approval, patient records were retrieved from the hospital electronic medical records system and analysed. Patients with non-segmental or unstable acrofacial vitiligo aged 12 years or older who had received oral tofacitinib for at least 24 weeks were included in the study. All patients underwent baseline routine blood investigations including hemogram, liver and renal function tests, lipid profile, HIV, Hepatitis B, Hepatitis C markers and pre-treatment screening for tuberculosis via chest radiographs and the interferon-gamma release assay (IGRA). Patients were followed up on a monthly basis. Vitiligo Area Severity Index (VASI) was assessed at baseline as well as at each follow-up visit. Complete hemogram, liver and renal functions tests and fasting lipid profile were performed during each follow-up visit. Any adverse events during the treatment period were noted.

Results:

16 patients (Male: Female 6:10) with a mean age of 37.4 years (range 14–59 years) were included. All patients noted some degree of improvement at 6 weeks. Mean VASI scores improved from 5.038 at baseline to 1.269 at 24 weeks, with 13 patients (81.25%) achieving at least 50% improvement. . Phototherapy was concurrently administered to all patients except one. Transient leukopenia was observed in 3 patients and one patient developed palmar warts. No otheradverse effectes were noted.

Conclusion:

Oral tofacitinib shows significant potential in the management of non-segmental vitiligo, especially in patients unresponsive to conventional therapies.

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Abstract N°: 6451

Genital bullous pemphigoid : A rare case report

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Introduction & Objectives:

Bullous pemphigoid (BP) can manifest in various regions of the body, including the genital region impacting a person's quality of life. Genital BP (GBP) is a rare subgroup of localized BP, which typically affects women and children. Scrotal BP is a small subset of this, and the only cases we are aware of are in children and are extremely rare in adult men. We hereby report a rare case of localized bullous lesions over genitalia in an elderly male patient.

Case report:

A 75-year-old male patient comes with a primary complaint of recurring fluid-filled lesions over the genitalia, with three to four such episodes in the previous seven months. The results of tzanck smear were inconclusive. His medical history included type 2 diabetes mellitus, and he denied any changes in his regular medication over the last year. No inciting history of drug use, trauma, secondary manipulation, or use of any local agents was present. Routine hematological and biochemical tests revealed no abnormality except raised blood sugars. Based on the clinical diagnosis of herpes genitalis, he was treated with oral valacyclovir with resolution of lesions after few days. However, the patient noticed recurrence of similar lesions over genitalia associated with itching and burning sensation and mild pain after 2 months.

Cutaneous examination revealed a large erythematous and edematous plaque over the penile skin, with multiple crusted lesions and smaller, serous-filled tense blisters were also observed over the erythematous plaque. A provisional diagnosis of autoimmune bullous disorder was suspected, and direct immunofluorescence was performed which showed features suggestive of pemphigoid group of disease. Indirect immunofluorescence study was performed on salt-split normal human skin with the serum from patient showing linear staining of basement membrane zone with IgG only on the epidermal side of the split ("roof" pattern). These features were diagnostic of bullous pemphigoid. The patient was then started on oral doxycycline 100mg twice daily with topical antibiotics and oral antihistamines. He noticed complete resolution of skin lesions with no recurrence

Discussion:

Localized BP has a clinical variation known as genital bullous pemphigoid (GBP). It occurs frequently in pediatric patients and primarily effects females. Typically, vesiculobullous skin lesions in the vulvovaginal region are the first sign of female GBP. Usually benign, it responds quickly to topical steroids. There have only been few case reports of localized GBP in males, making it incredibly uncommon. Differential diagnoses for genital bullous pemphigoid include other erosive disorders like bullous lichen planus, dermatitis herpetiformis, epidermolysis bullosa acquisita, linear IgA disease, mucosal pemphigoid, pemphigus vulgaris; drug reactions, various anogenital infections and contact dermatitis. Due to the unusual site and similarity in presentation, the diagnosis can be challenging.

Conclusion:

We are reporting the first case of adult GBP with the diagnosis confirmed by both direct and indirect

immunofluorescence.

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Abstract N°: 6464

Improving Alopecia Areata Care in Poland: Closing the Clinical Gap.

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Introduction & Objectives:

Alopecia areata (AA) is an autoimmune disease characterized by the immune system's targeting of hair follicles, leading to non-scarring hair loss, which affects up to 2% of the global population. The onset of symptoms may occur at any age. However, clinical observations suggest that the initial episode often manifests before the age of 5. Alopecia areata often causes significant psychological burdens among patients and significantly affects quality of life. It is associated with an increased risk of depression, anxiety, and emotional and social functioning disorders. The care of patients with alopecia areata in Poland requires multidisciplinary cooperation and the spreading of public awareness, which will promote modern and effective therapies. This is because currently Polish patients do not have access to reimbursed therapies. The aim of this study is to evaluate the current clinical strategies in Alopecia Areata diagnosis and management by dermatologists in Poland.

Materials & Methods:

A proprietary survey was created and conducted among the specialists and physicians specializing in dermatology and venereology in Poland. A group of 100 physicians, practicing in both hospital and private clinics, was enrolled in this study.

Results:

Among responders, 39% admit diagnosing alopecia via medical history and clinical examination only. The rest of the specialists use additional tools to analyze the patients' condition. Only 5% order a biopsy in the diagnostic process, and 13% perform a trichogram. 10% of the doctors participating in the survey also perform mycological tests on their patients to rule out fungal infections. 78% of respondents admit they use dermoscopy in the examination of a patient. 36% of respondents use scales to assess disease severity, with 78% of doctors using the SALT scale. Only 35% of doctors use tools that assess the psychosocial impact of the disease on patients' lives and functioning, with the DLQI being the most commonly used scale. As many as 30% of doctors participating in the survey do not use any imaging methods to monitor patients, including lesion imaging or dermoscopy. Analyzing the therapeutic aspects, the survey results show that 60% of the physicians surveyed prefer topical corticosteroids as the first therapeutic option for treating alopecia areata. The second popular therapeutic option is intralesional steroid injections, used mainly in patients older than 12 years, and calcineurin inhibitors and irritants such as anthralin, used in patients younger than 12 years.

Conclusion:

The findings reveal concerning gaps in the clinical management of alopecia areata (AA) among dermatologists in Poland. The underutilization of scales assessing disease severity and its impact on patients' quality of life underscores a critical need for enhanced education and awareness. Optimal therapeutic decisions rely heavily on accurate assessments using tools such as the Severity of Alopecia Tool (SALT) or Alopecia Areata Scale (AAS).

Addressing these deficiencies through educational initiatives and promoting the integration of comprehensive assessment tools into clinical practice is imperative to ensure more effective and patient-centered management of AA in Poland.

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Abstract N°: 6483

Wong type dermatomyositis with clinical features reminiscent of papuloerythroderma of Ofuji

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Introduction & Objectives:

Dermatomyositis (DM) is a rare autoimmune disorder characterized by inflammatory changes in the skin and muscles, leading to characteristic cutaneous manifestations and muscle weakness. Among the various subtypes of DM, the Wong type DM (WTDM) represents a distinct clinical variant of the disease with unique clinical and histological features mimicking other dermatoses, such as discoid lupus erythematosus (LE) and pityriasis rubra pilaris (PRP). The specific clinical presentation of WTDM is a slightly pruritic widespread eruption consisting of erythematous, keratotic follicular papules without accompanying myopathic symptoms. We recently observed a case of WTDM with clinical features reminiscent of papuloerythroderma of Ofuji.

Materials & Methods:

A 52-year-old man presented with a 6-month history of a skin rash involving the trunk and extremities. Topical therapy with clobetasol propionate, mometasone furoate, and emollients was ineffective. Physical examination revealed a symmetrically distributed erythematous eruption on the neck, chest, back, and external surfaces of the upper and lower extremities consisting of miliary papules with keratotic surface and slight central atrophy upon dermoscopy. The papules were coalescing into large erythematous plaques with a cobblestone-like appearance, which tended to spare the folds on the chest and abdomen, thus resembling the “deck-chair sign” in Ofuji’s papuloerythroderma. A slightly pronounced heliotrope erythema was observed on the face, as well as a positive Gottron’s sign and papules on his dorsal forearms and hands. The patient did not report any comorbidities or family burden.

Results:

Routine laboratory investigations showed slightly elevated creatine kinase and eosinophils. Immunological testing revealed ANA at a titer of 1:320, and positive AMA M2-1, anti-centromere B-1, anti- dsDNA-1, anti-Jo-1-1, and anti- Ro 52-2 antibodies. Histological examination demonstrated findings compatible with connective tissue disease. Direct immunofluorescence showed deposition along the dermo-epidermal junction of granular C3 together with few ovoid bodies (IgG, IgA, IgM) in the papillary dermis. Phototesting was negative for UVA and positive for UVB. Neurological consultation and EMG study showed no evidence of myogenic damage. Systemic treatment with methylprednisolone 32 mg/day, hydroxychloroquine 400 mg/day and topical photoprotection improved the patient’s complaints.

Conclusion:

The recognition and diagnosis of WTDM is challenging due to its rarity and clinical presentation mimicking a wide variety of other dermatoses. WTDM is also termed amyopathic DM as cutaneous manifestations overshadow muscle involvement which may divert attention from the underlying autoimmune process and delay the diagnosis. In conclusion, WTDM appears as a diagnostic dilemma and should be ruled out by prolonged patients’ monitoring in cases overlapping clinically with PRP, as well as LE or papuloerythroderma of Ofuji.

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Abstract N°: 6488

Unveiling the Enigma: Systemic Sclerosis Sine Scleroderma, Lichen Planus and Dermatomyositis in the Overlap Syndrome Landscape

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Introduction & Objectives: Overlap syndrome is an entity that requires two or more connective tissue disorders. Systemic sclerosis sine scleroderma is a rare subtype of systemic sclerosis that presents with internal organ manifestations but no cutaneous findings. Previous reports have documented that dermatomyositis may be associated with other autoimmune connective tissue diseases such as scleroderma, lichen planus, Sjögren's syndrome. A combined pathology has an impact on the clinical features, diagnosis, and treatment.

Materials & Methods: A 56-year-old female presented to the dermatology clinic for cyanosis of the last four fingers of both hands induced by cold or stress. Dermatoscopically, giant capillaries, hemorrhagic spots, and hyperkeratosis of the cuticle on a pink-pale background can be seen at the level of the proximal nail fold of fingers 2-4 of the hands bilaterally. It also presents photodisposed violaceous erythema on the neckline and shoulders. A skin biopsy was performed which established the diagnosis of dermatomyositis. A lung CT scan showed bilateral lung fibrosis. The diffusing capacity of the lungs for carbon monoxide is <50%. The antinuclear antibody (ANA) and anti-centromere-B tests yielded positive results with a value of 7.5 and 130 RU/mL, respectively. Considering Raynaud's phenomenon, abnormal nail fold capillaries, lung damage, and a positive ANA test lead to establishing the diagnosis of systemic sclerosis sine scleroderma. The patient's medical history includes lichen planus diagnosed histopathologically in 2006, psoriasis vulgaris clinically diagnosed in 1994, ocular and oral sicca syndrome since 2016, unbalanced and complicated type 1 diabetes with peripheral sensory-motor polyneuropathy and diabetic retinopathy since 1993, Hashimoto's autoimmune thyroiditis since 1994, and myasthenia gravis IIB Osserman since 2007.

Results: A rheumatologist evaluated the patient, and capillaroscopically, an active sclerodermic pattern was detected.** Systemic treatment with azathioprine 50 mg/day was initiated, then after a month, the dose was increased to 100 mg/day. Also, the patient followed repeated courses with iloprost 20, 1 ampoule/day, 5 days, low-dose aspirin, and pentoxifylline 400 mg/day. The patient's skin manifestations slowly began to resolve.

Conclusion: Approximately a quarter of patients with early-stage rheumatologic disease meet the criteria for an overlap syndrome. There is conflicting evidence about whether overlap syndromes are more treatment-resistant than either disease entity alone. The identification of overlap syndromes is essential since it aids in customizing treatment for specific components of the illness and determining the prognosis.

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Abstract N°: 6515

An unusual presentation of lupus tumidus mimicking cutaneous lymphoma: A case report

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Introduction & Objectives:

Tumidus lupus erythematosus (TLE) is an uncommon subtype of cutaneous lupus erythematosus which is usually confined to skin without systemic involvement. Autoimmune workup is often negative. Only a few cases have been reported. We report an unusual presentation of lupus tumidus.

Materials & Methods:

A 78-year old female presented with multiple asymptomatic skin lesions over face, scalp, upper limbs, and trunk for four weeks. Skin lesions initially appeared on the face and then progressed to involve the torso and upper limbs. Systemic inquiry was unremarkable. There was no history of connective tissue diseases. The drug history was insignificant.

Examination revealed multiple well defined erythematous non tender papules, nodules, and plaques distributed over both sun-exposed and sun-protected areas.

Antinuclear Antibody (ANA) was positive (1:100) and the Extractable Nuclear Antigen-antibody panel was negative except for the weakly positive Anti-U1RNP antibody. The antiphospholipid antibody panel was negative. Other haematalogical, biochemical and imaging studies were negative. Incisional skin biopsy revealed mild epidermal hyperkeratosis, atrophy, follicular plugging, attenuated rete ridges, and basement membrane thickening. Vacuolar degeneration with apoptotic keratinocytes was seen in the dermo-epidermal junction. Dermis had increased mucin and moderate superficial perivascular chronic inflammation. A diagnosis of lupus tumidus was made.

The patient improved with topical and systemic corticosteroids, oral hydroxychloroquine, and photoprotection.

Results:

TLE may have polymorphic presentations that mimic other clinical conditions. Diagnosis is based on clinicopathological correlation. TLE is characterized by erythematous, succulent, urticariform, non-desquamative plaques mainly over sun-exposed areas, Majority has photosensitivity.

Patients often have a low titre of ANA and negative other autoantibody panels. Histology shows a superficial and deep dense lymphocytic infiltrate in the perivascular and periadenexial regions and diffuse mucin deposition. It has a better prognosis compared to other variants of cutaneous lupus.

Conclusion:

Awareness of this entity and clinicopathological correlation is important in diagnosing TLE.

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Abstract N°: 6548

Polymorphic eruption of pregnancy mimicking gestational pemphigoid : an unusual clinical case.

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Introduction & Objectives: Polymorphic Eruption of Pregnancy (PEP) is a common and benign dermatosis that typically affects primiparous women during the third trimester of pregnancy. It usually presents as pruritic erythematous papules, eczematous lesions, targetoid appearance, or vesicles. However, the presence of blisters in PEP has been rarely described before. We present here a highly unusual case of PEP associated with the late development of blisters, posing a differential diagnosis challenge with bullous pemphigoid. **Materials & Methods:** A 21-year-old primiparous patient at 27 weeks of gestation, with no specific medical history, who had been experiencing urticarial papules and small vesicles on the limbs for the past month. An initial skin biopsy with the direct immunofluorescence (DIF) that was negative were suggestive of chronic eczema. **The patient was reevaluated two weeks after daily application of topical corticosteroids, with no significant clinical improvement. Dermatological examination revealed extensive confluent eczematous plaques sparing the face, palms, soles and mucous membranes. These were accompanied by tense clear-content blisters on the distal extremities, associated with scratch lesions, diffuse excoriated papules, and targetoid lesions on the back. All this occurred in the absence of fever. A second skin biopsy was performed, revealing a blister at the dermo-epidermal junction with an intact epidermal roof without intercellular edema. There was a perivascular infiltrate consisting of neutrophils and lymphocytes with eosinophilic epidermal exocytosis. The second DIF was negative.** The clinical presentation, histological findings, and immunological data supported a diagnosis of Polymorphic Eruption of Pregnancy (PEP). The patient was started on oral corticosteroid therapy at a dose of 40mg/day, leading to a significant clinical improvement. **Results:** Our case highlights that the development of blisters on the limbs can be a late polymorphic feature of Polymorphic Eruption of Pregnancy (PEP), the polymorphic appearance is mainly characterized by pruritic erythematous papules evolving into plaques, often accompanied by vesicles. The navel, face, palms, soles, and mucous membranes are typically spared. Histologically, there is usually perivascular and/or interstitial lymphocytic infiltration alongside eosinophils. To our knowledge, PEP with blister formation has been reported only twice before in recent literature. The presence of blisters in our patient raised suspicion of other potential diagnoses. However, the medical history, clinical criteria, histological findings, and specific tests did not support these alternatives. **Conclusion**: In conclusion, although the presence of blisters may raise suspicions of various pathologies, this case underscores the importance of considering the specific context of pregnancy and the characteristics of Polymorphic Eruption of Pregnancy (PEP) in the process of a differential diagnosis.

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Abstract N°: 6568

The experience of a Mediterranean country in the management of Pemphoid Gestationis

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Introduction & Objectives:

Gestational pemphigoid (GP) is a rare subepidermal bullous dermatosis, linked to the presence of anti-BP180 autoantibodies synthesized as a result of a breakdown in maternal-fetal immunological tolerance. It typically occurs in multiparous women during the second or third trimester of pregnancy, rarely in the postpartum period, and exceptionally post-abortion.

Materials & Methods:

This is a retrospective study, compiling cases of GP diagnosed in the dermatology and venereology department at CHU IBN SINA in Rabat over a period of 22 years; between January 2001 and December 2023. Various epidemiological, clinical, paraclinical, therapeutic, and evolutionary parameters were recorded.

Results:

We identified 33 patients, with an average of hospitalized cases per year ranging between 1 and 2. The average age at diagnosis of our patients was 33 years with extremes ranging from 23 to 45 years. 79% of patients were multiparous and 21% were primiparous. Among multiparous patients, the majority were in their second pregnancy (40% of cases) or third pregnancy (30% of cases), and 33% of them had previously experienced pruritic urticarial eruptions during previous pregnancies. The disease onset occurred in 45% of cases in the third trimester, in 33% of cases in the second trimester, with three cases starting in the first trimester and four cases (12%) in the postpartum period. In our series, there were no cases occurring post-abortion, but a history of early miscarriage preceding the current pregnancy was found in 15% of patients. All patients experienced intense pruritus preceding the cutaneous eruption by an average of one week.

The eruption consisted of urticarial papules with atypical targets, vesiculobullous lesions were found in 70% of cases, and pustules in 45%. The onset occurred peri-umbilically with centrifugal extension in 79% of cases; the face was affected in only two cases. Additionally, we also noted mucosal involvement in 21% of cases, consisting of cheilitis in 5 patients, endobuccal erosions in one patient, and genital mucosal erosion in only one patient.

Histology revealed subepidermal blisters with an eosinophilic infiltrate and some necrotic keratinocytes in 70% of cases. Direct immunofluorescence (DIF) performed in 40% of cases showed linear deposition of C3 along the dermo-epidermal junction, and indirect immunofluorescence (IIF) performed in 36% of cases demonstrated the presence of circulating autoantibodies to the dermal–epidermal junction (DEJ).

Systemic steroids (0.5 to 1 mg/kg/day) were used in 67% of cases, complicated by puerperal psychosis 15 days after initiation in one patient. Ten patients (30% of cases) showed improvement with very potent topical corticosteroids combined with antihistamines. Exacerbation in the postpartum period was noted in 4 patients, with ten cases of recurrence in subsequent pregnancies. Regarding fetal prognosis, 4 cases of prematurity, 5 cases of intrauterine fetal death, and 3 cases of hypotrophy were recorded.

Conclusion:

Our series aligns with the literature regarding the percentage of multiparity, predominance of occurrence in the 2nd and 3rd trimesters, clinical and histological data, as well as the positivity rate of DIF. Two risk factors for the occurrence of fetal and neonatal complications were identified in our study, namely the presence of severe inflammatory syndrome and severe eosinophilia in the mother, highlighting the importance of immediate management.

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Abstract N°: 6612

A case of keloidal morphea in a 40-year old moroccan female

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Introduction:

Morphea is a chronic autoimmune condition, characterized by sclerotic plaques in the skin and subjacent tissues. Keloidal morphea (KM) is one of the rarest variants. We describe a 40-year-old woman with morpheic lesions for 10 years along with the development of kelidal nodules within these lesions.

Case Description:

A 40-year-old female presented for management of hyperpigmented sclerotic plaques involving her neck, trunk, arms and axilla. These lesions slowly multiplied over the course of 10 years.

Shortly after, firm nodules occured within the hyperpigmented plaques. She did not show any evidence of systemic involvement such as Raynaud's phenomenon, diarrhea or shortness of breath

There was no history of keloid formation, and she denied any trauma at those affected sites.

Physical examination revealed hyperpigmented and slightly sclerotic plaques, scattered on the trunk, abdomen, back, axilla, upper arms and legs. Additionally, the patient presented multiple nodules, that were non tender, firm on the back, abdomen, arms and legs. Pseudopodes like extensions on both upper arms were noted. These lesions measured 5 to 40 mm and were not fixed to deep layers.

Periungual dermoscopy showed no abnormalities. Furthermore, there was no calcinosis, sclerodactyly, or telangiectasia. However, we noticed a thin scar at the lower abdomen, indicative of a caesarean section performed 2 years before the first manifestation of cutaneous lesions. This contrasting observation disproved the diagnosis of spontaneous keloidal formations, suggesting the possibility of KM.

Routine laboratory tests were normal and antinuclear antibodies, anticentromere, anti-dsDNA, Anti-Scl70 antibodies were negative.

Two biopsy specimens were obtained. In the hyperpigmented lesion, examination showed that the dermis is fibrous, made up of thick, horizontalized collagen bundles. Biopsy of keloidal lesions revealed an acanthotic and orthokeratotic epidermis and thickened bundles of collagen, which were found in the dermis. Both clinical presentation and histological examination were in line with the diagnosis of KM

Discussion:

KM is a very rare variant, which can be clinically mistaken for keloid formation. To this date, there a fewer than 60 reported cases of this form of morphea.

Clinically, it is characterized firm, raised lesions, which can vary in size. The lesions are painless, although often accompanied by pruritus.

Histological findings are variable, showing either aspects of morphea, or aspects of keloid scarring or a combination of the two. Our case fulfills the criteria for both clinical and histopathological diagnosis of KM. The diagnosis of true spontaneous keloids is highly unlikely given the presence of the thin scar from the cesarean section. Differential diagnoses include keloid/hypertrophic scar, sarcoidosis and sclerotic dermatofibroma.

Several treatment modalities have been described in the literature. Yet, the results have been variable.

There is no consensus for the treatment of KM. Our patient was treated with systemic steroids in combination with methotrexate. She reported a partial response with 12 weeks.

Conclusion:

This case highlights the importance of distinguishing KM from a hypertrophic scar or keloid to reveal an underlying systemic disease.

Studies are recommended to elucidate aspects of this condition as well as available treatment options.

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Abstract N°: 6655

Simultaneous diagnosis of Kikuchi Fujimoto Disease and Familial Chilblain Lupus. Two related pathologies?

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Simultaneous diagnosis of Kikuchi Fujimoto Disease and Familial Chilblain Lupus. Two related pathologies?

Introduction & Objectives:

Familial Chilblain Lupus (FCL) belongs to the type 1 interferonopathies in which Aicardi-Goutières syndrome (AGS) is the most representative entity. Kikuchi Fujimoto disease (KFD) is a rare condition with neither infectious nor neoplastic lymphadenopathies. Our objective was to report the first case of simultaneous FCL and KFD in the same patient and propose a possible association.

Materials & Methods:

Case report

Results:

We present the case of a 36yo woman referred to dermatology clinic with child-onset skin lesions of acral distribution persisting despite chronic use of steroids and vasodilators. Furthermore, over the past 3 years she has presented enlargement of lymph nodes with PET-Scan guided lymph node biopsy revealing necrotizing granulomatous lymphadenitis non-responsive to empirical treatment against tuberculosis. Ancillary tests showed neutropenia and mildly elevated ANAs. This patient was evaluated in an interdisciplinary medical board advising pertinency of genetic testing for autoimmune lymphoproliferative syndrome mutation without finding FAS abnormalities, but a TREX1 mutation instead. Histocytes where found in bone marrow biopsy. Diagnosis of AGS was ruled-out based on absence of neurological signs or basal-ganglia calcifications. Therefore, diagnosis of Familial Chilblain Lupus associated with Kikuchi-Fujimoto disease was made.

We report the first known case of FCL and KFD coexisting in the same patient. FLC belongs to a group of pathologies known as type 1 interferonopathies (of which AGS is the prototype disease) and is a rare condition characterized by early-onset chilblain lupus lasting several years. A missense mutation p.Asp18Asn in TREX1 is the most prevalent genetic variation, which was found in our patient. Concomitant, diagnosis of KFD was made, a clinical entity frequently misdiagnosed as lymphoproliferative disorders or chronic infections with lymph node involvement. Multiple lymphadenopathies, neutropenia and abnormal findings on PET-scan are core findings in this pathology. Biopsies revealed necrosis and histiocytes, and even though granulomas are not as typical, there have been reports with this presentation. An increase of transcriptional activity on Interferon type I (IFN-I) genes has been described, suggesting a viral etiology. Presence of positive ANAs titers, leucopenia and lymph node involvement beyond cervical groups have been associated to the chronic clinical evolution of the patient symptoms. We propose that the coexistence of these two pathologies is not explained by mere chance, but rather by the increased activity of IFN-I transcripts related to the TREX1 mutation, thus explaining the development of KFD in an atypical chronic fashion. This would not be the first time that a type 1 interferonopathy presents with signs usually associated with a viral disease, as is the case of AGS mimicking TORCH syndrome.

Conclusion:

The discussed patient suffers from two infrequent conditions that may be linked in its pathophysiology through similar overexpression of the IFN-I pathways. To our knowledge this association has not been made before and it’s an opportunity for future research.

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Abstract N°: 6708

Therapeutic targets for systemic Sclerosis: Mendelian randomization and colocalization study

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Introduction & Objectives: Systemic sclerosis (SSc) has one of the highest mortality rates among rheumatisms. It is difficult to treat due to its heterogeneity. Thus, the search for new druggable targets is crucial for the treatment of SSc.

Materials & Methods: In the present study, we used Mendelian randomization (MR) and co-localization to explore the potential therapeutic target of SSc. We used 2923 plasm proteins from 54,219 blood donors in the UKB dataset. Gwas data of SSc was sourced from the GWAS by López-Isac E et al. in 2019, including 9,095 cases and 17,584 controls. We conducted an MR analysis by integrating plasma protein data of UKB and GWAS data of SSc. Additionally, we used reverse causation, bayesian colocalization analysis, and phenotype scanning to further validate our results. We then explored pathways and mechanisms using GO and KEGG enrichment analysis. Protein-protein interaction (PPI) network was used to analyze the functional network interaction between plasma proteins and identified drug target genes. External validation was used to further validate results.

Results: MR analyses identified five blood proteins that may causally be associated with SSc. MERTK, MPI, and PTPRM can increase the risk of SSc. AIF1 and TIMP4 can decrease the risk of SSc. Bayesian colocalization analyses showed that MERTK, MPI, PTPRM and TIMP4 shared common causal variant with SSc. TIMP4 was validated using external validation.

Conclusion: Our study suggested that four plasma proteins (MERTK, MPI, PTPRM and TIMP4) were potential therapeutic target of SSc. These proteins' specific roles and mechanisms need further clinical validation.

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Abstract N°: 6727

Efficacy of Complex Therapy for Localized Scleroderma in Children: A Comparative Study

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Introduction & Objectives:

Morphea, also known as localized scleroderma, is an autoimmune connective tissue disorder that triggers a series of immune responses resulting in autoaggression, metabolic abnormalities in the extracellular matrix, and endothelial cell damage. Its incidence among children is approximately 1–3 per 100,000 annually, with a prevalence of 2 per 1,000 children. This study's primary goal was to evaluate the efficacy of different treatment approaches for morphea in children, using the Localized Scleroderma Assessment Tool (LoScAT) index to measure treatment outcomes.

Materials & Methods:

To assess and optimize treatment for pediatric morphea, this study divided patients into two groups:

- Group 1 (control, n=30): Received standard therapy.

- Group 2 (intervention, n=38): Received standard therapy with additional papain enzyme and 0.03% tacrolimus ointment.

Participants were randomized by age, gender, disease duration, and severity. Treatment efficacy was evaluated through the LoScAT index, which assesses activity, severity, and tissue damage associated with localized scleroderma.

Results:

In both groups, the linear form of morphea was the most common, with 53.3% and 57.9% incidence, respectively. The disease duration in most cases ranged from 1 to 5 years. Traditional therapy in Group 1 resulted in a reduction in erythema and edema, yet significant inflammation indicators persisted. The lesions exhibited a whitish hue, with a diminishing pinkish-lilac rim. A reduction in erythema by 1.3 times ($P<0.05$) from baseline was observed. In Group 2, erythema decreased by 2.1 times, indicating enhanced effectiveness with the additional treatment regimen.

In the sclerosis stage, Group 2 experienced a greater reduction in lesion density and a noticeable softening of the sclerotic areas, with some resolution in the peripheral zone. Group 1 displayed a modest reduction in compaction by 1.1 times ($P<0.01$), whereas Group 2 showed a 1.4 times decrease in lesion density. The appearance of new lesions dropped significantly in Group 2 by 3.62 times (from 0.87 ± 0.03 to 0.24 ± 0.04), while Group 1 only saw a 1.4 times decrease. Traditional therapy proved ineffective in cases of atrophy, with limited recovery of compacted regions despite some softening.

Conclusion:

Complex therapy incorporating papain enzyme and tacrolimus ointment significantly improved the clinical outcomes in children with morphea compared to traditional therapy alone. The enhanced treatment regimen led to a notable decrease in lesion size, density, and inflammation, with stabilized scleroderma progression, and in

some cases, regression. This study suggests a more robust therapeutic approach could yield more favorable outcomes for pediatric morphea patients. Further research is recommended to validate these findings and optimize treatment protocols.

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Abstract N°: 6754

Beyond the Skin's Surface: Deciphering the Enigmatic Overlap of Lupus Tumidus and Jessner's Lymphocytic Infiltration

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Introduction & Objectives:

Lupus tumidus (LT) and Jessner's lymphocytic infiltration of the skin (JLIS) are uncommon dermatological conditions characterized by similar clinical and histopathological features, posing a diagnostic dilemma for clinicians. While LT is considered a subtype of cutaneous lupus erythematosus (CLE), JLIS is classified as a benign lymphoproliferative disorder. Despite their distinct etiologies, the overlapping presentation often necessitates careful clinical evaluation and histopathological analysis for accurate diagnosis and appropriate management.

Materials & Methods:

We present the case of a 46-year-old male with a three-year history of papulo-erythematous eruptions and annular lesions on the thorax and face, which began shortly after initiating antihypertensive treatment. Despite experiencing temporary improvement with systemic corticosteroid therapy, the lesions recurred. However, due to the pandemic, the patient did not seek further dermatological care until June 2023, when suspicion of subacute cutaneous lupus erythematosus (SCLE) arose, and a skin biopsy was performed, yielding inconclusive results. Additional investigations, including an extended antinuclear antibody (ANA) profile, which was negative, and negative antiphospholipid antibody testing, were conducted. Referred to our department in January 2024, a repeat skin biopsy revealed features consistent with subacute cutaneous lupus erythematosus, despite negative direct immunofluorescence findings for granular deposits. In April 2024, due to a poor response to local corticosteroid treatment, suspicion of drug-induced eruption prompted another skin biopsy, revealing histopathological features suggestive of either lupus erythematosus tumidus or Jessner's lymphocytic infiltration of the skin.

Results:

Clinical examination revealed characteristic papulo-erythematous lesions with annular morphology on the posterior thorax, along with facial erythema and papules. Histopathological examination of skin biopsies demonstrated features consistent with SCLE, including interface dermatitis, perivascular, and perifollicular lymphocytic infiltrates, and dermal mucin deposition. However, negative direct immunofluorescence findings for granular deposits raised diagnostic uncertainty. Repeat biopsy findings suggested features overlapping with both lupus erythematosus tumidus and JLIS, adding complexity to the diagnosis. Thus, the integration of known diagnostic criteria and characteristics played a pivotal role in navigating the challenges of establishing an accurate diagnosis in this complex case.

Conclusion:

This case underscores the diagnostic challenges in distinguishing between SCLE and JLIS due to overlapping clinical and histopathological features. The necessity for repeated biopsies and comprehensive immunological evaluations highlights the importance of a multidisciplinary approach involving dermatologists, pathologists, and rheumatologists in reaching an accurate diagnosis and guiding appropriate therapeutic interventions tailored to

the specific entity. Increased awareness of these rare entities and their distinguishing characteristics is essential for optimizing patient care and preventing unnecessary intervention. Further research is warranted to elucidate the underlying pathogenic mechanisms and refine diagnostic criteria for these rare dermatological conditions.

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Abstract N°: 6810

IgG4-related disease presenting as panniculitis of the breast: case report and review of the literature

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Introduction & Objectives:

IgG4 related disease is a rare multi-system immune-mediated condition that is characterized by IgG4-positive plasma cell infiltrates (Stone JH, 2012). The clinical presentation of IgG4 related disease is variable and may affect almost any organ (Stone K, 2012;). Histology typically shows a dense lymphoplasmacytic infiltrate, storiform fibrosis and obliterative phlebitis (Deshpande V, 2012). Elevated serum IgG4 levels are seen in 60-70% of patients (Stone JH, 2012). Diagnosis is based on the 2019 ACR/EULAR classification criteria for IgG4 related disease (Walle Z, 2020).

Materials & Methods:

We present a 57-year-old female with a three-year history of a slowly progressing, indurated right breast plaque. The skin overlying the plaque was erythematous to violaceous and approximately 60 by 70mm in size. Initial histology showed lobular lymphocytic panniculitis. Repeat biopsies revealed chronic inflammation with fibrosis and an excess of IgG4+ cells with an elevated IgG4:IgG ratio. Serum IgG4 level was elevated at 2.54g/L.

Although her disease was initially responsive to corticosteroids, it continued to progress despite trials of hydroxychloroquine, dapsone and intralesional triamcinolone. She then received treatment with Rituximab.

Results:

Skin manifestations of IgG4-related disease are rare and usually present concurrently with systemic disease. Common features include papules, patches, plaques, nodules and purpura, and most often present on the head and neck region (Charrow A, 2016). Lesions may be present for months or years prior to diagnosis (Jalilian C, 2014).

IgG4 related sclerosing mastitis is a rare condition which clinically may mimic malignancy, recurrent abscess, or panniculitis (Bajad S, 2020; 35 (4); Bajad S, 2020; 35 (4); Zen Y, 2005;29; Cheuk W & 33:, 2009:33; Ogiya A, 2014;21).

Conclusion:

We present a case of IgG4 disease presenting as panniculitis of the breast, treated with rituximab and review the literature on manifestations of dermatological presentations of IgG4 disease.

This case highlights the need of awareness of the varied cutaneous presentations of IgG4 disease and the importance of clinicopathological correlation.

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Abstract N°: 6890

Wegener Granulomatosis: Atypical clinical presentation on vulvar mucosa

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Introduction & Objectives:

Granulomatosis with polyangiitis (GPA), formerly known as Wegener granulomatosis, is an uncommon form of multisystem autoimmune disease of unknown etiology, combining necrotizing vasculitis of small-sized blood vessels and extravascular granulomatous inflammation. GPA has a wide spectrum of clinical presentations, mostly affecting upper and lower airways, leading to recurrent respiratory infection as well as life-threatening necrotizing glomerulonephritis. A key feature of GPA is the presence of anti-neutrophil cytoplasmic antibodies against proteinase 3 (PR3-ANCAs) in approximately 90% of systemic forms and in 50% of localized forms, which are highly specific and, therefore, have a high diagnostic value. Dermatologic involvement, reported in 50%-60% of the patients with GPA, includes purpura, usually involving the lower extremities, ulcers, papules, vesicles, and subcutaneous nodules (granulomas) commonly involving the olecranon region.

Materials & Methods:

A 22-year-old woman was admitted to our clinic with a 4-month history of ulcerations on her face, trunk, and genital region. During that time, our patient had general symptoms, including periodically high fever, malaise, and weight loss. Upon admission, a physical examination revealed small ulcerations approximately 1x1 cm in diameter in the preauricular and deltoid region. A couple of circular, atrophic scars were seen on her forehead, chin, and trunk. In the genital area, around the vaginal introitus, deep ulcerations were noted. Due to tissue necrosis, the right labia minora was almost completely detached from surrounding structures.

Results:

A histopathological (HP) evaluation of skin lesions and nasal mucosa suggested eosinophilic granulomatosis with polyangiitis. MRI of the endocranium showed signs of pansinusitis and otomastoiditis. PR3-ANCA were present, and their concentration in the serum was more than ten times higher than reference values. Initially, the patient was treated with pulse doses of systemic corticosteroid therapy and methotrexate 15 mg weekly. Due to the ineffectiveness of treatment, rituximab was introduced to therapy, and complete epithelization was achieved. A thorough gynecological evaluation was conducted and reconstruction of labia minora is planned.

Conclusion:

GPA is a severe and complex disease that requires early diagnosis and treatment. Genitourinary system involvement in GPA occurs in less than 1% of cases. Despite its low prevalence, it should not be overlooked as a differential diagnosis in any age group due to its potential to cause significant organ damage. In everyday clinical practice, it is important to differentiate genital ulcerations due to GPA from other causes, such as infections or malignancies. In terms of management, the presence of genital ulcerations can be very challenging and requires a comprehensive approach.

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Abstract N°: 6901

Psoriasis in systemic lupus erythematosus. Pathogenetic and therapeutic implications

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Introduction & Objectives: Psoriasis has been described in cases of systemic lupus erythematosus (SLE). In a cohort of SLE patients retrospectively studied cases of psoriasis, mainly plaque psoriasis were identified. In this cohort psoriasis did not have an impact on disease activity and target organ damage accumulation. This rare coexistence raises pathogenetic questions and has therapeutic implications. The aim was to present the case of a female patient who had psoriasis and developed SLE.

Materials & Methods: The case of a female patient is presented who developed psoriasis at the age of 52. Topical treatment was applied. A year later the patient presented with severe fatigue, headaches, arthritis and positive ANA 1/2560. There was no evidence of enthesitis.

Results: SLE was diagnosed and hydroxychloroquine, along with methotrexate and prednisolone were administered. Liver enzymes increased and orally administered methotrexate was switched to subcutaneously administered methotrexate. Fatigue and headaches improved. Topical treatment was adequate for the management of psoriasis.

Conclusion: The case of a patient with psoriasis who was diagnosed with SLE is described. This very rare coexistence has been previously described. It raises the hypothesis of common pathogenetic mechanisms being involved in the pathogenesis of both psoriasis and SLE. It corroborates the idea that autoimmune mechanisms may be involved in the pathogenesis of psoriasis. This coexistence also has therapeutic implications. There is evidence that methotrexate administration may be beneficial in these very rare cases of psoriasis in the context of lupus. In cases previously described in the literature as well as in our case topical treatment seemed to be adequate for the management of psoriasis.

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Abstract N°: 6947

Serologic Biomarkers in Pemphigus Monitoring: C-reactive Protein, Macrophage Migration Inhibitory Factor, and Prolactin Levels Versus Autoantibody Assays

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Introduction & Objectives:

Evaluation and monitoring of pemphigus vulgaris (PV) typically involve autoantibody detection by enzyme-linked immunosorbent assay (ELISA) and indirect immunofluorescence (IIF). We aimed to determine the levels of antipemphigus immunoglobulin (Ig) G autoantibodies using ELISA and IIF (as standard biomarkers), and compare it to prolactin, macrophage migration inhibitory factor (MIF), and C-reactive protein (CRP) (as nonstandard biomarkers) to determine which of these non-standard biomarkers is appropriate for PV monitoring.

Materials & Methods:

The experiment was performed before and during therapy. Anti-Dsg immunoglobulin G autoantibodies were measured using ELISA and IIF (as standard biomarkers) versus prolactin, MIF, and CRP (nonstandard), before 1 and 3 months after the treatment. Before beginning the treatment, the severity of the disease was determined using the pemphigus disease area Index (PDAI).

Results:

We enrolled 60 newly diagnosed patients with PV (32 men and 28 women; mean age=43.8±14.2 years). Before treatment, the levels of anti-Dsg1, anti-Dsg3, and IIF were high and had a significant relationship with PDAI. PDAI also had a connection with the levels of CRP and prolactin. The anti-Dsg1, anti-Dsg3, IIF, and CRP titers decreased in patients treated with conventional (prednisolone plus azathioprine) and rituximab therapy during and after treatment.

Conclusion:

In conclusion, anti-Dsg1, anti-Dsg3, and IIF autoantibody titers remain standard biomarkers for assessing disease activity, severity, and PV monitoring. The trend of CRP was similar to that of anti-Dsg1, anti-Dsg3, and IIF. Thus, CRP may be used for PV monitoring.

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Abstract N°: 6991

Risk factors for fecal incontinence in patients with systemic sclerosis

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Introduction & Objectives: Systemic sclerosis is a connective-tissue autoimmune disease, characterized by a chronic inflammatory process, with varying degrees of tissue fibrosis and small vessel vasculopathy. The main organs affected are the skin and the gastrointestinal tract. Gastrointestinal involvement can occur from the oral cavity to the anorectum. Pelvic floor disorders, such as fecal incontinence, are under diagnosed in these patients. They can be severe in some cases and have a significant negative impact on quality of life. A more detailed study of the risk factors for fecal incontinence could guarantee earlier and more individualized multidisciplinary treatment. The present study aims to evaluate the frequency of fecal incontinence, its risk factors and impact on the quality of life of patients with systemic sclerosis, in addition to proposing a prediction score for this outcome in this population.

Materials & Methods: An observational, cross-sectional, prospective study was carried out in a reference center in the city of São Paulo, in which 120 patients followed at the Systemic Sclerosis outpatient clinic were submitted to validated questionnaires about fecal incontinence and its impact on quality of life. The Cleveland Clinic Fecal Incontinence Index, the Bristol stool consistency scale, the general quality of life index (SF-12), the Fecal Incontinence Quality of life index (FIQL) and, the Pelvic Floor Bother Questionnaire index (PFBQ) were used. Clinical and demographic data were collected from an electronic medical record: age, sex, BMI, comorbidities, skin photo type, continuous use medications, previous surgeries, parity and mode of delivery, clinical subtype, systemic sclerosis duration, clinical manifestations, internal organ involvement and autoantibody profile.

Results: Of the 120 patients evaluated, 45 (37.5%) had some degree of incontinence, 19 (42.2%) mild FI, 20 (44.4%) moderate FI and 6 (13.3%) severe FI. Each additional comorbidity and each additional year of illness increased the patient's chance of developing fecal incontinence by 41% and 6%, respectively. Patients with urinary incontinence were 9.52 times more likely to have associated fecal incontinence, regardless of other characteristics. There was a significant impact on the quality of life of these patients, since 31% of patients with fecal incontinence considered their health to be poor due to fecal loss, with suicidal ideation in 6.7% of cases. Using the logistic and additive mathematical formulas developed in this study, it was possible to objectively calculate the fecal incontinence predictor score for these patients.

Conclusion: Systemic sclerosis presents a high frequency of fecal incontinence, with a significant impact on the quality of life of these patients. In this study, the number of comorbidities, urinary incontinence and longer illness duration were associated with a greater risk of fecal incontinence. The prompt identification of risk factors for fecal incontinence through the predictive score in patients with systemic sclerosis can help the clinician to identify this comorbidity, providing individualized management for these patients, as proposed in the additive score.

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Abstract N°: 7017

Unmasking Brunsting-Perry Cicatricial Pemphigoid in a woman with a unique chronic ulceration mimicking a cutaneous carcinoma.

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Introduction & Objectives:

Brunsting-Perry pemphigoid (BPP) is a rare autoimmune blistering disorder classified within the spectrum of auto-immune bullous diseases. BPP is a heterogeneous disease with several target antigens (BP180, collagen type VII), responsible for vesiculo-bullous lesions restricted to the head and neck region. We herein present a case of BPP initially mimicking either a malignant etiology, such as a rapidly infiltrating squamous cell carcinoma, or a infectious etiology.

Materials & Methods:

In February 2023, a 55-year-old woman originated from Mali, presented with a 2-months old, unique, painless and ulcerated lesion of 7 centimeter on the right frontal scalp, covered by a thick crust. Her medical history comprised a heterozygous drepanocytosis and an overlap myositis on minimal therapy for years (5 mg/day of prednisone). Initial examination revealed a superficially ulcerated lesion with well-defined margins and purulent discharge. The rest of the skin and mucous membranes were not affected. No signs of giant cell arteritis were noted. A rapidly progressive cutaneous cancer or a skin infection was initially suspected.

Results:

Bacterial culture and herpes simplex virus PCR on local swab were negative. Histological examination of a first punch biopsy and of a subsequent excision biopsy were inconclusive, showing a non-specific ulceration with a mixed dermal infiltrate comprising plasma cells and no keratinocytic tumour proliferation. On the latter, the presence of intra-histiocytic parasites was suggested. Leishmaniasis was ruled out by negative results on the direct wet mount exam, PCR test, serology and blot test. Considering the immunocompromised status, other causes of common and opportunistic infection were ruled out by negative results of the following exams: serological diagnosis of syphilis, mycological direct wet mount exam and cultures, microscopic examination of auramine-stained smears, solid and liquid cultures for mycobacterial infection, and finally pan-infectious shotgun metagenomic sequencing on a biopsy sample. While performing the latter biopsy for genomic analyses, we noticed cicatricial changes around the lesion and discrete cleavage on small areas. New skin biopsies on this area and perilesional direct immunofluorescence (DIF) analyses finally demonstrated respectively subepidermal cleavage with blister formation without neutrophilic infiltrate and a linear IgG deposit along the dermo epidermal junction. On DIF examination, dermis and epidermis were split with IgG staining both the floor and the roof of the bullae. Indirect immunofluorescence, ELISA and immunoblotting did not demonstrate circulating auto-antibodies, notable against BP-180 or collagen type VII. However, multiple connective tissue antibodies were found positive (anti-RNP, dsDNA, Sm, SSA, SSB, NXP2).

Thus, we concluded to a BPP presenting as a single lesion. Treatment with daily clobetasol propionate cream allowed a partial remission at 3 month follow-up. Sulfasalazine was preferred to dapsone as second line considering the chronic anemia.

Conclusion:

Our case report highlights that BBP can mimic other diseases, including cutaneous malignancies such as squamous cell carcinoma, or a infectious** etiology**,** notably in the absence of skin bullae**.** Clinicians should thus maintain a high index of suspicion for autoimmune blistering diseases and performe a DIF in case**s** of chronic and unique erosion of the head and neck region.

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Abstract N°: 7029

Pemphigus and comedonic lupus erythematosus

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Introduction & Objectives:

Pemphigus erythematosus (PE), also known as Senear-Usher syndrome, is a rare autoimmune blistering dermatosis combining features of pemphigus foliaceus (PF) and lupus erythematosus (LE). PF usually occurs in the seborrheic areas and presents with flaccid bullae that rupture easily and turn into erythematous scaly and crusted plaques. The LE lesions usually consist of discoid plaques with “carpet-tack” scales characteristic of chronic cutaneous LE (CLE), but other manifestations of CLE may be observed, such as chronic disseminated CLE and malar erythema of acute skin lupus. We present a patient with PE manifesting with the rare cystic and comedonic form of CLE.

Materials & Methods:

A 52-year-old man was hospitalized for an eruption of 1-year duration consisting of non-pruritic erythematous, scaly and crusted oozing lesions on the scalp, face, ears, face, chest and back. For the last 4 years the patient suffered from the appearance of multiple papules and plaques that had been diagnosed and treated as “allergic contact dermatitis” to construction materials. Multiple follicular pustules, cysts, open comedones, and acneiform atrophic scars were seen on the face, ears and trunk. Former treatment with systemic methylprednisolone 60 mg/daily improved the rash but was discontinued due to gastric complaints.

Results:

Laboratory studies showed elevated white blood cells, gamma-glutamyl transferase, and complement 4 while ANA titer and complement 3 levels were normal. Histopathological examination revealed perivascular infiltrate in the upper and middle dermis. Direct immunofluorescence on perilesional skin demonstrated intercellular deposition of IgG and C3 in the epidermis and a homogeneous band of IgM at the dermo-epidermal junction (DEJ). ELISA Dsg 1 was strongly positive while ELISA Dsg 3 was negative. Phototest result was interpreted as normal. The diagnosis of PE was based on clinical and laboratory results. Reintroduced methylprednisolone 40 mg/day, azathioprine 150 mg/day, gastroprotector, clobetasol propionate cream, and photoprotection resulted in clinical improvement.

Conclusion:

The diagnosis of Senear-Usher syndrome in the present case was based on clinical, histologic, immunofluorescent and immune-serologic criteria, i.e. presence of in vivo bound and circulating anti-Dsg 1 antibodies combined with the characteristic immunopathologic finding at the DEJ. The clinical manifestations of the LE counterpart of the syndrome were compatible with the rarely described comedonic variant of CLE which can be easily mistaken for acne vulgaris, naevus comedonicus, as well as acneiform eruption induced by the long-term systemic corticosteroid treatment. All of these were ruled out in our patient. Therefore, comedonic CLE may be considered as a rare atypical manifestation of PE.

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Abstract N°: 7040

Thymoma, a Nexus of Autoimmunity: A Rare Association with Systemic Lupus Erythematosus

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Introduction & Objectives:

Thymoma, a tumour originating in the epithelial cells of the thymus, is often associated with various immunologic disorders. However, its link to systemic lupus erythematosus (SLE) is exceedingly rare, with only a few documented cases, estimated at 0.5%.

Herein, we report a unique case detailing this unusual association.

Case description:

A 60-years-old woman, had been diagnosed with myasthenia gravis and thymoma 14 years earlier managed with azathioprine (150 mg/day) and Ambenonium (50mg/day), and pernicious anemia 4 years prior, presented to our department with a 4-week history of skin rash. Physical examination revealed numerous fresh-red papules on her face and extremities, accompanied by purpura on her legs, photosensitivity and polyarthralgia involving knees and ankles. No mucosal eruption were observed. Skin biopsy demonstrated focal vacuolization, ymphocytic and neutrophilic lichenoid infiltration in the epidermis and septal and perilobular panniculitis without vascular involvement. Laboratory invistigations revealed lymphopenia (732), strong positive antinuclear antibody (ANA) at 1:320, while anti-DNA, complement levels (C3, C4, CH50) and renal exploration were within normal limits. Radiographs of the joints were unremarkable. Based on the clinical, histopathologic and laboratory findings, systemic lupus erythematosus was diagnosed (with a score of 14 points on EULAR/ ACR-19 criteria). The patient was initiated on prednisone (60 mg/day) with gradual tapering regimen leading to significant improvement in symptoms.

Conclusion:

The association between lupus and thymoma has been reported in 36 cases in the literature, with thymoma is benign in 59% of the cases. Clinical manifestations of lupus in these cases are often nonspecific except for a median age of onset 48 years, and sex ratio, 4:3 (Female-Male). The clinical outcome of the lupus does not appear to be influenced by the thymectomy. Thymoma may precede lupus by several years, occur concurrently, or be diagnosed later. Although the pathogenic link remains elusive, it’s hypothesized that the decline in thymic function associated with thymoma may lead to the proliferation of autoreactive T lymphocytes and activation of B cells. Patients with thymoma should be monitored post-thymectomy, as autoimmune diseases, including lupus, may manifest later. Conversely, the occurrence of lupus in patients around the age of 50 should raise suspicion for thymoma.

This case underscores the importance of recognizing the rare association between thymoma and systemic lupus erythematosus, prompting clinicians to consider comprehensive evaluations and long-term surveillance in patients with either condition. Further research is warranted to elucidate the underlying mechanisms and optimize management strategies for these complex presentations.

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Abstract N°: 7074

A case of cutaneous polyarteritis nodosa associated with hepatitis B infectionIntroduction

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Introduction:

Cutaneous polyarteritis nodosa (CPAN) is a rare type of vasculitis affecting small to medium-sized arteries within the dermis and subcutaneous tissue without systemic involvement. Numerous infectious and non-infectious factors have been linked to both the onset and recurrence of the condition. Here, we present the case of a CPAN in a 53-year-old woman, which revealed a hepatitis B infection.

Case report:

A 53-year-old woman, with a medical history of hypothyroidism and smoking, presented with recurrent episodes of fever and painful nodules in the extremities that had been evolving over the past year. Some nodules resolved spontaneously, while others progressed into ulcers. No respiratory infections or recent medication use were reported, and there was no history of Raynaud's phenomenon or malar rash.

On physical examination, bilateral painful subcutaneous nodules were observed on the lower limbs, mainly on the right side. Three ulcers were noted on the right lower limb, one measuring 5cm on the dorsum of the foot. No purpura, livedo, or atrophic scars were present, and vital signs were normal.

A biopsy revealed vascular wall necrosis and thrombosis in medium-sized vessels, with subacute inflammatory changes in the panniculus and perivascular neutrophilic infiltration. Further investigations were normal, but serological tests showed a positive hepatitis B surface antigen. Consequently, the patient was diagnosed with hepatitis B virus-associated cutaneous PAN.

Discussion:

PAN is a type of vasculitis that typically affects organs like the lungs, kidneys, and skin. When it's limited to the skin, it's called CPAN (Cutaneous PAN). CPAN mainly involves small and medium arteries at the dermal-subcutaneous junction without venous involvement. Symptoms include subcutaneous nodules, livedo reticularis, ulcers, and purpura. Diagnosis depends on characteristic skin lesions, the absence of systemic vasculitic features, and supportive histopathologic findings. Both PAN and CPAN can be triggered by underlying diseases, infections, or medication use.

The connection between viral hepatitis and vasculitis was first suggested by Paull R, who observed PAN in military officers. Mowrey and Lundberg further supported this link with 230 PAN cases, 16 of which had viral hepatitis. While hepatitis B is often linked to systemic PAN, the association with CPAN and HBV hasn't been consistently observed. In two large case series with 20 and 79 CPAN cases, all hepatitis B viral serologies were negative. It's hypothesized that immune complexes, due to immunoregulation disorders, mediate vascular damage.

Haut du formulaire

Bas du formulaire

It’s important to stress the existence of nosological ambiguity, with likely overestimation of numbers due to many case series being published prior to the revised classification of systemic vasculitis in 1994. Consequently, study populations may have included patients with eosinophilic granulomatosis with polyangiitis and microscopic polyangiitis.

Conclusion:

CPAN is a rare condition that can be the cause of recurrent ulcers on the lower limbs. Several triggering factors have been identified, including HBV, but the prevalence of this association remains unclear, as most studies addressing it are outdated. Therefore, there is a need for more recent studies on this subject.

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Abstract N°: 7136

Bullous pemphigoid in association with urothelial carcinoma

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Introduction & Objectives:

Bullous pemphigoid (BP) is an autoimmune blistering disease predominantly affecting the elderly. It is characterized by autoantibodies directed against two hemidesmosomal proteins, namely BP230 and BP180. There have been numerous reports on the association of BP with malignant tumours, mainly carcinomas of internal organs. Atypical clinical features such as figurate erythema, negative indirect immunofluorescence, or presence of antibodies against BP180 using Western immunoblotting, have been suggested as predictive for underlying neoplasia in patients with BP.

Materials & Methods:

A 77-year-old man was admitted to the dermatology department for a widespread blistering eruption of 3 months duration starting few days after a CT scan with iopamidol contrast for painless haematuria and difficulty in urinating. Further on the patient underwent a transurethral resection of a bladder tumour that was histologically identified as high-grade urothelial carcinoma with subepithelial infiltration. Few days after the surgery, new multiple blisters of various size and clear or haemorrhagic content appeared on the trunk and extremities, including the distal part of the hands and feet. In the abdominal area the vesiculo-bullous lesions had an annular arrangement at the periphery of erythematous urticaria-like plaques. Additionally, the patient had history of type 2 diabetes mellitus under metformin and vildagliptin treatment.

Results:

Routine blood tests showed hyperglycaemia, hyperuricaemia, CRP of 10.5 mg/l, and slight anaemia. Histology from a blister was compatible with BP, and the direct immunofluorescence on perilesional skin showed linear deposition of immunoglobulin G and complement C3 at the basement membrane (BM) zone. ELISA BP180 was strongly positive (> 200 RU/ml) whereas BP230 tested negative. Treatment with doxycycline 100 mg/24h, dexamethasone 7 mg/24h, fluconazole 100 mg, and topical corticosteroids lead to clearance of the blistering eruption within a month.

Conclusion:

In this case presentation three potential triggering factors for BP can be identified, i.e. the urothelial carcinoma, the contrast substance, or the gliptin antidiabetic therapy, however our attention is focused on the associated malignant disease. In some cases, BP may occur before or after the diagnosis of bladder cancer. Some studies suggest that common genetic and immunological factors may play a role in their pathogenesis and may be involved in the development of both conditions. Additionally, the BP180 antigen is also known to exist within the bladder wall. This raises the possibility that anti-tumour antibodies, including anti BP180 antibodies, cross-react with the cutaneous BM antigens. Although the link between BP and malignancies is still subject of debates, these hypotheses provide some possible mechanisms that may explain their association.

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Abstract N°: 7144

Juvenile generalized morphea: Case report

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Juvenile generalized morphea: Case report

Introduction & Objectives:

Morphea, also known as localized scleroderma, is a rare disorder, which classically presents benign and self-limited evolution and is confined to the skin and/or underlying tissues. It is a chronic connective tissue disease of unknown etiology. Patients with morphea typically present systemic symptoms such as malaise, fatigue, arthralgias, and myalgias, as well as positive autoantibodies.

Our case report explores juvenile generalized morphea, discussing its clinical features, diagnostic approach, and management strategies.

Materials & Methods:

In this report, we detail the case of a patient diagnosed with likely juvenile generalized morphea.

Results:

A 10-year-old child with a history of recurrent pharyngitis, who presented with pruritic lesions evolving for 7 months,initially erythematous then gradually became shiny brownish with a hypochromic center on the four limbs, back, trunk and abdomen. Inflammatory polyarthralgias and asthenia were reported. Clinical examination revealed circumscribed, well-defined, hyperpigmented plaques with a depigmented center and smooth surface, indurated and sclerosed. Dermoscopy revealed a pigmented network with an erythematous background and rainbow appearance in places, central whitish patches and a few scales.

Laboratory investigations revealed no abnormalities, notably the negativity of anti-nuclear antibodies and anti-Scl-70 antibodies.

The diagnosis of generalized morphea was retained due to the presence of more than 4 plaques over 3 cm in size, associated with arthralgia and asthenia, as well as the absence of Raynaud's syndrome, sclerodactyly and abnormalities on periungual dermoscopy.

The patient was treated locally with a combination of dermocorticosteroids and calcipotriol along with topical tacrolimus, resulting in significant clinical improvement.

Conclusion:

Generalized morphea is one of the most severe subtypes, characterized by widespread cutaneous involvement, and in some cases, extension to subcutaneous tissues and fascia. It is often confused with systemic scleroderma, and is distinguished by the absence of Raynaud's phenomenon, sclerodactyly and capillaroscopy abnormalities.

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Abstract N°: 7146

Reticular erythematous mucinosis: a diagnostic challenge.

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Introduction & Objectives:

Reticular Erythematous Mucinosis (REM) is an uncommon entity. Its clinical and histological overlap with other inflammatory dermatoses such as cutaneous lupus erythematosus can render its diagnosis challenging.

Materials & Methods:

We present an illustrative clinical case of REM and conduct a literature review on the clinical-pathological characteristics of this disease.

Results:

A 41-year-old male patient with no significant past medical history was referred to dermatology outpatient clinic for a 4-year-old erythematous-edematous plaque measuring 10 x 7 cm in the sternal area. An incisional biopsy was performed, revealing mucin deposition with interstitial distribution in the reticular dermis, accompanied by a mild superficial and mid-perivascular chronic inflammatory infiltrate, without epidermal alterations. Blood tests showed no abnormalities in the complete blood count, hemostasis, general biochemistry, or autoimmunity. The diagnosis of reticular erythematous mucinosis was established and treatment was initiated with hydroxychloroquine 200 mg daily, with good tolerance and complete remission of the lesion after 6 months of treatment

The exact pathophysiology of REM remains unknown. Clinically, it is characterized by erythematous plaques on the trunk, which can present in two forms: plaque-like and reticular. Histologically, it is characterized by a perivascular infiltrate in the superficial and mid dermis, along with mucin deposits in the dermis. In some cases, direct immunofluorescence may demonstrate immunoglobulin deposition in the epidermal basement membrane. This feature, along with the plaque-like clinical appearance, exacerbation after photoexposure, and response to antimalarial treatments, is shared with cutaneous lupus erythematosus. The treatment of REM can pose a challenge, with antimalarials being the preferred option

Conclusion:

REM is an uncommon entity with clinical and histological characteristics that overlap with cutaneous lupus erythematosus. A better understanding of its pathophysiology could help define the boundaries of this entity more clearly and perhaps pave the way for new therapeutic alternatives.

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Abstract N°: 7153

Hyperhidrosis overlaps with Raynaud's syndrome and therapeutic alternative

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Introduction: Raynaud's phenomenon manifests itself as transient ischemia of the extremities due to the vascular response to cold, stress or vibrational mechanisms. Through the symptoms of pallor, followed by cyanosis and distal hyperemia, they can be of primary, idiopathic or secondary etiology to an autoimmune disease, mainly rheumatological. Hyperhidrosis, on the other hand, is a disorder of overstimulation of cholinergic receptors in the eccrine glands, generating excessive, focal or generalized sweating, overcoming the thermoregulation mechanism through sympathetic autonomic fibers. While the focal, involves in a localized and symmetrical way the hands, feet and face, due to autonomic dysfunction worsened by physical, emotional and thermal stimuli, the generalized is related to autonomic disorders secondary to neurological, endocrine, metabolic, febrile or malignant pathologies. Both pathologies arise from dysfunctions of the sympathetic autonomic nervous system and can occur simultaneously. **Results:** Female, 24-years-old, complained of increased sweating in the armpits, groin, hands and feet for 8 years, with worsening during periods of extreme temperatures such as summer and winter and when she feels anxiety. She reports the use of oxybutynin 5mg/day, without improvement. Previously diagnosed with Raynaud's Syndrome. He uses oxybutynin hydrochloride associated with nifedipine 10mg/day, under follow-up with the rheumatologist. At another time, he used diosmin in association with hesperidin, without response (according to information collected). As a course of action, it was decided to exchange nifedipine for pentoxifylline, as the medication in use can worsen hyperhidrosis, as a compensatory mechanism. In addition to the change, we added deodorant with 15% aluminum with good sweat control, without changing the condition of Raynaud's Phenomenon. **Conclusion:** Both pathologies derive from changes in the sympathetic autonomic nervous system. In the patient's case, after a rheumatological diagnosis, sweating began. Oxybutynin is ineffective in 30% of patients, which is part of the statistics. Therefore, despite the functionality of nifedipine as the first choice for the treatment of Raynaud's phenomenon, its anticholinergic action worsened the symptoms of hyperhidrosis. As pentoxifylline is another option for sweat control without intervening in vascular pathology, a measure associated with 15% aluminum deodorant was ideal for the patient in question. Finally, another alternative would be the use of botulinum toxin type A (BoNTA), which has an effect on acetylcholine, norepinephrine, substance P, peptide related to the calcitonin gene and release of glutamate from nerve terminals, which determines new paths for treatment of hyperhidrosis in patients with Raynaud's phenomenon, without its complications, based on individualized medical practice.

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Abstract N°: 7172

Multiimmunity ratio in a patient with bullous penfigoid rheumatoid arthritis

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Introduction & Objectives:

Bullous pemphigoid (BP) is the most common bullous autoimmune disorder, usually in people >60 years old. It is characterized by autoantibodies IgG against dermo-epidermal binding proteins and initially presents with a polymorphic eruption that progresses to blisters and erosions. 25% of patients with the autoimmune disease develop other autoimmune disorders, known as “autoimmune diathesis”. Association with rheumatoid arthritis (RA) suggests a connection between these autoimmune conditions.

Materials & Methods:

A 61-year-old female, from Mexico City, presented with an acute history of multiple blisters and flictenas, on an erythematous base of irregular shape and well-defined edges, located in the neck, trunk, arms, and legs with predominance in folds.

Past medical history of rheumatoid arthritis 28 years of diagnosis with no previous treatment.

Blister biopsy was performed reporting positivity for IgG and C3C, positive immunohistochemistry: linear pattern in basal membrane 3+. Confirming the diagnosis of bullous pemphigoid.

After three weeks we requested general blood studies and complementary studies in search of an infectious focus, as well as immunological studies to determine associations, which are still pending, according to results we will regulate behavior to follow. The patient probably will benefit from rituximab treatment along with other biological drugs to control BP and rheumatoid activity.

Results & Conclusion:

Interleukin-6 (IL-6) is crucial for immune responses, but its overexpression is linked to autoimmune diseases and capillary leakage syndrome. Autoimmune bullous diseases are skin disorders caused by an autoimmune response against intercellular adhesion molecules. The pemphigoid group comprises conditions such as BP, associated with IgG autoantibodies against hemidesmosomes proteins at the dermo-epidermal junction. Blistering involves complement activation and inflammatory cells. These disorders are usually related and characterized by shared “autoimmune diathesis”.

The clinical significance of this case lies in examining how PA interacts with other autoimmune diseases, highlighting the need to recognize and diagnose promptly the joint occurrence of autoimmune disorders to make a difference in the patient’s treatment and quality of life.

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Abstract N°: 7173

Lichen planus pemphigoid

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Introduction & Objectives:

Lichen planus pemphigoid (LPP) is a rare autoimmune bullous dermatosis, especially in children. It is characterized by the coexistence of both lichen planus lesions and authentic bullous pemphigoid. We report a new case

Materials & Methods:

A 5-year-old girl presented with a pruritic dermatosis evolving for 3 months, characterized by papules and lichen-like plaques with bullous and post-bullous lesions. The bullous lesions consisted of tense blisters, arising on lichenoid lesions and on clinically uninvolved skin. The lesions were located on the trunk, all four limbs, perineum, and face.

The diagnosis of lichen planus pemphigoid was confirmed by both histopathological examination, which revealed features of lichen planus and subepidermal blistering, and ELISA testing, which detected anti-BP180 antibodies in the patient's serum.

General corticosteroid therapy was initiated, leading to clinical remission.

Results:

During LPP, blisters can appear on both lichenoid lesions and healthy skin. The blistering eruption typically follows that of lichen planus, although it can rarely be concurrent, but seldom precedes it. Histologically, the appearance combines typical features of lichen planus and a subepidermal blistering dermatosis. Direct and indirect immunofluorescence as well as ELISA reveal typical features of bullous pemphigoid. The main differential diagnosis remains bullous lichen planus, during which blisters form due to the intensity of lichenoid inflammation and basal membrane degeneration.

From a pathophysiological perspective, basal keratinocyte lesions generated by the lichenoid infiltrate unmask antigenic determinants or create new antigens, leading to the formation of autoantibodies and the onset of bullous pemphigoid.

The pediatric form of LPP is rare, with an average onset age of 12 years and a sex ratio of 3. Similar to adults, lichen planus precedes bullous lesions by a few weeks. Vesiculobullous lesions are common on the extremities, with palmoplantar involvement described in 50% of cases. The same histological and immunohistochemical lesions are observed as in adults. Dapsone is the treatment of choice for pediatric LPP, with systemic corticosteroid therapy as a second-line option. In our patient, systemic corticosteroid therapy at a dose of 1.5 mg/kg/day for 6 months resulted in remission.

Conclusion:

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Abstract N°: 7176

Morphea in the spectrum of autoimmune syndrome induced by silicone breast implants

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Introduction & Objectives:

Autoimmune conditions from the scleroderma spectrum, specifically morphea, systemic sclerosis (SSc), and eosinophilic fasciitis have occasionally been described in relation to silicone breast implants, regardless of the latter being placed for cosmetic augmentation, post-cancer breast reconstruction, or for gender-affirming surgery. A large cross-sectional study has shown an increased risk of developing any autoimmune or rheumatic disorder in patients with breast implants. Often, a myriad of systemic symptoms is present following silicone breast implantation that cannot meet the criteria of a definite rheumatic disease but may be suggestive of overlap syndromes. Two terms, namely “breast implant illness” and “autoimmune/inflammatory syndrome induced by adjuvants (ASIA)” have been introduced to designate this type of breast implant complications. We recently observed a patient with morphea and other systemic symptoms occurring after repeated breast implant surgery.

Materials & Methods:

A 39-year-old woman presented with complaints of erythematous to violaceous and brownish patches on her chest and back, persisting for 6 months. Eighteen months prior to the onset of symptoms she underwent a second breast implantation, because of corrupted and removed previous implants. The skin changes were accompanied by general fatigue, arthralgia, and swelling of the large joints and therefore, the second implants were also removed. Physical examination revealed several round, well-demarcated, erythematous, bruise-like and indurated plaques located on the trunk, upper, and lower extremities, and the lumbosacral region.

Results:

An extended scleroderma panel was positive for antinuclear antibodies at a titer of 1:1280, as well as for the specific autoantibodies RP11-97 and RP155-110. Additionally, elevated levels of anti-beta 2 glycoprotein I immunoglobulin (Ig)G and IgG antiphospholipid antibodies were found. Skin biopsy from distal dorsal phalangeal area was not supportive of SSc and direct immunofluorescence from normal skin was negative. The patient’s clinical presentation, in conjunction with laboratory findings, suggested an autoimmune etiology, possibly morphea, SSc, or scleroderma-like syndrome induced by silicone breast implants. Despite treatment with systemic corticosteroids and non-steroid anti-inflammatory drugs, symptom resolution was inadequate.

Conclusion:

This case underscores the complexity of autoimmune conditions, particularly in the context of potential triggers such as foreign body reactions to silicone implants known in the literature as ASIA syndrome. The challenge lies in the distinction between primary autoimmune diseases like SSc and secondary conditions induced by external factors. More research is needed to understand the connection between silicone implants and autoimmune reactions and to refine treatment approaches.

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Abstract N°: 7231

Pemphigus vegetans induced by indapamide

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Introduction :

Pemphigus vegetans is an autoimmune bullous dermatosis characterized by vegetative plaques primarily located in intertriginous areas. This rare form of pemphigus accounts for only 1 to 2% of all pemphigus cases.

In this report, we discuss a case involving a 70-year-old patient who developed pemphigus vegetans induced by indapamide.

case report:

A 70-year-old female patient, with a history of hypertension treated with Indapamide 1.5 mg/day for five years, presented pruritic bullous lesions in the inguinal region for the past five months. These lesions ruptured, giving way to exulcerative areas and gradually taking on a vegetative appearance.

Physial examination revealed erythematous-violaceous, malodorous and vegetative plaques occupying the entire inguinal region with a negative Nikolsky sign. Dry cheilitis with two pseudo-aphthoid endobuccal erosions, nasal erosions, and onychomadesis affecting the fingernails were also noted.

Complete blood count demonstrated hypereosinophilia at 1190 ; and anatomopathological tests and direct immunofluorescence showed acantholysis and intraepidermal cleavage with intercellular deposition of IgG and C3 within the intraepidermal space and on the surface of keratinocytes.

Indirect immunofluorescence was positive for IgG, with an intercellular pattern at a titer of 1280 ; and the pharmacological investigation pointed to indapamide as a potential trigger. Consequently, the diagnosis of indapamide-induced pemphigus vegetans was established.

Treatment involved oral corticosteroid therapy at 1 mg/kg/day, along with adjuvant immunosuppression (azathioprine 100 mg/day). Indapamide was discontinued, replaced by amlodipine.

The patient exhibited complete healing after two months of treatment, with no recurrence during a one-year and a half follow-up.

Discussion:

Pemphigus vegetans, a rare pemphigus variant, infrequently arises as a medication-induced condition.

Literature reports few cases of drug-induced pemphigus vegetans, and the onset duration between medication and lesion appearance varies widely.

It is crucial to investigate medication intake in any patient with pemphigus and to stop, if possible, any attributable medication.

This case stands out for its originality, considering the rarity of pemphigus vegetans induced by indapamide.

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Abstract N°: 7233

Bullous pemphigoid and Milia: A serie of seven cases

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Introduction & Objectives:

Milia are superficial keratin cysts seen as pearly white, measuring 1-2 mm in diameter. Milia are associated with diseases that cause subepidermal blistering, such as hereditary bullous epidermolysis (HBE), acquired bullous epidermolysis (ABE), bullous pemphigoid (BP), bullous lichen planus, and porphyrias. Bullous pemphigoid (BP) is the most common autoimmune blistering disease worldwide.

The aim of this study is to describe the occurrence and clinical-laboratorial findings of BP-milia association in our patients.

Materials & Methods:

A retrospective, descriptive, and monocentric study. Seven cases of BP with milia formation between January 2013 and May 2023 were included. BP confirmation was based on clinical, histological elements and linear fluorescence of IgG at the level of the basement membrane zone during direct and indirect immunofluorescence (DIF, IIF respectively). No cases underwent confocal microscopy.

Results:

Out of 147 BP cases collected, seven showed milia formation, corresponding to a prevalence of 4.76%. Six were males, with a median age of 66.8 years and an average diagnostic delay of 5 months. Neurological impairments were present in 3 patients. All patients presented typical BP lesions with histological examination revealing detached epidermis at the junction and eosinophilic inflammatory infiltrate at the dermal level. DIF showed junctional deposits of C3 and/or IgG. Indirect immunofluorescence for anti-BP autoantibodies (anti-BP180 and anti-BP230) returned positive with varying titers. Milia appeared within an average of 2.6 months. The eruption consisted of pearly white dome-shaped lesions measuring 1 to 2 mm in diameter, non-pruritic, localized at the sites of previous bullous scars in all cases except one where they appeared on healthy skin. The most common sites in our patients were the trunk and lower limbs, followed by the neck and face. Regression occurred within a few months without adjunctive ablative treatment.

Conclusion:

To our knowledge, the association of bullous pemphigoid and milium formation is a rare finding. In our series, the prevalence of this association is 4.76%, indicating the necessity of a careful differential diagnosis with HBE. Neurological impairments associated with BP were described in 3 of our patients. The neurological and BP profile suggests a link in the pathogenesis of BP, but no association with the appearance of milia has been found in the literature. The formation of milia during BP remains poorly understood, but an interaction of immunological predisposition and atypical interactions between hemidesmosomes and the extracellular matrix is suggested. Clinicians should be cautious in making an accurate diagnosis of the type of bullous dermatosis in the presence of these epidermal cysts.

To our knowledge, only one Brazilian study has revealed the association milia-BP. Our serie= is the first to shed light on this issue of milia, which presents more of an aesthetic problem for patients with bullous pemphigoid.

Further studies are necessary to understand the mechanism and provide therapeutic approaches to improve the quality of life of these patients.

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Abstract N°: 7296

Subcorneal pustular dermatosis in a 30-years old Omani female

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Introduction & Objectives:

Subcorneal pustular dermatosis (SPD), also known as Sneddon-Wilkinson disease, is a rare, relapsing, sterile pustular eruption of unknown etiology that develops most commonly in middle-aged or mature women. Here we describe a typical clinical presentation and supportive histological SPD, treated successfully with dapsone.

Materials & Methods:

A 30-year-old female presented to our clinic with a long-standing history of recurrent generalized pruritic pustular eruptions, resembling impetigo-like lesions in an annular distribution mainly affecting the trunk and extremities. Additionally, she reported swelling of both hands and feet. Her medical history revealed no significant medication use.

Over the past decade, she experienced repeated episodes of pustular lesions on the trunk, abdomen, upper and lower extremities, particularly in flexural areas, accompanied by aphthous ulcers. Wound cultures consistently showed heavy growth of Staphylococcus aureus. Each episode was managed with oral and topical antibiotics.

The dermatologic examination showed flaccid bullae and pustules (some displaying the classical appearance of half-pustular, half-clear fluid-filled blisters), situated on an erythematous base with fine scales over the trunk and the extremities. The face, palms, soles, and mucous membranes were spared. No lymphadenopathy or hepatosplenomegaly was presented.

Our differential diagnoses included Subcorneal Pustular Dermatosis (SPD), IgA-pemphigus, pustular psoriasis, and tinea. Because there was no history of exposure to a new drug; acute generalized exanthematous pustulosis (AGEP) was not considered in differential diagnosis. Cultures of the pustules were sterile. Biopsy was taken for light microscopy examination.

Investigations:

- CBC, LFT, UE and bone profile were within normal.
- Microscopic examinations of Skin biopsy demonstrated: acanthosis, mild orthokeratosis, the blisters are predominantly intraepidermal (subcorneal), with dermal interstitial neutrophils and papillary dermal edema. Possibility of subcorneal pustular dermatitis was though of.

So The diagnosis of SPD was made based on the clinical and histopathological findings.

Treatment: We kept patient on Dapson at a dosage of 100 mg daily and topical steroid twice daily.

2nd visit after 1 month patient came for F/U:

- All lesions resolved totally & no active eruptions
- Remaining are post-hyperpigmented patches mainly over posterior aspect of both hands and feet

Patient was well satisfied with the result of treatment

Results:

The history of recurrent pustular eruption with typical clinical picture along with the pathology findings all pointed toward the diagnosis of subcorneal pustular dermatoses which was further supported by the **significant** clinical improvement after dapsone.

Conclusion:

Subcorneal Pustular Dermatosis is a rare , relapsing, sterile pustular eruption disorder characterized by sterile pustular eruptions, often appearing as blisters filled with half-pustular, half-clear fluid. Initial treatment typically involves antineutrophilic medications as the primary therapeutic approach.

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Abstract N°: 7338

Bullous systemic lupus erythematosus

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Introduction & Objectives:

Bullous systemic lupus erythematosus (BSLE) is a rare autoimmune blistering disorder that typically manifests as an acute vesiculobullous eruption in patients with already known systemic lupus erythematosus (SLE). BSLE is more frequent in females than in males, reflecting the female preponderance in SLE and usually occurs in adults between the ages of 20 and 40 years, but may also affect children or elderly patients. The clinical manifestations of BSLE result from the disruption of epidermal-dermal adhesion secondary to autoantibody formation against type VII collagen, the major component of anchoring fibrils in the cutaneous basement membrane zone (BMZ).

Materials & Methods:

A 34-year-old woman presented with a 3-month history of widespread erythematous rash on the face, trunk, and upper extremities. Two weeks before the admission the patient developed a vesiculo-bullous eruption initially involving the vermilion border of the lips and subsequently affecting the upper part of the trunk in the absence of mucosal lesions or fever. Tense, clear, and haemorrhagic fluid-filled blisters, erosions, and crusting were present, as well as areas of postinflammatory hyperpigmentation and small superficial scars on the face. Additionally, she complained of persistent arthralgia. The patient was diagnosed with SLE three years before and was treated with hydroxychloroquine 200 mg daily, the latter being currently discontinued.

Results:

Routine laboratory parameters were within the normal range, except for mild anaemia. Extensive serologic workup revealed antinuclear antibodies at a titer 1:1280 and positive anti-dsDNA, Sm, Ro/SS-A antibodies and low complement (C)4. Histology showed subepidermal blistering with a prominent neutrophilic dermal infiltrate. Direct immunofluorescence demonstrated linear deposition of IgG as well as a granular band of IgM (++) and C3 along the BMZ. Based on the clinico-laboratory findings the diagnosis of BSLE was made. A rapid resolution of the blisters occurred following treatment with dapsone 50 mg daily. The blistering eruption responded quite favorably with no new lesions and satisfactory healing of pre-existing ones.

Conclusion:

BSLE is a rare blistering condition with a combination of distinctive clinical, histologic and immunopathologic features occurring in less than 1% of the patients with already diagnosed SLE. A rapid response to dapsone therapy is characteristic. The course of the disease is benign with remission within a year in most of the cases reported, including ours. Oral lesions are believed to occur in 30% of patients with BSLE. The most common presentations include ruptured and intact blisters along the lip vermilion, buccal mucosa and palate. In conclusion, we present a case of BSLE to illustrate and emphasize the need for an integrative diagnostic approach in such cases.

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Abstract N°: 7352

The association of alopecia and cutaneous lupus: 3 observations

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Introduction & Objectives:

Alopecia areata ,common cause of non-scarring acquired alopecia, is an autoimmune disease . It may be associated with a variety of comorbidities, especially autoimmune conditions. The coexistence of alopecia areata and systemic lupus erythematosus (SLE) has been reported; however, the link between these conditions has yet to be definitively proven. We present three cases of patients who were treated for alopecia areata and acquired cutaneous lupus. Our research is particularly interesting since it highlights the link between alopecia areata and autoimmune illnesses, specifically cutaneous lupus

.

Materials & Methods:

3 cases

Results:

Case 1:

Alopecia areata is a common cause of non-scarring acquired alopecia. It is an autoimmune disease that affects people who are genetically predisposed to it. It may be associated with a variety of comorbidities, especially autoimmune conditions. The coexistence of alopecia areata and systemic lupus erythematosus (SLE) has been reported; however, the link between these conditions has yet to be definitively proven. We present three cases of patients who were treated for alopecia areata and acquired cutaneous lupus. Our research is particularly interesting since it highlights the link between alopecia areata and autoimmune illnesses, specifically cutaneous lupus

Case 2:

54-year-old female patient with alopecia universalis and primary biliary cholangitis, with positive anti-MI2 antibodies, presented with papular erythematous scaly lesions of the neck,trunk, back and extremities, which had been present for 18 months. The skin biopsy was consistent with subacute lupus. Laboratory tests revealed positive antinuclear AAN antibodies, doubtful SAA antibodies, negative SSB and anti-native DNA antibodies, and autoimmune thyroiditis with positive anti-thyroglobulin antibodies. The patient was treated with hydroxychloroquine and dermocorticoids.

Case 3:

A 35-year-old patient with alopecia universalis presented with pruritic skin lesions under the breast that had been evolving for 5 years, with photosensitivity. A skin biopsy showed chronic lupus with positive direct immunofluorescence and linear deposits of IgG and IgM antibodies at the membrane level. The patient was treated with hydroxychloroquine and corticosteroid infiltration.

Discussion:

Alopecia areata is sometimes associated with other autoimmune or allergic diseases. To date, several cases of SLE associated with alopecia have been reported in 1.2% of cases. This association is more frequent in elderly and female patients, as in our observations. Indeed, CD4+ T lymphocytes play a central role in the genesis of both diseases, producing autoantibodies to several antigens, leading to the subsequent induction of autoimmunity. A genome-wide study has suggested that several genomic regions are significantly associated with alopecia, lupus erythematosus, and other autoimmune diseases, and that this association increases with age and gender. Female gender, age of onset over 40, and Jewish ancestry were identified as risk factors for the onset of SLE concomitant with alopecia. In some cases, alopecia may be associated with several autoimmune pathologies at the same time.

Conclusion:

Several autoimmune diseases can be associated with alopecia, and coexistence with lupus is not uncommon. A better understanding of the comorbidities associated with alopecia could help us to understand them better, and screen for them if necessary.

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Abstract N°: 7406

A case of bullous systemic lupus erythematosus

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Introduction & Objectives:

Bullous systemic lupus erythematosus (BSLE) is a rare autoimmune blistering disease that typically presents as an acute vesiculobullous eruption in a patient with known systemic lupus erythematosus (SLE).

The differential diagnosis of blistering in patients with SLE is broad and careful assessment is necessary to confirm the diagnosis. Our aim is to raise awareness of this rare disease and its management.

Materials & Methods:

We report a case of a woman diagnosed with SLE who presented with blistering lesions on her face, trunk and limbs.

Results:

This was a 38-year-old female patient diagnosed with SLE 10 years ago with positive antinuclear, anti-double-stranded DNA, anti-Sm, anti-RNP and anti-Ro antibodies. She was on treatment with belimumab for the last 15 days.

She consulted for asymptomatic lesions in the cervicofacial region, trunk and limbs of 2 months' duration. Examination revealed the appearance of tense vesicles and blisters on the normal-appearing skin of the trunk and extensor surfaces of the extremities. In addition, erosions could be seen on the oral mucosa. PCR for herpes simplex and zoster virus, anti-BP 180, anti-BP 230, anti-desmoglein 1, anti-desmoglein 3, anti-C7 and anti-transglutaminase IgA were requested, all of which were negative. Histopathology revealed a subepidermal blister and a superficial dermal inflammatory infiltrate with abundant neutrophils.

Based on anamnesis, physical examination, histology and serological tests, she was finally diagnosed with BSLE. Furthermore, a literature search was performed to correlate the lesions with the initiation of belimumab, with no findings. She required treatment with dapsone 50 mg daily.

Conclusion:

BSLE is a rare manifestation of SLE that occurs in less than 5% of patients with SLE.

BSLE mainly occurs in young women, with predominance among those with a dark phototype (V–VI), as in our patient. It is characterized clinically by tense vesicles or bullae not limited to photo-exposed areas, occurring predominantly on the trunk, the upper limbs, and the face, with mucous membrane involvement in 50% of the cases. Usually, lesion resolution occurring without scarring or formation of milia. Anti-C7 autoantibodies are involved in the pathogenicity of BSLE and are found in up to 69% of patients. The analysis of skin biopsies demonstrated subepidermal detachment, neutrophil-predominant dense infiltrate in the upper dermis, leukocytoclasis and absence of vasculitis. Dapsone is considered the treatment of choice based upon clinical

experience, the response is typically rapid with an efficacy of 90%.

The differential diagnosis includes other subepidermal autoimmune blistering diseases and other blistering disorders that can occur more frequently in patients with SLE. Autoimmune blistering diseases that share features with BSLE include epidermolysis bullosa acquisita, linear IgA bullous dermatosis, dermatitis herpetiformis, and anti-p200 pemphigoid, other subtypes of cutaneous lupus erythematosus and Rowell syndrome.

In addition, immunosuppressive therapy for SLE can increase risk for blistering infections, such as bullous impetigo, disseminated herpes simplex virus infections, and disseminated herpes zoster infections.

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Abstract N°: 7419

quality of life among patients with cutaneous lupus erythematosus

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Introduction & Objectives: Lupus erythematosus is a chronic or chronically relapsing, potentially multisystem disorder which often affects the skin. Cutaneous Lupus erythematosus (CLE) can occur concomitantly with SLE, or independently from SLE and can range in severity from mild, moderate or severe cutaneous disease. Clinical presentation of LE may be significantly different in people of different ethnicities. CLE can greatly impact the quality of life of the affected individual which in turn, can affect the patient's health seeking behavior, income-generating activities and other aspects of one's life. The objective of this study was to assess the quality of life among patients with cutaneous lesions associated with lupus erythematosus attending dermatology clinic at KCMC between September 2022 to March 2023.

Materials & Methods:

Both** new and previously diagnosed patients with CLE attending dermatology clinic were recruited in the study. Data was collected using structured questionnaire and validated DLQI tool. Bivariate and multivariate analysis were done to test for associations and confounders.

Results:

A total of 47 patients were requited in the study.** Chronic CLE was the most prevalent subtype observed. The mean DLQI of this cohort was 9.5 (± 6.36), where 40.4% of participants had great to very great impact on quality of life, 23.4% had moderate impact, whilst 36.2% had little to no impact on their quality of life owing to presence of CLE lesions. Factors significantly associated with a greater impact on quality of life were: type of medication used, positive diagnosis of SLE, painful lesions, photosensitivity and presence of facial lesion(s).

Conclusion:

CLE has a significant impact on quality of life on majority of its sufferers. It is thus of great importance for clinicians to assess the quality of life of individual patients to ensure holistic approach so as to improve overall health of the patients.

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Abstract N°: 7422

Association of cutaneous leishmaniasis and discoid chronic lupus : A local immune reaction ?

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Introduction & Objectives:

Cutaneous Leishmaniasis (CL) is a protozoan disease caused by Leishmania spp. and is endemic in Morocco. CL could be associated with other diseases, especially immune system disorders representing a diagnostic challenge. Herein, we describe a Moroccan patient with CL associated with Discoid chronic lupus erythematosus on the same location .

Case Report

A 52-year-old patient from Fkih Ben Saleh (an endemic area for leishmaniasis in Morocco), with no medical history, presented with papuloverrucous lesions on the face evolving for 3 years. Clinical examination revealed a violaceous papuloverrucous plaque with an atrophic center on the nose resting on an infiltrated base, annular papular lesions on the mandibular and zygomatic regions of the left cheek, a small verrucous lesion on the right cheek, and finally squamous papular lesions on the concha of the left ear. Dermoscopic examination showed erythema, starry white patterns at the periphery, lupoma grains, hyperkeratosis with a yellowish tear-like appearance, and telangiectasias. PCR identified a *L infantum* species and the patient was treated with intramuscular Glucantime. Given the lack of improvement of lesions of the lesions , a skin biopsy was performed and was consistent with a discoid cutaneous chronic lupus. There was no systemic involvement of lupus .Treatment with hydroxychloroquine resulted in a favorable outcome after 12 months of follow-up.

Discussion:

Our patient illustrate a rare case of CL occurring on the same location of Discoid lupus. This isotopic response suggest a local immune dysregulation involving the activation of type-1 interferon . Dermatologists must be aware of this phenomenon in order to establish an early and appropriate treatment.

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Abstract N°: 7492

Atorvastatin-induced pemphigus with Lyell's syndrome-like presentation

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Introduction & Objectives:

Pemphigus is an autoimmune intraepidermal and/or intraepithelial bullous disease characterized by autoantibodies that typically target desmoglein 1 and/or desmoglein 3. Induced pemphigus is a rare form. We report here a case of pemphigus vulgaris mimicking Lyell's syndrome, induced by atorvastatin.

Case Report:

An 82-year-old woman with a history of pemphigus herpetiformis in remission for 3 years presented with an acute bullous skin eruption evolving over one week. History revealed irregular intake of atorvastatin for 2 months as background treatment for an ischemic stroke. Physical examination revealed flaccid blisters and post-bullous erosions, extensive areas of skin detachment on the trunk and limbs involving 50% of the body surface area with a positive Nikolsky sign and a wet sheet appearance in some areas. Erosions of the oral and nasal mucosa were noted. Skin biopsy showed acantholysis with suprabasal detachment associated with some necrotic keratinocytes, while direct immunofluorescence demonstrated IgG deposition between keratinocytes. Atorvastatin was discontinued. The patient was started on high-dose systemic corticosteroid therapy. The clinical course was marked by the extension of skin detachments and the development of septic shock leading to the patient's death.**

Discussion:

Iatrogenic pemphigus is a specific form of pemphigus that can be triggered, exacerbated, or rarely induced by drug intake. Typically, it presents as erythematous or foliaceous pemphigus, less commonly as vulgar pemphigus. Mucosal involvement is rare. The main drugs implicated usually possess a thiol group, disulfide bridges, or a sulfur-containing ring. Increasingly, other medications are being reported to induce pemphigus. To our knowledge, only one case of atorvastatin-induced erythematous pemphigus has been reported in the literature. The precise mechanism of autoimmune reactions triggered by statins is not fully elucidated. Some authors suggest that statins, as proapoptotic agents, may release nuclear antigens into circulation, which could lead to the production of pathogenic autoantibodies. Our case is also notable for its clinical presentation, resembling Lyell's syndrome. This similarity has been reported predominantly in the context of paraneoplastic pemphigus.

Conclusion:

It is important to consider drugs as a possible triggering cause in patients newly diagnosed with pemphigus, as well as in those in whom the disease reappears after a long period of remission, especially in the elderly.

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Abstract N°: 7493

childhood scleromyositis : a case report

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Introduction & Objectives: Scleromyositis defines a syndrome where two distinct autoimmune diseases, systemic scleroderma and either polymyositis or dermatomyositis overlap. It is a common overlap syndrome found in adults, but rarely reported in childhood. We report a case of a pediatric scleromyositis.Haut du formulaire

Materials & Methods: A 13year- old young girl with no medical history, presented with 7 months history of skin sclerosis starting at the extremities, then extending to the trunk, roots of the limbs and the face, muscle weakness associated with generalized aches and pain , also claimed a dyspnea with minimal effort , a solid state dysphagia leading to weight loss and several episodes of false alimentary route .The physical examination revealed diffuse sclerosis resulting in sclerodactyly of the hands, puffy fingers where the fingers are locked in semi-flexed position. She had difficulties opening her mouth, flexion of the elbows and knees, also a poikiloderma of the back. She denied any history of Raynaud's syndrome. Blood tests results showed elevated levels of liver enzymes (ALT) and muscle markers (CPK), as well as an increase in LDH. Immunological test results reveal the presence of high-titer anti-ANA antibodies (1/1600) and a positive result for anti-PM-Scl antibodies(100), indicating possible connective tissue disease or autoimmune disorder. Electromyography (EMG) showed a myogenic pattern. Thoracic computed tomography (CT) reveals early diffuse interstitial lung disease . Functional exploration of the respiratory apparatus showed severe restrictive ventilatory deficit, severe damage to the alveolar-capillary membrane and reduced efficiency of bronchial drainage. Skin biopsy results show histological features compatible with scleroderma. Indeed, in the presence of cutaneous scleroderma confirmed by biopsy, as well as generalized muscle involvement confirmed by EMG associated with increase in muscle enzymes the most probable initial diagnosis was scleromyositis. She received a bolus of Solu-Medrol for 3 days followed by general corticosteroid therapy at a dose of 1 mg/kg/day.

Results: Scleromyositis is the most common of the overlap syndromes, combining two connective tissue diseases, namely systemic sclerosis and myositis (dermatomyositis or polymyositis). It typically occurs in adults, but some cases have been reported in childhood. The most frequently observed symptoms are specific to each autoimmune disease. The biological marker of this disease is an antibody directly targeted against the nuclear antigen PM-Scl. Also, it is usually used as a prognostic marker . The course of the disease is generally slow and tends to be benign, with corticosteroid therapy good response. In the pediatric form of scleromyositis, we find that myalgia usually presents first, followed by cutaneous sclerosis which is sometimes more significant, even severe. Visceral involvement seems to be moderate, sometimes absent, with pulmonary involvement being the most feared, followed by digestive complications. The diagnosis of scleromyositis is not always evident due to its rarity and variable presentation, and also due to the possible negativity of its main biological marker. However, this patient exhibits a more pronounced systemic involvement and no history suggesting Raynaud's syndrome accompanying systemic sclerosis.

Conclusion:

Our case illustrates a rare presentation of pediatric scleromyositis with a particular severe clinical presentation.

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Abstract N°: 7513

Five-year retrospective study of cutaneous lupus erythematous in a tertiary hospital in Portugal

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Introduction & Objectives:

Lupus erythematous is a chronic, relapsing condition that can affect all organ systems. Its pathogenesis is based on the chronic inflammation induced by immune complex production. Despite the advancing understanding of the disease, it is still unclear whether there is a relation between laboratorial and clinical findings in cutaneous lupus erythematous (CLE). The aim of this study was to analyse demographical, clinical and laboratorial characteristics of the different subtypes of CLE: acute, subacute and chronic.

Materials & Methods:

A retrospective observational study was conducted from all medical records referred to our Dermatology Clinic in a tertiary Hospital in Portugal, with histologically confirmed CLE, between January 1st 2019 and December 31st 2023.

Results:

A total of 103 patients were included, with an average age of 48.8 years (89-12) at diagnosis and a higher proportion of female patients 75.7%. Systemic Lupus International Collaborating Clinics' 2012 diagnostic criteria for systemic lupus erythematous (SLE) were met by 34.9% (n=36) of patients.

Among the patients with concomitant SLE, the most common CLE variant was chronic CLE, which was observed in 58.3% (n=21) cases, followed by subacute CLE, found in 30.6% (n=11) patients, and acute CLE in 11.1% (n=4) patients. Within the chronic CLE variant, discoid subtype was the most common (57.1%), followed by *lupus tumidus* (23.8%), lupus panniculitis (14.3%) and chilblain lupus (4.8%). From the group of patients who didn't meet SLE criteria, 85.1% (n=57) had chronic CLE (among these: 59.6% discoid, 36.8% *lupus tumidus*, 3.6% lupus panniculitis) and 14.9% (n=10) had subacute CLE.

No statistically significant differences were found between the two groups regarding laboratorial abnormalities (including cytopenias and inflammatory markers); although these were more frequently observed in chronic CLE forms. A similar pattern was observed for involvement of other organs (such as the kidneys, brain, serosae, among others), which all occurred more frequently in patients with CLE. All patients with simultaneous SLE had positive antinuclear antibodies and 55.5% (n=20) had positive anti-dsDNA. Whereas in the non-SLE group only 38.8% (n=26) had positive antinuclear antibodies and 1.5% (n=1) had positive anti-dsDNA antibodies. The difference between antibody positivity for the two groups was statistically significant ($p<0.05$).

Conclusion:

Although LE manifestations are very heterogeneous, cutaneous findings can be the key for early diagnosis. We provide data on how cutaneous involvement without SLE criteria and minimal laboratorial abnormalities is highly prevalent, reinforcing the need for a high clinical suspicion for diagnosis, ensuing a timely LE recognition and treatment. Interestingly, our data doesn't match what is usually found in literature, which could be explained by the fact that all patient data was gathered through the database of the Dermatopathology Laboratory.

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Abstract N°: 7524

Pruritic bullous dermatosis during pregnancy : About two cases

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Introduction & Objectives:

Pemphigoid gestationis is a rare supepidermal blistering dermatosis specific to pregnancy.

Materials & Methods:

We report a description of two cases of pemphigoid gestationis.

Results:

Observation 1:

A 47 years old multiparous patient hospitalized in the dermatology department for the management of a pruritic blistering dermatosis that developed during the third trimester of her fourth pregnancy. Skin examination revealed multiple erythematous urticarial plaques with polycyclic borders on the thighs, intergluteal region, legs, feet, arms, and face. Erythematous plaques with circinate and pseudo-circinate patterns were observed on the buttocks, thighs, and upper back. These lesions were surmounted by erosions, hemorrhagic crusts, vesicles and serous tense bullae; some of them had hemorrhagic content with a negative Nikolsky sign, involving an estimated skin surface area of 24%. Some vesicles were clustered in groups on the anterior surface of the left leg and on the lower back. A pigmented plaque with polycyclic borders and edematous erythematous margins, was present on the trunk, peri-umbilical region and lower back surmounted by erosions and hemorrhagic crusts. The palmo-plantar region was affected with a dyshidrotic appearance, while the oral and genital mucosa were spared. The histopathological examination revealed

an extensive subepidermal blister. Direct immunofluorescence was positive, showing linear staining of the dermo-epidermal junction with IgG, C3, and C1q. The diagnosis of pemphigoid gestationis was established and the patient was treated with high-potency topical corticosteroids applied daily. After 15 days, pruritus resolved, no new blisters appeared, and the lesions began to heal. Treatment was continued daily for another two weeks before tapering off.

Observation 2:

A 37 years old multiparous patient hospitalized in the dermatology department for the management of a pruritic blistering dermatosis that developed two months after her last pregnancy. Skin examination revealed multiple erythematous-violaceous plaques with polycyclic borders on the thighs, legs, feet, arms, forearms, abdomen, back and buttocks. These lesions were surmounted in some areas by erosions, hemorrhagic crusts, vesicles and serous tense bullae with a negative Nikolsky sign, involving an estimated skin surface area of 10 %. The palmo-plantar region was affected with a dyshidrotic appearance, while the oral and genital mucosa were spared. The histopathological examination revealed an extensive subepidermal blister. Direct immunofluorescence was positive, showing linear staining of the dermo-epidermal junction with IgG, C3, and C1q. The diagnosis of pemphigoid gestationis was established and the patient was treated with oral corticosteroids. After 16 days, pruritus resolved, no new blisters appeared, and the lesions began to heal. Treatment was continued for another

two weeks before tapering off.

Conclusion:

Pemphigoid gestationis is a rare autoimmune dermatosis that typically occurs in multiparous women during the second or third trimester of pregnancy, or in the immediate postpartum period. Diagnosis is confirmed by biopsy with direct immunofluorescence. There is no consensus on therapeutic strategy it commonly involves topical or systemic corticosteroid therapy, depending on severity.

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Abstract N°: 7550

combination of oculocutaneous albinism, systemic lupus and psoriasis in a single patient : case report

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Introduction & Objectives:

Albinism is a group of inherited conditions associated with an abnormality in the biosynthesis of melanin, a pigment produced in specialised cells of the skin, hair, iris, pigmented epithelium of the retina and inner ear, associated with a normal number and structure of melanocytes. The reduction in pigment is responsible for increased sensitivity to UV radiation and a predisposition to skin cancer. Oculocutaneous albinism (OCA) involves generalised cutaneous hypopigmentation and ophthalmological involvement.

Systemic lupus (SL) or acute systemic lupus erythematosus (ASLE) is a syndrome characterised by variable clinical manifestations of varying duration, associated in different ways from one patient to another, and multiple biological abnormalities. It is a long-term (chronic) disease of immunological origin, the exact causes of which are still unknown.

Materials & Methods:

We report here an extraordinary case of combination of oculocutaneous albinism, systemic lupus and psoriasis in a single patient.

Results:

The patient was 18 years old and presented with oculocutaneous albinism with a generalised decrease in pigmentation of the hair, skin and eyes on examination and nystagmus, decreased visual acuity and photophobia on ophthalmological examination. At the age of 14 years, the patient was diagnosed with systemic lupus with neurological, cutaneous and articular involvement, and recently presented with small pruritic, scaly, erythematous plaques on the upper limbs with Auspitz sign or positive bloody dew sign.

A skin biopsy was performed, which was consistent with psoriasis.

The patient had already been treated for systemic lupus with imurel for 4 years and for psoriasis with dermocoticoids with degression and UVB phytotherapy.

Conclusion:

The complex clinical presentation of this combination of conditions requires careful assessment and integrated management to optimise clinical outcomes and patient quality of life. Further studies are needed to better understand the underlying mechanisms of this association and to guide more effective therapeutic strategies.

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Abstract N°: 7551

A Systematic Review of Case Series and Clinical Trials Investigating Regenerative Medicine for the Treatment of Vitiligo

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Introduction & Objectives:

The aim of this study is to examine the efficacy and safety of various regenerative medicine treatments, such as cell therapy, platelet-rich plasma (PRP), plasma-poor platelet (PPP), plasma-rich fibrin (PRF), mesenchymal stem cells, stromal vascular fraction (SVF), exosomes, adipose-derived stem cells (ADSC), and stem cell-conditioned media (SC-CM), for treating vitiligo.

Materials & Methods:

We conducted a thorough search of major databases such as Pubmed, Scopus, and Web of Science, and selected 48 articles based on specific criteria. We used EndNote X8 and Google Sheets to review and extract data from the articles. After analyzing the studies, we categorized them accordingly.

Result:

Our study has found that all 48 studies (100%) and 96 intervention groups included have demonstrated the effectiveness of regenerative medicine treatment. The majority of studies conducted in the field of regenerative medicine for the treatment of vitiligo have focused on melanocyte-keratinocyte transplant with epidermal origin. The next treatments included platelet-rich plasma injection, hair follicle-derived cell therapy, transplantation of isolated melanocytes, and melanocyte-keratinocyte transplantation in the form of cell suspension in PRP, respectively. The highest rate of response to treatment in the intervention groups with melanocyte-keratinocyte transplantation of epidermal origin was observed in 56% of patients, with “repigmentation above 90%” being the achieved outcome. Among the groups treated with platelet-rich plasma (PRP), the highest response rate was 58.7% repigmentation. For the groups treated with hair follicle-derived cell therapy, the percentage was 80.15%±22.9 %, while for the groups treated with melanocyte-keratinocyte suspension in PRP, it was 75.6 ± 30%. In another study, complete repigmentation was observed in 50% of patients treated with this suspension. The highest percentage of repigmentation in the groups treated with isolated melanocyte transplantation of epidermal origin was more than 90% repigmentation in 83.33% of patients. For patients with isolated melanocyte transplantation with hair follicle origin, 43.33% of them reached this level of repigmentation.

Conclusion:

We have concluded that regenerative medicine plays an effective role in the treatment of vitiligo lesions. Furthermore, this treatment method is safe and does not cause serious complications. It can be used alone or in combination with other methods for treating vitiligo. To advance the treatment of vitiligo, we recommend conducting clinical trials on the unexplored branches of regenerative medicine.

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Abstract N°: 7663

Ocular pemphigus: a rare clinical variant without skin involvement

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Introduction & Objectives:

Pemphigus is an autoimmune disease that is characterized by the formation of antibodies against desmogleins, specifically type 1 and 3, which, as is known, are an essential part of the intercellular bridges that allow adequate adhesion, resulting in a barrier effect. So when adhesions are destroyed, blisters would form in the tissues where these proteins are found. We can find this type of proteins at the epidermal level, and at the level of mucous membranes and conjunctival tissue (predominantly type 3), with more than 97% of idiopathic etiology.

For its diagnosis, 2 biopsies are indicated, one for H&E and one for immunofluorescence, with the purpose of achieving the detection of epidermal alterations such as the level of the blister and the type of cellular infiltrate, and more importantly the demonstration of the antibodies attached to these proteins.

Materials & Methods:

We present the case of a 75-year-old female patient, who reports having had a diagnosis of scar entropion since 2015, requiring multiple surgeries which were ineffective as she presented a recurrence with a subsequent decrease in visual acuity. She decided in October. 2023 to go to our Hospital for an evaluation by the Ophthalmology service who decided to send to our Dermatology service for evaluation and support in the diagnostic approach of periocular erythema and conjunctival erosions with telangiectasias, with poor response to moisturizers and multiple ocular antibiotics; provisionally, an ocular rosacea was provisionally diagnosed. However, no suggestive data were found at the cutaneous level, both at the nasal and malar levels. A test was also performed in search of Demodex, which was negative. For this reason, our service makes the suggestion of performing a conjunctival biopsy by Ophthalmology, seeking to take a portion of the scar patches in the conjunctiva and concomitantly with a "normal" portion for histopathological study.

When the results of the H&E biopsy were obtained, no presence of superficial blisters was observed, with little lymphocytic infiltrate, only significant fibrosis was evident.

Results:

Through immunofluorescence, the presence of IgG and C3c antibodies against desmoglein 3 were detected giving an image in network pattern, allowing the diagnosis of ocular pemphigus to be established, so multidisciplinary management between dermatology and ophthalmology was initiated, based on prednisone and azathioprine, with the purpose of stopping the progression of the disease and reducing the risk of total corneal scarring.

Conclusion:

It is a peculiar case, due to the condition limited only to the conjunctival tissue, where only 3 cases have been reported until 2014. It is well known that within the spectrum of pemphigus vulgaris, ocular involvement can be found in 7 - 26% of patients, but it is unlikely that only the eyes are affected, without any other skin symptoms.

The case is relevant due to the limited condition with very few reported cases worldwide, which can sometimes be difficult to diagnose, requiring a diagnostic approach in conjunction with other specialties and knowing the multiple clinical variants found, as well as the challenge of not having first-line drugs such as rituximab due to high costs, requiring more easily accessible therapeutic management.

Currently, the patient is stable without new ocular lesions, with progressive reduction of prednisone and it is expected that azathioprine will be withdrawn soon.

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Abstract N°: 7683

"Clinical-epidemiological characteristics and description of the dermatopathological findings of pediatric patients with discoid lupus erythematosus¨

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Introduction & Objectives:

General

• To evaluate the clinical and epidemiological characteristics and describe the dermatopathological findings of pediatric patients with DLE.

Specific

• Calculate the incidence of DLE in pediatric patients, Identify the most affected genus of DLE in the pediatric population, Describe the time of evolution, topography, morphology, symptoms, histopathological findings.

Materials & Methods:

Retrospective, observational, descriptive study of a public hospital in the west of the country. All dermatology files and dermatopathology laboratory records with a diagnosis of lupus. "Time between January 1, 2011 and December 31, 2020." Descriptive statistics and measures of central tendency, frequency, mean. kappa index, by calculating the proportion of the histopathological variables.

Results:

35 patients were included for the study. The incidence of DLE in the pediatric population in the west of a public institution in the country was 0.32%, 32 cases per 100,000 inhabitants.

The mean age at diagnosis was 11.91 +/- 0.67 years, median 12 years.

The female sex was the most affected in 31%.

The median age at diagnosis was 13.3 months +/- 2.57 months.

The most frequently reported symptom was pruritus in 37%.

The diagnosis sent for histopathological study was DLE in 62%.

The most affected segment was the head in 100% and the most affected region of this was the cheeks in 28.6%.

The most frequent morphology found was plaque in 85.8%.

100% of the patients presented epidermal and dermal involvement in the histopathological analysis. The most observed epidermal finding was vacuolization of the basal layer (94.2%); and the dermal finding, the superficial lymphocytic infiltrate (94.2%).

At least one criterion proposed by Elman for the classification of DLE was present in 94.3% (n=33) of the patients8. The most frequent clinical criterion proposed by Elman was the presence of erythema 88.5% (n=31).

Conclusion:

In general, the findings were different between the adult population studied by Elman and the pediatric population of the IDJ.8 Vacuolization of the basal layer (kappa coefficient 0.83) could be the main diagnostic indicator of DLE in the pediatric population with a strength of agreement almost perfect. The thickening of the basal layer and the presence of follicular plugs could be the major histopathological criteria for the diagnosis of DLE in children and the presence of perivascular lymphocytic infiltrate, mucin deposition and thickening of the basement membrane, minor criteria. The histopathological diagnosis of DLE in the pediatric population is those patients who have the major criterion and 1 or 2 minor criteria.

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Abstract N°: 8058

Allogeneic CD19-targeted CAR-T therapy for refractory systemic lupus erythematosus (SLE)

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Introduction & Objectives:

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease associated with significant morbidity and mortality. It can affect various tissues and organs throughout the body, leading to a broad spectrum of clinical symptoms. The treatment approach for SLE primarily involves immunomodulation and immunosuppression. However, these drugs have considerable toxicity and are not effective in all patients. Conceptually, a deep depletion of CD19+ B cells and plasmablasts could trigger an immune reset in autoimmune diseases. Autologous CD19-targeted CAR-T therapies has been explored in several autoimmune diseases and reported promising efficacy in recent studies. In this study, a healthy donor-derived, multiplex genome-edited allogeneic CD19-targeted CAR-T product (BRL-303) was developed for refractory autoimmune diseases patients. Here we report the efficacy and safety profile of BRL-303 in treating patients with systemic lupus erythematosus (SLE).

Materials & Methods:

This is an investigator-initiated trial evaluated the safety and efficacy of BRL-303 in adult patients with refractory autoimmune diseases (NCT05859997, NCT05988216). Participants were screened and underwent lymphodepletion chemotherapy with cyclophosphamide (300 mg/m^2, day -5 to -4) and fludarabine (25 mg/m^2, day -5 to -3), then BRL-303 infusion at day 0 with a dose level of 1×106 cells/kg. After BRL-303 infusion, each patient underwent safety and systematic follow-up assessments according to the protocol.

Results:

As of August 16, 2024, a total of 6 patients with refractory systemic lupus erythematosus have been treated. These patients were unable to control disease progression despite treatment with corticosteroids and various immunosuppressants. After BRL-303 infusion, CART cells expanded in vivo in all subjects significantly within 7 days and reached a peak around 14-21 days. With expansion of BRL-303, B cells in peripheral blood of patients reached deep and persistent depletion about 1-3 months. The median follow-up time for the 6 subjects was 6 months (range: 1-9 months). After BRL-303 infusion, there was a significant decrease in SLEDAI scores and PGA in all subjects(6/6,100%), and all of them achieved SRI-4 remission. Moreover, all subjects showed a significant decrease in serum autoantibodies and increase in complement. Meanwhile, proteinuria significantly decreased in the subjects with lupus nephritis. In terms of safety, the patient's vital signs remained mostly stable throughout the monitoring period, 2 subjects developed grade 1 CRS on D4 and D5, which recovered soon without tocilizumab and steroid usage. No serious infection (Grade 3/4), no ICANS and GvHD observed in all subjects.

Conclusion:

Overall, SLE subjects treated with BRL-303 achieved significant clinical remission, with a controllable safety profile, offering a potential paradigm shift for SLE patients who are refractory to currently available treatments. Further observation of SLE and more autoimmune disorders treated with BRL-303 are ongoing in our study.

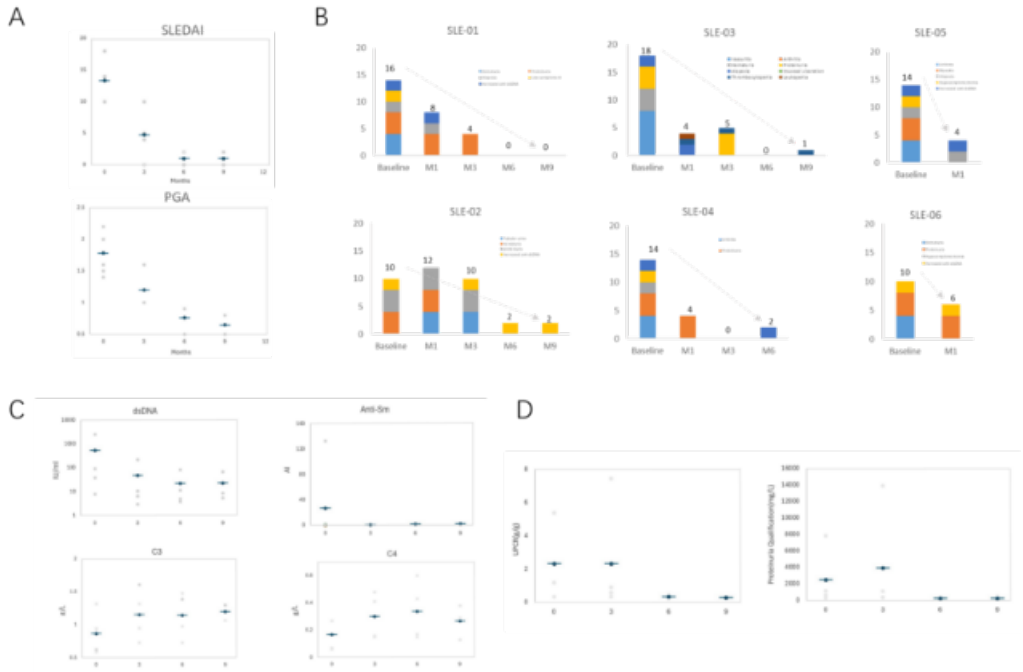

Figure 1 Effects of BRL-303 for SLE patient (A, SLEDAI-2K and PGA change befor and after BRL-303 treatment; B, SLEDAI-2K Score changs; C,Serological changes ; D, Urine protein changes)