

Abstract N°: 145**Omalizumab as a corticosteroid-sparing agent in the treatment of bullous pemphigoid**

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Introduction & Objectives: Bullous pemphigoid is the most common bullous autoimmune disease, characterized by the presence of autoantibodies directed against the hemidesmosomal proteins BP180 and BP230. Treatment is based on the use of topical and systemic corticosteroids and other immunosuppressive drugs. As this pathology typically affects elderly subjects, the presence of comorbidities can often complicate the therapeutic management. The objectives of the study were to evaluate the efficacy of omalizumab in disease control, allowing to reduce the dose of systemic corticosteroids administered to patients, as well as evaluate the effects of omalizumab on antibody titres against BP180 and BP230, IgE and eosinophils.

Materials & Methods: We retrospectively evaluated all the patients receiving treatment for BP in the Autoimmune Bullous Disease Department of the Dermatology Clinic, University of Pavia, IRCCS Policlinico San Matteo Foundation; among 222 subjects evaluated for BP in the last 10 years, we selected five patients who presented with corticosteroid-dependent BP, with a contraindication to the use of other immunosuppressive treatments and who could instead benefit from the use of a safer and selectively-acting agent like omalizumab.

Results: All the patients tolerated omalizumab without side-effects. The mean duration of treatment was 9.2 months (median: 8 months, range: 12 months), while follow-up lasted a mean of 17 months (median: 12 months, range: 28 months). It was possible to obtain a reduction in the dosage of systemic corticosteroids in 100% of the treated patients, furthermore a reduction in circulating IgE titres was observed in 40%, a reduction in antibody titres against BP180 and BP230 in 60% and of eosinophils in 80% of cases.

Conclusion:

In conclusion, we found that in 100% of cases it was possible to administer lower doses of steroids while controlling the disease, confirming the utility of omalizumab as an adjuvant therapy. These results should be seen in light of some limitations. First, the dosing protocol chosen for the administration of omalizumab was that indicated for chronic idiopathic urticaria. Second, all the patients included in our study had a long-standing history of BP and had previously received various other treatments before beginning omalizumab. Therefore, we believe that further studies evaluating the effects of this monoclonal antibody should be carried out.

Abstract N°: 200**A case of Unilateral Pemphigus Foliaceus**

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Introduction & Objectives:**Materials & Methods:****Results:****A Case of Unilateral Pemphigus Foliaceus**

Pemphigus Foliaceus (PF) is a rare autoimmune blistering disorder that typically presents with crusted scaly erosions in a seborrheic distribution. This is mediated by autoantibodies directed against desmoglein-1. We report an unusual presentation of this rare disease, a case of unilateral PF.

A 90-year-old presented with a 3-month history of painful erosions. The lesions were limited to the right side. There was no mucosal involvement. He had multiple comorbidities which included hypertension, gout, severe spinal stenosis at multiple cervical levels for conservative care and colon cancer with palliative stenting done.

Clinical examination revealed unilateral erythematous scaly plaques with flaccid blisters and superficial erosions on the face, upper limb, trunk and buttock. Nikolsky's sign was positive.

A skin biopsy was performed and histology demonstrated subcorneal acantholysis. Direct immunofluorescence showed intra-epidermal intercellular IgG and C3 depositions. Wound swab for herpes polymerase chain reaction (PCR) was negative for Herpes simplex virus 1, 2 and Varicella zoster virus. Intercellular epithelial staining on monkey oesophagus was demonstrated by indirect immunofluorescence. The clinical and pathological findings were consistent with a diagnosis of pemphigus foliaceus (PF).

The patient was started on prednisolone 20mg per day (~0.5mg/kg/day) and good clinical response was noted.

Unilateral PF is a rare entity. To the best of our knowledge, this is the first report on unilateral PF involving trunk and limbs. Literature review yielded only two cases of unilateral PF, both with lesions limited to the face. Previous reported causes for localized involvement of pemphigus include the prior application of imiquimod and ultraviolet radiation (UVR) exposure. Cases of unilateral bullous pemphigoid over the hemiplegic side due to cerebrovascular accidents have been described, and the concept of an immunocompromised district was introduced. Our patient did not have a history of stroke, however, he did have long standing spinal stenosis at multiple cervical levels. MRI C-spine done in 2013 showed severe degenerative changes at all cervical levels down to T1/2 level. There was severe stenosis of the neural foramina at multiple levels. This might result in an immunocompromised district which contributed to the unilateral distribution of PF in our patient.

In conclusion, a high index of suspicion is needed to diagnose rare conditions such as PF. Patients may present atypically with unilateral erosions, or over sun-exposed areas.

Conclusion:



Abstract N°: 229**Thyroid auto-antibodies and hormones in Egyptian vitiligo patients before and after systemic steroids versus NB-UVB treatment**

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Introduction & Objectives:

Vitiligo is strongly associated with a number of autoimmune disorders, including alopecia areata, diabetes mellitus, pernicious anemia, Addison's disease and Hashimoto's thyroiditis. Thyroid functional and autoimmune disorders have been reported in association with vitiligo, and it seems that the incidence of clinical and subclinical thyroid involvement is more common in vitiligo patients than healthy subjects. Our aim was to evaluate thyroid auto antibodies and hormones levels, in comparison to normal controls before and after treatment with systemic steroids and NB-UVB in Egyptian patients with non- segmental vitiligo.

Materials & Methods:

This case control study was carried out on 40 patients, with non- segmental vitiligo 20 patients received NB-UVB therapy 3 times weekly for 3 months, 20 patients received oral mini pulse of systemic steroids for 3 months and 20 age and sex matched volunteers as controls. Clinical diagnosis of vitiligo, vitiligo area scoring index (VASI score), Dermoscopic examination and photographic documentation were done before and after treatment. Blood samples were collected from all participants to estimate levels of TSH, T3, T4, thyroid auto antibodies (Anti-TPO and Anti-TG) and ANA before and after treatments in patients and once from controls using the ELISA technique.

Results:

Levels of TSH, Anti-TPO and Anti-TG were significantly elevated in vitiligo patients compared to controls ($p < 0.05$). After NB-UVB treatment there was no significant difference between group 1 and controls as regards TSH, Anti-TPO, and Anti-TG ($p \text{ value} > 0.05$). After steroids treatment there was no significant difference between group 2 and controls as regards Anti-TPO ($p \text{ value} > 0.05$), with significant difference as regards TSH and Anti-TG ($p \text{ value} < 0.01$).

Levels of TSH, Anti-TPO and Anti-TG (figure-1), showed highly significant decrease after NB-UVB treatment ($p < 0.01$) in group. After steroids treatment Anti-TG and TSH significantly decreased ($p < 0.05$) and Anti-TPO insignificantly changed in group 2 (figure-2).

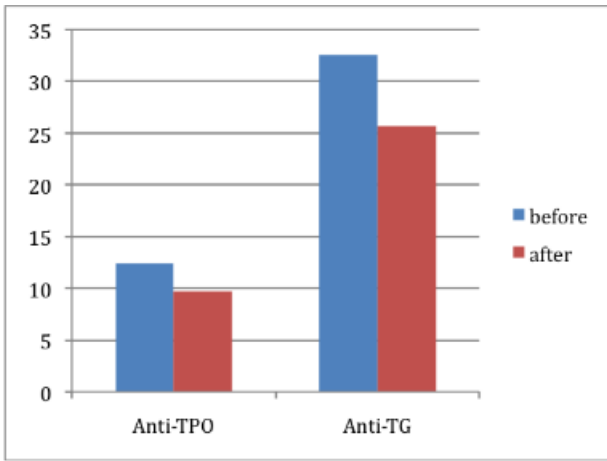


Figure 1: Comparison between anti-TPO and anti-TG before and after NB-UVB treatment.

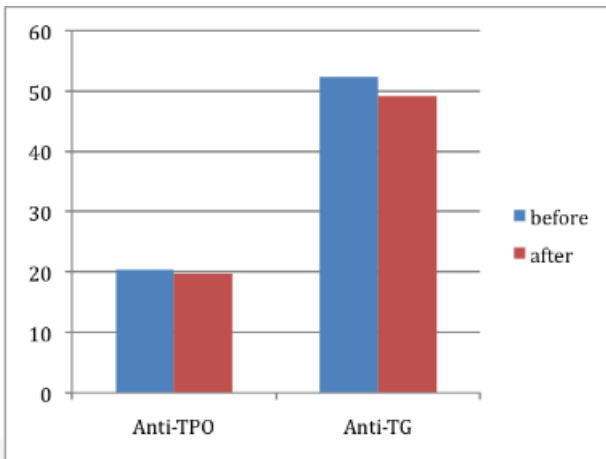


Figure 2: Comparison between anti-TPO and anti-TG before and after Steroids treatment.

Anti-TPO and Anti-TG were significantly higher in group 2 than group 1 (figure-3), however, TSH was significantly lower (figure-4) in group 2 than group 1 ($p < 0.05$). On comparing VASI score of patients before and after treatment in both groups, a statistically highly significant difference was observed ($p < 0.01$).

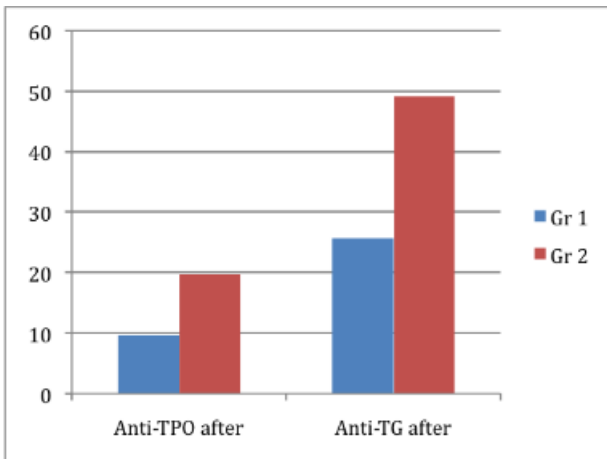


Figure-3: Comparison between group 1 and 2 as regards anti-TPO and anti-TG.

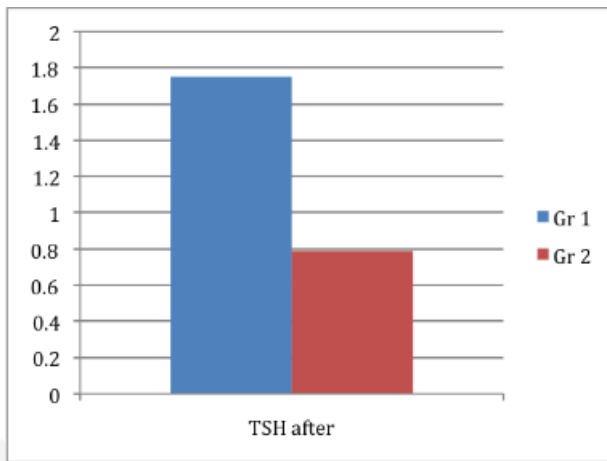


Figure-4: Comparison between group 1 and 2 as regards TSH.

Conclusion:

The rationale of the current study was evaluating thyroid auto antibodies and hormones in patients with non-segmental vitiligo before and after two main immunomodulatory therapeutic modalities; NB-UVB and systemic steroids. We supposed that vitiligo is commonly associated with autoimmune disorders and thyroid diseases. The results demonstrated significantly high levels of thyroid autoantibodies and TSH in vitiligo patients when compared to healthy controls. After treatment with NB-UVB and systemic steroids this difference was decreased more notably in NB-UVB group which suggests that NB-UVB is a safe effective treatment of vitiligo decreasing the risk of autoimmune thyroid diseases.



Abstract N°: 331**Clinicopathological Characteristics and Health-Related Quality of Life in Patients with Autoimmune Blistering Diseases at the Rizal Medical Center: A Cross-Sectional Study from 2017-2022**Dana Andrea Nery*¹, Jacqueline Michelle Melendres², Mae Ramirez-Quizon²¹Rizal Medical Center, Dermatology, Pasig, Philippines, ²Rizal Medical Center, Pasig, Philippines**Introduction & Objectives:**

Autoimmune blistering diseases (AIBDs) are a group of diverse disorders characterized by blister formation due to pathogenic autoantibodies directed against structural proteins of the skin and mucous membranes. Due to its relative rarity, there are limited studies on the prevalence, demographic and clinicopathological spectrum of various autoimmune bullous diseases from the Philippines.

The study aimed to determine the prevalence of autoimmune blistering diseases and their correlation to sociodemographic characteristics, clinicopathologic features and health-related quality of life (QOL) prior to and after treatment in Filipinos based in a tertiary hospital in the Philippines. Additionally, the study sought to determine the relationship between disease severity scores and health-related quality of life (HRQOL) measures.

Materials & Methods:

A total of 53 patients diagnosed and managed with AIBD at the Rizal Medical Center's Department of dermatology from 2017 to August 2022 were included in this retrospective cross-sectional study. Chart review was conducted to determine the sociodemographic characteristics, clinicopathologic features, and health-related quality of life scores. The diseases severity scores: Autoimmune Bullous Skin Disorder Intensity Score, Bullous Pemphigoid Disease Area Index, Pemphigus Disease Area Index (ABSIS/BPDAI/PDAI) in relation to Autoimmune Bullous Disease and the Treatment of Autoimmune Bullous Disease quality of life scores (ABQOL/TABQOL) were compared to determine if there was a correlation.

Results:

Out of 53 cases of AIBDs seen, there was a female preponderance of 35 (66.04%) in the study group. The age ranged from 2-91 years of age with most cases belonging to the > 60 years age group. Bullous pemphigoid (BP) was the most common disease (54.72%), followed by pemphigus foliaceus (PF) and pemphigus vulgaris (PV) (16.98%) (15.09%), respectively. Most of the histopathologic findings obtained from each patient corroborated with their clinical impression. Further confirmation was done through direct immunofluorescence (DIF) and enzyme-linked immunofluorescence antigen (ELISA). This study noted moderate to weak correlations between pre-treatment disease severity indices and ABQOL. This was exhibited by a positive monotonic association between (ABSIS) and (BPDAI) scores with (ABQOL).

Conclusion:

The study findings are consistent with the existing literature on the clinicopathologic profile of AIBD in other populations, suggesting that trends among the Filipino population are in line with population trends in other regions of the world. This study demonstrated that ABQOL was moderately correlated with disease severity indices, although different components of these indices showed varied responses to HRQOL indices. TABQOL was not very sensitive to treatment and did not correlate with disease severity indices.

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Abstract N°: 332

A Case Report on Erythroderma Secondary to Psoriasis and Pemphigus Foliaceus in a 57-Year-Old Filipino Female

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Introduction & Objectives:

Psoriasis and autoimmune blistering disorders are thought to be two distant diseases, although there have been a few cases of psoriasis associated with bullous disorders that have been reported worldwide. This report recognizes the possible challenges encountered during the diagnosis and management of this case with psoriasis and pemphigus.

Materials & Methods:

A 57-year-old Filipino female presented with a 1-year history of chronic erythematous plaques with eventual progression to erythroderma. Guided with a baseline skin biopsy of spongiotic dermatitis done at another institution, she was initially managed as a case of erythroderma secondary to atopic dermatitis. Treatment with systemic corticosteroids was initiated upon admission. However, the treatment course was complicated with the development of sepsis and multiple pustules on her trunk and extremities.

Serial skin punch biopsy and Serum enzyme-linked immunosorbent assay.

Results:

Laboratory findings Chest x-ray revealed bilateral opacities, suggestive of pneumonia. Complete blood count revealed normocytic, normochromic anemia with leukocytosis (hemoglobin 109, hematocrit 0.33, WBC 12.46). Liver enzymes were also elevated (ALT 65, AST 54). The patient was subsequently referred to ophthalmology and internal medicine for further evaluation. The patient was diagnosed with stage 2 hypertension; anemia secondary to chronic inflammation; corneal melt secondary to chronic inflammation and was managed accordingly.

Histopathologic findings revealed the overlying epidermis showed basketweave orthokeratosis, psoriasiform hyperplasia with diffuse epidermal spongiosis, hyper granulosis, marked papillary edema, thinning of the supra-papillary plates with underlying dilated blood vessels with mild to moderate superficial perivascular infiltrates of neutrophils, eosinophils, numerous plasma cells, and rare mast cells. This was suggestive of psoriasiform and spongiotic dermatitis.

Serial skin punch biopsies revealed psoriasiform hyperplasia with diffuse epidermal spongiosis and acantholysis in the stratum corneum. The finding of acantholysis prompted an investigation of pemphigus. Serum enzyme-linked immunosorbent assay detected elevated titers of anti-desmoglein-1 autoantibodies at > 200 RU/mL, normal value: (<20 RU/mL). Given these findings, a final diagnosis of erythroderma secondary to psoriasis and pemphigus foliaceus was made.

After completing treatment with systemic antibiotics, the patient was shifted back to cyclosporine (2.5mg/kg/day) with topical medications. (Clobetasol propionate ointment 0.05% with petroleum jelly). The patient was discharged with almost complete clearance of her lesions.

Conclusion:

This rare case elucidates a probable immunological link between pemphigus and psoriasis. Ultimately, this may aid in the better understanding and treatment of the concurrent entities.

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Abstract N°: 434

Pyoderma gangrenosum - atypical case

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Introduction & Objectives: The ulcerative variant is a classic manifestation of gangrenous pyoderma that occurs spontaneously or after trauma. Pyoderma gangrenosum appears suddenly and shows a predilection for the lower extremities, gluteus, abdomen, but it can appear on any part of the body.

Materials & Methods: The patient 20 years old, suffered multiple fractures as a motorcyclist. From the orthopedic-surgical point of view, he had several operations due to poly trauma. All the time he received prophylactic anticoagulant, antibiotic, analgesic therapy. An injury in the region of the right shoulder was treated with surgical debridement, which led to a worsening of the condition. The resulting ulceration increased very quickly and did not heal. On the right shoulder ulceration 12x15 cm, of irregular shape, with raised blue-gray edges. The bottom of the ulceration is covered with necrotic - purulent contents. The ulceration is surrounded by bright erythema and is extremely painful. From the laboratory findings, the patient maintained an accelerated SE of erythrocytes, in the blood count leukocytosis, the concentration of C-reactive protein was increased and followed the activity of the disease. Other laboratory findings, including hepatic and renal analyses, markers of viral hepatitis were within reference values.

Bacteriological smear of ulceration isolated: *Escherichia coli*. Histopathological finding of a skin section from the edge of an ulcer: signs of lymphocytic vasculitis. Chest X-ray et abdominal ultrasound not show any pathological changes. Consultative examinations: internist, infectologist and dermatologist. After all the examinations, the association of Pyoderma gangrenosum with other findings diseases was ruled out.

Results: Treatment and outcome of the disease: After the diagnosis, corticosteroid therapy was started in the patient. The initial dose was Methylprednisolone 80 mg/day, this dose was lowered by 10 mg per week in accordance with the improvement of the clinical picture up to a dose of 20 mg/day. Since then, the dose has been gradually reduced to a maintenance dose of 8 mg/day. Antibiotic therapy was carried out according to the ulcer swab (Meropenem, Vankomicin, Metroniazol), local treatment with antiseptic solutions and hydrocolloid dressings (dressing interval 3 days) until the formation of granulations, then topical corticosteroids under occlusion. After the therapy, complete epithelialization of the ulcer occurred in 60 days. The patient is still under regular dermatological control.

Conclusion: There is no definitive therapeutic regimen for Pyoderma gangrenosum. Our success is a quick diagnosis and therapeutically quick and favorable response. Pyoderma gangrenosum is difficult to diagnose and very often misdiagnosed.

The disease is diagnosed exclusively on the basis of the clinical picture and the course of the disease. Pyoderma gangrenosum should always be kept in mind in patients with painful, necrotic, rapidly spreading ulcerations. Pyoderma gangrenosum is a disease that cannot be defined by laboratory parameters. The pathohistological finding is not specific. Differential diagnosis should be distinguished from: other neutrophilic dermatoses, deep fungal infections, bacterial infections. It is important to detect and treat the underlying systemic disease if it is present, because the prognosis most often depends on it. A multidisciplinary approach is essential to achieve optimal treatment results.

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Abstract N°: 525

The efficacy and safety of Influenza vaccine in patients with pemphigus in the Covid-19 era: A randomized control trial

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Introduction & Objectives: There is still insufficient evidence about the efficacy and safety of the influenza vaccine on B-cell-mediated autoimmune diseases such as pemphigus. This study aims to assess the effects of the influenza vaccine on patients with pemphigus.

Materials & Methods: Sixty patients were randomly divided into vaccination and control groups. Patients were followed for 12 months, while flu symptoms (including fever, cough, rhinitis, and chest pain), flu PCR results, Anti-desmoglein 1, 3 antibodies, and PDAI (Pemphigus Disease Area Index) scores were evaluated at baseline and 6-month intervals.

Results: The cumulative and daily doses of prednisolone were not significantly different in the two groups. The incidence of flu symptoms in the vaccination group wasn't notably different from the controls. The recurrence rate of pemphigus wasn't significantly different between patients who received the flu vaccine and those who did not.

Conclusion: The results of this study violate the hypothesis and concern that influenza vaccination can lead to the recurrence of pemphigus.



Abstract N°: 605**Efficacy of rituximab in the treatment of epidermolysis bullosa acquisita**

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Introduction & Objectives

Epidermolysis bullosa acquisita (EBA) is a subepidermal bullous disorder characterised by skin fragility and mucosal erosions, mediated by antibodies that bind collagen VII. The condition is difficult to treat and often requires combination immunosuppressant therapy. Rituximab is a monoclonal anti-CD20 biologic therapy that has been reported in small case series to be effective in the treatment of recalcitrant EBA. In this case series we report the efficacy of rituximab in the treatment of 14 patients with severe EBA.**

Materials & Methods

Electronic records were used to identify all patients with a diagnosis of EBA who received rituximab in our centre from 2011-2021. EBA was defined as patients with skin fragility, positive direct immunofluorescence demonstrating linear IgG at the basement membrane with localisation to the base of salt split skin and a positive collagen VII ELISA. Clinic records for each patient's follow up visits over 600 days post treatment were then used to identify clinical response, collagen VII titre and prednisolone dosing. Complete remission (CR) was defined as the absence of new or established lesions on minimal therapy for 2 months, partial remission (PR) as transient lesions that heal within 1 week on minimal therapy and active disease (AD) as the development of new lesions. B cell quantification, immunoglobulin levels and reported adverse events related to rituximab were recorded for each follow up visit.

Results

14 patients were identified (male = 10 and female = 4) with a mean age of 58.1 years (age range 35-91 years old) and a mean disease duration of 12.8 years at the time of rituximab administration. All patients were on combination corticosteroid and immunosuppressant therapy, consisting of one or more of mycophenolate mofetil, intravenous immunoglobulin, azathioprine or dapsone. 4 patients also had positive BP180/230 antibodies. Rituximab induced CR in 6 of 14 patients who remained on low dose prednisolone and mycophenolate mofetil. PR was seen in 4 patients with AD in the remaining 4. A reduction in collagen VII titre of 78% was observed over the follow up period. The mean reduction in prednisolone dosing was 40%. Rituximab also allowed withdrawal of intravenous immunoglobulin therapy in 9 of 14 patients. No significant adverse events were reported.

Conclusion

Rituximab was well tolerated and resulted in clinical improvement in 71% of our patient cohort and enabled dose reduction of prednisolone and other immunomodulatory therapies in the majority of patients. This study represents the largest reported cohort of EBA patients treated with rituximab and supports its role in the management of this challenging disease.

Abstract N°: 626**Rituximab for the treatment of epidermolysis bullosa acquisita: a large case series**Shayan Dasdar^{*1}, Nika Kianfar¹, Hamidreza Mahmoudi¹, Kamran Balighi¹, Maryam Daneshpazhouh¹¹School of Medicine, Tehran University of Medical Sciences (TUMS), Tehran, Iran

Introduction & Objectives: Epidermolysis bullosa acquisita (EBA) is a rare bullous dermatosis triggered by IgG autoantibodies against collagen VII (COL7). Disease response to treatment is unsatisfactory, and finding the optimal treatment for EBA is still challenging. Rituximab (RTX) has shown potential as an alternative treatment, with some success reported in small numbers of cases. Herein, we report RTX's efficacy, safety, and treatment durability in a series of patients with EBA.

Materials & Methods: In this retrospective study, 15 patients with a diagnosis of EBA were included. The diagnosis was based on clinical and histopathological presentations, immunofluorescence on salt-split skin demonstrating linear floor deposition of IgG, and detection of anti-COL7 antibodies. RTX was administered based on two protocols: RTX 500 mg weekly for four weeks or RTX 1000 mg for two biweekly infusions. Due to the lack of a consensus statement on definitions of disease endpoints for EBA, definition outcome measures for mucous membrane pemphigoid were employed. The corticosteroid-sparing effect of RTX was investigated by recording prednisolone (PSL) dose tapering within 12 months of RTX therapy.

Results: The mean age of the patients at disease onset was 51.9 years. The median time from diagnosis to RTX administration was 15 months. Four patients received RTX as early treatment within six months of diagnosis. Eleven patients consumed PSL at RTX initiation, with a mean dose of 28.6 mg/d. Following the treatment with RTX, all patients showed clinical response initiated in a mean of 3.9 months. Disease control was obtained in 14 patients after the first cycle of RTX. Partial and complete remission occurred in four (26.7%) and five (33.3%) patients, respectively.

The mean PSL dose remarkably decreased to 17.95 mg/kg in the first month after RTX administration ($p = 0.012$). After 12 months, the mean reduction in PSL dose reached 84.1% ($p < 0.001$). Six patients experienced relapse within a mean duration of 9.5 months after the initial clinical response. Four of them were managed with additional cycles of RTX. At the last visit, six patients were in the control phase of the disease, four in partial remission, and five in complete remission. The mean follow-up time was 29.5 months after the first RTX cycle. Five RTX-related adverse events were noted, three mild infusion reactions, one severe infusion reaction, and one COVID-19 infection. RTX was not used as a single treatment in some patients, and the combinatory effect of other medications could have influenced the clinical response.

Conclusion: RTX would be a promising treatment option in EBA by inducing sustained complete or partial remission. Considering the potentially scarring nature of EBA, early treatment with RTX could be more beneficial in managing the disease.



Abstract N°: 652**Clinical Characteristics and Survival of Pemphigoid and Pemphigus Patients**

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Introduction & Objectives:

Pemphigoid and pemphigus are blistering diseases caused by autoantibodies against bullous pemphigoid antigen (BP180, 230) and desmoglein (1, 3), respectively. Both diseases differ in their clinical presentation, laboratory findings, disease progression and severity. Currently, there is limited research in Thailand regarding survival rates and prognostic factors for these diseases.

Materials & Methods:

The study was conducted on retrospective data of patients with pemphigoid and pemphigus who were treated and followed up at Naresuan University Hospital between 1 October 2012 and 30 September 2022.

The demographic data and clinical characteristic of patient was described and compared. Comorbidity disease and drug use prior to diagnosis were compared between pemphigoid and pemphigus patients. The survival rate and prognostic factors were compared between pemphigoid and pemphigus patients by using the Kaplan-Meier method, log-rank test and cox's regression.

Results:

During ten-year period (2012-2022), there were 34 pemphigoid patients and 46 cases pemphigus patients (pemphigus vulgaris 33 cases, pemphigus foliaceus 12 cases and pemphigus vegetans 1 case). Clinical characteristics of the two diseases that differed were older age in pemphigoid patients (75.3 vs 51.2), more presence of small blisters (73.5% vs 52.2%), and less lesions in the oral cavity (20.6% vs 41.3%). Regarding comorbidities, pemphigoid patients had a statistically significantly (p -value <0.05) higher incidence of hypertension, cerebrovascular disease, and hyperlipidaemia than pemphigus patients. For medications used prior to diagnosis, univariate analysis found that pemphigoid was associated with the use of certain medications before diagnosis: angiotensin receptor blockers (ARBs), calcium channel blockers (CCBs), statins, clopidogrel, biguanide, and non-steroidal anti-inflammatory drugs (NSAIDs). However, after adjusted in multivariate analysis by cerebrovascular disease and old age (more than 60 years), these were not statistically significant. For the outcome of treatment, the survival rate of pemphigoid and pemphigus at 1 and 5 years, there was a statistically significant difference (p -value = 0.001). The survival rate of pemphigus was 91.1% at 1 year (95% CI 77.9-96.6) and 82.2% at 5 years (95% CI 65.9-91.2). The survival rate of pemphigoid was 69.9% at 1 year (95% CI 51.2-82.5) and 47.7% at 5 years (95% CI 28.7-64.4). There was a difference of remission rate, but not statistically significant (p -value = 0.060). The remission rates of pemphigoid were 20.8% at 1 year (95% CI 10.5-38.8) and 41.6% at 5 years (95% CI 26.1-61.6), the remission rates of pemphigus were 47.3% at 1 year (95% CI 33.9-62.8) and 64.1% at 5 years (95% CI 49.9-78.1). In multivariable cox regression analysis, worse prognosis among pemphigoid and pemphigus patients was associated with having comorbidity disease (adj HR= 3.5, p -value=0.012) and being older than 60 years (adj HR= 2.5, p -value=0.034).

Conclusion:

Clinical characteristics of bullous pemphigoid and pemphigus were different such as age of onset, presence of blister and oral lesion. Neurological diseases were more common in bullous pemphigoid. Prior drug use was more common among bullous pemphigoid patients. Survival of pemphigoid patients were worse than pemphigus patients. However, these finding could be confounded by older age of pemphigoid patients.

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Abstract N°: 779**Self-assessment Pemphigus Vulgaris Activity Score (SA-PVAS): A new tool for patients to self-assess their disease severity**

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Introduction & Objectives:

Pemphigus Vulgaris (PV) is an autoimmune bullous mucocutaneous disease, highly affecting the quality of life of the patients. Many scoring indexes including Pemphigus Vulgaris Activity Score (PVAS) and Pemphigus Disease Activity Index (PDAI) have been used to determine the severity of the disease. Regarding the importance of using a tool to assess the PV severity by the patients themselves, we conducted this study to evaluate the accuracy of the translated simplified version of the PVAS called Self-Assessment PVAS (SA-PVAS) as a self-assessment tool in comparison with the PDAI score system as a gold standard. This study aimed to evaluate a simplified Self-Assessment PVAS (SA-PVAS) as a patient-reported measurement in comparison to PDAI measured by the physicians.

Materials & Methods:

At first, a Persian simplified version of PVAS was created (SA-PVAS) consisting of 7 multiple choice questions and assessed for validation and reliability by 64 PV patients. Then, the second group of PV patients, consisting of 81 cases, referring to pemphigus clinic from April to December 2018, were asked to score their disease using the validated SA-PVAS. The accuracy of patients' reported scores were compared with PDAI scored by the physicians. The effect of age, gender, disease duration, education, and place of residence (urban or rural) on SA-PVAS and PDAI correlations were also assessed. Moreover, a comparison between the reported severity of the disease by SA-PVAS and PDAI was made. To categorize the severity into mild, moderate, and severe; 25, 50, and 75 percentiles cutoffs were considered.

Results:

The entire designed questionnaire had very good reliability (Cronbach's alpha = 0.934) and acceptable factorial validity (skin, mucosal and total items had validations of 0.783, 0.64, and 0.638 respectively). There was a significant difference in the skin PDAI between men (mean = 5.47 ± 10.09) and women (mean = 0.96 ± 2.69) (P = 0.001). SA-PVAS indexes were not statistically different regarding gender. We observed no significant difference between age, disease duration, education, residency area and the indexes of both questionnaires. A strong (non-linear) correlation was observed between SA-PVAS and PDAI total (Spearman's = 0.6), skin (Spearman's = 0.766) and mucosal scores (Spearman's = 0.547) (P < 0.001). The two questionnaires had moderate agreements on reporting the severity of skin and total scores (k = 0.424, P < 0.005) and a poor non-significant agreement on mucosal severity (k = 0.172, P = 0.103).

Conclusion:

This study has an important implication for developing the Persian and simplified version of PVAS called SA-PVAS, as a self-assessment scoring system, for patients to report their disease activity which turned out to have a strong correlation with PDAI as a gold standard scoring system. Hence, SA-PVAS could act as a practical tool for

physicians to remotely follow and treat patients, particularly in conditions like COVID-19 outbreaks when the pandemic control protocols would restrict patients' physical presence in clinics and instead they show preference in reporting the disease condition from home.

Keywords: Pemphigus Vulgaris, SA-PVAS, PDAI, severity index, self-assessment.

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Abstract N°: 851**Cutaneous polyarteritis nodosa: a three-case-serie**

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Introduction & Objectives:

Cutaneous polyarteritis nodosa (cPAN) is a skin-limited variant of systemic polyarteritis nodosa (sPAN). It is a very rare necrotizing vasculitis of medium and small arterial vessels. We present a three-case-serie diagnosed with cutaneous polyarteritis nodosa with the aim of describing the clinical features, histologic findings, treatment and follow-up of the disease.

Materials & Methods: Medical history of all three patients

Results:

We present the data corresponding to the three patients in Table 1.

Table 1. Description of patients

	Patient 1	Patient 2	Patient 3
Sex / Age	Man / 43y	Woman / 29y	Woman / 73y
Medical history of interest	Smoker	Down syndrome; Hypothyroidism	Hypertension, dyslipidemia, cardiac valve disease
Time from first outbreak to diagnosis	5 years	6 months	1 year
Clinic			
Localization	Posterior region of left leg	Both forearms and lower third of both legs	Anterior part of legs and thighs
Erythematous nodules	+	-	+
Livedo reticularis	-	+	+
Purpura	-	-	-
Fever	-	-	-
Arthralgia	+	-	-
Myalgia	-	-	-
Mononeuritis	-	-	-
Ulceration	-	-	-
Histology	Vasculitis of small-medium caliber vessel + predominantly septal panniculitis	Vasculitis of small-medium caliber vessel	Vasculitis of small-medium caliber vessel + predominantly septal panniculitis
Treatment	Prednisone 30mg	Prednisone 30mg	Prednisone 30mg
Response	Recurrent outbreaks	Recurrent outbreaks	No new outbreaks after treatment
Follow-up time	3 months	60 months	14 months
Progression to sPAN	-	-	-

In our case series, we observed a female predominance (66%). The ages are disparate, ranging from the third decade of life to the eighth. The time from the onset of symptoms to diagnosis is one year or more in 2 of 3 patients, being the earliest diagnosis in the patient who was being followed up by Dermatology for nevus revision, which has influenced in an early diagnosis.

As for the clinical manifestations, 100% of the patients had involvement of the lower limbs, and only 1 patient had involvement of the upper limbs. Erythematous nodules are observed in two thirds, as well as livedo reticularis. None of the patients had ulceration, considered a prognostic factor in the severity and recurrence of cPAN.

Histology revealed in all patients a small-medium vessel vasculitis, accompanied by septal panniculitis in the two patients with erythematous nodules in the clinic.

Treatment with oral corticosteroids was decided in all of them, with a favorable response in only 1 case.

After a follow-up of 3-60 months, no signs or symptoms suggesting evolution from cutaneous to systemic variant have been detected.

Conclusion:

In this case series we describe the epidemiologic, clinical and histologic findings associated with cPAN. The presentation in the form of erythematous nodules or livedo reticularis coincides with other published cases. Ulceration, the main prognostic factor, was not present in any of our patients, which a priori implies a favorable prognosis. Although these patients should be followed closely for the appearance of systemic symptomatology, this is currently considered to be an exception, and it is recommended that in case of the appearance of systemic symptomatology, another different coincident vasculitis should be ruled out rather than assuming a shift towards the systemic form of cPAN. With this publication we want to contribute with three more cases to the description of the clinical findings of this rare variant of polyarteritis nodosa.

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Abstract N°: 899**A Case of Hypertrophic Discoid Lupus Erythematosus Mimicking Hypertrophic Lichen Planus: Dermoscopic and Histopathologic Clues to Diagnosis**Luddwi Achmad Rizky^{*1}, Rahadi Rihatmadja¹, Eyleney Meisyah Fitri¹, Windy Keumala Budianti¹¹Faculty of Medicine Universitas Indonesia – Dr. Cipto Mangunkusumo National Hospital, Dermatology and Venereology, Jakarta, Indonesia**Introduction & Objectives:**

Cutaneous lupus erythematosus (CLE) is classified into acute, subacute, and chronic forms. The most common form of the latter is discoid lupus erythematosus (DLE). A rare variant of DLE, hypertrophic discoid lupus erythematosus (HDLE) or verrucous DLE, is diagnostically challenging because of its resemblance to other dermatoses, one of them being hypertrophic lichen planus (HLP) which may also coexist with LE.

Materials & Methods:

A 56-year-old female presented with atrophic, partly scaly, violaceous to hyperpigmented plaques with indurated edges measuring between 1 cm² and 6 cm² of two-year duration on both sides of the forehead, left infraorbital area, left ear, and chest. The plaque on the chest was noticeably more hypertrophic. Patchy alopecia marred several areas of the scalp, although pruritus and photosensitivity were less obvious. The patient had worked as a street vendor for 30 years without adequate sun protection. She was suspected to suffer from CLE, but the possibility of HLP on the chest was explored by investigating its dermoscopic and histopathologic features, and compared them with the description in the existing literature.

Results:

Dermoscopic evaluation of the hypertrophic plaque on the chest revealed follicular keratotic plugs, follicular red dots, telangiectatic vessels, speckled brown pigmentation, structureless whitish areas, and white scales, consistent with the histologic findings which comprised hyperkeratosis, acanthosis, keratin plugs, lymphocytic infiltrate at the dermo-epidermal junction, prominent vacuolar alteration, thickened and disrupted basement membrane, and pigment incontinence. Mucin deposition, lymphocytic infiltrates resembling germinal center, and Civatte bodies (CB) were also observed in the dermis. Those dermoscopic and histopathologic features are frequently encountered in DLE. However, certain features such as marked hyperkeratosis accompanied by congestive margins and central atrophy in this patient, along with follicular plugging and acanthosis, are clinically more attributable to HDLE, according to previous studies. While CB may probably be found in lichen planus (LP), the latter is pruritic, and should demonstrate Wickham striae clinically and under dermoscopy. Furthermore, patients with HDLE are not notably more susceptible developing systemic lupus erythematosus (SLE) compared to those with classic DLE lesions.

Conclusion:

HDLE may mimic HLP, but certain dermoscopic and histopathologic characteristics, namely hyperkeratosis with follicular plugging, congestive margins and central atrophy, and Wickham striae, should enable differentiation between both diseases.

Abstract N°: 968**A pilot study characterising chronic graft-versus-host disease morphology longitudinally in a patient from retrospective systematic analysis of clinical photographs**

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Introduction & Objectives:

Characterization of skin chronic graft-versus-host disease (GVHD) is challenging, due to the varied morphology, time-consuming nature and expert skill needed for accurate assessment and documentation. Our objective was to retrospectively evaluate progression of cutaneous cGVHD in a single patient over multiple time points using high-quality medical photographs and avatar mapping with proprietary anatomy mapping software (1).

Materials & Methods:

After receiving approval by the lead authors' Institutional Review Board, we selected a patient with cutaneous cGVHD who had undergone serial photography in a professional medical photography studio over several years. Five of the most comprehensive photography sets spaced over time were selected for analysis. A blinded researcher reviewed all photographic images taken of this patient from onset of acute GVHD to end of life, with quality check by a GVHD dermatologist. For each visit, combinations of epidermal, sclerotic and pigmentary morphologies were annotated on a customized version of a surface anatomy mapping application (Anatomy Mapper®). Later, clinician and patient impressions regarding disease severity at timepoints correlating with photographs were documented.

Results:

We identified 23 sets of photographs from the index patient, a 59-year-old Fitzpatrick 1-2 skin type male. He had received a matched unrelated peripheral blood stem cell transplant for myelodysplastic syndrome and fludarabine and melphalan conditioning. The most heavily affected sites were his abdomen, including isomorphic plaques at the beltline, back, and distal extremities. In general, morphology transitioned from epidermal, to sclerotic and epidermal, to sclerotic and pigmentary disease. Standardized avatars were found to correlate subjectively with patient and clinician impression of change and with medication requirement (Figure 1). For example, we observed steady improvement after the introduction of oral ruxolitinib.

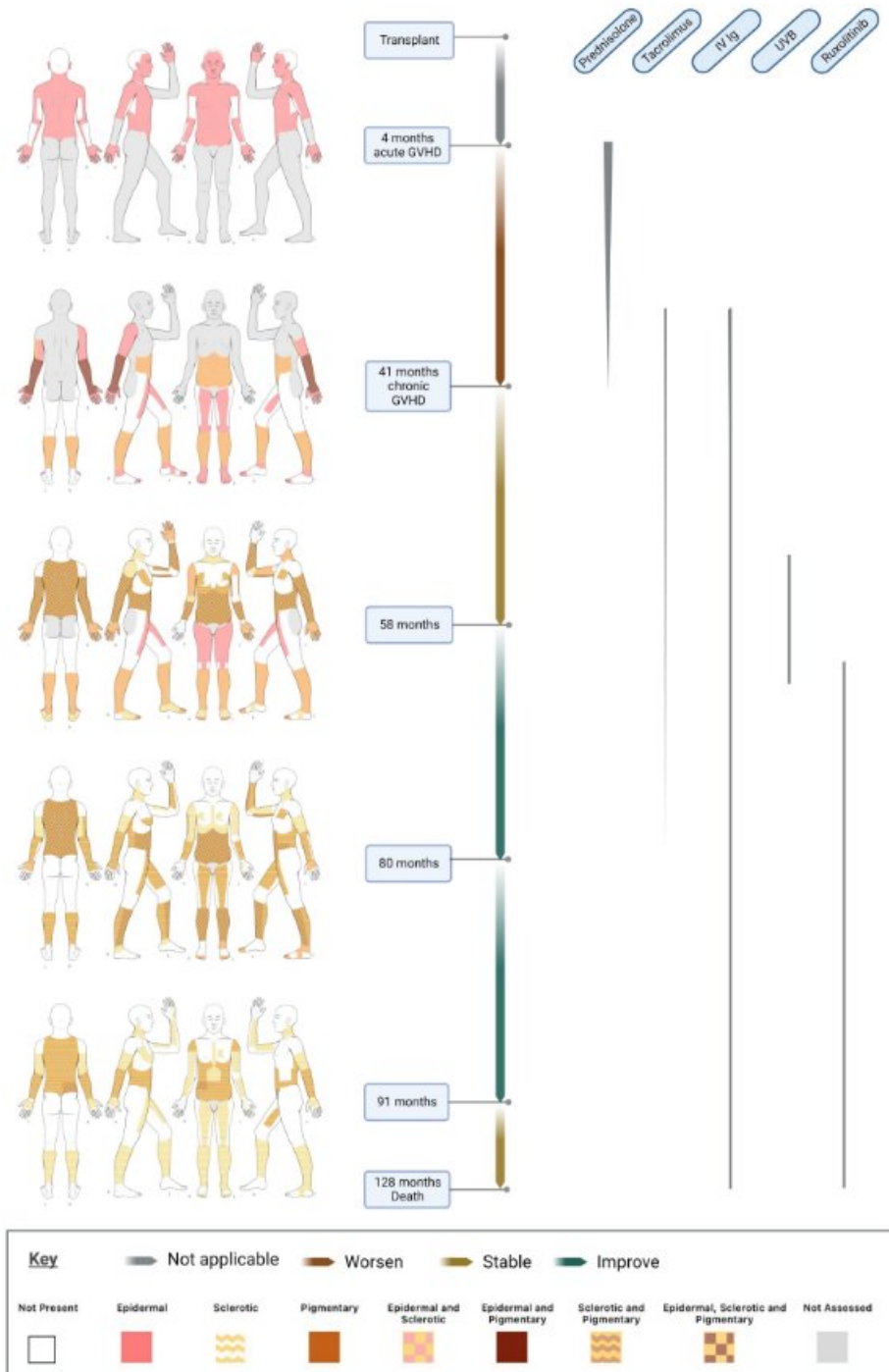
Conclusion:

This study is limited by having only a single assessor interpret morphology from photographs, though this limitation was mitigated by review by a GVHD expert. While variations in photographic technique were noted, such as with lighting, the avatar enabled standardized viewing despite variations in camera angles and missing data. In conclusion, this pilot study shows that documenting findings from medical photography using standardized avatar annotation enables visualization of progression of cGVHD morphology over time.

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Figure 1. Visual representation of progression of cutaneous graft versus host disease (GVHD) morphology in one patient over time. Dates refer to time (months) following allogeneic hematopoietic cell transplantation. Created with AnatomyMapper® (avatars) and Biorender.com© (flow diagram). IV Ig = intravenous immunoglobulins; UVB = narrowband ultraviolet B phototherapy.



Abstract N°: 1069**Immunological profile by enzyme-linked immunosorbent assay anti-BP230 and anti-BP180 of 24 patients with bullous pemphigoid**

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Introduction & Objectives:

The detection of anti-BP230 and BP180 antibodies by enzyme-linked immunosorbent assay (ELISA) techniques confirms the diagnosis of bullous pemphigoid (PB).

We aimed to describe the immunological profile by ELISA BP230 and BP180 test of patients with PB diagnosed in the region of Oran (western Algeria).

Materials & Methods:

We retrospectively analyzed the serum samples of 24 patients with PB diagnosed during the period from January 2014 to December 2015 at three dermatology departments in the region of Oran (Western Algeria).

Results:

The average age of our patients was 70 years old with a sex ratio (F/M) of 0.71. Neurological history was found in 25% of cases. Involvement of the oral mucosa was objectified in 29.2% of cases. Hyper eosinophilia was present in 60% of cases. It was greater than 1500 elements in 29.2% of cases. At histology, subepidermal detachment was objectified in all patients. The contents of the blister were predominantly PNE in 33.3% of cases. The margination of eosinophil was found in 83.3% of patients. The direct immunofluorescence was positive in all patients; with IgG type only in 16.7% of cases, C3 type only in 9.3% of cases and mixed (IgG+C3) in 75% of cases. The ELISA test was positive for BP230 in 66.7% of cases and for BP180 in 83.3%, both tests were positive in the same patient in 54.16% of cases. The mean titer of anti-BP230 and BP180 antibodies was 92.7 ± 77.2 RU/ml and 134.2 ± 75.5 RU/ml, respectively.

Conclusion:

In our study, the combination of the two ELISA tests BP230 and BP180 confirmed the diagnosis of BP in 96% of case.

Abstract N°: 1130**Epidermolysis Bullosa Acquisita in a child with severe mucosal involvement - Case report**Loubna Zreik¹, Issam Tablit¹, Samira Zobiri¹¹CHU Mustapha, Dermatology**Introduction & Objectives:**

Epidermolysis bullosa acquisita (EBA) is an acquired subepidermal bullous disease characterized by** IgG autoantibodies directed against type VII collagen. Its occurrence in childhood is very rare.** We report here the case of a child with EBA involving multiple mucous membranes.

Materials and Methods:

An 8 year-old-boy, presented with a 5 year-history of non-pruritic disseminated recurrent bullous lesions, mainly occurring on acral regions and sites of trauma, associated with skin fragility and mucosal involvement.

Physical examination revealed a boy with failure to thrive, presenting multiple tense serous-filled vesicles on a non-inflamed skin, located on the extensor surfaces of acral regions, associated with numerous atrophic scars, milia and hypo- or hyperpigmentation. He also had erosions on oral, nasal and anal mucous membranes. Otherwise, he was suffering from bilateral blepharitis with major photophobia mainly in the right eye. Ophthalmological examination showed bilateral keratitis with corneal ulcer in the right eye. Nasofibroscope demonstrated nasal synechiae without laryngeal lesions. Digestive endoscopy was not performed.

Histopathological examination showed subepidermal blister and dermal multinucleated cells infiltrate. Direct immunofluorescence (IF) revealed linear deposits of IgG, IgA and C3 at the basement membrane zone (BMZ). Indirect IF showed circulating IgG anti-BMZ antibodies at the dermal side of salt-split skin. ELISA demonstrated autoantibodies against type VII collagen. Diagnosis of EBA was made and a combination of dapsone (25 mg/d) and prednisone (0.5 mg/kg/d) was started.

Results:

EBA is a rare autoimmune dermatosis affecting the skin and mucous membranes triggered by autoantibodies against collagen VII.** EBA has no sex predilection and can occur anytime from childhood to old age.

EBA is characterized by the presence of tense bullae, erosions and skin fragility. The presence of milia is a valuable feature for considering of this diagnostic possibility. EBA has two main clinical forms: inflammatory and mechanobullous. While the mechanobullous phenotype is typically seen in adults, the inflammatory or bullous pemphigoid-like form is more commonly observed in children. The frequency of mucosal involvement is quite different between adults and children. In children, it is mentioned in the vast majority of the reported cases. The most frequently affected mucosae in children are the mouth and genitalia.

Our patient met the criteria for EBA as indirect IF and ELISA showed IgG at the dermal side of salt-split skin and IgG-binding type VII collagen. The clinical features were consistent with the mechanobullous EBA-form. Because of an early-onset, our patient was misdiagnosed as dystrophic epidermolysis bullosa for 5 years. Although the severity of mucosal involvement in our patient is reminiscent of cicatricial pemphigoid, it still belongs to the current spectrum of EBA.

Overall, the prognosis and response to treatment in EBA are better in children. The drugs used are prednisone and

dapsone, which are more effective when combined. In severe cases, immunosuppressants, intravenous immunoglobulin, and rituximab can be used. Our patient seemed to respond well to the treatment.

Conclusion:

Our report emphasizes the difficulty in making the diagnosis of EBA in childhood. Only an accurate diagnosis allows an early appropriate treatment which may avoid the severe outcome of mucosal lesions in EBA.

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Abstract N°: 1205**The content of antinuclear antibodies in patients with vulvar lichen sclerosis**Delya Fozilovna Porsokhonova^{*1}, Nurjanar Ilesova¹, Khalida Khalidova¹, Yulduz Askarovna Alimukhamedova¹¹Republic Center Dermatovenereology and Cosmetology, STD and Reproductive disorders, Tashkent, Uzbekistan

Introduction & Objectives: Vulvar lichen sclerosis (VLS) is a chronic relapsing dermatosis that occurs with periods of exacerbation and remission and, without proper treatment, can lead to dysplasia and malignancy. There is no specific etiology of this dermatosis, but there are many factors that, together or separately, lead to the development of the disease. The most common theory of VLS formation is the theory of autoimmune inflammation, according to which the formation of antibodies against extracellular matrix 1 (ECM1) protein and the basement membrane of the skin can contribute to the progression of the disease. Autoantibodies to ECM1 are detected in 74% of patients with VLS. In this connection, it was of interest to determine antinuclear antibodies (ANA) in the blood serum of patients with VLS.

Materials & Methods: Under our supervision there were 35 patients with a diagnosis of vulvar lichen sclerosis at the age of 18 to 75 years. The design of the study included a comprehensive clinical and laboratory examination of patients in order to characterize the autoimmune mechanism of the development of the disease. Antinuclear antibodies of class G to denatured single-stranded DNA (ss-DNA) and native double-stranded DNA (ds-DNA) in the blood serum of patients were determined by ELISA. Patients were also examined for urogenital infections by PCR.

Results: The study revealed an increase in IgG to ss-DNA in 11 (31.4%) patients, an increase in IgG to ds-DNA in 8 (22.9%) patients. A simultaneous increase in IgG to ss-DNA and ds-DNA was observed in 9 (25.7%) patients. In 7 (20.0%) patients, there was no increase in the content of antinuclear antibodies. In general, an increase in ANA was noted in patients with the presence of human papillomavirus type 16/18 and *Ureaplasma urealyticum* (Parvo), which may indicate the triggering role of these pathogens in the formation of an autoimmune process. We have not recorded the dependence of the increase in ANA on age.

Conclusion: Thus, an increase in IgG to denatured DNA (ss-DNA), and IgG to native DNA (ds-DNA) was observed in almost 80% of patients with VLS, which may indicate the development of autoimmune inflammation and become an available informative indicator characterizing the onset of the autoimmune mechanism. In patients with VLS. More frequent detection of human papillomavirus type 16/18 and *Ureaplasma urealyticum* (Parvo) may indicate their role in the development of chronic inflammation of the vulva and the triggering of cellular dysplasia.



Abstract N°: 1238

Pemphigus vegetans mimicking tinea pedis

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Introduction & Objectives:

Pemphigus is a group of autoimmune blistering diseases with acantholysis, affecting skin and mucosa. There are several types of pemphigus depending on the level of the split.

Pemphigus vegetans is similar to pemphigus vulgaris but causes vegetative plaques that usually develop in intertriginous areas such as groins and armpits.

Materials & Methods:

We present the case of a patient with pemphigus vegetans that had been previously misdiagnosed *astinea pedis*, and to review the main characteristics of this entity.

Results:

A 70 year old man was referred to the Dermatology department with a 6 month history of erythematous annular plaques on both feet. He had previously received antifungal treatment on suspicion of fungal infection, without response.

On examination he presented large annular plaques with pustular edge and marked exudation, located in the interdigital spaces and dorsum of the feet. Cultures and skin biopsy were performed, and we indicated empirical antibiotic and antifungal therapy. Two weeks later, the patient presented with a torpid evolution of the lesions and the appearance of several infiltrated erythematoviolaceous vegetative masses in both inguinal and axillary folds, as well as whitish plaques on the back and lateral side of the tongue.

Skin mycological cultures were negative and histological study revealed spongiosis and acantholysis with intraepidermal blisters, pseudoepitheliomatous hyperplasia and papillomatosis, with a dense eosinophilic infiltrate. Direct immunofluorescence showed deposition of IgG and C3 within the intraepidermal space and on the surface of keratinocytes. Blood tests showed eosinophilia and positive anti-desmoglein 1 and 3 antibodies.

With diagnosis of vegetative Hallopeau variety of pemphigus vegetans, we prescribed treatment with oral prednisone with very good response, subsequently associating mycophenolate mofetil.

Conclusion:

Pemphigus vegetans is one of the rarest clinical variants of pemphigus, characterized by the formation of vegetative plaques in intertriginous areas and oral mucosa. Diagnosis is challenging due to the variable presentation and presence of verrucous vegetations.

There are two recognized forms of pemphigus vegetans that differ in clinical presentation, evolution and prognosis. Hallopeau variant presents an indolent course and associates pustules and vegetative plaques. On the other hand, Neumann type is more severe and refractory to treatment, with major affectation of oral mucosa.

Systemic corticosteroids represent the first line of treatment, and other immunosuppressive agents (azathioprine, dapson, mycophenolate mofetil or cyclophosphamide) can be associated. In case of absence of complete clinical response, rituximab can be used as a second-line treatment.

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Abstract N°: 1269**Assessment of the level of C-reactive protein in the clinical course in patients with acantholytic pemphigus.**

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Introduction & Objectives: Acantholytic pemphigus (AP) is one of the severe dermatosis, accompanied by damage to the skin and / or mucous membranes, the leading role in the pathogenesis of which is assigned to autoimmune reactions leading to the harmful effect of circulating auto-antibodies that are directed against the components of the desmosomes. In the development of the immune response, along with antibodies and cytokines secreted by lymphocytes, acute-phase serum proteins are also involved, the concentration of which rapidly increases during the infectious process.

The aim of our research was to assess the state of C-reactive protein in the blood serum of patients with acantholytic pemphigus.

Materials & Methods: We examined 57 patients with pemphigus aged 18 to 71 years. All patients underwent clinical, cytological, biochemical and immunological studies. Determination of the level of C-reactive protein (CRP) in blood serum was determined by the method of solid-phase ELISA study. All patients were consulted by related specialists: therapist, endocrinologist, etc.

Results: Among 57 patients with AP, the vulgar form was 51 patients (89.5%), the seborrheic form was 4 (7.01%) and the vegetative form was 2 (3.5%), respectively.

The results of the ELISA study showed that among 57 patients in 50 patients in the blood serum there was an increase in the concentration of CRP, which amounted to 87.7% of cases, which indicates the severity of the inflammatory reaction in the body of patients with acantholytic pemphigus.

Analysis of the quantitative characteristics of C-autoantibodies (AAT) to native DNA in the blood serum of patients with pemphigus revealed an increase in its concentration by 3.8 times compared with the control group and averaged 18.3 ± 0.5 IU/ml and had a statistically significant character ($P < 0.05$).

Taking into account the prescription of the disease up to 1 year, the average CRP level was 21.4 ± 0.6 IU/ml, with a prescription of 1-5 years - 18.6 ± 0.8 IU/ml and more than 5 years - 17.2 ± 1.3 IU/ml, respectively ($P < 0.05$). A sharp increase in the level of CRP in patients with acantholytic pemphigus causes the initial stage of the acute inflammatory response of the body to the production of autoantibodies. With an increase in the prescription of the disease, the level of CRP was also in the range of high concentrations, however, compared with the prescription of up to 1 year, it decreased by 1.2 times ($P < 0.05$).

Conclusion: The results obtained indicate the development of severe inflammation in patients with acantholytic pemphigus and an increase in the level of CRP indicates the activation of autoantibodies, contributing to the development of complications of both a bacterial and / or viral, fungal nature. In our opinion, the data obtained have diagnostic and prognostic significance in the clinical course of the disease, and the determination of the CRP titer will contribute to the further choice of adequate treatment.

Abstract N°: 1441**Bullous pemphigoid and neurologic comorbidities: cause or consequence?**Lula María Nieto Benito¹, Ricardo Suarez-Fernandez¹¹Gregorio Marañón General University Hospital, Dermatology, Madrid, Spain**Introduction & Objectives:**

Bullous pemphigoid (BP) is the most prevalent autoimmune bullous disease, with a prevalence that is not negligible, despite being considered a rare disease. In recent decades, there has been a significant increase in the incidence of this disease, with the presence of neurological comorbidities being one of the possible explanations for this increase. The aim of this study was to analyze the neurological comorbidities of patients with BP in our setting.

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 the type of neurological comorbidity if it was present, were collected.

Results:

Of the total of 257 patients with BP, 102 (39.7%) had at least one neurological disease at the time of diagnosis of BP. The mean age of this subgroup of patients was 81.66 years (± 9.06 SD) and 58.8% were men. Senile dementia, Alzheimer's disease and cerebrovascular disease were the most frequent (**Figure 1**), each of them being present in 26 patients. We observed that patients with BP and neurological disease had higher mortality, greater disease activity (higher anti-BP180 NC16A autoantibody titers) and disease severity. The characteristics of the patients with BP and neurological disease are listed in **Table 1**. Despite not being included in our study, BP has been associated with Central Nervous System (CNS) neoplasms, as well. The neuroinflammation present in these diseases and the existence of neuronal isoforms of BP180 and BP230 would explain the connection between BP and neurological diseases.

Conclusion:

Neurodegenerative diseases are the main comorbidity of patients with BP and, due to the neuroinflammation that produces them, they predispose, *per se*, to the development of this disease, which will be more severe and more fatal when compared to patients with bullous pemphigoid without any neurological history.



Abstract N°: 1448**New treatment prospects for female patients with lichen sclerosus.**

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Introduction: Lichen sclerosus (LS) is an acquired chronic inflammatory dermatosis commonly affecting the vulvar and perianal regions. The incidence of LS is increased among women in recent decades. Actual therapeutic approaches of LS involve the long-term use of the high-potent topical corticosteroids, emollients, and topical calcineurin inhibitors. The treatment requires high, not always achievable compliance. Hence methods are sought, which would prolong the remission and reduce the topical steroid burden.

Objectives: To evaluate the efficacy of fractional thermolysis with an ER:YAG laser in combination with platelet-rich plasma (PRP) in the treatment of vulvar LS.

Materials & Methods: Eligible patients were 10 women (31 to 68 years, mean age 48.1 ± 1), LS lasts 5.2 ± 1.3 years. Previous treatment included 0.05% clobetasol ointment and a moisturizer. Patients involved areas were treated with 2940 nm ER:YAG laser (Asclepion MCL 31) and following PRP. Patients were followed 4 times for 180 days: first and second (30 ± 2 day) treatment sessions, first (60 ± 2 day) and second (180 ± 2 day) follow-ups. Efficacy was evaluated by the Dermatology Clinical Sign Score (DCSS), the Visual Analogue Scale for pruritus (VAS) and the Dermatology Life Quality Index (DLQI)).

Results: VAS was initially 4–5 (mean 3.9 ± 1.9). By the second visit VAS decreased to 1.3 ± 0.6 ; on following visits VAS did not exceed 0.5. Before the treatment DCSS was 7 ± 0.2 and after - 1.8 ± 0.1 . At follow-ups patients showed the affected area reduction, the lesions completely resolved in 4 women. Only 6 patients continued to use moisturizers and topical steroids. DLQI dropped from 11.28 ± 2.57 to 1.49 ± 0.31 . Patients reported moderate pain during the procedures. No side effects were noted.

Conclusion: The results showed the high efficacy of combination therapy. ER:YAG laser in combination with PRP allows to induce long-lasting remission and to reduce the topical steroid burden.



Abstract N°: 1539

A Clinico-Histopathological evaluation of Trichloroacetic Acid in treatment of Alopecia Areata

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Introduction & Objectives:

Trichloroacetic acid in a concentration of 35% (TCA) is a medium-depth chemical peeling agent that cause controlled keratocoagulation and denaturation of the proteins within the epidermis and upper dermis. The proposed mechanism of action utilizes stimulating of growth factors, proinflammatory cytokines and chemokines during the obtaining of therapeutic wound and its healing process. There are very few publications on this treatment modality and up to our knowledge this is the first work assessing efficacy of TCA in treatment of AA with clinico-histopathological correlation of treatment results.

The aim of the current work was to assess clinically, trichoscopically, and pathologically the efficacy and safety of TCA 35% peeling agent for the treatment of alopecia areata.

Materials & Methods:

We perform prospective cross-sectional open-label single-center interventional study which included 40 patients diagnosed clinically, dermoscopically, and pathologically as alopecia areata.

Inclusion criteria

- \1) Patients diagnosed with Alopecia areata multilocularis (including Ophiasis pattern of alopecia areata) and Alopecia totalis aged from 16 to 50 years of both genders.
- \2) Wash out period from previous treatment (if any) 3 months.

Exclusion criteria

- \1) Age less than 16 and more than 50 years old.
- \2) Alopecia areata of the face or any other body areas.
- \3) Pregnant women.
- \4) Patients with the psychiatric disorders, chronic diseases of heart, kidney, liver, or blood.

All participants were subjected to evaluation by McDonald Hull and Norris grading system for density, pigmentation, and texture of growing hair at first visit, before each session and after 3 months of follow up period. Serial photographs were done, and scoring was assessed by two non-treating blinded dermatologists to minimize potential bias.

Dermoscopic evaluation: All participants were examined using Dermoscope, and was evaluated by non-treating blinded dermatologist (at initial visit, before every session and after 3 months of follow up period) to assess the signs of disease such as exclamation marks, dystrophic hairs, yellow dots, and black dots.

Histopathological evaluation: A 4-mm punch biopsy was carried out at the first visit and 4 weeks after the last session.

Patient satisfaction was measured using the quartile scale (slight improvement <25%, moderate improvement 25%–49%, significant improvement 50%–74% and marked improvement \geq 75%). Participants made grading 4 weeks after last treatment session.

Results:

After completion of research there was response to treatment in 30 patients (75%). The rest 10 subjects (25%) had no response. Response was graded according to McDonald Hull and Norris grading system.

In the present study the higher percentage of response was in subjects with active alopecia (14 patients, 100%) in comparison to subjects with inactive form of disease (16 patients, 61.5%) with statistically significant difference.

Histopathological evaluation of our study showed marked reduction of lymphocytes infiltrate around the bulb region of anagen hair follicles (swarm of bees) in active case of alopecia areata.

Conclusion:

Chemical peeling with topical TCA 35% is an effective, safe treatment for alopecia areata. It is easy to perform in the outpatient clinic. Peeling agents is a good option in patients with wide area of the scalp affection and alopecia totalis.



Abstract N°: 1605**Bullous pemphigoid secondary to an orf nodule: a still unrecognized complication!**

Zineb Zeggwagh¹, Meriam Meziane¹, Karima Senoussi¹

¹Department of Dermatology and Venerology, University Hospital Center Ibn Sina-University Mohamed V, Rabat, Morocco.

Introduction & Objectives:

Orf's nodule is a rare viral zoonosis caused by parapoxvirus infection. Diagnosis is often clinical.

Several complications can occur following orf's disease: fever, lymphadenopathy and bacterial infection. Post-Orf autoimmune bullous dermatoses are much rarer with only 11 cases in the literature. We describe a case of bullous pemphigoid secondary to orf's disease which is considered a very rare complication.

Materials & Methods:

Case report:

A 56-year-old man, presented 8 days after Eid Al Adha, a nodule in the right middle finger gradually increasing in size. In view of this lesion and the context of occurrence (handling of the sheep during Eid Al Adha) the diagnosis was in favour of orf. Three weeks after the appearance of the orf nodule, he consulted for a diffuse pruriginous bullous eruption, initially located in the right arm then generalizing to the trunk, abdomen and all four limbs. Dermatological examination found an erythematous ulcerated nodule that was well limited, firm, deeply infiltrated, measuring 2 cm and located on the right middle finger. Multiple tense blisters with clear contents on noninflamed skin affecting the four limbs, the abdomen and the back. Some erosions on the limbs and abdomen. The Nikolsky sign was negative. The rest of the examination was normal.

Skin biopsy revealed a subepidermal blister with a mixed inflammatory cell dermal infiltrate. Direct immunofluorescence showed a linear deposition of C3 along the basement membrane. Indirect immunofluorescence revealed the presence of anti basal membrane antibodies positive at 40. The diagnosis of bullous pemphigoid complicating an orf nodule was retained. Treatment with topical steroids (clobetasol propionate 0,05%) was rapidly effective with disappearance of the bullous lesions in a few days. There was no relapse 1 year of follow-up.

Results:

Orf is a zoonosis caused by a virus from the parapoxvirus family. It's transmitted to humans through direct contact with infected animals, or indirectly through contaminated offal or knives, which is our patient's mode of transmission. Post-Orf autoimmune bullous dermatoses are much rarer with only 11 cases in the literature. The first cases of bullous pemphigoid secondary to an orf nodule were described in 1995 by J K Murphy et al, where they reported 5 cases occurring after 2 to 3 weeks. In our case, the bullous lesions appeared about 3 weeks after the appearance of the orf nodule, which is consistent with the literature (2-4 weeks).

The clinical, histopathological, immunofluorescence results, as well as the improvement under topical steroids and the absence of relapse confirmed the diagnosis of orf-induced bullous pemphigoid.

The relationship between orf and bullous pemphigoid has not yet been fully elucidated.

The physiopathological mechanism of autoimmunity induced by parapoxvirus infection is still poorly understood, hence the interest of reporting other cases of this complication.

The patient was treated by topical steroid with a total improvement .

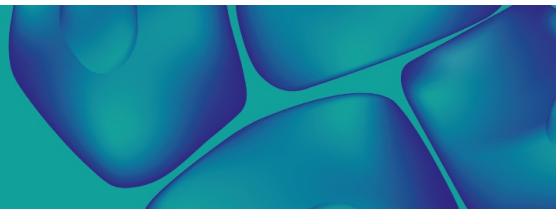
In the case described in the literature, they also treated their patient by topical steroid but some patient received prednisone, azathioprine or dapsone or colchicine or ciclosporine.

Conclusion:

We report a case of bullous pemphigoid secondary to an orf nodule which is a very rare complication. The relationship between this two disease has not been elucidated completely yet. Further case reports regarding this clinical condition are needed.

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Abstract N°: 1617

Ulcerated Lesions of the Oral Mucosa: Clinical and Histologic Review of Lichen planus.

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Introduction & Objectives: Around 25% of adults experience mouth ulcers, which can be caused by various factors, including lichen planus, a chronic inflammatory and autoimmune disorder affecting the oral cavity. According to literature, differential diagnosis of oral lichen planus can be aided by the use of direct immunofluorescence (DIF). The diagnostic criteria for oral lichen planus, which were first published in 1978, are still being refined, with the most recent diagnostic algorithm proposed in 2019 based on clinical signs and medical history. Our study aimed to develop a new multivariate predictive model by combining medical history and DIF.

Materials & Methods: The study included patients who presented to the Department of Dermatology in 2019-2022 with erosive lesions in the oral cavity or were referred there by their dentists. The following variables were collected: DIF IgG, DIF IgA, DIF IgM, DIF C3, DIF F1, DIF F2, histopathology, gender, age on the day of lesion onset, stress during the study period etc. Statistical analysis was performed using Statistica 13. For neural networks we used default parameters of the Statistica software.

Results: The study group consisted of 80 patients: 63 (78.8%) women and 17 (21.2%) men. Lichen was confirmed by histopathology in 4 (5.0%) of the study participants and not confirmed in 57 (71.2%); it was not excluded in 30 subjects (37.5%) and excluded in 31 (38.8%). The incidence of DIF IgG, DIF IgA, DIF IgM, DIF C3, DIF F1, DIF F2 positivity did not differ significantly between either subjects with confirmed or unconfirmed lichen, or between subjects with lichen excluded or not excluded. Data Mining module suggested four significant predictors to create a multivariate model for dependent variable 'lichen planus not excluded by histopathology' and none for 'lichen confirmed'. It were: stress at onset (0.017), white patches under a tonque (p=0.029), erosions on mandibular gingiva (0.041), and erosions under a tonque (0.049). Neural networks created on this basis had 74% correct classifications for learning, 85% for testing, and 71% for validation (Figure 1).

Conclusion: In some populations, DIF is not a significant predictor of the diagnosis of lichen planus, regardless of whether strict diagnostic criteria for this disease were used in histopathological examination or whether results potentially indicative of lichen planus were also included. By using neural networks, interview data can establish a diagnosis with approximately 70% certainty compared to histopathology as the reference test. Further optimization of variables included in the model may allow for the creation of a clinically useful tool.

Fig. 1. ROC curve for 'lichen not excluded'.

Tab. 1. DIF results in patient subgroups distinguished by histopathology (HP) result.

N=63	Lichen <u>confirmed on</u> HP		p-value	Lichen not excluded on HP		p-value
	No (n=57)	Yes (n=4)		No (n=33)	Yes (n=30)	
DIF IgG (-)	53 (93.0)	4 (7.0)	0.635	29 (50.9)	28 (49.1)	0.594
DIF IgG (+)	3 (100.0)	0 (0.0)		2 (66.7)	1 (33.3)	
DIF IgA (-)	55 (93.2)	4 (6.8)	0.787	31 (52.5)	28 (47.5)	0.297
DIF IgA (+)	1 (100.0)	0 (0.0)		0 (0.0)	1 (100.0)	
DIF IgM (-)	53 (93.0)	4 (7.0)	0.635	29 (50.9)	28 (49.1)	0.594
DIF IgM (+)	3 (100.0)	0 (0.0)		2 (66.7)	1 (33.3)	
DIF C3 (-)	45 (91.8)	4 (8.2)	0.327	25 (51.0)	24 (49.0)	0.832
DIF C3 (+)	11 (100.0)	0 (0.0)		6 (54.6)	5 (45.4)	
DIF F1 (-)	40 (90.9)	4 (9.1)	0.212	25 (56.8)	19 (43.2)	0.185
DIF F1 (+)	16 (100.0)	0 (0.0)		6 (37.5)	10 (62.5)	
DIF F2 (-)	42 (91.3)	4 (8.7)	0.253	26 (56.5)	20 (43.5)	0.173
DIF F2 (+)	14 (100.0)	0 (0.0)		20 (35.7)	9 (64.3)	



Abstract N°: 1656**Progressive hemifacial atrophy: a case report of localized scleroderma**Gaia Fasano^{*1}, Cataldo Patruno¹, Serena Federico¹, Carmen Volpe¹, Stefano Dastoli¹¹Magna Græcia University, Dermatology, Catanzaro, Italy

Introduction & Objectives: Progressive hemifacial atrophy (PHA) is facial distortion with multiple etiology among which localized scleroderma. Linear scleroderma, called *en coupe de sabre* (ECDS), and PHA belong to the spectrum of localized scleroderma and can often coexist. Clinical manifestations begin in the first two decades of life and slowly progress until quiescence is reached. PHA is more often seen in female sex. Etiology is still not clear; trauma, autoimmunity and infection have been suggested as trigger factor. PHA is associated with multiple extracutaneous findings, of which neurologic complications are the most common.

Materials & Methods: A 41 years-old woman presented, from the age of 25, slowly progressive hemifacial atrophy of zygomatic area. After initial presentation the disorder self-limited and became stationary during the next 5 years. The atrophy affected subcutaneous tissue, fat, and muscle without involvement of osseocartilaginous tissue and neurological structures. The atrophic skin was hyperpigmented, shiny and firm with sclerosis and induration of the area. PHA was not associated with ECDS and other neurological or ocular findings. The diagnosis of PHA was clinical and supported by histopathology and imaging. Biopsy showed hyperpigmentation, dermal sclerosis, fat atrophy, decrease in adnexal structures, perivascular plasma cells and lymphocytes. Laboratory analysis such as complete blood count, protein electrophoresis, anti-nuclear antibody, rheumatoid factor, anti-Scl-70, C-reactive protein, anti-dsDNA antibody, extractable nuclear antigen screening, were normal. Brain Magnetic resonance imaging (MRI) and computer tomography (CT) showed absence of neurological and skull abnormalities.

Results: Considering the long clinically stability, the absence of functional alterations, and patient's wishes it was decided to carry out periodic follow up without any reconstructive surgery.

Conclusion: The lack of an etiology consensus and explanation of pathogenesis is due to the rarity of the disease. No standardized criteria for the diagnosis are available.



Abstract N°: 1692**Bosentan versus nifedipine in the treatment of vasculopathy in Vietnamese systemic sclerosis patients: A randomized clinical trial**Phat Trinh¹¹Vinmec Times City International Hospital, Internal Medicine, Hanoi, Viet Nam

Introduction & Objectives: Vasculopathy is a major cause of morbidity and mortality in systemic sclerosis. This study was performed to compare the efficacy of bosentan versus nifedipine in the treatment of vasculopathy in Vietnamese systemic sclerosis patients.

Materials & Methods: We randomly assigned patients in a 2:1 ratio to receive 62.5 mg of oral bosentan twice daily for 4 weeks followed by 125 mg twice daily for 12 weeks or receive 20mg of oral nifedipine twice daily for 16 weeks, respectively. The primary outcomes were the degree of change in Raynaud's Condition Score (RCS), appearance of new digital ulcers (DUs) and change in WHO functional class. Secondary outcomes included the change in the nailfold capillaries disease stage and systolic pulmonary arterial pressure (sPAP) values.

Results: At week 16, patients treated with bosentan had an improved RCS, the mean difference between two groups was 0.8 ± 0.2 (95 percent confidence interval [95% CI], 0.4 to 1.1, $p < 0.001$) and an improved WHO functional class, a mean treatment effect of 35.6% in favor of bosentan (95% CI, 13.4 to 57.7%, $p < 0.05$). Bosentan treatment was associated with a 58% reduction in the number of new DUs compared with nifedipine (mean \pm standard error: 0.22 ± 0.42 vs 0.52 ± 0.59 new ulcers, $p < 0.05$). sPAP value was decreased by 4.1 ± 3.8 mmHg (95% CI, 3.0 to 5.3, $p < 0.001$) in the bosentan group, versus 1.0 ± 2.9 mmHg (95% CI, -0.2 to 2.1, $p > 0.05$) in the nifedipine group. Both treatments did not improve nailfold capillaries disease stage. Headache was the most common adverse event in both groups

Conclusion: Bosentan significantly improved Raynaud's phenomenon, limited the occurrence of new DUs, significantly reduced symptoms of PAH and sPAP values and all were better than nifedipine.



Abstract N°: 1695**Bosentan versus nifedipine in the treatment of vasculopathy in Vietnamese systemic sclerosis patients: A randomized clinical trial**Phat Trinh¹¹Vinmec Times City International Hospital, Internal Medicine, Hanoi, Viet Nam

Introduction & Objectives: Vasculopathy is a major cause of morbidity and mortality in systemic sclerosis. This study was performed to compare the efficacy of bosentan versus nifedipine in the treatment of vasculopathy in Vietnamese systemic sclerosis patients.

Materials & Methods: We randomly assigned patients in a 2:1 ratio to receive 62.5 mg of oral bosentan twice daily for 4 weeks followed by 125 mg twice daily for 12 weeks or receive 20mg of oral nifedipine twice daily for 16 weeks, respectively. The primary outcomes were the degree of change in Raynaud's Condition Score (RCS), appearance of new digital ulcers (DUs) and change in WHO functional class. Secondary outcomes included the change in the nailfold capillaries disease stage and systolic pulmonary arterial pressure (sPAP) values.

Results: At week 16, patients treated with bosentan had an improved RCS, the mean difference between two groups was 0.8 ± 0.2 (95 percent confidence interval [95% CI], 0.4 to 1.1, $p < 0.001$) and an improved WHO functional class, a mean treatment effect of 35.6% in favor of bosentan (95% CI, 13.4 to 57.7%, $p < 0.05$). Bosentan treatment was associated with a 58% reduction in the number of new DUs compared with nifedipine (mean \pm standard error: 0.22 ± 0.42 vs 0.52 ± 0.59 new ulcers, $p < 0.05$). sPAP value was decreased by 4.1 ± 3.8 mmHg (95% CI, 3.0 to 5.3, $p < 0.001$) in the bosentan group, versus 1.0 ± 2.9 mmHg (95% CI, -0.2 to 2.1, $p > 0.05$) in the nifedipine group. Both treatments did not improve nailfold capillaries disease stage. Headache was the most common adverse event in both groups

Conclusion: Bosentan significantly improved Raynaud's phenomenon, limited the occurrence of new DUs, significantly reduced symptoms of PAH and sPAP values and all were better than nifedipine.



Abstract N°: 1703**Drug-induced lupus erythematosus tumidus during treatment with atorvastatin: a case report**Katarina Trcko¹¹Maribor University Medical Centre, Department of Dermatovenereology, Maribor, Slovenia**Introduction & Objectives:**

Lupus erythematosus tumidus (LET) is a rare photosensitive inflammatory disease of the skin, that is classified as a separate subtype of cutaneous lupus erythematosus. It has a benign course as it only rarely correlates with systemic autoimmune disease and does not induce scarring or postinflammatory hyper- or hypopigmentation. The diagnosis of LET is based mainly on clinical and histopathological findings. Drug-induced LET is rare. We report a case of LET in a patient with hyperlipidemia who developed LET on the face and back a few months after the start of atorvastatin therapy.

Materials & Methods:

We report a case of LET in a 56-year-old female who was treated with atorvastatin.

Results:

A 56-year-old woman presented with a 2-month history of multiple mildly itchy erythematous plaques on her face and upper back. The eruption appeared one month after starting treatment with atorvastatin. The patient had no previous history of skin disease or photosensitivity. Physical examination revealed several erythematous papules and plaques on the face and on the upper back. Routine laboratory tests including complete blood count, chemistry, immunochemistry (alfa-fetoprotein, CEA, CA19-9, CA-15-3, CA-125) and serum protein electrophoresis were within the normal limits, The erythrocyte sedimentation rate was elevated (76 mm/h, normal <28 mm/h). A complete autoantibody screening panel revealed positive antinuclear antibodies (ANA), H+, and titer 1:160 was negative.

Histopathologic examination of the tissue revealed a combination of interface dermatosis and moderate periadnexal, perivascular interstitial lymphoplasmacytic infiltrate accompanied by mucin deposition in the dermis. Immunofluorescence examination showed discrete focal granular deposits of IgM and C3 in the wall of small vessels and IgG and IgM along the dermo-epidermal border. Based on the clinical characteristic and pathohistological findings, a diagnosis of LET was made.

The patient discontinued treatment with atorvastatin and started treatment with mometazon cream. After a few weeks of local treatment, the skin changes disappeared and to date, patient has not had a relapse.

Conclusion:

Drug-induced LET is a relatively rare disease, in which LET is triggered by medication. In the literature, there are only a few drugs described that can induce LET. To our knowledge, this is the first case of statin-induced lupus erythematosus tumidus. The most likely mechanism by which atorvastatin can trigger LET is photosensitivity. Early diagnosis is crucial for its management since discontinuing the causative drug is the most important step. Identifying the cause of LET may be difficult because many patients take multiple medications. It is important for clinicians to be aware of any potential adverse effects associated with statins.

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Abstract N°: 1794

Hypocomplementemic Urticarial vasculitis presenting as an erythema gyratum repens - like eruption associated with incomplete SLE

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Hypocomplementemic Urticarial vasculitis presenting as an erythema gyratum repens - like eruption associated with incomplete SLE

Introduction:

Hypocomplementemic Urticarial vasculitis (HUV) as rare form of vasculitis characterised with urticarial lesion, hypocomplementemia and systemic manifestation. The histopathologic pattern of UV is that of leukocytoclastic vasculitis (LCV), consisting of fibrinoid necrosis of dermal vessels walls and neutrophilic perivascular inflammatory infiltrates. Lesions of UV last more than 24 h, often persisting for several days, and may leave behind a residual hyperpigmentation that is not seen in chronic idiopathic urticaria. Patients with low levels of complement have a higher risk of complications and are either classified as having HUV when systemic involvement is little to none or HUVS when systemic involvement is significant. Erythema gyratum repens (EGR) is a paraneoplastic disorder that presents as erythematous annular and serpiginous lesions compared with grain of wood. EGR is associated with an underlying malignancy in more than 80% of cases. There is also 'EGR-like eruption' without internal malignancy and it has been increasingly reported recently. First case unassociated with malignancy was described in 1978 by Barber et al. In a patient with pulmonary tuberculosis.

We present a rare case of Urticarial vasculitis with cutaneous lesions resembling EGR, in a patient with hypocomplementemia and incomplete lupus according to ARA criteria.

Case report:

Our patient is 69-year-old woman, presented with a 10 month history of multiple pruritic** urticaroid papular lesion which in a few days formed erythematous annular lesions on her low extremities, body and upper limbs. Initially, the eruption appeared as a small papule forming concentric erythematous rings, resulting in a wood-grain appearance. The individual lesion formed residual hyperpigmentation. She has a history of DM type II, Hypertension, Hypothyroidism and receive appropriate therapy for them. She denied any systemic symptoms at the moment, but at the beginning of the changes refers mild weakness. She came with elevated AFA and D-dimer and she was placed on OAT. An immunological screen identified elevated levels of antiSSA, antiSSB 84, anti U1-snRNP antibodies, ANA ++ 1:160 with no features of systemic lupus erythematosus and low Complement C3 and C4. She had urinary symptoms and we isolated Escherichia coli from the urine culture. Skin biopsy revealed leukocytoclastic vasculitis with a dense neutrophil infiltrate, small numbers of eosinophils and lymphocytes, nuclear dust, and extravasated red blood cells. Laboratory studies revealed the normal blood count and CRP and normal ranges of the tumor markers and virus markers. Chest radiography is without findings. We apart from OAT also gave her a Chloroquine, corticosteroids and antibiotics. The patient was followed up after discharge from our clinic and did not show any additional symptoms or exacerbation of the dermatological status.

Conclusion:

We presented rare case of hypocomplementemic Urticaria vasculitis with erythema gyratum repens - like eruption in a patient with seropositive SLE, but without clinical symptoms of lupus and absence of underlying malignancy.

The obvious diagnosis of Erythema gyratum in this case is hidden in Urticaria vasculitis. Not always what we see is definitive, which makes us dermatologists constantly alert and ready for challenges.

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Abstract N°: 1827

Treatment of Severe Pyoderma Gangrenosum with Adalimumab and Low-Intensity Laser Therapy: A Case Report

Matheus Alves Pacheco* , Ariel Rosa , Monique Schmitz , Mateus Krahl , Iara Bernardy , Athos Martini

Introduction & Objectives:

Pyoderma gangrenosum (PG) is a rare and potentially severe disease characterized by painful ulcerative skin lesions. Treatment of PG can be challenging, and immunobiologic therapy has shown promising results in severe or refractory cases. Low-intensity laser therapy has also been reported as an effective complementary treatment for promoting wound healing. The objective of this case report is to highlight the use of adalimumab therapy and low-intensity laser treatment in a severe case of PG.

Materials & Methods:

A patient with a one-year history of severe pain in the lower limbs and bilateral leg ulcers resulting from a third-degree burn accident was included in the study. Initially, the patient received Ceftriaxone followed by prednisone, colchicine, and ciclosporin. Immunobiologic therapy with infliximab was initiated but discontinued due to a severe adverse reaction. The patient was then switched to adalimumab therapy, with a loading dose and maintenance regimen. Low-intensity laser therapy was performed using a specific laser device, with a combination of red and infrared lasers. The treatment protocol involved photobiomodulation, application of methylene blue, and irradiation with red LED. Dressings were applied using non-branded barrier creams and hydrofiber dressings.

Results:

The patient showed initial clinical improvement with adalimumab therapy but experienced an anaphylactic reaction. Switching to adalimumab successfully continued the treatment. Over the course of one year, approximately 70 sessions of low-intensity laser therapy were performed, resulting in wound healing and improvement of the ulcers. The combination of adalimumab therapy and low-intensity laser treatment showed positive outcomes in managing the severe and refractory PG ulcers.

Conclusion:

This case report demonstrates the potential efficacy of adalimumab therapy and low-intensity laser

treatment in severe and refractory cases of PG. Adalimumab proved effective after an adverse reaction to infliximab, and low-intensity laser therapy contributed to wound healing. These treatment modalities provide valuable options for managing PG ulcers, especially when conventional therapies are not effective. Further research is needed to validate these findings and optimize the use of adalimumab and low-intensity laser therapy in PG treatment.

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Abstract N°: 1908**Online Education Significantly Improved Dermatologists' Knowledge of Vitiligo Management**Elaine Bell¹, Marinella Calle¹, Markus Boehm²¹WedMD Global, 14-17 Market Place, LONDON, United Kingdom, ²Dermatology, Munster, Germany**Introduction & Objectives:**

Vitiligo is an acquired depigmentation disorder of the skin that can have a profound impact on patients' lives. Optimal management of vitiligo can be challenging for clinicians.

We assessed whether online accredited education could improve dermatologists' understanding of the impact of vitiligo and the treatment options for vitiligo, and could enhance their confidence in managing patients with vitiligo.

Materials & Methods:

Dermatologists participated in a 30-minute, chapterized online video activity entitled 'Optimizing Outcomes in Vitiligo Management'. Responses to 3 multiple-choice, knowledge questions and 1 self-efficacy, 5-point Likert scale confidence question were analyzed. Educational effect was assessed using a repeated-pair design, pre-/post-assessment. A paired samples t-test was conducted for significance testing on overall average number of correct responses and for confidence rating. A series of McNemar's tests were conducted at the question level (5% significance level, $P < .05$). Cohen's d with correction for paired samples estimated the effect size of the education on number of correct responses ($< .20$ modest, $.20$ -. 49 small, $.59$ -. 79 moderate, $\geq .80$ large). The activity launched on 6 May 2022 with data collection through 21 November 2022.

Results:

- Dermatologists ($n=67$) showed significant improvements in their knowledge of:
 - the impact of vitiligo on patients' lives (51% correct responses at baseline rising to 84% post-activity; $P < .001$)
 - treatment options for facial vitiligo (46% correct responses at baseline rising to 70% post-activity; $P < .001$)
 - first-line treatment recommendations from the British Association of Dermatologists for achieving repigmentation (60% correct responses at baseline rising to 84% post-activity; $P < .001$)
- The average percentage of correct responses increased from 52% at baseline to 79% post-activity ($P < .001$), and there was a large educational impact (Cohen's $d=0.87$)
- The proportion of dermatologists answering all 3 questions correctly rose from 15% at baseline to 51% post-activity
- After completing the activity, 51% of dermatologists reported improved confidence in optimizing outcomes for patients with vitiligo, resulting in a notable confidence shift of 59%
- 98% of responders felt the education would improve their performance, resulting in improved patient outcomes

Conclusion:

This online activity significantly improved dermatologists' knowledge of the burden of vitiligo and treatment

options for vitiligo. Given the evolving treatment landscape for vitiligo, dermatologists would benefit from additional education to improve their knowledge of emerging therapies and translate this knowledge into clinical practice and improved patient outcomes.

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Abstract N°: 1969

Pemphigoid of the Pulmonary System (POPS): New Clinical Entity

A. Razzaque Ahmed¹, Thomas Kinane²

¹Tufts University School of Medicine, Dermatology, Boston, United States, ²Mass General Hospital/Harvard Medical School, Pulmonary Medicine, Boston, United States

Introduction: Mucous Membrane Pemphigoid (MMP) is an autoimmune mucocutaneous blistering disease involving multiple mucous membranes and skin. The hallmark of this disease is that as the blisters heal there is irreversible scarring, which has devastating and catastrophic sequelae and consequences.

Objective: Describe involvement of lower airway or pemphigoid of the pulmonary systems (POPS) in eleven patients.

Materials & Methods: Eleven patients with MMP involving the trachea and bronchi are described. All had biopsies for histology and direct immunofluorescence which showed immunoreactants deposition at BMZ. Clinical presentation, course of disease, demographics symptoms, bronchoscopic studies, treatments used and clinical outcome were noted.

Results: Mean age at onset was 20 years 82% of patients were under forty years, at the time of disease onset. All patients had three to five mucosa and skin involvement. 100% had oral involvement, 82% had skin and ocular involvement and 36% had anogenital and esophageal involvement. All reported shortness of breath and persistent coughing. Some have coughed up tissue. Bronchoscopy revealed simultaneous active disease (erosions, blisters and tissue sloughing) and scarring. It was the only procedure to make diagnosis. The interval between bronchoscopy and death was a mean of 1.5 years. POPS occurred late in the course of the disease. Several patients required surgical intervention and tracheostomy. Rituximab was the only drug that produced remission in the patients who received it. Mean follow-up was only 11 months, so relapse rates could not be assessed. Mortality rate 45%, entirely disease related, due to respiratory failure, associated with pulmonary infection and sepsis in some patients.

Conclusion: POPS has several features that differentiate it from the usual MMP patients. Hence identifying it as a separate clinical entity is clinically beneficial. Dermatologist should consider pulmonary involvement in MMP patients with respiratory symptoms. Early intervention with bronchoscopy may save lives.



Abstract N°: 1979

Intractable Stomatitis in CLL - a Diagnostic and Therapeutic Challenge

Anika Rajput Khokhar¹, Marisa Klemp¹, Farzan Solimani¹, Rose Moritz¹

¹Charité – Universitätsmedizin Berlin, Department of Dermatology, Venereology and Allergology, Berlin, Germany

Introduction & Objectives:

A 64-year-old patient with known chronic lymphocytic leukemia (CLL) presented in our department with mucocutaneous lesions that had been present for 5 weeks, accompanied by progressive dyspnea, and weight loss. Erosive mucositis manifested orally, genitally and conjunctivally, while the skin presented polymorphous cutaneous eruptions including erosions and partly lichenoid, partly targetoid-like plaques.

Materials & Methods:

A skin biopsy was taken. Histology showed erythema multiforme-like interfacial dermatitis. Serologically, we detected antibodies against skin antigens such as desmoglein 3 (35 U/ml) and BP180 (99 U/ml) by enzyme-linked immunosorbent assay (ELISA); yet direct immunofluorescence (DIF) was negative. Given this peculiar serological constellation and the underlying presence of a CLL we performed an indirect immunofluorescence (IIF) on monkey bladder epithelium, which was positive. Based on these results, we made a diagnosis of paraneoplastic pemphigus (PNP) in CLL.

Results:

The patient received prednisolone iv. 2mg/kg body weight. Due to protracted treatment response, therapy with intravenous immunoglobulins was initiated. Cutaneous but not mucosal manifestations improved. Upon complete evaluation, we extended the therapy with rituximab 1000 mg.

Eventually, a new abdominal lymph node conglomerate was histologically confirmed and evaluated with bone marrow cytology as a progression of the known CLL. With increasing respiratory insufficiency, the patient died of pneumogenic sepsis in intensive care despite escalated anti-infective therapy.

Conclusion:

PNP is a rare paraneoplastic disease that should be considered for differential diagnosis in severe erosive stomatitis. It normally occurs in patients with underlying hematologic malignancies but can also be seen in the setting of solid tumors. The histologic picture is often not very specific, direct immunofluorescence sometimes remains negative, and the particular serology complicates the diagnosis. Autoantibodies to plakins are characteristic, although antibodies to desmogleins, desmocollins, or BP180/230 may also be present. In addition, IIF on monkey or rat bladder epithelium, a tissue rich in plakins, is essential.

An often fatal complication is pulmonary involvement with bronchiolitis obliterans. Even with correct diagnosis and timely systemic therapy, fatal courses can rarely be prevented with a mortality rate of up to 90%.

This didactic case illustrates the clinical presentation of PNP and the diagnostic workout needed to rapidly diagnose this severe skin disease to enable the prompt therapeutic response required.

Abstract N°: 1981**Worldwide Presence of Pemphigoid Diseases: Molecular Explanation for Incidence and Clinical Spectrum**A. Razzaque Ahmed¹, Pedro Reche²¹Tufts University School of Medicine, Dermatology, Boston, United States, ²University Complutense of Madrid, Immunology, Madrid, Spain

Introduction & Objectives: Pemphigoid diseases affect the skin and mucosal tissues and have a large spectrum of clinical presentation. Study possible factors that may partially explain incidence of pemphigoid diseases worldwide and the basis for the clinical spectrum observed using molecular and computer generated models.

Materials & Methods: Data available on HLA studies in patients with immunopathology confirmed diagnosis of bullous pemphigoid (BP), mucous membrane pemphigoid (MMP), and BP secondary to DPP4-i drugs in diabetic patients formed the data base. T-Cell epitopes within BP-180, BP-230, Beta 4 and alpha 6 integrin restricted HLA DQ 7 were predicted by a computer model (RANKEP Server) and their tertiary structure models subjected to MODELLAR energy optimization methods, selecting the best dope scores. Superimposition of tertiary structures and molecular graphic representation obtaining using Pymol molecular graphic system were used to determine presentation of antigen to T-Cell receptor.

Results: 19 studies between 1990 and 2020 were examined. 904 BP patients from seven countries and 335 MMP patients from five countries had HLA DQB1*03:01 in statistically significant incidence compared to age and sex matched controls ($P < 0.00001$). Similar observations were made in DPi4; associated BP in diabetic patients in countries where studied. HLA-DQB1*03:01 presence in overwhelming majority of pemphigoid patients was observed in the US, Europe, and Japan. Several tertiary structures of peptide DQ7 complexes were obtained by homology modeling. The peptides had high affinity binding to HLA DQB1*03:01 for T-Cell presentation.

Conclusion: Pemphigoid diseases occur worldwide because of a common MHC Class II gene association, in patients in many countries, with different ethnic and racial backgrounds. Clinical phenotype may depend on which peptide amongst the putative antigens is presented to the T-cell receptor, via the MHC Class II genes. The peptide used may determine the clinical phenotype.



Abstract N°: 1983

IgM Deficiency in Autoimmune Blistering Disease: Treated and Untreated Patients and long-term follow-up

A. Razzaque Ahmed¹, Merve Aksoy²

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Introduction & Objectives: IgM has been considered to play a role in autoimmunity and cancer for several decades. IgM deficiency has been reported with rituximab treatment and in many autoimmune diseases.

Study IgM levels in patients with several autoimmune blistering diseases and follow them post recovery to determine the persistence or recovery of IgM deficiency. Patients were also studied before initiation of any systemic therapy and those on systemic therapy.

Materials & Methods: 403 patients were screened for IgM deficiency with pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid, and mucous membrane pemphigoid and epidermolysis bullosa. Treatments included rituximab and oral prednisone, dapson, methotrexate, azathioprine, mycophenolate mofetil and cyclophosphamide. Follow-up for IgM deficiency was done at every 3-4 months interval for a mean of 36 months (24-54). 55 patients with same diagnosis on initial evaluation were studied for IgM deficiency. In all patients of serum IgA, IgG and B-cells were measured, throughout study period.

Results: Of the 403 patients, 279 patients had IgM deficiency. The level of deficiency was comparable in different diseases. Treatment used did not influence the level of IgM deficiency. Untreated patients the level of IgM deficiency was similar to those under treatment. The follow-up period did not influence IgM recovery. Serum IgA, IgG and B-Cell levels were always normal.

Conclusion: This study provides preliminary evidence that IgM deficiency may have a role in pathogenesis and clinical course of autoimmune blistering disease. This affect can occur in the bone marrow where check points are involved in B-cell development. Its specific role and influence warrants methodical and detailed investigation, because it may be a model to study other autoimmune diseases and cancers.

Abstract N°: 2105**Safety, Pharmacokinetics, and Pharmacodynamics of IMG-004, a Selective Reversible Bruton's Tyrosine Kinase Inhibitor in Healthy Participants**Zi Lin¹, Yancong Shen¹, Chi-Yuan Wu², Run Lei¹, Yu Wang³, Aswin Nair⁴¹Imagene Biopharmaceuticals Technology (Shanghai), China, ²Imagene LLC, United States, ³Imagene Biopharmaceuticals (Hangzhou), China, ⁴MeDaStats LLC**Introduction & Objectives:**

Bruton's tyrosine kinase (BTK) is a clinically validated therapeutic target for the treatment of autoimmune disorders (e.g., chronic spontaneous urticaria). Newer generation non-covalent and reversible BTK inhibitors would potentially enhance the kinase selectivity and reduce off-target effects which would theoretically improve the safety profile for long-term treatment. IMG-004 is a highly selective, reversible, non-covalent BTK inhibitor. Here we report the results from a first-in-human single-dose study that aims to evaluate the safety, pharmacokinetics (PK), and pharmacodynamics (PD) of IMG-004 in healthy participants.

Materials & Methods:

The study was a single ascending dose design with five oral dose cohorts of IMG-004 or placebo at 30 mg to 600 mg (n=8, 6 active:2 placebo per cohort). Adverse events, laboratory parameters, vital signs, electrocardiograms, and plasma/urine PK were assessed. The PD effect of IMG-004 was evaluated using an ex vivo basophil activation assay expressed as percent of CD63+ basophils.

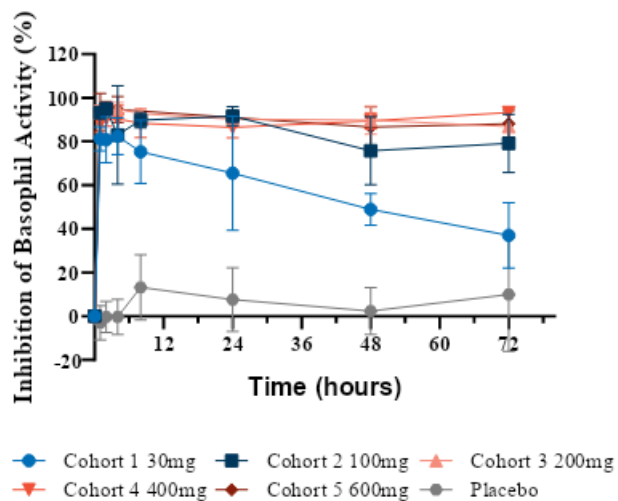
Results:

IMG-004 was well tolerated with no dose-limiting or serious treatment-emergent adverse events (TEAE). All TEAEs reported were of mild intensity. IMG-004 was rapidly absorbed following the oral administration with peak concentrations occurring around 2 hours post-treatment and was eliminated mainly through the hepatic pathway with a mean terminal half-life ranging from 26.0 to 37.0 hours. Based on the geometric means reported, the AUC_{0-inf} of IMG-004 appeared to increase in a dose-proportional manner from 30 mg to 600 mg, while C_{max} appeared to increase in a dose-proportional manner from 30 mg to 400 mg, and less than dose-proportional manner from 400 mg to 600 mg. IMG-004 demonstrated robust and dose-dependent target engagement in the ex vivo basophil activation assay. A maximum (approximately 95%) inhibition of basophil activation was observed at the dose levels between 100 mg and 600 mg, with sustained inhibition (80%-90%) being observed up to 72 hours post-treatment (Figure 1).

Conclusion:

IMG-004 was well tolerated at doses up to 600 mg. The long half-life and sustained PD effect support further exploration of efficient and convenient dose regimens in future clinical development. IMG-004 represents a promising potential therapeutic agent for the treatment of autoimmune diseases.

Figure 1 Mean (±SD) Inhibition of Basophil Activity Over Time



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Abstract N°: 2118**Overlap syndrome with skin involvement**

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Introduction & Objectives:

Overlap syndromes are diagnosed in patients who experience clinical features and laboratory findings of multiple autoimmune diseases. This is a challenging case of a patient with a fifteen-year history of rheumatoid arthritis who was diagnosed with both systemic lupus erythematosus (SLE) and dermatitis herpetiformis (DH). Sixteen years later she was also diagnosed with Sjogren's Syndrome.

Materials & Methods:

A 49-year-old woman with a fifteen-year history of rheumatoid arthritis (with positive rheumatoid factor), presented with a polymorphous exanthema consisting of vesiculobullous lesions, erythematous plaques and crusted skin lesions. The lesions were initially limited to the face, superior thorax and superior limbs, with a later extension on the abdomen and inferior limbs. The patient described severe pruritus and arthralgia involving the knees and small joints of the hands and feet. She has also been experiencing recurrent diarrhea. Blood tests and biopsies from a vesiculobullous lesion and perilesional skin were obtained. Sixteen years later she accused xerostomia, xerophthalmia and was referred to otorhinolaryngology and ophthalmology department for a multi-disciplinary approach. A minor salivary glands biopsy was performed.

Results:

The blood work revealed an increased erythrocyte sedimentation rate, leukopenia with neutropenia and lymphopenia. The immunological profile was specific to lupus, with positivity for the antinuclear antibody, the anti-double-stranded DNA, anti-Ro/SSA and anti-La/SSB antibodies. The patient met 5/11 of the 1997 Updated American College Rheumatology classification criteria for SLE. She was tested negative for anti-U1RNP antibody hence Sharp Syndrome was excluded. The path report showed a subepidermal bulla with a collection of predominant eosinophilic infiltrates into the dermal papillae. Direct immunofluorescence (DIF) revealed a lupus band in IgM and IgG and a granular IgA deposition on top of dermal papillae. The patient was diagnosed with SLE and DH. A gluten-free diet was recommended and therapy with dapsone and hydroxychloroquine was initiated. After six months dapsone was removed and the favorable clinical outcome was maintained. The Schirmer II test was positive and the minor salivary glands biopsy was suggestive of a salivary component of Sjogren's syndrome.

Conclusion:

This rare association of diseases made the case challenging. The patient had mildly articular symptoms since her rheumatoid arthritis diagnosis and the presence of rheumatoid factor in LES patients is not uncommon, therefore her articular symptoms could have been the first clinical manifestation of lupus. The initial clinical presentation with bullous lesions required a differential diagnosis between bullous systemic lupus erythematosus and a primarily blistering disease. Although the presence of predominant eosinophilic infiltrates into the dermal papillae on the path report was more specific for bullous pemphigoid, DIF established the accurate diagnosis, having a sensitivity and specificity close to 100% for DH. All new symptoms in a patient with an autoimmune field should be carefully investigated for other autoimmune diseases and long-term, multi-disciplinary management is mandatory.

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Abstract N°: 2184**Chronic cutaneous lupus erythematosus and paraneoplastic dermatomyositis - Report of an unusual overlap syndrome**

Kaique Arriel*¹, Daniel Simao de Andrade¹, Thais Kohatsu Yanase¹, Ana Clara Maia Palhano¹, Thaisa Bosquiroli Brandalize¹, Dimitri Luz Felipe Da Silva¹, Artur Duarte¹

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Introduction & Objectives:

Lupus erythematosus (LE) and dermatomyositis (DM) are examples of the connective tissue diseases (CTD), autoimmune disorders that occur with a chronic inflammatory process directed at the connective tissue. Although isolated cutaneous involvement is more frequent, these diseases can have severe systemic involvement. Currently the CTD are classified using clinical and serological criteria, but in clinical practice it is common to see patients who do not meet criteria for a specific form of CTD (undetermined connective tissue disease), as well as patients who have clinical manifestations of 2 or more CTD, characterizing the overlap syndrome. The most frequent overlap syndromes involve the association of the several CTD with Sjogren's syndrome.

We intend to report an unusual case of overlap between chronic cutaneous lupus erythematosus (CCLE) and paraneoplastic dermatomyositis (PNDM).

Case report:

A woman, 52-year-old, who had been followed up at a dermatology outpatient clinic for 20 years due to CCLE, leading to cicatricial alopecia of approximately 50% of the scalp. The patient had been in relative control of the disease over the past 5 years, making regular use of hydroxychloroquine and azathioprine. In the last year, she presented worsening of lesions on the scalp, appearance of erythematous-scaly plaques on the upper limbs and back, facial erythema, heliotrope, Gottron's papules and paresis in the proximal muscles of the limbs. At this time, the patient was being investigated for a lump in the left breast, which was later confirmed to be a breast cancer with pulmonary metastasis. In view of the worsening of the CCLE, the appearance of lesions characteristic of DM and the diagnosis of breast cancer, azathioprine was suspended and prednisone 40mg/day was prescribed; new laboratory tests were requested; and a new skin biopsy was performed. Laboratory tests showed leukopenia, increase in creatine phosphokinase, polar dotted cytoplasmic pattern reactive ANA 1/160 and reactive anti-cardiolipin antibody. Biopsies showed images compatible with the diagnostic hypothesis of DM. In view of the clinical, laboratory and histopathological findings, the diagnosis of overlap syndrome with association of CCLE and paraneoplastic dermatomyositis (PNDM) was confirmed. After 15 days using 40 mg/day of prednisone, the patient already showed significant improvement in the skin lesions. Subsequently, after the initiation of chemotherapy, prednisone was gradually withdrawn and the patient has been in control of the condition since then.

Discussion:

DM has been found as a paraneoplastic manifestation in 15 to 30% of adult patients with this CTD. There are reports in the literature of association with various neoplasms such as hematological, breast, colorectal, lung, ovarian and renal cancers. The overlap syndrome with association between LE and DM is a rare entity, difficult to diagnose and with few reports in the literature. The presence of specific cutaneous lesions, myopathy and specific autoantibodies (anti-Jo1) can help in the diagnosis of DM. We emphasize here that, in patients with CTD, the worsening of the condition or the change in the pattern of manifestation of the disease always indicates the need

to carry out a new investigation, with new serologies, screening for infections and neoplasms. The diagnosis of DM in adults is a factor that alone determines the need for neoplastic screening due to its high association with malignancies.

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Abstract N°: 2237

identification of immunological factors predicting clinical relapse in pemphigus- a prospective bicontinental study

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Introduction & Objectives: There is a paucity of prospective studies to identify immunological parameters that can predict clinical relapse in pemphigus vulgaris (PV) and foliaceus (PF). The aim of this bicontinental study was to periodically assess immunological parameters in patients with pemphigus in remission so that immunological events preceding clinical relapse are better understood.

Materials & Methods: 105 patients with PV/ PF (India, n= 75; Bulgaria, n=15; Greece, n=15) either in complete remission off therapy (n=41) or on minimal therapy (n=64), were included. At baseline, clinical history was assessed and immunological investigations including direct immunofluorescence (DIF), serum IgG against desmoglein (Dsg) 1, IgG, IgG1, and IgG4 against Dsg 3, IgG against the extracellular domains 1 and 2 of Dsg 3, IgG against muscarinic (M3)-AChR, and peripheral CD19+CD27+ memory B cells/plasma cells were performed by ELISA and by flow cytometry, respectively. These investigations were repeated at an interval of 3 months up to 12 months (up to 24 months in some patients) or until relapse, whichever was earlier. DIF was repeated at month 12 and at the time of relapse.

Results: 29 of 105 patients (28%) experienced a relapse. Anti-Dsg 1 at baseline was positive in a significantly higher proportion of relapsed compared to non-relapsed patients. Compared to the immunological parameters assessed at the visit immediately preceding relapse, a significant increased number of patients with positive anti-Dsg1 (38% vs 31.1%, p=0.01), anti-Dsg3 (51.7% vs 41.4%, p=0.01) as well as IgG positivity by DIF (85.7% vs 25%, p<0.001) was observed at the time of relapse.

Conclusion: Regular monitoring of anti- Dsg 1 and anti-Dsg 3 serum levels during the course of the disease in particular when remission has already occurred may predict relapse. Performing a DIF before complete tapering-off of pemphigus medication appears to be advisable.

Acknowledgement: The study was supported by the European Academy of Dermatology and Venereology under project proposal research funding (2017; to D.D.) and structural funding through the Excellence Cluster 2167 *Precision Medicine in Chronic Inflammation* (to E.S.)

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Abstract N°: 2271**Muscle biopsy features of dermatomyositis**Elyamani Lamis¹, Zerrouki Nassiba¹, Zizi Nada¹, Dikhaye Siham¹¹Department of Dermatology, Venereology and Allergology, CHU Mohammed VI, Oujda, Dermatology, Oujda**Introduction & Objectives:**

Dermatomyositis (DM) is an idiopathic inflammatory myopathy (IIM). Histopathologic features on muscle biopsy are very important criteria for establishing the diagnosis as histology characterizes each subtype of IIM. The aim of this study is to describe the histopathological features of muscular biopsy in dermatomyositis' patients.

Materials & Methods:

We retrospectively reviewed the case records of 27 patients diagnosed with dermatomyositis from June , 2014 to June, 2022 in the Dermatology and Internal medicine departements of Mohammed VI Hospital in Oujda city.

Results and discussion:

Twenty seven patients were included, with a mean age of $49,5 \pm 21$ years. Twenty patients were female and seven patients were male. Histopathological features found in our study were mainly perifascicular atrophy in 68% and inflammatory infiltrate in 68%;the inflammatory infiltrate was perivascular in 64% and perimysial in 52%; the endomysial location of the inflammatory infiltrate was not noted in any patient. Vacuolar degeneration in 48%, interstitial fibrosis 40%, loss of double striation 40%, muscle fiber necrosis 24%, muscle fiber regeneration 20%, vitreous necrosis 16%, and microangiopathy 4%. The main pathological hallmark of DM is perifascicular atrophy (PFA) characterised by atrophic, degenerating and regenerating fibers at the periphery of the fascicle that may affect two to ten layers and strongly support a diagnosis of DM, even in the absence of inflammation, wish is consistent with the results of our study where perifascicular atrophy was the most frequent histopathological feature. However, PFA is a late finding and is found in only 50% of adult cases when biopsied early in the course of illness. However, the ENMC criteria indicate PFA as the diagnostic feature on muscle biopsy for definite diagnosis of DM. In DM, inflammatory infiltrates occur predominantly at perivascular sites or within the interfascicular septae , rarely in the endomysium wish is consistent with our results. Indeed, in our study, the inflammatory infiltrates was perivascular and perimysial. The endomysial location of the inflammatory infiltrate was not noted in any patient. An early histological feature is the involvement of intramuscular blood vessels; the angiopathy is characterized by the deposition of immunoglobulins and complement that cause a reduction in number of capillaries with endothelial hyperplasia. In our study, the angiopathy was the less frequent histopathological feature. This could be explained by the delayed diagnosis, since angiopathy represents an early histological feature that disappears during the course of the disease.

Conclusion:

Histopathological features of muscular biopsy are very useful for differentiating dermatomyositis from other IIM. However, clinical and immunological features in association with muscle biopsy features are very important to establish the right diagnosis

Abstract N°: 2317**Complete response of multicentric reticulohistiocytosis treated with tofacitinib.**

Júlia Mercader Salvans^{*1}, Maria Arteaga-Henriquez¹, Judit Algarra Sahuquillo¹, Luísa Santos e Silva Caldeira Marques¹, Miguel Quetglas Valenzuela¹, Maria Mar Pestana Eliche¹, Eva Fagundo Gonzalez¹, Vania Alexandra Lukoviek Araya¹, Rosa Nieves Rodríguez Rodríguez², Sagrario Bustabad Reyes³, Francisco Guimera¹

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Introduction & Objectives:

Multicentric reticulohistiocytosis (MRH) is a rare systemic disease caused by an inflammatory histiocytic infiltrate which primarily affects the skin and articular synovial. It is considered a therapeutic challenge because of its low incidence and multiple treatments used with varying outcomes.**

Materials & Methods:

Case Report: A 42-year-old woman diagnosed with Sjögren's syndrome presented with a 6-month history of pruritic nodular lesions. For that reason, she had consulted a private dermatologist and had been prescribed chloroquine and prednisone. Physical examination revealed bilateral yellowish erythematous papulonodules in her dorsal hands. A skin biopsy was performed, showing a dermal infiltrate of mononuclear and multinucleate histiocytes with an eosinophilic, finely granular 'ground-glass' cytoplasm. On immunohistochemistry, the cells expressed CD68, being negative for S-100, confirming the diagnosis of MRH. Hematological, biochemical, and serological tests were normal, and a malignant neoplasm-association was excluded. Methotrexate was started but the patient developed hypertransaminasemia after 2 weeks. She was then prescribed adalimumab but skin lesions continued progressing and the patient developed disabling joint pain, even after intensified treatment. Finally, tofacitinib 5mg/12h was started.

Results:

A progressive joint pain improvement was noticed and pruritus and skin lesions were completely resolved.**

Conclusion:

MRH is a non-Langerhans cell histiocytosis of unknown etiology. Erosive arthritis mimicking rheumatoid arthritis, erythematous nodules on the face and arms, together with erythematous patches in photoexposed areas are the main clinical manifestations. About 20-30% of MRH are associated with a malignant neoplasm. First-line treatment is based on oral corticosteroids associated with methotrexate or other immunosuppressive drugs. In refractory cases, anti-tumor necrosis factor alpha agents, such as adalimumab, have been reported with variable results. Recently, isolated cases of patients treated with JAK inhibitors (JAKi) with successful outcomes have been published.

We present a patient with MRH successfully treated with tofacitinib. JAKi are presented as an effective treatment alternative for patients with MRH.

Abstract N°: 2342**A Case of Dyshidrosiform Pemphigus in Accompany Guillain-Barré Syndrome and Nail Involvement**Ezgi Akin¹, Emel Bulbul Baskan¹, Şaduman Balaban Adım²¹Bursa Uludag University, Dermatology and Venereology, ²Bursa Uludag University, Pathology**Introduction & Objectives:**

Pemphigus vulgaris (PV) is an autoimmune blistering disease. Pemphigus vulgaris may occur anywhere on the skin surface as a primary lesion.(1) In this case report we present the uncommon clinical presentation of pemphigus vulgaris as dyshidrosiform eruption on the feet and nail involvement.

Materials & Methods:

A 51-year-old female patient presented with a 3 year intermittent, draining lesions in her hands, nails and feet. The patient had a past history of Guillain-Barré disease in 2018, diabetes, hypertension, chronic kidney disease. First she was treated with topical corticosteroids and antibiotics as well as multiple oral antibiotics. She had a partial remission with this treatment but soon recurred with accompanying severe pain and pruritus. The patient was admitted to the dermatological ward. On dermatologic examination she had foul smelling, purulent draining plaques with crust, scale and erythema on her both feet, toes and soles. Also she had subungal hyperkeratosis, yellow green discoloration in her toenail and right fingernail. Mucous membranes were not involved. Laboratory studies revealed mild elevation of C-reactive protein. Swab culture and tissue culture taken from the feet isolated Staphylococcus aureus. X-rays of the feet did not reveal evidence of osteomyelitis.

Results:

Because of the isolation of Staphylococcus aureus the patient was given a 14-day course of piperacilin-tazobactam and teicoplanine. Topical antiseptic dressings, cream including corticosteroids, antibiotics and mousturizers were applied daily. Direct microscopic examination of the nails revealed dermatophyte hyphea and she was treated with terbinafine. Histology of the skin taken from the soles revealed suprabasal dermatosis with acantholysis. Direct immunofluorescence showed intercellular deposits of IgG and C3c within the epidermis. These findings were compatible with pemphigus vulgaris. After 14 days treatment she had a remission.

Conclusion:

Although the classic clinical picture is generalized with mucosal involvement, localized and unusual presentations have been described in literature . Dyshidrosiform pemphigus, however, is rarely seen. Only 3 case report seen in literature. (2) This is the first case without previos history of pemphigus.

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Abstract N°: 2585

BMS-986326 is a novel IL-2/CD25 fusion protein that induces highly selective and durable expansion of regulatory T cells in vitro and following single doses in healthy volunteers

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Introduction & Objectives:

Low-dose interleukin (IL)-2 has shown signs of clinical efficacy in immune-mediated diseases via expansion of regulatory T cells (Tregs), which are essential in maintaining immune tolerance.¹ BMS-986326 is an investigational IL-2/CD25 fusion protein designed for greater Treg selectivity and a longer half-life through slow dissociation from an inactive, non-covalent homodimer to active monomers. We report results from an in vitro study of BMS-986326 in whole blood from healthy volunteers (HVs) compared with patients with immune-mediated disease, and an ongoing first-in-human single-ascending dose (SAD) study to evaluate the safety, pharmacokinetics (PK), and pharmacodynamics (PD) of BMS-986326.

Materials & Methods:

Treg IL-2 receptor (IL-2R) proximal signaling was evaluated by measuring phosphorylated STAT5 (pSTAT5) after 15 min stimulation of whole blood ex vivo with BMS-986326 from both HVs and patients with immune-mediated disease (eg, systemic lupus erythematosus [SLE], atopic dermatitis). Total CD25 staining and pSTAT5 induction potency (% positive cells) in Tregs were compared between HVs and patients with SLE.

In a phase 1 SAD study (NCT04736134), HVs were randomized to receive either a single dose of IV BMS-986326 or placebo (PBO; 6 treated and 2 PBO per dose). AEs were monitored over a 55-day follow-up. Blood samples were collected at prespecified times for PK/PD analyses (Treg, T conventional [Tcon], and CD8+ T-cell count [106/L], and pSTAT5 % in Tregs). A power model assessed dose-proportionality.

Results:

In vitro, mean (SD) Treg half maximal effective concentrations of BMS-986326 for the induction of pSTAT5 were 0.72 (0.37) nM for HVs (n = 12) and 1.4 (1.1) nM for patients with SLE (n = 11). More patients with SLE had a CD25-low Treg profile, indicating CD25 deficiency, than HVs (median [SD], 56% [16%] and 43% [10%], respectively). BMS-986326 induced pSTAT5 with a comparable maximal signal and an approximately 2-fold reduction in potency in Tregs from patients with SLE vs HVs.

In the SAD study, 32 male HVs aged 19–50 years were randomized to PBO or 1 of 4 ascending BMS-986326 doses (A, B, C, D). All AEs were mild/moderate; 13 (54%) and 5 (50%) HVs experienced AEs when treated with BMS-986326 across doses or PBO, respectively. Eosinophilia was seen in HVs receiving doses C or D. PK data showed linear dose-proportional increases in BMS-986326 exposure (half-life: 4–6 days). There were dose-dependent increases in Treg expansion; doses A, B, C, and D showed peak mean fold change (SE) vs baseline of 1.5 (0.3 [day 28]), 1.8 (0.3 [day 11]), 2.6 (0.3 [day 28]), and 5.4 (1.0 [day 15]), respectively. These increases were maintained by all HVs until the last time point (day 55): mean fold change (SE) of 1.1 (0.2), 1.5 (0.3), 1.4 (0.2), and 2.7 (0.1) for doses A, B, C, and D, respectively. All doses showed increases in pSTAT5 % in Tregs vs baseline following a single dose of BMS-986326. Minimal changes on Tcon and CD8+ T-cell populations were observed.

Conclusion:

In whole blood from patients with immune-mediated disease, BMS-986326 induced comparable maximal IL-2R signaling to that in blood from HVs.

In a SAD study in HVs, BMS-986326 was generally well-tolerated with robust, selective, durable, and dose-dependent Treg expansion. Further clinical studies are warranted to investigate how BMS-986326 may restore immune homeostasis by Treg expansion in patients with immune-mediated dermatologic diseases.

Reference:

1He J, et al. *Ann Rheum Dis* 2020;79:141–9.

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Abstract N°: 2613**The anti-interferon-beta neutralizing antibody PF-06823859 significantly improves molecular and histological profiles in the skin of patients with dermatomyositis in a 12-week study**

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Introduction & Objectives:

Type 1 interferon (IFN) driven gene expression is elevated in the skin and blood of patients (pts) with dermatomyositis (DM) and serum IFN β protein levels correlate with disease activity. PF-06823859 is a potent, selective, humanized immunoglobulin G1 neutralizing antibody against human IFN β . We assessed the pharmacological and clinical effects of PF-06823859 by measuring IFN activity in skin and blood, and assessed the relationships between IFN-induced gene expression, histological scores, and clinical outcomes in adults with DM.

Materials & Methods:

This double-blind, placebo (PBO)-controlled, Phase 2 study (NCT03181893) enrolled pts (18–75 years) with moderate-to-severe DM. Pts with skin disease predominant (SP) DM (Cutaneous Dermatomyositis Disease Area and Severity Index activity [CDASI-A] score ≥ 14 ; had failed ≥ 1 standard of care systemic treatment), were randomized to PBO, intravenous (IV) PF-06823859 150 mg or 600 mg. Pts with muscle disease predominant DM met one of these inclusion criteria: (1) Manual Muscle Testing (MMT-8) $\leq 136/150$ and Physician Global Assessment (PhGA) ≥ 3 cm on a 0–10 cm visual analogue scale (VAS) or (2) sum of PhGA, Patient Global Assessment, Extramuscular Global Assessment ≥ 10 cm (each using 0–10 cm VAS), with refractory disease, and were randomized to PBO or IV PF-06823859 600 mg. Change from baseline (CFB) at Week (Wk) 12 in a 13-gene IFN gene signature (13GS) from lesional skin (SP cohort) and blood (both cohorts) was evaluated by linear mixed model with CFB modeled by treatment (skin) or treatment, visit, and their interactions (blood), adjusted by baseline (BL) 13GS. Gene signature improvement (GSI) at Wk 12 in lesional skin was calculated from the CFB of 13GS in lesional skin divided by the averaged difference of BL lesional and non-lesional skin (SP cohort). Histological scores (SP cohort) were assessed by fitting a cumulative link mixed model by treatment, visit, and interactions. Correlations between BL 13GS and BL CDASI-A, and between BL histological scores and BL CDASI-A (SP cohort) were assessed by fitting a linear model.

Results:

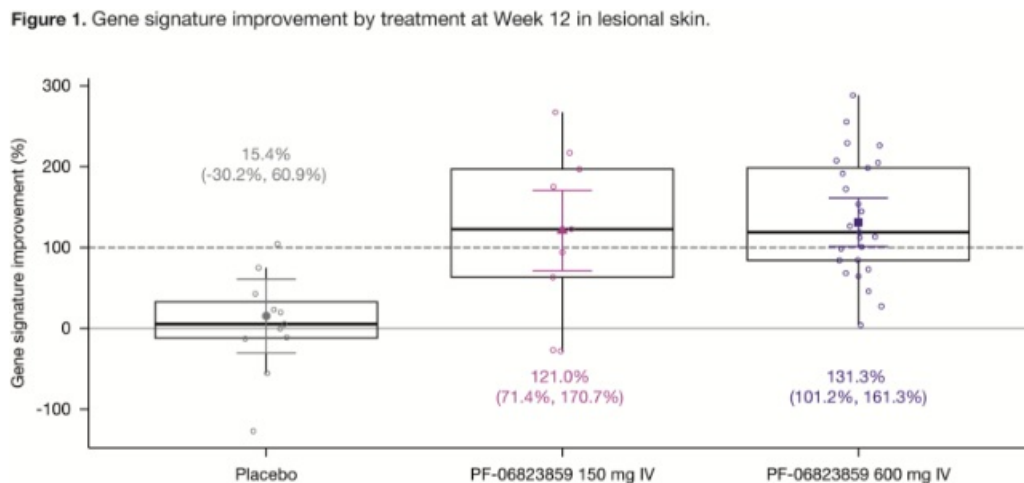
Samples (skin: 11, 26, 11; blood: 13, 37, 22 in PF-06823859 150 mg, 600 mg and PBO, respectively) from 72 pts were included. CFB at Wk 12 in 13GS from lesional skin was greater for PF-06823859 150 mg (-80.5%; $p < 0.001$) and 600 mg (-83.0%; $p < 0.001$) groups than PBO (-18.7%; $p = 0.250$), indicating pharmacodynamic activity. Similar but weaker trends were seen in blood for PF-06823859 150 mg (-38.2%; $p = 0.003$) and 600 mg (-16.5%; $p = 0.038$) groups, with no significant changes for PBO (-5.5%; $p = 0.661$). GSI at Wk 12 in lesional skin indicated recovery of 13GS vs averaged BL levels with PF-06823859 150 mg (121.0%; $p < 0.001$) and 600 mg (131.3%; $p < 0.001$) but not

PBO (15.4%; $p=0.250$; Fig. 1). PF-06823859 600 mg showed PBO adjusted statistical significance in CFB at Wk 12 in acute disease activity assessment ($p=0.012$), dyskeratotic keratinocytes ($p=0.012$) and T cell infiltrate by CD3 immunohistochemistry (IHC; $p=0.033$), indicating histological improvements. At BL, 13GS values in skin and blood correlated with CDASI-A. BL histological scores for acute disease activity assessment, dyskeratotic keratinocytes and T cell infiltrate by CD3 IHC also correlated with CDASI-A (all $p<0.01$).

Conclusion:

IFN β neutralizing antibody PF-06823859 improved molecular and histological outcomes in pts with DM after 12 wks. These findings support further investigation of IFN β as a treatment target in pts with DM.

Figure 1. Gene signature improvement by treatment at Week 12 in lesional skin.



13GS, 13-gene IFN gene signature; IFN, interferon; IV, intravenous. The 13GS include CMPK2, CXCL10, ESPT11, HERC5, IFI44, IFI44L, IFI6, IFIT1, IFIT3, ISG15, MX1, OAS3, and RSAD2 genes, and was assessed per sample by averaging the log2 (counts per million) across these 13 genes. n=11 for placebo; n=9 for PF-06823859 150 mg; n=25 for PF-06823859 600 mg. Gene signature improvement is defined as the negative change from baseline of the 13GS in lesional skin divided by the averaged difference of baseline lesional and non-lesional skin. The dashed line at 100% gene signature improvement marks full recovery of the 13GS from lesional skin to the averaged baseline non-lesional level. Box whisker plots represent the median and quantiles of the overlaid datapoints (circles) of each group. Coloured error bars indicate 95% confidence intervals calculated from the fitted linear model.



Abstract N°: 2617

Interventions for bullous pemphigoid (Cochrane Review)

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Introduction & Objectives: Bullous pemphigoid (BP) is the most common autoimmune blistering disease. Oral steroids are the standard treatment. We updated the Cochrane review first published in 2002.

To assess treatments for BP.

Materials & Methods: We updated searches of the following databases to November 2021: Cochrane Skin Specialised Register, CENTRAL, MEDLINE and Embase. We searched five trial databases to January 2022, and checked the reference lists of included studies for further references to relevant randomised controlled trials (RCTs). Our selection criteria were RCTs of treatments for immunofluorescence-confirmed BP.

Results: We identified 14 RCTs (1442 participants). The main treatment modalities assessed were oral and topical steroids and the oral anti-inflammatory antibiotic doxycycline. Mortality was reported in most studies but adverse events and quality of life (QoL) were not well reported.

Almost all studies involved different comparisons, and two were placebo-controlled. The results are therefore based on a single study for each comparison except azathioprine. Most studies involved small numbers of participants. We assessed the risk of bias for all key outcomes as having some concerns or high risk, due to missing data, inappropriate analysis, or insufficient information.

Clobetasol propionate (CP) cream applied over whole body vs oral prednisone (P) probably increases skin healing at day 21. At 21 days (d), skin healing was seen in 99% of the CP group and 92% of the P group. Death at 1 year (y) occurred in 27% assigned to CP and 36% assigned to oral p. CP cream may reduce risk of severe complications by d 21 vs oral P.

Mild regimen of topical CP vs the standard regimen probably does not change skin healing at d 21. Mortality at 1 y (38%) is similar with both regimens.

Doxycycline (D) (200 mg/d) vs P (0.5 mg/kg/d) induces less complete skin healing at 6 weeks (74% vs 91% patients). D vs P probably decreases mortality at 1 y (2.4% vs 10%). D vs P improved QoL at 1 y. D vs P probably reduces severe or life-threatening treatment-related adverse events at 1 y.

It is unclear whether nicotinamide plus tetracycline vs P affects skin healing or mortality because there is only very low certainty evidence from 1 small RCT. Fewer adverse events were reported in the nicotinamide group.

It is unclear whether azathioprine (A) plus P vs prednisone affects skin healing or mortality because there is only very low certainty evidence from 2 RCTs. Adverse events were reported in a total of 42% participants assigned to A plus P and 34% assigned to P.

It is unclear whether A plus methylprednisolone vs dapsone plus methylprednisolone affects skin healing or mortality because there is only very low certainty evidence from 1 RCT.

Conclusion: CP cream applied over the whole body is probably similarly effective and may cause less mortality than oral P when treating BP. Lower dose CP cream applied over the whole body is probably similarly effective

compared with standard dose CP cream and has similar mortality. D is less effective but causes less mortality than P for treating BP.

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Abstract N°: 2640**A Social Media Analysis of Pemphigus**Gaurav Pathak^{1, 2}, Rithi Chandy^{1, 2}, Vidisha Naini^{1, 2}, Shazli Razi³, Steven Feldman^{2, 4, 5, 6}

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Title: A Social Media Analysis of Pemphigus**Introduction & Objectives:**

The increased use of social media platforms has allowed for greater outreach and discussion of medical information to the public. Pemphigus is a rare spectrum of autoimmune mediated blistering diseases that are associated with diagnostic delays, indicating a greater need for awareness of symptoms. We aim to assess the frequency and manner in which pemphigus is discussed on social media and to characterize the sources generating content.

Materials & Methods:

To identify the social media footprint of pemphigus, the popular social media applications Instagram, Facebook, Twitter and YouTube were searched in April 2023. The common search phrase #pemphigus was queried on Facebook, Twitter and Instagram and YouTube. Information regarding support groups was ascertained on Facebook groups. Information regarding the sender, level of engagement, and type of content (educational, promotional, personal, or recruitment) was analyzed.

Results:

The majority of the YouTube content analyzed (60%) was created by physicians and 100% of content was educational. Instagram had a high proportion of non-human pemphigus content (49%), and organizations were the most common publishers (76%). Physicians contributed 50% of Twitter pemphigus content with the highest engagement, with very few patient-made tweets (6%) and 12% of content was for non-human pemphigus. The majority of Facebook support groups were for pemphigus vulgaris (80%) and most Facebook pemphigus posts were for general awareness.

Conclusion:

Current use of social media for pemphigus revolves around educational content and establishing support systems. There is a need for more human-centered pemphigus content by trusted healthcare professionals and more patient made content. There were some support groups for pemphigus vulgaris, but very few support groups for pemphigus foliaceus and other subtypes.

Abstract N°: 2655**Inflammatory cytokines and the interaction with sleep quality: an investigation on immune aspects between vitiligo and sleep**

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Introduction & Objectives: Vitiligo is a chronic autoimmune skin disease, with a great impact on the quality of life of the patients, which can interfere with their sleep quality. Sleep, in turn, influences the immune system, and regulates inflammatory cytokines, that have a crucial role in the pathogenesis of vitiligo. The interaction between vitiligo and sleep, therefore, may have a bidirectional pathway. Pro-inflammatory cytokines, such as interleukin (IL)-6 and IL-17A, and the anti-inflammatory IL-4 and IL-10 have been described as substrates with an interaction with the circadian rhythm, and with relevant role in the vitiligo immunopathogenesis. The objectives were to analyse cytokine profiles of a vitiligo and a control samples; to compare these data between groups; and to verify the occurrence of any association between these substrates and sleep patterns.

Materials & Methods: The study enrolled 30 patients with vitiligo attending in a Dermatology Service, and 35 individuals comprising a control group. Quality of life and sleep questionnaires were assessed: the Dermatology Life Quality Index (DLQI), the Short-Form Health Survey (SF-36), the Pittsburgh Sleep Quality Index (PSQI) and the Insomnia Severity Index (ISI). Blood collection was performed to measure 7 cytokines through a 7-plex kit: interferon- γ (INF- γ), IL-4, IL-6, IL-10, IL-12 p40, IL-17A and tumour necrosis factor- α (TNF- α). For statistical analysis, ANOVA test was used to compare the cytokine levels between groups; and Generalized Linear Model to analyse the effect of cytokines in the sleep scores of both sleep questionnaires, and in patients with vitiligo treated with ultraviolet B narrow band (UVB-NB) phototherapy.

Results: The PSQI total score and sleep efficiency (PSQI domain 4) were statistically worse in the group with vitiligo. Total PSQI > 5 suggests poor sleep quality; the PSQI total means were 9.07 (vitiligo group) and 6.93 (control group) ($p=0.01$). The final scores of the SF-36 and ISI were numerically worse in the vitiligo sample, although the statistical differences were not significant between groups. The SF-36 domain "body pain" was worse in the vitiligo sample ($p=0.04$). The DLQI mean score in the vitiligo group indicated a classification between mild and moderate impact on quality of life (5.57). The cytokine levels were not statistically different between groups, nor in the analysis with PSQI scores. The analysis of the cytokines with ISI, in turn, showed that higher scores (more severe insomnia) were related to increased levels of IL-17A in the vitiligo group ($p=0.007$). Increasing levels of IL-4, IL-6 and IL-10 were associated with previous phototherapy in the vitiligo sample ($p=0.03$; $p=0.03$ and $p=0.04$, respectively).

Conclusions: Poor sleep quality predominated in the vitiligo group. Body pain may be related to hyperalgesia due to worse sleep, impairing quality of life of the individuals with vitiligo. Increasing levels of the pro-inflammatory IL-17A have been associated to insomnia symptoms (elevated ISI scores). Higher levels of IL-4, IL-6 and IL-10, which have circadian behaviour, were related to UVB phototherapy, highlighting the effect of this treatment on the immune system. Sleep disturbance and worse vitiligo may have common pathways related to cytokine regulation, and this interaction should be further examined in respect of vitiligo pathways and clinical management of the disease.

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Abstract N°: 2679

Patients' desire for interdisciplinary management of axial spondyloarthritis: A qualitative interview study in Germany

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Introduction & Objectives: Axial spondyloarthritis (axSpA) is associated with extra-articular manifestations of the eyes, gastrointestinal tract, and skin, including psoriatic skin lesions in 20% of axSpA cases, making it difficult to differentiate from psoriatic arthritis. The worldwide diagnostic delay for axSpA is 6.7 years, accompanied by numerous misdiagnoses, physician changes, and a large disease burden for those affected. However, early specific therapy is essential for the lives of affected persons. Thus, the aim of the study was to explore the healthcare journey of axSpA patients.

Materials & Methods: A total of 31 patients with axSpA in the patient population of a rheumatology practice in southern Germany were identified by purposive sampling and invited to participate by mail. Semi-structured interviews were conducted via video call with 15 voluntary patients by one interviewer using a pilot-tested interview guide between September and October 2021. The audio-recorded interviews were transcribed verbatim and analysed according to Kuckartz's qualitative content analysis. Three transcripts were coded by two independent researchers the rest from the interviewer using a uniform inductively and deductively established coding manual. The qualitative research software MAXQDA was used for data management and analysis.

Results: Fifteen patients (aged 24-67 years, 9 women) participated. Three main themes emerged: "onset and initial healthcare contact", "diagnostic delay and diagnosis", and "treatment and medical care". Not all patients were able to identify the onset of their disease. In retrospect, they complained of having been suffering from severe, stabbing, recurring and often inexplicable back pain. Nevertheless, this pain alone did not always lead the participants to consult a physician; in some cases, the initial contact was based on another medical concern, including skin conditions. Patients described the often years-long path to diagnosis as particularly stressful and painful. They complained about not being taken seriously and about frequent misdiagnoses and described barriers to early diagnosis (e.g., insufficient interdisciplinary exchange among specialists). Some patients also complained about the lack of medical information and an interdisciplinary disease management. Of great importance to the participants were their personal treatment goals (e.g., desire to have children).

Conclusion: The study allowed a comprehensive insight into the individual healthcare journey of persons with axSpA and highlighted the enormous burden of the patients especially due to the diagnostic delay. Awareness regarding axSpA should be increased in specialist disciplines such as dermatology and interdisciplinary cooperation in the care of patients should be promoted by establishing networks for axSpA management. This can reduce the burden of axSpA and ensure individualized treatment, tailored to personal treatment goals.

Abstract N°: 2739

SRP-positive necrotising myopathy with cutaneous involvement

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Introduction & Objectives:

Idiopathic inflammatory myopathy, classified into four major categories: dermatomyositis, polymyositis, inclusion body myositis and the immune-mediated necrotising myopathies (IMNM) is a rare condition, estimated to affect 2–7 people per 1,000,000. We present a case of IMNM with positive SRP autoantibodies and a skin manifestation.

Materials & Methods:

A 61-years old patient was admitted to the Dermatology Department due to skin lesions that had been present for two years accompanied by an enormous fatigue, muscle pain affecting the proximal muscles of the upper and lower limbs, permanent hair loss, itch and burning sensation of the skin. He also complained of losing 10 kg of body weight in the last 6 months. Apart from atherosclerosis of the lower limbs and common femoral arteries angioplasty 3 years earlier, he denied any chronic diseases.

Physical examination revealed scaly plaques with scarring and dyspigmentation most prominent on the sun-exposed areas, scarring alopecia and linear infiltrated erythema on the dorsal surfaces of the fingers.

Laboratory tests showed anemia, neutrophilia, lymphopenia, elevated erythrocyte sedimentation rate, ketones and urobilinogen in general urine test, elevated liver enzymes, CRP, LDH, phosphocreatine kinase, CK-MB activity, myoglobin, aldolase level and coproporphyrins in urine. ANA test was positive with a titer of 1:320, granular pattern but with no specific antibodies, in myositis panel SRP 54 antibodies were detected. Antibodies specific for dermatomyositis or scleroderma were negative. A fecal occult blood test was positive. Elevated PSA level and rectal examination showed a suspicion of prostate cancer. Photoprovocation tests were positive. Direct immunofluorescence from the perilesional skin was negative. Histopathological examination of the skin suggested the diagnosis of scleroderma. Abdominal ultrasonography revealed an enlarged liver and smooth-walled gallbladder - urolithiasis. Chest X-ray showed no abnormalities.

The treatment of systemic glucocorticosteroids was administered which improved the skin condition, but with no improvement in muscle pain, limb weakness and swallowing difficulties.

During the extended workup the capillaroscopy examination suggested disease from the scleroderma spectrum - a period of late changes. Muscle electromyography showed advanced primary myogenic damage. Biopsy of the muscle indicated dermatomyositis, anti-synthetase syndrome or related diseases. Electron microscopy of the muscle also confirmed the ultrastructural image supported the diagnosis of inflammatory myopathy found in the conclusion of the optical microscopy diagnosis.

Despite the treatment, the course of the disease was complicated by prosthetic vascular graft infection and bleeding in groin areas. The patient had to undergo amputation of both lower limbs due to high risk of sepsis. Unfortunately, gradual deterioration of the patient's general condition eventually resulted in patient's death.

Results & Conclusion:

Taking into consideration the clinical picture, the diagnosis of SRP-positive necrotising myopathy was made. There are not many cases describing skin lesions in the course of SRP-positive necrotising myopathy in the literature. Although rare, dermatologists should be aware of differential diagnosis of myositis and its possible cutaneous manifestation.

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Abstract N°: 2777**Adjuvant therapy of severe and/or refractory bullous pemphigoid with immunoadsorption - A prospective monocentric exploratory pilot study -**

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Introduction & Objectives: Bullous pemphigoid (BP) is the most common autoimmune blistering disease in western countries. It is mediated by circulating and tissue-bound autoantibodies against the hemidesmosomal protein BP180 (type XVII collagen). While remission is achieved in the majority of BP patients by first-line therapy, the guidelines recommend therapy with the anti-CD20 antibody rituximab, high-dose intravenous immunoglobulins, or immunoadsorption (IA) in refractory patients. The aim of the study was to investigate the safety and efficacy of IA in severe and/or refractory BP patients.

Materials & Methods: 10 BP patients (3 women, 7 men; mean age 69.7 years; range 52-81 years) were treated with adjuvant IA on three consecutive days. For all patients, a central venous catheter was employed. In addition, all patients received therapy with oral prednisolone (0.5 mg/kg/day, tapered), dapsone (1.5 mg/kg/day) and topical mometasone furoate ointment (lesional application, twice daily).

Results: The primary endpoint of this prospective pilot study was the number and severity of adverse events (AEs). A total of 56 AEs occurred during the 12-month follow-up (1 to 12 AEs per patient). The majority of AEs corresponded to severity grade 2 (50%), 15 AEs were classified as severe AE, of which 8 (angina pectoris, eczema, BP relapse, hypertensive crisis, and unclear infection) were possibly related to the study medication, and in 7 (gout, bronchial carcinoma, norovirus infection, bursitis olecrani, pelvic ring fracture, intensive care treatment, fall) a relationship with IA seemed unlikely. Two months after IA, 50% of patients showed complete remission on therapy, and after 6 months the number was 90%. In all patients, serum levels of autoantibodies to BP180 dropped significantly after IA, by an average of 85% of baseline immediately after 3rd day of IA, and by 68% 4 weeks later.

Conclusion: IA is a relatively safe and effective therapy, even in older age, for patients with severe or refractory BP.



Abstract N°: 2864**Complete remission after rituximab therapy in an HIV-positive patient with pemphigus foliaceus**Insaf Moubine*¹, Kenza Baline¹, Soumia Chiheb¹¹CHU Ibn Rochd, Dermatology, CASABLANCA**Introduction & Objectives:**

Pemphigus foliaceus (PF) is a life-threatening autoimmune blistering disorder characterized by diffuse skin involvement and the presence of circulating antibodies against desmoglein-1. For moderate to severe pemphigus, B-cell depletion therapy with anti-CD20 rituximab has become the first-line therapy. Active infections are some of the reported contraindications in the dermatological guidelines for the use of rituximab therapy. We describe a case of corticosteroid-dependent PF in a patient with a history of HIV infection who achieved complete remission after rituximab therapy.

Materials & Methods:**Results:**

A 45-year-old female patient with a history of HIV discovered incidentally, evolving for 3 years on antiretroviral therapy, and followed since 2005 for confirmed pemphigus foliaceus on histology and direct and indirect immunofluorescence, received corticosteroid therapy at 1.5 mg/kg/day associated with azathioprine, which was stopped due to leukopenia, then replaced by dapsone with good evolution but with frequent relapses upon tapering. The patient was readmitted for a relapse of the disease without any history of drug intake or preceding infectious episode. Clinical examination found several post-bullous erosions covered with impetiginous crusts located on the trunk, back, and limbs, with involvement of the face and hyperpigmented scar lesions. In view of disease progression and corticosteroid dependence, the decision was made to start rituximab. Before starting the treatment, the patient's HIV viral load was undetectable and the CD4 count was 763 cells/mm³. The patient received 1000mg on days 1 and 14, with complete remission at 6 months. HIV viral load and absolute CD4 counts remained stable during therapy.

Conclusion:

Despite an increase in the use of rituximab for the treatment of pemphigus, there are few dermatological guidelines for prescribing therapy in patients with simultaneous viral infections. Information indicating rituximab avoidance in HIV patients arises from conflicting research involving combination chemotherapy regimens for HIV-associated non-Hodgkin lymphoma. The main concern in the treatment of patients with VIH is the increased risk of infection, particularly viral reactivation. In fact, rituximab therapy does not appear to cause considerable lymphopenia in pemphigus patients, with no changes in absolute CD4+ T-cell count. Although additional studies are required to confirm these findings, the use of rituximab in HIV-positive individuals with pemphigus should be considered in order to prevent the numerous side effects associated with conventional immunosuppressive medications.



Abstract N°: 2868

Pseudotumor Cerebri in Systemic Lupus Erythematosus

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Introduction & Objectives:

Systemic lupus erythematosus (SLE) is a disease that may involve different organ systems and manifest a large variety of clinical syndromes .

Pseudotumor cerebri (PTC), also known as idiopathic intracranial hypertension (IIH), is a syndrome characterized by the occurrence of raised intracranial pressure in the absence of a mass lesion, hydrocephalus, focal structural abnormalities and biochemical or cytological abnormalities.

PTC is included among the rare neuropsychiatric manifestations of SLE.

We report a case of a patient with systemic lupus erythematosus and idiopathic intracranial hypertension.

Materials & Methods:

A 20-year-old woman previously diagnosed with SLE, came to the emergency room with a week history of headache, fever and blurred vision. The neurologic and physical examinations were normal. On eye examination, she had visual acuity of 10/10 in both eyes and no afferent pupillary defect. Fundoscopic examination revealed severe bilateral papilledema.

Intracranial pressure measurement was not performed, and a MRI angiography was normal.

In the absence of an alternative cause for the intracranial hypertension, the presence of lupus flare-up markers suggested a lupic origin.

A treatment with methylprednisolone 1mg/kg/d combined with mycophenolate mofetil 2g/d was started. During the following days symptoms of intracranial hypertension disappeared, papilledema gradually resolved and visual examination improved.

Results:

Pseudotumor cerebri (PC) is included among the rare neuropsychiatric manifestations of SLE. The most common presenting symptoms were headache (82.8%) and visual manifestations (45.7%).

The pathogenetic mechanism of PTC in SLE is not yet clear. Immune-mediated injury within the arachnoid villi and the resultant decrease in cerebrospinal fluid absorption and/or thrombotic obliteration of cerebral arteriolar and venous systems due to a hypercoagulable state are among the most probable mechanisms.

In the cohort provided by Hershko and co-workers, the prevalence of IIH was 1.5% (only hospitalised patients), whereas in the general population the prevalence is 1–19 per 100,000. This fact made them suggest that the two diseases are related.

Almost all of the cases reported were managed with steroids. Most of the patients responded successfully to

steroids alone, or in combination with other drugs.

Conclusion:

we present a case of a neurological condition in a patient with SLE.

several studies suggest that these two entities are related. this association is more than coincidental.

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Abstract N°: 2938**Single-Cell RNA Sequencing Revealed the Heterogeneity of Immune Cells in Han and Zang Systemic Lupus Erythematosus Patients**

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Introduction & Objectives: Systemic Lupus Erythematosus (SLE) is an autoimmune disease with unclear pathogenesis. The purpose of this study was to reveal the heterogeneity of immune cells in SLE patients of Han and Zang nationality by single-cell RNA (sc-RNA) sequencing.

Materials & Methods: We applied single-cell RNA sequencing (scRNA-seq) to 94,102 peripheral blood mononuclear cells (PBMCs) from six patients with SLE (3 Zang, 3 Han) and six healthy controls (3 Zang, 3 Han). The immune cell subsets and susceptibility genes involved in the pathogenesis of SLE were analyzed. Finally, real-time quantitative PCR (RT-qPCR) was applied to confirm the results of sc-RNA sequencing data analysis.

Results: At the end, 24 cell clusters were assessed and defined to be 15 cell types. On the basis of CD4 Naïve, CD4 Tcm, CD8 CTL, and Memory BC, 9 cell subtypes were further determined. For the Tibetan samples, the proportions of CD4 Naïve RPS4Y1 cells, CD4 Naïve cells, Memory BC CD24 and Memory BC were significantly different between the SLE and control samples. For Han nationality samples, the proportion of CD8 CTL MAL cells was significantly different between the SLE and control samples. Via pseudotime analysis, we detected variable differentiation states of CD8 CTL MAL cells, CD8 CTL GZMK cells, and CD4 Naïve cells in the SLE and control groups. Abundance and diversity analyses revealed that T-cell receptor (TCR) abundance was significantly higher in Tibetan SLE samples than in the control group, while TCR diversity of SLE samples was significantly lower than the control group. B-cell receptor (BCR) abundance was higher in Tibetan and Han SLE samples than in the control group, while diversity of control group's BCR was higher. Finally, the results of RT-qPCR indicated the reliability of scRNA sequencing data analysis.

Conclusion: In summary, we explored the immune cellular heterogeneity of SLE patients of Han and Zang nationality based on single-cell RNA sequencing and bioinformatics approaches, providing new perspectives for immunological characteristics of SLE among different ethnic groups.



Abstract N°: 2993**A Novel treatment for salt and pepper pigmentation in scleroderma.**Aarya Sree^{*1}, Preethi Nagaraj², Padmanabha Shenoy³¹India, Dermatology, Cochin, India, ²India, Dermatology, cochin, India, ³India, rheumatology, Cochin, India**Introduction & Objectives:**

Pigmentary alteration is a common yet under reported manifestation in Systemic sclerosis (SSC). Dyspigmentation can either be diffuse hyper pigmentation or mixed hyper and hypo pigmentation known as salt and pepper pigmentation (SPP). The literature on treatment of these lesions are scarce . As SPP is common in the exposed areas, it can cause significant impairment in quality of life and social embarrassment especially amongst those with type IV, V and V skin photo types. One of the hypotheses of SPP is perifollicular sparing of the melanocytes due to the preserved capillary networks in the perifollicular region a process akin to vitiligo . Excimer light has been shown to be effective in the treatment of vitiligo . Here we present a case report of scleroderma associated SPP being treated with 308-nm Monochromatic Excimer light.

Materials & Methods: none

Results: A 32 year old female patient with progressive systemic sclerosis of 7 years duration since the first Raynaud's symptom presented to us with skin thickening, salt and pepper pigmentation, Interstitial lung disease (ILD). She had a Modified Ronan's Skin score of 27 and had SPP on upper part of chest, back and both lateral sides of face. She was positive for anti-topoisomerase I antibody and was initially started on Mycophenolate mofetil (MMF) . With MMF although ILD stabilised and pruritus improved, the patient noticed increase in SPP and began to isolate herself from social occasions. After a detailed discussion with the patient and family ,a shared decision to try Excimer lamp (308 nm) therapy was arrived . The treatment was scheduled thrice in a week .The initial dose was 150 millijoules/cm² (mj/cm²) which was increased by 10% on subsequent visits until patient developed photo toxic reaction like mild erythema which lasted less than 24 hours without any blistering .The erythema over the lesional area was treated with topical corticosteroid and dose was slowly up titrated by 15 mj/cm² (10% of baseline dose)once its subsided. The maximum dose was given over the back and chest were 1000 mj/cm²and 850mj/cm² respectively. Improvement in SPP was documented from 6th session and she attained near normal skin pigmentation after 40 sessions(4 months)

Conclusion: The benefits of Excimer light in the treatment of vitiligo depends on the number of sessions rather than the frequency. Studies have reported re -pigmentation of >75% of the lesions . Excimer lamp use a mix of a rare gas and a reactive gas (Xenon chloride) to create a complex that breaks apart and releases energy as UV radiation.The monochromatic incoherent non directional 308 nm wavelength light of Excimer lamp induces photo biological reactions and results in activation of latent melanocytes⁶ .This leads to migration of the active melanocytes to the nearby depigmented skin and causes repigmentation in vitiligo. Besides this,Excimer light is also a strong inducer of T cell apoptosis and upregulates endothelin-1 production from keratinocytes..Though their mechanism of action is not explored in SSc, it is likely to be based on the above principles. This is the first report to the best of our knowledge that has resulted in complete reversal of SPP in diffuse systemic sclerosis. Even though the treatment may not prevent new lesions, its efficacy in this case definitely can be an effective treatment modality in Scleroderma. Larger and well planned studies exploring the effects of topical photo therapy in SPP would shed more light in this context.

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Abstract N°: 3044**Experience sharing on the treatment of severe bullous pemphigoid with low-dose systemic corticosteroids combined with Dupilumab**Xin-Gang Wu¹, Aie Xu¹¹Affiliated Hangzhou Dermatology Hospital of Zhejiang University School of Medicine, Dermatology, Zhejiang**Introduction & Objectives:**

To investigate the clinical efficacy of low-dose systemic glucocorticoids combined with dupilumab in the treatment of severe bullous pemphigoid (BP) patients.

Materials & Methods:

Clinical data of 10 patients with severe BP treated with low-dose systemic glucocorticoid combined with Dupilumab in our hospital were analyzed, and compared with 10 patients with severe BP treated with other regimens in our hospital before. BPDAI score, NRS (Simple Pruritus Score), eosinophil percentage/absolute value, time to stop new blisters, average length of hospital stay, skin disease quality of life index (DAQI) Evaluate the therapeutic effect based on indicators such as the time from hormone reduction to maintenance dose (10mg) and total hormone dosage.

Results:

Before treatment, there was no significant difference in the BPDAI score, NRS score, and eosinophil percentage/absolute value indicators between the low-dose systemic glucocorticoid combined with Dupilumab treatment group (combined treatment group) and the control group; After treatment, the NRS, eosinophil percentage/absolute value, and average hospital stay in the combined treatment group were significantly reduced compared to the control group, and all values were statistically significant; The duration of cessation of new blisters and average hospitalization days in the combined treatment group were significantly shorter than those in the control group, and the difference was statistically significant; The DAQI score of patients in the combined treatment group during hospitalization decreased significantly compared to the control group, and the difference was statistically significant. The time for hormone reduction to maintenance dose in the combined treatment group was shortened compared to the control group, and the total hormone dosage was significantly reduced compared to the control group, both of which were statistically significant.

Conclusion:

The combination of low-dose systemic glucocorticoids and Dupilumab is effective in the treatment of severe bullous pemphigoid. The combination of Dupilumab can quickly control the condition of severe BP, significantly improve the quality of life of patients, and contribute to the rapid reduction of systemic glucocorticoids.



Abstract N°: 3176

Nasal skin inflammation and necrosis after two failed surgical nose reconstructions in a female patient with a very rare diagnosis of relapsing polychondritis and successful posterior rhinoplasty using costal cartilage

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Introduction & Objectives:

Relapsing Polychondritis (RP) is a very rare disease characterized by inflammation of the cartilage of the nose, ears and airways. Many patients develop deformities with saddle nose being a functional, aesthetic and psychological challenge. The medical community is often apprehensive to perform nose surgical corrections on patients affected by RP, as procedural experience and literature on the subject is scarce. There is a tremendous need to share experiences, results and complications on these procedures. Dermatological manifestations of RP may be a direct consequence of the natural history of the disease or as in this case a consequence of the surgical procedure.

Materials & Methods:

We present the case of a 43-year-old female patient who was diagnosed with RP after several years of being studied for recurrent upper respiratory problems originally managed as asthma. About 6 months after the initial diagnosis the nasal septum was impacted by the disease and its destruction created a saddle nose deformity affecting the functionality with breathing limitations and a severe aesthetic and psychological burden.

At first a reconstructive rhinoplasty was performed by a plastic surgeon using a porous polyethylene implant. The aesthetic results were somehow acceptable, but the functionality was still severely affected; so, another surgeon operated a second time inserting a calvaria graft fixing it with surgical screws. During the recovery the aesthetics and functionality of the nose were severely impacted once again, and the patient developed a severe inflammation of the skin over the nose with erythema, pain, edema and skin necrosis and exposure of the surgical screws.

An experienced rhynologist intervened by studying the few previously published cases together with his vast experience. He instead suggested a different surgery: a new nasal reconstruction using costal cartilage without the use of surgical screws.

Results:

The successful surgery brought regained functionality of the nose. The skin infection was properly managed and hyperbaric oxygen was prescribed as adjunctive treatment to promote wound healing and prevent infection. The patient has been followed for a year now without presenting any new skin or breathing complications and manifests improved quality of life.

Conclusion:

RP is a very rare disease; the incidence is 1 to 2 cases per one million patients at risk. Literature on the topic is scarce, with cases related to the dermatological complications and the correct management of nose deformities in RP even more rarely published. Nasal reconstructions require the use of cartilage, and the procedure should only be done by highly experienced rhynologists with vast knowledge, not only of the aesthetics, but of the

functionality of the nose and the possible dermatological consequences.

In this case the skin and posterior necrosis likely developed as a foreign body reaction to the use of materials such as the surgical screws. Coastal cartilage should be the medium used for such surgeries since it is almost always spared by the RP and foreign body reaction is unlikely. In the scant previously published cases the consensus is that these patients can present dermatological reactions to the use of allogenic materials. Our experience with this case confirmed the fact that successful surgical corrections with coastal cartilage can be performed in RP with very positive functional and aesthetic results and minimizing dermatological complications.

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Abstract N°: 3185

Risk factors for malignancy in adult-onset dermatomyositis

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Introduction & Objectives:

Adult-onset dermatomyositis is an idiopathic inflammatory myopathy characterized by symmetric proximal muscle weakness and pathognomonic cutaneous features. There is a well-established association between adult-onset DM with serious comorbidities including malignancy. The objective of this retrospective study was to describe the prevalence and types of malignancies and analyze for independent risk factors for malignancies in Canadian participants with adult-onset dermatomyositis.

Materials & Methods:

This study was approved by the institutional review board. We conducted a retrospective chart review of patients with DM at Hamilton Health Sciences and St. Joseph's Healthcare Hamilton in Hamilton, Ontario between January 1, 2010 until December 31, 2020. These two institutions serve as tertiary care centers for Southern Ontario. Included participants were clinically diagnosed with DM at age 18 years or above and had their diagnosis listed in at least two separate medical documents. Juvenile-onset participants were excluded. Baseline characteristics of the study population were summarized with descriptive statistics. Continuous variables were compared between cohorts with and without malignancy using the Mann Whitney U test. Categorical variables were compared using Fisher's exact test. A multivariable regression analysis identified independent risk factors for malignancy. All statistical analyses were completed in RStudio 1.4 (Boston, MA) with two-sided p values and significance determined a priori as $p < 0.05$.

Results:

The study population ($n=92$) was 80% female and the mean age at diagnosis was 51 years (range 18-88 years). Malignancy was reported in 41% ($n=38/92$) of subjects. The most common malignancies were breast (10%; $n=9$), cutaneous melanoma ($n=4$), lung ($n=3$), and colorectal ($n=3$). The majority of participants (63%, $n=24$) received a diagnosis of malignancy within 5 years of their DM diagnosis. Most malignancies (42%; $n=16/38$) postdated the diagnosis of DM by a mean 6.44 years, 34% ($n=13$) of malignancies predated the diagnosis of DM by a mean 5.92 years, and 8% ($n=3$) of malignancies were diagnosed simultaneously with DM. Data with respect to mortality was available for 92% ($n=35/38$) of the participants with a malignancy, and 46% ($n=16$) were deceased at the time of chart review.

Baseline characteristics of the cohorts with and without malignancy were compared. Significantly more participants with malignancy were deceased at the time of chart review ($p=0.01$). The cohort with malignancy was significantly older (mean age 57.8 years) than the cohort without malignancy (mean age 47.0 years, $p=0.0001$) at the time of diagnosis. Multivariable regression analysis found greater age at the time of diagnosis to be the only significant predictor of malignancy in our study population (hazard ratio 1.04, 95% confidence interval 1.023, 1.062, $p=1.8 \times 10^{-5}$).

Conclusion:

This is the largest dedicated study of Canadian participants with adult-onset dermatomyositis. Increased age at the time of diagnosis was the only statistically significant risk factor for malignancy. Larger prospective studies with standardized reporting of variables will help identify other risk factors for malignancy in this patient population and may provide evidence for malignancy screening guidelines.

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Abstract N°: 3193**Rare cutaneous manifestation of IgG4 disease**

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Introduction & Objectives:

We present a case of a young female patient with progressive swelling in the nasal dorsum, eyelids, bilateral malar region, and unilateral strabismus. The objective of this report is to discuss the clinical, imaging, pathological, and laboratory parameters that led to the diagnosis of IgG4-related disease in this patient and highlight the rare cutaneous involvement of this condition.

Materials & Methods:

A thorough clinical examination was performed, including imaging studies such as face tomography. A skin biopsy was obtained for histopathological analysis. Laboratory investigations, including serum IgG4 levels, were also conducted to aid in the diagnosis.

Results:

The face tomography revealed soft tissue infiltration consistent with the clinical findings. The histopathological examination demonstrated dermal plasmacytic inflammatory infiltrate, storiform fibrosis, obliterative phlebitis, lymphoid follicles, and IgG4-positive plasmacytic infiltrate. Additionally, elevated serum IgG4 levels were detected. Based on these findings, a diagnosis of IgG4-related disease was made.

Conclusion:

IgG4-related disease is a rare immune-mediated fibroproliferative disorder characterized by inflammation, fibrosis, and lymphoplasmacytic infiltration rich in IgG4-positive plasma cells. Although cutaneous involvement is uncommon, it should be considered in the differential diagnosis. The diagnosis of IgG4-related disease relies on the correlation of clinical, imaging, laboratory, and histopathological findings. Corticosteroids are the first-line treatment, and Rituximab is used in refractory cases. In this case, the patient showed a favorable response to corticosteroid therapy, maintaining a stable clinical condition.



Abstract N°: 3216**Characteristics and management of patients with alopecia areata and selected comorbid conditions: results from a survey in 5 European countries**

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Introduction & Objectives: Alopecia areata (AA) is a chronic autoimmune condition that causes non-scarring hair loss and can impose a high burden on patients. Previous studies have shown that AA is associated with various comorbidities, such as autoimmune, atopic, and psychological disorders [1, 2]. However, there is little evidence on the characteristics and therapeutic management of AA in patients who present with these comorbidities. This analysis aims to describe disease characteristics and treatment patterns related to AA in patients with selected comorbid conditions, including autoimmune, atopic, and psychological disorders.

Materials & Methods: Data from a point-in-time medical chart review conducted in France, Germany, Italy, Spain, and the United Kingdom between October 2021 and June 2022, were analysed. Physician-adjudicated adults with mild, moderate, and severe AA, were enrolled. Data on demographics, AA clinical characteristics, comorbid conditions, as well as information related to AA therapies were collected and analyzed descriptively.

Results: In all, 239 dermatologists provided data for 2083 patients. The mean age for the three comorbidity groups was 37.6 years (SD=12.1) and 56% of respondents were women (n=313). Physician-rated severity was mild for 299 patients (14%), moderate for 936 (45%), and severe for 848 (41%). Overall, 27% (n=558) of patients had at least one of the selected comorbid conditions. Within the three comorbidity groups, more patients had severe AA and less patients had mild AA. Scalp hair loss was the primary sign reported across the three comorbidity groups (autoimmune=91%, atopic=91%, psychological=88%). Overall, a higher percentage of patients in all three comorbidity groups than in the entire population presented with AA-related signs and symptoms beyond scalp hair loss. The current use of topical and systemic corticosteroids was similar across the groups. The use of topical immunotherapy, conventional systemic immunosuppressants and oral JAK inhibitors tended to be higher in patients with autoimmune, atopic, and psychological comorbidities, with the exception of the use of conventional systemic immunosuppressants in patients with atopic comorbidities which was similar to the overall population.

Conclusion: This analysis provided insights into the burden and management of AA in patients presenting with auto-immune, atopic, and psychological comorbidities, in five European countries. Patients with autoimmune, atopic, and psychological comorbidities had a more severe AA, characterized with a higher prevalence of AA-related signs and symptoms, beyond scalp hair loss. Overall, these patients were more frequently receiving topical immunotherapy, conventional systemic immunosuppressants and oral JAK inhibitors for the treatment of AA.

\1. Egeberg, A., et al., *Comorbidities of alopecia areata: a population-based cohort study*. Clin Exp Dermatol, 2021. **46**(4): p. 651-656.

\2. Lee, S., et al., *Comorbidities in alopecia areata: A systematic review and meta-analysis*. J Am Acad Dermatol, 2019. **80**(2): p. 466-477.e16.

Table 1: Demographic, disease, and comorbidity characteristics of enrolled patients

Physician-reported concomitant condition groups ¹	Overall ²	Atopic comorbidities	Auto-immune comorbidities	Psychological / psychiatric comorbidities
	n=2083	n=255	n=195	n=221
Age (years), mean (SD)	35.7 (11.5)	35.8 (12.1)	39.9 (12.1)	39.2 (12.5)
Sex (female), n (%)	997 (48%)	129 (51%)	122 (63%)	134 (61%)
BMI, mean (SD)	24.3 (3.2)	24.3 (3.3)	24.0 (3.5)	24.2 (3.8)
Current level of disease severity*, n (%)				
Mild	299 (14%)	19 (7%)	15 (8%)	15 (7%)
Moderate	936 (45%)	107 (42%)	83 (43%)	85 (38%)
Severe	848 (41%)	129 (51%)	97 (50%)	121 (55%)
Current signs and symptoms related to AA n (%)				
Scalp hair loss	1802 (87%)	232 (91%)	177 (91%)	195 (88%)
Eyebrow hair loss	566 (27%)	88 (35%)	71 (36%)	98 (44%)
Eyelash hair loss	363 (17%)	57 (22%)	51 (26%)	62 (28%)
Scalp itching	324 (16%)	50 (20%)	44 (23%)	55 (25%)
Body hair loss	247 (12%)	37 (15%)	38 (19%)	35 (16%)
Facial hair loss (moustache or beard)	210 (10%)	31 (12%)	30 (15%)	26 (12%)
Nail damage	208 (10%)	46 (18%)	37 (19%)	46 (21%)
Scalp irritation	206 (10%)	34 (13%)	30 (15%)	39 (18%)
Scalp burning or stinging	204 (10%)	36 (14%)	32 (16%)	42 (19%)
Scalp pain	176 (8%)	31 (12%)	20 (10%)	32 (14%)
Eye irritation	64 (3%)	13 (5%)	11 (6%)	14 (6%)
Other	5 (<1%)	1 (<1%)	1 (1%)	2 (1%)
Don't know	12 (1%)	0 (0%)	0 (0%)	0 (0%)
None of the above	133 (6%)	8 (3%)	6 (3%)	10 (5%)
Percent of patients who have never been prescribed treatment for AA, n (%)				
Patient has never been prescribed treatment for AA	31 (1%)	4 (2%)	1 (1%)	1 (<1%)
Current treatment for AA³, n (%)				
	n=2052	n=251	n=194	n=220
Topical corticosteroid	693 (34%)	80 (32%)	77 (40%)	69 (31%)
Systemic Corticosteroids (oral and intravenous regimen)	483 (24%)	55 (22%)	41 (21%)	52 (24%)
Conventional Systemic Immunosuppressants (azathioprine, cyclosporine A, and methotrexate)	433 (21%)	51 (20%)	47 (24%)	80 (36%)
Intralesional injected corticosteroid	430 (21%)	45 (18%)	33 (17%)	48 (22%)
Topical calcineurin inhibitor (tacrolimus, pimecrolimus)	261 (13%)	45 (18%)	29 (15%)	41 (19%)
Topical Immunotherapy (1-chloro, 2,4-dinitrobenzene [DNCB], squaric acid dibutylester [SADBE], 2,3-diphenylcyclopropanone [DPCCP], and other)	212 (10%)	34 (14%)	31 (16%)	32 (15%)
Oral JAK inhibitor (baricitinib, ruxolitinib, tofacitinib, and other oral JAK inhibitor)	205 (10%)	33 (13%)	29 (15%)	38 (17%)

¹ **Atopic comorbidities** includes patients diagnosed with atopic dermatitis, atopy, asthma, allergic rhinitis, urticaria, angioedema, allergic contact dermatitis, atopic keratoconjunctivitis, other allergic conditions and/or nasal polyps), **Auto-immune comorbidities** includes patients diagnosed with rheumatoid arthritis, Crohn's disease, ulcerative colitis, lupus, psoriasis, thyroid disease, vitiligo, celiac disease, diabetes (type 1), connective tissue disease, psoriatic arthritis, ankylosing spondylitis and/or non-radiographic axial spondylitis) and **Psychological/psychiatric comorbidities** includes patients diagnosed with depression, anxiety, insomnia, bipolar disorder, attention deficit disorder and/or bipolar disorder).

² 1,117 patients (54%) had no reported comorbid condition, including but not limited to those in the selected groups of comorbid conditions.

³ Patients could list more than one AA treatment.

*Disease severity was determined by the dermatologists according to their own definition of the terms "mild," "moderate," and "severe".



Abstract N°: 3248

Bullous pemphigoid in a patient with complex partial epilepsy and newly onset treatment with levetiracetam

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Introduction:

Bullous pemphigoid (BP) has been traditionally associated with chronic neurologic diseases and certain drug intake. Although these etiologies are clearly defined in scientific publications, in the real world both may contribute to the development of BP.

Results:

A 70-year-old female patient presented to the emergency department complaining of intensely pruritic skin lesions for four days. She had a history of complex partial status epilepticus, diabetes mellitus and hypertension. She had initiated treatment with levetiracetam 39 days prior to the skin symptoms onset. On physical examination, she showed tense bullae on an erythematous base over the trunk and limbs. Nikolsky sign was negative. She had no mucosal involvement.

Serum analysis for BP180 and BP230 antibodies was positive for the former. A skin biopsy demonstrated a subepidermal blister with eosinophils and direct immunofluorescence revealed a linear deposition of C3 along the dermoepidermal junction, confirming the diagnosis of bullous pemphigoid.

Levetiracetam was discontinued and the patient was started on Prednisone 0.5 mg/kg/day and clobetasol propionate 0.05% cream. However, minor blisters continued to appear, so azathioprine was initiated at low doses (0.5-1 mg/kg/day), with no new blisters development after 1 month of treatment. Nowadays corticosteroids have been discontinued and azathioprine is being tapered, with no flare-up of skin lesions.

BP patients present neurologic diseases more frequently than healthy individuals, including epilepsy, dementia and stroke, probably because of a cross reaction between epidermal and neuronal BP180. These disorders usually precede BP, and are diagnosed on average 6.7 years before BP onset. Even though there are large reviews focused on the relationship between BP and neurologic diseases, none of them dismiss the possible role of concomitant neurologic medications that patients may be taking.

On the other hand, the list of drugs which could contribute to BP development is endless, but the strongest associations are described with dipeptidyl peptidase-4 and checkpoint inhibitors. Just two cases of levetiracetam-induced BP have been reported to date, occurring in 70 and 31-year-old patients. As a difference with other drug induced bullous pemphigoid, these cases related to levetiracetam show a shorter latent period (approximately 2 months) and apparently resolve with the discontinuation of the culprit medication. Nonetheless, the underlying mechanism of both drug reactions is yet to be described.

Conclusion:

Even though the incidence of BP in patients diagnosed with epilepsy or treated with levetiracetam is low, it is important to be aware of this relationship in order to perform an early diagnosis and start treatment as soon as possible, and therefore to improve its prognosis.

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Abstract N°: 3258

Ocular Mucous Membrane Pemphigoid (OMMP): an Italian real life monocentric case series study

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Ocular Mucous Membrane Pemphigoid (OMMP): an Italian *real life* monocentric case series study

Introduction & Objectives: OMMP is a rare and *high risk* autoimmune bullous disease with a calculated overall incidence of 0.7-0.8 cases/million/year. The difficult therapeutic management and the average delay in diagnosis (ranging from 225 days to 6.4 years) make this pathology often highly disabling for the patients. Herein, we would like to describe our experience from April 2014 to March 2023 in managing twenty-two patients (5 M and 17 F) diagnosed with OMMP.

Materials & Methods: During the screening phase for each patient we performed biopsies for optical microscopy (OM) and direct immunofluorescence (DIF), serological tests (anti-BP180/230, DSG-1/3 ELISA “enzyme-linked immuno sorbent assay”), complete blood chemistry, ABQOL questionnaire (QoL) and disease grading using Foster’ staging system. The complete medical history and the Karnofsky Performance Status (KPS) were also recorded. The diagnosis of OMMP was established based on clinical findings, direct immunofluorescence of conjunctiva and/or buccal mucosal biopsies, and serology. All patients received topical and systemic treatments up to the 3rd line according to the European guidelines (S3) on diagnosis and management of MMP. The FU phase included a visit every 4-6 months until rescheduling because of clinical signs and symptoms’ remission, relapse, adverse events (AEs) and/or patient choice. The ophthalmological staging and a quality-of-life re-evaluation were performed at the last dermatological visit both for patients in systemic maintenance therapy and those in topical one.

Results: The median age at diagnosis was 69.5 y.o. (range 43-88) with a 3-year average delay from the OMMP’s onset. DIF and serological autoantibodies detections resulted negative in 9 (40.9%) and 16 (72.7%) patients, respectively: 2 of them acquired anti- BP180/230 seroconversion. 50% of the cohort was in Foster’ stage III and IV. Seven patients described an OMMP *sensu stricto*, while* the others were characterized by an extra-ocular manifestation, mainly oropharyngeal (59%). Ocular pemphigoid anticipated the development of extra-ocular lesions in 11 cases (69%). In addition, the ocular disease presentation resulted asymmetric in 8 patients, with more severe clinical picture affecting the right eye in three cases and the left one in five. This association was not reported when the disease interested only the mucosal sites. Systemic immunosuppressive therapy required up to 6 different traditional drugs: dapsons and prednisone were the most used molecules. Three patients underwent therapy with rituximab, omalizumab and intravenous immunoglobulins. Among ADRs (Adverse Drug Reactions) only one patient experienced pharyngodynia complicated by haemoptysis with serological CRP rising (8.7 mg/L). The average Foster staging calculated for the entire cohort at the beginning and end of FU was reduced by a certain number. Likewise, ABQOL questionnaires showed a decrease in terms of disease’s discomfort and negative impact on the quality of life.

Conclusion: Our study confirms the therapeutic efficacy of the traditional immunosuppressive therapies commonly used in the dermatological management of OMMP. However, the early diagnosis today could be

considered the Achille's heel of this pathology, even though it represents in fact the only "therapy" capable of avoiding disabling and irreversible ocular complications for the patients.

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Abstract N°: 3261**A case report of a patient with coexisting Morphea and Generalised Granuloma annulare**

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A case report of a patient with coexisting Morphea and Generalized Granuloma annulare

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Introduction & Objectives:

Granuloma annulare is a benign, self-limiting granulomatous disease. Recent studies showed that the incidence among the population is 0.01-0.04%, most commonly occurring in individuals under 30 years of age. The localized annular and subcutaneous forms occur more frequently in children and young adults. The disseminated variant is more common in adults, constituting about 8-15% of granuloma annulare cases. The female-to-male ratio is approximately 2:1. Morphea, also known as localized scleroderma, is characterized by fibrosis of skin and subcutaneous tissue. The increase in collagen production resulting from skin fibrosis can arise from endothelial cell injury, immunological factors (for example, relating to T-lymphocytes) and inflammatory activation and dysregulation of collagen production.

Materials & Methods:

We present a 74-year old woman, who suffered from a pruritic erythematous rashes involving her trunk and extremities. The lesions originally appeared a year prior, initially involving the lower extremities and gradually spreading to the trunk and upper extremities. Previous treatment administered by rheumatologists regarding elevated anti-IgG antibody values (134, the upper reference limit being 15) included a 25-day course of Doxycycline tablets 100 mg, orally twice daily. Prior topical treatment was made with Clobetasol propionate 0,05% cream with unsatisfactory results. The patient reported the erythema had only intensified. Somatic status was without deviations.

Dermatological status: Physical examination revealed multiple cutaneous lesions. Pathological cutaneous process affected the trunk, upper and lower extremities. The lesions were presented by erythematous-violaceous annular plaques with raised margins and a pale central depression (granuloma annulare) and rashes with well-defined oval patches with a central ivory white area surrounded by an erythematous-violaceous rim - "lilac ring" (plaque-type morphea).

Skin appendages, peripheral lymph nodes and visible mucosa showed no pathological changes.

Results:

Routine laboratory investigations: Complete blood count, biochemistry, Lyme IgM/IgG antibodies, autoimmune panel (antinuclear antibodies and anti-ds-DNA), erythrocyte sedimentation rate, thyroid hormones and antibodies, and urine analysis showed no deviations.

Histopathological testing on a skin sample confirmed both diagnoses Morphea and Granuloma annulare.

Based on data from patient's medical history, dermatological status and results from the investigations we made the diagnosis morphea combined with granuloma annulare – generalized form. According to guidelines for these diseases we decided to start therapy with Chloroquine phosphate tab. 200 mg p.o. administered once daily. Topical therapy included Tacrolimus 0,1% ointment, applied in tapering doses over the course of 5 weeks.

Conclusion:

In conclusion we presented a clinical case of a rare form of Granuloma annulare generalized variant combined with Morphea. Only a few cases in the literature have described the coexistence of morphea and granuloma annulare. The last one was described in 2019.

Abstract N°: 3293**Paraneoplastic bullous pemphigoid secondary to cutaneous squamous cell carcinoma**Karolina Zarańska¹, Barbara Radecka², Kamila Czerw³, Grażyna Wąsik¹¹Provincial Hospital in Opole, Clinical Department of Dermatology, Opole, Poland, ²Oncology Center in Opole, Oncology Department, Opole, Poland, ³Oncology Center in Opole, Radiotherapy Department, Opole, Poland

Introduction & Objectives: Cutaneous squamous cell carcinoma (cSCC) is the second most common form of skin cancer. The incidence of cSCC is growing, mostly in fair-skinned individuals, with UV-damaged skin, usually over the age of 50. Treatment recommendations are generally related to the stratification of high-risk and low-risk tumors, but surgical therapy is the most effective in general. Like other malignant diseases, the presence of SCC can be related to a paraneoplastic phenomenon. The appearance of bullous pemphigoid (BP) in the context of cSCC is an exceptional and strongly suggests the relationship between these two diseases.

Materials & Methods: We present a case of bullous pemphigoid and cutaneous squamous cell carcinoma, with a simultaneous clinical course, which suggests a paraneoplastic phenomenon.

Results: A 62-year-old man was admitted to the dermatology department with a 3-week history of non-prurigo oval-shaped eczematous and urticarial lesions, some with excoriations in the center. The lesions were distributed on the trunk and symmetrically on flexural surfaces of the lower limbs, with no mucosal involvement. In addition, physical examination revealed enormous ulceration on the scalp size 10x7cm, infiltrating the frontal bone. Based on the clinical features, the suspicion of Sweet syndrome and erythema multiforme was raised. A skin biopsy from one of the eczematous lesion was performed, which was non-specific. A biopsy from ulceration on the scalp confirmed cSCC. Afterwards the patient was referred to the oncology department. Because of the infiltration of deeper tissues, the computer tomography was performed and stage T4N0M0 (according to American Joint Committee on Cancer, AJCC) was diagnosed. Surgical treatment was not feasible at that moment because of high-risk localized tumors and a poor aesthetic result. Modulated Radiation Therapy started out with the progression of cSCC in the form of metastasis to regional, parotid lymph node during it. Concomitantly, the patient presented pruritic bullae arising on erythematous inflamed skin. In the physical examination the Nikolsky sign was negative. The direct immunofluorescence microscopy (DIF) showed linear deposits of IgG4 and C3 along the dermoepidermal junction. DIF was consistent with bullous pemphigoid. Immunotherapy with cemiplimab, monoclonal antibody targeting programmed cell death 1 (PD-1) receptor, was contraindicated because of BP coexistence. Suboptimal oncological therapy started, in the form of renewed local radiotherapy and chemotherapy initially with fluorouracil and cisplatin, then with paclitaxel. Further follow-up was impossible because of the loss of contact with the patient after the third dose of paclitaxel.

Conclusion: Bullous pemphigoid (BP) constitutes the most common autoimmune subepidermal blistering disease of the skin and mucous membranes which occurs mainly in the elderly. It is associated with tissue-bound and circulating autoantibodies directed against BP antigen 180 and BP antigen 230. The diagnosis is based on a combination of criteria encompassing clinical features, compatible light microscopy findings and positive direct immunofluorescence microscopy (DIF) findings. Both, BP and cSCC had a parallel clinical course which suggests a paraneoplastic phenomenon in this reported case. Different types of malignant diseases can present with bullous pemphigoid, but the paraneoplastic significance of this association is still unclear.

Abstract N°: 3299**Importance of immunofluorescence in rare cases of bullous dermatoses**Diana Aniculaesa¹, Adriana-Ionela Patrascu^{1, 2}, Elena - Larisa Butnaru¹, Laura Solovastru^{1, 2}¹Saint Spiridon County Hospital, Dermatology, Iași, Romania, ²Universitatea de Medicină și Farmacie „Grigore T. Popa” din Iași, Iași, Romania**Introduction & Objectives:**

Linear IgA bullous dermatosis (LABD) is a rare autoimmune blistering disease that occurs in both children and adults. In adults can have a varied presentation, making it difficult to distinguish from other autoimmune vesiculobullous diseases. Patients may present with scattered, tense bullae on a background of non-inflamed skin, while others may develop a more herpetiform appearance to their lesions with prominent erythema beneath the vesicles or bullae. Pruritus is common and may be severe. The presence of IgA deposits along the basement membrane can usually be identified using direct immunofluorescence microscopy (DIF). The histological and clinical features of this disorder may mimic those of dermatitis herpetiformis.

Immunohistochemistry is a powerful technique that exploits the specific binding between an antibody and antigen to detect and localize specific antigens in cells and tissue, most commonly detected and examined with the light microscope.

Materials & Methods:

We report the case of a 39 old women patient presenting with isolated and grouped tense serum-filled vesicles and bullae on healthy skin or on an erythematous base, mostly in a rosette pattern. The patient did not present mucosal lesions or fever, and there was no impairment of the patient's general condition, was associated only pruritus.

Results:

An anatomopathological examination of a bullous lesion showed subepidermal cleavage with neutrophilic inflammatory infiltrate. Direct immunofluorescence of perilesional skin showed intense linear deposition of IgA and IgG at the basement membrane.

Dermatitis herpetiformis has very similar findings on immunohistochemical staining, with the main difference being the granular deposition of IgA, instead of linear deposition as in LABD. Bullous pemphigoid is characterized by linear deposits of IgG along the epidermal basement membrane zone. Unfortunately, at times, there can be a mixed linear deposition of both IgG and IgA at the basement membrane, creating a diagnostic conundrum. This is a rare case of bullous dermatoses where immunofluorescence showed linear deposit of IgA at basal membrane and also IgG, which is observed only in 25 % of the cases.

Conclusion:

LABD has a diverse presentation and has many causes, being a rare disease. This disorder is best managed by an interprofessional team. We talk about the importance of performing DIF to confirm the diagnosis of the disease and to establish an appropriate treatment.



Abstract N°: 3312**The Great Imitator Strikes Again: Generalized acute cutaneous lupus erythematosus simulating an erythema multiforme**

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Introduction & Objectives:**Materials & Methods:****Results:****Introduction:**

Generalized acute cutaneous lupus is an atypical form of presentation that only occurs in 5-10% of cases of systemic lupus erythematosus. It presents as a generalized maculopapular rash predominantly in photo-exposed areas with palmoplantar and mucosal involvement. This generalized form appears similar to a drug-induced rash which may delay the diagnosis.

Case report:

A 51-year-old Mexican male with history a history of chronic arthralgias for which he refers consumes allopurinol (300mg/day). He presented to our dermatology department with a 10-day history of multiple oral ulcers with a diffuse rash on the trunk, and fever.

His physical examination revealed a symmetrical dermatosis disseminated to the face, trunk, and extremities, consisting of a confluent, erythematous-purple maculopapular rash, predominantly in the face, central back, with palmar-plantar involvement, some of them had a "target" like lessons, labial mucosa, buccal mucosa and lips ulcers and extensive hematic scab.

Laboratory findings showed pancytopenia (Hemoglobin 12.0 g/dl, platelets 91.59 thousand/ μ L, lymphocytes 3.29 thousand/ μ L), therefore he was admitted to hospitalization for further evaluation. Serologies for HIV, VDRL, and Herpes virus were negative. A skin biopsy reported an epidermis with hyperkeratosis and acanthosis, in the dermis there is a superficial perivascular inflammatory infiltrate, predominantly lymphocytic. According with the skin biopsy findings and the persistence of fever and arthralgia, a rheumatological profile was performed, where ANA's and anti-dsDNA were positive, C3 hypocomplementemia, and the rest of the laboratory test were negative.

Based on the EULAR/ACR 2019 criteria, acute generalized cutaneous lupus erythematosus with systemic involvement was diagnosed. Renal and neurological involvement was ruled out. He began treatment with Prednisone 1 mg/kg/day and Hydroxychloroquine 200 mg every 24 hours, presenting clinical improvement, he was discharged with immunosuppressive treatment.

Discussion:

The atypical clinical presentations of lupus have to be taken into account since the prognosis, treatment, and severity of the disease of erythema multiforme and systemic lupus erythematosus are completely different.

The topography of the erythema multiforme lesions (extremities, photo-exposed areas, and palmoplantar

involvement with a severe mucosal involvement) and the initial morphology of this pathology led us to have this diagnosis initially. Given the clinical findings, Rowell syndrome was ruled out since it does not meet the clinical and serological Torchia's criteria.

The early diagnosis of Lupus impacts on survival since a delay in diagnosis of more than 6 months is associated with a high activity of the disease and in male patients the systemic damage is more serious with a predominance of renal disease and hemolytic anemia.

Conclusions:

It is important to know the atypical presentations of lupus erythematosus, for its early management and to prevent serious systemic complications, especially in populations in which this pathology is infrequent.

Abstract N°: 3408**Safety, tolerability, and pharmacokinetics of TPM203, a tolerizing Topas particle mixture in pemphigus vulgaris: preliminary results from a phase 1, first in man study (EudraCT Number: 2019-001727-12)**

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Introduction & Objectives:

Pemphigus vulgaris (PV) is an autoimmune disorder of the mucous membranes and skin caused by IgG autoantibodies against desmoglein (Dsg) 3 and Dsg1. Growing evidence shows that in PV the production of the pathogenic autoantibodies is largely dependent on the presence of autoreactive T cells.

T cell recognition of a set of epitopes of Dsg3 is restricted by the HLA class II allele HLA-DRB1*04:02, which is prevalent in PV. TPM203 is a mixture of 4 different Topas Particle Conjugates (TPCs), each coupled to a distinct Dsg3 T cell epitope, shown to induce tolerance against Dsg3 in an HLA-DRB1*0402-transgenic mouse model of PV.

Materials & Methods:

We report data from the single ascending dose part of a phase 1, open label study in which four cohorts of three patients each received TPM203 at 0.12, 0.36, 1.2 and 3.6 μmol of total TPC-bound peptide. Patients were followed-up for 12 weeks. Patients had to be in complete clinical remission or with low to moderate clinical disease activity and positive for anti-Dsg3 antibodies as well as for peripheral blood CD4+ T cells specific for at least one of the Dsg3 peptides present in TPM203. No immunosuppressive/immunomodulatory treatment was allowed, other than ≤ 10 mg/d of prednisone equivalent.

Results:

After a single TPM203 administration, 11 out of 12 patients reported at least one adverse event (AE), none of which was severe or serious. Three PV related AEs (lip erosion, oral mucosal erosion, and oral blood blister) were reported as mild events. Worsening of PV was reported in two additional patients following COVID vaccination (one mild and one moderate). During the study, no clinically relevant changes in laboratory parameters were observed.

After TPM203 infusion, detectable levels of the different TPCs could only be found in patients at the two highest doses (from 0.5 h up to 2.5 hours post administration for the highest administered dose).

As to exploratory pharmacodynamic parameters, when pooling all 12 patients, a trend toward a decrease in anti-Dsg3 IgG was found in patients receiving the first three dose levels. The phenotypical analyses of T and B cells revealed the following trends: decrease (% change, 95% CI) in CD27+ memory B cells, increase (% change, 95% CI) in Th2 and Tfh2, decrease (% change, 95% CI) in Th17 cell compartment. Furthermore, a trend towards an increase in Tregs was also observed. A decrease of memory B cells and Th cells (Th17, Th17.1, and Th1 cells) was

associated with diminished anti-Dsg3 autoantibody levels in 5/12 patients.

Conclusion:

In conclusion, escalating single doses of TPM203 were safe and well tolerated in PV patients with no or low/moderate disease activity. Despite the limitations of the study (small patient number, single TPM203 administration, and short observation period), these preliminary data could indicate that this T cell-targeted approach might modulate the immune response in PV patients by increasing Treg cells and decreasing Th17 and memory B cells.

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Abstract N°: 3506

Efficacy of latanoprost, fractional CO2 laser and platelet rich plasma in treatment of localized vitiligo.

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Introduction & Objectives:

Several treatment options are described for vitiligo. Latanoprost, a prostaglandin PGF2 α , has been recently studied for treatment of vitiligo. We aimed to study the the effectiveness of topical latanoprost 0.005% in treatment of vitiligo, versus latanoprost combined with fractional CO2 laser or fractional CO2 laser &PRP in treatment of localized stable vitiligo.

Materials & Methods:

This prospective, parallel group comparative study included 60 patients with localized stable vitiligo. Patients were randomly divided into 3 groups. Group A patients applied topical latanoprost drops once daily for 2 months; group B patients received once daily topical latanoprost drops and 4 fractional CO2 laser sessions at 2 weeks interval; and group C patients received oce daily topical latanoprost drops and 4 fractional CO2 laser sessions combined with platelet rich plasma spaced 2 weeks interval. Clinical assessment of improvement was done by calculating baseline (VASI0) and 3 months after the final session (VASI 3) by an independent observer.

Results:

Mean VASI score dropped from 0.64 ± 0.53 before treatment to 0.39 ± 0.36 after treatment in group A ($p = 0.001$), from 0.69 ± 0.42 before treatment to 0.15 ± 0.16 after treatment in group B ($p = 0.001$) and from 1.05 ± 0.88 before treatment to 0.09 ± 0.09 after treatment in group C ($p = 0.001$). The percentage improvement of VASI score at 3 months after the end of treatment was highest in group C ($91.69\% \pm 2.10\%$) followed by group B ($79.84\% \pm 8.84\%$) and lowest in group A ($44.84\% \pm 9.19\%$). This difference was statistically significant ($p < 0.001$). Erythema was observed in one patient (5%) in group A and B and 2 patients (10%) in group C. Oedema was observed in 2 patients (10%), one patient (5%) and 3 patients (15%) in groups A, B and C respectively. Post-laser crusting was encountered in 3 patients (15%) and 4 patients (20%) in groups B and C respectively. Peri-lesional hyperpigmentation was observed in 2 patients in each of the three groups. The patient reported treatment satisfaction ranged from 5-8 in group A with mean of 6.20 ± 1.01 . It ranged from 6-9 in group B with mean of 7.20 ± 1.08 and ranged from 8-10 in group C with mean of 8.80 ± 0.77 ($p < 0.001$).

Conclusion:

Latanoprost is a known prostaglandin F2 alpha analogue that has been primarily for treatment of glaucoma. One of the reported side effects associated with its usage is hyperpigmentation of periocular skin and eye lashes. This suggested a possible indication for its use in treatment of vitiligo. Previous studies reported its effectiveness in skin regimentation alone or in combination with phototherapy or micro needling as a transdermal delivery system.

Fractional CO2 laser is used for treatment of refractory non-segmental vitiligo. The therapeutic effect is probably mediated by the release of various cytokines and growth factors stimulating the proliferation and migration of melanocytes from the adjacent normal skin. Similarly, platelet rich plasma (PRP) which is high concentrate of various growth factors, is believed to act through stimulation of regimentation via stimulation and proliferation of

melanocytes.

We conclude that topical latanoprost 0.005% enhances the therapeutic efficacy of fractional CO2 laser and PRP in treatment of localized stable vitiligo.

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Abstract N°: 3507**Decreased serum level of soluble PD-L1 in discoid lupus erythematosus, but not in subacute cutaneous lupus erythematosus – does it hold any clinical significance?**

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Introduction & Objectives:

Cutaneous lupus erythematosus (CLE) is an autoimmune skin condition that presents in various clinical forms, such as discoid lupus erythematosus (DLE) and subacute cutaneous lupus erythematosus (SCLE). The altered function of the programmed cell death 1/programmed cell death ligand 1 (PD-1/PD-L1) pathway has been suggested as a contributing factor in the development of CLE. This study aimed to investigate the presence of soluble forms of PD-1 (sPD-1) and PD-L1 (sPD-L1) in untreated DLE and SCLE.

Materials & Methods:

The levels of sPD-1 and sPD-L1 were measured using an enzyme-linked immunosorbent assay in the serum of 21 DLE, 18 SCLE, 13 systemic lupus erythematosus (SLE) patients, and 20 healthy controls (HCs). Mann-Whitney U-test and Spearman's rank correlation were performed to assess differences between patient groups and HCs and to examine any relationship between the severity of skin symptoms and sPD-1/sPD-L1 levels.

Results:

Regarding sPD-1 levels, no significant differences were observed between the DLE and SCLE groups or when compared to HCs. However, significantly lower levels of sPD-L1 were found in the DLE group compared to both the SCLE and HC groups ($p=0.027$ and $p=0.009$, respectively). In SLE patients, significantly higher levels of sPD-1 were detected compared to HCs ($p=0.002$). No association was found between the severity of skin symptoms and sPD-1/sPD-L1 levels.

Conclusion:

These findings suggest that alterations in the inhibitory effect of sPD-L1 on T-cell activity could potentially explain the differences between DLE and SCLE.



Abstract N°: 3650

Focal pneumopathy in Dermatomyositis: an uncommon entity

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Introduction & Objectives:

Dermatomyositis is a rare autoimmune disease affecting mainly skin and muscle, but that may be associated with other visceral manifestations. Pulmonary involvement, in particular, is variable in frequency and potentially severe. Besides the classical forms, focal pneumopathy is a much less common type, but of primary importance due to its potential severity and the need for appropriate treatment.

Materials & Methods:

We report the case of a 44-year-old female patient with history of pulmonary tuberculosis treated 25 years ago; admitted for erythema of the face and back of the hands evolving for 4 months and associated with arthralgias and NYHA class II dyspnea, without cough or chest pain or any other associated sign. Clinical examination revealed cutaneo-muscular signs suggestive of dermatomyositis. Complete workup revealed elevated muscle enzymes, a negative immunological panel, and a myogenic pattern on the electroneuromyogram. Chest CT revealed a left anterobasal alveolar condensation, biopsies performed during bronchoscopy showed nonspecific fibroinflammatory alterations, and tests for Mycobacterium tuberculosis, Aspergillus and Pneumocystis jirovecii were negative. The patient was initiated on prophylactic antibiotic therapy and corticosteroids 1mg/kg/d after excluding infectious origin, with improvement of the dyspnea and the scannographic lesion after 2 months.

Discussion:

The originality of our work lies in the rarity of focal lung involvement in dermatomyositis.

Dermatomyositis is a rare and potentially serious multisystemic disease. Pulmonary involvement, found in 30-50% of cases, mainly affects groups with anti-MDA5 antibodies and must be detected as it is a major cause of mortality. More frequent in adults, it is typically due to Interstitial lung disease, hypoventilation due to respiratory muscle impairment, or aspiration pneumonia related to swallowing disorders. However, there are uncommon and consequently little-known forms with a similar prognosis.

Organized pneumonia, rarely associated with dermatomyositis, is variable in manifestation, ranging from asymptomatic forms to dyspnea, cough, and even acute respiratory symptoms.

High resolution CT scan is the radiological examination of choice, showing areas of subpleural condensation and linear opacities located preferentially in the lower lobes and posterior regions. Histological signs consist of non-specific inflammatory lesions.

Lastly, the first-line treatment is based on corticosteroids at a dose of 1mg/kg/d, in combination with an immunosuppressant in case of non response.

Conclusion:

The Dermatomyositis - Organized Pneumonia association is rare. Data concerning it are limited because of this rarity and the absence of diagnostic confirmation in certain cases. However, it remains important to know about it

in view of its prognostic impact and the adaptation of therapy it implies.

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Abstract N°: 3814**Mortality, and causes of death in patients with pemphigus in Tehran, Iran**Nastaran Namazi¹¹Shahid Beheshti University of Medical Sciences, Skin Research Center, Tehran, Iran**Mortality, and causes of death in patients with pemphigus in Tehran, Iran**

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Introduction & Objectives: Pemphigus consists of a group of rare autoimmune bullous diseases that affect the skin and mucous membranes. pemphigus includes three major forms: pemphigus vulgaris, pemphigus foliaceus, and paraneoplastic pemphigus. Before the advent of systemic corticosteroids, pemphigus vulgaris was usually a fatal disease, pemphigus foliaceus had a better prognosis. Rituximab (RTX), a monoclonal antibody against the CD20+ B cells has been approved for the treatment of patients with PV. Previous studies have confirmed the high efficacy and safety profile of RTX in patient. We aimed to estimate the overall mortality, causes of death among pemphigus patients who were admitted to Shohada-e- Tajrish and Loghman-e-Hakim hospitals in Tehran, Iran. The cause-specific mortality was determined among patients with pemphigus after administrating of rituximab (RTX) compared with those not receiving Rituximab.

Materials & Methods: 480 patients who were admitted either to Shohada-e- Tajrish or to Loghman-e-Hakim Hospitals in Tehran, Iran between October 2010 and October 2022 were included in the study. The diagnosis of all of patients was confirmed by direct immunofluorescence and pathological studies. All variables such as age, sex, type of pemphigus, presence of comorbidities, medications, and cause of death were assessed, and the effects of RTX treatment on the patient's mortality were evaluated.

Results: The prevalence of pemphigus was slightly higher in women compared to men (N=262, 54.58 % vs. N=218, 45.41 % respectively). Pemphigus vulgaris (PV) was the most common type (N= 474 , 98.75%) followed by pemphigus foliaceus (N= 4 , 0.83 %) and paraneoplastic pemphigus (N=2 , 0.41 %). The most common comorbidities were Hypertension (N=98 , 20.41%) and Diabetes mellitus (N= 93, 19.37%).The overall mortality was 20 (4.16%). 15 patients out of 20, were under treatment with high dose systemic corticosteroids and immunosuppressive agents (75 %) while 5 patients had received at least 500 mg of RTX and low dose systemic corticosteroids (25%).

Conclusion: The mean age of the disease was found to be a decade earlier than other parts of the world, with an almost higher preponderance of women. The most common comorbidities were Hypertension and Diabetes mellitus. These results showed the efficacy of Rituximab in treating patients with moderate to severe PV, while tapering off of corticosteroid therapy. Overall, RTX treatment had a positive effect on remission of patients.

Abstract N°: 3985**Spanish multicenter retrospective study of real-life experience with Omalizumab for the treatment of bullous pemphigoid.**

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Introduction & Objectives:

Bullous pemphigoid (BP) is the most common autoimmune blistering disease. This disorder typically affects the elderly, leading to high morbidity and a severe impact on the quality of life. Current treatment strategies depend on the severity of the disease and patients' comorbidities. Oral corticosteroids and other immunosuppressive therapies are frequently contraindicated in this population. The use of doxycycline and dapsone is often ineffective for moderate to severe disease. In this scenario, the use of new drugs such as Dupilumab or Omalizumab is showing promising results.

Our objective is to assess the real-life efficacy of Omalizumab for the treatment of BP in different Spanish hospitals. We would also like to describe patients' general health status, BP subtypes and IgE levels, trying to define the patients who would benefit the most from this treatment.

Materials & Methods:

We carried out a multicenter retrospective study including patients from 15 tertiary hospitals in Spain. Patients diagnosed of BP who received omalizumab for at least 3 months were included. The study was approved by the ethical committee of Doctor Peset University Hospital in Valencia, Spain.

Results:

A total of 36 patients were included. The mean age at diagnosis was 79.6 years with 57% of male patients. The vast majority of the patients were fragile with several comorbidities. The mean G8 geriatric screening tool (used to assess the general status) was 10.3 indicating a poor general health status.

The mean follow-up period from the BP diagnosis was 4.5 years. All patients had used other systemic therapies before Omalizumab (35% azathioprine, 35% doxycycline, 30% methotrexate, 30% dapsone, 15% mycophenolate, 3% Dupilumab, 3% iv Ig, 5% rituximab). Most patients presented with bullae and erosions (75%), while 25% had with urticarial lesions. To assess the severity and extent of BP prior to Omalizumab we used the body surface area (BSA) involvement, BSA <20: 52% of patients; BSA 20-40: 15% and BSA >40: 33%.

Omalizumab was administered subcutaneously at a dose of 300 mg every 4 weeks in most patients, however, in 20% of patients doses were increased (450-600mg every 4 weeks). The mean duration of treatment was 13.5 months, with a mean of 2.8 months to reach the maximal response. Regarding the response, 17% of patients did not present any improvement while the remaining 83% experienced any kind of treatment response (43% complete response on treatment (CR), 25% good response (defined as >50% BSA improvement), 17% partial response (defined as <50% BSA improvement)). Although mean IgE serum levels prior to treatment were 555 kU/L (0-200), we did not find any correlation between higher levels and a better response. All the patients tolerated Omalizumab without side-effects reported.

Conclusion:

Although Omalizumab is only approved for asthma and spontaneous idiopathic urticaria, it has shown good results in other dermatological conditions such as urticarial vasculitis or BP. Recent systematic reviews report CR rates ranging from around 55 to 68% in BP patients. Our series demonstrates a treatment response in 83% of patients; however, only 43% achieve a CR. We have to take into account that other immunosuppressive drugs frequently prescribed in BP raise concerns about adverse effects in elderly patients and often prove ineffective. In this scenario, Omalizumab seems to be a promising therapy for BP, offering a good response rate and favorable safety profile.



Abstract N°: 3995**Adverse pregnancy outcomes in Pemphigoid gestations**Sophie Preuß¹, Ralf Ludwig^{1, 2}¹University Medical Center Schleswig-Holstein, Department of Dermatology, Allergology and Venerology, Lübeck, Germany, ²Institute of Experimental Dermatology, Lübeck, Germany**Adverse pregnancy outcomes in Pemphigoid gestationis****Introduction & Objectives:**

Pemphigoid gestationis (PG) is a rare autoimmune blistering disease that affects pregnant women. Clinical presentation is characterized by pruritus and blistering skin lesions that typically occur during the second or third trimester of pregnancy or immediately postpartum. PG is caused by a disruption of immunotolerance against the hemidesmosomal antigen BP180. Topical corticosteroids are the primary treatment, and in refractory cases, systemic glucocorticoids or other immunosuppressive agents may be used. So far, data from small cohort studies suggest an increased risk for preterm labor and fetal growth restriction in PG patients, hinting towards impaired placental function. However, as PG is rare, large sample studies exploring other adverse pregnancy outcomes (APO) are lacking. Thus, the aim of this study was to examine the impact of PG on the risk of APO in a substantial cohort.

Materials & Methods: This retrospective matched cohort study was conducted using the US Collaborative Network of TriNetX. All females aged 12-55 were screened for the presence of the diagnostic code for PG (O26.4). Participants with PG were matched 1:1 with participants without PG using propensity score matching for demographic variables and comorbidities (smoking, obesity, diabetes mellitus, hypertension, and chronic kidney disease).

Baseline characteristics were described using means and standard deviations or percentage and compared using student t-test or Pearson chi-square test. Participants were analyzed for the onset of the following outcomes 12 months after the diagnosis of pregnancy: Preterm labor, pre-eclampsia, HELLP syndrome, eclampsia and gestational diabetes. Survival analyses were conducted using the Kaplan-Meier method, and outcome distribution differences between the two groups were assessed using a log-rank test. Hazard ratios (HR) for the risk of the outcomes were obtained using the Cox regression model. To account for multiple comparisons, a Bonferroni adjustment was applied, considering p-values less than 0,002 as statistically significant.

Results:

The study population comprised 27695 participants in each group. Mean age in the PG-group and non-PG-group was 27,3 ± 5,95 and 27,3 ± 5,96 years, respectively.

No significant variability in demographic variables or the prevalence of smoking, obesity, diabetes mellitus, hypertension and chronic kidney disease were found between the two groups. Compared to participants without PG, those with PG had a higher risk of preterm labor (hazard ratio (HR), 9,62; 95% confidence interval (CI), 8,34-11,09; $P < 0,0001$), pre-eclampsia (HR, 2,92; 95% CI, 2,56-3,35; $P < 0,0001$), HELLP syndrome (HR, 4,79; 95% CI, 3,78-6,07; $P < 0,0001$), eclampsia (HR, 2,72; 95% CI, 2,13-3,47; $P < 0,0001$) and gestational diabetes (HR, 8,02; 95% CI, 6,31-10,19; $P < 0,0001$).

Conclusion:

In conclusion, this study demonstrates that PG can significantly affect the course of pregnancy, increasing the risk for multiple APO. These APO pose a potential threat to the lives of both the mother and fetus. Thus, results from this study imply that mothers with PG should be closely monitored for APO. Managing PG should involve a multidisciplinary approach, with close collaboration between dermatologists, obstetricians, and neonatologists.

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Abstract N°: 4031**Recalcitrant infantile bullous pemphigoid**

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Introduction:

Bullous pemphigoid (BP) is an autoimmune subepidermal blistering dermatosis primarily affecting older adults. In children, it is far less common. We report a case of infantile BP that presents a therapeutic difficulty.

Case presentation:

A female infant presented at the age of 12 months with a profuse pruritic bullous skin rash associated with oral and genital mucosal involvement. The biological evaluation revealed a hypereosinophilia of 3400/mm³. Skin histology showed a subepidermal bulla. The direct immunofluorescence was negative. Bullous pemphigoid (BP) was identified by Indirect immunofluorescence on salt-split skin which revealed the presence of antibodies against the basement membrane on the epidermal side.

Dermocorticoid therapy in combination with prednisone 0.5 mg/kg/day resulted in an incomplete and transient remission. Prednisone was increased to 1 mg/kg/day with good response however, flare-ups appeared when corticosteroid medication was tapered off. The addition of dapsone (2.5 mg then 5 mg/kg/d) only partially improved this cortico-dependence (relapse below the 0.3 mg/kg/d threshold). There was also a delay in the growth rate (-4 DS) and weight (-2 DS) and the development of a cushingoid appearance.

Discussion:

Bullous pemphigoid in children is uncommon and is distinguished by preferential palmoplantar and mucosal involvement. It is classically treated successfully with corticosteroid therapy (local and/or general). In case of cortico-resistance or cortico-dependence, the therapeutic strategy is not codified. Cases documented in the literature received a variety of therapies, including ciclosporin, azathioprine, mycophenolate mofetil, intravenous immunoglobulins, rituximab, and dapsone, which was used most frequently.

In our case, recourse to immunosuppressive treatment became necessary, because of the corticoid dependence observed despite the use of high-dose dapsone, and the occurrence of corticosteroid side effects (iatrogenic Cushing's disease and severe growth delay)

Conclusion:

Our observation reports a rare case of chronically evolving infant BP with cortico-dependence requiring the use of immunosuppressive therapy.

Abstract N°: 4033**Analysis on Integrated lncRNA and mRNA Expression in Systemic Lupus Erythematosus by Transcriptomics**Limei Yuan¹, Wenting Cao¹, Binbin Yang¹, Danqi Deng¹¹The Second Affiliated Hospital of Kunming Medical University

Introduction & Objectives: Systemic lupus erythematosus (SLE) is an autoimmune disease. SLE patients with a high SLE disease active index (SLEDAI) may easily suffer from unexpected complications. Several investigations have reported dysregulated gene expression in SLE patients, but gene expression patterns in patients with high SLEDAI scores remain unknown. To reveal the integrated expression of mRNA and lncRNA in SLE.

Materials & Methods: Transcriptomics and bioinformatics analysis was employed to reveal the integrated expression of mRNA and lncRNA in 13 SLE patients and 8 healthy controls.

Results: 4184 mRNAs and 766 lncRNAs were expressed in the SLE patients. Among the genes that were expressed differentially ($P < 0.05$), 348 of 383 mRNAs were up-regulated, 56 of 59 lncRNAs were up-regulated. The majority of differentially expressed mRNAs and lncRNAs was up-regulated in SLE patients. KEGG analysis demonstrated that the up-regulated mRNAs were enriched predominantly in the SLE signaling pathway. GO analysis revealed that the up-regulated mRNAs were mainly enriched in neutrophil degranulation and activation. A key module was identified to be associated with SLE by Weighted Gene Co-Expression Network Analysis. 4 lncRNAs (LINC01484, AL133467.1, AC012236.1 and AC010149.1), targeted at 187 mRNAs in the key module, and 35 of them were found to be neighbored to the 4 lncRNAs. The expression of LINC01484, AL133467.1, and AC012236.1 was significantly higher in SLE patients than that in control by q-RT PCR analysis.

Conclusion: The study reveals a comprehensive expression profile of lncRNAs and mRNAs in SLE patients. LINC01484, AL133467.1, and AC012236.1 may be potential lncRNA bio-markers of SLE.



Abstract N°: 4079

Comedogenic lupus: a rare variant of chronic cutaneous lupus erythematosus - case series

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Introduction & Objectives: # To report clinicopathological characteristics of 2 new cases of comedogenic lupus (CL), a rare variant of chronic cutaneous lupus erythematosus. **Materials & Methods:**

Retrospective study of patients with clinical and histopathological diagnoses of comedogenic lupus between the years 2010-2022.

Results:

Patient 1. A 51-year-old female patient with hypothyroidism presented with pruritic lesions as comedones distributed on both elbows, ears, the preauricular region, knees, and palms with erythematous-violaceous peripheral halos, which appeared after sun exposure. The patient also reported asthenia, arthralgia, and mild hair loss. A punch biopsy showed follicular plugging, perivascular dermatitis, and lymphomonocytic inflammatory infiltrate. The patient was positive for antinuclear antibodies at 1:40, thrombocytopenia, and decreased serum complement C3 levels. These findings were consistent with CL but did not meet the criteria for systemic lupus erythematosus (SLE). The patient was treated with oral prednisone, oral hydroxychloroquine (HCQ), and avoidance of sun exposure, resulting in great improvement in the lesions and resolution of thrombocytopenia after two months of treatment.

Patient 2. A 47-year-old female patient, smoker, diagnosed with discoid lupus erythematosus (DLE) for two years. She developed multiple open comedones on previous DLE plaques on the auricular area. She denied systemic symptoms. A punch biopsy revealed hyperkeratosis, liquefaction degeneration of the basal layer and large follicular plugs. ANA was negative, without clinical and laboratory criteria for SLE. The patient was treated with oral HCQ, topical corticosteroids and avoidance of sun exposure, resulting in a resolution of the lesions after one month.

Conclusion:

CL is a rare form of CCLE of unknown etiology. It predominates in women between the 3-4 decades of life, although only 23 cases have been described in the literature. The clinical manifestations include comedones, erythematous papules, and punctate scars affecting sun-exposed areas, usually itchy.

The histological findings are similar to those seen in DLE, including hyperkeratosis, epidermal thinning, liquefaction degeneration of the basal layer, thickening of the basement membrane and lymphocytic inflammatory infiltrate in the papillary and periadnexal dermis.

Differential diagnoses include acne vulgaris, comedogenic nevus, and Favre-Racouchot disease.

The treatment can be challenging, and photoprotection is essential in all cases. Topical therapy with retinoids and the use of topical and/or intralesional corticosteroids may contribute to improvement. However, most cases require systemic therapy with HCQ being considered the first line of treatment.

Due to its rarity and little knowledge of CL by dermatologists, the diagnosis can be delayed, with a negative

impact on quality of life, since it is a dermatosis with the potential for significant unaesthetic complications.

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Abstract N°: 4236

Impressive ulceronecrotic skin lesions revealing a Henoch-Schönlein purpura

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Introduction & Objectives:

Rheumatoid purpura or Schönlein-Henoch purpura (HSP) is a systemic vasculitis of small vessels related to (IgA) tissue deposits and circulating immune complexes that mainly affects children .

Systemic involvement is frequent in adults forms and cutaneous manifestations tend to be more commonly complicated by necrotic or hemorrhagic bullous . Although ,ulcerative forms still exceptional.

We report a case of adult rheumatoid purpura revealed by necrotic plaques with multiple and extensive superinfected ulcerations of the lower limbs

Case report :

A 49-year-old woman, presented to emergency with a two-month history of evolutive cutaneous lesions of the lower limbs associated to a severe oligoarthropathy of ankles and knees. Her past medical history included insulin-dependent diabetes and ischemic cardiomyopathy. She had no traumatism and her social and family history were unremarkable.

The clinical examination revealed a purpuric vasculitic rash with multiple polycyclic ulcerations , with pigmented edges and variable depth and sizes , covered with a greenish yellow crusts and necrotic patches. The rest of the examination showed no particularities.

The laboratory blood findings revealed a neutrophilic leucocytosis with a white cell count of 14000/uL ,inflammatory markers were raised (C-reactive protein (CRP) 62 mg/L and erythrocyte sedimentation rate 109mm/h). 24h Urine sample was positive for blood (263000/ml), and protein (0.96g/24h).

The rest of the full blood count, glomerular filtration rate, urea and electrolytes, liver function tests and coagulation panel were normal. Autoimmunity tests were within normal ranges.

At the bacterial and fungal cultures the isolated microorganism was identified as sensible Escherichia coli.

Cutaneous biopsy showed leukocytoclastic vasculitis with perivascular IgA and C3 deposition .

Based on these investigation findings ,a diagnosis of ulceronecrotic rheumatoid purpura was suspected .

Given the biological renal involvement, a renal biopsy was required and revealed a Henoch -Schönlein purpura nephritis (HSPN) . The diagnosis was confirmed.

The patient received 14 days of C3G 2g/day for with Gentamycin 3mg/kg/day for 5 days with excision of necrotics tissues. Over the next two weeks, the infection improved and treatment with 1mg/kg of oral prednisone could be started. After 20 days the skin involvement has clearly improved and proteinuria has started to normalize.

Discussion:

Henoch-Schönlein purpura (HSP) is the most common systemic vasculitis in children . Adults are less affected

,cutaneous manifestations of HSP are typically benign and self-limiting, although they can include rarer skin complications such as haemorrhagic, bullous, necrotic lesions.

Leg ulcers are a less frequent presentation of HSP and reflect the severity and extent of the disease. The precise pathogenesis is not yet known but may be explained by skin necrosis due to vasculitis.

Management is twofold and is based on both treatment of the vasculitis with corticosteroids, immunosuppressants, colchicine, dapsone, or others, depending on the indication, and local treatment of the ulcer to promote rapid and aesthetic healing.

Conclusion:

Although exceptional, rheumatoid purpura remains an etiology of leg ulcers that should not be overlooked. The association of leg ulcers with purpuric lesions and systemic signs, mainly articular, digestive and especially renal, should also raise the diagnosis of IgA vasculitis .

Abstract N°: 4265**Pemphigus foliaceus with an erythema gyratum repens -like pattern.**

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Introduction:

The clinical presentation of Foliaceous pemphigus is pretty well described and typical, it involves the development of superficial erosions with scaling and crusting within the seborrheic areas: scalp, face, chest and upper back, nevertheless it is known by its clinical polymorphism.

Erythema gyratum repens (EGR) on the other hand is a rare cutaneous eruption characterized by concentric erythematous moving bands forming a wood-grain appearance. It is considered to be a paraneoplastic eruption in 70% of the time, but some cases are related to benign diseases. Some dermatoses can mimic EGR and therefore called EGR-like eruptions.

Here we report the case of a superficial pemphigus of particularly spectacular presentation.

Case report:

A 68-year-old man, presented with erosions and scaly, erythematous patches initially located on the back and arms, subsequently extended to buttocks and inner thighs, without any real blisters ever seen, furthermore there were no mucosal involvement. On the 4th day of his hospitalization in our department, physical examination revealed new plaques arranged on erythematous annular lesions forming concentric rings, expanding centrifugally, suggesting an erythema gyratum repens (Nikolsky test was positive). Skin histology showed acantholysis with sub corneal blistering, direct immunofluorescence revealed intercellular IgG/C3 deposition in the epidermis, with a honeycomb fluorescence pattern. A whole blood workup was done including tumor markers, computed tomography (CT) scan, Anti-BP180, Anti BP230, were all negative, Anti-SIC were positive. The final diagnosis of foliaceus pemphigus was therefore established. Treatment with prednisone (1,5mg/kg/day) and dapsone 100mg/day was initiated along with daily showers and bandages to avoid infection. The eruption completely cleared within 3 months

Discussion:

We report an atypical case of bullous dermatosis presenting as EGR, revealing pemphigus foliaceus. Diagnosis of Foliaceous pemphigus was made on clinical features (an exfoliative dermatitis made with erythematous scaly patches and erosions, the absence of tense blisters and genital or oral involvement) and histopathological analysis. Our patient presented an atypical clinical presentation with EGR features, annular arrangement of plaques or blisters with an EGR-like eruption is reported in very few cases of bullous dermatosis such as linear IgA dermatosis, bullous pemphigoid, bullous lupus erythematosus, anti p-200 pemphigoid but no case of an EGR-like foliaceus pemphigus was ever published to our knowledge. In our case, the negativity of paraclinical tests in search of associated malignancy and the quick response to dapsone and corticosteroids, were sufficient to exclude an authentic EGR.

Conclusion:

To conclude, we report an atypical clinical presentation of foliaceus pemphigus, with an EGR pattern, successfully

treated by corticosteroids and dapsone, our case illustrates the heterogeneous clinical presentation of pemphigus foliaceus.

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Abstract N°: 4405**Bullous lupus: a rare manifestation of systemic lupus erythematosus**

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Introduction:

Bullous lupus (BL), a subgroup of subepidermal autoimmune bullous dermatoses, is a rare cutaneous manifestation of systemic lupus erythematosus (SLE). It mainly affects young women. We report a new case.

Case presentation:

A 20-year-old female patient, with SLE since the age of 12, presented with a systemic flare-up (lupus nephropathy stage II, hematologic and ocular involvement) of her lupus, treated with oral prednisone at an initial dose of 1 mg/kg/d. One month later, pseudo-urticarial plaques with vesiculo-bullae appeared on the neck, trunk and upper limbs. Skin histology revealed a subepidermal blister associated with a polymorphic dermal inflammatory infiltrate. Direct immunofluorescence showed linear deposits of IgG and C3 at the dermal-epidermal junction. Indirect immunofluorescence for anti-basal membrane antibodies was positive. The diagnosis of LB was made and the patient was prescribed 100 mg/day of dapsone. The evolution was marked by the complete disappearance of the bullous lesions in a few days.

Discussion:

Bullous lupus is uncommon non-specific skin lesion of SLE. It primarily affects young women. Clinically, it is distinguished by bullae or vesiculo-bullae, on erythematous or healthy skin. The lesions occur on covered or exposed areas. This bullous manifestation may occasionally be the first sign of lupus.

Histologically, it presents as subepidermal blisters with an inflammatory infiltrate of neutrophils and eosinophils. The cleavage is located in the superficial dermis. Direct immunofluorescence is positive, with IgG, IgM, or IgA deposits at the dermo-epidermal junction. Anti-collagen type VII antibodies are frequently used to identify BL. It is frequently associated with lupus nephritis, as observed in our patient's case. BL shows a remarkable response to dapsone, with the disappearance of the bullae without scarring.

Conclusion:

Bullous lupus illustrates the great clinical polymorphism of lupus disease. It is often associated with serious visceral damage, especially renal. Its treatment of choice is dapsone.

Abstract N°: 4419**Pyoderma gangrenosum and systemic lupus erythematosus: when inflammatory pathology meets autoimmune disease**Billel Merrouche¹, Nour el Houda Mezhoudi¹, Houria Sahel¹¹University Hospital Center Lamine Debaghine, Department of Dermatology, Bab El Oued, Algiers, Algeria**Introduction:**

An uncommon neutrophilic dermatosis known as pyoderma gangrenosum (PG) causes inflammatory and aseptic skin ulcerations. The association of PG with systemic lupus erythematosus (SLE) is rarely described in the literature. We report a new case.

Case presentation:

A 20-year-old female patient, followed for SLE since the age of 12, presented with a systemic flare-up (renal, hematological, ocular and cutaneous bullous lupus) of her lupus, treated with oral prednisone at an initial dose of 1 mg/kg/d and dapsone at 100 mg/d. Four weeks later, she developed a deep left cervical ulceration, measuring 5 cm in diameter with an increased inflammatory border at the site of a skin biopsy. The diagnosis of PG was retained and the patient was restarted on general corticosteroid therapy at 1mg/kg/d with guided wound healing. After two months, the ulceration had completely healed.

Discussion:

A pathology is present in about 50% of PG patients. It is most often a digestive or rheumatological inflammatory disease. Its association with lupus is rare and poorly understood. PG mostly affects young individuals and typically manifests during the chronic course of a previously diagnosed lupus, as in the case of our observation. The pathogenesis of both diseases is complex, a dysfunction of neutrophils is suggested by some authors as a common pathophysiological mechanism. Our patient developed a unifocal ulceration at the site of a biopsy, consistent with a pathergy phenomenon. The latter is reported in approximately

one-third of PG cases. The prognosis of lupus does not appear to be worsened by PG. The treatment of

PG is usually based on drugs that control neutrophil activity (colchicine, dapsone, corticosteroids and cyclin). Our patient was successfully treated with systemic corticosteroids.

Conclusion:

Although uncommon, the association of SLE and PG should be known. It shows the transition between autoimmunity and inflammatory pathology.



Abstract N°: 4447**Case report: Expect the unexpected – what can be uncovered by Dermatitis Herpetiformis**

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Introduction & Objectives:

Dermatitis herpetiformis (DH) is a chronic, pruritic, autoimmune blistering disorder associated with gluten-sensitive enteropathy. In addition to celiac disease, other diseases like autoimmune thyroid disease, type I diabetes mellitus, and pernicious anemia may occur at increased frequencies in patients with DH. Studies have also observed the link between DH and an elevated risk of cancer, particularly due to the higher frequency of leukemias and lymphomas, as well as the link between celiac disease and an increased risk of non-Hodgkin's lymphoma.

Materials & Methods:

This presentation is based on a case report of a patient diagnosed and treated in our dermatovenereology clinic.

Results:

We present the case of a 63-year-old male patient with a history of treated gastric ulcer and no chronic medication at the time of admission. The patient came to our clinic with a generalized rash consisting of papules, grouped vesicles, and erythematous plaques with erosions on the surface, covered by hematic crusts in some places, blisters in the periphery. The rash was accompanied by intense itching and had been evolving for approximately one year. The patient had previously undergone multiple topical (dermatocorticoids, antiparasitary) and systemic treatments (antihistamines, systemic corticosteroids), but had no medical documentation in this regard. In April 2022, a skin biopsy was performed in another hospital, and the histopathological examination raised the suspicion of bullous pemphigoid. Following another skin biopsy that was performed in our department in March 2023, the histopathological features were also highly suggestive of bullous pemphigoid (pre/post bullous stage), but further immunofluorescence testing was recommended. The immunohistochemical examination revealed granular deposition of IgA at the dermo-epidermal junction, without deposition of IgG, C3c, IgM or fibrinogen, which established the positive diagnosis of dermatitis herpetiformis. Laboratory studies identified inflammation (ESR 33 mm/h), an elevated neoplastic screening marker (CEA 9.82 ng/mL) and positive celiac disease antibodies (immunoglobulin A anti-tissue transglutaminase). The patient also reported gastrointestinal symptoms: abdominal bloating, cramping and pain. A truly unexpected finding emerged during the abdominal ultrasound, which identified at the level of the medio-renal parenchyma a tumor of approximately 3.8/3.3 cm, round-ovalar, with an irregular contour, heterogeneous echostructure with anechoic areas, and intense color doppler signal, being intensely vascular.

Conclusion:

One notable aspect of this case is the rare association between paraneoplastic DH and solid tumors. This association is more commonly observed in patients with hematologic disorders, such as lymphomas and leukemias. Therefore, clinicians should always consider conducting a comprehensive neoplastic screening in

patients diagnosed with DH, as well as including the disease in the differential diagnosis when a patient with a solid organ neoplasia presents with a pruritic, blistering eruption. Another noteworthy aspect is the patient's medical history, as some population-based studies have indeed revealed an association between DH and bullous pemphigoid.

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Abstract N°: 4462**Potential use of anti-IL23 drugs in autoimmune blistering diseases**

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Introduction & Objectives:

Despite knowledge of the epidemiological association between psoriasis and bullous pemphigoid (BP), the possible existence of a common pathway, and the consequent possibility of employing a targeted treatment against it, remains an unknown.

Materials & Methods:

We present a case of a patient with psoriasis, psoriatic arthritis and BP refractory to several lines of treatment, which finally resolved completely with guselkumab.

Results:

A 48-year-old man who had been followed for 12 years for moderate plaque psoriasis only treated with topical corticosteroids presented with a worsening of his psoriasis and an outbreak of BP (confirmed after a biopsy, a direct immunofluorescence test and a serology).

Azathioprine and cyclosporine failed to achieve control. Methotrexate was effective but it had to be discontinued due to thrombocytopenia. Etanercept was started due to the onset of psoriatic arthritis and the response was positive for 6 years, but it was stopped because of the finding of chondroid neoplasm of uncertain malignant potential.

A combination of omalizumab and doxycycline was prescribed. Nevertheless, the BP was not controlled and the psoriasis worsened to PASI 10, so after 7 months he switched to guselkumab monotherapy. One month after the first dose, the patient presented PASI 0, and one month later the BP lesions disappeared. To date, 2 years after the initiation, he continues with guselkumab and remains with PASI 0, no radiologic signs or symptoms of arthritis, and no new outbreaks of BP.

The association between psoriasis and BP is described in the literature at epidemiological level. The pathophysiological substrate could be the architectural alteration of the basement membrane found in psoriasis, both in the plaque itself and in apparently healthy skin. The most relevant findings are the disintegration and irregular distribution of laminin and fibronectin 3, which lead to a weakening of the basement membrane that may facilitate its damage in case of intercurrent with BP.

Furthermore, it seems that elevated levels of IL-23 are not only elevated in psoriasis, but also in BP. IL-23 could play a relevant role in that it produces an overexpression of matrix metalloproteinase (MMP)-9, which weakens the basement membrane.

The mechanism whereby guselkumab has been effective in our case remains unknown. A first option could be that the regulation of the IL-23 axis directly controls BP. Another possibility is that guselkumab improves psoriasis primarily, thus achieving a stabilization of the basement membrane that avoids the concurrence of BP.

Conclusion:

The relation between anti-IL23 drugs and BP is not defined. However, there are data that support the biological plausibility that they may be effective in this pathology.

Therefore, they could be considered in the future as a therapeutic alternative in multirefractory cases, as long as more studies are carried out to evaluate their safety and efficacy for this indication.

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Abstract N°: 4499

Clinico dermoscopic correlation of discoid lupus erythematosus on glabrous skin : prospective study of 12 cases

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Introduction & Objectives:

Discoid lupus erythematosus (DLE) is the most common subtype of chronic cutaneous lupus erythematosus and accounts for 50 to 85% of cases. Although dermoscopy is of interest in pigmented lesions and inflammatory lesions, such as differentiating scalp lupus from other causes of scarring alopecia, there is limited data regarding the use of dermoscopy in DLE. However, there is limited data regarding the dermoscopic criteria of discoid lupus of the hairless skin.

Objective: to describe the dermoscopic aspects characterizing cutaneous discoid lupus and to correlate them with disease progression.

Materials & Methods:

Prospective study conducted between September 2022 and March 2023 that included all patients with a histologically confirmed diagnosis of cutaneous DLE examined by Dermlite DL4 dermoscopic in polarized mode.

Results:

During this period: 12 patients were collected, of which two were male, and ten were female. The average age was 30 years (ranging from 14 to 43 years). The cheeks were the most frequently affected site (8 patients), followed by the scalp (2 patients), the trunk (1 patient) and the legs (1 patient). Our patients were subdivided into two groups: the first group of four patients had an evolution of less than 6 months with erythematous lesions, the second group of eight patients had an evolution of more than 6 months with mainly dyschromic lesions.

The first group had all a peri follicular whitening halo, and a background erythema. Three patients had follicular keratotic plugs in the form of yellow dots with perifollicular desquamation in one patient. Thick scales associated with polymorphic vessels were present in one patient.

In the second group, epidermal atrophy was present in all patients associated with perifollicular honeycomb pigmentation in 7 patients, and dotted telangiectasias in one patient and polymorphic in 6 patients. White areas without structures were present in 5 patients.

Conclusion:

In the present study, we report the frequency of dermoscopic criteria seen in DLE localized to areas other than the scalp and their possible correlation to disease progression. Perifollicular whitish halo, follicular keratotic plugs, and thick or perifollicular scales were the main signs of recent and still active lupus. This is similar to other series, which have demonstrated these signs in both newly diagnosed patients and those followed for several years who are in relapse. These studies suggest that the whitish halo may correspond to perifollicular fibrosis on histopathology.

In contrast, old and stable discoid lupus is characterized by perifollicular pigmentation giving a honeycomb appearance, whitish areas without structures as well as telangiectasias. This can be explained by the healing

process in DLE which is initiated at the level of the hair follicle with destruction of the elastic sheath and then progressively extends to involve the dermis between the follicular units resulting in fibrosis which corresponds dermoscopically to the whitish areas without structures.

The results of our study confirm those of the literature. The different dermoscopic aspects of DLE underline the interest of this non-invasive tool in the positive diagnosis of DLE and its evolution, thus allowing the clinician to evaluate the therapeutic response.

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Abstract N°: 4575**Comedonic lupus: a rare variant of cutaneous lupus erythematosus**Mafalda Pestana¹, Alexandre João², Joana Barbosa²¹Centro Hospitalar Universitário de Lisboa Central, Dermatology and Venereology, Portugal, ²Centro Hospitalar Universitário de Lisboa Central, Dermatology and Venereology**Introduction & Objectives:****Materials & Methods:****Results:**

A healthy 21-year-old man was referred to our Dermatology clinic for an asymptomatic nasal cutaneous lesion with two years of evolution. Dermatological examination showed a 4cm erythematous plaque localized in the right side of the nasal pyramid, with irregular and well-defined borders, with multiple scattered opened comedones, telangiectasias and fine scale on the surface. A punch biopsy was performed, with histopathological examination showing atrophy and hyperkeratosis of the epidermis, basal hydropic degeneration, perivascular and periadnexal dense lymphocytic infiltrate and dilated follicular infundibulum, with follicular plugging and fibrosis. Given the clinical and histopathological findings, the patient was diagnosed with comedonic lupus. He did not meet clinical or laboratory criteria for systemic lupus erythematosus. Daily photoprotective measures were adopted and treatment with hydroxychloroquine 400mg/day and bethametasone dipropionate cream applied once a day was initiated, with significant improvement of the lesions at three months of follow-up.

Cutaneous lupus erythematosus (CLE) is an autoimmune disease subdivided into acute, subacute, and chronic forms according to its clinical manifestations. Chronic CLE predominantly presents as discoid lupus, but it may present with less common variants that can delay the diagnosis. Comedonic lupus (CL) is a rare and underdiagnosed variant of chronic CLE, that most commonly affects women between the third and fourth decades of life. Clinically, it is characterized by erythematous infiltrated papules and plaques, comedones, cysts, and acneiform scars in sun-exposed and seborrheic areas. Lesions can be asymptomatic or variably itchy. Differential diagnosis includes several benign dermatoses such as acne vulgaris, milium cysts, trichoepithelioma, nevus comedonicus, and Favre-Racouchot syndrome. The diagnosis is confirmed by skin biopsy, with histological findings similar to those seen in discoid lupus. Hyperkeratosis, epidermal thinning, vacuolar degeneration of the basal layer and periadnexial mononuclear infiltrate are present, with

dilated follicular ostia, prominent follicular plugs and comedones. Treatment of this form of CLE can be challenging, with photoprotection being essential as recommended in the other variants. Hydroxychloroquine is considered the first line of treatment, supplemented with topical or intralesional corticosteroids. Oral and topical retinoids may be used as an alternative.

By presenting this case, the authors raise awareness for this unusual and understudied variant of CLE. A delayed diagnosis and inappropriate treatment may have a negative impact on the quality of life, since comedonic lupus has the potential for significant disfigurement.

Conclusion:

Abstract N°: 4576

Retrospective Observational Study of Mortality Risk Associated with Oral Corticosteroid Use Among Patients Diagnosed with Bullous Pemphigoid

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Introduction & Objectives: Bullous pemphigoid (BP) is an autoantibody-driven blistering skin disease. Despite known toxicities, oral corticosteroids (OCS) generally remain first-line therapy for BP patients requiring systemic immunosuppression. The purpose of this study is to quantify the long-term mortality risk associated with early OCS exposure using US-based claims data.

Materials & Methods: This retrospective analysis included adults diagnosed with BP by a dermatologist in the Optum® Clinformatics® database from 1/1/2016 to 6/30/2022 with at least 1-year of continuous enrollment prior to the first BP claim (index date). OCS exposure was classified based on initiation within 90 days of the index date. Patients were followed from the index date to the earliest of death, end of insurance enrollment, or end of study. Baseline patient characteristics were summarized and unadjusted Kaplan-Meier (KM) methods were used to estimate overall survival. Crude incidence rate ratio (IRR) between OCS groups was reported. An extended Cox model was fitted to adjust for fixed baseline covariates (age, gender, payer, Charlson Comorbidity index [CCI], baseline OCS use), and time-varying concomitant medication use (intravenous immunoglobulin, immunosuppressive therapy, plasmapheresis, dapsons, doxycycline, topical corticosteroids). Hazard ratio was reported for mortality risk comparing the OCS-exposed and non-OCS exposed groups. Two sensitivity analyses were conducted: 1) initial OCS exposure by daily prednisone equivalent categorized as none, low (<15 mg), medium (15-30 mg), and high (>30 mg); and 2) time-varying assignment of OCS exposure at the time of initiation.

Results: Among 2,164 adult BP patients identified, 983 (45%) started OCS within 90 days of the index date (OCS exposed). Mean age was 79 years (SD 11) at index, 52% were female, mean follow up time was 1.62 years (SD 1.27), and mean CCI score was 3 (SD 3). Baseline clinical characteristics were similar between exposure groups (Table 1). Unadjusted KM analysis showed survival difference between the groups ($p=0.005$). Two-year survival rate was 71% (95% CI: 68-75%) for OCS-exposed group and 77% (95% CI: 75-80%) for the non-OCS exposed patients (Figure 1). Crude IRR of mortality between OCS-exposed and non-exposed groups was 1.29 (95% CI: 1.08-1.54). The adjusted Cox model identified a significantly increased hazard of mortality among OCS-exposed patients compared to non-OCS exposed patients, with an adjusted HR of 1.35 (95% CI: 1.10-1.66). Sensitivity analyses demonstrated significantly increased mortality with medium- and high-dose OCS exposure (Table 2). Modeling incorporating time-varying OCS exposure showed findings similar to the primary analysis (HR=1.37, 95% CI: 1.10-1.69).

Conclusion: Medium to high dose OCS use during early diagnosis was associated with a significantly increased risk of mortality in BP in this large US claims data study. This finding demonstrates the unmet medical need for safer, targeted treatment options for BP patients.

Table 1. Patient Demographics and Baseline Characteristics*

	Overall (N = 2,164)	OCS Use (N = 983)	No OCS Use (N = 1,181)
Age at Diagnosis, mean (SD)	77 (11)	77 (11)	77(11)
Female, n (%)	1,133 (52%)	527 (54%)	606 (51%)
Follow-up Time (Years), mean (SD)	1.6 (1.3)	1.6 (1.3)	1.7 (1.3)
Health Insurance Type, n (%)			
Commercial	294 (14%)	133 (14%)	161 (14%)
Medicare	1,870 (86%)	850 (86%)	1,020 (86%)
US Region, n (%)			
North Central	624 (29%)	264 (27%)	360 (30%)
Northeast	409 (19%)	187 (19%)	222 (19%)
South	734 (34%)	333 (34%)	401 (34%)
Unknown	6 (0.28%)	2 (0.20%)	4 (0.34%)
West	391 (18%)	197 (20%)	194 (16%)
CCI, mean (SD)	3 (3.0)	3 (3.1)	2 (3.0)
Comorbidities, n (%)			
Myocardial Infarction	165 (7.6%)	79 (8.0%)	86 (7.3%)
Congestive Heart Failure	424 (20%)	195 (20%)	229 (19%)
Peripheral Vascular Disease	617 (29%)	303 (31%)	314 (27%)
Cerebrovascular Disease	398 (18%)	180 (18%)	218 (18%)
Dementia	296 (14%)	158 (16%)	138 (12%)
Chronic Pulmonary Disease	552 (26%)	283 (29%)	269 (23%)
Rheumatic Disease	106 (4.9%)	58 (5.9%)	48 (4.1%)
Peptic Ulcer Disease	42 (1.9%)	25 (2.5%)	17 (1.4%)
Mild Liver Disease	111 (5.1%)	53 (5.4%)	58 (4.9%)
Diabetes without Chronic Complications	702 (32%)	336 (34%)	366 (31%)
Diabetes with Chronic Complications	428 (20%)	209 (21%)	219 (19%)
Hemiplegia or Paraplegia	70 (3.2%)	35 (3.6%)	35 (3.0%)
Renal Disease	534 (25%)	256 (26%)	278 (24%)
Any Malignancy	330 (15%)	140 (14%)	190 (16%)
Moderate or Severe Liver Disease	10 (0.46%)	3 (0.31%)	7 (0.59%)
Metastatic Solid Tumor	80 (3.7%)	39 (4.0%)	41 (3.5%)
AIDS/HIV	3 (0.14%)	1 (0.10%)	2 (0.17%)

*Baseline comorbidities reported in the 1 year prior to index date. Baseline OCS patients were defined as patients who started OCS within 90 days of index date, and non-OCS patients were defined as patients not receiving OCS OR starting OCS after 90 days.

†Comorbidities included as part of the CCI based on ICD codes.

AIDS: acquired immunodeficiency syndrome; CCI, [Charlson Comorbidity Index](#); HIV, human immunodeficiency virus; ICD, International Classification of Diseases; OCS: oral corticosteroid; SD, standard deviation; US: United States.

Figure 1.

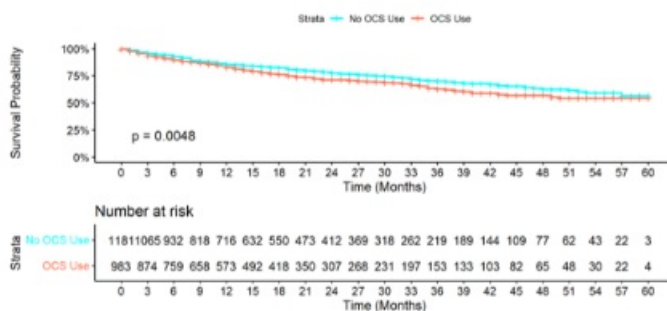


Figure 1. Unadjusted Kaplan-Meier analysis of survival probability between OCS exposure groups.

OCS: oral corticosteroid (budesonide, cortisone, dexamethasone, hydrocortisone, methylprednisolone, prednisolone, prednisone, clobetasol). BP: bullous pemphigoid.

Table 2. Sensitivity Analyses on the long-term mortality risk associated with initial OCS exposure dose in BP patients. Extended Cox proportional hazards model adjusted for fixed baseline covariates, including age, gender, payer, [Charlson Comorbidity index](#), baseline use of corticosteroids, and time-varying concomitant medication use such as intravenous immunoglobulin, immunosuppressive therapy (cyclosporine, tacrolimus, methotrexate, mycophenolate, azathioprine, cyclophosphamide, rituximab), plasmapheresis, dapsone, doxycycline, and/or topical steroids during follow-up.

Initial OCS Daily Dose	HR	95% CI
None	REF	REF
Low	1.06	0.75, 1.49
Medium	1.34	1.03, 1.73
High	1.58	1.22, 2.05

BP: bullous pemphigoid, HR: hazard ratio, CI: confidence interval, OCS: oral corticosteroid (budesonide, cortisone, dexamethasone, hydrocortisone, methylprednisolone, prednisolone, prednisone, clobetasol), REF: reference group

Abstract N°: 4604**Parry-Romberg Syndrome associated with ipsilateral segmental vitiligo**Omayma Khadiri¹, Maryem Aboudourib¹, Ouafa Hocar¹, Said Amal¹¹University hospital of Marrakech, Department of Dermatology venereology, Marrakech

Introduction & Objectives: The Parry-Romberg Syndrome (PRS) is characterized by hemifacial atrophy which can be associated with neurological, ophthalmological but also orthodontic signs which can jeopardize the functional and vital prognosis. The interindividual variability of this syndrome makes it all its complexity. The etiology is unknown. We report the observation of a young woman with an SPR characterized by the association of facial hemi-atrophy and ipsilateral segmental vitiligo.

Observation : A 16-year-old girl, with no significant history or notion of consanguinity, consulted for facial asymmetry that had been evolving for 18 months. Parents reported the first appearance of skin depigmentation, involving the left upper limb, at age 6. The lesions progressed, eventually also affecting the skin on the left side of the neck. The lesions stabilized in 1 year under tacromilus. At the age of 14, the patient observed progressive atrophy of the left cheek. The clinical examination revealed a visible left facial hemiatrophy including the skin and the subcutaneous tissue without cutaneous induration opposite, and without dental impact. The amelanotic lesions had a left cervical dermatomal distribution. The ophthalmological and oral examinations were without anomalies. Complete blood count, sedimentation rate, C-reactive protein, anti-TPO, anti-TG, antinuclear and anticardiolipin antibodies were all negative. Cerebral magnetic resonance imaging revealed no focal lesions or signs of neurovascular incident. Systemic treatment with a combination of corticosteroids at a dose of 0.5 mg/kg/day and methotrexate at a dose of 12.5 mg/week was initiated.

Discussion: The peculiarity of this observation is the association of hemifacial atrophy and ipsilateral segmental vitiligo, which is rather rare in Parry-Romberg syndrome (PRS). In the literature, three cases of ipsilateral segmental vitiligo associated with atrophy in PRS have been described, as well as one case of PRS associated with anti-native DNA antibodies (1-3). This association supports the autoimmune origin of PRS. The autoimmune hypothesis is further supported by the existing link between PRS and en coup de sabre scleroderma. The pathogenesis of segmental vitiligo is not clear, and various hypotheses have been proposed, including dysfunction of the sympathetic nerve and an immune-related mechanism. It should be noted that segmental vitiligo is rarely associated with various autoimmune diseases, as is the case with non-segmental vitiligo (4). Cases of simultaneous coexistence of progressive hemifacial atrophy with ipsilateral segmental vitiligo are characterized by onset in children and young adults, rapid progression followed by a long period of subsequent stabilization, and a dermatomal pattern.

Conclusion: Our case is one of the rare reported examples of the coexistence of these two pathological entities. Further studies are needed to explain the coexistence of the common pathomechanism in PRS with ipsilateral vitiligo.

Abstract N°: 4642**Can Methotrexate Be Employed as Monotherapy for Bullous Pemphigoid? Analysis of Efficiency And Tolerance of Methotrexate Treatment in Patients with Bullous Pemphigoid**Agnieszka Zebrowska¹, Anna Wozniacka¹, Magdalena Wojtczak¹¹Medical University of Lodz, Department of Dermatology and Venereology, , Lodz, Poland**Introduction & Objectives:**

BP is a common autoimmune blistering disease. While it is most frequently observed in elderly patients, it may appear at any age. European Academy of Dermatology and Venerology consensus states that the treatment of choice for bullous pemphigoid is systemic glucocorticosteroid therapy. Bearing in mind that long-term steroid therapy is associated with numerous side effects, an effective and safer treatment regimen for these patients is still being sought. Considering the side effects of steroid therapy and the risk of mortality in older patients during long-term steroid therapy, methotrexate (MTX) seems to be a good alternative treatment in elderly patients with BP. The main aim of this study was to analyze the efficiency of MTX treatment and tolerance to it. It compares the effects of MTX monotherapy with a combined regimen of MTX and systemic steroid therapy (GKS).

Materials & Methods:

A retrospective analysis was performed of the medical reports of patients with diagnosed bullous pemphigoid. The study included 40 patients with moderate or severe disease, and who had continued ambulatory treatment for at least six months. The patients were divided into two groups: one treated with methotrexate in monotherapy, or with combined methotrexate and systemic steroid therapy.

Experimental data was collected in MS Excel. The mean value and standard deviation of the studied population were calculated. Qualitative data was analyzed using the χ^2 test, and quantitative data using Student's t-test. Additionally, Pearson's correlation coefficient (r) was determined. Differences were regarded as significant at a level of $p < 0.05$. All analyses were performed using Statistica 13.1 software.

Results:

A slightly better survival rate was noted in the methotrexate group. No significant differences were observed between the groups in time to achieve clinical remission. The combination therapy group (GKS +MTX) demonstrated more frequent disease recurrence and exacerbations during treatment, and higher mortality rate. None of the patients in either group presented with severe side effects related to methotrexate treatment. Our findings indicate that patients with BP should be treated with MTX in monotherapy. In addition, the dermatologist should focus on reducing systemic steroid use during treatment to reduce the risk of side effects related to steroids and mortality rate.

Conclusion:

Treatment of bullous pemphigoid with methotrexate in monotherapy is an effective and safe therapeutic method for elderly patients.

Abstract N°: 4691**Pemphigus vulgaris with scalp onset**

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Introduction & Objectives:

Although the scalp is usually affected in pemphigus vulgaris, hair loss is rarely seen. We present a case of a patient that consulted with an initial form of presentation on the scalp, showing hair traction positivity as an important marker of disease activity.

Materials & Methods:

Case report.

Results:

A 40-year-old male patient consulted for itchy scalp lesions of six weeks' evolution. He reported that the first lesion appeared in the frontal region and then lesions began to appear in the vertex and temporal region.

On physical examination, he presented multiple adherent scabs and wart-like plaques that fell off when pulled, along with anagen hairs, leaving an erosive base. Given the clinical suspicion of pyodermitis vs. false tinea vs. pemphigus vulgaris, a biopsy was taken from one of the lesions and treatment with mupirocin cream associated with minocycline 100 mg/day was indicated.

The pathology revealed erosion and marked acantholytic changes that left a single basal layer in sectors, with extension to pilosebaceous adnexa, accompanied by inflammatory lymphocytic infiltrates, some eosinophils, plasma cells, and erythrocyte extravasation. Given the histopathological images linkable to pemphigus vulgaris, a new biopsy was performed for direct immunofluorescence of the perilesional skin of one of the scalp erosions.

Two weeks later, small isolated erosions appeared in the right axilla and upper trunk, a small blister in the right scapular region, and erosions in the oral mucosa. It also presented positive traction of anagen hairs in affected regions of the scalp, being a marker of disease activity.

Direct immunofluorescence revealed positive IgG with a "chicken wire" epidermal pattern, positive C3 focal intercellular at the level of the epidermal basal layer, and positive IgG at the level of the hair follicle, consistent with pemphigus.

With the definitive diagnosis of pemphigus vulgaris, treatment with meprednisone 60 mg/day was indicated, with which he presented an excellent response with disappearance of the cutaneous-mucosal lesions and negative scalp hair traction. Currently, the dose of meprednisone is being reduced in order to add mycophenolate mofetil as a corticosteroid-sparing agent.

Conclusion:

In our case, the patient debuted with an initial form of presentation on the scalp, later adding skin lesions and oral mucosa. Hair traction is important as a marker of disease activity. We highlight the importance of clinical suspicion and the value of pathological anatomy and direct immunofluorescence to confirm the entity, in order to install treatment as early as possible.

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Abstract N°: 4725

Localized bullous pemphigoid developing twice after orthopedic surgery

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Introduction & Objectives:

Bullous pemphigoid (BP) most commonly presents as widespread, itchy, tense blisters in older patients. Localized bullous pemphigoid is a less common form of BP that can be more difficult to diagnose. We present the case of a 60 year old man who presented with pruritic tense bullae twice after orthopedic surgery. This case illustrates the potential for localized BP to be triggered by surgical procedures.

Materials & Methods:

A 60-year-old male with a history of right femur fracture for which he received retrograde nailing, presented with a pruritic bullous eruption in the right knee and leg, 15 days after surgery. The skin biopsy showed bulla with eosinophilic infiltrates and linear immunoglobulin IgG, and C3 deposition in the basement membrane with direct immunofluorescence. The diagnosis of a localized form of bullous pemphigoid was made.

Prednisone 0.5 mg/kg was started daily and the lesions disappeared by 3 months. Unfortunately, the fracture did not heal during this period and the patient underwent a bone graft. Few days after surgery, the patient developed new tense blisters around the surgical site, right thigh and knee. Prednisone was again introduced and the lesions resolved 2 months later.

Results:

Localized BP has previously been described in the context of radiation, phototherapy and burns. Development of this entity after surgery has rarely been reported in the literature. It is thought that tissue damage interferes with the humoral mediated immunity, causing alteration of BP180 and BP230 antigens.

The fact that localized BP occurred twice suggests *undoubtedly* a link with surgery. To our knowledge, this is the second case in which such a phenomenon re-occurred after 2 surgical procedures in the same patient.

Proposed mechanisms of localized BP in this context include koebnerization. But in our case, we think that the trigger is the localized edema, a likely consequence of the orthopedic surgeries, which can explain the pathogenesis of this phenomenon since the blisters extended beyond the surgical sites.

Conclusion:

Prompt recognition of localized BP is important so that appropriate treatment can be started.

Abstract N°: 4735

Scleroderma in coup de sabre, aesthetic improvement on the scalp with local bimatoprost 7 years follow up, case report

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Introduction & Objectives:

Linear sclerosis *en coup de sabre* is a rare autoimmune disease that involves focal atrophy and sclerosis of the head and face and affects predominantly children and women.

The early lesion appears as an erythematous or violaceous linear indurated atrophic plaque and subsequently lesions progress to hypopigmented sclerotic deep furrows where alopecia appears.

Bimatoprost, a synthetic prostamide F2 α analog originally approved for the treatment of ocular hypertension and open-angle glaucoma, is now FDA approved as a 0.03% solution to be applied once daily to increase eyelash growth. There have been occasional reports on its use on scalp alopecia.

Results:

We report an important aesthetic improvement on a severe case of local scleroderma with 7-year follow-up with bimatoprost 0.03%.

29-year-old woman with no history of previous diseases consulted due to a linear atrophic lesion involving the left scalp, forehead, eyebrow, and orbita. She was evaluated for systemic disease with no relevant data. She was treated with 3 infiltrations of triamcinolone locally with improvement on the furrow atrophy but no significant response over the hair of the scalp or eyebrow.

She had an important post-inflammatory hyperpigmentation treated with local hydroquinone 4%.

She followed a daily regimen of local bimatoprost with significant response after 4 months with newly hair growing on the previously atrophic area and a twice-a-week regimen over the past 6 years maintaining response.

Conclusion:

Bimatoprost 0.03% on a daily basis regimen might be considered as an option for scalp hair regrowth.



Abstract N°: 4738**Sinusitis with perforation of the nasal septum and strawberry gingivitis with SARS-COV2 infection as the first manifestation of granulomatosis with polyangiitis. A diagnostic challenge.**

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Introduction & Objectives:

Strawberry gingivitis is a rare manifestation of granulomatosis with polyangiitis, but its clinical presentation is highly suggestive of the disease. Therefore, recognizing specific signs, such as strawberry gingivitis, is crucial for timely diagnosis and definitive management of this life-threatening disease.

Materials & Methods:

We present the case of a 31-year-old female patient with no relevant history who came to the emergency room with severe headache, fever, cough, runny nose, epistaxis, and symmetrical polyarthralgia of three months' evolution. She had been treated previously for recurrent upper respiratory tract infections, the first of which was a confirmed diagnosis of mild SARS-COV2. When she did not show improvement, despite multiple antibiotics and symptomatic treatments, she went to a specialist otolaryngologist, who requested a computed tomography (CT) scan showing a tumor in the nasal septum. They took a biopsy of the tumor, which only ruled out malignancy without being conclusive. Meanwhile, the patient persisted with the symptoms, worsening the headache, accompanied by fever, asthenia, adynamia, facial edema, epistaxis, and rhinorrhea with abundant purulent discharge and two skin ulcers.

Our dermatology service¹ was consulted for presenting these two ulcers in the anterior and posterior pelvis, resembling pyoderma gangrenosum, of 1 cm in diameter each. Physical examination also revealed intensely painful strawberry gingivitis. Complementary studies revealed high titers of anti-neutrophil cytoplasmic antibodies with a cytoplasmic staining pattern (c-ANCA), and enzyme-linked immunosorbent assay for anti-proteinase-3 antibodies was positive. Serum creatinine level and urinalysis were average. A head CT scan revealed a tumor and perforation of the nasal septum. With the above, we established the diagnosis of granulomatosis with polyangiitis (GPA).

Results / Conclusion:

GPA and its complications can only be prevented with early diagnosis and treatment, which requires a high suspicion of the disease. Autoimmune diseases in patients with COVID-19, even after recovery from the infection, represent an even more significant diagnostic challenge due to the coincident respiratory symptoms. Infections can be the initial manifestation of GPA. The persistence of symptoms, without improvement, should make us consider differential diagnoses. The dermatological approach to GPA lies in the importance of identifying dermatoses and integrating them to achieve a timely, multidisciplinary diagnosis and treatment that avoids fatal complications, influencing a better prognosis for the patient.

Abstract N°: 4743**Clinical Feature of Dermatomyositis with Anti-SRP Antibody and Anti-Ro52 Antibody**Daisuke Suzuki^{*1}, Chika Ohba¹, Yuta Norimatsu¹, Makoto Sugaya¹¹International University of Health and Welfare, Dermatology, Chiba, Japan

The autoimmune inflammatory myopathies are a heterogeneous group of diseases with variable clinical presentation and features; muscle inflammation, skin rashes and systemic complications including interstitial lung disease (ILD), cardiac involvement and malignancy. They are classified into dermatomyositis (DM), polymyositis (PM), inclusion body myositis and necrotizing autoimmune myositis. Accumulated studies revealed various autoantibodies associated with myositis. A positive result of myositis-specific antibodies will usefully provide diagnostic and prognostic information guide selection of therapy, and prompt surveillance for potential organ involvement and other features, such as cancer, throughout the disease course. We herein report a case of DM with rare antibodies; anti-SRP antibody and anti-Ro52 antibody.

A 77-year-old male patient presented with faint erythema over the knuckles. He had suffered from ILD for two years and also was aware of fatigue in thighs on exertion. Serological examination showed slight elevation of creatinine kinase (CK: 523 U/L (normal 59-248) and Krebs von den Lungen-6 (KL-6: 568 U/mL (normal 0-500)), and he was suspected to have a DM, though autoantibodies against ARS, MDA-5, TIF1- γ , Mi-2 were not detected. His eruptions were too faint to make a diagnosis of DM at first, however his skin manifestations became apparent in winter; eruptions over the knuckles became more reddish and indurated, and telangiectatic vessels in the proximal nailfolds became prominent, while ILD and muscle inflammation including KL-6 and CK value were followed without significant change. Skin biopsy was performed from the dorsal hand, and mild vacuolar degeneration of dermo-epidermal junction and slight mucinous depositions were revealed histologically. Based on the findings described above, he was diagnosed with DM.

In order to confirm the diagnosis, comprehensive examination for myositis-specific antibodies was performed and anti-SRP antibody and anti-Ro52 antibody were detected. Anti-SRP antibody is usually detected in PM patient and known to be associated with severe muscle necrosis, however the patient was diagnosed with DM and his muscle inflammation was mild. Only a few DM patients with anti-SRP antibody had been reported so far; skin manifestations varied and ranged from erythema or papules on fingers to digital necrosis, but muscle inflammation was mild in almost all cases. Although PM patients with anti-SRP antibody tend to have severe necrotizing myopathy, this tendency may not be applicable to the DM patients with anti-SRP antibody. Ro52 is part of the SS-A/Ro protein, and anti-Ro52 antibody is commonly detected in patients with systemic lupus erythematosus and Sjögren's syndrome. In autoimmune inflammatory myopathies, anti-Ro52 antibody have been reported the association with ILD severity, however ILD of the patient was mild and non-progressive.

As described above, this DM patient with rare autoantibodies had atypical clinical findings in ILD and myopathy that were different from the symptoms expected from these autoantibodies. We feel pleased if we contribute the investigation of the clinical feature of the rare variant of DM.



Abstract N°: 4748

Clinicopathologic Characteristics and Health-Related Quality of Life in Patients with Autoimmune Blistering Diseases in a Tertiary Hospital: A Cross-Sectional Study from 2017 to 2022

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¹Rizal Medical Center, Pasig, Philippines

Introduction & Objectives:

Autoimmune blistering diseases (AIBDs) are a group of diverse disorders characterized by blister formation due to pathogenic autoantibodies directed against structural proteins of the skin and mucous membranes. Even though the disease affects the elderly in most cases, AIBDs may spontaneously arise and may be observed in children, and young adults especially if there is a genetic predisposition.

Our general objective is to describe the sociodemographic characteristics, clinicopathologic features, and health-related quality of life of Filipinos with autoimmune blistering diseases at a tertiary hospital from 2017 to 2022.

Materials & Methods:

This was a retrospective analytical cross-sectional study conducted through a review of records of patients seen at a tertiary hospital from 2017-2022.

Descriptive statistics was used to summarize the general and clinical characteristics of the subjects. Frequency and proportion were used for categorical variables. Shapiro-Wilk test was used to determine the normality distribution of continuous variables. Continuous quantitative data that met the normality assumption was summarized using mean and standard deviation (SD), while those that do not were described using median and range. Pearson's product-moment correlation or Spearman's rank correlation was used also used. Null hypothesis was rejected at 0.05 α -level of significance. STATA 15.0 was used for data analysis.

Results:

A total of 92 patients diagnosed with Autoimmune blistering disorders were seen at a tertiary hospital, Department of Dermatology between the years 2017 to August 2022. However, this study included only 53 patients based on the study's inclusion criteria.

The demographic, associated comorbidities and treatment profile of patients with bullous pemphigoid (BP), chronic bullous disease of childhood/linear IgA bullous disease (CBDC/LABD) epidermolysis bullosa acquisita (EBA) pemphigus foliaceus (PF), pemphigus vulgaris, pemphigus vegetans are summarized in this study, including the Sociodemographic characteristics, associated comorbidities, and treatment profile by AIBD type (N=53), Immunologic findings per diagnosis, Disease severity and HRQOL before and after treatment, and Correlation of disease severity scores with quality of life measures (ABQOL and TABQOL).

Conclusion:

Trends among the Filipino population are in line with population trends in other regions of the world.

Bullous pemphigoid, pemphigus vulgaris, and pemphigus foliaceus are the most common autoimmune blistering diseases within our population, with most patients older than 40 years of age. This study demonstrated that ABQOL was moderately correlated with disease severity indices, although different components of these indices

showed varied responses to HRQOL indices. TABQOL was not very sensitive to treatment and did not correlate with disease severity indices. Based on these findings, we would recommend specifying at which disease phase (induction/remission), ABQOL/TABQOL scores were obtained.

The majority of our cases demonstrated consistency between clinical, histopathological, and immunofluorescence findings, highlighting the value of appropriate investigations in vesiculobullous disorders to reach a conclusive diagnosis. Financial constraints need not hinder diagnosis – an important factor to consider in lower-to-middle income countries.

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Abstract N°: 4754**An atypical case of bullous pemphigoid in an 11-year-old Filipino female**Bernadette Iglesia*¹, Mae Quizon¹¹Rizal Medical Center, Dermatology, Pasig, Philippines**An atypical case of bullous pemphigoid in an 11-year-old Filipino female****Introduction & Objectives:**

Bullous pemphigoid is the most common autoimmune blistering disease in the elderly but is rare in children. Bullous pemphigoid in adolescence is much more uncommon since pediatric bullous pemphigoid usually occur during early childhood. This is also supported by local data, wherein only 66 cases of bullous pemphigoid in childhood were recorded in the Philippines from 2011 to 2022. We present an atypical case of bullous pemphigoid in childhood that posed a challenge in the diagnosis of this autoimmune bullous disorder in the pediatric population.

Materials & Methods: N/A**Results:**

We present an atypical case of bullous pemphigoid in childhood that posed a diagnostic dilemma in the pediatric population. Clinically, there was extensive acral involvement, as well as involvement of the vermilion border of the lips and the genitalia. There were no mucosal lesions. Dermoscopy findings showed yellowish-pink translucent areas and erythematous, distorted pigment network, with prominent follicular openings.

Due to the age of the patient and the clinical presentation resembling a “cluster of jewels”, the initial assessment was chronic bullous disease of childhood (CBDC). Histopathology showed a subepidermal split, with karyorrhexis and band-like infiltrates heavily composed of neutrophils, in contrast to the typical findings of eosinophilic infiltrates in bullous pemphigoid. Direct immunofluorescence showed a linear pattern of immunoglobulin G and C3 deposition along the epidermal–dermal junction, which narrowed down the differentials to bullous pemphigoid, epidermolysis bullosa acquisita (EBA) and bullous systemic lupus erythematosus (BSLE), all of which are rare in the pediatric population. Serological tests for BP180, BP230, and type VII collagen were done to differentiate bullous pemphigoid from other subepidermal bullous diseases. The patient was negative to BP180 but reactive to BP230, which is usually the case in atypical presentations of bullous pemphigoid, with no mucosal involvement and with milder symptoms. The set of clinical and laboratory data confirmed the diagnosis of childhood bullous pemphigoid.

A combination of topical steroids, systemic steroids (prednisone, 1mg/kg/day) and dapsone (2mg/kg/day) were given, with excellent response to dapsone due to the histopathology of neutrophilic-predominant bullous pemphigoid. Relapses were noted at decreased doses of prednisone (0.8mg/kg/day) and when off-medications. On the 7th month of treatment, the patient remained lesion-free. Complete remission is expected by the 9th month of treatment.

Conclusion:

Bullous pemphigoid, despite being rare in children, should still be part of the differential diagnoses in children presenting with blistering conditions. Childhood variants of bullous pemphigoid can present with atypical clinical, histopathological and serological features, hence, the importance of complete history and work-up. As compared

to adults, bullous pemphigoid in childhood has a favorable course but needs long-term follow-up before complete remission is achieved.

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Abstract N°: 4783**Cutaneous microvascular occlusion syndrome as the first manifestation of catastrophic Lupus-associated Antiphospholipid Antibody syndrome: A rare presentation**

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Cutaneous microvascular occlusion syndrome as the first manifestation of catastrophic Lupus-associated Antiphospholipid Antibody syndrome: A rare presentation**Introduction & Objectives:**

Background: Antiphospholipid syndrome (APS), defined by thrombotic events or obstetric complications in the presence of persistently high antiphospholipid antibodies, is characterized by a wide variety of clinical presentations and the effects of vascular occlusion can impact almost any organ system or tissue. Since adult-onset APS classification criteria are not well verified in pediatrics (where pregnancy-related problems are rare), estimating childhood prevalence is challenging. Stroke and pulmonary embolism are thromboembolic events occurring in children that can cause considerable long-term morbidity. Children with APS are more prone to recurrent thromboembolism than adults. Cutaneous symptoms are prominent and typically represent the first clue of APS. Although dermatologic findings are exceedingly heterogeneous, it is essential to consider which dermatological symptoms justify the investigation of antiphospholipid syndrome and the required further management.

Materials & Methods:

We describe a seven-year-old boy with retiform purpura and acral cutaneous ischemic lesions as the first clinical presentation of antiphospholipid syndrome in the setting of systemic lupus erythematosus.

Results:

-

Conclusion:

APS in pediatrics, is associated with a variety of neurologic, dermatologic, and hematologic symptoms. Therefore, it is essential for paediatricians to be aware of the rare appearance of Catastrophic APS as an initial indication of APS.

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Abstract N°: 4802**Chilblain Lupus Erythematosus as initial presentation of systemic lupus erythematosus.**

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Introduction:

Chilblain Lupus Erythematosus (CLE) is a rare subtype of chronic cutaneous lupus erythematosus (CCLE), which is sometimes related to systemic lupus erythematosus (SLE). It has been reported that up to 25% of patients with chilblain meet the criteria for SLE at the time of diagnosis or later. CLE presents with clinical findings of chilblain in the context of a patient diagnosed with cutaneous lupus or systemic lupus erythematosus (SLE). The lesions are located bilaterally in distal regions such as ears, nose, fingers, and toes. It is characterized by the presence of erythematous-violaceous plaques and nodules, ulcerations, and edema, and the lesions are usually painful or pruritic. It appears as a consequence of intermittent or prolonged exposure to cold. There are few cases of CLE published in the literature. The case of a male patient with an initial presentation of chilblain is reported, who met the criteria for SLE during his hospitalization, making the diagnosis of CLE.

Clinical case:

A 48-year-old man with no significant pathological history was hospitalized due to 4-month history of generalized arthralgia and myalgia, pancytopenia, and a dermatosis, which began in the fingers of both hands and spreading to the palms and toes, characterized by erythematous-violaceous plaques, ulcers and hematic crusts, and painful hyperkeratosis, in addition to presenting onychodystrophy, onychorrhexis, and subungual hemorrhages. Subsequently, a second dermatosis located in sun-exposed areas of the neck, anterior thorax, and ears characterized by erythematous-violaceous, indurated, scaly, atrophic, irregular plaques was added to the picture. Due to the clinical suspicion of CCLE, laboratory tests were requested, and reported anemia with hemoglobin of 6.8 mg/dl, and creatinine of 0.7 mg/dl, in addition to positive antibodies for Anti-nRNP 24 U/ml, Anti-Sm 7 U/ml, Anti-SS-A 8 U/ml, and Anti-dsDNA 50 U/ml, Anti-Nucleosomes 62 U/ml, Anti-Histones 15 U/ml, positive antinuclear antibodies with homogeneous pattern 1: 5,120, and cytoplasmic 1: 640.

Two skin biopsies were performed, and histopathological diagnosis corresponded to interface dermatitis with leukocytoclastic vasculitis compatible with SLE.

With the above, the diagnoses of CCLE and SLE were integrated according to the ACR/EULAR 2019 criteria. Treatment with prednisone and hydroxychloroquine was started with immediate clinical improvement.

Discussion:

The clinical diagnosis is based on the criteria created by the Mayo Clinic after studying a small group of people with CLE, requiring two major criteria and one minor criterion.

Clinically it presents as erythematous or violaceous papules or plaques in acral areas. The fingers and toes are the most affected sites, with ulceration and necrosis in some cases; less frequently, it affects palms and soles, presenting with hyperkeratosis and fissures; ulcerations on the toes can develop scars. Other sites include the ears, nose, and, very rarely, the trunk. Itching may be an initial symptom, later accompanied by pain.

Conclusion:

There is not much literature on chilblain lupus erythematosus, hence the objective of raising awareness of this disorder, both its clinical presentation and the possibility of underlying SLE and a better prognosis if recognized early, responding favorably to classic SLE treatment, as well as how to contribute to future research.

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Abstract N°: 4811**One-Year Observational Experience From A Combined Dermatology-Rheumatology Clinic**

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Introduction & Objectives:

Immune-mediated inflammatory disorders are associated with increased morbidity and mortality, decreased quality of life, increased risk of associated comorbidities, and reduced socioeconomic status. There is a need for increased access to care for such patients. Combined dermatology and rheumatology clinics offer one-stop experience for patients; increase access to care; streamline comprehensive treatment plans; provides high-quality experience for trainees, specialists, and patients, and improve patient outcomes.

In this observational cohort study, we examine the diagnostic accuracy in all patients seen at the University of New Mexico (UNM) combined rheumatology and dermatology clinic over the course of one year.

Patients seen in this clinic have systemic autoimmune or inflammatory disorders with cutaneous manifestations, and notably an alternate diagnosis is made in about 25% of patients seen.

Materials & Methods:

One-year patient data from the UNM combined rheumatology-dermatology clinic was added to a database to gather information on types of diagnoses seen as well as alternative diagnoses made. Patients were seen as referrals from separate dermatology or rheumatology clinics at UNM or primary care physicians. Diagnoses were made or confirmed by the two sub-specialists of the clinic.

Results:

Patients were divided into 8 groups separated by diagnoses (n=61 patients): sclerosing disorders (n= 11), psoriasis with arthritis (n=13), lupus (n= 12), vasculitis (n=2), idiopathic inflammatory myopathies (IIM; n= 2), mixed connective tissue disease (MCTD; n=1), undifferentiated connective tissue disease (UCTD; n=3), and miscellaneous disorders (n=17). Alternative diagnoses were made in 15/61 patients (24.6%).

A change in diagnoses was observed in the psoriasis with arthritis group (n=2), the UCTD group (n=2), and miscellaneous disorders (n=11). In the psoriasis group, 2 patients suspected to have psoriatic arthritis were instead diagnosed with rheumatoid arthritis and ankylosing spondylitis. Two patients in the UCTD group were suspected of having SLE based on serologies but did not meet SLE criteria.** In the miscellaneous group 7 patients were referred for suspected lupus diagnosis, but did not meet SLE criteria and were diagnosed with other disorders (3 with TNF-inhibitor induced antibodies; 1 hypertrophic lichen planus; 1 chronic urticaria; 1 autoimmune progesterone dermatitis, and 1 contact dermatitis); 1 patient referred for suspected ANCA-associated vasculitis but found to have generalized livedoid vasculopathy; 1 patient with HLA-B27 positivity referred for psoriatic arthritis but found to have post-traumatic arthritis; one patient referred for UCTD but found to have oculopharyngeal muscular dystrophy, and 1 patient referred for dermatomyositis, has a myopathy not otherwise specified.

Conclusion:

The one-year analysis of this multidisciplinary clinic revealed that alternate diagnosis was made in nearly 25% of patients with complex medical diagnoses. In an analysis of the patients who had alternative diagnosis after being evaluated in our interdisciplinary clinic, the vast majority were patients referred for lupus (9/15), but none were found to meet SLE criteria, or have cutaneous lupus. We hypothesize that this second-opinion in real-time review also acts as a patient safety mechanism that helps reduce error and provides patients with evidence-based treatment for their precise clinical diagnoses.

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Abstract N°: 4835**Environmental Factors for the Development of Autoimmune Blistering Disease: A Case-control Study**Corey Stone^{*1, 2}, Grace Bak², Daniel Oh², Cathy Zhao², Supriya Venugopal², Dedee Murrell^{2, 3}¹Bond University, Robina, Australia, ²St George Hospital, Kogarah, Australia, ³UNSW Sydney, Sydney, Australia**Introduction & Objectives:**

Autoimmune blistering disease (AIBD) are chronic and potentially life-threatening skin conditions caused by autoantibodies targeting skin adhesion proteins. Although genetic predispositions have been identified, the impact of environmental factors initiating or exacerbating these diseases is incompletely understood. Various environmental factors have been suggested as potential triggers of immune dysregulation including food, medications, viruses, UV exposure, trauma, and emotional stress. To date there are no published case-control studies with age, gender, and location matched controls investigating the potential link between environmental exposures and the development of AIBD.

This case-control study aims to determine the environmental factors that contribute to the development of autoimmune blistering disease, particularly pemphigus vulgaris (PV) and BP (bullous pemphigoid).

Materials & Methods:

Patients with bullous pemphigoid and pemphigus vulgaris (diagnosed with serum antibodies, histology, and direct immunofluorescence) were interviewed using a questionnaire on prior environmental and occupational exposures between 2009 and 2017. Controls matched for age, sex and area of residence, were sourced from the Australian Electoral Commission and undertook the same questionnaire. Data was analysed using SPSS software to compare exposure factors between cases and controls.

Results:

There was a total of 25 PV cases vs. 72 controls and 29 BP cases vs. 84 controls. Environmental factors significantly associated with PV included consumption of leeks (p .025), mustard oil (p .049), tomatoes (p .032), multivitamins (.009), and alcohol (p .039). Additionally, stress (p .002) and total lifetime sunburn (p <.001) were also associated with development of the disease. Environmental factors significantly associated with BP included consumption of celery (p .035), green/herbal tea (p .002), glucosamine (p .019), fish oil supplements (p <.001), and calcium supplements (p <.001). Stress (p .006) and the use of household de-greasants were also associated with disease onset.

Conclusion:

Our case-control study has identified several potentially causative environmental factors for the development of pemphigus vulgaris and bullous pemphigoid suggesting the need for further research in this area.



Abstract N°: 4848**Current approaches and management trends of localized scleroderma**Beatrice Balaceanu-Gurau*¹, Mara Mihai^{1, 2}, Adina Gheorghe², Alexandra Timofte¹, Calin Giurcaneanu^{1, 2}¹Elias University Emergency Hospital, Dermato-oncology, Bucharest, Romania, ²Carol Davila University of Medicine and Pharmacy, Bucharest, Romania**Introduction & Objectives:**

Morphea is a rare idiopathic autoimmune disease that mainly affects the dermis and subcutaneous tissue, but can also spread to the underlying muscles, joints and bones. At the cutaneous level, this can lead to single or multiple inflammatory or sclerotic plaques. Sclerosing scars can progress with significant induration of the integument, causing the development of disfiguring scars, with decreased joint mobility and functional impairments. There are also patients who develop more persistent or recurring involvement. Unlike systemic sclerosis, morphea does not cause sclerodactyly, nail fold capillaries, fibrosis or vascular damage to internal organs. Regarding morphea management, there are many therapeutic options, but the evidence supporting them is limited, with patients being followed-up and treated with topical products, phototherapy, or systemic immunosuppressive therapy.

The objectives of the study consisted in the analysis of the main risk factors, clinical manifestations, comorbidities, diagnostic tools and treatment methods, in order to bring up to date information on the pathogenesis and evolution of this condition and to optimize the early diagnosis of localized scleroderma.

Materials & Methods:

Although morphea is a well-known and defined pathology in current research studies, there is only a small number of observational studies published in recent years. We performed a longitudinal, retrospective, observational study on a group of 68 international patients, followed between January 2012 and October 2020. The aim of our study was to carry out a detailed analysis of hospitalized patients with localized scleroderma.

Results:

Morphea occurs most frequently in females, the average age diagnosis being 54.67 years. The average duration of this affliction turned out to be 3.76 years, underlining its chronic course. The prevalence of other associated autoimmune diseases is increased, and the coexistence of several morphea-independent comorbidities causes a poor monitoring of the dermatological condition, leading to the presentation of patients in very late stages of the disease. As other specialized studies prior confirmed, any type of radiation, especially radiotherapy, represent a risk factor for the appearance of morphea in 8.82% of patients included in our study.

Cutaneous lesions can be found in any anatomical region, but mainly in the thighs, chest and abdomen. Laboratory investigations do not have an increased specificity for the diagnosis of morphea. Histopathological examination is the most useful paraclinical investigation to confirm the clinical suspicion. The most common recommended therapies were topical ones (Vaseline and Tacrolimus), followed by systemic ones (Penicillin G and Corticosteroids) and phototherapy.

Conclusion:

In conclusion, the results of our study support the importance of a proper management of morphea, with an early and correct diagnosis to prevent complications, cosmetic and functional sequelae and also to increase patient's quality of life.

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Abstract N°: 4966**Evaluation of the efficacy and safety of azathioprine in the treatment of severe alopecia areata, a cohort of 10 years' follow-up of the patients**Susan Farshi^{*1}, Parvin Mansouri¹, Mojgan Saghazadeh²¹Tehran University of Medical Sciences, ²Azad University**Introduction & Objectives:**

Alopecia areata is an autoimmune disease and T cells may have a key role in the pathogenesis of the disease. Several immunosuppressive drugs have been used with different results. The aim of this study was to evaluate the efficacy and safety of long-term azathioprine use as a systemic monotherapy for moderate to severe alopecia areata.

Materials & Methods:

A total of 63 patients (27 females (42.9%) and 36 males (57.1%)) with minimum six months' history of alopecia areata were included. The extent of scalp hair re-growth during and yearly until the completion of the 10-years treatment evaluated by the Severity of Alopecia Tool (the SALT score) and the primary endpoint was the percent change in SALT score during treatment. The daily drug intake was calculated as 2mg/Kg of body weight.

Results:

Mean duration of current episode scalp hair loss was 34.10 (34.10±39.16) months. Mean re-growth percentage was 92.69 (92.69±9.08). Mean hair loss percentage was 74.2 (74.2±27.8) before the treatment compared to 5.2 (5.2±8.6) after 10years azathioprine treatment. This showed a highly significant statistical difference (Paired t test, CI 95%= 55.9-75.3).

Mean hair loss score (S0-S5) before treatment was 5.56(5.56±1.3) and after 10years azathioprine treatment was 0.67(0.67±0.53). Assessment showed significant difference from baseline score (Wilcoxon Signed Rank Test, P<0.0001).

Conclusion:

This study showed that azathioprine can be used as a safe and effective systemic drug in the treatment of recalcitrant and severe alopecia areata.



Abstract N°: 4974

Role of novel biomediators of BMP signalling in correspondence to NB-UVB phototherapy in vitiligo patients.

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Introduction & Objectives: Vitiligo is a chronic autoimmune disorder characterized by loss of functional melanocytes leading to depigmentation of skin. NB-UVB (Narrow band ultraviolet B therapy) is considered as one of the most effective and safest repigmentation therapy for vitiligo. However, there are few patients who do not respond to this NB-UVB therapy. BMP (Bone morphogenetic protein) signaling pathway is widely studied in NB-UVB therapy. Amongst various types of BMP molecules, such as BMP2, BMP4, BMP5, BMP6, BMP7; BMP4 and BMP6 are known to be involved in regulating the process of melanogenesis. BMP-6 promotes melanogenesis whereas BMP-4 has been shown to inhibit it. Hence, this study was designed to understand BMP signaling in vitiligo patients undergoing NB-UVB phototherapy and to achieve this, following objectives were designed:

\1) To check BMP-4 and melanin synthesis gene expression in patients undergoing

NB-UVB therapy.

\2) To check the expression of BMP antagonists i.e, Noggin, Gremlin, Sclerostin in these patients.

Materials & Methods: Non Segmental Vitiligo (NSV) patients between age (>18) to (<60) years were recruited. The duration of study is for 24 weeks. Consenting patients were treated with whole body NB-UVB phototherapy thrice a week with initial dose of 25mJ/cm² and increased by 20% at each visit upto maximum of 3000mJ/cm². Skin biopsy was collected before and after phototherapy. The gene expression of BMP4, Noggin, Gremlin Sclerostin and melanin synthesis genes before and after NB-UVB therapy were checked by RT-PCR analysis.

Results: In patients which were not responding to phototherapy, BMP signalling molecule i.e, BMP-4 was significantly increased after NB-UVB phototherapy whereas the expression of Noggin, Sclerostin, Gremlin were reduced after phototherapy. The expression of melanin synthesis genes was also reduced in these patients after phototherapy.

Conclusion: The pilot study revealed that expression of BMP-4 was upregulated in vitiligo patients undergoing phototherapy in which no pigmentation was observed.

Further, the inhibitors of BMP were downregulated. So this study might indicate the potential use of BMP inhibitors in repigmentation of vitiligo. Further extensive studies at both transcriptional and translational level are still required to validate these findings.



Abstract N°: 5042**Case of systemic lupus erythematosus with lupus nephritis presenting with painful swelling over axillary region - lupus panniculitis: a rarity**Senkadhir Vendhan*¹, Biju Vasudevan¹, Shekhar Neema¹, Kashif A W²¹Armed Forces Medical College, Pune, Dermatology, ²Armed Forces Medical College, Pune, Pathology**Introduction:**

Lupus panniculitis is a rare type of chronic cutaneous lupus erythematosus manifesting as deep subcutaneous nodules. Areas of predilection include the upper arms, shoulders, face, scalp, hips, axilla, buttocks, and breasts. The age of presentation ranges from 30 to 60 years. We hereby present a case of 23 years old lady, a case of lupus erythematosus with lupus nephritis presenting with a painful swelling on both the sides of the axilla and groin managed successfully with intravenous immunoglobulin.

Case report:

23-year-old unmarried woman, with a known case of lupus erythematosus with lupus nephritis who sought consultation due to a 6-week history of indurated plaques over both sides of the axilla, groin, left breast, and neck. Lesions were associated with pruritus, pain, and tenderness. She eventually experienced fever, abdominal pain, and weakness. She was admitted with an initial assessment of extrapulmonary pulmonary tuberculosis vs. malignancy probably hematologic vs. breast. A complete blood count revealed anemia and leukopenia. Peripheral blood smear revealed no immature cells. ESR, CRP, and ANA were elevated and decreased C3 and C4 levels. A computerized tomographic scan of the chest revealed a soft tissue tumor over the axilla with malignant features. Skin punch biopsy showed plasma cell infiltrates with nuclear dust extending into a portion of the subcutaneous fat with hyaline fat necrosis. She was treated with hydroxychloroquine 200 mg once a day, prednisone 35 mg/d, and injection of methylprednisolone 1gm for 3 days with no response following which, IVIG@2gm/kg and steroids were tapered gradually. She responded well to the treatment and was discharged.

Discussion:

Lupus panniculitis can produce breast nodules that mimic breast carcinoma both clinically and radiologically. This case highlights the importance of lesional biopsy as the cornerstone of cutaneous lupus erythematosus diagnosis. IVIG works well in cases with diffuse cutaneous involvement, recalcitrant cases, or in case of infections.

Conclusion:

When lupus panniculitis presents in combination with SLE, it is considered a marker for less severe SLE. But in our case, it was associated with severe type. Lesions of lupus panniculitis may resolve with hyperpigmentation or depressed scars.

Abstract N°: 5043**Conception and Conduction of the German Vitiligo Registry VitiBest.**

Caroline Gewiss*¹, Nesrine Ben-Anaya¹, Markus Böhm², Matthias Augustin¹

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Introduction & Objectives: Vitiligo is a chronic autoimmune-mediated skin disease leading to a loss of function and progressive destruction of melanocytes. Clinical characteristics are sharply demarcated white macules affecting the skin and mucosa in varying degrees of spread and severity, depending on the subtype of vitiligo. The visual changes can lead to significant psychosocial impairment and a severe reduction in the quality of life of those affected. Vitiligo is also associated with an increased rate of other autoimmune diseases (e.g. thyroid disease and alopecia areata) and chronic inflammatory skin conditions. The aim is to conceptualize and establish a patient registry for vitiligo patients in Germany. The registry recruits all patients with vitiligo diagnosed and treated in the participating centers.

Method: Multiple investigational sites in Germany will recruit at least 800 patients over a period of three years with no upper limit on the number of patients included and each patient will be observed for 2 years. A core data set of variables assessed by both the physician and the patient has been specified. Multiple time points of documentation are permitted. VitiBest is a disease registry based on electronic, web-based data collection in a standardized eCRF. Continuous analyses of recruitment and baseline data are possible. Consensus reports, expert recommendations, and annual updates from the registry are the communication tools.

Results: The registry will provide for the first time real life data in Germany on vitiligo itself, comorbidities, psychosocial impact, efficacy as well as the safety aspects of given therapeutics. It will facilitate the development of predictive models and lead to multiple benefits at both clinical and research levels.

Conclusion: A registry for vitiligo patients is an important and scientifically accepted method to receive answers to many clinical and epidemiological questions concerning vitiligo.



Abstract N°: 5066

Pemphigus Vegetens : epidemiological, clinical, and histopathological features

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Introduction & Objectives:

Pemphigus vegetans (PVeg) is a rare variant of pemphigus vulgaris, representing only 2% of all cases of pemphigus. It is characterized by two clinically recognized forms known as the Hallopeau type and the Neumann type.

We aimed to describe epidemiological, clinical, and histopathological features of PVeg our patients.

Materials & Methods:

A retrospective study was conducted from January 2010 to December 2022, including all patients with PVeg.

Results:

A total of ten cases of PVeg were collected, with a sex ratio of 0.42 and a mean age of 49.18 years \pm 15.6. The mean interval between the diagnosis of pemphigus and the onset of vegetative lesions was 10 months \pm 22. The oral mucosa was the most commonly affected site (n=6). Vegetations were also observed on the trunk, elbows, face (n=5), inguinal and axillary folds, umbilicus (n=4), and lips (n=1). Histologically, suprabasal acantholysis (n=7) and subcorneal acantholysis (n=3) were noted. Clinically, a classical subtype could be identified in 8 cases, with Neumann's PVeg in 4 cases and Hallopeau's PVeg in 4 cases. All patients received treatment with systemic corticosteroids, and immunosuppressive therapy was additionally administered in 3 cases. PVeg was challenging to classify in 2 cases. An atypical tumor-like form was found in a 51-year-old patient with a 25-year history of superficial pemphigus. The patient presented with multiple papillomatous vegetating tumors on the scalp, neck, and back, without prior bullous or pustular lesions. Treatment with fractional CO₂ laser therapy resulted in a favorable outcome. Another atypical form was observed in a 33-year-old patient with a prior diagnosis of pemphigus vulgaris, who subsequently developed bilateral vegetating plaques on the big toes.

Conclusion:

PVeg is typically characterized by the development of vegetative lesions predominantly in flexural and periorificial areas. Histological examination reveals hyperplastic epidermis with intramalpighian leukocyte microabscesses and suprabasal acantholysis. Our case series highlights the clinical heterogeneity of PVeg, including frequent mucosal involvement, its co-occurrence with pemphigus vulgaris and superficial variant of pemphigus. Additionally, we observed in one case the presence of vegetations without p bullous or pustular lesions. Histologically, both suprabasal and superficial acantholysis can be observed. Patients can present with the classic forms of PVeg (Hallopeau and Neumann), as well as with unclassifiable variants. Systemic treatment is similar to that for Pemphigus

Vulgaris, with high-dose steroids being the first choice therapy. Immunosuppressive agents may allow a steroid-sparing effect.

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Abstract N°: 5074**Rheumatoid arthritis - dermatomyositis - systemic lupus erythematosus overlaps in a middle-aged Asian woman: a rarity.**

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¹Armed Forces Medical College, Pune, Dermatology, ²Armed Forces Medical College, Pune, Pathology

Introduction:

Overlap syndromes are rare inflammatory rheumatic conditions that satisfy the classification criteria for at least two connective tissue diseases that include systemic lupus erythematosus, rheumatoid arthritis, systemic sclerosis, polymyositis/dermatomyositis, and Sjögren syndrome occurring at the same time or at different times in the same patient. In our case report, we describe the case of a 51-year-old lady fulfilling the criteria for rheumatoid arthritis, dermatomyositis, and systemic lupus erythematosus managed successfully with IVIG.

Case report:

A 51-year-old lady presented with multiple joint pains, swelling and restriction of movements of 30 years duration, redness and burning over sun-exposed areas and weakness of extremities and trunk of 1-year duration, diffuse thinning/loss of hair of 6 months duration. Confluent hyperpigmented plaques with scaling over the face including the nasolabial fold with relative sparing of the nasal bridge. Involvement of trunk & upper limbs in the form of diffuse erythema and scaling with surface telangiectasias. Heliotrope rash, V sign, Shawl sign, and Samitz sign over nailbed were noted. Diffuse alopecia with erythema and scaling, thinning of hair, and rough lustreless hair (lupus hair). Erythematous to violaceous plaques over the extensor surfaces of the metacarpophalangeal and interphalangeal region sparing the knuckles were present. Livedo reticularis was noted over the bilateral thigh region. The antinuclear antibody profile was positive with a 2+ nuclear, speckled pattern. ENA profile showed Ro 52 Positivity and TIF 1 gamma strong positivity. C3 and C4 levels were reduced. The patient was managed with IVIG, and a follow-up of the patient showed decreased hyperpigmentation scaling and improvement in weakness.

Discussion:

Dermatomyositis is a rare idiopathic chronic acquired autoimmune inflammatory myopathy with skin manifestations that vary in severity. Incidence is 1 per 100 000 population, with peaks at ages 5–15 years in children and ages 45–60 years in adults. A disorder can only be considered an overlap syndrome if the patient meets the separate diagnostic criteria for more than one autoimmune connective tissue disease. The reported incidence of overlap syndrome ranges from 11% to 40% in patients diagnosed with dermatomyositis. Our patient fulfilled 2010 ACR/EULAR criteria for Rheumatoid arthritis with a score of 8 points [1-3 small joints(3), High positive RF/anti-CCP (3) Abnormal ESR (1), duration of symptoms > 6 weeks (1)], Bohan Peter criteria as probable dermatomyositis (proximal and symmetrical weakness, elevation of serum skeletal muscle enzyme levels, typical cutaneous lesions and ACR/EULAR criteria for SLE with a score of 17 points[ANA - Positive, Fever (2), Alopecia(2), Arthritis(6), Leukopenia(3), Low C3 + C4(4)].

Conclusion:

Our patient met the criteria for Rheumatoid arthritis, Dermatomyositis, and Systemic lupus erythematosus, which is a rare overlap syndrome reported in the literature. Our patient showed a reduction in erythema, scaling, and weakness following intravenous immunoglobulin injection, and she is currently being monitored without experiencing any negative side effects.

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Abstract N°: 5088

Pemphigus hepetiformis : A 22-year single-center experience

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Introduction & Objectives:

Pemphigus herpetiformis (PH) is a rare variant of pemphigus, which poses diagnostic challenges. It is distinguished by clinical manifestations resembling dermatitis herpetiformis and immunological findings consistent with pemphigus.

The purpose of our study was to describe the epidemiological, clinical, and immunological features, as well as the management approaches employed for patients with PH.

Materials & Methods:

We conducted a retrospective study that included seven patients diagnosed with PH between January 2010 and December 2022. The medical records of these patients were thoroughly examined to extract essential information, including patient characteristics, clinical aspects, histologic data, and follow-up data

Results:

We included a total of seven female patients with a mean age of 35 years in our study. The average duration before the initial medical evaluation was 12.4 months. All patients presented with a combination of annular erythematous plaques and grouped vesiculobullous lesions, often in a herpetiform configuration, accompanied by severe pruritus primarily affecting the trunk (6 cases) and limbs (5 cases). Mucosal involvement was observed in one patient. Nikolsky's sign was positive in 2 cases. Histopathological examination revealed eosinophilic spongiosis in 5 cases and acantholysis in 4 cases. Suprabasal cleavage was observed in 5 cases, while subcorneal cleavage was detected in 2 cases. Direct immunofluorescence consistently demonstrated intercellular deposition of immunoglobulin G and C3 in the epidermis. ELISA testing revealed reactivity against Dsg1 in all patients, while only one patient exhibited anti-Dsg3 antibodies. Systemic corticosteroids were administered to all patients at a dosage of 1 to 1.25 mg/kg daily. The average time until disease control was achieved was 20 days. In two cases, additional treatment with methotrexate (7.5 mg/week) and azathioprine (100 mg/day) was required, showing favorable progress. Relapses occurred in two patients after 24 and 36 months of treatment.

Conclusion:

Skin lesions in PH typically exhibit similarities to dermatitis herpetiformis, manifesting as erythematous, vesicular, bullous, or papular lesions. These lesions commonly appear in a herpetiform pattern and are accompanied by severe pruritus, as observed in our patients. Oral mucosal involvement is rare and was observed in only one patient. Positive Nikolsky's sign may be present in certain cases. Histologically, PH is characterized by the presence of eosinophilic spongiosis and intraepidermal eosinophilic pustules, accompanied by acantholysis. Direct immunofluorescence studies confirm the diagnosis by revealing intercellular deposition of immunoglobulin G (IgG) and complement component C3 in the epidermis. The management of PH primarily involves the use of systemic corticosteroids as the main treatment approach. Additional immunosuppressive agents may be considered in refractory cases or as steroid-sparing agents. Given the rarity of PH, further research and clinical studies are needed to better understand its pathogenesis, optimal management strategies, and long-term

outcomes

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Abstract N°: 5144**Beyond direct immunofluorescence: the role of serological investigations in the diagnosis of bullous Pemphigoid**Carlo Pipitò*¹, Maria Efenesia Baffa¹, Roberto Maglie¹, Stefano Senatore¹, Marzia Caproni¹, Giuseppe Pipitò², Emiliano Antiga¹¹University of Florence, Department of Health Sciences, Florence, Italy,²University of Bari, Department of Computer Science, Bari, Italy**Introduction & Objectives:**

Bullous Pemphigoid (BP) is the most frequent autoimmune sub-epidermal blistering dermatosis. The diagnosis is based on a combination of criteria encompassing clinical features, positive direct immunofluorescence microscopy (DIF) findings and serological tests for the detection of circulating IgG anti basement membrane zone autoantibodies. The study aims to determine which cases can be diagnosed in the absence of direct immunofluorescence (DIF) microscopy test, looking for a threshold value of BP180 NC16A enzyme-linked immunosorbent assay (BP180 NC16A ELISA) to predict the direct immunofluorescence result with 100% specificity and to assess the diagnostic accuracy of the different tests available for the diagnosis of bullous Pemphigoid.

Materials & Methods:

This paired, multivariable diagnostic accuracy study analyzed data from 245 consecutive patients with suspected bullous Pemphigoid referred to the Florence Center for Blistering Diseases from secondary and tertiary care hospitals. Eligible participants were patients with paired data on at least (1) a skin biopsy specimen for the direct immunofluorescence (DIF) microscopy test; (2) indirect immunofluorescence on a human salt-split skin substrate (IIF SSS) test; and (3) BP180 NC16A and BP230 enzyme-linked immunosorbent assay administered between January 1, 2018, and October 1, 2022. Samples were taken from patients at the time of first diagnosis. In our analysis, we used statistical models to assess the diagnostic usefulness of combining multiple diagnostic tests (such as DIF, IIF SSS, BP180 NC16A ELISA, and BP230 ELISA) in the initial diagnosis of bullous Pemphigoid. Additionally, we incorporated clinical variables like age, sex, presence of pruritus, and blisters into our analysis.

Results:

Of the 245 patients analyzed, 130 participants received a diagnosis of Pemphigoid, with 115 controls. The DIF microscopy was the most sensitive and specific test (Decision Tree model: 94.6%; 100% CI, 89.8%-100%. Logistic regression model: 94.2%; 100% CI, 96.2%-100%) whereas IIF SSS and BP180 NC16A ELISA were less sensitive and specific. About the results of the multivariable logistic regression analysis and decision tree analysis of combined tests, The DIF microscopy plus IIF SSS combined test (Decision Tree model: 97.8%; 100% CI, 95.4%-100%. Logistic regression model: 97.8%; 100% CI, 94.4%-100%) and the DIF microscopy plus BP180 NC16A ELISA (Decision Tree model: 98.5%; 100% CI, 97.7%-100%. Logistic regression model: 97.7%; 100% CI, 97.7%-100%) were the most sensitive and specific diagnostic test. Regarding combined serological tests, BP180 NC16A ELISA plus IIF SSS was the most sensitive and specific test (Decision Tree model: 86.7%; 95.8% CI, 85.4%-100%. Logistic regression model: 87.7%; 100% CI, 95.1%-100%). Additionally, the analysis allowed us to determine the value of BP180 above which it is possible to predict the direct immunofluorescence result with 100% specificity.

Conclusion:

The results found that direct immunofluorescence remains the gold standard in diagnosing bullous Pemphigoid.

The addition of serological tests improves the diagnostic accuracy of DIF microscopy. In some instances, utilizing the BP180 NC16A ELISA alone may enable diagnosing patients exhibiting compatible clinical features.

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Abstract N°: 5280

Milia Formation Within Resolving Bullous Pemphigoid Lesions: A Case of Bullous Pemphigoid with Secondary Milia Formation in a 41-year-old Filipino Male

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Introduction & Objectives:

Milia are benign superficial keratinous cysts clinically described as pearly white, dome-shaped papules 1-2mm in diameter. There are two types: primary milia, which occurs spontaneously, and secondary milia, which occurs from various secondary processes including blistering diseases. Secondary milia formation is commonly associated with epidermolysis bullosa acquisita and mucous membrane pemphigoid, however, a few case reports of milia have been linked to bullous pemphigoid. Here we report a case of secondary milia formation within bullous pemphigoid lesions

Materials & Methods:

A 41-year-old Filipino male presented with a 1-month history of tense, clear fluid-filled vesicles and bullae on erythematous urticarial base some with erosions and crusts on the trunk and upper and lower extremities. Diagnosis was bullous pemphigoid based on clinical features, histopathology, direct immunofluorescence, and Anti-BP 180.

Results:

Patient was started on oral Prednisone, Doxycycline, and Niacinamide along with topical Clobetasol Propionate ointment. After 8 weeks of treatment, patient presented with asymptomatic pinpoint white papules within resolving vesicles and bullae. Dermoscopy findings and histopathology report confirmed that the papules are milia formation, in this case, secondary to bullous pemphigoid. No additional treatment for milia was needed due to its benign and self-limiting nature.

Conclusion:

This case provides further evidence of secondary milia formation in bullous pemphigoid thus the presence of milia should not be limitedly associated to certain blistering diseases.



Abstract N°: 5487**Efgartigimod: A Novel FcRn Antagonist in the Treatment of Autoimmune Diseases**Kris Van de Walle^{*1}, Beatriz Wittlin¹, Bianca Balbino¹, Peter Ulrichts¹, Peter Verheesen¹, Ivaylo Stoykov¹¹argenx BVBA, Ghent, Belgium**Introduction & Objectives:**

Immunoglobulin G (IgG) autoantibodies are key in autoimmune disease pathogenesis, including pemphigus and bullous pemphigoid (BP), immune thrombocytopenia (ITP), and neurological conditions: generalized myasthenia gravis (gMG), chronic inflammatory demyelinating polyneuropathy (CIDP) and myositis. The objective was to determine whether reducing IgG level by inhibiting the neonatal Fc receptor (FcRn) could lead to improved outcomes in patients with autoimmune diseases.

Materials & Methods:

FcRn is the central regulator of IgG homeostasis. Efgartigimod, a human IgG1-derived Fc-fragment and FcRn antagonist, outcompetes endogenous IgG binding, reduces recycling and increases IgG degradation, including all IgG subtypes, without impacting other immunoglobulins or albumin, making it a potential treatment for IgG-mediated disorders.

Results:

Based on the clinical benefit in the Phase 3 ADAPT study, efgartigimod was approved for treatment of adult patients with anti-acetylcholine receptor antibody positive gMG in several countries. In a phase 2 adaptive trial in pemphigus vulgaris and foliaceus, efgartigimod was well tolerated and exhibited an early effect on disease activity and outcome parameters, providing support for further evaluation in an ongoing Phase 3 study (ADDRESS). The Phase 3 ADVANCE study in ITP met the primary endpoint with higher, sustained platelet response with efgartigimod vs placebo. Other ongoing studies include another Phase 3 study in ITP (ADVANCE SC), and Phase 2/3 studies in CIDP (ADHERE), myositis (ALKIVIA) and BP (BALLAD). Across all completed studies to date, efgartigimod reduced total IgG, including pathogenic IgG, corresponding to clinical improvements in each population. Efgartigimod safety is consistent, with comparable treatment-emergent adverse event (TEAE) rates to placebo (ADAPT 77.4% efgartigimod/84.3% placebo; ADVANCE 93.0% efgartigimod/95.6% placebo; 85% of participants open-label pemphigus study). Most TEAEs were mild to moderate in severity. Efgartigimod was well tolerated in the ADAPT+ extension study, with no increase in TEAE incidence rates or infections with up to 19 treatment cycles.

Conclusion:

FcRn inhibition by efgartigimod is a promising therapeutic option for autoimmune diseases mediated by pathogenic IgG autoantibodies.



Abstract N°: 5514**Amyopathic dermatomyositis: the first manifestation of a prostate adenocarcinoma**Catarina Cerqueira¹, Ana Lopes¹, Carlos Nogueira¹, Maria Guimaraes¹, Catarina Araújo¹, Maria Celeste Brito¹¹Hospital de Braga, Dermatology and Venereology**Introduction & Objectives:**

Dermatomyositis is an autoimmune disease characterized by proximal inflammatory myopathy and characteristic skin rashes. In approximately 20% of patients, there is absence of muscle involvement, known as amyopathic dermatomyositis and in this group, the presence of an underlying neoplasm is less frequent (14% of patients compared to 30% in dermatomyositis with muscle involvement).

Materials & Methods:

We present the clinical case of a 74-year-old male patient with a history of hypertension, dyslipidemia, and benign prostatic hyperplasia, referred to the Dermatology clinic for psoriasiform lesions on the elbows and scalp evolving for 5 months, and erythema on the face, neckline, and forearms evolving for 2 months. On physical examination, he presented with heliotrope rash, poikiloderma erythema on the anterior thoracic region as well as on the abdominal and dorsal regions, outer aspect of the forearms, and anterior aspect of the thighs, along with ulcers on the back of the hands. Additionally, he had Gottron's papules and erythematous, scaly, psoriasiform, pruritic lesions on both elbows and the scalp. There were no other associated complaints, such as muscle weakness or asthenia. Due to clinical suspicion of dermatomyositis, an incisional skin biopsy of the neckline region was performed. He was prescribed prednisolone 60 mg/day, hydroxyzine 25 mg/day, photoprotection, and emollients. The skin biopsy revealed atrophic epidermis, dermal elastosis lesions, and a perifollicular lymphocytic infiltrate, consistent with dermatomyositis. Laboratory analysis showed aldolase levels of 13.4 U/L, ANA 1/160, positive anti-TIF-1 γ antibodies, positive anti-Ro52 antibodies, and a PSA level of 31 ng/ml. Paraneoplastic studies were requested, including upper and lower gastrointestinal endoscopy, thoracoabdominopelvic computed tomography, and thyroid ultrasound. An Urology consultation was also requested due to elevated PSA levels. Given the worsening of the condition, human immunoglobulin therapy was initiated at a dose of 2.5 mg/kg for 3 days, every 4 weeks, resulting in improvement of erythema and absence of ulcers. Prostate magnetic resonance imaging was performed, and the prostate biopsy revealed adenocarcinoma. Staging of the neoplasm is pending.

Results:**Conclusion:**

With this study, we emphasize the importance of screening for paraneoplastic syndromes in all adult patients with amyopathic dermatomyositis.



Abstract N°: 5546**Overview of the Safety Profile from Efgartigimod Clinical Trials in Participants with Diverse IgG-Mediated Autoimmune Diseases**

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Introduction & Objectives:

Efgartigimod is a first-in-class, human IgG Fc fragment that inhibits the neonatal Fc receptor (FcRn) and outcompetes endogenous IgG binding. This results in reduced recycling and increased degradation of IgGs, including pathogenic IgG autoantibodies. FcRn inhibition by efgartigimod is a rational therapeutic option for IgG-mediated autoimmune disorders. The objective was to assess the safety profile of efgartigimod across different IgG-mediated disorders.

Materials & Methods:

Intravenous (IV) efgartigimod safety was assessed in generalized myasthenia gravis (gMG) in phase 2, phase 3 (ADAPT) trials, and a 3-year open-label extension (ADAPT+) trial. It was also evaluated in a phase 3 trial (ADVANCE) in primary immune thrombocytopenia (ITP) and an open-label phase 2 trial in pemphigus (vulgaris and foliaceus). These studies examined different dosing regimens (10–25 mg/kg) of efgartigimod, including cyclical dosing in gMG and continuous weekly dosing in ITP and pemphigus.

Results:

Across all indications and doses studied, efgartigimod demonstrated a consistent safety profile, with comparable treatment emergent adverse event (TEAE) rates to placebo (ADAPT 77.4% efgartigimod/84.3% placebo; ADVANCE 93.0% efgartigimod/95.6% placebo; 85% of participants in the open label pemphigus study). Most TEAEs across studies were mild to moderate in severity. Discontinuation rates due to adverse events were consistently low across studies (3.6% efgartigimod group/3.6% placebo in ADAPT; 3.5% efgartigimod/2.2% placebo in ADVANCE; and 3% of pemphigus study participants). Efgartigimod was well tolerated in ADAPT+, with no increase in TEAE incidence rates or infections with repeated efgartigimod cycles (up to 19). Efgartigimod treatment did not reduce serum albumin levels or increase cholesterol levels.

Conclusion:

Efgartigimod is well tolerated across indications and doses studied. Most TEAEs, including infections, were mild or moderate in severity and did not increase in frequency with recurrent dosing.



Abstract N°: 5582

Comparison of Dipeptidyl Peptidase-4 Inhibitor-Induced Bullous Pemphigoid and “Conventional” Bullous Pemphigoid: A Retrospective Study of Clinical and Laboratory Characteristics

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Introduction & Objectives:

Bullous pemphigoid (BP), mostly seen in the elderly population, is the most common autoimmune bullous disease. In addition to factors such as genetic predisposition, neuropsychiatric disorders, physical agents and UV light playing a role in its etiopathogenesis, a group of antidiabetics, dipeptidyl peptidase-4 (DPP4) inhibitors (gliptins) have recently been shown to increase the risk of BP. The aim of this study was to investigate the clinical and laboratory differences between gliptin-induced and normal BP patients.

Materials & Methods:

BP patients treated in our clinic from January 2010 to May 2023 were included.

Age, sex, comorbidities, treatments used for comorbidities, clinical features of BP (date of diagnosis, disease severity, distribution of lesions, existence of a prebullous phase and pruritus), biopsy results, IgE levels, complete blood count results, comorbidities and medications used for comorbidities, and treatments given for BP were noted for each patient.

Patients using gliptins were separated as a different group and compared to patients not using gliptins.

Data were analyzed using independent sample t-test for parametric distribution, Kruskal-Wallis test for non-parametric data, and chi square test for correlation analysis.

Results:

Out of 105 patients included, 11 were taking gliptins at time of diagnosis. There was no statistically significant difference between sex, current age, age at diagnosis, presence of pruritus, IgE levels, and eosinophil count. Female patients constituted the majority of both groups (63.6% for gliptin group and 59.6% for the non-gliptin group) ($p=0.79$). 90.9% of gliptin using patients presented with a prebullous phase, this percentage was lower in the non-gliptin group (50.0%) ($p=0.01$). The disease severity was significantly higher in the non-gliptin group ($p=0.007$). There was no statistically significant difference between the two groups in terms of involvement areas (body, extremities, face and mucosae) ($p=0.84$).

Patient comorbidities and medications used for these comorbidities were also analysed. The frequencies of hypertension and hyperlipidemia were statistically significantly higher in the gliptin using group (Table 1). Among medications used for comorbidities, statins, angiotensin receptor blockers (ARB), insulin and insulin secretagogues were used significantly more frequently in BP patients using gliptin (Table 2).

	No gliptin		Gliptin using		p value
	Number of patients	Percentage (%)	Number of patients	Percentage (%)	
Cardiovascular disorder	6	6.4	1	9.1	0.733
Hypertension	31	33.0	7	63.6	0,045*
Hyperlipidemia	1	1.1	3	27.3	0,000*
Dementia	7	7.4	1	9.1	0.845
Chronic kidney disease	4	4.3	1	9.1	0.507

Table 1. The frequencies of comorbidities in the patient groups.

	No gliptin		Gliptin using		p value
	Number of patients	Percentage (%)	Number of patients	Percentage (%)	
Statins	0	0.0	2	18.2	0,001*
Diuretics	10	10.6	2	18.2	0.476
Calcium channel blockers	8	8.5	1	9.1	0.948
ACE inhibitors	5	5.3	1	9.1	0.61
Angiotensin receptor blockers	3	3.2	5	45.5	0,000*
Alpha blockers	3	3.2	1	9.1	0.333
Beta blockers	4	4.3	1	9.1	0.476
Anti-platelet agents	5	5.3	1	9.1	0.61
Insulin	12	12.8	6	54.5	0,001*
Insulin secretagogues	1	1.1	2	18.2	0,001*
Metformin	7	7.4	2	18.2	0.228

Table 2. The frequencies of medications used for comorbidities in the patient groups. (ACE: Angiotensin converting enzyme)

Conclusion:

This is one of the first studies to compare clinical characteristics of gliptin using BP patients with other BP patients. Our results suggest that BP patients using gliptins more frequently present with a more severe disease and a prebullous phase. An important finding was that gliptin using patients had higher frequencies of having hypertension and hyperlipidemia, and using statins, ARB, insulin and insulin secretagogues. The sample size of this study is limited to suggest a definitive correlation, and studies with larger cohorts are warranted. However, we believe it may be worthy for endocrinologists to keep these findings in mind and choose patients carefully when starting gliptins for the treatment of diabetes.

Abstract N°: 5605**Lichen Planopilaris Outside the Scalp: A Case Report**

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Introduction & Objectives:

Lichen planopilaris (LPP) is considered a follicular form of lichen planus, typically manifesting as patchy areas of cicatricial alopecia on the scalp. Alopecia in other hair-bearing areas of the skin seems to be an uncommon feature of classic LPP, although commonly described in other variants (with cicatricial alopecia of the eyebrows in frontal fibrosing alopecia and non-cicatricial alopecia of the axillae and groin in *Graham-Little-Piccardi-Lasseur* syndrome).

Materials & Methods:**Results:**

We report a case of a healthy 36-year-old man with ongoing pruriginous hair loss on the lower legs, forearms, and right eyebrow, progressing for a full year prior to the first evaluation. The patient reported no hair regrowth after therapy with topical minoxidil and pimecrolimus. The physical examination showed large patches of cicatricial hair loss in the affected regions, with peripheral perifollicular erythema and hyperkeratosis on dermoscopy. There was no alopecia or similar findings on scalp hair. A punch biopsy was performed on the right leg, showing peri-infundibular mucoid fibrosis and a perifollicular lymphocytic infiltrate, with lymphocytes permeating the infundibular interface and rare apoptotic keratinocytes, findings suggestive of LPP. There were no alterations in the full blood count, liver enzymes and renal/thyroid functions. The antinuclear antibodies, as well as the screening for syphilis, HIV and viral hepatitis were negative. Treatment was initiated with a combination of topical corticosteroids and topical tacrolimus, as well as oral minoxidil, with an improvement of pruritus and cessation of hair loss, although no hair regrowth was observed in the affected regions, as expected. The patient maintains regular follow-ups, with no signs of scalp disease up to three years after the first symptoms.

Conclusion:

LPP with no scalp findings and exclusive body hair loss appears to be a rare occurrence and we could only find one similar case in the pre-existing literature. The natural history of LPP with these characteristics is unknown and the diagnosis requires a high clinical suspicion, making the report of these cases extremely relevant.

Abstract N°: 5649

Neutrophil Ageing and change in the expression of the homing marker CXCR4 in patients with Pemphigus Vulgaris.

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Introduction & Objectives: Pemphigus vulgaris (PV) is a fatal autoimmune blistering disease marked by high levels of autoantibodies for desmogleins (Dsg1 and Dsg-3). Despite a clear aetiology, the triggers for the production of autoantibodies are still unknown. Neutrophils, the prime cells of immune defence, are poorly defined in pemphigus pathogenesis. CXC-motif chemokine receptor 4 (CXCR4) accumulates on neutrophil cell surfaces with their age, which dramatically changes migration. Since CXCR4 expression allows them to arrive at the site of inflammation rather than returning to the bone marrow. This ageing process makes neutrophils more likely to release DNA fibres known as neutrophil extracellular traps (NETs), which can expose self-antigens and promote autoantibody generation. To test this role of neutrophils in pemphigus, we designed the present pilot study to compare neutrophil ageing and NETs-forming capacity among neutrophils in PV patients compared to healthy controls.

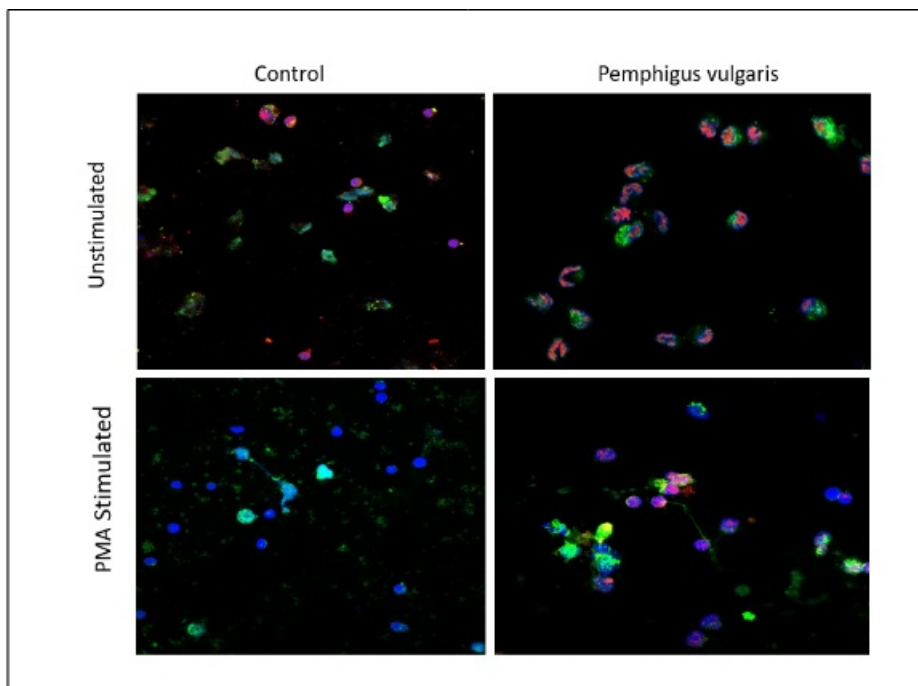
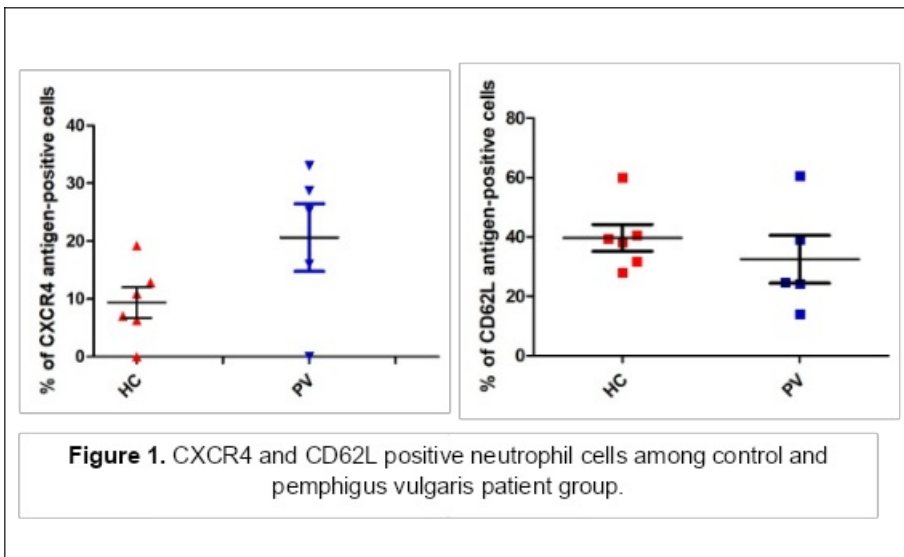
Materials & Methods: Patients with active disease were recruited after assessing their anti-DSG1 & anti-DSG3 levels using a dermatology profile ELISA (IgG) kit. Healthy individuals who consented to participate in the study were also enrolled. About 5 ml of blood from patients and healthy individuals was collected in an EDTA vial and processed for FACS and NETosis analysis. The expression of CD11b, CXCR4, and CD62L was analysed through FACS. For NETosis, the neutrophils were stimulated with phorbol myristate acetate (PMA) for 6 hours and stained with DAPI, anti-MPO-FITC, and anti-Histone H3-Cy3 antibodies to visualise NETs. Images were acquired using confocal microscopy. The number of NETs was calculated using ImageJ software. GraphPad Prism was used to conduct the statistical analysis.

Results: The mean ages of recruited patients and healthy controls were (45±7.1years) and (42±6.1years), respectively. The recorded levels of anti-DSG1 and anti-DSG3 were (6.59±6.6 & 7.61±1.4) respectively, in PV patients (normal range 0.0-1.0). The total percentage of neutrophils was about 48% in the control group and 35% in PV patients. The observed circulating CXCR4 positive cells were high in pemphigus vulgaris patients (20.6±13.1%) in comparison to healthy controls (9.38±6.5%), while the levels of CD62L showed the reverse trend (PV 32.5±18% vs. HC 39.7±11.1%). In addition, compared to healthy controls, the neutrophils of PV patients had a greater potential for NET formation.

Conclusion: The higher expression of CXCR4 and low CD62L markers in PV patients compared to controls indicates the accumulation of aged neutrophils. These aged neutrophils displayed a higher NET forming potential compared to the ones isolated from HC. Both of these changes might be a reflection of the active disease state. Further, comprehensive data is needed to validate this association.

Figure 1. CXCR4 and CD62L positive neutrophil cells among control and pemphigus vulgaris patient group.

Figure 2. Immunofluorescence microscopy (IFM) of neutrophils forming NETs in control and pemphigus vulgaris group. DAPI-blue, MPO-FITC- green, and histone H3-Cy3-red.



Abstract N°: 5691**Post bullous milia formation in vildagliptin-induced recalcitrant bullous pemphigoid**Gulsen Akoglu¹, Ahmet Tecik¹, Elçin Kadar²

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Introduction & Objectives: Milia formation within healing bullous lesions of epidermolysis bullosa acquisita and mucous membrane pemphigoid is commonly observed; however, only a few cases of resolving bullous pemphigoid (BP) are associated with milia.

Materials & Methods: A 76-year-old female patient with pruritic widespread tense blisters and erosions on the trunk and extremities lasting for two months was referred to our dermatology outpatient clinic. Mucous membranes were intact. She had insulin-dependent diabetes mellitus for 50 years, hypertension, and coronary artery disease. Medical history revealed ongoing vildagliptin therapy for the last year to control hyperglycemia.

Results: Histopathological examination of the skin revealed subepidermal bullae and linear C3 (+) and IgG (+) depositions at the basement membrane in direct immunofluorescence. Immunofluorescence examination of salt-split skin substrate showed linear epidermal IgG (++) and dermal linear C3 (+) and IgG (+) and globular IgA (+) deposits. Serum anti-BP180 antibody was positive; anti-BP230, anti-collagen VII and envoplakin antibodies were negative; IgE levels were within normal limits. Screening tests did not detect any associated internal malignancies. Due to uncontrolled hyperglycemia, systemic steroid therapy was avoided. Systemic doxycycline combined with topical clobetasol propionate 0,05% ointment and omalizumab injections did not benefit. Methotrexate therapy caused grade 1 neutropenia. Since the patient had pneumonia, antibiotics were administered and intravenous immunoglobulin (2 g/kg, 4 weeks apart, 3 cycles) was considered best for BP therapy. The number of new blister formations remarkably reduced within a few weeks following the first IVIG infusion. Additionally, two doses of rituximab (1000 mg, two weeks apart) were administered. Healed areas of the skin presented prominent milia formation.**

Conclusion: Although uncommon, milia formation may accompany the healing process of recalcitrant gliptin-associated BP.



Abstract N°: 5734**Localized bullous pemphigoid after total knee arthroplasty**

Sara De Benito Mendieta¹, Giulia Greta Dradi¹, Marta Menéndez Sánchez¹, Diego De la Vega Ruiz¹, Joseph Griffiths Acha¹, Alejandra Méndez Valdés¹, Enrique Gómez de la Fuente¹, Elena García Zamora¹, Miguel Vela Ganuza¹, Reyes Gamo Villegas¹, José Luis López Estebanz¹

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Localized bullous pemphigoid after total knee arthroplasty**Introduction & Objectives:**

Bullous pemphigoid is an autoimmune blistering disease presenting with tense blisters, erosions and crusting with intense pruritus. It is usually a widespread disease found in elderly patients and in occasions, a trigger is identified such as drugs, infections or traumatism.

Materials & Methods:

A 79-year-old male with tense blisters and crusting over the right knee is referred to the dermatology clinic. The lesions were pruritic and localized around the scar of a recent total knee arthroplasty, with no other lesions upon physical examination. Two biopsies showed a subepidermal blister with little inflammation and a negative immunofluorescence. Lesions remitted with oral steroids but reappeared when the dosage was decreased. A few months later, after a re-intervention for an arthroplasty exchange he presented with a new episode of blistering over the scar and with emergence of new urticarial lesions on the trunk and upper extremities. The urticarial lesions showed eosinophilic spongiosis. Indirect immunofluorescence was positive for anti-basement membrane antibodies. Patch testing was performed with a standard spanish battery (GEIDAC), benzoyl peroxide, vancomycin and gentamycin, which were negative. The patient was managed with oral and topical steroids and doxycycline for almost a year until remission was achieved.

Results:

We present the case of a patient with recurring blistering around the scar of a total knee arthroplasty, which were followed by generalized urticarial plaques after a second surgery. The histological study showed two distinct pattern: a subepidermal blister and eosinophilic spongiosis, both compatible with bullous pemphigoid in two different phases (bullous and urticarial). Indirect immunofluorescence was positive for anti-basement membrane antibodies, which is considered a potent marker for this disease.

Differential diagnosis included allergic contact eczema to the prosthesis, which was excluded with patch testing. Cases of localized bullous pemphigoid after a variety of surgical interventions have been described in the literature, including knee arthroplasties. However, a disease course with such extensive lesions is exceptional and only two similar cases have been reported. In only one case, an underlying sensitization to acrylates was found. Localized bullous pemphigoid is hypothesized to be due to a disruption of the dermo-epidermal junction through the surgical act, causing an immune disbalance with formation of autoantibodies.

Conclusion:

We present a case of localized bullous pemphigoid after a total knee arthroplasty with secondary generalization after the second surgery. When presented with bullous lesions around a recent scar we propose to take into

account this possibility in the differential diagnosis, after ruling out more frequent entities such as bullous impetigo and allergic contact eczema. Lastly, we want to emphasize the importance of carrying out patch testing in these patients due to the possibility, even if low, of an allergic underlying mechanism.

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Abstract N°: 5760**Transition of pemphigus vulgaris to pemphigus herpetiformis following adjuvant treatment with mycophenolate mofetil**Elia Valeva¹, Kossara Drenovska¹, Martin Shahid¹, Lyubka Stoyanova-Miteva¹, Snejina Vassileva¹¹Medical University of Sofia, Department of Dermatology and Venereology, Sofia, Bulgaria**Introduction & Objectives:**

Pemphigus is a group of autoimmune blistering disorders mediated by circulating and *in vivo* bound autoantibodies against desmosomal proteins. Multiple variants of the disease exist, including pemphigus vulgaris (PV), foliaceus (PF), herpetiformis (PH), etc. PV is generally considered to be a more severe subtype often requiring prolonged treatment with systemic corticosteroids that are well known for their numerous side effects. Other immunosuppressive agents, namely mycophenolate mofetil (MMF), are often added for their steroid-sparing effect. We report the case of PV transition to PH following MMF administration given to optimize a long-lasting steroid therapy.

Materials & Methods:

A 50-year-old Caucasian man with a 4-year history of mucosal-dominant PV affecting the oral cavity presented with a widespread, intensely pruritic herpetiform eruption involving the skin of the face, trunk and proximal parts of the extremities. The observed annular erythematous plaques and peripheral vesicles have appeared one week after the administration of MMF 2.0 g/daily added in order to minimize the cushingoid effects of the current maintenance steroid treatment with 8 mg methylprednisolone daily. The chronology and the change in the clinical presentation were suggestive of PV transition to PH. The diagnostic procedures included routine laboratory tests, histology, direct immunofluorescence (DIF) microscopy, indirect IF (IIF), and ELISA for anti-desmoglein (Dsg) autoantibodies.

Results:

Routine laboratory parameters were within normal ranges. Histology demonstrated spongiosis and intraepidermal vesicles. DIF showed intercellular deposition of immunoglobulin G (IgG) and complement C3 in the epidermis. IIF detected circulating IgG anti-epithelial cell surface antibodies at a titer of 1:320 on monkey esophagus substrate. ELISA tested strongly positive for Dsg3 and negative for Dsg1. MMF was suspected as the potential trigger of this pemphigus relapse and was therefore discontinued. Methylprednisolone 40 mg/day i.m., systemic antibiotics, antimycotics and topical high-potency corticosteroids led to progressive improvement of the blistering eruption.

Conclusion:

PH is a peculiar subtype of pemphigus clinically resembling dermatitis herpetiformis but displaying the immunopathologic features of pemphigus. PH has been reported in association with malignancies, infections, or drug intake. Transition between the various pemphigus subtypes, namely that of PH into PV or PF and *vice versa*, has been only rarely described. MMF is a first-line steroid-sparing agent known for its efficacy and relative safety in the treatment of moderate-to-severe PV. No other reports of such PV transition to PH under MMF treatment nor PH-like MMF-induced drug reactions have been described so far. Further studies of the immunopathology of PH should be conducted to elaborate on the atypical nature of this unusual case.

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Abstract N°: 5784**Wells syndrome**

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Introduction & Objectives:

Wells syndrome or eosinophilic cellulitis is a rare dermatosis of unknown etiology and pathogenesis. Many triggering factors have been proposed: insect bites, drugs, allergic contact dermatitis, an underlying myeloproliferative disorder, and infections. Typical lesions include recurrent, infiltrated erythematous plaques resembling cellulitis. It is characterized by great clinical variability and diagnosis is based mainly on the characteristic histopathology picture of eosinophilic infiltrate of the dermis.

A 57-year-old female was admitted to the Department of Dermatology due to erythematous, edematous lesions located on the trunk and upper limbs which had been present for about a year. The lesions were recurrent and self-limiting. Laboratory tests showed increased parameters of inflammation, eosinophilia and elevated tumor markers (CA19.9, CEA). In the histopathological examination, the inflammatory infiltrates were dominated by neutrophils with a large admixture of eosinophils. Based on the clinical presentation and the histopathological examination, a diagnosis of Wells syndrome was made. The patient was treated with glucocorticoids with a good response. The lesions subsided leaving mild hyperpigmentation. No new lesions appeared during treatment and 6 months of follow-up.

Conclusion:

Prognosis of Wells syndrome is excellent. Lesions usually resolve over weeks or months, but they may recur later. Systemic corticosteroids and dapsone are the two main treatments, apart from the etiological treatment of a triggering or associated disease.



Abstract N°: 5797**Bullous pemphigoid triggered by cardiac pacemaker implantation**Teodor Kozhuharov¹, Kossara Drenovska¹, Martin Shahid¹, Lyubka Stoyanova-Miteva¹, Snejina Vassileva¹¹Medical University Sofia

Introduction & Objectives: ##### Bullous pemphigoid (BP) is a subepidermal autoimmune blistering disease (AIBD) of the elderly which is mediated by autoantibodies targeting the structural components of the cutaneous basement membrane zone (BMZ). The exact cause of BP remains unknown, but various genetic, immunologic, endocrine, or environmental factors and drugs may contribute to its development. BP following insect bites, thermal burn, mechanical trauma, or surgical procedures, including implantation of medical devices has been rarely reported. We present a case of BP after cardiac pacemaker implantation.

Materials & Methods:

A 68-year-old man was admitted to our clinic for the acute onset of tense haemorrhagic blisters following a pacemaker implantation five days ago. The patient's medical history included arterial hypertension, ischemic heart disease, and myocardial infarction for which he had undergone coronary stenting, mitral and tricuspid valve replacement, and phlebotomy two weeks earlier. The clinical observation upon admission revealed tense hemorrhagic bullae on the trunk and left leg, strictly localized along the postoperative scars from the recent surgical procedures. The chronology, the clinical appearance and the specific lesions' distribution were suggestive of a subepidermal AIBD. The diagnostic procedures included routine laboratory tests, histology, direct immunofluorescence (DIF) microscopy, indirect IF (IIF), and epicutaneous patch tests.

Results:

The laboratory findings revealed anemia, mild leucocytosis, and elevated erythrocyte sedimentation rate. Histological examination demonstrated subepidermal clefting with tissue eosinophilia. DIF microscopy detected linear deposits of immunoglobulin G (IgG) with an n-serration pattern and complement C3 along the BMZ. IIF on monkey esophagus revealed anti-BMZ antibodies in the patient's serum with a titer of 1:640 and epidermal staining on salt-split skin substrate. Patch testing with standard European series of allergens was negative. The clinico-laboratory results were consistent with the diagnosis of BP. Systemic therapy with methylprednisolone 20mg/24h i.v., ceftriaxone 2g/24h i.v., as well as topical corticosteroids, antibiotic creams and braunovidon dressings resulted in complete healing of the cutaneous lesions.

Conclusion: ##### BP occurring after implantable devices surgery, including cardiac pacemaker implantation, has been rarely observed, the possible relationship between them being suspected in cases of well-defined chronology. The exact mechanisms of that association remain unknown but Koebner phenomenon after the surgical trauma, implant component allergy, or antigen unmasking during surgery and activation of autoimmunity could be discussed as potential triggering or exacerbating factors for BP. However, further research is needed to establish a definitive link between BP and pacemaker implants and to better understand the underlying mechanisms of this association.

Abstract N°: 5856**Recalcitrant Hemorrhagic Lesions in a Patient with Pemphigus Vulgaris: A Challenging Case**Pelin Ertop Doğan^{*1}, Merve Topuz¹, Emel Hazinedar¹, Rafet Koca¹¹Zonguldak Bülent Ecevit University Medical Faculty, Dermatology, Zonguldak, Türkiye**Introduction & Objectives:**

Pemphigus vulgaris (PV) is a rare autoimmune disease characterized by the formation of blisters and erosions on the skin and mucous membranes. Antibodies targeting the cell-to-cell adhesion molecules, particularly desmogleins, result in acantholysis and blister formation. There are triggering factors including human herpes simplex virus (HSV) infections leading to antibody production. There are controversial proposed mechanism regarding the role of HSV in the course of disease in PV patients. In this case report, we present a PV patient with persistent lesions complicated by a herpes virus infection.

Materials & Methods:

A 43-year-old male patient presented to our clinic with widespread erosions in the oral cavity and the body. After histopathological and immunofluorescence examinations, a diagnosis of pemphigus vulgaris was made. Despite treatment with systemic steroids and azathioprine, the patient's lesions could not be controlled. Due to the COVID-19 pandemic, monthly intravenous immunoglobulin therapy was added. The lesions regressed with this treatment and the patient was followed up for 5 months with stability. However, he suddenly presented with widespread lesions on the face and neck. His physical examination revealed erosions with hemorrhagic crust and fissures with angulated margins. The systemic steroid dose was increased up to 1 mg/kg. Meropenem was initiated for the patient, as the wound culture showed growth of *Pseudomonas aeruginosa*. However, the patient's lesions persisted even worsened despite these interventions. Subsequently, a Tzanck smear from the lesion revealed multinuclear giant cells, and PCR examination confirmed the presence of HSV-1. The systemic steroid dose was reduced, and intravenous acyclovir (3x10 mg/kg) was initiated. On the 6th day of acyclovir treatment, existing lesions showed improvement. As the lesions completely resolved, the intravenous acyclovir was discontinued on the 21st day, and the patient was switched to oral valacyclovir 500 mg once daily for prophylaxis.

Results:

The relationship between HSV and PV is not clearly understood and remains uncertain. While some studies suggest triggering effect of HSV due to possible molecular mimics in pemphigus or HSV as a complication of immunosuppressive therapy and impaired skin barrier, other studies have not found a significant correlation between the two entities. Additionally, clinical studies have shown that PV patients who were HSV-positive during the onset had a more severe relapsing disease course and shorter times to the first relapse. Regardless of the nature of this relationship, HSV superinfection should be considered, especially in cases of resistant pemphigus. HSV infection in pemphigus patients can be unrecognised due to clinical similarities. However, the presence of fissures, hemorrhagic crusts, erosions with angulated margins, and linear erosions should raise suspicion of HSV superinfection.

Conclusion:

In our patient, treatment resistance and presence of hemorrhagic crusts, fissures and angulated margins suggested the HSV superinfection.

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Abstract N°: 5859

Pemphigoid gestationis: a Single Center Experience

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Introduction & Objectives:

Pemphigoid Gestational (PG) is a rare autoimmune subepidermal bullous disease occurring mainly in multiparous women during the 2nd or 3rd trimester of pregnancy. The aim of our work is to determine the epidemiological, clinical, therapeutic and evolutionary profile

Materials & Methods:

A retrospective study of PG cases collected between 2004 and 2022 at dermatology and venerology department of the Mohammed VI Universal Hospital of Marrakesh.

Results:

They were 24 pregnant patients, 22 multiparous and 2 primiparous. The mean age was 31.5 years, ranging from 19 to 48 years. The onset was the 3rd trimester in 60% of cases, 2nd trimester in 20%, and postpartum in 20%. Similar occurrence during subsequent pregnancies were found in three patients.

Pruritus was constant, followed by tense bullous lesions in 95% of cases, urticaria rash in 75%, and cocaroid disposition in 4 patients. Nikolsky's sign was constantly negative.

Lesions were initially localized in periumbilical region, then generalized to the trunk and the limbs in 95% of cases. The face was affected in 7 cases, and the mucous membranes in one.

Blood count revealed hypereosinophilia in 62%. Histological examination of skin biopsy showed a subepidermal bulla in all cases, a monocytic dermal infiltrate in 85% of cases, lymphocytic in 10%, and eosinophilic in 5%. Direct immunofluorescence showed a positive linear deposition of C3 along the dermal-epidermal junction in 100%, and a deposition of C3 and IgG in four patients.

Treatment was mainly based on systemic corticosteroid therapy at a dose of 0.5 mg/kg/d in 85% of patients and topical corticosteroids alone in 15%. Antihistamines were also usually associated.

Follow-up found recurrences after delivery in three patients. Fetal prognosis was generally good, with one case each of fetal and neonatal deaths. We have also found a case of anamios with a polymalformative syndrome, but this occurrence was probably due to the use of a teratogenic drug during the first trimester.**

Discussion:

PG is a rare autoimmune skin disorder that occurs during pregnancy. Typical lesions of PG consist of severely pruritic papules, urticarial plaques, vesicles, or tense bullae. They usually begin around the umbilicus and spread to the abdomen and thighs. There is no mucosal involvement.

Direct immunofluorescence confirms the diagnosis by showing constant positive linear C and occasionally IgG.

Treatment aims to reduce pruritus and prevent new blisters. Its strategy depends on the severity of the disease.

According to current recommendations, patients with mild symptoms should be treated with potent or very potent topical corticosteroids. In more severe cases, oral corticosteroids are necessary

Fetal prognosis is generally good, but some fetal risks such as hypotrophy and prematurity are possible.

Our results are comparable to published series, with some particular aspects: the predominance of occurrence in multiparous women and an earlier onset and more severe forms in subsequent pregnancies. Moreover, note the relatively high involvement of the face and the absence of the two main fetal risks: prematurity and hypotrophy

Conclusion :

PG is one of the specific pregnancy dermatoses that generally responds to local or oral steroids . It's an added stress factor of pregnancy, hence, a therapeutic approach must take into account psychological consequences

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Abstract N°: 5964**Coexistence of systemic sclerosis and oral lichen planus in a female patient.**

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Coexistence of systemic sclerosis and oral lichen planus in a female patient.**Introduction & Objectives:**

Systemic sclerosis is an immune-mediated rheumatologic disorder with cutaneous manifestations, characterized by fibrosis of the skin, internal organs, and vasculopathy.

Lichen planus is an immune-mediated inflammatory condition that affects mucosa, skin, and appendages.

Our objective is to describe the clinical case of a female patient with systemic sclerosis who subsequently developed oral lichen planus, since this association is rare and there are no reported cases in the literature.

Materials & Methods:

A 58-year-old female, with a family history of type 2 diabetes mellitus, systemic arterial hypertension, dyslipidemia, acute myocardial infarction, leukemia and rheumatoid arthritis.

Patient's personal history included systemic arterial hypertension treated with losartan, type 2 diabetes mellitus treated with dapagliflozin, hypothyroidism treated with levothyroxine, mixed anxiety-depression disorder treated with sertraline and olanzapine, dyslipidemia treated with ezetimibe. She presented 3 deep vein thrombosis episodes, managed with warfarin, enoxaparin, acetylsalicylic acid, and saphenectomy of the right pelvic limb on one occasion. Previous diagnosis of limited systemic sclerosis (CREST syndrome, antinuclear antibodies 1:1000 centromeric pattern) 22 years ago, under treatment with azathioprine and chloroquine.

She started 3 years ago with painful ulcers in the oral cavity and upper lip.

Results:

Examination of the oral cavity revealed whitish patches, affecting both cheeks, gums, and upper lip; distributed in a lace pattern.

A 3 mm punch biopsy of the mucosa of the upper lip was performed. On histology, the lesion reported hyperkeratosis, vacuolization of the basal epithelial layer and lymphocytic inflammatory infiltrate in a band in the dermis, compatible with lichen planus.

Viruses, use of amalgams and medication causes were discarded. She was treated with medium-high potency topical steroids with good response.

Conclusion:

This patient presented two immune-mediated disorders, systemic sclerosis and oral lichen planus. Although each one of these entities are developed in different mechanisms, both involve genetic, environmental and immune

factors. Oral lichen planus causes include viruses, medications, and certain autoimmune disorders.

This case is of great interest since there are no reports to date that describe the coexistence of systemic sclerosis and oral lichen planus. We think that related immune causes could play an important role between these two entities.

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Abstract N°: 5973**A peculiar reticular rash of the extremities: think of reticular erythematous mucinosis**Kenza Tahri Joutei Hassani¹, Zakia Douhi¹, Hanane Bay Bay¹, Sara Elloudi¹, Meryem Soughi¹, Fatima Zahra Mernissi¹¹Hassan II university hospital center**Introduction & Objectives:**

Reticular erythematous mucinosis (REM) syndrome is a condition first described by Steigleder et al. in 1974 as a plaque-like form of cutaneous mucinosis. REM is a rare, chronic and persistent disorder that affects patients of all ages and both sexes, but most cases are middle-aged women. The aetiology of REM remains unknown. REM has been considered by some authors to be an idiopathic, primary form of cutaneous mucinosis, while by others as a disorder closely related to or associated with cutaneous lupus erythematosus, in particular with lupus erythematosus tumidus. The photosensitivity and photodistribution of skin lesions observed in many patients suggest that sun exposure may play a role in the pathogenesis of REM. Clinically, REM syndrome manifests as the persistent erythema in a reticular pattern and/or confluent erythematous papules and plaques that lack scales or other surface changes. Skin lesions are typically located on the central area of the chest or on the upper back but may also spread to the upper abdomen. We report the case of a 30-year-old woman presenting a REM on the extremities.

Materials & Methods:

We report the case of a 30-year-old patient presenting a reticular rash on the extremities diagnoses as REM. The patient was examined by a DermLite 4 dermoscope and the diagnosis was confirmed by histology.

Results:

We report the case of an otherwise healthy 30-year-old woman who presented 5 months prior to consultation a non pruritic erythematous rash of the upper and lower limbs without involvement of the trunk or face, diagnosed as Pityriasis rosea of and put under topical corticosteroids during 1 month without improvement. The patient reported exacerbation of the rash with heat, a photosensitivity without any other systemic signs. She didn't take any form of hormonal contraception. Clinical examination found erythematous macular plaques slightly infiltrated sometimes with reticulated arrangement of the upper and lower limbs. There were no lesions on the face, trunk or mucosis. Dermoscopy showed dotted vessels on a slightly erythematous background with perifollicular distribution in some spots. Histology found a perivascular, and occasionally perifollicular, mononuclear cell infiltrate with increased dermal mucin deposition without epidermal changes confirming the diagnosis of REM.

Conclusion:

Reticular erythematous mucinosis is a rare dermatological condition characterized by distinctive clinical and histopathological features. While its exact etiology and pathogenesis remain elusive, advancements in diagnostic techniques and therapeutic options have contributed to better understanding and management of REM. Increased awareness among healthcare professionals is crucial for early recognition, accurate diagnosis, and appropriate treatment of this uncommon condition.

Abstract N°: 5988**Resistant pemphigoid gestationis : efficacy of plasmapheresis**Maissa Abid¹, Khadija Sellami¹, Kmar Turki¹, Emna Bahloul¹, Mariem Amouri¹, Sonia Boudaya¹, Hamida Turki¹¹Hedi Chaker hospital, Sfax, Department of dermatology, Sfax, Tunisia**Introduction & Objectives:**

Pemphigoid gestationis (PG) is a rare autoimmune blistering disease that occurs in the 2nd or 3rd trimester of pregnancy. Its treatment can be challenging because of concerns over the use of systemic treatment during pregnancy. Also, recurrence is frequent when corticotherapy is discontinued. We report a case of severe resistant form that responded successfully to plasmapheresis.

Materials & Methods:

A 34-year-old primiparous woman with a twin pregnancy developed a generalised bullous rash at the 28th week of gestation. The diagnosis of PG was confirmed by histological and immunological examination. Progression on prednisone at a dose of 1mg/kg/d resulted in remarkable improvement until delivery. A recurrence was noted a few days after delivery, while the patient was on 0.5mg/kg/d of prednisone. Increased doses of corticosteroids and the combination of disulone were ineffective. In view of the generalisation of bullous lesions, 3 sessions of plasmapheresis were carried out at 3-day intervals. The improvement was spectacular with disappearance of bullous lesions and there was no subsequent relapse with progressive discontinuation of corticosteroids in 3 months.

Results:

Pemphigoid gestationis is a unique entity of blistering diseases occurring during pregnancy. Potent topical steroids in moderate forms and systemic corticosteroids in severe forms are the first-line treatments. Complete remission is achieved in 80% of cases. In resistant cases, other therapeutic modalities can be tried, in particular cortisone sparing agents. The most frequently used are IVIGs alone or in combination with systemic corticosteroids. Azathioprine and dapsone may be used secondarily after delivery or abortion. However, cyclosporine can be used during pregnancy.

There were few published cases regarding the efficacy of plasmapheresis as an alternative treatment for recurrent PG.

Conclusion:

Plasmapheresis has only been tried in PG in a few cases in the literature, it could possibly be an excellent alternative in recurrent cases despite several therapies.



Abstract N°: 6006

Does IgG4-related disease impact on metastasis?- a case of disseminated cutaneous metastasis of laryngeal squamous cell carcinoma.

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Introduction & Objectives:

Materials & Methods:

Results:

A 55 year old gentleman presented with a 3 week history of widespread tender subcutaneous nodules associated with lethargy. Physical examination identified numerous tender, smooth mobile nodules located to the left forearm, right flank and bilateral thighs.

This gentleman's complex medical background included; multisystemic IgG4 related disease involving the liver, lung and oesophagus requiring; oesophagectomy and gastric transposition; and medical management with Azathioprine and Prednisolone. Additionally, he had a recent diagnosis of poorly-differentiated laryngeal squamous cell carcinoma treated with debulking surgery and adjuvant radiotherapy.

Deep incisional biopsy of a cutaneous lesion, demonstrated a large infiltrative tumour, extensively replacing the dermis and subcutaneous tissue. Histology suggested a poor-differentiated acantholytic carcinoma, with focal IgG4 staining.

We postulate that the subsequent rapid disseminated metastasis of his laryngeal SCC was secondary to the immunomodulatory effects of his underlying IgG4 related disease. In patients with known IgG4 related disease, there is over expression of PD-1 (programmed cell death 1) and PD-L1 (programmed cell death ligand 1)^{3,4}, which increases the chance of malignancy. Similarly in squamous cell carcinoma⁵, PD-1 and PD-L1 are over expressed facilitating a possible escape mechanism which acute onset of widespread cutaneous metastasis.

Due to role of PD-1, would Cemiplimab⁶ have been an option for treatment in this patient with IgG4 malignancy?

Conclusion:

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Abstract N°: 6015**Hypoxia-inducible factor-1 α (HIF-1 α) as a biomarker to indicate alterations in the microcirculation in patients with systemic sclerosis.**

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Introduction & Objectives:

Systemic sclerosis is an autoimmune disease characterized by tissue fibrosis and microangiopathy. Vascular changes such as a decrease in capillary density diminish blood flow and impair tissue oxygenation. Reliable ways to monitor disease activity and predict disease progression are important in the process of patient selection for clinical trials and to optimize individual patient outcomes. Hypoxia-inducible factor-1 (HIF-1) is a dimeric protein complex that plays an integral role in the body's response to hypoxia. Our study aimed to explore the potential abnormalities in the plasma concentration of hypoxia-inducible factor-1 α and its potential correlation with disease activity and vascular abnormalities in patients diagnosed with systemic sclerosis.

Materials & Methods:

Blood plasma levels of hypoxia-inducible factor-1 α were measured in patients with systemic sclerosis (n = 50) and in healthy individuals (n = 30) using commercially available ELISA test kits.

Results:

The results show a marked increase in hypoxia-inducible factor-1 α levels in patients with systemic sclerosis (3.042 ng/ml [2.295-7.749]) when compared to the control group (1.969 ng/ml [1.531-2.903] p<0.01). Patients with both, diffuse cutaneous SSc (2.803 ng/ml, IQR 2.221-8.799) and limited cutaneous SSc (3.231 ng/ml, IQR 2.566-5.502) displayed an increased serum hypoxia-inducible factor-1 α levels compared to the control group (p<0.01). We found a notable increase in hypoxia-inducible factor-1 α plasma concentration in patients with an "active" pattern (6.625 ng/ml, IQR 2.488-11.480) compared to those with either an "early" pattern (2.739, IQR 2.165-3.282, p<0.05) or a "late" pattern (2.983 ng/ml, IQR 2.229-3.386, p<0.05). Patients with no history of digital ulcers had significantly higher levels of hypoxia-inducible factor-1 α (4.367 ng/ml, IQR 2.488-9.462) compared to patients with either active digital ulcers (2.832 ng/ml, IQR 2.630-3.094, p<0.05) or healed digital ulcers (2.668 ng/ml, IQR - 2.074-2.983, p<0.05).

Conclusion:

Our results indicate that hypoxia-inducible factor-1 α may serve as a biomarker in assessing microcirculatory changes in individuals with systemic sclerosis.

Abstract N°: 6047**Risk factors associated with paraneoplastic dermatomyositis**Elyamani Lamis¹, Zerrouki Nassiba¹, Zizi Nada¹, Dikhaye Siham¹¹Department of Dermatology, Venereology and Allergology, CHU Mohammed VI, Oujda, Dermatology

Introduction & Objectives: Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by an inflammatory infiltrate primarily affecting the skeletal muscle and skin. It can also affect other organs such as lungs and joints [1]. DM is a rare disease, it's more common among females [1] and in low geographical latitude countries [2]. The association between malignancy and dermatomyositis has been widely described and confirmed by numerous epidemiological studies.

The aim of this study is to determine the risk factors associated with paraneoplastic dermatomyositis.

Materials & Methods:

We retrospectively reviewed the case records of all patients diagnosed with dermatomyositis in the Dermatology department of Mohammed VI University Hospital in Oujda city. All patients with a diagnosis of DM fulfilling Bohan and Peter's criteria were included. We defined two groups ; a group of paraneoplastic dermatomyositis and a group of non paraneoplastic dermatomyositis. We compared age,sex,skin necrosis,dysphagia,**erythrocyte sedimentation rate(ESR)**, reactive protein C (CRP) value and anti-TIF-1 gamma antibodies between the 2 groups.

Results:

Twenty seven patients were included, with a mean age of $49,5 \pm 21$ years. Twenty patients were female (74,1%) and seven patients were male (25,9 %). The diagnosis of cancer was retained in 8 patients (29.6%). The mean age of patients with paraneoplastic dermatomyositis was 57.7 years (+/- 16.6). We noted a female predominance with a sex ratio F/H of 1.6. The most frequent cancer in our serie was nasopharyngeal cancer in 37.5%, followed by breast cancer in 25%, larynx, endometrium and stomach cancer in 1 case each . Nasopharyngeal cancer was the most frequent cancer in men, while breast cancer was the most frequent type of cancer in women. The difference was statistically insignificant ($p > 0.05$) between the 2 groups for mean age, sex, skin necrosis, dysphagia, and anti-TIF-1 gamma antibody positivity. In addition, we noted a statistically significant difference ($p < 0.05$) for sedimentation rate and C-reactive protein level. It should be noted that the elevation of ESR and CRP was not explained by any intercurrent infection.

Conclusion: The association of dermatomyositis with cancer is frequent and represents the main cause of mortality. A paraneoplastic screening is systematically performed, especially in the presence of clinical and paraclinical risk factors.



Abstract N°: 6050**A rare presentation of lichen planus pemphigoides**Rym Zmiti¹, Ismahene Souissi¹, Mariem Tabka², Fatima Alaoui³, Ines Chelly³, Maryam Sellami², Mourad Mokni¹¹La Rabta, dermatology, Tunis, ²La Rabta, immunology, mourouj, ³La Rabta, anatomopathology, Tunis**Introduction & Objectives:**

Lichen planus pemphigoides (LPP) is a rare autoimmune blistering muco-cutaneous disease of the pemphigoid family of diseases. It is characterized by the development of vesiculobullous lesions on or adjacent to areas of lichen planus (LP). LPP primarily affects the skin and oral involvement alone is rare. It is hard to clinically differentiate between mucosal LPP and mucous membrane pemphigoid (MMP) with any degree of certainty. We present a rare case of LPP with an exclusive mucosal involvement (LPPM).

Case report:

A 68-year-old man with a personal history of a hypertension treated with captopril50, presented with a history of “blisters and inflammation of the gums and cheeks” and dysphagia. The physical examination revealed a keratotic lesion e left buccal mucosa and erosive lesions on the lateral side of the tongue. Two to six mm ulcers were present in the buccal gingiva. No cutaneous lesions were found. In 2021 we performed a biopsy of his oral mucosa that showed a thickened epithelium with compact orthokeratosis, lymphocytic exocytosis, and some necrotic keratinocytes, a lymphocytic band consistent with LP. The direct immunofluorescence (*DIF*) study showed linear deposition of IgG and C3 and IgA at the epithelium-connective tissue interface. ELISA revealed positive anti-BPAG1/ BPAG2. He was treated by oral corticoids (1mg / kg / day) and dexamethasone rinses. Nonetheless, lesions in the oral cavity kept persisting and, after 1 year, simultaneously, ocular lesions characterized by hyperemia and xerophthalmia appeared. He was seen by an ophthalmologist and a DIF of a biopsy of the conjunctival mucosa was performed and the diagnosis of ocular cicatricial pemphigoid was made. A treatment based on dapsone and corticosteroid was started and the evolution was marked by a worsening of the lesions. One year later, he presented with an ankyloblepharon and keratinization of the ocular surface that might lead to cecity. He underwent an amniotic membrane transplants for fornix reconstruction. A monthly bolus of Cyclophosphamide (600 mg/m² CS) was then prescribed.

Conclusion:

The pathogenesis of LPPM is not well established. There is some hypothesis that states the LPP as a variant of bullous pemphigoid (BP) as they both recognize the antigen BP180. Moreover, it has been suggested that the lichenoid inflammation itself may actually promote the exposure of the antigenic epitopes that induce an autoreactivity which is responsible of the development of an autoimmune response, targeting proteins of the epidermal basement membrane. The LPP with a purely mucous involvement has been described only in 23 and in only 3 patients the conjunctival mucosa was involved. More studies are needed to clarify the underlying pathophysiologic mechanisms when two different and unrelated immunologic disorders follow one another or seemingly occur together.

Abstract N°: 6087**Navigating the Therapeutic Challenges of Chronic Gestational Pemphigoid: A Case Report**

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Introduction & Objectives:

Gestational pemphigoid is a rare autoimmune blistering disease of pregnancy and postpartum period. Pathophysiologically, there is a breakdown of maternal-fetal immune tolerance, leading to the production of autoantibodies directed against placental proteins and cross-reactivity with the dermoepidermal basement junction membrane protein BP180 in the maternal skin.

Materials & Methods:

Case report and review of articles in Medline

Results:

A 38-year-old woman, gravida 2, para 1, at 6 weeks of gestation, presented to our emergency department with pruritic erythematous patches, vesicles, and bullae originating from the periumbilical region and spreading throughout the skin. Skin biopsy revealed a subepidermal blister with eosinophils, and direct immunofluorescence demonstrated a linear deposition of C3 and IgG along the basement membrane, consistent with the diagnosis of gestational pemphigoid. During pregnancy, she was treated with topical and systemic corticosteroids. The baby was born without any skin lesions. There was an improvement of the dermatosis during the initial postpartum period, but 4 months later, there was a recurrence and extension of the disease, with scattered bullae on the breast, back, thighs, and forearms, as well as extensive erosions on the oral mucosa and scalp. There was no association between disease exacerbation and the initiation of hormonal contraception or resumption of menstrual periods. The condition became chronic, with a failure to respond to oral prednisolone treatment cycles or experiencing relapses during tapering over the next three years. The anti-BP180 antibody titer remained elevated. Treatment with azathioprine was ineffective. Treatment with dapsone (100mg/day) was started one year ago, which provided reasonable control of the dermatosis without any documented adverse effects or toxicity to date.

Conclusion:

Chronic gestational pemphigoid is associated with therapeutic refractoriness. Due to the limited number of reported cases in the literature and considering the similar pathophysiology to bullous pemphigoid, an equivalent approach was adopted, which successfully controlled the dermatosis.

Dapsone exerts its anti-inflammatory action by reversibly inhibiting a neutrophil myeloperoxidase that generates a reactive oxygen species from the neutrophil's cytotoxic system, causing tissue damage during inflammation. Neutrophil-derived reactive oxygen species infiltration into the dermis is responsible for blister formation (in a murine model). Currently, this drug has demonstrated efficacy in multiple autoimmune blistering diseases, including pemphigoid group disorders. Furthermore, its use is considered safe during pregnancy and breastfeeding.

This case of chronic gestational pemphigoid highlights the therapeutic challenge associated with this condition. Reporting new cases and successful therapies is important to define the management approach and clarify the prognosis.

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Abstract N°: 6110**A case of bullous pemphigoid induced by Covid-19 infection**

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¹La Rabta Hospital , Dermatology department , Tunisia

Introduction & Objectives:

Bullous pemphigoid (BP) is an autoimmune bullous disease caused by circulating autoantibodies toward the hemidesmosomal antigens BP180 and BP230. In literature, cases of BP during the pandemic were mainly associated with the Covid-19 vaccine and little with the infection itself. Herein, we report and discuss the particularities of a case of BP following a covid-19 infection.

Results:

The patient is a 55 year-old male with no notable medical history , referred to the dermatology department for suspicion of an auto-immune bullous disease.

He had a severe itchy blistering eruption over the trunk, the face and the neck, with tense bullae and crusted erosions. Involvement of the oral mucosa was noted. The patient reported common symptoms including fever, cough, dyspnea, and hyposmia occurring before the eruption. A skin biopsy and direct immunofluorescence test confirmed the diagnosis of BP.

Indirect immunofluorescence test for circulating pemphigoid antibodies found positive AntiPB180 and negative anti PB230.

Covid polymerase chain reaction test confirmed an ongoing SARS-COV2 infection. Screening for internal malignancies found no abnormalities. The patient was treated with systemic steroids (prednisone 0.75mg/kg/day) and moderate potency topical steroids and emollients to relieve itch and dryness. A rapid resolution was observed and a long lasting remission was obtained with 5 mg of prednisone on a 2 year follow up.

Conclusion:

It is known that drugs, injuries, or skin [infections](#) are considered as facilitating factors in genetically predisposed individuals for BP. Many viral infections are suggested to contribute to the outbreak of the disease such as human herpes virus, cytomegalovirus, Epstein-Barr virus, HHV-6 and hepatitis B and C viruses. It is stipulated that these organisms can trigger the onset of BP through molecular mimicry or antigen spreading. SARS-COV2 infections

Treatment for BP is usually needed for several years however prompt discontinuation of the above mentioned environmental agents might result in rapid improvement or even cure of the disease. In our patient, the Covid-triggered BP had some particularities, which are: the young age of onset, mucosal involvement, and rapid response to treatment.

Abstract N°: 6117**Epidermolysis bullosa acquisita mimicking linear IgA bullous disease**

Wejden Fakhfekh¹, Mariem Tabka¹, Fatima Alaoui¹, Ismahene Souissi¹, Maryam Kallel Sellami¹, Mourad Mokni¹

¹Rabta hospital

Introduction & Objectives:

Subepidermal* autoimmune* bullous diseases (SABDs) are characterized by sub epidermal blistering and loss of dermal-epidermal adhesion induced by autoantibody and immune cell infiltrate at the dermal-epidermal junction and upper dermis. Distinction between the different entities can be difficult because of the wide and overlapping clinical spectrums of these diseases. Here, we present a case of an epidermolysis bullosa acquisita (EBA) mimicking linear IgA bullous dermatosis (LABD).

Results:

A 17-year-old male patient with no medical history presented with a 15-days history of bullous pruritic eruption. Skin examination showed numerous vesicles, tense blisters and erosions arising on normal-appearing skin. Most lesions had a herpetiform arrangement. The main involved areas were: the face especially the forehead and the perioral area, the axillae, the groin and the pubis. He had also some lesions on the trunk and the thighs. Oral mucosa was slightly affected on the inner side of the lower lip and the lateral edges of the tongue. Physical examination was otherwise unremarkable. On clinical presentation, LABD was the most likely diagnosis. A skin biopsy was performed. Histopathological examination revealed a sub epidermal blister with a mild inflammatory dermal infiltrate composed of neutrophils and eosinophils. Direct immunofluorescence showed linear deposits of IgG, C3 with high intensity and IgA with medium intensity along the epidermal basement membrane. BP180 and B230 antibodies were negative in ELISA. Indirect immunofluorescence on split skin, anti-basement membrane autoantibodies of the IgG class were detected, bound to the dermal side. The diagnosis of EBA was considered. The patient was initially treated with colchicine 1mg per day. Since he had partial improvement, we prescribed general corticosteroid therapy 0.5mg/kg/day. The progression was favourable with full remission.

Conclusion:

Our case illustrates an autoimmune bullous disease characterized by tense blisters in a herpetiform arrangement on normal-appearing skin associated to a mild mucosal involvement. Anti-basement membrane autoantibodies mainly of the IgG class bound to the dermal side were found. Three diagnoses may be discussed: anti-laminin 332 mucous membrane pemphigoid, anti-p200 pemphigoid and EBA LABD-like. The first diagnosis was unlikely in view of the young age of our patient and the minor involvement of the mucosae. Anti-p200 pemphigoid remains a possible diagnosis, laminin gamma-1 chain antibodies might have been found with immunoblotting. In our patient, a LABD-like EBA was considered. Several subcategories of EBA phenotypes have been described including the LABD-like EBA where patients present with linear IgA deposits in the basement membrane zone on direct immunofluorescence. IgA autoantibody titers by indirect immunofluorescence are usually low and may be absent. In several reports, immunoelectron microscopy was used to confirm the diagnosis of the LABD-like EBA. The IgA autoantibodies appear to react with type VII collagen, which is the major antigen in patients with EBA. In immunoblot, serum-IgA may bind the type VII collagen molecule. Unfortunately, we do not have these techniques to better support the diagnosis.



Abstract N°: 6119**drug induced linear iga bullous dermatosis, mimicking toxic epidermal necrolysis - a case report**Svjetlana Ponorac¹¹University Medical Center Ljubljana, Department of Dermatovenereology , Ljubljana, Slovenia**Introduction:**

Toxic epidermal necrolysis (TEN) is a rare, potentially fatal mucocutaneous drug reaction, occurring 1-3 weeks after starting the drug. It is characterized by widespread epidermal necrosis and associated with significant morbidity and mortality. Early diagnosis and treatment is critical in improving patients' outcomes. More than 100 drugs have been associated with TEN, but antibiotics, allopurinol, non-steroidal anti-inflammatory drugs and anticonvulsants are considered "high risk" drugs.

Linear IgA dermatosis (LAD) is an autoimmune bullous dermatosis, characterized by deposition of IgA autoantibodies along the basement membrane zone of the skin and/or mucosae. Tense vesicles in a characteristic »string of pearls« pattern favour the extensor side of the limbs, trunk and buttocks, but can mimic other bullous diseases, like dermatitis herpetiformis, bullous pemphigoid, pemphigus vulgaris, erythema multiforme and TEN. Drug-induced LAD comprises almost 40% of LAD cases in adults and occurs within 2 days to 4 weeks after introduction of the offending drug. LAD with extensive erosions and positive Nikolsky sign is infrequent and almost exclusively drug induced, with vancomycin being the most frequent culprit drug. Skin biopsy for histopathological and DIF assay is indispensable for the diagnosis. We present a case report of drug induced LAD, mimicking TEN.

Results:

A 58-year old male patient with metastatic melanoma and newly introduced pembrolizumab (eight weeks prior) was admitted to the hospital due to cellulitis in the right axilla, which complicated with septic shock and appearance of extensive mucocutaneous lesions. Before the occurrence of the rash he received a 7 day course of vancomycin, metronidazol and ciprofloxacin. The latter two were stopped just before the rash, but vancomycin was continued and amoxicillin with clavulanic acid was also introduced. Patients' drug history included also several days of amoxicillin with clavulanic acid and clindamycin v just before admission. On examination, erythema, flaccid bullae and epidermal detachment with red oozing dermis were affecting approximately 50% of body surface area (extremities, trunk, flexures, buttocks). Singular tense vesicles were noted on the abdomen and erosions with hemorrhagic crusts on the genital and oral mucosa. TEN was suspected, thus antibiotic treatment was stopped and high dose i.v. methylprednisolone initiated. Skin biopsy was performed and histopathology revealed absent epidermis, dense neutrophilic and lymphohistiocytic infiltrate in the upper dermis, concordant with TEN. Direct immunofluorescence results showed linear IgA deposition along the basement membrane. Indirect immunofluorescence was negative for antibodies against BP180 , BP230, Dsg1 and Dsg3. Diagnosis of drug induced linear IgA dermatosis, mimicking TEN was established, with multiple suspected culprit drugs (vancomycin, amoxicillin with clavulanic acid, metronidazol and ciprofloxacin). Initial improvement of skin lesions was observed after a week of methylprednisolone, however, the patient died due to sepsis and multiorgan failure.

Conclusion:

In clinical scenarios, suggestive of TEN, it is advisable to routinely perform skin biopsy for histopathology and direct immunofluorescence, in order to not miss TEN's many mimickers, including LAD. Establishing a causal drug relationship may be challenging, as patients are frequently exposed to many concurrent medications.

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Abstract N°: 6163**Importance of the diagnosis and management of discoid lupus: A clinical case with atypical distribution.**Veronica Martinez Garcia*¹¹ISSSTE Hospital Regional "Gral. Ignacio Zaragoza", Ciudad de México, Mexico**Introduction & Objectives:**

Systemic lupus erythematosus is a multisystemic disease with one of the widest clinical spectra and with a wider range of presentations. One of the most frequently affected organs is the skin, and among the subtypes, we find discoid lupus. Discoid lupus is an entity that is not as universally recognized clinically as the manifestations of acute or subacute cutaneous lupus, so the reporting of cases and their distribution variations is considered relevant and helps to discern among other differential diagnoses.

Materials & Methods:**Results:**

A 35-year-old woman with a history of autoimmune hemolytic anemia in childhood was evaluated for disseminated dermatosis that began in the anterior thorax and progressively affected the upper limbs, hands, posterior thorax, face, and lower limbs. Lesions are characterized by violet macules, papules, and pustules, some of which were in the crust phase. Painless lesions, not pruritic, no scratch marks. It highlights a condition in the atrial and periocular pavilion, peri buccal, in addition to ulcers in the oral and nasal mucosa, not bleeding. Normal liver, kidney, and thyroid function tests. Immunological tests without alterations. A skin biopsy was performed that reported nonspecific perivascular inflammation. For subsequent rheumatological evaluation, the diagnosis of discoid lupus was determined and management with prednisone was initiated, presenting gradual improvement of lesions and adequate clinical response.

Conclusion:

There is still controversy about the classification of cutaneous lupus subtypes since discoid lupus lesions can sometimes be classified as psoriasiform or polycyclic lesions in up to 32% of cases. The clinical characteristics suggestive are the round shape in the lesions and affection of photo-exposed areas, in addition to the hyperpigmentation and the resulting scar, all the above presents in the case presented. Among the potential diagnostic criteria proposed by Fabbri et al. are those mentioned above and the most frequent anatomical location in the face, atrial pavilion, and scalp is mentioned. The case discussed here presented a widespread distribution of the lesions, involving the photo-exposed areas, but also fewer common areas such as upper and lower limbs, anterior and posterior thorax, and even hands, so your report is considered relevant.

From the case presented here, we can conclude that discoid lupus can present a wide variety of symptoms and distribution of lesions in the skin. Although not as recognized as other forms of cutaneous lupus, its report and study are relevant to differentiate it from other differential diagnoses. The diagnosis of discoid lupus can be challenging due to the diversity of clinical presentations and the possibility of being confused with other skin conditions, such as psoriasis. Case reporting and research in this area are critical to improving the understanding and management of this multisystemic disease.

Abstract N°: 6227

Different faces of Lupus Erythematosus: Case series

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Different faces of Lupus Erythematosus: Case series

Introduction & Objectives:

Lupus erythematosus (LE) is an autoimmune disease with a wide range of clinical manifestations.** We report nine uncommon presentations.

Materials & Methods:

CASE	AGE/SEX	CLINICAL FEATURES	INVESTIGATIONS	DIAGNOSIS	TREATMENT
1	12/M	Fever, dusky macular lesions over face, trunk, upper limbs, oral ulcers since 7 days. HIV disease on antiretroviral therapy (ART) since 07 years.	Antinuclear antibody (ANA): positive Antihistone antibodies: positive Anti ds DNA: positive Skin biopsy suggestive of LE	Drug induced acute cutaneous LE	Tablet Hydroxychloroquine, syrup Prednisolone, revised ART Topical steroids & sunscreen
3	60/F	dusky Erythematous scaly plaques over scalp, face, upper limbs, multiple oral, genital ulcers since 01 month. Scarring alopecia. On antituberculous (AKT) therapy since 03 months for pulmonary kochs	ANA: positive Skin biopsy suggestive of LE	Drug induced subacute lupus erythematosus (SCLE)	Tablet Hydroxychloroquine Prednisolone tablet, revised AKT. Topical steroids & sunscreen
4	28/F	Pulmonary kochs on AKT since 3 months with papulosquamous dusky plaques on trunk, upper & lower limbs, face with oral ulcers since 1 month	ANA: positive Skin biopsy suggestive of LE	Drug induced SCLE	Tablet Hydroxychloroquine, Prednisolone tablet, revised AKT. Topical steroids & sunscreen

5	23/F	Malar rash, targetoid lesions over upper limbs, palms, face, oral ulcers, swelling of ankle joints since 14 days	ANA: positive (speckled pattern); Rheumatoid factor: positive Skin biopsy suggestive of LE	Rowell's syndrome	Tablet Hydroxychloroquine, Prednisolone tablet Topical steroids & sunscreen
6	23/F	6 months of gestation with painful erythematous plaques with targetoid lesions on palms and soles with oral ulcers	ANA: positive, Antiphospholipid antibodies: negative Skin biopsy suggestive of LE	Systemic lupus erythematosus (SLE) in Pregnancy	Topical steroids & sunscreen, Tablet Hydroxychloroquine, Tablet Prednisolone
7	48/F	HIV positive on ART since 4 years with Tender erythematous indurated plaque over left arm lateral aspect since 6 months. Raynaud phenomenon and oral ulcers since 3 years.	Hemoglobin: 2.5g/dl Peripheral smear : anisopoikilocytosis, Spherocytes + RBC agglutination ++ ANA: positive; Anti ds DNA: positive Coombs (direct): positive Skin biopsy suggestive of lupus panniculitis	Lupus panniculitis in a HIV positive patient with autoimmune hemolytic anemia	Tablet Prednisolone, Cap Cyclosporine, Tab Hydroxychloroquine, Topical steroids & sunscreen
8	19/F	Fever, malar rash, photosensitivity, multiple tense vesicles over face, oral mucosal ulcers, myalgia, swelling of ankle joints of	ANA: positive; Anti ds DNA: positive; low complement levels (C3, C4); Skin biopsy suggestive of LE	SLE	Tab Hydroxychloroquine, tablet Prednisolone, Tablet Azathioprine, Topical steroids & sunscreen

		20 days duration			
9	10/F	Recurrent painful ulcerative lesions healing with scarring over distal upper & lower limbs, oral ulcers, since 06 months	ANA: positive Anti ds DNA: positive Anemia, leucopenia low platelet count. low complement levels (C3, C4); Skin biopsy suggestive of Vasculitis	Pediatric SLE	Tab Hydroxychloroquine, tablet Prednisolone, tablet Azathioprine, Topical steroids & sunscreen

Results & Conclusion:

Cutaneous manifestations of lupus erythematosus are varied. Awareness of the common, rare and atypical forms is

crucial for managing such cases.

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Abstract N°: 6234

Graham-Little-Piccardi-Lassueur syndrome: two case reports and dermoscopy findings

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Introduction & Objectives:

Graham-Little-Piccardi-Lassueur syndrome (GLPLS) is a rare variant of lichen planopilaris (LPP). GLPLS is characterized clinically by the triad of progressive cicatricial alopecia of the scalp, non-cicatricial alopecia of the trunk, including axillae and pubis, and a lichenoid follicular eruption. Most frequently LPP appears in postmenopausal females and scarcely in males. Herein we describe the presence of the syndrome in a female with onset in adolescence, as well as in a young male, both cases representing unusual clinical scenarios.

Materials & Methods:

Results:

Case Report 1

A 26-year-old man presented with a clinical history of progressive, multifocal, scarring alopecia of the scalp over the last six years, along with a non-cicatricial alopecia on the arms, axillae, abdomen and pubic hair. Physical examination also revealed confluent lichenoid papules located mainly in the chest and abdominal area. Past medical history of idiopathic thrombocytopenia was reported. Trichoscopic findings in scalp area were highly suggestive of LPP and included perifollicular hyperkeratosis and peripilar casts, white areas representing fibrosis and diffuse erythema with arboriform vessels. Different body parts trichoscopically were characterized by the absence of follicular openings. Histopathology of scalp biopsy revealed degeneration and atrophy of the hair follicles, fibrosis in the papillary dermis and perifollicular lymphocytic infiltration, findings compatible with lichen planopilaris.

Case Report 2

A 42-year-old woman presented with a longstanding history of more than 25 years of progressive cicatricial alopecia of the scalp, along with a pruritic papular eruption on her trunk and upper extremities, and loss of hair in the pubic and axillar area. Clinical examination of the scalp revealed multiple cicatricial alopecic areas with irregular aspect and a follicular hyperkeratotic exanthema in the chest, axillar and inframammary area. Hypotrichosis without skin atrophy was observed in different body parts, especially upper limbs and pubis.** Dermoscopy findings were similar to the case discussed above, with certain additional findings observed on the borders of the affected areas of the scalp and including milky-red areas and follicular plugging, representing signs of disease activity.** Histopathology reported follicular plugging of all hair follicles and perifollicular infiltration from small T4 and T8 cells in the scalp, and features of interface dermatitis in a hyperkeratotic lesion of the trunk.** The patient was treated over the past years with topical and systemic corticosteroids, acitretin, cyclosporine and hydroxychloroquine without experiencing significant improvement.

Conclusion:

PLGLS should be suspected in patients with progressive cicatricial scalp alopecia combined with alopecia in other body regions and follicular hyperkeratotic lesions on the trunk and extremities, irrespectively of age and sex.

Dermoscopy could be extremely helpful in establishing the features of the diagnostic triad characterizing PLGLS, keeping in mind that the dermoscopic findings vary according to the severity, stage of evolution and disease activity.

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Abstract N°: 6250**Dermatomyositis impact on quality of life**Elyamani Lamis¹, Zerrouki Nassiba¹, Zizi Nada¹, Dikhaye Siham¹¹Department of Dermatology, Venereology and Allergology, CHU Mohammed VI, Oujda, Dermatology

Introduction & Objectives: Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by an inflammatory infiltrate primarily affecting the skeletal muscle and skin[1]. Dermatology Life Quality Index (DLQI) and Skindex-29 are validated skin-specific measure of quality of life (QoL) that measure the effect of dermatologic disease on QOL and consists of questions;each of which can be answered by marking choices [2].The aim of our study is to describe the impact of dermatomyositis on patients' quality of life (QOL) according to DLQI and SKINDEX scores.

Materials & Methods: We retrospectively reviewed DLQI and Skindex scores from the case records of all patients diagnosed with dermatomyositis from June , 2014 to June, 2022 in the Dermatology Derpartement of Mohammed VI University Hospital. All patients with a diagnosis of DM fulfilling Bohan and Peter's criteria were included.

Results: Twenty six patients were included, with a mean age of $47,3 \pm 19$ years (range: 12–82 years). Twenty patients were female (76,9 %) and six patients were male (23,1 %). The mean DLQI score of our patients was 14 ± 3 ; reflecting an important effect on quality of life, with extremes ranging from 12 to 22 . The distribution of patients according to the impact on quality of life was as follows: An extremely important impact was found in (7.6%) and an important impact on quality of life was found in (92.4%). For the Skindex score, the average of the Skindex symptoms was $59,9\% \pm 5$, Skindex emotions $53,3\% \pm 16$; Skindex functioning was $55,1\% \pm 17$. The cutaneous manifestations of dermatomyositis can have a significant impact on patients' quality of life. Although the symptoms of myositis are significant contributors to QOL impact of dermatomyositis, they did not impact the DLQI or the Skindex score, which are skin disease specific measures of QOL.The above results indicate that DM has a significant impact on QoL. In our study, the impact on the quality of life of patients based on the DLQI and the SKINDEX was superior to the different series in the literature. In the study of Goreshi [2], the average DLQI of the patients was 7.6 and in the study of Hundley [3] it was estimated at 10.7. For the skindex score , in Goreshi's study [2] ; the Skindex symptoms mean score was 44.9% , the Skindex emotions 50.4%; the Skindex functioning was 28.2% , while in the study of Hundey [3] ; the Skindex symptom mean score was 16.5%+ , the Skindex emotion 32.8%; and the Skindex functioning was 18.4%. The cutaneous manifestations of dermatomyositis have a major impact on patients' lives . This significant impact can be explained by many causes : pruritus , the facial localization of the dermatologic disease wish is an important aesthetic prejudice and the remittent course of the disease; also dermatomyositis' patients may be an underserved in terms of being evaluated by clinicians for psychological well-being.Pruritus has been noted as a frequent clinical feature of dermatomyositis, but its impact and severity has not been thoroughly discussed.

Conclusion: Studies of patients with dermatomyositis have traditionally focused on the impact of the muscle disease, only few studies have studied the impact of the dermatologic manifestations on QOL. The data impressively show a profound impact of the dermatologic aspects of dermatomyositis on quality of life as a result of physical discomfort and impairment along with emotional distress which incites to a psychological care of dermatomyositis' patients.

Abstract N°: 6372**A Rare Case Of Rupioid-type Psoriasis Vulgaris And Systemic Lupus Erythematosus Overlap In A 15-year-old Filipina**Kelsie Kirsty Santos^{*1}, Lily Lyralin Tumulad¹, Arunee Siripunvarapon¹¹East Avenue Medical Center, Dermatology, Quezon City, Philippines**Introduction & Objectives:**

Rupioid-type psoriasis vulgaris and systemic lupus erythematosus belong to a group of immune-mediated inflammatory diseases that involve the activation of different T-cell subtypes. The coexistence of these diseases is very rare, with no reported cases in the Philippines. Due to the lack of relevant studies and the rarity of this overlap, there are no known guidelines for managing it. Consequently, diagnosis and therapeutic challenges arise, especially since treating one of these diseases may worsen the other.

Materials & Methods:

A 15-year-old Filipina presented with a six-month history of onychogryphosis and generalized, pruritic, conical, and hyperkeratotic plaques with adherent scales, accompanied by fever and joint pains. Histopathology results revealed psoriasis and vacuolar interface changes. The lupus panel test and direct immunofluorescence findings were consistent with lupus erythematosus.

Results: **

The patient was started on topical corticosteroids with little improvement. She was then given oral methotrexate (15 mg) for one week. However, six days after taking a total of 15 mg of methotrexate, the patient developed oral ulcerations, abdominal pain, and new lesions on the legs, for which she did not seek consultation immediately. Upon emergency room consultation, ultrasound revealed hepatosplenomegaly with a highly elevated liver profile and kidney function test, pointing to a possible low-dose methotrexate toxicity. The patient subsequently deteriorated after a few hours, which eventually led to her demise.

Conclusion:

This overlap syndrome is an understudied category in comparison to other well-defined diseases. It poses a new challenge in understanding the relationship and devising appropriate treatment strategies to prevent life-threatening complications and/or comorbidities arising from concurrent disease activity. Moreover, this case serves as a constant reminder that health is a multifaceted concept that cannot be confined to a single component. External factors, such as access to healthcare and health education, can also exert a significant influence on the patient's health outcome.



Abstract N°: 6375**Psoriatic like keratinocyte internalization of *Staphylococcus aureus* and *Staphylococcus epidermidis* an in-vitro study.**Daljit Kaur¹, Vinod Sharma¹, Sunil Dogra¹, Muttu Kumaran¹¹PGI HOSPITAL CHANDIGARH, Dermatology, Chandigarh, India**Introduction:**

A widespread chronic inflammatory skin condition known as psoriasis is characterised by epidermal acanthosis, hyperkeratosis, and immune infiltration. Dendritic cells (DC), T cells, mast cells, neutrophils, monocytes, macrophages, and even keratinocytes have been identified as participating in the pathogenesis of psoriasis. Skin microbes, their invasion, colonisation, and interaction with the immune system are important to determine a disease state. Dermatological conditions have been linked to the makeup of the skin microbiota. But the commensal and pathogenic microbes that regulate the localised immune response are not well defined, especially in psoriasis. Hence, this study was designed to check whether keratinocytes have different susceptibilities to hold this microbial balance in psoriasis using an in vitro cell model.

Materials & Methods: *Staphylococcus aureus*, *Staphylococcus epidermidis*, and HaCat cells were used for this study. HaCaT cells were allowed to adhere overnight and then treated with (20uM; Imiquimod) for 24 hours at 37oC. The next day, cells were washed and approximately, 1.08x10⁸cfu/ml of *Staphylococcus aureus* or *Staphylococcus epidermidis* were added; separately. This coculture was maintained for 3hr at 37oC. During this period, the total CFU/ml was counted and compared. The bacterial count was defined as non-adhered, adhered, and internalised. For non-adherent count, the media at different time intervals were plated on agar plates. For the adherent count, the media was removed and the culture plate was vortexed to release the adhered bacteria, then plated on agar plates. While for internalised counts, cells were lysed with triton-x100 and plated on agar plates.

Results: The experiment was split into three sections based on incubation times of one hour, two hours, and three hours. Bacterial density was 1.08x10⁸ cfu/ml prior to incubation. Bacteria that were not attached to the cells after one hour, two hour and three hour were 6.4x10⁷, 5.2x10⁷, and 5.8x10⁷ cfu/ml, respectively. The cfu/ml of adhered bacteria was 6x10⁴, 8x10⁴, 8.02x10⁴ at one, two and three hours, respectively. The count for internalised bacteria after an hour was zero (no colony), 1.3x10⁸cfu/ml after two hours, and 1.4x10⁸ cfu/ml after three hours.

Conclusion:

The present study demonstrates the differential susceptibility of IMQ treated and non-treated HaCaT cells towards *Staphylococcus aureus*, and *Staphylococcus epidermidis*. We are generating more and conclusive data for this pilot study.



Abstract N°: 6533**Efficacy and safety of coacillium in children and adolescents with moderate to severe alopecia areata: a randomized, double-blind, multicentre, phase 2-3 trial**

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Introduction & Objectives:

Alopecia areata (AA) is an autoimmune disease driven by the trafficking of cytotoxic T lymphocytes, from the circulation to peri- and intra-follicular regions, mediated mainly by adhesion molecules expressed by activated endothelial cells (EC). AA is characterized by rapid onset of hair loss, often with chronic relapsing courses due to tissue-resident memory T cells, primed by interaction with activated EC. Two JAK inhibitors were recently approved for AA, representing a major advance. However, being indicated for severe AA in adults and adolescents only, patients with moderate AA, the largest population, and children, remain unserved. Also, early intervention after first manifestation in infancy might possibly prevent disease progression or chronicity. Last, discontinuation of JAK-inhibitors' treatment leads to disease relapse. Coacillium is a botanical drug composed of *Allium cepa*, *Citrus limon*, *Theobroma cacao* and *Paullinia cupana*. Its multiple components have shown to act positively on both hair follicle cycling and EC activation. We investigated coacillium safety and efficacy in children and adolescents with moderate to severe AA.

Materials & Methods:

A randomised, double-blind, multicentre, phase 2-3 trial (RAAINBOW) was conducted at 12 sites in 4 countries. Patients aged 2 to 18 years with Severity of Alopecia Tool (SALT) score of 25-50 (moderate AA) and 50-95 (severe AA) were randomly assigned to coacillium cutaneous solution 22.25% twice-daily (N=71), or placebo (N=36)(2:1). The treatment period of 24 weeks was followed by a treatment-free period of 24 weeks to evaluate disease relapse after treatment discontinuation. No concomitant treatment for AA was allowed. The primary outcome was the relative change in SALT score from baseline to week 24, and patients achieving SALT \leq 20 was evaluated. Protocol is registered with ClinicalTrials.gov, NCT03240627.

Results:

A total of 62 patients met inclusion criteria (coacillium N=42, placebo N=20). Average age was 11 years old, mean time since onset of AA was 3 years, 45% female, 60% had severe AA, 40% moderate AA, 52% experienced their first episode of AA and 48% their second flare or more. After 24 weeks of treatment, the mean change in SALT score in coacillium group was +22.87%, versus -8,00% in placebo group. Treatment effect was 30,87% ($p < 0.0001$). Improvement in quality-of-life endpoints was positively correlated with treatment effect. After treatment discontinuation, SALT score of coacillium-treated subjects continued to improve, from 43.6 to 29.0. At week 48, 47% of coacillium-treated completers reached SALT score \leq 20, versus 9.1% for placebo ($p = 0.0031$). In the coacillium group, no adverse event was serious, all were local, transient, mild or moderate, except one case of severe transient eczema.

Conclusion:

In this phase 2-3 trial involving children and adolescents with moderate to severe AA, coacillium cutaneous solution 22.25% twice-daily was superior to placebo after 24 weeks of treatment, and well tolerated. Most drug-responders experienced sustained improvement during the 6-month treatment-free follow-up period, potentially a paradigm shift in the treatment of AA. To our knowledge, coacillium is among the first drugs to show sustained remission off-treatment in an autoimmune-mediated disease, without immune-altering side-effects. Coacillium might be a suitable treatment option for children and adolescents with moderate to severe alopecia areata.

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