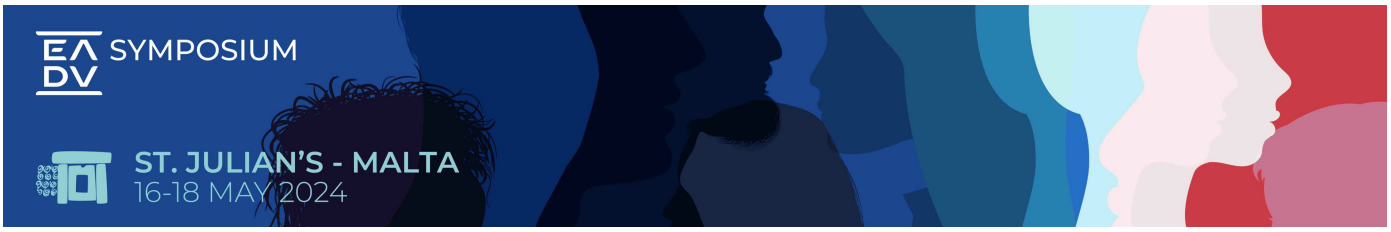

Abstract N°: 112

Radiotherapy- induced Pemphigus Foliaceous treated with Rituximab: a case report

Stephanie Farrugia*[1], David Pisani[1], Daniel Micallef[1], Michael John Boffa[1]

[1]Mater Dei, L-Imsida, Malta

Introduction & Objectives

Pemphigus foliaceus (PF) is a rare autoimmune bullous disease caused by IgG4 subclass antibodies directed against desmoglein-1, which is a vital glycoprotein for intercellular keratinocyte adhesion. Radiotherapy may rarely induce new presentations of PF, termed radiotherapy-induced pemphigus foliaceus (RIPF). Treatment may include corticosteroids, mycophenolate mofetil, cyclophosphamide, azathioprine, methotrexate, ciclosporin, dapsone, intravenous immunoglobulins and plasmapheresis.

Materials & Methods

A 78 year old Caucasian lady presented to the dermatology department with a 1 month history of extensive erythema over the left breast with severe crusting and a few erosions especially at the periphery. This occurred 2 months post radiotherapy treatment for grade 2 invasive left breast carcinoma of no special type. Around the same time, she was also found to have a type A thymoma, diagnosed histologically. Mucosal involvement was absent.

Desmoglein-1 antibody levels were raised and desmoglein-3 antibody levels were normal. Skin biopsy from the left breast revealed epidermal erosions with a sparse superficial dermal perivascular mixed inflammatory infiltrate with neutrophil polymorphs and rare eosinophils. Acantholysis was present focally in the upper epidermis. Immunofluorescence showed intraepidermal deposition of IgG and to a lesser extent C3, between the keratinocytes.

Results

Initial treatment included topical clobetasol propionate 0.05% ointment and a tapering course of oral prednisolone but response was poor. After careful consideration, she was treated with 2 doses of rituximab 1g IV 2 weeks apart. The patient responded very well to this treatment.

Discussion & Conclusion

The mechanism of RIPF is not well defined, however may involve disruption of the sulfhydryl group on the pemphigus antigen, which then leads to an antibody reaction. In addition, radiation-sensitive T-suppressor cells are suppressed, interfering with immune surveillance.

Some key features which help in the recognition of RIPF are: onset in the irradiated area after radiotherapy with possible secondary extension of the PF to other areas of the skin, a latency period of at least several weeks up to one year between radiotherapy and PF onset, and an irradiation dose of 40-70 Gy.

Radiotherapy could also have elicited antigenic properties and caused alterations in localized immunity which led to autoantibody production, which could be related to her underlying thymoma. Autoimmune bullous conditions may be associated with thymomas through a yet unclear mechanism involving T-cell dysregulation.

Rituximab is a monoclonal anti-CD20 antibody depleting B-cells, which offers an effective treatment option for therapy-resistant PF, however the ideal dosage regimen in RIPF is not yet established. Currently, the lymphoma protocol (375 mg/m^2 rituximab weekly for 4 weeks) and the rheumatoid arthritis protocol (2 doses of 1,000 mg rituximab administered 2 weeks apart) are used, with similar response rates. A low dosage regimen has been proposed given the low B-cell burden

in autoimmune diseases.

Through our case we would like to highlight that radiotherapy could potentially induce autoimmune skin conditions such as PF and document the potential response of RIPF to rituximab. The literature suggests that patients should be closely followed up after rituximab treatment as relapse of pemphigus may occur, even years after treatment.

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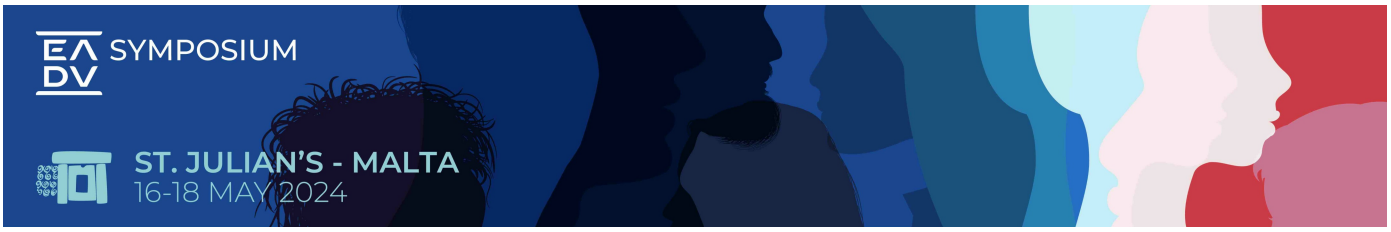


Abstract N°: 198

Serration pattern analysis in the diagnosis of pemphigoid diseases

António Manuel Sequeira Santos*[1], Dario Didona[1], Michael Hertl[1], Bárbara Betonico Berg[1]

[1]Phillips Universität Marburg

Introduction & Objectives:

Autoimmune blistering dermatoses are a diverse group of rare disorders in which patients will present with blisters and erosions of the skin and mucous membranes. These include pemphigoid diseases, which are characterized by antibodies against various basement membrane proteins. Pemphigoid diseases include bullous pemphigoid (BP) and epidermolysis bullosa acquisita (EBA), among others. Because of the inconsistent clinical features resembling other pemphigoid disorders, the differentiation of EBA from the other diseases cannot be based on clinical characters, and sophisticated laboratory techniques, not available to every dermatologist, are needed. Generally, the diagnostic gold standard uses direct immunofluorescence (DIF) of the skin or mucous membranes and is routinely performed in medical laboratories. All pemphigoid diseases are characterized by a linear deposit of immunoglobulins and/or complement elements along the dermoepidermal junction in the DIF. In 2004 the concept of serration pattern was described. In most cases, one can further classify the linear deposits along the basement membrane using the serration pattern and distinguish some of the pemphigoid diseases. This is based on the location of the autoantigens in the basement membrane. The u-serrated pattern is pathognomonic for the pemphigoid disease with autoantibodies against type VII collagen: EBA or bullous systemic lupus erythematosus. In contrast, the n-serrated pattern is found in all other pemphigoid diseases with antibodies against hemidesmosomal components above the lamina densa.

Identifying patients with EBA is important for optimal treatment, to prevent complications and to evaluate prognosis. Studies show that after training, clinicians and pathologists can assess the serration pattern and distinguish patients with EBA from other pemphigoid diseases. We aim to make the differential diagnosis between BP and EBA by identifying the serration pattern using DIF and optimize this process so that it can be used in the routine diagnosis of these diseases.

Materials & Methods:

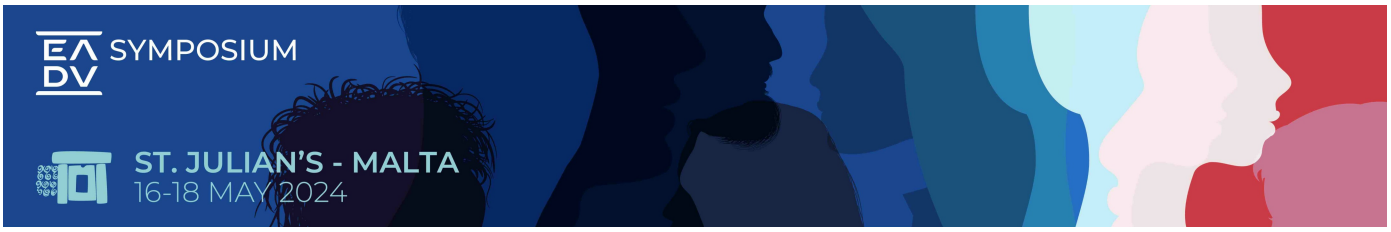
We retrospectively identified and reanalyzed the DIF of patients with pemphigoid diseases. After optimizing our protocol for the preparation of the DIF, we tried to identify the serration pattern for each patient and confirmed the diagnosis. For analysis we took photography of DIF using a Zeiss LSM 700 confocal microscope. Furthermore, we developed a computer software to identify this pattern automatically and compared it with independent observers.

Results:

We were able optimize our protocol do more easily identify the serration pattern in the DIF. Moreover, it is possible to identify this serration patterns using a software.

Conclusion:

Our results show that it is possible to use the serration pattern to further increase our diagnostic capacities of blistering diseases. The use of software to automatically identifiy patterns may diminish the human error that can be associated with image analysis. This was however acoomplisched with a optimized protocol and using a confocal microscope, which is not available in the standard laboratory.



Abstract N°: 224

Refractory Anti-MDA-5 Antibody Negative Ulcerative Dermatomyositis Responsive to Mycophenolate Mofetil

Sumeyye Ozer[1], Banu Farabi*[2], Rebecca Kann[3], Kenneth Shulman[2], Mehmet Fatih Atak[4], Shoshana Marmon[5]

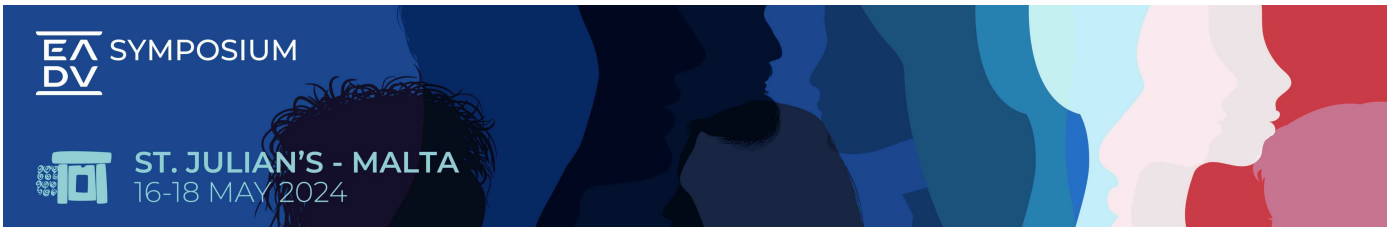
[1]Rao Dermatology, Dermatology, New York, United States, [2]New York Medical College/Metropolitan Hospital Center, Dermatology, New York, United States, [3]New York Medical College, Dermatology, New York, United States, [4]New York Medical College/Metropolitan Hospital Center, Internal Medicine, New York, United States, [5]New York Medical College/South Brooklyn Health, Dermatology, New York, United States

Introduction & Objectives: Dermatomyositis (DM) is an autoimmune disorder characterized by distinct immune profiles and various clinical presentations. Herein, we present a unique case of myopathic DM with unconventional ulcerative lesions.

Materials & Methods: A case of anti-MDA-5 negative dermatomyositis with extensive skin ulcers

Results: A 52-year-old woman with ongoing myopathic DM was referred to dermatology due to worsening draining sores, despite prior treatment with multiple antibiotics, azathioprine, and prednisone therapy. Upon dermatologic examination, we observed Gottron papules, the shawl sign, as well as multiple purulent ulcerative nodules, and one draining abscess. The patient started on doxycycline treatment while other lab tests were pending. Her immune profile tested positive for ANA, anti-Ro52, and anti-beta-2 glycoprotein, but negative for anti-MDA-5 antibodies. A biopsy was done from the edge of the ulcers along with tissue cultures. Cultures grew Staphylococcus hominis, a skin flora bacteria. The histopathological findings were consistent with DM showing epidermal atrophy with mild vacuolar interface change with sparse inflammatory infiltrate, and no acid fast bacili was detected. Despite persistence after doxycycline use, the ulcers responded well to high-dose mycophenolate mofetil along with prednisone therapy. This positive response to immunomodulator therapy supported the autoimmune nature of the ulcerative lesions.

Conclusion: Ulcerative DM is a rare amyopathic variant typically associated with Anti-MDA-5 and rapidly progressive interstitial lung disease (RPILD). It often presents with ulcers on Gottron's papules. Our case stands out due to the atypical locations of the ulcers, the co-occurrence of myopathy, and anti-Ro52 positivity, while the absence of Anti-MDA-5 antibodies and lung involvement is notable. This type of ulceration and infected abscess could potentially result from cutaneous calcification, which is also uncommon in adult DM, although we did not find an evidence for subcutaneous calcifications in biopsy specimens. Previous reports in the literature have highlighted delayed MDA-5 positivity and the coexistence of Anti-Ro52 antibodies and RPILD. Identifying delayed MDA-5 positivity or potential lung involvement through screening could improve prognosis of DM. This report underscores the importance of a comprehensive approach when evaluating ulcerative lesions in dermatomyositis patients.



Abstract N°: 264

Immunobullous diseases with ear involvement: A systematic review

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[1]Keck School of Medicine, University of Southern California, Rick and Tina Caruso Department of Otolaryngology - Head and Neck Surgery, Los Angeles, United States, [2]Keck School of Medicine, University of Southern California, Department of Dermatology, Los Angeles, United States

Introduction & Objectives:

Autoimmune bullous diseases (AIBD) present with various sites of disease manifestation on both the skin and mucosa. Ear, nose and throat (ENT) lesions have been previously documented with ear involvement being only sporadically reported. The ear is rarely assessed in pemphigus and pemphigoid patients and consequently, may lead to an inaccurate evaluation of disease activity and/or severity. We aimed to explore the prevalence of auricular involvement and extend the knowledge about a rarely reported anatomic area.

Materials & Methods:

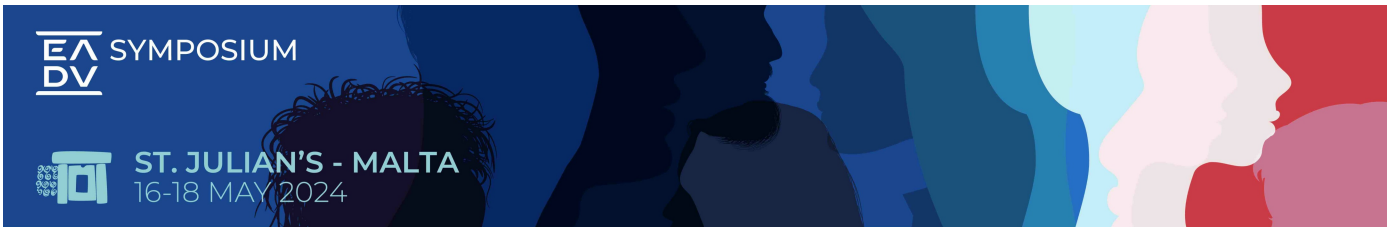
This systematic review was conducted following the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. Literature from the inception of the database until 11 December 2023 was explored using PubMed. Keywords were “pemphigus” or “pemphigoid” combined with “ear”. Inclusion criteria were peer-reviewed, English language articles about AIBD cases with ear involvement. Reviews, guidelines, and basic research studies were excluded. Collected data were checked by a second author, and any disagreement or data inconsistency were resolved by discussion.

Results:

At the end of our selection process, we included 11 papers published between 1971 and 2023. These comprised 5 case reports and 6 case series (n = 175 individuals with AIBDs; 170 [97.1%] pemphigus vulgaris, 3 [1.7%] mucous membrane pemphigoid, and 2 [1.1%] bullous pemphigoid). Overall, 36 out of a total of 175 AIBD study patients (20.6%) presented with ear involvement, with one patient having a localized disease. In the case series, the frequency of ear involvement ranged from 7.5%-26.8%. Overall, the affected auricular areas were the ear mucosa (n=16, 44.4%), external auditory canal (n=8, 22.2%), pinna (auricle) (n=6, 16.7%), tympanic membrane (n=3, 8.3%), Eustachian tube (n=1, 2.8%), and ear lobes (n=1, 2.8%). The types of ear lesions included erosions (n=19, 52.8%), external lesions (n=9, 25.0%), scarring (n=1, 2.8%), blistering (n=1, 2.8%), postero-superior retraction pockets (n=1, 2.8%), cholesteatoma (n=1, 2.8%), inflammation (n=1, 2.8%), erythematous plaques with blood crust (n=1, 2.8%), pigmentation (n=1, 2.8%), and depigmentation (n=1, 2.8%). The symptoms associated with the ear lesions were otalgia (n=24, 66.7%), ear block (n=10, 27.8%), hypoacusis/reduced hearing (n=5, 13.9%), deafness (n=2, 5.6%), discharge/drainage (n=2, 5.6%), and chronic otitis media (n=1, 2.8%).

Conclusion:

Our systematic review showed that over the past 52 years, only 36 AIBD patients with ear involvement have been described in the literature with pemphigus vulgaris being most frequently reported. The most commonly affected auricular areas, lesions, and symptoms were mucosa, erosions, and otalgia, respectively. These findings emphasize the need for ENT examination in patients with AIBD in addition to the routine dermatological examination for improved management.



Abstract N°: 315

McDuffie's hypocomplementemic urticarial vasculitis : case report

Rharib Kholoud[1], Maryem Aboudourib[2], Ouafa Hocar[1], Amal Said[1]

[1]CENTRE HOSPITALO-UNIVERSITAIRE MOHAMMED VI MARRAKECH, DERMATOLOGY VENEROLOGY, MARRAKECH,
[2]CENTRE HOSPITALO-UNIVERSITAIRE MOHAMMED VI MARRAKECH, MARRAKECH

Introduction & Objectives:

Hypocomplementemic urticarial vasculitis (HUV), is a rare systemic vasculitis, of unknown etiology, affecting small vessels. It is characterized by urticarial lesions, hypocomplementemia and variable organ damage, mainly joint and ocular, but also digestive, pulmonary and renal.

Materials & Methods: Case report

We report the case of a 57-year-old femal patient, followed for high blood pressure under treatment, admitted for acute renal failure with chronic urticaria.

The patient was conscious, hemodynamically and respiratory stable, with infiltrated purpuric lesions on the skin such as echymosis and vibices, painless and non-itchy.

Paraclinical examinations revealed negative hepatitis B and C serologies, negative cryoglobunemia, elevated rheumatoid factor, negative anti-DNA and ANA antibodies, with C3, C4 and C1q hypocomplementemia. The level of anti-C1q antibodies was high.

Histological examination and IFD appearance were compatible with leukocytoclastic vasculitis with IgM deposition.The renal biopsy revealed endocapillary glomerulonephritis.

The patient was put on colchicine 1 mg per day with general corticosteroid therapy at a dose of 0.5 mg per kilo. The evolution was marked by a disappearance of the skin lesions with a correction of his renal function. The particuliarities of our case reside in the rarety of the hypocomplementemic urticarial vasculitis. The treatment of McDuffie's hypocomplementemic urticarial vasculitis is not codified.

Results:

Hypocomplementemic urticarial vasculitis is a systemic vasculitis characterized by small vessel involvement, urticarial lesions, hypocomplementemia and variable organ involvement. Renal damage is mainly glomerular. Biology may generally show proteinuria, positive rheumatoid serology, negative antinuclear antibodies and hypocomplementemia. Anti-C1q antibodies are detected in only half of patients, whereas low or collapsed C1q levels seem to represent a more sensitive marker for the diagnosis of VUH. Skin biopsy reveals leukocytoclastic vasculitis. Renal biopsy often suggests glomerulonephritis. However, hydroxychloroquine, colchicine or dapsone seem to have satisfactory effectiveness in the first line.

Conclusion:

Hypocomplementemic McDuffy urticarial vasculitis is a rare multisystem disease that can be mild to fatal. Although the majority of cases occur in the fifth decade, it should be suspected in younger patients who present with vasculitis, low complements, and negative antibodies.

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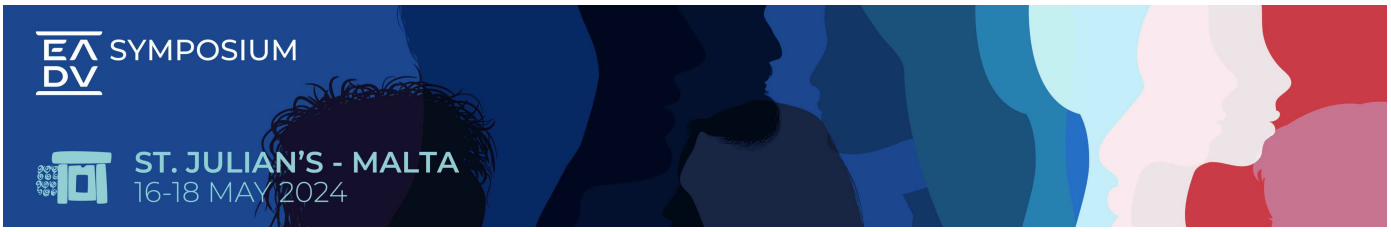


Abstract N°: 344

PDE4DIP is highly mutated in patients with dermatomyositis: a single centric prospective analysis

Zhuangli Tang*[1], Xiaoyong Man[1]

[1]Second Affiliated Hospital, Zhejiang University, Dermatology, Hangzhou, China

Introduction & Objectives:

Dermatomyositis, an auto inflammatory disease which often involves skin and muscles, is relatively rare. Pathogenesis of dermatomyositis is still largely unknown. Currently, liquid biopsy including cell-free DNA is widely used in connective tissues such as systemic lupus erythematosus, etc. Nevertheless, cell-free DNA analysis of dermatomyositis is quite limited. Herein, we aimed to conclude highly mutated cell-free DNA and try to reveal underlying mechanism of dermatomyositis.

Materials & Methods:

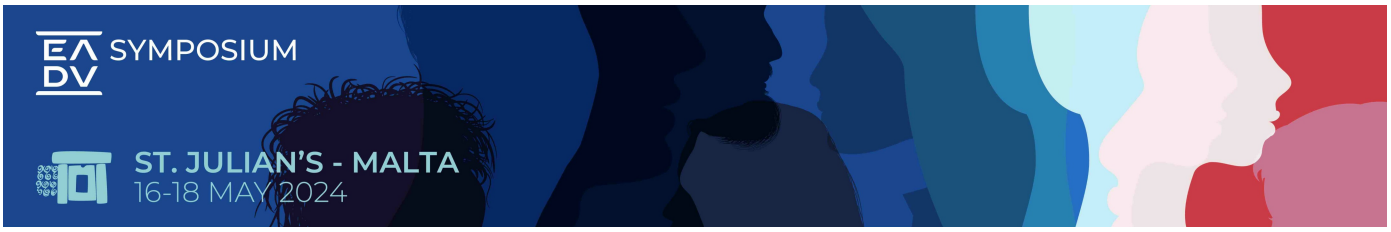
This is a single centric prospective analysis spanning from September, 2022 till December, 2023 and was approved by the ethic committee of our hospital. We prospectively enrolled 17 untreated patients diagnosed as dermatomyositis on the basis of 2017 EULAR/ACR criteria. Peripheral blood samples were collected and subsequent cell-free DNA analysis including concentration, fragment length, mutation categories were conducted. Waterfall plot was measured and the mostly mutated genes were summarized.

Results:

PDE4DIP ranked the mostly mutated gene with a mutation rate of 12/17 followed by*BCLAF1* and *BRCA2*. Among all the identified cell-free DNA, nine genes were found to mutate in dermatomyositis. According to public databases, PDE4DIP mediates RNA degradation and is involved in cAMP/PKA signaling pathway. Currently, PDE4DIP is reported to play a role in malignancies and skeletal muscles diseases. Whether or not and how PDE4DIP joins the pathogenesis of dermatomyositis shall be further clarified.

Conclusion:

Cell-free *PDE4DIP* is highly mutated in patients with dermatomyositis but whether and how it functions in the occurrence and development of dermatomyositis shall be further studied.



Abstract N°: 416

Analysis of the psycho-emotional portrait of patients with lichen sclerosus of the anogenital area.

Maria Proskurina[1], Nadezhda Chernova[2], Olga Zhukova[3], Irina Zadorozhnaya[1]

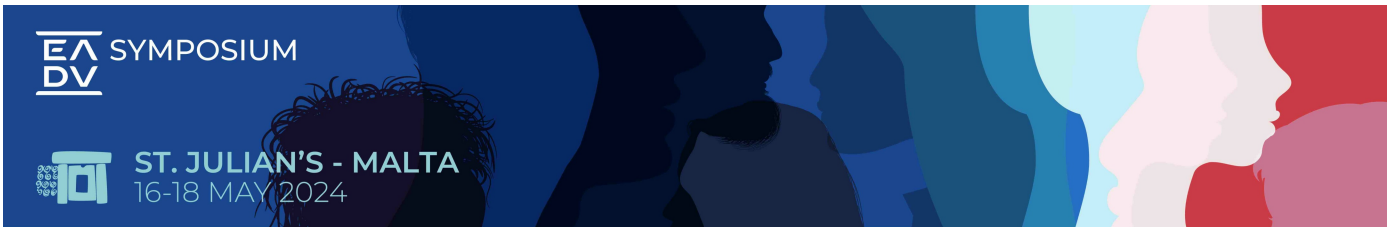
[1]Moscow Scientific and Practical Center of Dermatovenereology and Cosmetology, Department of Dermatology and Venereology, Moscow, Russian Federation, [2]Moscow Scientific and Practical Center of Dermatovenereology and Cosmetology, Head of the scientific department of anogenital dermatosis and infections, Moscow, Russian Federation, [3]Moscow Scientific and Practical Center of Dermatovenereology and Cosmetology, Chief doctor, Moscow, Russian Federation

Introduction & Objectives: Lichen sclerosus (LS) of the vulva greatly affects the psycho-emotional and sexual health of women. Recent research suggests that women with LS have an increased incidence of dyspareunia and anxiety about loss of relationships, and the desire to restore experiences of intimacy and sexual pleasure is the main motivation for surgical interventions for their condition. The purpose of our study is determine the degree of influence of the disease lichen sclerosus of the anogenital area on the psycho-emotional state of patients and on their sexual function.

Materials & Methods: Outpatient records of 45 patients, assessment of anamnestic data, clinical manifestations, determination of quality of life on the dermatological quality of life index (DIQL) scale; determination of the level of anxiety and depression using the Hospital Anxiety and Depression Scale (HADS); assessment of sexual activity using The Female Sexual Function Index (FSFI) scale.

Results: We have examined and interviewed 45 women with lichen sclerosis of the anogenital region and a control group of 45 healthy women from May 2022 to October 2023 years. The average age of the patients ranged from 21 to 76 years (mean 54 ± 7 years). At the same time, 16 (36%) women were of reproductive age (18-45 years), 9 (20%) people were in the menopause period (46-55 years), and 20 (44%) people were in the postmenopausal period (after 56 years). More than half of the surveyed women with LS (25 people) complained of sexual dysfunction, and 35 people complained of sleep disturbances. With a more detailed study of the anamnesis using the determination of quality of life on the Dermatological Quality of Life Index (DIQL) scale, we determined that the average value was 6.86 ± 2.87 and in the group of healthy women it was 1.86 ± 2, which indicates a moderate impact of the disease on the patient's life. Determining the level of anxiety and depression using the Hospital Anxiety and Depression Scale (HADS) revealed that before treatment, anxiety was rated by patients at 6.86 ± 2.74, which is not defined as subclinical anxiety; depression 6.2± 1, which is not regarded as subclinical depression. But when conducting a survey, we found out that women with LS had the following data: subclinical severe anxiety - 12 people, clinically significant anxiety – 7 people, subclinical severe depression – 13 people, clinically significant depression – 4 people. Assessing sexual activity using The Female Sexual Function Index (FSFI) scale allows you to determine the main components of women's sexual function: sexual desire, sensitivity and excitability, lubrication (vaginal moistening), orgasmicity, satisfaction with sexual life, coital and/or post-coital discomfort/pain. There is no quantitative assessment of the test results; the maximum positive number of points when answering each question in the questionnaire is considered optimal. The maximum score for each indicator is -6, the minimum is 0. During the study, we obtained the following data: IHF in women with LS: desire 2.43 ± 1.36, arousal 1.99 ± 1.86, lubrication 2.06 ± 2 .14, orgasm 2.09 ± 2.19, satisfaction 2.77 ± 1.8, pain 1.91 ± 1.97.

Conclusion: Lichen sclerosus disease has a negative impact on the self-esteem of patients. Studies also report decreased sexual activity, less satisfaction with sexual activity, depression, and decreased quality of life. The prevalence of sexual dysfunction among patients with LS is 55%.



Abstract N°: 428

An unexpected neoplasm in a malignancy-related disease

Ana Simón Gozalbo*[1], Inmaculada Balaguer Franch[1], María Gamo Guerrero[1], Emilio Del Río Pena[1], Ainhoa Roca Martiartu[1], Pablo De la Cueva Dobao[1], Raquel Carrascosa de Lome[1]

[1]Hospital Universitario Infanta Leonor, Dermatology, Madrid, Spain

Introduction & Objectives:

Dermatomyositis (DM) is an idiopathic inflammatory myopathy that encompasses skin symptoms, skeletal muscle involvement and a broad spectrum of autoantibodies. It can associate systemic manifestations and neoplasms. Up to 18% of patients lack traditional myopathy which defines a subtype called amyopathic DM (ADM).

Materials & Methods:

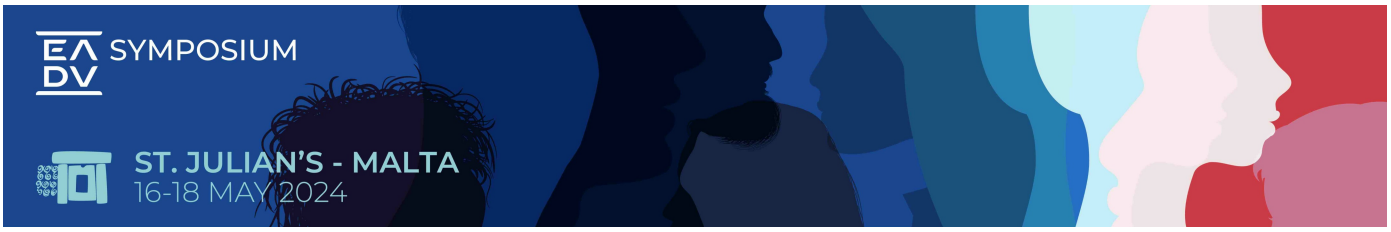
We present the case of an 88-year-old woman diagnosed with ADM who was monitored for 10 years in our Dermatology department. Malignancy screening tests were repeatedly negative, and skin lesions resolved with oral azathioprine. 15 years after DM diagnosis, she returned to our clinical setting showing erythematous plaques over the dorsum of the hands, periungual erythema and dystrophic cuticles. The examination also revealed poikiloderma on both hips and erythematous, scaly plaques on the scalp. No muscle weakness was determined.

Results:

Treatment with oral prednisone was started. The blood test showed a significant increase in CA 19.9 of 11207 U/ml (normal < 37) and the anti-small ubiquitin-like modifier-1 activating enzyme (antiSAE-1) antibodies were also positive. A thoracic abdominopelvic scan was performed displaying a hypodense lesion in the pancreatic head that was blocking the common bile duct with focal hepatic lesions. These findings were strongly suggesting primary neoplasia with metastatic liver disease. She was finally diagnosed of stage IV pancreatic adenocarcinoma. Palliative treatment was applied and the patient finally died.

Conclusion:

Patients with ADM seem to be at risk of developing internal malignancy. SAE antibodies are only seen in an 8% of patients with DM and they are related to severe cutaneous disease with minimal myopathy. Their association with malignancy is still unknown. The risk to develop neoplasia occurs during the first year after DM diagnosis, being highly unlikely beyond 5 years. We present a patient with ADM, late development of neoplasia and SAE-1 antibodies. In ADM, where we can only rely on dermatological findings, we must always be alert to new clinical findings, even if they are far from diagnosis, and search for the tumor again.



Abstract N°: 467

Bullous Pemphigoid Associated with Influenza Vaccination: A Case Report and Systematic Review of Literature

Nikolaos Chaitidis[1], Aikaterini Tsentemeidou[1], Georgios Mantios[1], Aikaterini Papadopoulou[1], Dimitra Kiritsi[1], Efstratios Vakirlis[1], Eleni Sotiriou[1]

[1]School of Medicine, Aristotle University of Thessaloniki, First Department of Dermatology and Venereology

Bullous pemphigoid associated with influenza vaccination: a case report and systematic review of literature

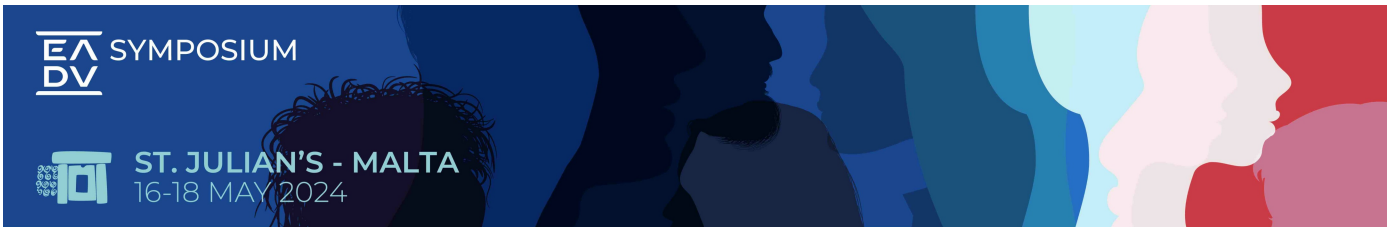
ABSTRACT

Introduction & Objectives: Blistering skin diseases associated with influenza vaccination are uncommon, with only a limited number of cases reported to date. A notable proportion of these cases involved bullous pemphigoid. Our objective was to report a case of bullous pemphigoid in an octogenarian temporally associated with influenza vaccination and to conduct a systematic literature review on cases of blistering skin diseases linked to influenza vaccination.

Materials & Methods: We conducted a literature search for studies reporting cases of de novo vesicular, bullous, or pustular eruptions temporally associated with influenza vaccination. Descriptive statistics were employed to present the pertinent data.

Results: Our patient was treated with oral prednisolone at an initial dose of 0.3 mg/kg/day with a tapering regimen, reaching a maintenance dose of 0.1 mg/kg/day in approximately one month. The patient remained lesion-free while on the maintenance dose at the three-month follow-up. Our review revealed that the most common blistering disease associated with influenza vaccination was bullous pemphigoid, and it responded well to treatment. Other common diseases included bullous vasculitis and neutrophilic dermatoses, which presented more treatment challenges. Most patients were elderly, with a male predominance and a latency period ranging from hours up to five weeks. There were no reported cases of mortality.

Conclusion: Blistering skin diseases temporally associated with influenza vaccination typically exhibit a favorable response to treatment.



Abstract N°: 555

Financial implications and healthcare utilisation in Cutaneous Lupus Erythematosus in Ireland

Claudine Howard-James*[1], Bairbre Wynne[2]

[1]St James's Hospital, Clinical Research Facility, Dublin, Ireland, [2]St James's Hospital, Dermatology Department, Dublin, Ireland

Introduction & Objectives: Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disorder, European prevalence approximately 40 per 100,000 (1). Cutaneous lupus erythematosus (CLE) is the most common manifestation of the disorder, appearing in 72-85% of patients with SLE (2).

This study plans to analyse healthcare attendance patterns and patient costs, direct and indirect, associated with CLE. Direct costs include prescription items. Indirect/out-of-pocket costs include sunscreen and hairpieces. This study will also assess disease impact on quality-of-life (QOL).

Materials & Methods: Patients with CLE diagnosis attending combined Dermatology/Rheumatology/Nephrology Lupus OPD at our public tertiary centre were invited to participate. Ethical approval was secured and informed consent obtained. Participants completed an anonymous paper survey, analysed to identify costs and healthcare utilisation patterns associated with CLE as well as demographics and QOL impact.

Results: A total of 25 patients completed the survey; median age was 53 and 92% (23 patients) were female. Median age at diagnosis was 37 and first presentation of lupus was cutaneous in 20/25. Self-reported overall disease classification was severe in six patients and self-reported disease activity in the past three months was high/flaring in seven. Healthcare utilisation was assessed in the categories of Emergency Department (ED), General Practitioner (GP) and Outpatient Department (OPD) attendance. All participants were recruited at combined Lupus OPD, and 23/25 endorsed attendance at another OPD appointment in the past year. Of this cohort, 13 patients attended their GP in the past year with a mean of 5.3 visits, and two reported 15-20 GP visits. Three patients attended ED for lupus-associated presentations. In the past three months, despite 23/25 participants holding medical cards which contribute towards prescription costs, nine patients spent >€200 on items prescribed for lupus. We found that those with severe disease had higher burden of prescription costs. In the non-prescription category; the most common expenditure was sunscreen, and eight patients endorsed costs >€100. Five participants reported treatment access was limited by expense. Quality-of-life impact (QOLI) was assessed by basic multiple-choice question, in which 23 patients reported some QOLI, and by Dermatology Life Quality Index (DLQI), with scores ranging from 0-30.

Conclusion: Cutaneous Lupus Erythematosus (CLE), the most common manifestation of SLE, has a significant financial impact in addition to the biopsychosocial burden. Expenditure was explored by direct prescription expenses and indirect costs such as sunscreen. Further research is needed to optimise financial and quality-of-life implications in acute flares and chronic CLE management.

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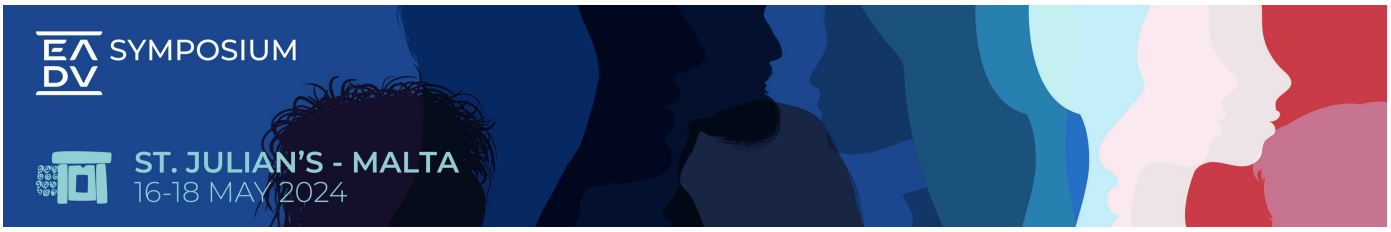

Abstract N°: 676

Flagellate erythema in idiopathic dermatomyositis

Jeewoo Choi[1], Jin Ju Lee[1], Rosa Kim[1], Heera Lee[1], Ji Yeon Byun[1], You Won Choi[1], Joo Young Roh[1], Hae Young Choi[1]

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Introduction & Objectives:

Dermatomyositis is an inflammatory myopathy of unknown etiology that is challenging to diagnose due to its clinical heterogeneity. In addition to the pathognomonic findings such as Gottron's papules, Gottron's sign, and heliotrope rash, it can show up in a various skin symptoms. However, a flagellate erythema which seems to be related to disease activity is only found in less than 5% of these patients.

Materials & Methods:

Results:

Case report

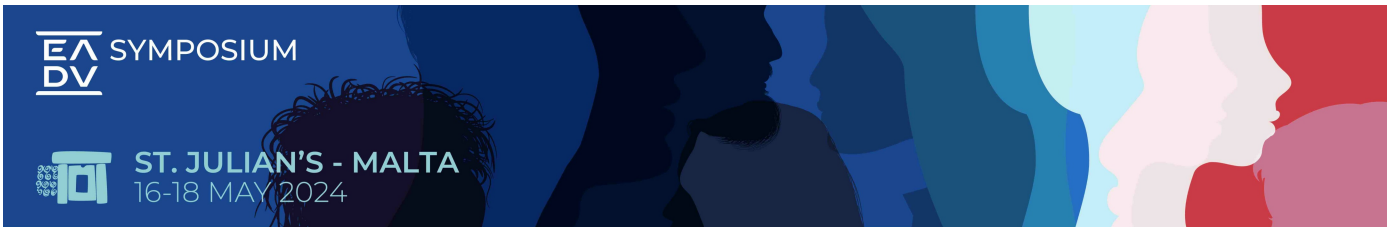
We report a case of a 45-year-old Korean man who was treated with tenofovir for hepatitis B and liver cirrhosis. He came to our department with severely pruritic red to purple maculopatches and acute proximal muscular weakness started a month ago. He reported owning a cat and using vegetarian cat litter, but denied consumption of mushrooms or use of bleomycin. Multiple erythematous streaks were observed extending widely from the confluent patch on the buttocks. Persistent pruritic flagellated streaks spreading from the Shawl sign, V sign, and Holster sign are observed. In addition to heliotrope rash, violaceous periorbital edema and gottron papules, erosive or crusted lesions were also present on the upper back.

In the laboratory studies, ferritin, creatine kinase (CK), lactate dehydrogenase (LDH), aspartate aminotransferase (AST), alanine aminotransferase (ALT), and aldolase were elevated, Fluorescent antinuclear antibody test (FANA) was weakly positive and Anti Jo-1 antibody was negative. Muscle biopsy showed muscle fiber necrosis with regeneration and minimal inflammation. Skin biopsy showed interface dermatitis and superficial dermal perivascular lymphoplasmacytic infiltration. Additional tests, such as computed tomography and ultrasound, were performed, but they found no indications of a related neoplasm or calcification. The patient was treated with systemic corticosteroids, azathioprine and methotrexate 15mg/week. After 3 months of treatment, his skin and muscular symptoms resolved, and 2 years and 6 months later, the patient is still being treated with azathioprine and methotrexate for maintenance.

Conclusion:

Flagellate erythema is a distinctive streaky rash that resembles whip marks. It was first identified as a side effect of bleomycin, but can also be associated with the consumption of undercooked shiitake mushrooms, and less commonly, with rheumatologic diseases and various types of chemotherapy. The exact mechanism is unknown, but it is thought to be related to physical injuries such as minor trauma or sun exposure. Histopathology findings are nonspecific, and may include basal cell vacuolization, papillary dermal edema, and superficial dermal lymphocytic infiltration. Direct immunofluorescence results are controversial. Currently, the association with malignancies or pulmonary fibrosis is unclear. However, some authors have reported an association with pulmonary complications or paraneoplastic etiologies. Conventional therapy for dermatomyositis, such as corticosteroids, has been shown to be effective.

Here, we provide flagellate erythema, a rare manifestation with a distinctive skin morphology that contributes to clinical features of dermatomyositis.



Abstract N°: 716

Energy-based devices for the treatment of cutaneous lesions in patients with lupus erythematosus and dermatomyositis

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[1]Hanyang University College of medicine, Dermatology, Seoul, Korea, Rep. of South

Introduction & Objectives:

The use of energy-based devices (EBDs) for the treatment of various aesthetic and dermatological conditions is increasing. However, data regarding its efficacy and safety in patients with autoimmune connective tissue disease (CTD) are limited. This study aimed to assess the outcomes of EBD treatment in Korean patients with CTD, specifically lupus erythematosus (LE) and dermatomyositis (DM), and review its efficacy and safety.

Materials & Methods:

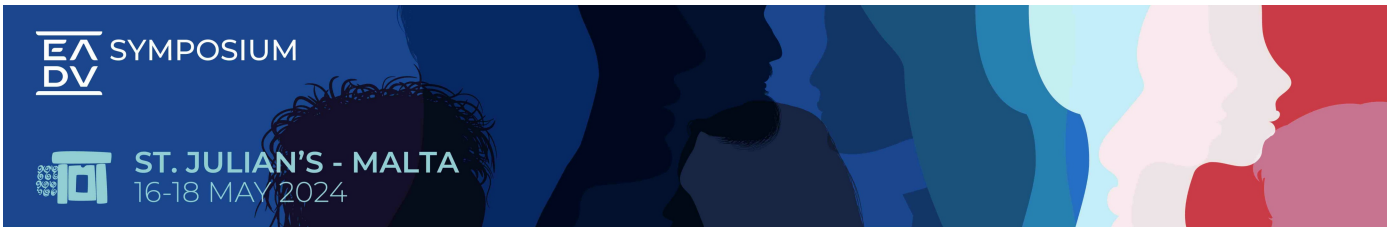
We conducted a retrospective study involving 26 patients with CTD, including those with LE and DM, who were treated at out hospital between December 2015 and November 2023. The use of various laser modalities was assessed, including pulsed-dye laser (PDL), intense pulsed light (IPL), long-pulse Nd:YAG (LPNY), Q-switched Nd:YAG (QSNY), and ablative fractional laser (AFL). Outcomes were evaluated by two independent dermatologists using a 5-point modified Investigator's Global Assessment Scale, which encompasses parameters such as erythema, dyspigmentation, and other morphological characteristics. Adverse events and patient cosmetic satisfaction were recorded using a visual analog scale.

Results:

Among twenty-six patients, 20 patients with LE and 6 patients with DM were treated using various EBD modalities. The treatments were well-tolerated, and significant improvements were observed in erythema, dyspigmentation, and overall skin morphology in patients with LE and those with DM. Some patients with LE also show improved follicular activity. A few transient side effects were reported; however, none of the patients exhibited long-lasting side effects or disease reactivation during follow-up. Overall, the cosmetic outcomes were satisfactory, with higher ratings reported in patients with LE than in patients with DM.

Conclusion:

EBD treatment, including laser therapy, can yield favorable cosmetic results for patients with LE and DM. These treatments are generally safe and well-tolerated. Further larger scale controlled studies are warranted to validate these findings and establish optimal treatment protocols.



Abstract N°: 862

Atypical lupus tumidus-like presentation of VEXAS syndrome

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Introduction & Objectives:

VEXAS syndrome is an autoinflammatory disorder described in recent years in the literature. The acronym stands for Vacuoles, E1 enzyme, X-linked, Autoinflammatory and Somatic. This syndrome is characterised by recurrent fevers, Sweet syndrome-like skin manifestations, lung involvement, polychondritis and hematological disorders. We report a case of VEXAS syndrome presenting as a lupus tumidus-like eruption.

Materials & Methods:

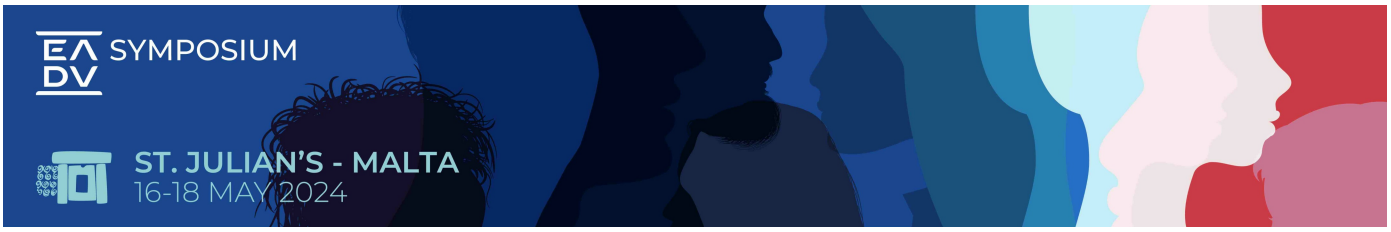
An 80-year-old Caucasian male was referred for evaluation of a 3-month history of recurrent skin lesions. The patient associated the eruption to newly prescribed sildolosin by urologists. Physical examination showed multiple erythematous nummular and annular plaques with infiltrated borders and central clearing on the trunk, limbs and face. He had no fever or systemic symptoms, but referred moderate pruritus. Laboratory findings were unremarkable. Repeated biopsies revealed a perivascular lymphohistiocytic infiltrate with mild dermal mucin deposits and with negative direct immunofluorescence, consistent with cutaneous lupus tumidus. Skin lesions responded very well to systemic and topical corticosteroids, but flare control was resistant to hydroxychloroquine, methotrexate, mepacrine, dapsone and combinations of the aforementioned. The patient then developed an anemia, which was initially attributed to dapsone, showing no additional symptoms or findings.

Results:

After discussion with dermatology colleagues and a literature search, VEXAS syndrome was included in the differential diagnosis; a peripheral blood smear showed vacuoles in neutrophils and genetic testing revealed a mutation in the UBA1 gene consistent with VEXAS syndrome.

Conclusion:

A wide range of cutaneous presentations have been reported in VEXAS syndrome, the majority being Sweet syndrome-like eruptions, cutaneous vasculitis and periorbital angioedema among others. We report an unusual presentation as a lupus tumidus-like eruption, with no other initial signs in other organs suggestive of VEXAS syndrome. As dermatologists, we should be aware of the broad variety of skin manifestations that can present in this novel and yet somewhat unknown condition.



Abstract N°: 869

Digital necrosis: a rare complication in systemic lupus

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Digital necrosis a rare complication in systemic lupus

Introduction & Objectives:

The onset of systemic lupus erythematous after 50 years is a rare event. The symptomatology is often atypical with a rather benign evolution. Digital gangrene in patients with systemic lupus is infrequent, reported mainly in subjects of average age with a long duration of their disease. We report the observation of a 63-year-old patient with an SLE whose initial manifestation was digital necrosis

Materials & Methods:

63-year-old woman, without profession, admitted for cyanosis of the fingers. This patient had a history of type 2 diabetes. The clinical history of the patient dates back to four days before admission, by the sudden installation of a cyanosis of the extremities of the upper limbs without phenomenon of Raynaud nor any other sign. The clinical examination found: a coldness with cyanosis of the fingers without trophic disorders or sclerodactyly or pulp ulceration. An arterial doppler of the upper limbs was performed urgently but the results were not very contributory with an arterial vasospasm without thrombosis or parietal abnormality. Capillaroscopy was normal. The hemogram showed anemia at 9.3. The rate of sedimentation was accelerated to 75 mm L. There was hypoalbuminaemia and hypoprotidemia with 24 h proteinuria at 0.60 g / 24 h.. Antinuclear, anti-DNA and anti-SSA antibodies were positive at levels of 1/1280, 28 and 64 respectively. Anti-cardiolipin, anti-β 2-glycoprotein antibodies, lupus anticoagulant, anti-SSB, anti- Sm, anti-RNP, anti-topoisomerase I and anticentromer antibodies as well as neutrophil antibodies were negative. The diagnosis of lupus disease was made. A bolus of methylprednisolone with relay 1 mg / kg / d of prednisone equivalent was initiated. The treatment also included calcium channel blockers and anti-coagulant therapy. In the absence of clinical improvement, the use of other vasodilator treatments was necessary based on a prostacyclin analogue. The evolution was unfavorable with rapid installation of extensive digital gangrene. The patient was placed under an immunosuppressant type cyclophosphamide 0.7 g / m 2, and the patient is deceased

Results:

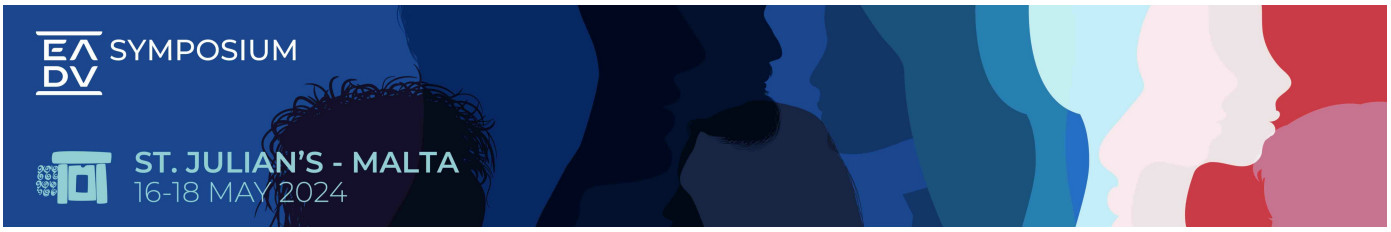
SLE remains a rare autoimmune disease in the subject age. Its diagnosis is based on a combination of clinical signs And immunological tests, according to the ACR classification Patients with late onset SLE have More insidious, with a delay in diagnosis, and a Decrease of the female preponderance, as reported Pu et al. A rare series of 6 lupus patients Of late onset after 75 years has been reported in the Literature.

The 6 patients in this series were Female, with an average age at diagnosis of 78 years. Skin lesions and haematological disturbances predominated To the diagnosis. Corticosteroids and hydroxychloroquine Were the most widely used treatments, but immunosuppressants Were used for 3 patients of this series.

Mortality is more Frequent in elderly patients with a pathology Lupus of late revelation, with a mean survival to 10 years 71% compared with 95% in young patients, according to The Boddaert et al. Septic shocks wereThe most frequent causes of death. In the very A multidisciplinary approach is recommended.

Conclusion: The digital gangrene is an exceptional complication of the LES with late declaration. His knowledge is

essential in order to institute an early and aggressive treatment with corticosteroids and immunosuppressants to prevent amputation.



Abstract N°: 924

Ultrasonographic findings of morphea lesions in comparison with normal appearing skin

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Introduction & Objectives:

Morphea is an inflammatory skin disorder with different manifestations from isolated superficial plaques to multiple lesions with or without deeper involvement. Morphea is characterized by thickening of the dermis, subcutaneous tissue, or both, caused by excessive collagen deposition. Ultrasonography has been shown to be useful in monitoring skin changes in therapeutic trials. In order to evaluate the role of ultrasonography in evaluating morphea, we compared histologically confirmed morphea lesions' ultrasonographic imaging data with symmetrical uninvolved skin.

Materials & Methods:

All patients with histologically confirmed primary cutaneous morphea who referred to our dermatology clinic in a period of one year entered the study. Sonographic evaluations were performed on the lesions and the symmetrical uninvolved sides. Dermal and epidermal thickness was measured, and dermal and epidermal echogenicities were recorded, too. Statistical analysis of group differences was performed by using the Paired Sample T-Test. A P-value of less than 0.05 was considered statistically significant.

Results:

Twenty seven patients with morphea lesions met the requirements for inclusion in the study. Statistical analysis showed that only epidermal density of the lesion (Mean: 156.68, SD: 27.00) was significantly higher compared with the nonlesional skin (Mean: 148.91, SD: 27.86) (P-value=0.042). Epidermal thickness in the morphea lesion (Mean: 126.75, SD: 34.24) was lower compared with the nonlesional normal skin (Mean: 133.87, SD: 45.01), but the difference was not significant (P-value=0.277). Dermal thickness in the morphea lesion (Mean: 1107.97, SD: 414.37) was higher compared with the nonlesional normal skin (Mean: 1094.65, SD: 331.06), the difference, however, was not significant (P-value=0.869). The echo-density of dermis in the morphea lesion (Mean: 49.13, SD: 18.97) was lower compared with the nonlesional normal skin (Mean: 52.22, SD: 25.33), but the difference was not significant (P value=0.325) (Figure 1).

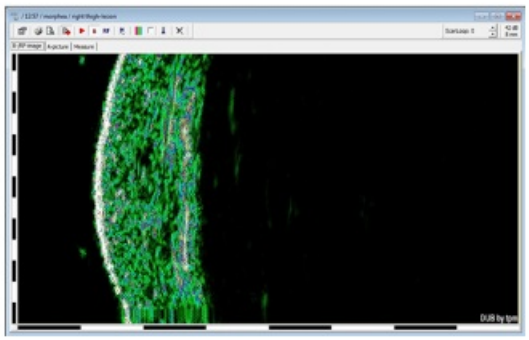
Conclusion:

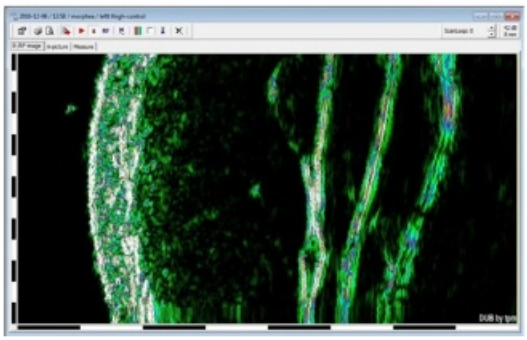
Ultrasound sonography can detect important differences between active localized scleroderma lesions and normal tissue, and may provide valuable information in assessment of the morphea disease. Greater sample size could demonstrate all significant changes.

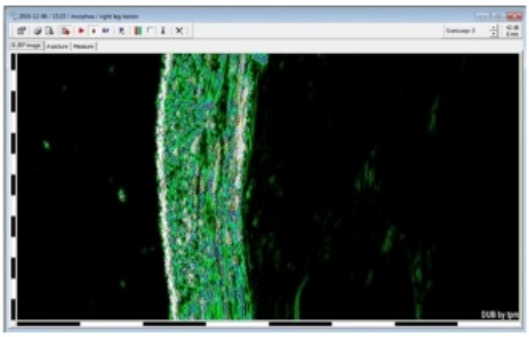
Figure 1. Two examples for comparison of ultrasound imaging in the lesions and uninvolved skin

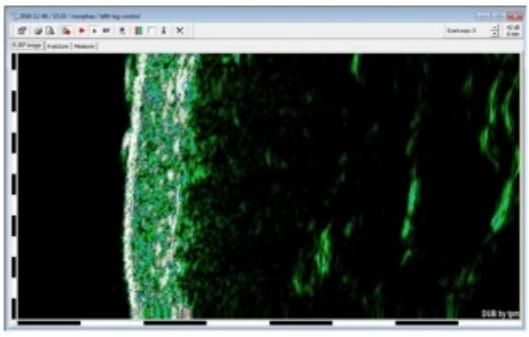
Lesion

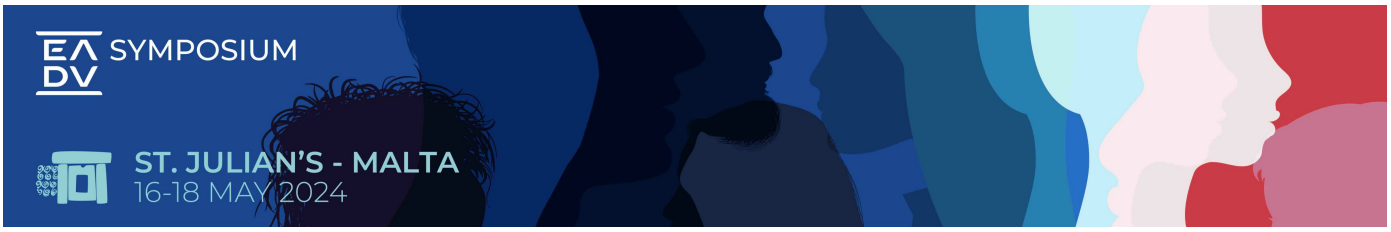
Control











Abstract N°: 926

Analysis of the detection of autoantibodies to native DNA class G in blood serum in patients with vitiligo.

Ixlos Shukurov[1], Shakhnoza Mavlyanova[2]

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Introduction & Objectives: According to research, the pathogenesis of vitiligo is an autoimmune process, accompanied by the appearance of depigmented lesions on the skin and mucous membranes as a result of the death of melanocytes.

The purpose of our research was to assess the levels of autoantibodies to native DNA class G in the blood serum of patients with vitiligo.

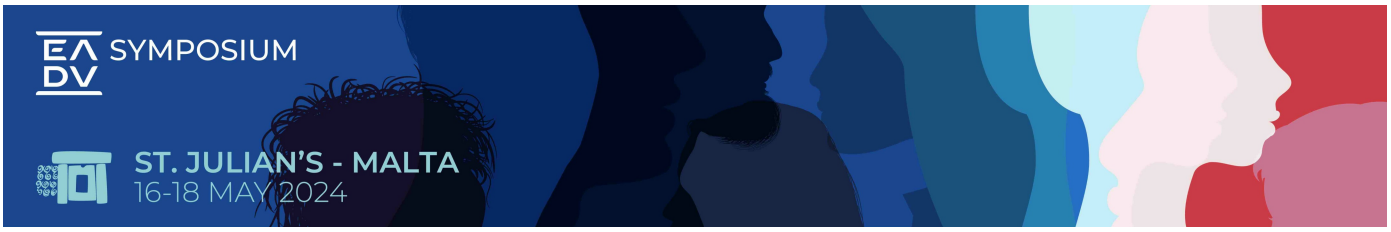
Materials & Methods: 114 patients with vitiligo aged from 7 to 62 years were examined. Among them, there were 44 males and 70 females. All patients underwent clinical, biochemical and ELISA studies. Determination of the level of autoantibodies (AAT) class G (IgG) to native single-stranded (DNA-SS) and double-stranded (DNA-DS) DNA in blood serum was determined by solid-phase ELISA studies. (Vector-Best company). All patients were consulted by related specialists: therapist, endocrinologist, etc. The control group consisted of 27 healthy individuals of the appropriate age.

Results: According to the clinical form, among 114 patients, the non-segmental form of vitiligo was 92 patients (80.7%), segmental - 22 (19.3%), respectively.

The results of an ELISA study on the detection of AAT in patients with vitiligo showed that among 114 patients, 61 patients had an increase in the level of AAT IgG to DNA in the blood serum, which amounted to 53.5% of cases. Moreover, the frequency of detection of IgG antibodies to double-stranded DNA (anti ds DNA) was observed in 61 out of 114 (53.5%), while single-stranded DNA was detected in 60, which amounted to 52.6% of cases. In the group of healthy individuals, among 27 individuals, only one had an increased level of class G AAT to DNA-SS, which amounted to 5.8% of cases.

A quantitative characteristic of the level of AAT in the blood serum of patients with vitiligo, depending on the clinical form, showed that in patients with a non-segmental form, the level of IgG autoantibodies to two ds-DNA increased by 3.6 times compared with the indicators of healthy individuals and compared with the indicators of patients with segmental form form – 1.06 times and on average amounted to –37.1±0.8 IU/ml. The same trend was noted in the level of IgG to single-stranded DNA (anti-ss-DNA) in the group of patients with vitiligo with a non-segmental form and averaged 37.2±0.7 IU/ml and the data obtained were statistically significant. (P <0.05).

Conclusion: The data obtained confirm the development of autoimmune processes in patients with vitiligo, which is of diagnostic and prognostic significance for the purpose of early detection of comorbid diseases and prescribing appropriate treatment.



Abstract N°: 937

Pemphigoid gestationis as a potential marker of choriocarcinoma intraplacentale

Melita Vuksic Polic[1, 2], Lana Lukenda[1], Lorena Stanojevic[1], Tena Zovkic[1]

[1]Josip Juraj Strossmayer University of Osijek, Faculty of Medicine, Osijek, Croatia,[2]University Hospital Osijek, Department of Dermatology and Venereology, Osijek, Croatia

Introduction & Objectives:

Choriocarcinoma is a rare trophoblastic malignancy of the placenta. It can be divided into two categories: gestational, which is more common, and non-gestational. Pemphigoid gestationis can be a rare paraneoplastic dermatosis. It can often be mistaken for several pruritic rashes that occur during pregnancy.

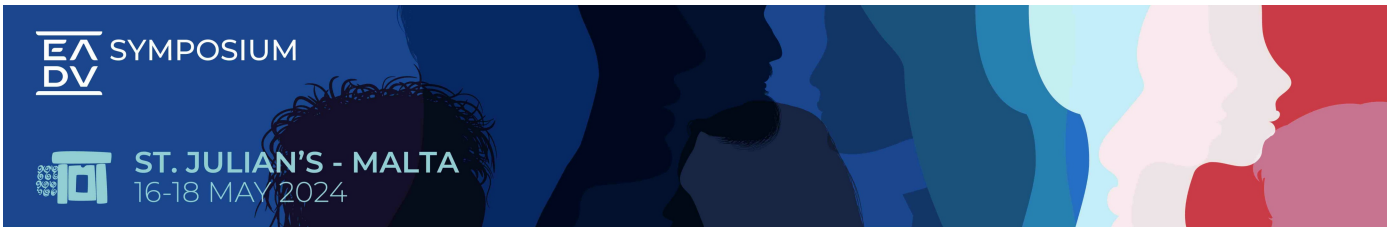
This case report aims to show the rare but important connection between pemphigoid gestationis and gestational choriocarcinoma.

Results:

In February 2023, a 26-year-old female patient with no significant past medical history, a multipara, reported to the Department of Dermatology and Venereology at 34 weeks gestation. The patient presented with confluent erythematous plaques in the umbilical region and smaller disseminated erythematous plaques on the abdomen, accompanied by intense pruritus. She was prescribed topical and systemic therapy, including corticosteroids and oral antihistamines. Two weeks later, despite the prescribed therapy, the clinical status worsened. The patient had disseminated papulovesicular morphs and plaques on the torso, as well as bullous morphs on the limbs. Bullous pemphigoid was clinically suspected, which was also histologically proven. In March 2023, the patient vaginally delivered a live, female baby weighing 3330g. The placenta was also delivered and pathohistologically diagnosed as intraplacental choriocarcinoma. Metastases in the lungs were also proven by diagnostic treatment. The multidisciplinary team recommended a hysterectomy, bilateral adnexectomy, and partial omentectomy, which were successfully performed on April 3rd, followed by polychemotherapy. Dermatological therapy included systemic corticosteroids, antihistamines and topical corticosteroids treatment. In addition, oncological therapy was also carried out: methotrexate in 5 cycles. The patient's post-operative gynecological examination was without abnormal findings. She was discharged against medical advice, with a recommendation to continue dermatological and oncological treatment.

Conclusion:

In conclusion, it is important to distinguish pemphigoid gestationis from other pruritic rashes in pregnant women, as it may be a first sign of choriocarcinoma. Particular attention should be directed towards multiparas. Making an early diagnosis can improve the overall outcome of this disease. Moreover, pemphigoid does not affect most fetuses, but in rare cases, transient lesions can occur, which can end up with complications and neonatal mortality.



Abstract N°: 1030

Linear IgA Bullous Dermatosis triggered by Pregnancy in a woman with Ulcerative Colitis

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Introduction & Objectives:

Linear IgA Bullous Dermatosis (LABD) is an autoimmune blistering dermatosis that can be associated with dysimmune or malignant conditions. An association with inflammatory bowel diseases (IBD) is also possible.

We report the case of a patient with ulcerative colitis (UC) who developed LABD during her second pregnancy.

Observation / Results:

A 28-year-old woman with a history of ulcerative colitis (UC) treated with mesalazine for two years, and who had her second delivery six months ago, consulted for painful erosive lesions on the areolas and nipples, which appeared during a flare-up of her IBD.

Her first pregnancy, which occurred four years ago, was uneventful, and she had no complaints during the postpartum period. However, during the second trimester of her recent pregnancy, the patient reported the onset of pruritic erosive lesions on the outer thighs and buttocks. These lesions had slightly improved with the use of topical corticosteroids.

Clinical examination revealed hyperpigmented scars and erythematous papules on the buttocks, but the mucous membranes were normal.

Histological examination of a skin biopsy fragment showed dermo-epidermal cleavage, and direct immunofluorescence demonstrated a linear deposition of IgA along the basement membrane, without IgG deposition. These findings confirmed the diagnosis of LABD.

Treatment with prednisolone at a dose of 40 mg per day, in combination with mesalazine, led to improvement in gastrointestinal symptoms within three days and healing of the cutaneous lesions after one month. Stabilization of the UC and dermatosis over two months allowed for a gradual reduction in the corticosteroid dose, which was maintained at 30 mg per day.

Discussion:

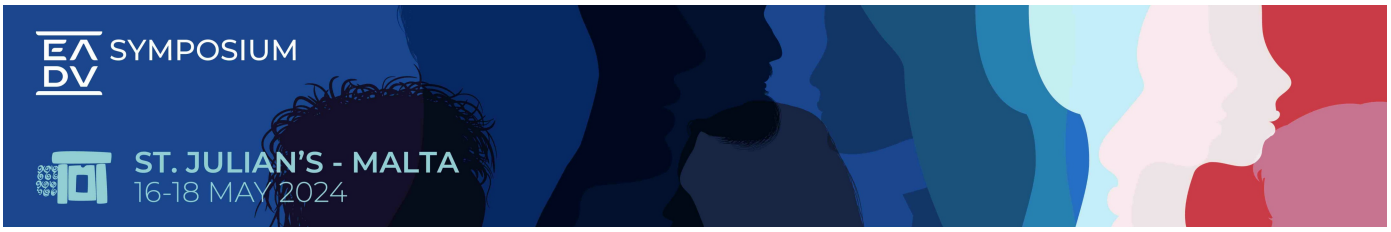
LABD can rarely be a complication of UC. According to recent data, 36 cases of UC associated with LABD have been reported up to 2022, with a male-to-female sex ratio of 1.25. In the majority of cases, the onset of UC preceded that of the dermatosis, as in the case of our patient. Indeed, intestinal inflammation leads to the exposure of intestinal antigens that cross-react with skin antigens, stimulating the autoimmune response to the skin basement membrane.

The occurrence of LABD for the first time during pregnancy has rarely been reported in the literature. Hormonal changes due to pregnancy have been implicated in the pathophysiology of the dermatosis. Moroever, the pathophysiological mechanism of LABD during pregnancy is similar to that of gestational pemphigoid. Aberrant expression of HLA class II molecules in the placenta induces the production of IgA autoantibodies against the foeto-placental unit.

The use of dapsone is often necessary to stabilize the dermatosis. However, it was not used in our patient due to its passage into breast milk.

Conclusion:

The diagnosis of LABD should be considered in cases of blisters or erythematous erosions in patients with IBD. Pregnancy and UC can trigger the onset of LABD through immunopathogenic changes.



Abstract N°: 1042

The experience of a Mediterranean country in the management of morphea disease

Darghal Hanane*[1], Lina Benchekroun[1], Karima Senoussi[1], Mariame Meziane[1]

[1]Hôpital Ibn Sina , dermatology, Rabat

Introduction & Objectives:

Morphea, or localized scleroderma, is defined by a sclerotic condition of the skin that can extend to the subcutaneous tissues but without Raynaud's phenomenon or visceral involvement. Some clinical forms may have functional and aesthetic implications, hence the interest in early management at the inflammatory stage.

Materials & Methods:

This is a retrospective monocentric descriptive study focusing on a series of 57 cases of morphea collected within the dermatology department of the Ibn Sina University Hospital Center in Rabat.

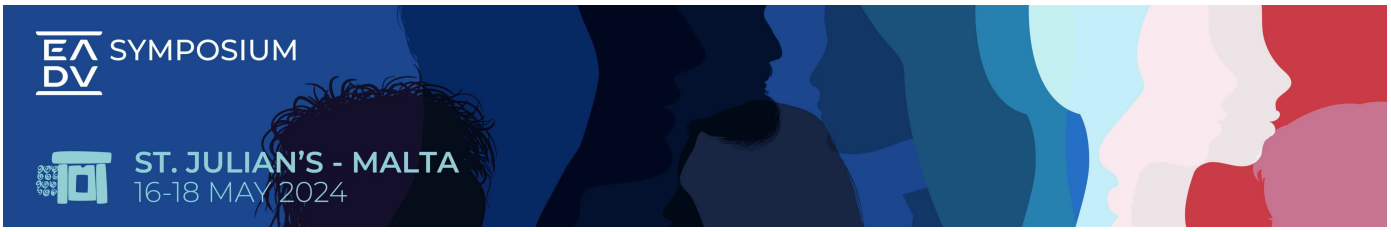
Results:

The male-to-female sex ratio was 3.3. The mean age of our patients was 45 years with extremes ranging from 5 to 80 years. 53.6 % of patients were in the 3rd age group, between 40 and 80 years old. Medical history was present in 13 out of 57 patients, mainly represented by autoimmune diseases found in 10 of them. Clinical forms were diverse. We identified (41%) plaque morphea, (38%) linear morphea, (19%) generalized morphea, (2%) deep morphea, and 1% bullous morphea. We found an association of two or more subtypes in 28.6 % of patients in our series, with the most common association being plaque and linear morphea.

Among the 57 patients collected, 4 did not receive treatment. The evolution was assessed in 45 patients. Functional sequelae were mainly observed in the linear form.

Conclusion:

Although the prognosis of localized scleroderma is generally good, some clinical forms can have functional and aesthetic repercussions, especially on the face and limbs near the joints. General corticosteroid therapy associated or not with an immunosuppressive or antifibrotic treatment seems necessary for linear and deep forms.



Abstract N°: 1045

A rare association of IgA vasculitis with aorto-mesenteric clamp syndrome and renal nutcracker syndrome: A case report

Hind Majdoul*[1], Fouzia Hali[1], Kenza Barbri[1], Bouchra Baghad[1], Soumiya Chiheb[1]

[1]Ibn Rochd University Hospital Center, Dermatology and Venerology Department, Casablanca, Morocco

Introduction & Objectives:

IgA vasculitis is a systemic vasculitis of small vessels related to tissue deposits of immune complexes containing type A immunoglobulins. It mainly affects children, and is less common in adults. The short-term prognosis depends on the severity of the digestive involvement, whereas the long-term prognosis depends on the renal damage.

Aorto-mesenteric clamp syndrome is a rare cause of upper gastrointestinal obstruction, due to extrinsic compression of the third portion of duodenum, between the superior mesenteric artery and the aorta.

We report the case of a rare association of IgA vasculitis with both aorto-mesenteric clamp syndrome and renal nutcracker syndrome in a teen patient.

Materials & Methods:

Results:

Case presentation:

A 16-years-old patient, with a pathological history of testicular ectopy with spontaneous resolution at the age of 3, presented for 3 weeks diffuse abdominal pain, initially isolated, then complicated by early post-prandial vomiting and a rapid alteration of his general condition. The patient consulted a general practitioner who, after a normal biological check-up, prescribed a treatment combining an antibiotic, an anti-emetic and an anti-spasmodic. Not only did the patient not improve, but also the clinical presentation was further aggravated by the cessation of materials and gas, as well as the appearance of a petechial purpuric rash, sloping, involving all four limbs distally, without any notion of fever, nor photo-phonophobia or consciousness disorder.

The patient was admitted in General surgery department, and underwent radiological investigations, including an abdominopelvic CT scan showing an upper gastrointestinal obstruction secondary to a superior mesenteric artery syndrome associated with a renal nutcracker syndrome. The laboratory tests revealed an isolated inflammatory syndrome.

The contribution of dermatologists was requested during the patient's hospitalization in General surgery department, for the diagnosis and management of the above mentioned skin lesions. Our examination revealed infiltrated petechial purpuric macules, purplish-red in color, located on the hands and feet and extending to the distal part of the forearms and legs. The diagnosis of vasculitis was considered after excluding a case of purpura fulminans or thrombocytopenic purpura; therefore, a skin biopsy for histopathological analysis and direct immunofluorescence, along with an immunological assessment were performed in that sense, which results concluded to an IgA vasculitis.

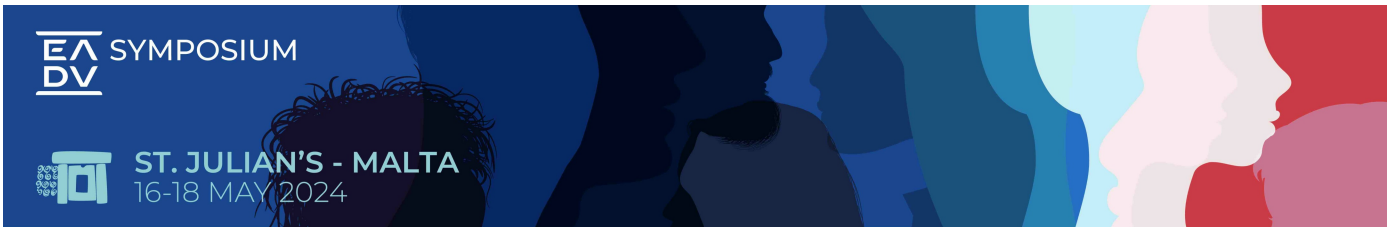
The patient underwent a surgical procedure for his occlusion, while the skin lesions progressed well under short cure dermocorticoids.

Conclusion:

IgA vasculitis is known to be associated with gastrointestinal involvement in more than 50% of cases; abdominal pain

being the most common manifestation, but severe complications such as intussusception, appendicitis or pancreatitis were also reported. Systemic corticosteroid therapy should be started at an early stage whenever digestive symptoms are severe, otherwise, the delay in medical treatment will lead to serious consequences requiring emergency surgery.

An anatomical condition mimicking aorto-mesenteric clamp syndrome might cause duodenal involvement during IgA vasculitis. Indeed, the duodenum being frequently affected, its parietal inflammation could lead to a reduction in both aorto-mesenteric angle and aorto-mesenteric distance, especially in thin patients.



Abstract N°: 1089

Beauty is only skin deep, morphea may not – case presentation of deep localized scleroderma with severe thorax deformity

Natalia Tekiela[1], Agnieszka Guberniak[1], Karina Polak[2], Bartosz Miziołek[2], Beata Bergler-Czop[2]

[1]Students Scientific Association at the Department of Dermatology, Medical University of Silesia, Katowice, Poland,[2]Chair and Department of Dermatology, Medical University of Silesia, Katowice, Poland

Introduction & Objectives:

Deep scleroderma (DS) is a rare (0.4–2.7 cases/100,000 persons/year) form of localized scleroderma (LoSc, morphea) characterized by the presence of deep sclerotic lesions affecting the muscles, fasciae, subcutaneous tissue and deeper layers of the dermis typically occurring in the paravertebral line, with potential triggering factors including infection, injury, radiation or drugs. Due to lack of adequate awareness among healthcare professionals, the diagnosis may be delayed, which may result in irreversible anatomical changes. The authors present a case of deep morphea in an unusual location including a severe deformity of the anterior thorax, the course of its' treatment and discuss possible therapeutic methods.

Materials & Methods:

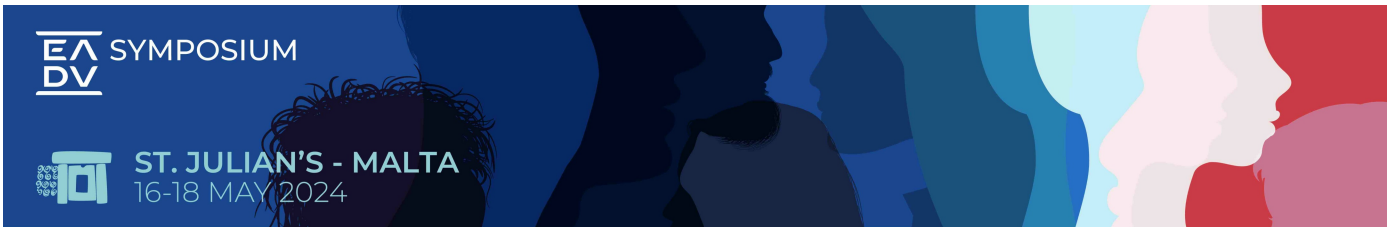
A 67-year-old woman with DS resulting in a severe deformity of the anterior wall of the thorax was referred to the dermatology clinic due to concomitant skin infection. Symptoms of DS were first observed in 1984, initially taken for breast cancer, then panniculitis non febrilis; the performed biopsy confirmed the diagnosis of DS. On admission, the patient presented with multiple, diffuse sclerotic lesions including infiltrating nodules, ill-defined dimpling scars and presence of post-inflammatory discoloration in the anterior chest region with no nipples involvement. MRI confirmed characteristic fibrous typical of DS and CT scan revealed bilateral sclerotic breasts skin infiltration. Over the years, the patient has been treated in various ways for the recurrent skin infections in the affected region, yet no systemic treatment targeting DS was used. Bacterial swab revealed methicillin-susceptible S. aureus. The patient received systemic targeted antimicrobial therapy with gradual improvement and after 7 days discharged from the hospital. During follow-up, s.c. methotrexate (MTX) 15mg/week, oral prednisone 0.5-1mg/kg body weight/day with gradual dose reduction, and mycophenolate mofetil (MMF) 2g/day were introduced, resulting in limiting the progression of the disease.

Results:

Successful treatment of concomitant infection with introducing systemic treatment, limiting the progression of the disease, was introduced. If DS is diagnosed, rapid treatment is required in order to prevent further damage. The treatment of choice is intravenous glucocorticosteroids (methylprednisolone) at a dose of 500-1000 mg/day for 3 days; an alternative to parenteral treatment is oral prednisolone therapy. It is recommended to administer 0.5-2 mg/kg/day of prednisolone for 2-4 weeks, followed by a gradual reduction of the dose. Glucocorticosteroids can be used in monotherapy or in combination with MTX (recommended dose 12,5-25mg per week continued for at least 12 months). MTX intolerance or drug resistance is an indication to consider MMF (1-2 g/day).

Conclusion:

Due to lack of adequate awareness among healthcare professionals, the diagnosis of DS is usually delayed, which may result in irreversible anatomical changes. This care report is meant to raise awareness of the DS, that should routinely be differentiated from subcutaneous tissue inflammation and the subcutaneous deep lupus erythematosus and treated immediately after diagnosis.



Abstract N°: 1118

Bullous pemphigoid and Milia: A case series of seven Moroccan patients

Saliha Jebbouje[1], Hali Fouzia[1], Bouchra Baghad[1]

[1]chu Ibn Rochd Casablanca , dermatology

Introduction & Objectives:

Milia are superficial keratin cysts seen as pearly white, measuring 1-2 mm in diameter. Milia are associated with diseases that cause subepidermal blistering. Bullous pemphigoid (BP) is the most common autoimmune blistering disease worldwide. The aim of this study was to describe the occurrence and clinical-laboratorial findings of BP-milia association in our patients.

Materials & Methods:

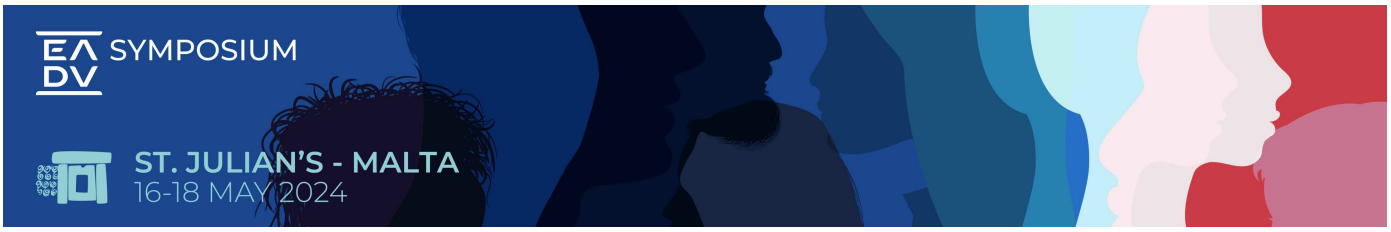
A retrospective, descriptive, and monocentric study was conducted in the department of dermatology of Ibn Rochd Hospital between January 2013 and May 2023 including patients with both BP and milial lesions. BP confirmation was based on clinical, histological elements and linear fluorescence of IgG at the level of the basement membrane zone during direct and indirect immunofluorescence (DIF, IIF respectively).

Results:

Out of 147 BP cases collected, seven showed milia formation, corresponding to a prevalence of 4.76%. Six were males, with a median age of 66.8 years and an average diagnostic delay of 5 months. Neurological impairments were present in 3 patients. All patients presented typical BP lesions with histological examination revealing detached epidermis at the junction and eosinophilic inflammatory infiltrate at the dermal level. DIF showed junctional deposits of C3 and/or IgG. Indirect immunofluorescence for anti-BP autoantibodies (anti-BP180 and anti-BP230) returned positive. Milia appeared within an average of 2.6 months. The eruption consisted of pearly white dome-shaped lesions measuring 1 to 2 mm in diameter, non-pruritic, localized at the sites of previous bullous scars in all cases except one where they appeared on healthy skin. The most common sites in our patients were the trunk and lower limbs, followed by the neck and face. Regression occurred within a few months without adjunctive ablative treatment.

Conclusion:

To our knowledge, the association of bullous pemphigoid and milium formation is a rare finding. In our series, the prevalence of this association is 4.76%, indicating the necessity of a careful differential diagnosis with hereditary epidermolysis bullosa. Neurological impairments associated with BP were described in 3 of our patients. The neurological and BP profile suggests a link in the pathogenesis of BP, but no association with the appearance of milia has been found in the literature. The formation of milia during BP remains poorly understood, but an interaction of immunological predisposition and atypical interactions between hemidesmosomes and the extracellular matrix is suggested. Clinicians should be cautious in making an accurate diagnosis of the type of bullous dermatosis in the presence of these epidermal cysts.



Abstract N°: 1122

Patient-Reported Quality of Life, Psychosocial, and Work Productivity Impacts among Patients with Clinically Distinct Alopecia Areata Severity Profiles

Kent Hanson[1, 2], Jenny Austin[3], Peter Anderson[3], Simran Marwaha[3], Samantha Kurosky[2], Alexandre Lejeune[4], James Piercy[3], Ernest Law*[2]

[1]University of Illinois Chicago, Pharmacy Systems, Outcomes and Policy, Chicago, United States,[2]Pfizer Inc, New York, United States, [3]Adelphi Real World, Bollington, United Kingdom, [4]Pfizer Inc, Paris, France

Introduction & Objectives:

Alopecia areata (AA) is an autoimmune disease characterized by non-scarring hair loss that often leads to substantial psychosocial burden. The clinical presentation of AA varies and may result in differential impact on patients' quality of life (QoL), disease perception, and work productivity. The aim of this study was to identify discrete AA severity profiles and evaluate the association between each profile and patient-reported outcomes (PROs).

Materials & Methods:

This analysis used data from the Alopecia Areata Disease Specific Programme, a survey that collected data through medical record review and patient-completed assessments (PCA) from dermatologists and adult patients in France, Germany, Italy, Spain, and the United Kingdom. Patients recruited by dermatologists were invited to complete a PCA including PROs such as the Alopecia Areata Patient Priority Outcomes (AAPPO), Work Productivity Activity Index - AA (WPAI-AA), Skindex-16, Patient Satisfaction With Hair Growth (P-Sat), and Hospital Anxiety and Depression Scale (HADS). Latent class analysis (LCA) was used to identify distinct, unobservable classes using response options from the AAPPO. Logistic and linear regression models, adjusted for age, sex, and current treatment, were used to evaluate the relative association between class assignment and PROs.

Results:

A five-class model was selected based on model fit statistics and disease state knowledge. Each class exhibited distinct profiles based on patient-reported hair loss (HL) and emotional symptom and activity limitation (ES/AL) burden **(Figure 1)**: (1) very mild HL, very mild ES/AL; (2) mild HL, mild ES/AL; (3) moderate HL, very severe ES/AL; (4) severe HL, moderate ES/AL; (5) very severe HL, severe ES/AL. Compared to the mildest class (class 1), on average, class 3 reported significantly more overall work impairment due to AA (ß = 16.3 [95% confidence interval, 7.8 - 24.7]) **(Figure 2)**. Overall Skindex-16 scores were highest for class 3 (ß = 38.7 [95% confidence interval, 33.2 – 44.2]), followed by class 5 (ß = 37.6 [95% confidence interval, 26.8, 48.3]). Compared to class 1, class 3 reported significantly more anxiety and depression as measured by the HADS (Anxiety: ß = 5.9 [95% confidence interval, 4.9, 7.0]; Depression: ß = 5.7 [95% confidence interval, 4.6 – 6.9]). Across all items of the P-Sat, class 5 reported the least satisfaction with therapy.

Conclusion:

Five clinically distinct AA severity profiles were identified based on AAPPO responses. Classes with moderate to extensive HL were associated with moderate to high psychosocial impacts. Generally, severe classes had decreased quality of life, greater work productivity impairment, and less satisfaction with care. Although classes 4 and 5 included patients with the most HL burden, class 3 experienced the most impact on most PROs, suggesting AA can have a profound impact on well-being and daily activities, even in patients with less extensive hair loss.

Figure 1. Mean AAPPO domain/item response scores by identified class

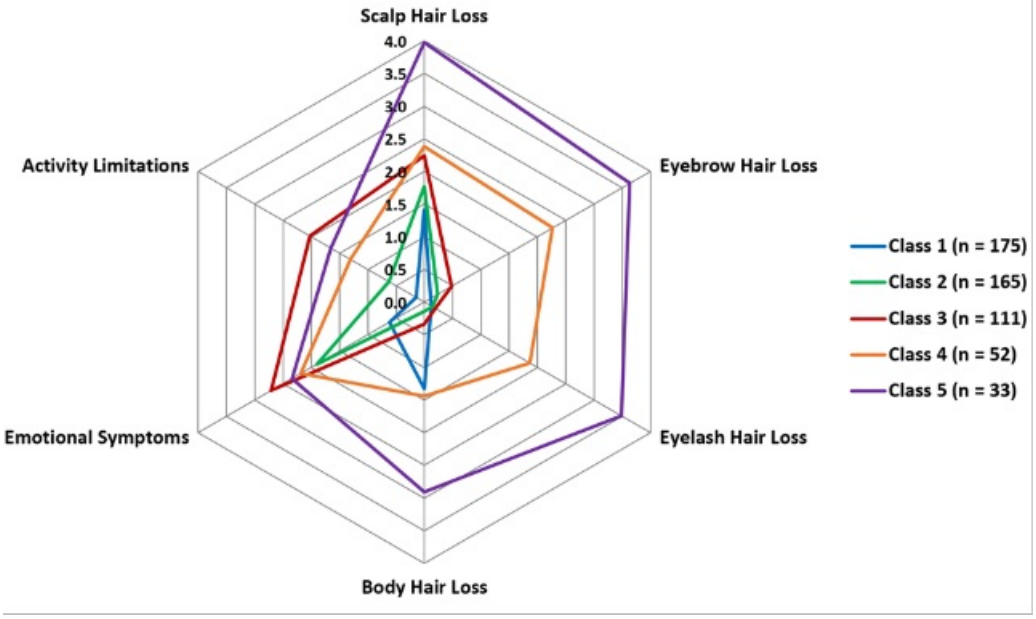
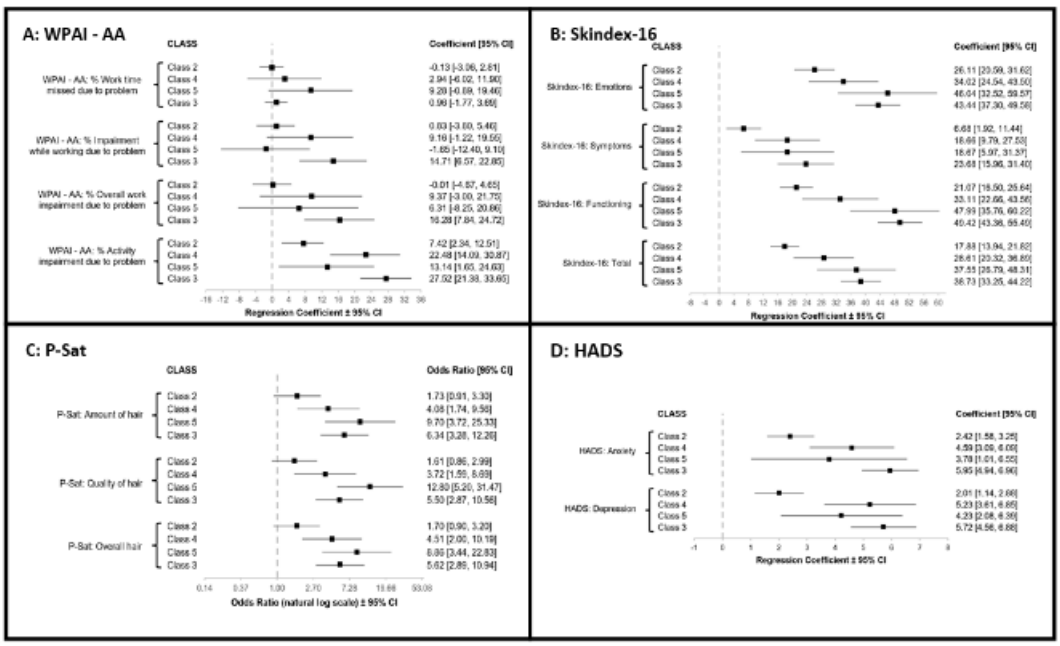
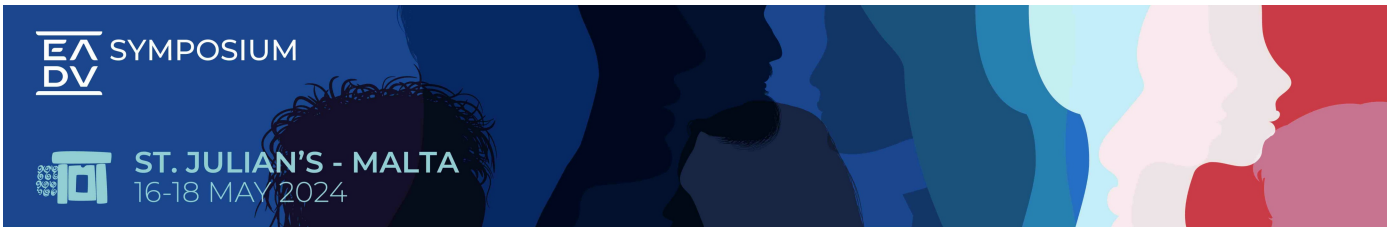


Figure 2. Class associations with patient-reported outcomes (relative to class 1)





Abstract N°: 1124

Indurated plaque with ulceration on the dorsum of the right forearm and shoulder: A diagnostic challenge

Saliha Jebbouje[1], Hali Fouzia[1], Bouchra Baghad[1]

[1]chu Ibn Rochd Casablanca , dermatology

Introduction & Objectives:

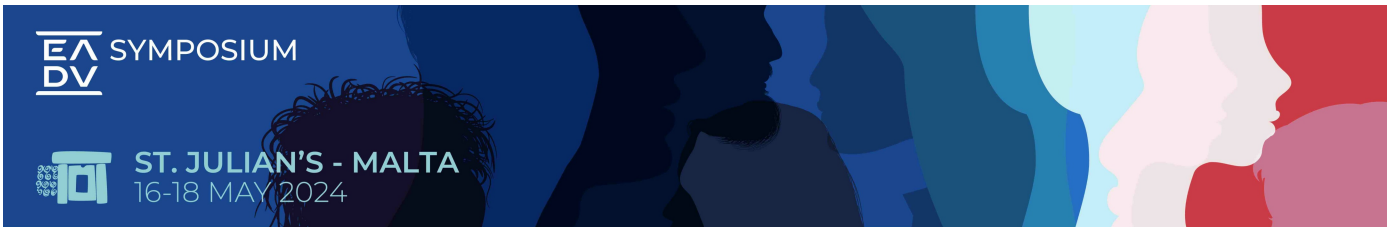
Morphea is a skin condition characterized by excessive deposition of collagen in dermal and/or hypodermic thickening. Lupus erythematosus panniculitis (LEP) is manifested by infiltrated plaques with lesions that ulcerate in 30% of cases. Finding both LEP and morphea in the same lesion is uncommon, especially when associated with systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APLS).We report a challenging case of morphea-lupus panniculitis-LES-APLS in a patient

Case report:

A 22-year-old female with deep morphea under strong topical corticosteroids for 3 years presented ulcerations on an indurated plaque of the posterior face of the right forearm and the left shoulder. She initially noticed deep subcutaneous nodules which evolve into ulceration. Some of these nodules which spontaneously resolved left depressed lipoatrophic areas.Standard X-ray of the right upper limb did not show calcium deposits. The biological assessment showed the presence of lymphopenia at 920, haemolytic anemia at 10.3, positive anti-nuclear antibodies (ANA) and anti-ds-DNA antibodies, false syphilitic serology VDRL + 8 and positive anti-phospholipid antibodies. Cutaneous biopsy revealed dense fibrosis of the dermis and hypodermis associated with lymphocytic panniculitis. Direct immunofluorescence does not objectify immunoglobulin or complement deposits. These findings were in consistence with morphea-lupus panniculitis associated with SLE and APL syndrome. The patient was treated with prednisone (1mg/kg/day), synthetic antimalarials (400mg/day), methotrexate (15mg/week), salicylic acid (75mg/day) and PUVA therapy. These treatments resulted in complete resolution within 6 months with no recurrence after a follow up of 6 months.

Conclusion:

We report a historical case of morphea-lupus panniculitis overlap associated with systemic lupus erythematosus and APL syndrome.The differentiation of LEP occurring without typical skin lesions of LE from deep morphea can be difficult because both diseases may show lipoatrophy, scleroderma-like lesions and deep subcutaneous nodules and/or plaques. They may also share some histopathological findings such as lymphocytic panniculitis, lymphoid nodular structures in the fat, broadening of fibrous septa of fat lobules and lymphocytic vasculitis. In our case both co-existing of signs of morphea and lupus panniculitis also, positive ANA and anti-ds-DNA and APL, the excellent response to the established treatment strongly suggested the overlap syndromes in our patient.



Abstract N°: 1145

Paraneoplastic IgA pemphigus as an early sign of diffuse large B-cell lymphoma relapse

Katarina Peljhan*[1], Neven Franjic[2], Sandra Peternel[1]

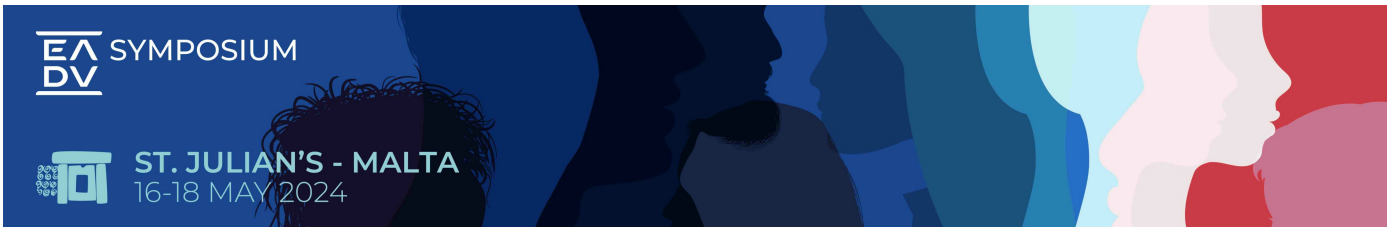
[1]Clinical Hospital Center Rijeka and Faculty od Medicine University of Rijeka, Department of Dermatovenereology, Rijeka, Croatia, [2]Clinical Hospital Center Rijeka and Faculty od Medicine University of Rijeka, Department of Internal medicine, Division of Hematology

Introduction & Objectives: IgA pemphigus is a rare autoimmune blistering skin disorder that has been reported in association with multiple myeloma, hematological malignancies, lung cancer and other solid tumors as well as with several chronic diseases such as rheumatoid arthritis and Sjogren syndrome. Here we present a case in which the appearance of IgA pemphigus preceded and possibly indicated a diffuse large B-cell lymphoma (DLBCL) relapse with a fatal outcome.

Results: A 76-year-old female patient with previous medical history of scalp psoriasis and non-Hodgkin lymphoma (NHL), DLBCL subtype, presented with a generalized rash lasting for a little over a month. Clinical examination revealed widespread bright red annular plaques with peripheral scaling. A skin biopsy was performed with psoriasis and erythema annulare centrifugum as differential diagnoses. General pathologist interpreted the histopathological findings as being consistent with psoriasis, due to pronounced epidermal changes and epidermal neutrophilic infiltrate, however, in consultation with a dermatopathologist, more emphasis was put on discrete signs of acantholysis in the upper epidermis, and a differential diagnosis of pemphigus foliaceus or IgA pemphigus was suggested. Direct (IgG, IgA, IgM, C3, fibrinogen and C1q) and indirect (IgG, IgA, IgM, C3, ICAb) immunofluorescence analyses were negative, while ELISA was negative for IgG against desmoglein 1 and 3, BP180, BP230 and envoplakin. Nevertheless, based on the correlation of clinical and histopathological findings, a diagnosis of IgA pemphigus, subcorneal pustular dermatosis-type was made. The patient was initially treated with prednisone 40 mg/day and topical corticosteroids which lead to significant clinical improvement. Acitretin 25 mg/day was then added, allowing prednisone taper to 15 mg/day with gradual resolution of skin lesions. Simultaneously, the patient underwent hematological workup because of a newly enlarged lymph node in her left axilla which appeared one month after the skin rash. This was confirmed to be DLBCL infiltrate and was treated with radiotherapy, however, in the following two months there was further progression of the disease, with CT scans showing conglomerates of enlarged lymph nodes around the aorta and hepatoduodenal ligament along with ascites and spleen infiltrates. The patient was started on a new polychemotherapy regimen but unfortunately has deceased soon thereafter.

Conclusion:

IgA pemphigus is a rare entity in which careful clinicopathological correlation is required for a diagnosis to be made due to significant overlap with other blistering diseases, pustular psoriasis and other dermatoses. It should also be noted that negative results of immunofluorescent analyses do not necessarily exclude a diagnosis of IgA pemphigus. Special regards should also be made for a patient's medical history. Specifically, a new onset of IgA pemphigus in an NHL patient should prompt the clinician to carefully examine the possibility of an aggressive relapse of the preexisting malignant disease.



Abstract N°: 1152

Dermatomyositis and morphea: a rare association.

Gabriela Benjuia[1], Sabrina Merenzon[1], Vicoria Saieg[1]

[1]Hospital General de Agudos J. M. Ramos Mejía, Buenos Aires, Argentina

Introduction & Objectives: dermatomyositis and morphea: a rare association.

Dermatomyositis(DM) is a rare disease typically characterized by proximal muscle weakness, accompanied by skin manifestations such as heliotrope erythema, Gottron papules, poikiloderma, and telangiectasias. Additionally, patients may experience dysphagia and nail disorders.

On the other hand, morphea is characterized by fibrosis and skin hardening, presenting as superficial plaques. Linear scleroderma, another variant, involves fibrosis affecting the dermis, subcutaneous tissue, and occasionally bone.

Although the coexistence of both diseases is rare, cases of overlapping manifestations have been observed. Early diagnosis and appropriate treatment can significantly influence the progression of both conditions and improve the patient's quality of life.

Materials & Methods: The diagnostic process involves gathering medical and personal history, conducting laboratory tests, and performing a skin biopsy.

Results:

Case report:** A 32-year-old patient with a history of DM since 2022, managed with methotrexate, folic acid, hydroxychloroquine, and meprednisone, presented with progressive dysphagia and proximal muscle weakness lasting over a month. He responded well to treatment with cyclophosphamide and methylprednisolone.

During hospitalization, blood laboratory tests revealed a CPK value of 3042 (normal range: 10-120 mcg/L), and electromyography indicated a recruitment pattern with minimal decreased effort.

On physical examination, bilateral pain in the quadriceps during stair movement was noted, although gait remained intact. Cutaneous examination revealed hyper- and hypopigmented macules on the face, a V sign on the neckline, and rounded hyperpigmented macules on the torso and back. A 12x5 cm hyperpigmented erythematous-violaceous plaque was observed on the left flank, demonstrating reduced flexibility. A skin biopsy confirmed the lesion as morphea.

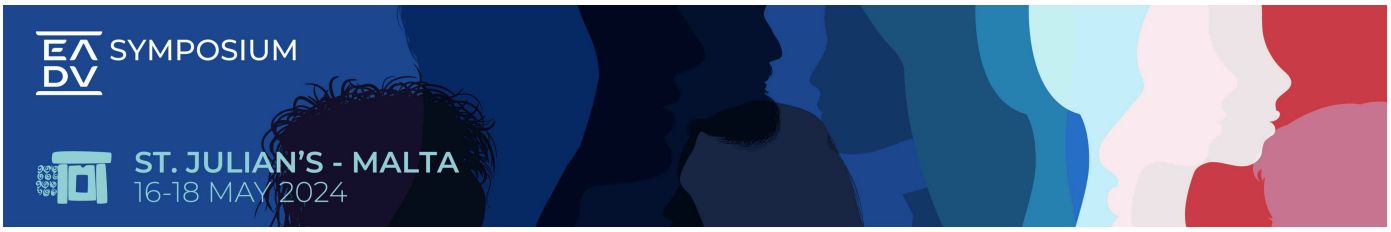
Conclusion: Acquired connective tissue diseases can occasionally manifest concurrently, a phenomenon referred to as "overlap syndrome." This condition has a prevalence of 20-25% in the population, wherein two or more systemic autoimmune diseases coexist simultaneously. Among the most commonly encountered combinations are antiphospholipid syndrome (APS) secondary to systemic lupus erythematosus (SLE), DM coexisting with SLE, rheumatoid arthritis (RA) alongside systemic sclerosis (SS), and polymyositis in conjunction with SS.

In patients presenting with challenging-to-treat rheumatological conditions, it is crucial to first rule out associated pathologies. This is particularly important for patients with positive antibodies and compatible symptoms, given the low incidence of such conditions. Sometimes, diagnoses are missed or only identified once symptoms or clinical signs manifest.

In the majority of cases described, the treatment yielding the best results for this condition involved a combination of immunosuppressants and corticosteroids, as seen in the previously described case.

In the current case, the patient has continued attending subsequent clinical check-ups, reporting improvement in

dysphagia and muscle weakness initially presented. Presently, the patient is undergoing treatment with methotrexate, azathioprine, and meprednisone.



Abstract N°: 1159

Demographics and Clinical Characteristics among Patients with Distinct Psychosocial Burden Profiles Related to Vitiligo

Kent Hanson[1, 2], Jenny Austin[3], Peter Anderson[3], Simran Marwaha[3], Samantha Kurosky[2], Roni Adiri[4], James Piercy[3], Ernest Law*[2]

[1]University of Illinois Chicago, Pharmacy Systems, Outcomes and Policy, Chicago, United States,[2]Pfizer Inc, New York, United States, [3]Adelphi Real World, Bollington, United Kingdom, [4]Pfizer Pharmaceuticals Israel Ltd, Herzliya, Israel

Introduction & Objectives:

The impact of vitiligo often extends beyond the skin, translating to increased psychological burden and impaired quality of life. Impact may vary by patient depending on factors such as skin depigmentation level, areas affected, and disease stability. This study aimed to 1) identify classes of patients based on the impact of vitiligo on their emotional/psychological and social functioning and 2) describe patient characteristics and patient-reported outcome (PRO) responses for each profile.

Materials & Methods:

Secondary data consisting of dermatologist-abstracted medical record reviews and patient-completed assessments (PCA) from dermatologists and adult and adolescent (age 12-17 years) patients with vitiligo in the United States, France, Germany, Italy, Spain, and the United Kingdom (Vitiligo Disease Specific Programme) were analyzed. Patients were invited to complete a PCA including PROs such as the Vitiligo Patient Priority Outcomes (ViPPO), Work Productivity Activity Index-Vitiligo (WPAI-Viti), Vitiligo-specific Quality-of-Life instrument (VitiQoL), EQ-5D-5L, and Hospital Anxiety and Depression Scale (HADS). A latent class analysis approach was used to identify distinct classes using ViPPO response options; each class represented a profile based on patient-reported emotional/psychological and social functioning. Patient characteristics and PRO responses were analyzed descriptively.

Results:

A three-class model was selected based on model fit statistics and disease state knowledge. Among the 530 patients included, most were grouped into class 1 (64%), followed by classes 2 (24%) and 3 (12%). Class 3 reported the most emotional/psychological and social functioning impact, while classes 1 and 2 reported mild and moderate degrees of impact, respectively (**Figure 1**). Classes 1 and 2 were most likely to have limited to moderate disease extent per physician-assessment, class 3 had a relatively higher proportion of patients with moderate to severe disease (**Table 1**). Class 3 had the highest mean body surface area involvement and had the highest proportion of patients with face involvement (n = 32 [49%]). Across all four PRO measures, class 3 reported more severe impact per HADS, WPAI–Viti, VitiQoL, and EQ-5D-5L **(Table 2)**.

Conclusion:

Three clinically distinct profiles were identified based on emotional, psychological, and social functioning responses. In general, classes with more severe impacts had more severe disease per physician assessment, visible areas of lesion involvement, and greater work productivity and quality of life impairment. However, a large proportion of patients in class 3 (severe psychological/emotional and social burden) also had limited/mild physician-reported disease severity, suggesting potential discordance between physician- and patient-perceived severity.

Figure 1. Mean ViPPO domain response scores by identified class

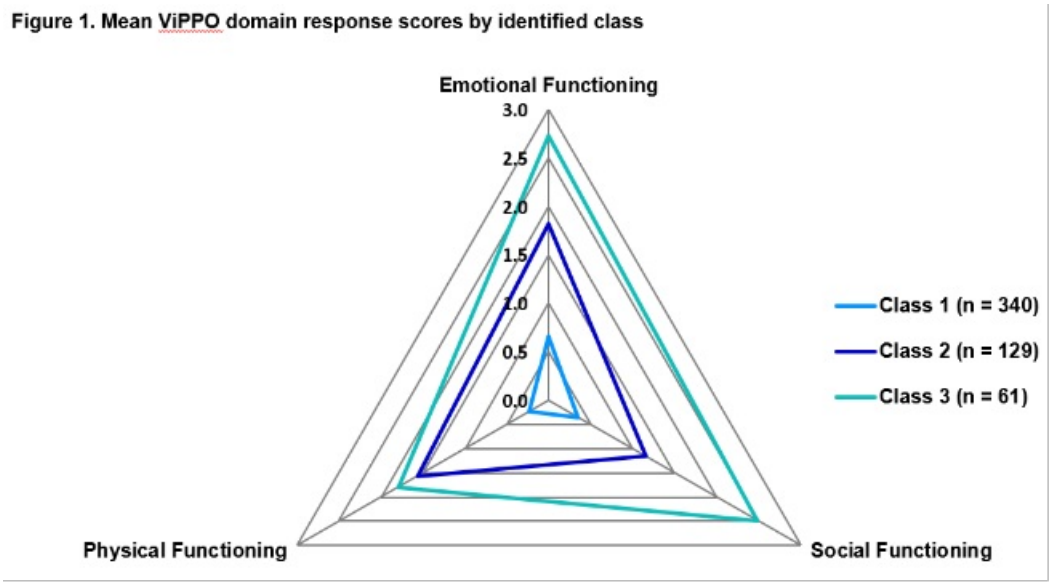
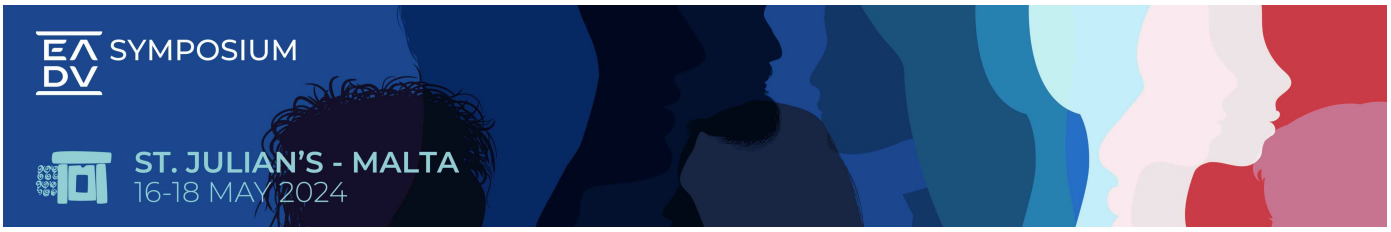


Table 1. Clinical characteristics by class

	Class 1 (n = 340)	Class 2 (n = 129)	Class 3 (n = 61)
Mean time since diagnosis (years)	3.59 (4.93)	2.79 (5.11)	4.15 (5.91)
Current vitiligo activity, n (%)			
Active/ unstable vitiligo	120 (35.3)	40 (31.0)	18 (29.5)
Chronic vitiligo	191 (56.2)	74 (57.4)	31 (50.8)
Refractory vitiligo	29 (8.5)	15 (11.6)	12 (19.7)
Most common areas currently affected, n (%)			
Hands	136 (40.0)	63 (48.8)	32 (52.5)
Face – rest of face	112 (32.9)	55 (42.6)	30 (49.2)
Face – eye area	109 (32.1)	44 (34.1)	30 (49.2)
Neck	73 (21.5)	31 (24.0)	16 (26.2)
Lower arms	63 (18.5)	21 (16.3)	18 (29.5)
Elbows	65 (19.1)	23 (17.8)	10 (16.4)
Genitalia/Groin	65 (19.1)	21 (16.3)	12 (19.7)
Physician-assessed current severity			
Limited extent / mild	209 (61.5)	70 (54.3)	23 (37.7)
Moderate extent	115 (33.8)	52 (40.3)	25 (41.0)
Extensive / severe	14 (4.1)	4 (3.1)	8 (13.1)
Very extensive / very severe	2 (0.6)	3 (2.3)	5 (8.2)
Mean % BSA currently affected	11.33 (9.58)	11.19 (11.9)	18.48 (15.69)

Table 2. Mean PRO Responses by Class

	Class 1 (n = 340)	Class 2 (n = 129)	Class 3 (n = 61)
HADS - Anxiety	2.8 (3.05)	6.65 (3.74)	9.9 (3.53)
HADS - Depression	2.23 (3.07)	5.57 (3.69)	8.46 (4.01)
WPAI - % Activity impairment due to problem	8.36 (13.39)	23.5 (19.61)	43.91 (21.24)
WPAI - % Overall work impairment due to problem	7.15 (11.08)	21.44 (21.12)	25.45 (21.94)
WPAI - % Impairment while working due to problem	6.06 (9.89)	19.57 (17.94)	23.57 (19.67)
WPAI - % Work time missed due to problem	2.7 (13.9)	4.93 (15.25)	8.82 (22.95)
VitiQol	18.05 (15.57)	39.95 (16.94)	61.5 (11.81)
EQ-5D-5L Index	0.95 (0.08)	0.84 (0.19)	0.71 (0.24)



Abstract N°: 1184

Keloidal scleroderma and generalized morphea in systemic sclerosis patient

Esther Kim[1], Juhee Han*[1]

[1]Seoul St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Department of Dermatology, seoul, Korea, Rep. of South

Keloidal scleroderma and generalized morphea in systemic sclerosis patient

Introduction & Objectives:

Cutaneous manifestations in patients with Sjogren's syndrome are diverse, and the coexistence of connective tissue disorders can present diagnostic and therapeutic dilemmas. We present a case of a 43-year-old female with Sjogren's syndrome, exhibiting a unique clinical presentation of multiple yellowish to erythematous nodules on both legs, ultimately diagnosed as a rare combination of keloidal scleroderma and generalized morphea.

Materials & Methods:

The patient's clinical history revealed a two-year progression of nodular lesions on both legs, accompanied by Sjogren's syndrome. Histopathological examination demonstrated distinctive findings including an acanthotic epidermis with basilar hyperpigmentation, eosinophilic amorphous material deposition, a proliferation of myofibroblasts, thickened collagen bundles and dilated vessels in the dermis. Congo red staining was negative.

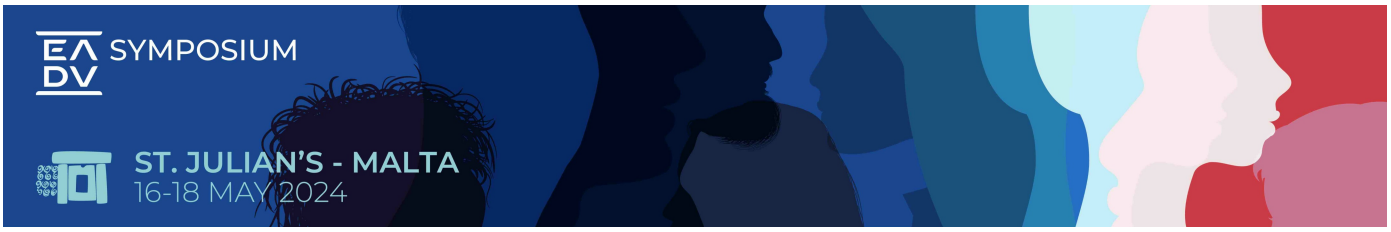
Results:

Based on these findings, she was diagnosed with keloidal scleroderma and generalized morphea.

The coexistence of keloidal scleroderma and generalized morphea in the setting of Sjogren's syndrome posed diagnostic challenges due to the rarity of this presentation. The patient's therapeutic regimen included intralesional steroid injections and oral prednisolone to address both the inflammatory and fibrotic components of the lesions.

Conclusion:

The challenges of managing such a complex case underscore the need for a tailored, multidisciplinary approach. This case also highlights the importance of recognizing and addressing cutaneous manifestations in patients with autoimmune disorders, emphasizing the intricate interplay between Sjogren's syndrome, keloidal scleroderma, and generalized morphea. A multifaceted treatment strategy, including localized and systemic interventions, is essential for optimal management in such complex dermatological scenarios.



Abstract N°: 1229

Rituximab therapy for refractory Ocular Mucous Membrane Pemphigoid

Santoshdev Rathod*[1], Jeta Buch[1], Swati Brahmbhatt[1], Ashish Jagati[1]

[1]Smt. NHL Municipal Medical College, Ahmedabad, India

Introduction & Objectives:

Mucous membrane pemphigoid (MMP) is a heterogeneous group of rare, chronic, subepithelial autoimmune blistering diseases (AIBDs) with predominant involvement of mucous membranes that can be sight-threatening and life-threatening.Rituximab (RTX) has demonstrated its efficacy in severe ocular MMP refractory to conventional immunosuppressants

Materials & Methods:

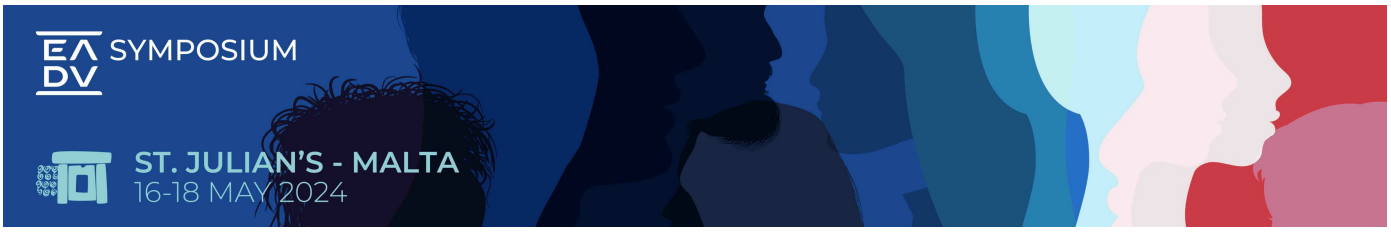
We are presenting two case reports of refractory MMP who had shown continued disease activity and partial response to conventional immunosuppressives given in adequate dosage for optimal time duration. Case 1 was a 52-year-old female with ocular and cutaneous MMP of thirteen-year duration with progressive loss of vision in left eye and diminution of vision in right eye with ocular examination showed hazy cornea in both eyes with symblepharon formation. Case 2 was a 47-year-old female with ocular cicatricial pemphigoid with corneal opacity in both eyes and cutanoeus lichen planus. Due to persistence disease activity in the eye and threat to loss of vision. Both patients were given Rituximab infusion as per lymphoma protocol (500 mg) every week after ruling out contraindication to Rituximab therapy.

Results:

Following one cycle of induction of Rituximab infusion (4 doses of 500mg weekly Rituximab infusion) both the patients responded favorably and visual acuity scores improved on follow up. The cutaneous disease also responded favorably.

Conclusion:

The treatment options for rare autoimmune blistering disorders like MMP include corticosteroids combined with mycophenolate mofetil or cyclophosphamide to suppress the immune system. MMP often progresses despite treatment leading to development of permanent scarring and disability in patients, Rituximab therapy given as per lymphoma protocol can be versatile treatment choice for such patients.



Abstract N°: 1248

paraneoplastic pemphigus : a 10 year retrospective study

Hajar El Bokhari[1], Fouzia Hali[1], Bouchra Baghad[1], Soumia Chiheb[1]

[1]ibn rochd university hospital, dermatology and venerology

Introduction & Objectives:

Paraneoplastic pemphigus (PNP) is a rare autoimmune disease accounting for 3-5% of all pemphigus cases. It was first described in 1990 by Anhalt and colleagues as a bullous disease associating erosive stomatitis, polymorphic eruption and known or revealed neoplasia at the time of PNP diagnosis. We report a series of cases.

Materials & Methods:

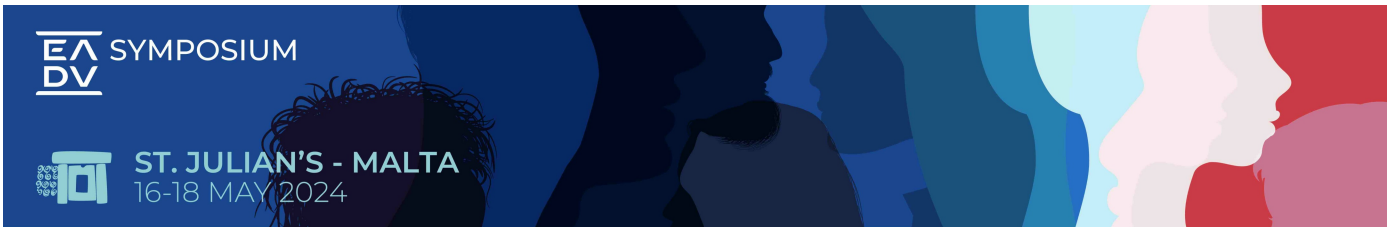
Retrospective study of PNP cases hospitalized in the dermatology department over a 10-year period from 2013 to 2023.

Results:

A total of eight patients were included in the study (5 men and 3 women). Mean age was 70.37 years (59-102 years). The mean duration of evolution before diagnosis of pemphigus was 11 months (1-60 months). The site of onset was cutaneous in 7 patients and mucosal in 2, and the combination of cutaneous and mucosal involvement was found in 3 patients. Cutaneous involvement was polymorphous, with flaccid bullae and post-bullous erosions (6 cases), tense bullae (1 case), lichenoid lesions (1 case), vesiculobullous lesions (2 cases) and erosive cheilitis (3 cases). These cutaneous lesions were located on the trunk (n=8), limbs (n=4), face (n=2), folds (n=2) and scalp (n=2), the skin surface involved was (>50%) in 3 patients, (20 -50%) in 2 patients and (<5%) in 2 patients, mucosal involvement was oral in 3 patients, genital in 2 patients and conjunctival in 2 patients. Histological findings were available in 4 patients, showing acantholytic suprabasal detachment with C3 and IgG interkeratinocyte deposits, and indirect immunofluorescence showed antibodies to intercellular substance in all patients. The neoplasms found in our series were breast (3 cases), prostate (2 cases), lung (1 case), larynx (1 case) and chronic lymphocytic leukemia (1 case). Neoplasia was diagnosed before the onset of pemphigus in 5 patients and after pemphigus in 3 patients. The prescribed treatment was oral corticosteroid therapy (7cases), azathioprine (2cases) and treatment of the neoplasia, which included chemotherapy in 3 patients, radio-chemotherapy in 2 patients and surgery in one patient. Progression was good in 6/8 and 2 patients were lost to follow-up.

Conclusion:

Paraneoplastic pemphigus (PNP) is a distinct autoimmune blistering disease that can affect multiple organs other than the skin. It occurs in association with certain neoplasms, among which lymphoproliferative diseases are most commonly associated. Contrary to the literature, PNP in our series was associated more with solid cancers than with hemopathies in particular breast cancer in women and prostate cancer in men. Therapeutic management combines the management of pemphigus and treatment of underlying neoplasia.



Abstract N°: 1264

Challenges in Diagnosis and Management of Generalized Morphea: A Case Report

Ivânia Soares[1], Inês Pereira Amaral[1], Madalena Pupo Correia[1], Luís Soares-Almeida[1, 2], Paulo Filipe[1, 2]

[1]Hospital de Santa Maria, Lisboa, Portugal, [2]Lisbon School of Medicine, Lisboa, Portugal

Introduction & Objectives:

Generalized morphea is a rare form of localized scleroderma characterized by widespread skin thickening and hardening. It is considered rare due to its low incidence compared to other forms of scleroderma. Despite its rarity, generalized morphea is important to recognize and understand due to its potential to cause significant disability and disfigurement. The condition can impact various organs and systems beyond the skin. Early diagnosis and appropriate management are crucial to prevent complications and improve outcomes for affected individuals. Additionally, research into the underlying mechanisms of generalized morphea and the development of effective treatment strategies are essential to better manage this condition and improve the quality of life for patients.

Materials & Methods:

Database and clinical reports from our institution.

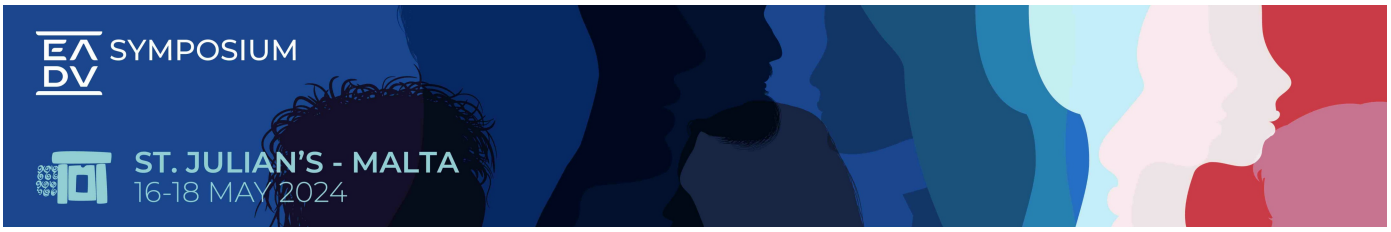
Results:

We present the case of a 34-year-old woman, with Fitzpatrick skin type IV, and known comorbidities of depressive syndrome treated with sertraline. She was referred from internal medicine to dermatology emergency with erythematous-violaceous, infiltrated, poorly defined plaques of varying sizes (ranging from 5 cm to 30 cm in the largest axis) disseminated on the back, abdomen, upper limbs, and bilateral mammary region, evolving over one year. The lesions had a progressive and additional onset without associated symptoms. The patient denied fever, chest pain, arthralgia, fatigue, Raynaud's phenomenon, or familial mucocutaneous diseases. On physical examination, besides the described plaques, brownish patches suggestive of post-inflammatory hyperpigmentation were observed in the initially affected areas.

A skin biopsy histopathological examination was consistent with morphea, and analytically, positive ANA autoantibody levels were noted, excluding other autoimmune diseases associated with morphea, namely thyroiditis and diabetes mellitus. With the diagnosis of generalized morphea assumed, she was evaluated by Rheumatology, which ruled out systemic sclerosis and initiated therapy with prednisolone 20 mg in a progressive reduction scheme and methotrexate 12.5 mg/week. After starting treatment, there was apparent stabilization of the dermatosis, with some plaques evolving into patches and no new lesions appearing to date. However, it is known from the literature that the response to treatment is limited, and the disease can be persistent, hence follow-up is crucial.

Conclusion:

This case underscores the challenges in diagnosing and managing morphea, particularly its generalized form. The treatment initiated aimed at halting disease progression and achieving stabilization, acknowledging the limited response to therapy highlighted in existing literature. Long-term follow-up is essential to monitor disease activity and response to treatment, as morphea can be chronic and recurrent. Further research into more effective therapeutic approaches is warranted to improve outcomes for patients with generalized morphea.



Abstract N°: 1344

efficacy of oral tofacitinib in alopecia areata,alopecia totalis and alopecia universalis

Shahmoona Faisal*[1]

[1]Hamdard medical university Taj medical complex, Dermatology, Karachi, Pakistan

Introduction & Objectives: Tofacitinib, an effective JAK inhibitor, has gained increasing interest in recent years among dermatologists for the management of refractory alopecia areata. One of the most prevalent autoimmune diseases is alopecia areata (AA), a kind of non-scarring alopecia. Variants that are persistent or severe have a major detrimental effect on the patient's quality of life and may disfigure them. Therefore, the purpose of this study was to evaluate the effectiveness of oral tofacitinib in treating alopecia areata, alopecia totalis, and alopecia universalis.

Materials & Methods: This interventional study was conducted at Hamdard medical university Taj medical complex using a non-probability consecutive sampling technique. The ethical approval was obtained from the Institutional Review Board. The duration of the study was about 6 months after approval of synopsis. The sample size was 50. All patients of both genders diagnosed with alopecia areata, alopecia totalis, and alopecia universalis, aged 5 years to 50years, and who were fit for treatment with oral tofacitinib, were included in the study. After the evaluation of patients, a 5 mg once daily dose in pediatric group and twice daily dose of oral tofacitinib in adults was started. Assessment of treatment response was done at 8 weeks, 12 weeks, and 24 weeks after the start of treatment. Improvement was assessed by a change in the Severity of Alopecia tool (SALT) from the baseline. SPSS version 23.0 will be used for data analysis. Quantitative variables were presented as mean and standard deviation, while qualitative variables were recorded as frequency and percentages. Chi-square test was applied to compare the scores of SALT at8, 12, and 24 weeks percentage change in SALT score from baseline.

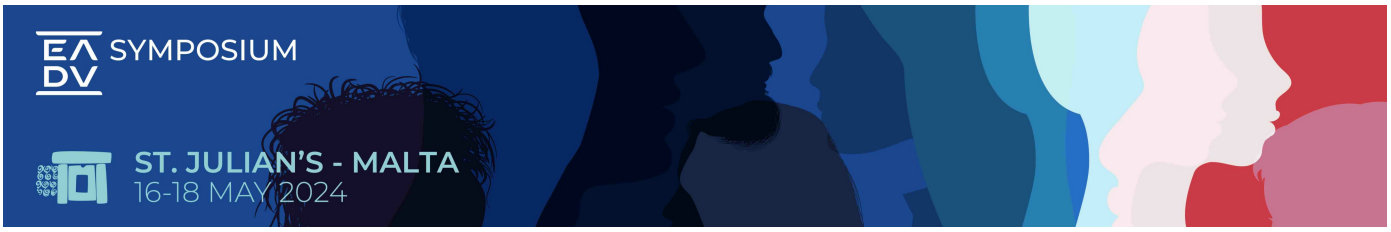
Results: A total of 50 patients with alopecia areata, who were or are being treated with tofacitinib. The mean age of the patients was 25.64 ± 11.77 years. Of them, 23(46.0%) were males, and 27(54.0%) were females. The majority of the patients, 39(78.0%) had alopecia areata, with 9(18.0%) patients having alopecia universalis, and 2(4.0%) having alopecia totalis. The mean duration of the disease was 44.94 ± 64.77 months, and the mean duration of therapy was 7.42 ± 2.45 months. The mean (SD) pretreatment scalp hair loss was 61.88 ± 24.08 %, and the mean scalp involvement at 24-weeks follow-up was 11.06 ± 24.85%. Fortunately, there were no side effects reported in any patient. The mean regrowth rate was 88.90 ± 24.52%. Moreover, a statistically significant difference was observed between the changes in SALT score from baseline and the scores of SALT at 8, 12, and 24 weeks ($p < 0.001$).

Conclusion: This study concluded that administration of oral tofacitinib to alopecia areata patients had significantly improved hair growth. Additionally, it has been revealed to be a potentially effective and safe therapy for the management of severe, refractory disease because no side effects were observed.

Keywords: Autoimmune hair disorder, Alopecia areata, Tofacitinib

Authors: Dr Syeda Shahmoona Tirmizi(FCPS)

Tirmizi SS, (Associate professor dermatology Hamdard medical university Taj medical complex)



Abstract N°: 1368

Perforating Dermatoses: Presentation of a Case of Reactive Perforating Collagenosis Associated with Diabetes

Meryame Hammouch[1], Fatima-Zahra El Fatoiki[1]

[1]CHU ibn rochd, dermatology, casablanca, Morocco

Introduction & Objectives:

Perforating dermatoses comprise four distinct entities: elastosis perforans serpiginosa, reactive perforating collagenosis (RPC), perforating folliculitis, and Kyrle's disease. RPC is characterized by transepidermal elimination of collagen fibers and is often associated with superficial skin trauma. Two types of RPC are distinguished: a rare hereditary form that manifests early in childhood, and a more common acquired form that occurs in adulthood and is often associated with other conditions such as diabetes, renal insufficiency, solid tumors, lymphomas, and AIDS. Here, we present a case of RPC associated with diabetes.

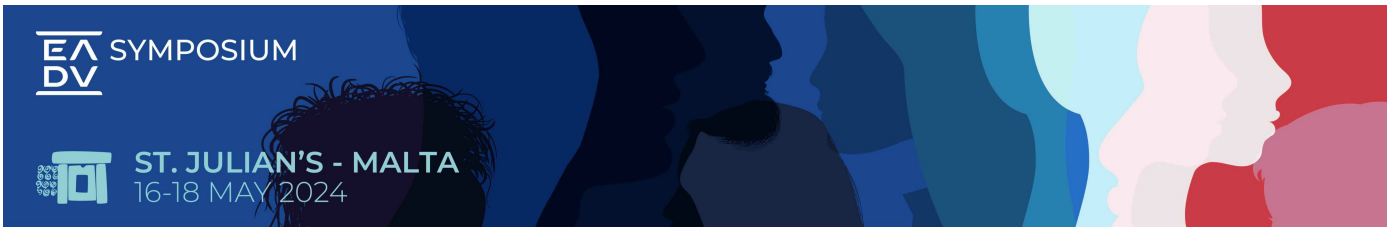
Case report:

A 60-year-old patient, hypertensive and type II diabetic under treatment, presents with pruritic lesions persisting for three months. The lesions initially appeared on the abdomen and extended to the upper and lower limbs. Dermatological examination reveals keratotic papulo-nodular lesions, approximately 1 cm in diameter, umbilicated, and confluent in some areas. Crusted, adherent, and necrotic central depressions along with a circular erythematous border are also observed. The skin lesions are generalized, with a predilection for the extensor surfaces of the limbs. Histological examination confirms the diagnosis of reactive perforating collagenosis, showing epidermal depression with the expulsion of collagen fibers to the surface.

Conclusion:

Acquired perforating collagenosis is an extremely rare disease, and its exact incidence is unknown. It can affect both men and women, with a slight predominance in men. It typically occurs around the average age of 57. This condition is often associated with diabetes, chronic renal failure, and/or hyperuricemia. When pruritic, keratotic papular skin lesions are present in a patient with pre-existing associated conditions, the diagnosis of acquired perforating collagenosis may be considered. However, only a correlation between histological and clinical findings can confirm the diagnosis. The pathogenesis of hereditary RPC remains unknown, while that of acquired RPC in diabetics is better understood and is thought to be related to collagen glycation.

Pruritus is the primary symptom reported in RPC, and its management is essential to prevent further dissemination through the Koebner phenomenon.



Abstract N°: 1426

the association between the single nucleotide polymorphism of st18 gene and the severity of pemphigus vulgaris in vietnamese population

Long Phan*[1], Tam Huynh[1]

[1]Pham Ngoc Thach University of Medicine, Department of Dermatology, Ho Chi Minh City, Viet Nam

Introduction & Objectives:

Pemphigus vulgaris (PV) is a serious autoimmune blistering disease caused by autoantibodies directed against antigens on the surface of keratinocyte adhesion. Recently, ST18 polymorphism has been proven to play a role in apoptosis and inflammation disorders, two processes of direct relevance to the pathogenesis of PV. Our study aimed to investigate the relationship between ST18 gene single nucleotide polymorphism (SNP) with PDAI score and severity of PV among Vietnamese people.

Materials & Methods:

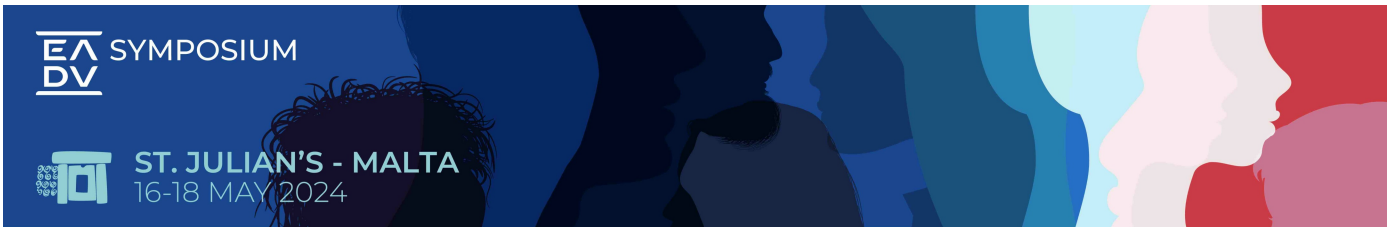
A total of 34 PV patients with diagnoses confirmed by clinical manifestations, histopathology, and direct immunofluorescence from February 2023 to October 2023. The genotype for two ST18 SNPs rs2304365 and rs4074067 were determined by Sanger sequencing.

Results:

The results indicated that the patients carrying the minor allele (T) of rs2304365 had higher PDAI scores and more severe disease than the major allele (C). No remarkable association between the SNP rs4074067 and PV severity.

Conclusion:

Thus, the present study confirmed the relationship between the SNP variations of ST18 gene may predict the severity of PV disease.



Abstract N°: 1427

the association between the single nucleotide polymorphism of st18 gene and the severity of pemphigus vulgaris in vietnamese population

Long Phan*[1], Tam Huynh[1]

[1]Pham Ngoc Thach University of Medicine, Ho Chi Minh City, Viet Nam

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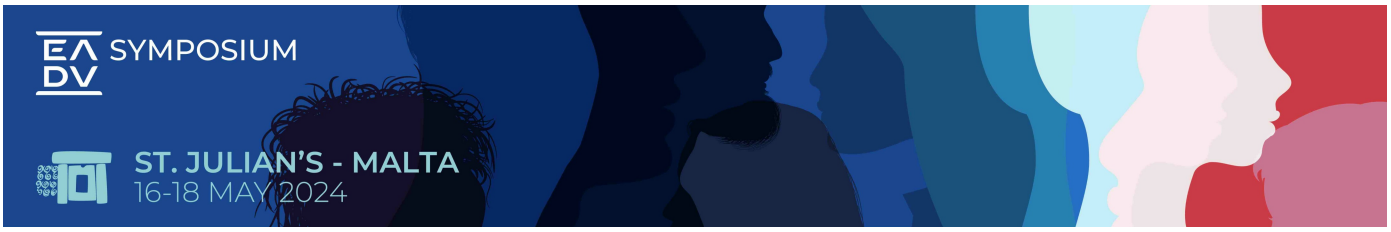


Abstract N°: 1441

Epidemiological-clinical profile of pemphigus in Algiers

Iberraken Lydia[1], Billel Merrouche[1], Hamchaoui Farida[1], Houria Sahel[1]

[1]Chu Maillot, Dermatolgy, Algiers, Algeria

Introduction & Objectives:

Pemphigus is a rare autoimmune bullous dermatosis. The aim of this study was to describe the epidemiological, clinical, therapeutic characteristics of patients with pemphigus in Algiers.

Materials & Methods:

Retrospective study, based on the hospitals records of patients hospitalized for pemphigus between January 2020 and June 2023 at the Dermatology Department of Bab El Oued University Hospital, in Algiers.

Results:

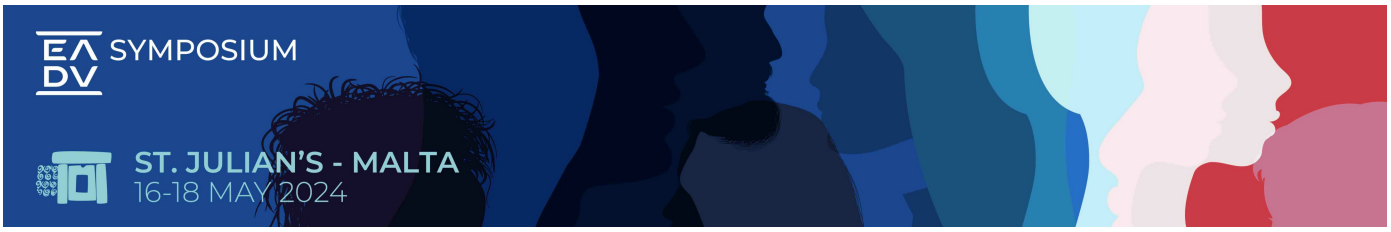
27 cases were collected, including 18 women and 9 men (sex ratio: 0.5). The mean age was 50 years with extremes (16-75 years). More than half the patients had a medical history. No medications or autoimmune diseases were found. There were 21 cases of deep pemphigus (vulgar: 19 cases, vegetative: 2 cases), 4 cases of superficial pemphigus (seborrheic), 1 case of paraneoplastic pemphigus, 1 case of pemphigus herpetiformis. There were no cases of induced pemphigus. Pruritus was present in 59.3% of cases. Nikolsky's sign was positive in 21 patients. The mode of onset was mucosal in 2/3 of patients. Involvement of the oral mucosa was found in 20 patients, the genital mucosa in 9, the ocular mucosa in 10, and the esophageal mucosa in 4. Nail involvement was found in two patients. Severity was assessed by PDAI score, with moderate forms accounting for 70% and severe forms for 30%.

Histology was characteristic except in 1 patient, IFD was positive in 88.9% and inconclusive in 11.1%. IFI was performed in 18 patients: positive in 90%.

Oral corticosteroid therapy was prescribed at a dose of 0.5 to 1.5 mg/kg/d prednisone in all patients except one, in whom local corticosteroid therapy was sufficient. The addition of azathioprine was necessary in 70% of patients. Rituximab was used in 5 patients, and IV immunoglobulin in 2. The average hospital stay was 2 months, with a favorable outcome in 85% of cases. 4 deaths were noted. The main complications observed were: infections (16 cases, including 7 herpetic superinfections and 6 bacterial superinfections), cortico-induced diabetes (16 cases), and one case of deep-vein thrombosis causing pulmonary embolism and resulting in the patient's death.

Conclusion:

This study of Algerian pemphigus reveals an epidemiological profile different from the spectrum of North African pemphigus.



Abstract N°: 1448

Gliptin-Associated and Non-Gliptin-Associated Bullous Pemphigoid Patients' Disease Characteristics and Treatment Responses: A Retrospective Case-Control Study

Erhan Topal*[1], Ferhan Bulut Demir[1], Tulin Ergun[1]

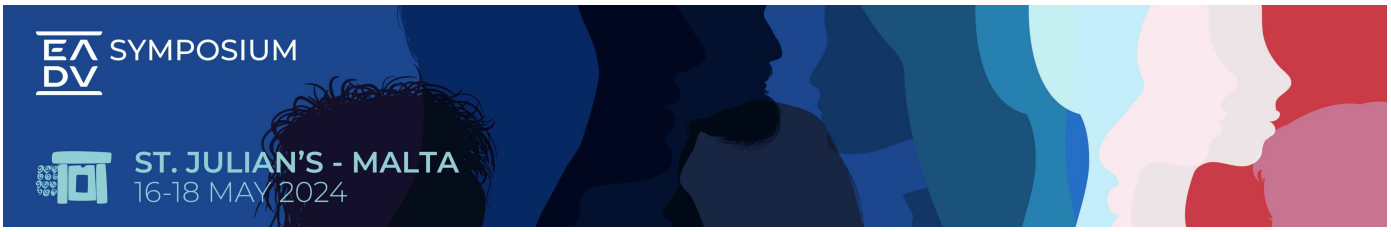
[1]Marmara University School of Medicine , Dermatology, Türkiye

Introduction & Objectives: Bullous Pemphigoid (BP) is an autoimmune blistering disease characterised by subepidermal blister formation. It is commonly observed in the elderly. Although BP can develop spontaneously, it can also be triggered by certain drugs. Over the past decade, there is mounting evidence that BP is associated with certain antidiabetic drugs, namely dipeptidyl peptidase inhibitors (DPPis), also known as gliptins. It is found that drug related BP is most commonly triggered by DPPis. Moreover BP associated with DPPi may differ from spontaneously occurring BP in terms of clinical manifestations and treatment response. In cases associated with DPPi use, it is seen that erythema may be less pronounced, blisters may be smaller and antibodies against a different epitope of BP180 may develop. However data on the differences between BP associated with DPPi and classical BP are extremely limited. Hence, this study aims to investigate the demographic, clinical, laboratory findings, and treatment responses of patients with gliptin-associated (GA) and non-gliptin-associated (NGA) BP.**

Materials & Methods: Patients diagnosed with BP and followed up at our clinic between January 2011 and June 2023 were retrospectively screened. Cases were divided into two groups: those with gliptin-associated BP and those with non-gliptin-associated BP. The GA group included patients using gliptin derivatives and the NGA BP group included all BP-diagnosed patients without a history of gliptin use. The groups were compared in terms of age, gender, comorbidities, presence of itching and mucosal involvement, presence of eosinophilia, use of treatment agents (immunosuppressive or biological agents), and clinical course (complete/near-complete remission, partial remission, chronic disease).**

Results: A total of 118 patients were included, comprising 46 patients classified as GA BP and 72 patients classified as NGA BP. The mean ages of the NGA BP and GA BP cases were 73.97 ± 14.27 and 70 ± 8.82 years, respectively, and this difference was statistically significant ($p = 0.017$). However, gender, presence of itching, eosinophilia, and treatment agents used were similar between the groups. Although not statistically significant, both mucosal involvement (17.4% vs. 29.2%) and use of immunosuppressive agents (21.7% vs. 29.2%) were lower in GA BP. When the two groups were compared in terms of clinical course, the proportion of patients with complete, near-complete, or partial remission was 82.2% (37/45) in the GA BP group and 71.9% (51/71) in the NGA BP group ($p > 0.001$).

Conclusion: In our study, consistent with the literature, significant differences in age and clinical course were observed between GA BP and NGA BP cases. The age at diagnosis of patients with GA BP was 3 years lower than that of NGA BP patients. Additionally these patients showed significantly better clinical course and treatment response compared to NGA BP patients. The main limitations of our study were its retrospective and single-centre nature and the absence of universally accepted criteria for evaluating clinical response in BP. Nevertheless our study is one of the few studies in our country examining the relationship between gliptin use and BP, with the largest patient group. Identifying the relationship between gliptin derivatives and Bullous pemphigoid in diabetic patients is among the priority research topics. Additionally, understanding the pathophysiology and at-risk population of this relationship is crucial.



Abstract N°: 1462

Post Covid-19 rRapid onset and expansion of vitiligo with subsequent stabilization post Covid-19 disease

Despoina Exadaktylou[1], Kanella Kalapothakou[2], Panagiota Chroni[2], Eleni Keramida[3], Pa Korkolopoulou[4], Ni Katsoulas[4], Panagiota Spyridonos[5], Georgios Gaitanis[6]

[1]Western Attica General Hospital "Aghia Varvara" , Department of Dermatology,, Aghia Varvara, Greece,[2]Western Attica General Hospital "Aghia Varvara" , Department of Dermatology,, Agia Varvara, Greece, [3]Western Attica General Hospital "Aghia Varvara" , Department of Dermatology,, Agia Varvara , Greece, [4]School of Medicine, National & Kapodistrian University of Athens, First Department of Pathology, Athens, Greece, [5]Faculty of Medicine, School of Health Sciences, University of Ioannina, Department of Medical Physics, Ioannina, Greece, [6]Faculty of Medicine, School of Health Sciences, University of Ioannina, Department of Skin and Venereal Diseases, Ioannina, Greece

Introduction & Objectives:

COVID-19 infection has been associated with cutaneous autoimmune-mediated disorders including vitiligo.

The pathogenesis is poorly understood but the spike protein of SARS-CoV-2, seems to be of central role as it binds to ACE receptors in target cells.

Materials & Methods:

A 60-year-old man of Egyptian origin presented with a three- month history of extensive vitiligo on face, torso, buttocks, upper and lower limbs covering >30% body surface area (BSA). According to the patient history, the achromic/hypochromic lesions appeared three weeks after a mild Covid-19 disease and rapidly progressed within few days reaching the BSA area at presentation. Subsequently the disease stabilized without appearance of new lesions or expansion of the initial ones. Our patient had no family or personal history of vitiligo and reported only Hashimoto's thyroiditis.

Laboratory examinations including complete blood count, ESR, Biochemical testing (serum glucose, urea, creatinine, AST, ALT, γGT, ALP, etc.), Immunological testing (ANA, RF, C3, C4, c-ANCA, p-ANCA), thyroid tests (TSH, fT3, fT4) and coagulation tests, were within normal limits and only serum angiotensin converting enzyme (SACE) was increased at 60 nmol/ml (normal range < 40 nmol/mL). Clinical photos to record the expansion of the disease were taken.

Results:

The histology confirmed the clinical diagnosis of vitiligo. Our patient did not wish to undergo any treatment and was subsequently followed up every three months for one year without any changes in the lesions or the SACE levels. The stabilization of the disease was confirmed with image analysis.

Conclusion:

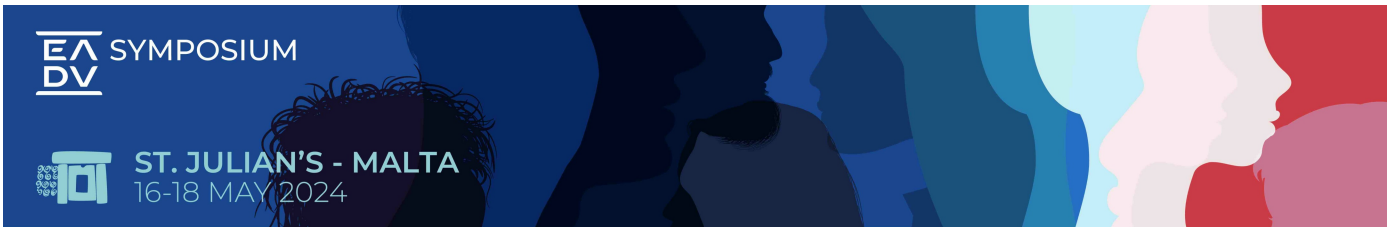
Vitiligo is an autoimmune dermatosis and has been reported following viral illnesses such as HIV, CMV, hepatitis C, and recently SARS-CoV-2 infection.

The pathogenesis of COVID-19-associated vitiligo seems to involve the SARS-CoV-2 spike protein that binds to angiotensin converting enzyme (ACE) receptors in target cells. In human skin the renin angiotensin system is completely expressed, angiotensin is locally synthesized while the relevant receptors are also expressed in epidermal cells, including melanocytes.

Likewise, ACE gene polymporhisms in Egyptians have been reported in association with vitiligo.

In our report, we present a patient of Egyptian origin with no personal or family history of vitiligo, who developed rapidly widespread vitiligo post SARS-CoV-2 infection with elevated SACE serum levels.

Further research is needed to clarify the complex immunologic pathways of vitiligo and the association of autoimmune disorders with viral infections.



Abstract N°: 1463

Understanding the immune interplay: psoriasis and its associated autoimmune diseases

Alexandra Petruta Savu[1, 2], Elena-Daniela Serban[1, 2], Costina Cristiana Mutu[2], Cezara-Diana Vaida[2], Stefana Bucur[1, 2], Maria Magdalena Constantin[1, 2]

[1]Carol Davila University of Medicine and Pharmacy, Bucharest, Romania,[2]Colentina Hospital, IInd Department of Dermatology, Bucharest, Romania

Introduction & Objectives:

Psoriasis, a prevalent chronic inflammatory skin conditions, often coexists with other autoimmune and autoinflammatory diseases, amplifying the complexity of its clinical management.

Materials & Methods:

Case presentation and literature review.

Results:

In this regard, we present a series cases of psoriasis with other autoimmune and autoinflammatory disorders to highlight diagnostic challenges, therapeutic considerations, and clinical implications of managing these conditions together.

Case 1: A 51-year-old male with a history of ulcerative colitis (UC) developed psoriasis vulgaris. Initially treated with mesalamine and corticosteroids for UC, partial remission was achieved. Due to worsening of UC and the development of psoriasis vulgaris, ustekinumab therapy was initiated, leading to rapid improvement in both gastrointestinal and cutaneous symptoms

Case 2: A 34-year-old female with history of psoriasis vulgaris developed non-segmental vitiligo. Methotrexate was added to her treatment plan due to limited response and exacerbation of both conditions. Significant improvement in both psoriasis and vitiligo lesions was observed during follow-up after initiating methotrexate therapy.

Case 3: A 51-year-old female initially diagnosed with psoriasis vulgaris, who later presented with symptoms suggestive for hidradenitis suppurativa (HS). Methotrexate therapy was ineffective, so secukinumab was initiated, resulting in significant improvement in both conditions.

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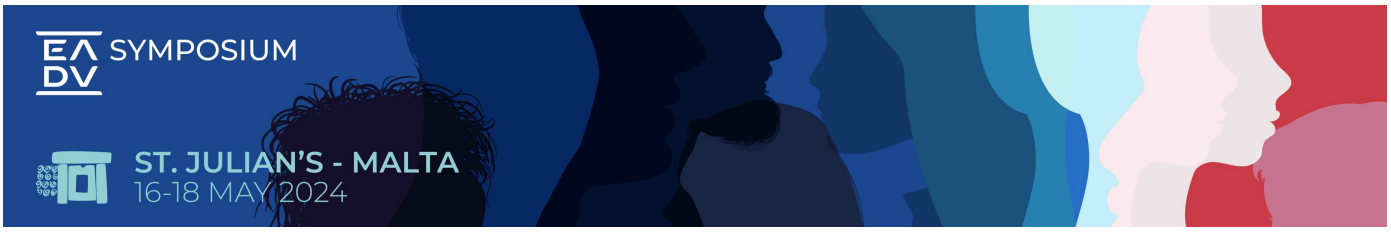
Case 4: A 79-year-old female initially diagnosed with psoriasis vulgaris and psoriatic arhritis mutilans, and later diagnosed with systemic lupus erythematosus. Due to the association of this disorders, it was decided to initiate treatment with methotrexate and the patient experienced significant improvement in both joint symptoms and cutaneous manifestations.

Case 5: A 54-year-old patient with history of non-segmental vitiligo and guttate psoriasis (without active lesions) developed discoid lupus erythematosus. Given this progression, treatment with plaquenil was initiated to address the emerging lupus discoid.

Conclusion:

By presenting these cases, we aim to highlight the complex interplay between psoriasis and other autoimmune or autoinflammatory diseases, emphasizing the importance of comprehensive assessment and personalized treatment

approaches. Furthermore, this series underlines the importance of further research to elucidate the shared genetic, immunological, and environmental factors contributing to the co-occurrence of these conditions, with the ultimate objective of improving patient outcomes and quality of life.



Abstract N°: 1484

Immunobullous Diseases Among The Geriatric Population In Lagos Nigeria: Diagnostic Challenges And Management.

Adaeze Ikebudu*[1]

[1]Lagos, Internal medicine, Lagos, Nigeria

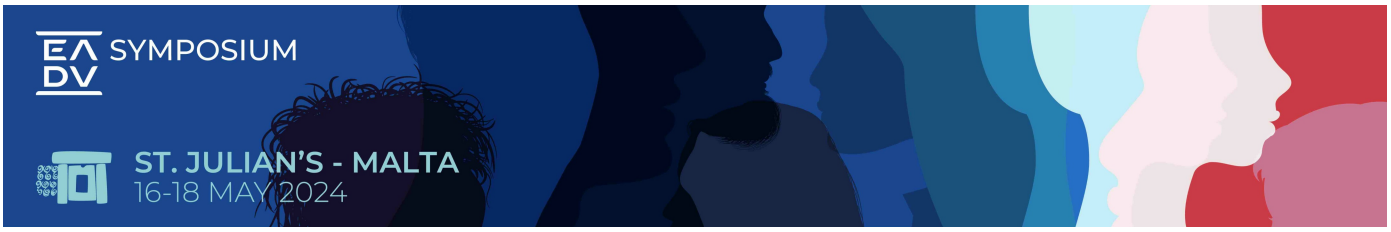
Introduction & Objectives: Studies from Nigeria revealed that a low percentage (2%) of geriatric individuals have immunobullous diseases probably on account of a low index of suspicion, and lack of access to quality health care. Geriatric individuals are prone to immunobullous diseases that may be linked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

The objective is to document the disease patterns and the frequency of occurrence among the geriatric population.

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

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

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



Abstract N°: 1519

Effects of cold argon plasma treatment in patient with recalcitrant pemphigus vulgaris

Danijela Milcic[1, 2], Marko Demenj[1], Marija Malinić[1], Maja Vilotijevic[1], Marija Tomanovic[1], Mirjana Milinkovic[1, 2]

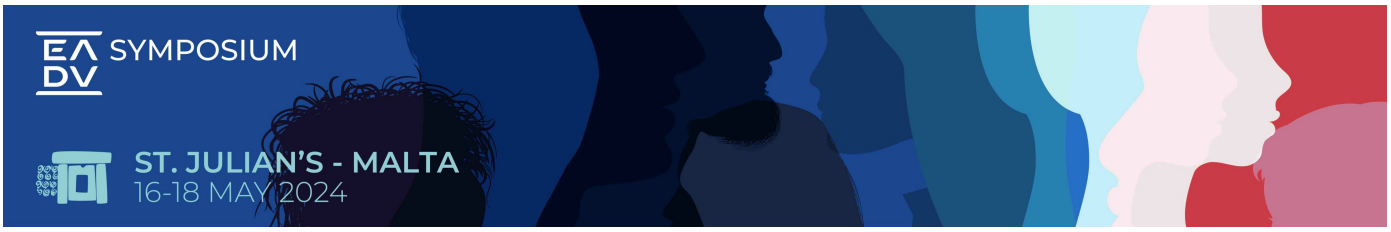
[1]University Clinical Center of Serbia, Clinic of Dermatology and Venereology, Belgrade,[2]University of Belgrade, Faculty of medicine, Belgrade, Serbia

Introduction & Objectives: Pemphigus implies a group of rare and severe autoimmune blistering diseases involving the skin and mucous membranes. The treatment includes the use of systemic corticosteroids and immunosuppressive agents, intravenous immunoglobulins, plasmapheresis, as well as monoclonal antibodies directed to CD20. However, there are the cases of recalcitrant lesions, which represent a unique challenge for the treatment. Cold atmospheric plasma (CAP) treatment has antimicrobial, tissue-stimulating, blood-flow-stimulating and pro-apoptotic effects. Argon is frequently used working gas during this treatment. Based on good therapeutic results, CAP can be used as effective therapy for disinfection and treatment of chronic ulcerations, as well as the treatment of tumors, actinic keratosis, scars, atopic eczema etc.

Materials & Methods: We present a 34-year old Caucasian woman with oral and genital blisters and erosions. The disease started after a serious family tragedy, the suicide of a sister. The patient was also developed serious mental disorder. The diagnosis of pemphigus vulgaris was established with standard tests. We introduced combined pulses therapy with dexamethasone and cyclophosphamide (C) in high doses, and oral prednisolone and low doses of C between pulses. After the first pulse therapy the patient had COVID-19 infection, and pemphigus progressed rapidly on the skin and mucous membranes. We introduced intravenous immunoglobulin (IVIG) combined with pulses dexamethasone therapy and oral azathioprine, while C was excluded because of hepatotoxicity. Plasmapheresis was performed as well twice, but it was also excluded due to side effects. The patient was hospitalized for four months and during treatment she developed severe complications like skin infections and sepsis, pathological fractures of nine vertebrae, pulmonary embolism… The skin lesions improved significantly, except for erosions on the face that did not epithelize for the next 6 months. Then, we introduced treatment with argon plasma, applying it on areas of 4–5 cm2, which made up in total 9 treatment zones on the face erosions. We preformed 3-minute treatment for every one of the nine treatment zones, 3 times weekly during 4 weeks.

Results: We achieved fantastic therapeutic effects, epithelization of about 80% of extensive erosions of the face, and a remaining 20% epithelialized during next few weeks.

Conclusion: Afterwards we continued therapy with low doses of prednisolone and azathioprine, and our patient is still in remission of the disease.



Abstract N°: 1589

Churg-strauss syndrome : a vasculitis often forgotten by dermatologists

Yassine Berrada[1], Younes Tamim[1], Meriam Meziane[1], Nadia Ismaili[1], Laila Benzekri[1], Karima Senoussi[1]

[1]Centre hospitalier Ibn Sina, Dermatologie venerologie, rabat

Introduction :

Eosinophilic granulomatosis with polyangiitis (EGPA), or Churg-Strauss syndrome, is characterized by necrotizing granulomatous inflammation, rich in eosinophils, mainly affecting small to medium-sized vessels.

We report the case of a patient diagnosed with Churg-Strauss syndrome presenting with urticarial plaques.

Observation :

A 60-year-old woman presented with complaints of recurrent episodes of purpuric urticarial lesions with slight pruritus, localized to the neck, and evolving for 4 months, leaving behind residual hyper-pigmented macules.

Past surgical history includes a nasal polypectomy 20 years ago.

Past medical history is significant for ischemic cardiopathy, first diagnosed 6 years ago, and late onset asthma.

The clinical picture was accompanied by febrile digestive symptoms including abdominal pain, nausea, vomiting, and intermittent diarrhea. Other cutaneous symptoms included painful subcutaneous nodules, oral and genital ulcers, Raynaud's phenomenon, and chronic paresthesia of the extremities.

Investigations including a complete blood count, enteroscopy, and jejunal biopsy revealed persistent hypereosinophilia (>4000/uL), exudative enteropathy, and chylous ascites without an identified cause. A skin biopsy showed leukocytoclastic vasculitis with eosinophils, confirming urticarial vasculitis. ANCA testing was negative, while a nasal sinus CT scan showed mucosal polyps. EMG revealed axonal sensorimotor polyneuropathy.

The diagnosis of EGPA was confirmed by clinical, biological, and histopathological results. The patient was treated with corticosteroids combined with cyclophosphamide with good outcomes.

Discussion :

Cutaneous involvement in Churg-Strauss syndrome is characterized by the appearance of vasculitic purpura, urticarial eruptions, subcutaneous nodules, and ulcers, occurring in about 2/3 of patients, reminiscent of the symptoms described in our patient.

The diagnosis of EGPA relies, in the presence of vasculitis signs, on four out of six following characteristics: asthma, eosinophilia, neuropathy, pulmonary infiltrates, paranasal sinus abnormalities, and eosinophilic vasculitis.

Treatment typically involves glucocorticoids and immunosuppressants.

Conclusion:

Our experience with this patient illustrates an important message: to consider vasculitis in the presence of any fixed urticarial lesion because without treatment, the outcome can be fatal.

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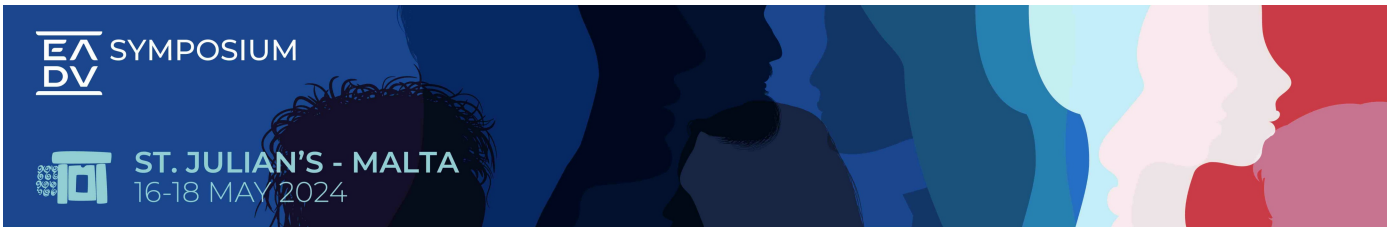

Abstract N°: 1643

Localized bullous pemphigoid induced by radiation therapy: A case report

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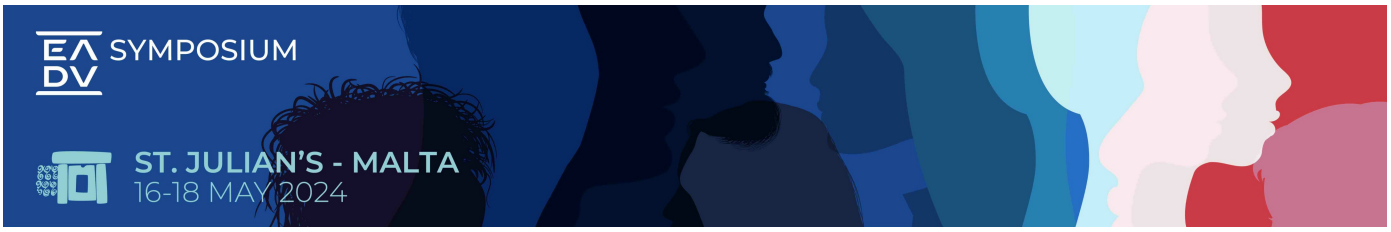
Introduction & Objectives: Bullous pemphigoid is an autoimmune bullous disease that mainly affects older people. It is characterized by bullae formation resulting from the antigen-antibody reaction when autoantibodies bind to the hemidesmosome complex of the skin in the basement membrane zone.The exact cause of pemphigoid is unknown, but it may be induced by certain drugs and rarely by local burns, ultraviolet phototherapy and radiotherapy.

In this report we present a case of radiation-induced bullous pemphigoid in erderly woman who was treated for vulvar squamous cell carcinoma.

Materials & Methods: A 77-year-old woman who was diagnosed with vulvar squamous cell carcinoma underwent radiotherapy for three months after vulvectomy. Five months later, she visited the hospital because of bullae on suprapubic area. Although she was treated with the topical corticosteroid cream, changes continued to persist and spread on the left upper leg and the erosion worsened. Then she was reffered to our department where laboratory tests, patohistological analysis and direct immunofluorescence (DIF) were taken. Patohistological analyisis show a subepidermal split with lymphocytes and lot of eosinophils. DIF examination of perilezional skin demonstrated linear IgG deposits and granulo-linear C3 deposits in the basement membrane zone. Patient was treated with short-term systemic corticosteroids and topical corticosteroids. After few days, the bullae spread ceased and the dosage od systemic corticosteroids was reduced. During the hospitalization, the lesion site was cleansed daily with saline and a hydrogel dressing was applied. She had a complete resolution of the bullae two months later and few postinflamatory hyperpigmentation were seen at the last follow-up.

Results: Radiation-induced pemphigoid is characterized by bullae formation at the irradiated site beyond which the bullae can spread. Although the association between radiotherapy and bullous pemphigoid has been well described, the underlying pathophysiology remains unconfirmed.

Conclusion: Multiple hypotheses have been described and one theory is that radiation-induced apoptosis of skin epithelial cells causes them to release BP 180 and BP 230 antigens, which are recognized by Langerhans cells and then started the production of autoantibodies. The development of bullous pemphigoid long time after radiation therapy seems to be explained by the “immunocompromised district” concept. There is an explanation for the occurrence of localized skin diseases secondary to local dysregulated immunity in the context of damaged areas which include irradiated skin. These condition cause an alteration of lymph circulation and damage to peripheral nerves which can alter the signaling of neuromediators. An abnormal neuroimmune interaction leading to infections or neoplasms, or leading to autoimmune disease such as bullous pemphigoid.



Abstract N°: 1646

A Case Report of Diagnostic and Treatment Challenges for Multicentric Reticulohistiocytosis

Neringa Borodicaite[1], Tadas Raudonis[1], Ruta Bleifertaite[2]

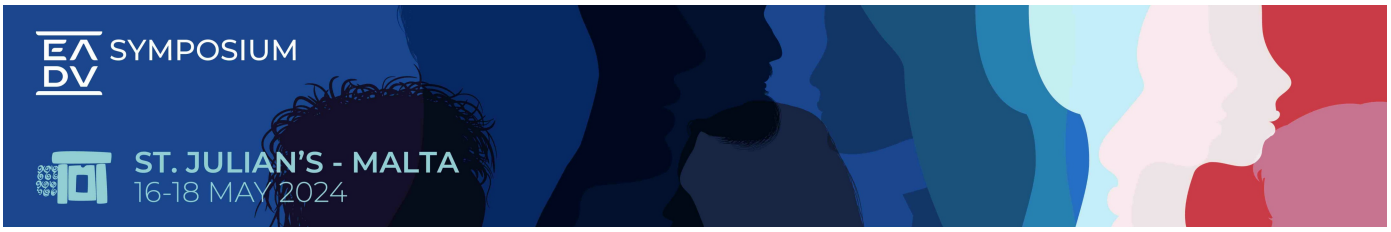
[1]Vilnius University, Centre of Dermatovenereology, Vilnius, Lithuania, [2]Vilnius University Faculty of Medicine, Vilnius, Lithuania

Introduction and Objectives: Multicentric reticulohistiocytosis (MRH) is a rare systemic disease which is associated with papulonodular skin lesions and rapidly developing arthritis. Objectives include outlining the 35 years old women's medical history, diagnostic challenges of MRH, and its subsequent management strategy.

Materials and Methods: In 2022, the patient experienced significant stress. In September 2023, she presented to a rheumatologist with swelling and pain in her knees, wrist and fingers, and reddish, tender, non-scaly papulae and nodules on her fingers and around the nails. The patient underwent extensive medical examination, which included broad spectrum laboratory investigations, Schirmer's test, nailfold videocapillaroscopy, lung CT scan and gas transfer test. She was inspected for immunodeficiency. Toxoplasmosis, hepatitis and HIV tests were performed. Biopsies from knee joint, skin, nasal polyps, minor salivary glands and trepanobiopsy were performed.

Results: Laboratory analysis revealed elevated anti-Ro52 antibodies, indicative of systemic autoimmune involvement. Immunodeficiency was ruled out. Lung CT scan and gas transfer test did not show any significant abnormalities. Schirmer's test did not show any abnormalities. Biopsies from knee joint, nasal polyps, minor salivary glands and trepanobiopsy yielded normal results. Further investigations ruled out infectious etiologies. In September 2023, initial skin biopsy showed features of skin sarcoidosis and the patient was referred to a dermatologist for the treatment. In December 2023, the patient was inspected by a dermatologist and a multicentric reticulohistiocytosis was suspected. Lesions pathohistology slides from the initial skin biopsy performed in September 2023 were checked again and a diagnosis of MRH was confirmed. The treatment plan for the patient consisted of injections of triamcinolone to the skin lesions, topical clobetasol ointment, oral methylprednisolone and oral hydroxychloroquine. During the control visit, the patient noted reduction of joint pain while skin lesion improvement was not significant.

Conclusions: The comprehensive diagnostic approach helped unveil the complexity of the patient's condition, leading to the final diagnosis of MRH. Difficulties making the right treatment plan were faced due to the rarity of the disease. Despite the challenges, the tailored management plan, including different forms of corticosteroids and oral hydroxychloroquine, showed positive outcomes in reducing the joint pain and slightly improving skin lesions. This case emphasizes the importance of a multidisciplinary approach in determining the right diagnosis and successfully managing rare skin diseases.



Abstract N°: 1681

Therapy-Resistent Mucous Membrane Pemphigoid Affecting the Oral Cavity in a Patient with a Hematological Malignancy: A Case Report.

Zdenka Šitum Čeprnja[1], Tina Gogić Salapić[1], Mara Drnas[1], Adela Markota Čagalj[1]

[1]University Hospital Centre Split, Dermatology and Venereology, Split, Croatia

Introduction & Objectives:

Mucous membrane pemphigoid disease (MMP) is a diverse set of vesiculobullous, chronic, autoimmune disorders with blister development that tend to affect various bodily mucous membranes. Constant eroding of the oral cavity, scarring of the conjunctiva, esophageal strictures, blockage of the airway, and vaginal mucosa scarring are major problems. Autoantibodies against basement membrane zone constituents, primarily targeting BP180 (type XVII collagen), laminin 332 (10%–20%), and type VII collagen (<5%), are the hallmarks of MMP.

Materials & Methods:

Results:

56-year-old man was referred to our Department of Dermatology and Venereology due to a sudden onset of blistering lesions on his trunk and extremities with expansion to the oral cavity, genital mucosa, palms, and soles. He also presented with eye redness and purulent eye discharge. His medical history revealed that he is receiving therapy for arterial hypertension and depression, as well as that he suffers from chronic lymphocytic leukemia, which until then did not require therapy. A biopsy was obtained from lesional and perilesional tissue and sent for histopathological examination and direct immunofluorescence. Correlation of clinical and histological features directed us toward the diagnosis of subepidermal blistering disorder.. Linear deposits of C3 and IgG were detected at the dermo-epidermal junction zone in direct immunofluorescence. Serum samples were tested by indirect immunofluorescence and revealed linear deposits of IgG at the dermal side (blister bottom) of the salt-split skin, and the diagnosis of MMP was confirmed. Systemic corticosteroid therapy (metilprednisolone 1 mg/kg) was used in combination with a corticosteroid-sparing agent (azathioprine 1.5 mg/kg) and with combined topical corticosteroid, antifungal, and antibacterial therapy, almost without any success. There was also an unsuccessful treatment with 5-day intravenous immunoglobulins treatment. Erosions inside the oral cavity were the lesions that caused the patient the most concern. Many complications had to be faced, such as fever, electrolyte imbalance, low albumins, feeding difficulties, weight loss, and depression worsening. Finally, hematologists revised the stage of the patient's chronic lymphocytic leukemia and decided to treat the underlying neoplasm with a Bruton tyrosine kinase inhibitor, ibrutinib. Not only did the total number of leukocytes decreased, but most of the mucous and skin lesions also healed.

Conclusion:

When treating MMP lesions, a multidisciplinary approach is frequently required for optimal results. In order to prevent long-term problems from this condition, it is very important to establish an early diagnosis and a successful treatment strategy that includes systemic or topical corticosteroid therapy along with additional therapeutic techniques such as periodontal therapy and appropriate oral hygiene practices. It is crucial to understand that, in the event that a dermatosis is resistant to therapy, one should constantly consider the possibility of underlying malignant disorders.

Abstract N°: 1691

Examination of BP180 and BP230 antibodies in Bullous Pemphigoid Patientsundergoing Rituximab Treatment

Kamran Balighi[1], Farbod Torkamand[1], Maryam Daneshpazhouh[1], Maryam Ghiasi[1], Hamidreza Mahmoudi[1], Srishti Mohapatra[2], Pooya Khanmohammad Beigi*[2]

[1]Razi Hospital, Tehran University of Medical Sciences. , Autoimmune Bullous Disease Research Centre, Tehran, Iran, [2]Misdiagnosis Association and Research Institute- MARI, Department of Dermatology, California, United States

Introduction & Objectives: Bullous Pemphigoid (BP) is the most common autoimmune blistering disease. Autoantibodies target BP230 and BP180 proteins. Immunosuppressants used to treat BP increase susceptibility to infections. The study aimed to examine the effect of rituximab on BP patients and to correlate increasing BP180 titers with disease severity scored using the Bullous Pemphigoid Disease Area Index (BPDAI).

Materials & Methods: This open-label, single-arm clinical trial examined 43 confirmed BP patients, who were identified through IgG/C3 direct immunofluorescent analysis, were considered for the study. G Powered software was used along with ANOVA for further statistical analysis.

Results:

Of the total patients (n=43), 15 (35%) were males and 28 (65%) females. The age range of the participants spanned between 33 and 94 years with a mean age of 62.70 years. Following BPDAI criteria, 8 individuals (18.6%) exhibited mild disease, 32 (74.4%) had moderate disease, and 3 (7%) had severe disease.

A significant positive correlation (r=0.336, p=0.014) was observed between serum BP180 levels and BPDAI scores (Fig 1a). BP230 levels showed no significant relationship with disease severity (r=0.109, p=0.243) (Fig 1b).

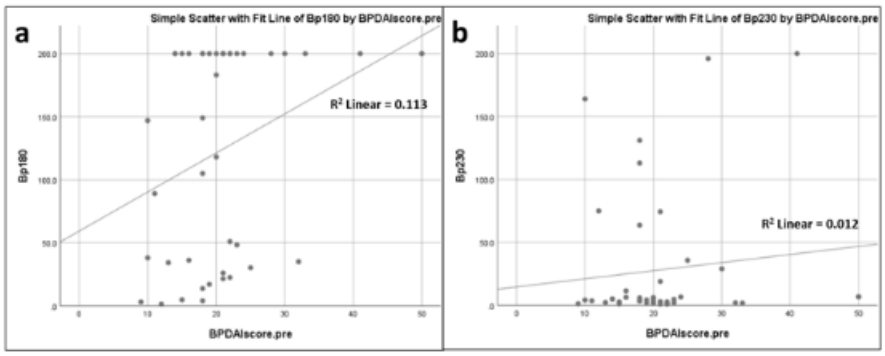


Fig 1. Association of disease severity with antigen levels in the serum.

Six months post treatment, a significant decrease in BPDAI scores (p=0.000) was observed. While the mild group witnessed the maximum change in the BP180 levels, the moderate groups showed the least change (Fig 2).

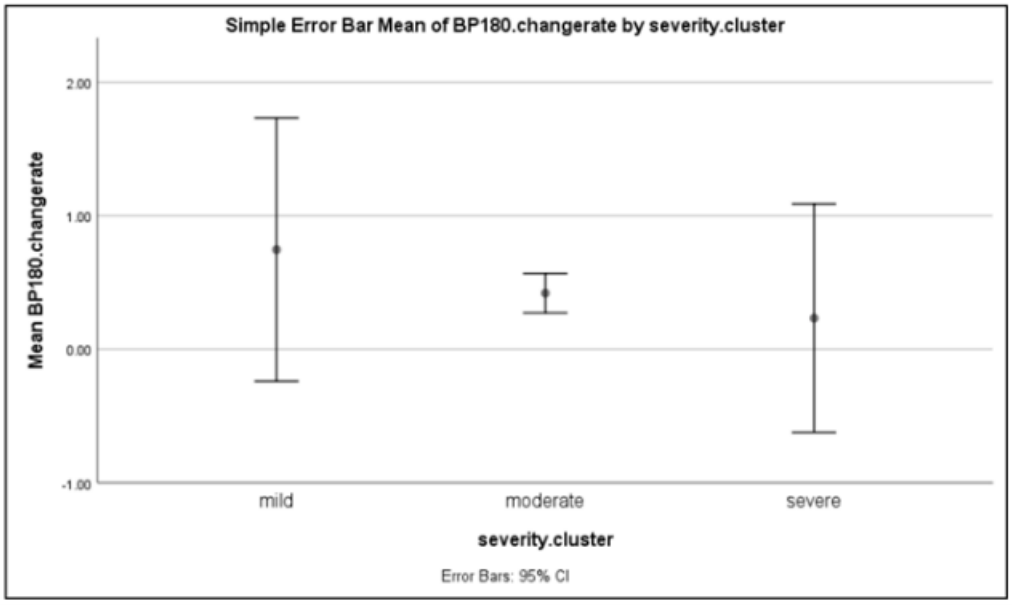


Fig 2. Levels of BP180 after six months of treatment with Rituximab in the three groups of disease severity.

A significant relationship between disease severity and prednisolone dosage was observed (p=0.002). This correlation persisted at three months (p=0.000), indicating a significant relationship between prednisolone dose reduction and BP180 levels after three months (r=0.459, p=0.002). Figures 3a and 3b show the average dosage of prednisolone at one and three months of rituximab treatment, respectively. Conversely, changes in topical Clobetasol dose within the first month showed no significant relationship with disease severity (p=0.139). The dose change was insignificant at three months (p=0.969). Fig 3c and 3d show the average dosage of topical clobetasol at one and three months of rituximab treatment, respectively.

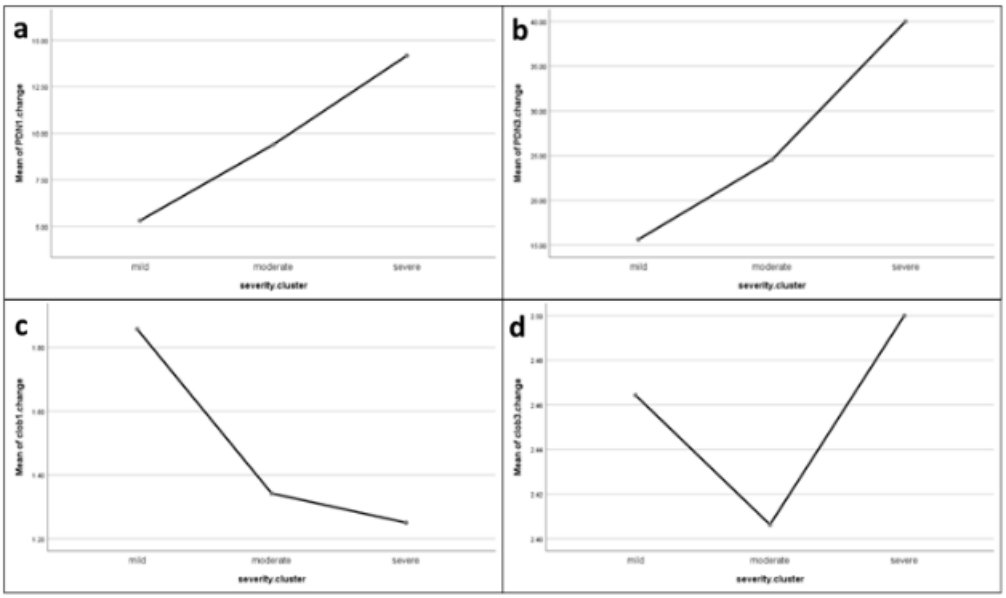
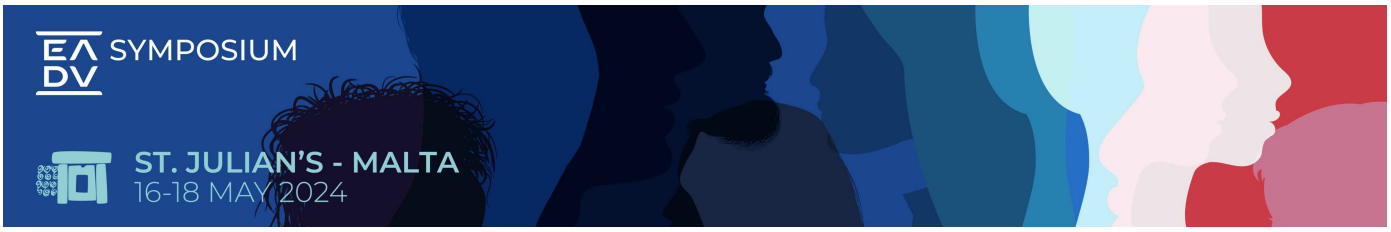


Fig 3. Effect of rituximab treatment on the dosage of prednisolone and clobetasol across different disease severity groups.

Conclusion: BP180 levels indicate BP severity and treatment response. Despite the efficacy of rituximab in reducing BPDAI scores, the significance of BP180 levels persisted, indicating a relationship between antibody levels and treatment response. Higher baseline BP180 levels correlated with reduced systemic steroid doses, suggesting enhanced efficacy of rituximab in patients with elevated initial BP180 levels.



Abstract N°: 1732

A rare case of Bullous Pemphigoid on preexisting lesions of Lichen Sclerosus et Atrophicus

Vesna Reljic[1, 2], Tijana Orlić[1], Olivera Drulovic[1], Jelena Vukovic[1], Dubravka Zivanovic[1, 2]

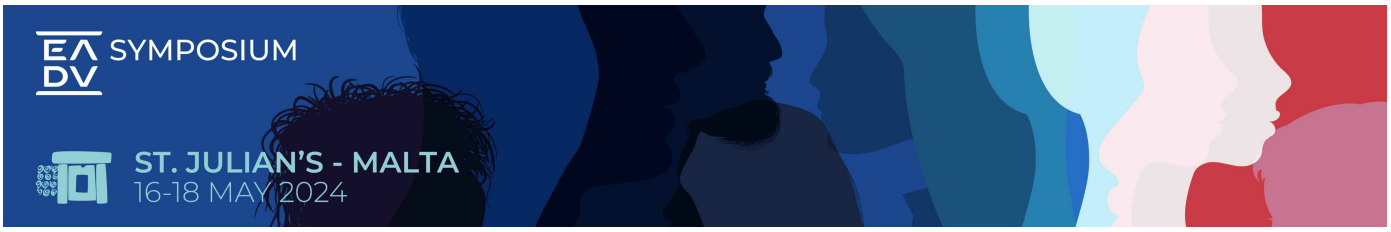
[1]Clinic of Dermatovenereology, University Clinical center of Serbia, 3rd female unit, Belgrade, Serbia,[2]Faculty of Medicine, University of Belgrade, Belgrade, Serbia

Introduction & Objectives: Bullous pemphigoid (BP) is an autoimmune blistering disorder that typically affects the elderly and can be associated with substantial morbidity. Although the etiology is not completely understood, it has been reported that BP might appear on sites of skin damaged due to physical factors or inflammation. We report an unusual case of BP developing on the lesions of pre-existing and histologically confirmed lichen sclerosus et atrophicus (LSA).

Materials & Methods: A 69-year-old woman was admitted to our clinic with an 8-month history of sclerotic and atrophic plaques on her back, breasts and both of her thighs. They were acompanied by itching and pain. Three months later blisters appeared on site of the plaques. At the time of admission, physical examination revealed reddish and whitish indurated plaques with follicular plugging, approximately 25x20 cm in diameter, symmetrically extending over proximal thighs. Several erosions with crusts were seen. There were no genital lesions. Similar plaques were also seen on her lumbar and mammary region, but without erosions. There was no family history of blistering diseases. Personal history of autoimmune diseases was also denied. A biopsy was taken from an indurated plaque of the left thigh.

Results: Histopathological examination revealed hyperkeratosis, interface dermatitis and flattened rete ridges in the epidermis. In dermis, hyalinosis of papillar and upper reticular derm was seen, as well as focal infiltrate composed of lymphocytes. Direct immunofluorescence (DIF) obtained from the perilesional skin showed a linear deposition of immunoglobulin G and C3 complement along the basement membrane zone. Indirect immunofluorescence test was negative. Enzyme-linked immunosorbent assay found no elevated levels of antibodies to collagen type VII that excluded the possibility of epidermolysis bullosa acquisita. The diagnosis of BP arising on the site of LSA was established. Treatment included methotrexate 15 mg weekly along with local treatment with 0.05% clobetasol propionate ointment. At the time of writing the patient has a good response and no blisters.

Conclusion: The occurrence of blisters in patients with LSA can cause a dilemma whether they represent a bullous variant of same disease or another unrelated autoimmune diagnosis. The occurrence of BP on LSA lesions is rare. We can assume that LSA served as a predisposing factor for the development of BP. Detailed diagnostics including histopathology and DIF may be needed to conclude between these two different but potentially related conditions.



Abstract N°: 1741

A case of Epidermolysis Bullosa Acquisita, imitating Chronic Bullous Disease of Childhood (or linear IgA dermatosis).

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Introduction & Objectives:

Childhood epidermolysis bullosa acquisita (EBA) is a very rare autoimmune skin blistering disease, with fewer than 50 cases reported in the literature. Four subtypes have been described: classic mechanobullous, linear IgA bullous dermatosis-like, bullous pemphigoid (BP)-like, and mucous membrane pemphigoid-like. IgA dermatosis-like EBA is characterized clinically by "string of pearls" or "clusters of jewels" appearance, commonly on an inflammatory background. Oral and genital mucosae are frequently affected in pediatric cases of EBA. The presence of antibodies against type VII collagen is characteristic.

Materials & Methods:

We present a case of a 16-year-old boy with a vesicular rash involving the face, neck and trunk. On examination, there were multiple vesicles overlying erythematous plaques and scattered small erosions, across the face and upper trunk. Discrete blisters in an annular configuration, like "string of pearls" were also noted over the neck bilaterally.

The patient was otherwise in a good medical condition, taking no regular medication. He had no family history of autoimmune or cutaneous diseases. Full blood count was normal with no leukocytosis or eosinophilia, as well as serum chemistry. The immunologic blood examinations were negative.

The differentials of linear IgA dermatosis, bullous pemphigoid, epidermolysis bullosa acquisita and bullous systemic lupus erythematosus were considered, and two punch biopsies were performed and sent for hematoxylin/ eosin staining and direct immunofluorescence.

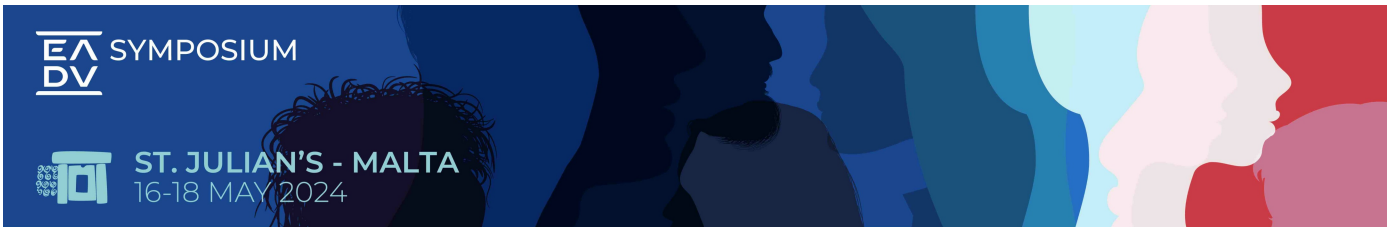
Results:

Histology revealed a subepidermal split and a superficial perivascular inflammatory infiltrate with prominent eosinophils. Direct immunofluorescence (IF) displayed linear deposition of C3 and IgG along the basement membrane zone. This finding was against a classical IgA dermatosis, although the clinical picture was very typical for that. The first working diagnosis was initially childhood bullous pemphigoid (BP), and systemic prednisone of 25mg was initiated (0.40 mg/kg). However, the patient did not respond to the treatment. Moreover, the clinical picture was not typical for childhood BP. Therefore, a sample was sent to perform a salt split skin test. The IF of the salt split skin examination revealed localization to the base of the blister and anti-collagen VII antibodies later proved positive, consistent with a diagnosis of epidermolysis bullosa acquisita (EBA). Given the clinical and histological features, it was classified as IgA dermatosis-like EBA and in addition to prednisone, dapsone 50mg daily (0.75mg/kg) was initiated. Topical treatment included clobetasol propionate 0.05% twice daily. The patient was reviewed at two weekly intervals, and at two-month follow-up, the lesions had resolved with residual dyspigmentation.

Conclusion:

We present a case of IgA dermatosis-like EBA, because of its rarity and to consider this in the differential of autoimmune

bullous diseases in adolescence. Evaluation of salt split skin test should be performed to distinguish between BP and EBA, and to follow, subsequently, the appropriate medical treatment.



Abstract N°: 1771

Eruptive Hypertrophic Discoid Lupus Erythematosus Mimicking Keratoacanthoma: A Clinical and Histopathological Diagnostic Challenge

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Introduction & Objectives

Chronic hypertrophic discoid lupus erythematosus (CHDLE) is a rare form of chronic discoid lupus erythematosus. It manifests itself with hyperkeratotic lesions affecting photoexposed areas. The clinical course is typically marked by chronicity and resistance to therapy. It presents a diagnostic challenge that can be aided by histological examination. In this study we present a case that highlights the importance of considering this pathology in the differential diagnosis of multiple papulo-squamous dermatoses.

Materials & Methods

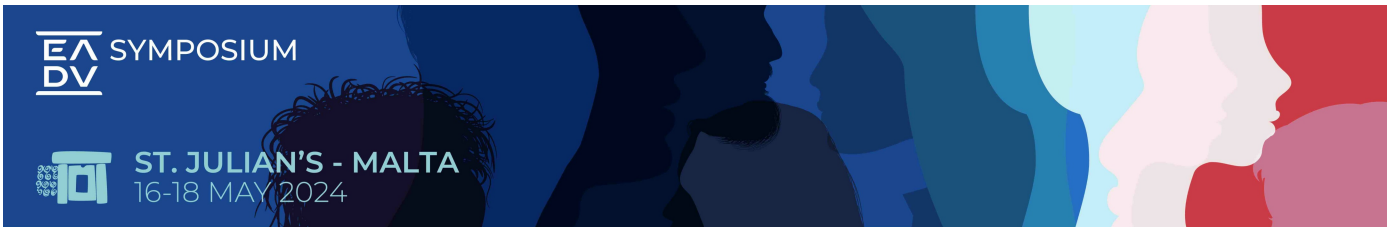
We report a case of CHDLE.

Results

A 47-year-old female patient with a history of systemic lupus erythematosus (SLE) but without regular follow-up presented with painful lesions on her arms that had been evolving over the past year. Physical examination revealed multiple tumors with a keratotic center and a peripheral rim covered with normal-looking skin, following a linear path on both upper limbs, including the shoulders, forearms, and hands. Additionally, the patient exhibited scarring alopecia and arthralgias. Dermoscopic examination revealed a hyperkeratotic central plug surrounded by a hypertrophic labrum, with some lesions showing thrombosed vessels due to excoriation. Differential diagnoses considered included multiple keratoacanthomas, Muir Torre's syndrome, skin tuberculosis or CHDLE. Biopsies from lesions on the forearm and shoulder were performed. Histopathological examination revealed pseudoepitheliomatous epidermal hyperplasia, parakeratosis, isolated necrotic keratinocytes, focal hypergranulosis; dilated vessels in the dermis, discrete mononuclear inflammatory infiltrate, focal lichenoid and surrounding the superficial plexus, fibrosis, and intense solar elastosis, consistent with CHDLE. Furthermore, the patient was diagnosed with hemolytic anemia and tested positive for antinuclear antibodies (1/640 homogeneous pattern) and negative for dsDNA and Sm antibodies, confirming the diagnosis of SLE. Treatment was initiated with hydroxychloroquine 200 mg every 12 hours and clobetasol cream 0.05% with occlusive dressing. After two months of follow up, the lesions improved, leaving hypopigmented and atrophic scars.

Conclusion

We present a case with multiple CHDLE lesions, that clinically and histopathologically resemble keratoacanthomas, along with features of SLE and discoid lupus erythematosus on the scalp aiding in the correct diagnosis. This disease poses a therapeutic challenge, but treatment with antimalarials and clobetasol cream proved effective in improving the lesions and resulting in hypopigmented atrophic macules. Approximately 5% of patients may progress to a systemic form of the disease, and long-standing CHDLE lesions have the potential to develop into squamous cell carcinoma, emphasizing the importance of clinical and histopathological follow-up.



Abstract N°: 1778

Alopecia areata and Hashimoto's thyroiditis. Treatment with JAK inhibitors.

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Title

Alopecia areata and Hashimoto's thyroiditis. Treatment with JAK inhibitors

Introduction & Objectives:

Alopecia areata is a rather common disease causing local or generalized hair loss. The disease may have an autoimmune etiology. It may be accompanied by Hashimoto's thyroiditis. Hashimoto's thyroiditis is an autoimmune thyroid disease which may be accompanied by hypothyroidism or may occur in the context of normal thyroid function. The aim was to describe a group of patients with alopecia areata and Hashimoto's thyroiditis and the response to treatment in a rather aggressive case of alopecia areata with JAK inhibitors.

Materials & Methods:

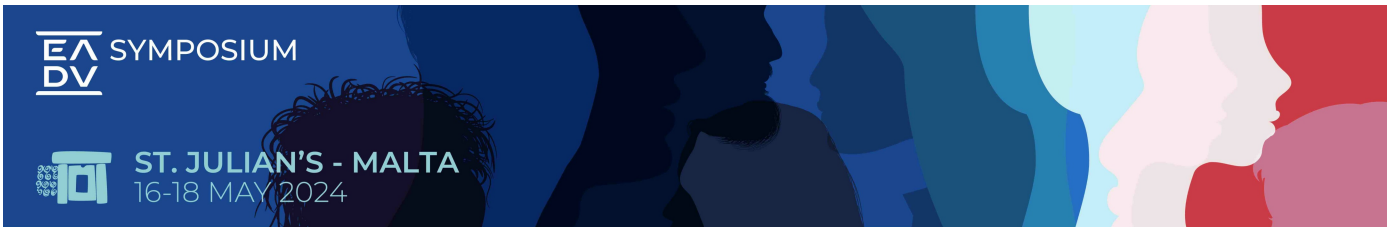
A group of 5 female patients is described who presented with alopecia areata. During laboratory evaluation the presence of anti-Tg and anti-TPO antibodies was detected. TSH levels were elevated in 3 of the 5 patients while they were normal in 2 of the patients.

Results:

During follow-up thyroxine was administered in the patients who had hypothyroidism. Lesions of alopecia areata in the scalp improved in 4 of the patients. In one of the patients, a female aged 18 years hair loss was in the scalp continued. Thereafter, treatment with a JAK inhibitor was initiated with a good response as hair loss stoped.

Conclusion:

In conclusion, alopecia areata is a rather common disease which may be of autoimmune etiology. It may be accompanied by Hashimoto's thyroiditis and in most cases has a rather bening course, as lesions improve over time. In patients with aggressive hair loss JAK inhibitors may be used therapeutically and have a beneficial effect. JAK inhibitor treatment is a novel and exciting mode of treatment for patients with aggressive forms of alopecia areata.



Abstract N°: 1786

The association of alopecia and cutaneous lupus: 3 observations

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Introduction & Objectives:

Alopecia areata is a common cause of non-scarring acquired alopecia. It is an autoimmune disease that affects people who are genetically predisposed to it. It may be associated with a variety of comorbidities, especially autoimmune conditions. The coexistence of alopecia areata and systemic lupus erythematosus (SLE) has been reported; however, the link between these conditions has yet to be definitively proven. We present three cases of patients who were treated for alopecia areata and acquired cutaneous lupus. Our research is particularly interesting since it highlights the link between alopecia areata and autoimmune illnesses, specifically cutaneous lupus.

Materials & Methods: 3 cases**

Results:

Case 1:

A 40-year-old female patient with an autoimmune polyendocrinopathy consisting of autoimmune thyroiditis and Addison's disease, associated with plaque-like alopecia of the scalp, presented with erythemato-squamous papular lesions on the face and photosensitivity associated with a relapse of her alopecia. The patient was treated with hydroxychlorochin and corticosteroid infiltration.

Case 2:

54-year-old female patient with alopecia universalis and primary biliary cholangitis, with positive anti-MI2 antibodies, presented with papular erythematous scaly lesions of the neck,trunk, back and extremities, which had been present for 18 months. The skin biopsy was consistent with subacute lupus. Laboratory tests revealed positive antinuclear AAN antibodies, doubtful SAA antibodies, negative SSB and anti-native DNA antibodies, and autoimmune thyroiditis with positive anti-thyroglobulin antibodies. The patient was treated with hydroxychloroquine and dermocorticoids.

Case 3:

A 35-year-old patient with alopecia universalis presented with pruritic skin lesions under the breast that had been evolving for 5 years, with photosensitivity. A skin biopsy showed chronic lupus with positive direct immunofluorescence and linear deposits of IgG and IgM antibodies at the membrane level. The patient was treated with hydroxychloroquine and corticosteroid infiltration.

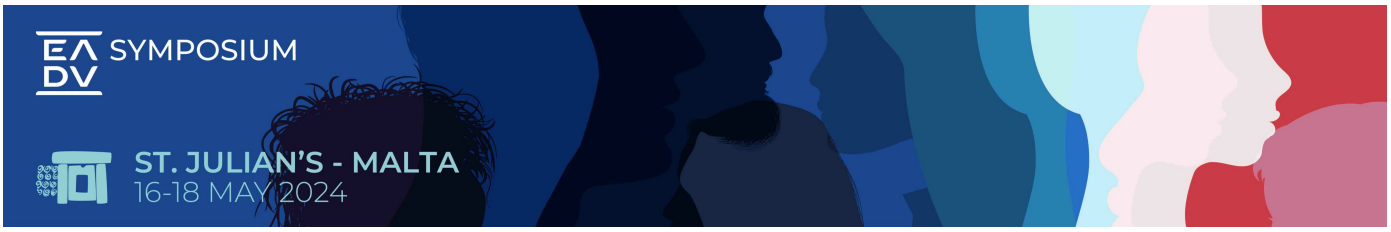
Discussion:

Alopecia areata is sometimes associated with other autoimmune or allergic diseases. To date, several cases of SLE associated with alopecia have been reported in 1.2% of cases. This association is more frequent in elderly and female patients, as in our observations. Indeed, CD4+ T lymphocytes play a central role in the genesis of both diseases, producing autoantibodies to several antigens, leading to the subsequent induction of autoimmunity. A genome-wide study has suggested that several genomic regions are significantly associated with alopecia, lupus erythematosus, and other autoimmune diseases, and that this association increases with age and gender. Female gender, age of onset over 40, and Jewish ancestry were identified as risk factors for the onset of SLE concomitant with alopecia. In some cases, alopecia may

be associated with several autoimmune pathologies at the same time.

Conclusion:

Several autoimmune diseases can be associated with alopecia, and coexistence with lupus is not uncommon. A better understanding of the comorbidities associated with alopecia could help us to understand them better, and screen for them if necessary.



Abstract N°: 1824

Extragenital Lichen sclerosus et atrophicus : A case report

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Introduction & Objectives:

Lichen sclerosus (LS) is a chronic inflammatory skin disease commonly affecting the anogenital area. However, the cutaneous involvement remains less known and studied.

Materials & Methods:

We report a case of scleratrophic lichen in a woman with purely cutaneous involvement, highlighting the importance of making this diagnosis in the absence of obvious genital involvement.

Results:

A 41-year-old woman with a history of hypothyroidism and diabetes who had for 8 years, dépigmented lesions slightly pruriginous, gradually increasing in size, initially affecting the back and trunk, then extending to the lower limbs.

Clinical examination showed multiple macules, slightly sclerotic in the centre, 1 to 5 mm, ivory-white in colour and confluent in patches on the back, trunk and lower limbs.

Oral and genital mucosa were normal. The rest of the clinical examination was normal.

However, histology showed an epidermal atrophy associated to basal vacuolization and moderate dermal fibrosis with lym- phocytic infiltrate orienting to the diagnostic of LSA cutané.

Complete blood count, routine blood biochemistry, antinuclear antibody (ANA) and hepatitis B and C serologies were within normal limits.

the patient was treated with methotrexate at a dose of 25mg/week with an emollient and there was slight improvement of the lesions on back. A close follow up was planned.

LS was first described by Hallopeau in 1887 and it received multiple names such as kraurosis vulvae, white spot disease, leukoplakia and lichen sclerosus et atrophicus. The final term “lichen sclerosus” was accepted in 1976 by the International Society of the Study of Vulvovaginal Disease

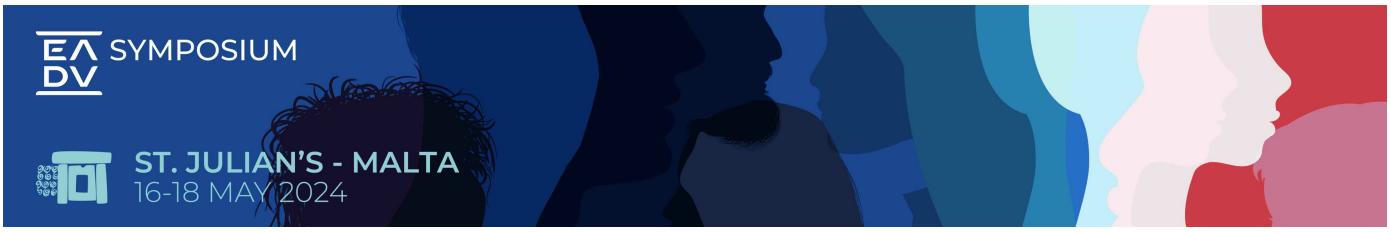
(LS) is an underdiagnosed inflammatory mucocutaneous condition affecting the anogenital areas. Purely extra-genital localization is only seen in 2.5% of cases . Extragenital atrophic lichen sclerosis occurs mainly in post-menopausal women , mainly in the submammary region, neck, shoulders, inner thighs, wrists and upper back. The pathophysiology may involve several factors: hormonal, genetic, infectious (Borrelia burgdorferi) or traumatic.

Clinically, the lesions take the form of whitish or pearly-white, “porcelain-like”, atrophic patches, mainly affecting the trunk, back, roots of limbs and folds. Pruritus is inconsistent. Blaschkolinear and bullous clinical forms have been described. Diagnosis is based on skin histology, which reveals atrophy of the squamous epithelium with horizontalization of the basal layer, follicular hyperkeratosis, and above all the presence of a subepithelial band of fibrous or oedematous collagen in the superficial dermis, devoid of elastic fibres when stained with orcein .

The treatment of CLS is not codified and consists of local treatment (potent topical DC or intralesional corticosteroid, calcipotriol, tacrolimus), more or less combined with general treatment (synthetic antimalarials, retinoids, general corticosteroid therapy, methotrexate or ciclosporin) depending on the course of the disease under local treatment, and UVB phototherapy can be also used.

Conclusion:

Extragenital lichen sclerosus et atrophicus is a rare disorder that can be difficulte to differentiate clinically from a number of disorders of hypopigmentation and sclero-atrophic disorders, and hence requires skin biopsy.Unlike genital LS, extragenital LS is not associated with a risk of carcinomatous transformation.



Abstract N°: 1829

Exploring Rarity: A case study of bullous Wells syndrome or the coexistence of pemphigus and eosinophilic cellulitis

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Introduction & Objectives:

Wells syndrome, also known as eosinophilic cellulitis, is a rare inflammatory skin disorder of unknown etiology characterized by initially erythematous-edematous and later indurated plaques, and sometimes blistering lesions on the skin. Typically presenting as recurrent granulomatous dermatitis with eosinophilia, it carries a favorable prognosis with appropriate treatment. However, the bullous presentation of Wells syndrome is rare, with only 10 cases reported in the literature.

Pemphigus vulgaris (PV) and Wells syndrome represent distinct entities, each with unique clinical features and diagnostic criteria. However, the coexistence of these two conditions in a single patient poses a diagnostic challenge and highlights the intricate interplay between immune dysregulation and cutaneous manifestations.

Materials & Methods:

We present the case of a 58-year-old man with a history of bullous lesions and a previous diagnosis of PV established at another clinic two years prior. The patient presented to our department in December 2023 with new cutaneous lesions, including multiple annular erythematous-edematous plaques with central clearing and a raised border, recurring for 2 months, and post-bullous erosions in the oral mucosa. The patient reported discomfort and intense pruritus associated with the cutaneous lesions. Clinical examination, skin biopsy, and specific antibody testing for pemphigus were performed to confirm the diagnosis. Additionally, laboratory investigations revealed a slight increase in peripheral blood eosinophil count, prompting further evaluation for eosinophilic cellulitis. The skin biopsy confirmed persistent eosinophilic infiltration in the dermis, consistent with the diagnosis of recurrent Wells syndrome. Treatment with topical and systemic corticosteroids was initiated, and the patient was closely monitored for response to therapy.

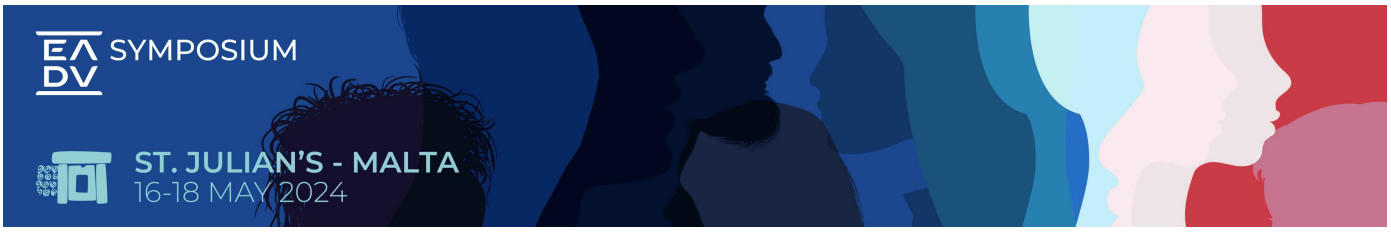
Results:

The patient's clinical and paraclinical presentation was consistent with a diagnosis of Wells syndrome. Notably, specific antibody testing revealed positivity only for anti-desmoglein 1 antibodies, suggestive of an overlap with PV. Treatment with corticosteroids, both topical and systemic, resulted in a favorable response, with resolution of the cutaneous lesions and improvement of oral mucosal erosions. However, two weeks after completion of corticosteroid therapy, the patient experienced recurrence of the cutaneous lesions, exhibiting features typical of eosinophilic cellulitis. This time, more indurated plaques were observed alongside residual post-inflammatory hyperpigmented patches.

Conclusion:

This case presents a diagnostic dilemma regarding the underlying etiology of the patient's cutaneous lesions. The possibility of bullous Wells syndrome with idiopathic positive anti-desmoglein 1 antibodies in an atypical context is considered. Alternatively, the recurrence of PV coexisting with Wells syndrome, which responded well to corticosteroid therapy, raises questions about the role of anti-desmoglein 1 antibodies in disease pathogenesis. Further investigation and monitoring are needed to guide optimal management of the patient's complex dermatologic condition. Consideration of bullous Wells syndrome in the differential diagnosis is crucial for patients presenting with bullous lesions, prodromal

pruritus, and significant peripheral eosinophilia.



Abstract N°: 1878

COVID-19-triggered scar sarcoidosis

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Introduction & Objectives:

The severe acute respiratory syndrome coronavirus 2 infections may lead to several side effects, including dermatological conditions such as urticarial rash, confluent erythematous rash, papulovesicular exanthema, chilblain-like acral pattern, livedo reticularis, racemosa-like patterns, and pityriasis rosea-like eruption.

Materials & Methods:

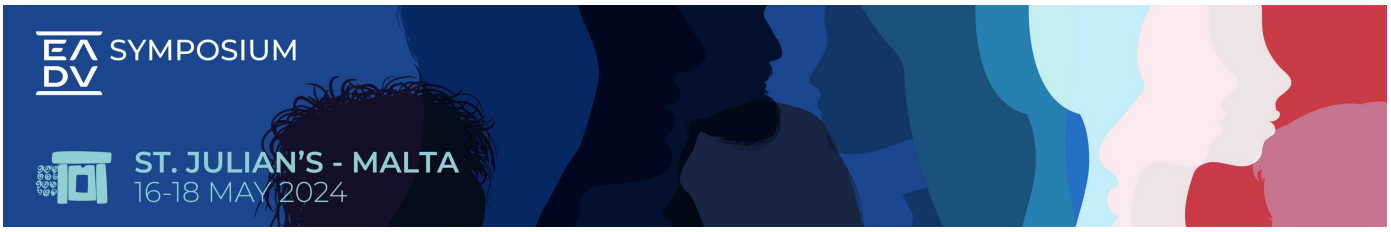
We report a case of scar sarcoidosis that appeared 10 days after infection with the SARS-CoV-2.

Results:

A 48-year-old man, with a history of COVID-19 infection 1 month before consultation, presented due to a subcutaneous tumor-like lesion in the area of his left eyebrow. Two years earlier, the patient experienced mechanical injury in the affected region with subsequent treatment of the scar with a CO2 laser and hyaluronic acid filler. Due to the unclear image of the ultrasound examination, magnetic resonance imaging (MRI) and punch biopsy were performed leading to a scar sarcoidosis diagnosis. Further diagnostics did not reveal the signs of extracutaneous sarcoidosis. Treatment consisted of intralesional administration of triamcinolone at a dilution of 10 mg/ml, which resulted in significant improvement and remission of the lesion. There were no signs of recurrence after 6 months of follow-up.

Conclusion:

This case shows a rare phenomenon of scar sarcoidosis triggered by COVID-19 infection, successfully treated with intralesional administration of triamcinolone. The underlying causes of this association remain to be elucidated



Abstract N°: 1881

Induced bullous pemphigoid : About two cases

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[1]La Rabta hospital, dermatology, [2]La Rabta hospital, Histopathology, [3]La Rabta hospital, Immunology

Introduction & Objectives:

Bullous pemphigoid (BP) is an autoimmune subepidermal blistering disease characterized by autoantibodies targeting hemidesmosomal proteins, primarily BP180 and BP230. The disease's pathogenesis has been linked to several triggering factors, including infections, physical agents, and drugs.

Materials & Methods:

Results:

Case report 1:

A 30-year-old man with a history of recurrent recreational drug and psychotropic use presented with a non-pruritic vesiculobullous eruption with mucosal involvement. The physical examination showed the presence of multiple tense bullae with clear or haemorrhagic content, arranged in rosettes on normal-appearing skin. Erosions with haemorrhagic crusts were mainly present in the large folds and trunk. Multiple erosions of the oral mucosa were also noted. The Nikolsky sign was negative. The patient reported taking hydroxyzine, heroin and levomeprazine two weeks before the eruption. The skin biopsy showed subepidermal blistering, apoptotic keratinocytes, and an eosinophilic infiltrate. Direct immunofluorescence revealed linear IgG and C3 deposits along the dermal-epidermal junction and granular perivascular IgG and C3 deposits. Indirect immunofluorescence was also performed. The patient responded rapidly and consistently to topical corticosteroids, with no signs of recurrence during several months of follow-up.

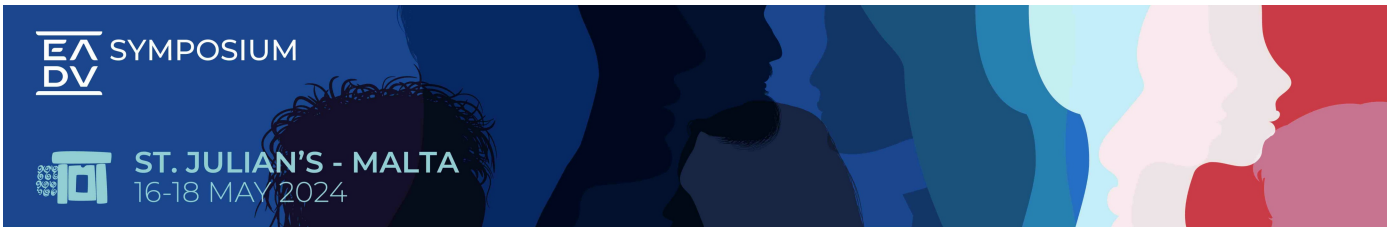
Case Report 2:

A 56-year-old man with no significant medical history presented with a non-pruritic bullous eruption on his legs two weeks after topical application of Marrubium vulgare. Examination revealed tense bullae arising on normal skin predominantly in flexural zones, upper extremities, and the trunk, with additional involvement of the oral mucosa. Nikolsky's sign remained negative. The skin biopsy confirmed the presence of subepidermal blistering with an eosinophilic infiltrate. Direct immunofluorescence revealed a linear deposition of IgG and C3 along the basement membrane zone. Indirect immunofluorescence showed anti-basemement antibodies type IgA and type IgG on the epidermal side. Treatment with topical corticosteroids resulted in a rapid clinical response.

Conclusion:

The pathogenesis of induced BP can be difficult to establish, but there are noticeable differences between idiopathic BP and induced BP that can offer valuable diagnostic clues. Induced BP typically affects younger individuals and is often associated with polypharmacy. Clinically, lesions develop on previously unaffected skin and may mimic other entities such as erythema multiforme or pemphigus. Patients with induced BP often experience involvement of their palms and soles, and may exhibit a positive Nikolsky sign. Histopathology frequently reveals eosinophilic infiltrates and necrotic keratinocytes. Treatment is usually effective, and patients typically do not experience a relapse once the causative substance has been eliminated.

The presented cases offer compelling evidence to support the diagnosis of induced BP. However, establishing a direct causal relationship between the administered drugs or substances and the disease remains challenging.



Abstract N°: 1903

Efficacy and tolerance of rituximab in the treatment of pemphigus herpetiformis

Kenza Khachani[1], Rasha Moumna[1], Maha El Maati[1], Najoua Ammar[1], Syrine Hamada[1], Nadia Ismaili[1], Meriam Meziane[1], Laila Benzekri[1], Karima Senoussi[1]

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Introduction & Objectives:

Pemphigus herpetiformis is a rare entity of the autoimmune disease pemphigus, with particular clinical, histopathological, and immunological characteristics. This subtype has an indolent course and it is known to have a good response to corticosteroids and dapsone. In some cases, these therapeutic options are not enough to maintain a remission of the disease, needing biological agents.

The objective of this study is to describe, a rare case of a relapsing pemphigus herpetiformis treated initially with corticosteroids then with rituximab.

Materials & Methods:

We report an uncommon case of a young woman presenting a recurrent pemphigus herpetiformis responding to biological therapy.

Results:

A 27-year-old female patient, with no particular history, presented in August 2020 with a 5 months history of a non itchy skin lesions and painful oral lesions. Physical examination revealed multiple bullous and erosive lesions initially on the trunk, then on the back and members, without Nikolsky's sign. Several erosions were also seen in the oral mucosa. Histological examination with direct immunofluorescence confirmed the diagnosis of a pemphigus herpetiformis. She had been treated with oral prednisone 1.2mg/kg/day with a progressive decline until August 2021 after a complete remission of the disease.

In November 2023, the patient was admitted to the hospital because of a pruriginous relapse of the bullous disease. Physical examination revealed the presence of vesicular and bullous lesions in an annular disposition on the trunk, the back and the upper portion of the limbs, without Nikolsky's sign. Mucous examination revealed multiple oral erosions. Anemia, peripheral eosinophilia and hypoalbuminemia were found in blood tests. We took a biopsy with a histological study showing intraepidermal bubble with acantholytic cells and eosinophilic spongiosis. Direct immunofluorescence revealed an intercellular IgG deposits located in the lower half of the epidermis, giving a "fishnet" appearance, without C3, IgA or IgM deposits. Indirect immunofluorescence showed the presence of circulating IgG antibodies targeting desmoglein 1 and 3.

The diagnosis of a pemphigus herpetiformis was retained. A treatment based on the association of rituximab cures, 1g every 6 months, and oral prednisone at a dose of 1mg/kg/day led to a clinical improvement with healing of skin and mucosal lesions after one month and a half of treatment. The good evolution led to a rapid decline of oral prednisone. The remission is maintained for four months without any side effect.

Conclusion:

Pemphigus herpetiformis is a rare pemphigus subtype that usually respond well to systemic steroids, dapsone and immunosuppressive agents. However, our case shows that biological therapy such as rituximab can also control this disease. Larger studies are needed to identify the role of rituximab in this disease.