

## Coexistence of pyoderma gangrenosum and sweet's syndrome in a patient with B-cell non-Hodgkin lymphoma

Dorsaf Mzoughi<sup>\*1</sup>, Malek Ben Slimane<sup>1</sup>, Faten Rabhi<sup>1</sup>, Kahena Jaber<sup>1</sup>, Mohamed Abderraouf Dhaoui<sup>1</sup>

<sup>1</sup>Military Hospital of Tunis, Dermatology department, Tunisia

**Introduction & objectives:** Pyoderma gangrenosum (PG) and sweet's syndrome (SS) are the most commun neutrophilic dermatoses (ND). They usually present with recurrent ulcers and tender erythematous plaques, respectively. Forms of transition, overlap or combination of these two ND in the same patient have been described in some cases.

**Materials & Methods:** We herein report a case of a patient with B-cell non Hodgkin lymphoma who developed two types of ND: PG and SS.

**Results:** A 53-year-old male patient with a history of B-cell non-Hodgkin lymphoma receiving chemotherapy since 8 months, presented with erythematous painful plaques located on the trunk and neck which appeared two weeks ago as well as an ulcerated plaque on the right arm of a three-month history. The patient was recovered as he had fever and an impaired general condition. The dermatological examination showed multiple , well-demarcated erythematous plaques infiltrated in places ,measuring 3 to 10 cm in diameter located on the trunk and the lateral sides of the neck. Some of the plaques had a dusky centre. There was also a large ulcerated circular plaque of 9 cm of diameter with an erythematous , well-demarcated and indurated border located on the right arm. The plaque was topped with thick dark-brown crusts. Blood tests showed leukopenia and elevated levels of C reactive protein. Histopathological examination showed at the ulcerated plaque an ulcerated epidermis and an inflammatory infiltrate of the dermis rich with neutrophils with leukocytoclastic vasculitis consistent with the diagnosis of PG. Skin biopsy from an erythematous plaque of the abdomen revealed an oedematous dermis with a dense infiltrate composed of neutrophils without vasculitis supporting the diagnosis of SS. The patient was treated with oral corticosteroid . Unfortunately , he died of septic shock two weeks later.

Conclusion: Some patients may present with overlapping clinicopathological features of different ND.

Coexisting PG and SS are the most commonly described cases in the literature, usually associated with haematological disorders (malignancies or myelodysplastic syndrome) and less frequently with chronic inflammatory bowel diseases especially ulcerative colitis. The first case of SS and PG occurring concurrently without malignancy was reported in 1997 by Lear and Byrne .Other associations of ND have been reported: PG and subcorneal pustular dermatosis , PG and aseptic spleen abscess.Two cases reported the coexistence of three ND in a single patient : PG , SS and erythema elevatum diutinum in a patient with seronegative rheumatoid arthritis and association of PG , SS and granuloma faciale in a patient with myelodysplastic syndrome.In summary,our case associates two types of ND and illustrates the clinicopathological continuous spectrum that may occur in such entities.



## Pyoderma Gangrenosum revealing a chronic inflammatory bowel disease

Yousra Habibi<sup>1</sup>, Baline Kenza<sup>2</sup>, Skali Hayat<sup>2</sup>, Chiheb Soumia<sup>2</sup>

<sup>1</sup>CHU Ibn Rochd, Casablanca, Morocco, <sup>2</sup>CHU Ibn Rochd

## Introduction & Objectives:

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis that presents with rapidly developing, painful skin ulcers hallmarked by undermined borders and peripheral erythema. Epidemiological studies indicate that the average age of PG onset is in the mid-40s, with an incidence of a few cases per million person-years. PG is often associated with a variety of other immune-mediated diseases, most commonly inflammatory bowel disease (IBL) and rheumatoid arthritis. The cause of PG is not well understood, but PG is generally considered an autoinflammatory disorder.

We report a particular case of a male patient who developed painful ulcers with rapid

deterioration on the left arm for 3 months along with intermittent episodes of diarrhea and rectal bleeding.

#### Materials & Methods:

#### **Results:**

This is a 51 years old male patient, without specific pathological antecedents who presented 3 months before his first consultation painful, rapidly evolving cutaneous ulcers with undermined, irregular, erythematous-violaceous edges on the left elbow, wrist and neck. In addition, he presented weight loss and episodes of diarrhea and rectal bleeding.

On clinical examination, large ulcerative plaques with ill-defined, violaceous, and undermined borders with cribriform scarring on the left elbow, wrist and neck.

The histological study of the skin biopsy demonstrated a diffuse neutrophilic infiltrate accompanied by a few lymphocytes and eosinophils. There was no growth of bacteria, fungi, or mycobacteria from tissue cultures, leading to the diagnosis of pyoderma gangrenosum.

A biological assessment was required, which showed low hemoglobin (10 g/dl reference range 13-17g/dl) normal leukocyte count (7000 /uL; reference range, 4000-10 000 /uL) and elevated C-reactive protein levels (43.86 mg/dL reference range, <5 mg/dL). Antinuclear antibody, rheumatoid factor, and antineutrophil cytoplasmic antibody levels were within normal limits. Colonoscopy and cross-sectional imaging showed varying degrees of inflammation (including erythema, erosions, and ulcers) next to areas of normal-appearing mucosa. The histological study objectified infiltrates from lymphocytes, plasma cells, granulocytes, basal lymphoplasmacytosis, distortion of the crypt architecture with shortening and disarray of the crypts leading to the diagnosis of Crohn disease.

The patient was treated with systemic prednisone, 30 mg/day in addition to management of his chronic inflammatory intestinal disease, and experienced complete healing of skin lesions.

## **Conclusion:**

PG is a reactive non-infectious inflammatory dermatosis falling under the spectrum of the neutrophilic dermatoses. PG is often associated with a variety of other immune-mediated diseases, most commonly inflammatory bowel disease and rheumatoid arthritis. IBL, neoplasia or rheumatological disease should be investigated.





# Advancements in Non-Invasive Skin Sampling: Clinical Conditions Characterization via the Assessment of Skin Surface Cytokine Biomarkers

Meital Portugal\*<sup>1</sup>, Miriam Oron<sup>2</sup>, Dror Cohen<sup>1</sup>, Ze'evi Ma'or<sup>3</sup>, Ron Kohen<sup>4</sup>

<sup>1</sup>DermAb.io, Haifa, Israel, <sup>2</sup>DermAb.io, Haifa, <sup>3</sup>The Skin Research Institute, the Dead-Sea & Arava Science Center, Masada, Dead Sea, Israel, <sup>4</sup>Hebrew University, The Myers Skin Research Laboratory, Faculty of Medicine, School of Pharmacy, Institute for Drug Research, The Hebrew University of Jerusalem, Jerusalem, Israel

## Introduction & Objectives:

The skin is increasingly recognized as a highly biological active organ with significant interactions with internal organs and the immune system. Cytokines play a crucial role as mediators between skin and immune system, regulating immune responses to preserve overall skin health. Given their involvement in both local and systemic immune processes, the expression of skin cytokines can serve as valuable indicators for the presence and progression of various systemic pathological processes and disordered conditions. Since the epidermal skin layer actively releases various cytokines, the emergence of non-invasive skin sampling methods might present a promising and sustainable approach for detecting these skin cytokines. These methods offer convenient means to gain valuable insights into the characteristics of clinical conditions.

This study focuses on non-invasively measuring cytokine levels directly from the skin surface using skin wash sampling to characterize different pathological processes: psoriasis, diabetes type 2, rosacea, and chronic kidney disease (CKD).

## Materials & Methods:

Skin surface wash sampling of the cytokines IL-1 $\beta$ , IL-8, and IL-10, was obtained from both healthy subjects and unhealthy patients, encompassing individuals of both genders, aged from 18 to 80 years. A well, 1 cm in diameter, containing 1 ml of sterile phosphate-buffered saline (PBS) solution was placed on skin surface. After 30 minutes of static period, the extracted PBS solution was collected from the well for further cytokine levels analysis using ELISA assays.

## **Results:**

Skin surface wash sampling results show distinct cytokine profiles in different pathological processes, healthy controls, affected and unaffected areas (Figures 1-3). In diabetes, IL-1 $\beta$  and IL-8 levels were elevated in lesional areas **(LS)**, while IL-10 levels were decreased in non-lesional skin **(NL)**. Psoriatic lesions showed elevated levels of IL-1 $\beta$  and IL-8. Rosacea patients had lower IL-10 levels in both lesional and non-lesional areas. CKD patients exhibited significantly lower IL-10 levels compared to healthy individuals.

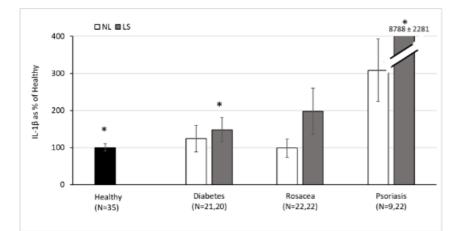
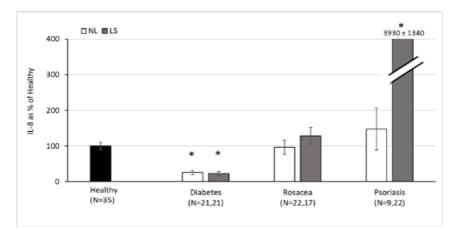


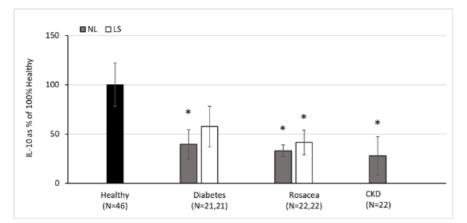
Figure 1: Skin surface IL-1β levels in different clinical conditions

Levels of IL-1 $\beta$  in healthy volunteers and in patients suffering from diabetes, Rosacea and Psoriasis. Samples were collected from the forearm. Samples from affected lesional areas in diabetes and Rosacea patients were collected from the feet and face respectively. All samples were collected with a PBS solution as described in the methods section. IL-1 $\beta$  was measured using ELISA as described in the methods section. The values presented are the mean  $\pm$  SEM. One-tail Student t -test was applied. Differences between average values were considered significant for P < 0.05 vs. Healthy controls.



#### Figure 2: Skin surface IL-8 levels in different clinical conditions

Levels of IL-8 in healthy volunteers and diabetes, Rosacea and Psoriasis patients. Samples were collected from the forearm. Samples from affected lesional areas in diabetes and Rosacea patients were collected from the feet and face, respectively. All samples were collected with PBS solution as described in the methods section. IL-8 was measured using ELISA as described in the methods section. The values presented are the mean  $\pm$  SEM. One-tail Student t -test was applied. Differences between average values were considered significant for P < 0.05 vs healthy controls.



#### Figure 3: Skin surface IL-10 levels in different clinical conditions

Level of IL-10 in healthy volunteers and diabetes, Rosacea and CKD patients. Samples were collected from the forearm. Samples from affected lesional areas in diabetes and Rosacea patients were collected from the feet and face respectively. All samples were collected with PBS solution, as described in the methods section. IL-10 was measured using ELISA, as described in the methods section. The values presented are the mean  $\pm$  SEM. One-tail Student t-test was applied. Differences between average values were considered significant for P < 0.05 vs healthy controls.

### **Conclusion:**

These findings demonstrate the potential of non-invasive skin sampling for understanding various pathological processes. Cytokine profiles obtained from skin surface wash method can serve as "alert biomarkers" for predicting disease progression, facilitating early detection. Furthermore, this method offers a cost-effective approach for pre-screening molecules and formulations in clinical studies. Skin surface wash sampling holds promise as a valuable tool that can be exploited and elaborated for additional biomarkers analysis and omics to optimize disorders characterization, by profiles of sub-populations, aiding in improving disease management.

\*Part of this study results is supported by the EU FP7 under the project SuperFlex no. 609198

#### **Conclusion:**



## An Unusual Presentation Of Morphea: A Case Report

Asiya Albadi<sup>1</sup>, Maryam Al Khamisani<sup>2</sup>

<sup>1</sup>Oman Medical Specialty Board, Oman, <sup>2</sup>HOD of Dermatology Department, Al Buraimi Hospital, Oman

## An Unusual Presentation Of Morphea: A Case Report

#### Introduction & Objectives:

Morphea, also known as localized scleroderma, is a chronic inflammatory connective tissue disorder with variable clinical presentations, that affects both adults and children.

Morphea clinically presenting as indurated short cords is rarely reported in literature.

We report a case of an unusual presentation of morphea, and review the clinical, histopathological presentation, and the treatment of this case.

## Materials & Methods:

A 24-year-old female, presented with insidious onset of multiple asymptomatic tiny skin-colored shiny papules forming short linear cords on anterior neck extending laterally both sides of many years duration. No history of joint pain, oral ulcers, or photosensitivity. No significant past medical, allergic, and drug history.

On examination, there were multiple skin-colored tiny linear papules forming short linear cords on anterior neck extending laterally on both sides. No similar lesions were found elsewhere on the body. There were no signs suggesting systemic sclerosis such as abnormal nail fold capillaries, no sclerodactyly, distal digital pitting/ulcers, or Raynaud phenomenon.

Biopsy showed thinned out epidermis, the dermis showed thickened eosinophilic collagen bundles in the papillary and reticular dermis along with perivascular lymphocytic inflammation. Alcian blue stain showed mild mucin deposition between the collagen bundles. Histopathological findings confirmed localized scleroderma.

#### **Results:**

Based on these clinical and pathological findings, the diagnosis of morphea was made. The patient was started on mometasone furoate cream once daily alternating with tacrolimus 0.1 % ointment for 1 month and to be followed up.

#### **Conclusion:**

This case highlights a rare entity of localized scleroderma with only one previously reported similar case in the literature and should raise awareness of this rare presentation.



#### Clinical Outcomes of Secukinumab in Moderate to Severe Hidradenitis Suppurativa: Real-world Response Rates

Gabriele Roccuzzo<sup>1</sup>, Federica Repetto<sup>1</sup>, Silvia Giordano<sup>\*1</sup>, Cristina Sarda<sup>1</sup>, Simone Ribero<sup>1</sup>, Pietro Quaglino<sup>1</sup>

<sup>1</sup>A.O.U. Citta della Salute e della Scienza di Torino, Medical Science, Torino, Italy

**Introduction & Objectives:** The SUNRISE and SUNSHINE trials have demonstrated the promising potential of the anti-IL17 agent secukinumab in ameliorating skin lesions associated with moderate-to-severe hidradenitis suppurativa (HS), achieving clinical responses in up to 46% of cases. Primary outcomes encompassed achieving a clinical response (Hidradenitis Suppurativa Clinical Response – HiSCR), reducing disease severity measured by IHS4 (International Hidradenitis Suppurativa Severity Score System), and attaining the new score IHS4-55 (indicating a 55% reduction in the IHS4-55 score).

**Materials & Methods:** In a retrospective study, we examined 24 patients from an Italian dermatological center who, having failed or shown contraindications to at least one anti-tumor necrosis factor (TNF)-alpha, received treatment with secukinumab. Drug survival was assessed, and Cox regression and logistic regression analyses were employed to explore potential clinical predictors of drug survival and clinical response.

**Results:** Among the 24 patients, 9 were male and 15 were female, with a median age of 38 and a median BMI of 25.6. Notably, 75% were active smokers. At the time of therapy start, 13 patients were classified as Hurley 2 and 11 as Hurley 3. The median number of previous therapy lines was 2 (range 1-5). As for the latent class analysis (LCA) subsets, LCA1, LCA2, and LCA3 accounted for 25%, 41.7%, and 33.3% of the patients' cohort, respectively. At the initiation of therapy, the overall median IHS4 was 18 (range 2-56), specifically 15 for Hurley 2 and 30 for Hurley 3 patients (p=0.03). All patients had been pre-treated with adalimumab, and discontinuation had occurred in 17 cases (70.8%) due to therapy inefficacy and 9 (37.5%) cases due to side effects. After a median follow-up of 24 months, HiSCR and IHS4-55 endpoints were achieved by 13 out of 24 patients (54.2%). The baseline IHS4 disease burden was negatively correlated with achieving HiSCR-IHS4-55 (OR 0.88, 95% CI 0.79-0.98, p=0.021), and patients with Hurley 2 were found to be more inclined to reaching the endpoints compared to those with Hurley 3 (p=0.045). The median drug survival was 16 months, with 13 cases of drug discontinuation due to inefficacy and 1 case due to secukinumab-related side effects. The prior use of systemic steroids (recorded in 20.8% of patients at baseline) was negatively associated with drug survival in univariate analysis. Conversely, achieving HiSCR-IHS4-55 was positively associated with drug survival, with an HR of 0.21 (95% CI 0.06-0.68, p=0.010), irrespective of the timing.

**Conclusion:** This research underscores the effectiveness of the anti-IL17 agent secukinumab in enhancing outcomes for certain patients with moderate-to-severe HS. The primary determinant of response to anti-IL17 treatment is the baseline disease burden, with no other clinical parameter displaying comparable predictive potential in our cohort. Noteworthy is the observation that the recently introduced score IHS4-55 demonstrates analogous discriminatory power to HiSCR in discerning responders from non-responders. Despite some encouraging findings, the overall rates of response and drug survival remain suboptimal, emphasizing the need for novel predictors of response and the advancement of new, effective therapeutic strategies.



## A Systematic Review on the use of Botulinum Toxin in the Treatment of Rosacea

Foteini Moniati<sup>\*1</sup>, Christos Costa<sup>2</sup>, Constantina Chatzimatthaiou<sup>1</sup>, Michalis Chatzimatthaiou<sup>3</sup>, Marianna Vassiliou<sup>1</sup>, Marios Chatzimatthaiou<sup>1</sup>

<sup>1</sup>Barts and The London School of Medicine and Dentistry, London, United Kingdom, <sup>2</sup>Imperial College London, United Kingdom, <sup>3</sup>Barts Health NHS Trust, London, United Kingdom

## Introduction & Objectives:

Rosacea, characterized by chronic inflammation, typically manifests with persistent erythema and intermittent flushing. Managing this condition poses challenges, with often unsatisfactory outcomes and a tendency for recurrence (1). In recent times, botulinum toxin has emerged as a novel therapeutic approach for rosacea (2). The aim of this systematic review is to assess the efficacy and safety of botulinum toxin on the treatment of rosacea, offering a more comprehensive and detailed evaluation.

## Materials & Methods:

PubMed, Embase and Cochrane Library were searched from inception until 31st December 2023 to identify the relevant studies for the use of Botulinum Toxin on patients with Rosacea. Prospective, retrospective studies and randomized controlled trials (RCTs) published in the English language and accessible as full-text articles in peer-reviewed journals were considered for the analysis.

## **Results:**

A total of twelve studies were included in the final review, involving a total of 212 participants. Among the examined studies, 5 were non-randomized prospective studies, 5 were prospective case series and 2 were characterized as randomized controlled trials (RCTs). Post treatment, each study demonstrated diverse degrees of improvement in patients' signs and symptoms, accompanied by diminished scores on the Clinician's Erythema Assessment (CEA). This improvement persisted for an extended duration, and any adverse effects reported were mild and self-limiting.

## **Conclusion:**

Botulinum toxin generally exhibits satisfactory effectiveness and safety in addressing rosacea; even though these findings are constrained by factors such as small sample size, imperfect study design, and short follow-up periods.

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## Pityriasis lichenoides secondary to CAR-T cell therapy in a patient with systemic B-cell lymphoma

Iuliana Frunze<sup>1</sup>, Hélène Bugaut<sup>1</sup>, François Le Pelletier<sup>2</sup>, Véronique Morel<sup>3</sup>, Stephane Barete<sup>1</sup>

<sup>1</sup>Pitié-Salpêtrière Hospital, APHP, Sorbonne Université, Dermatology Unit, Paris, France,<sup>2</sup>Pitié-Salpêtrière Hospital, APHP, Sorbonne Université, Anatomical Pathology Unit, Paris, France, <sup>3</sup>Pitié-Salpêtrière Hospital, APHP, Sorbonne Université, Hematology Unit, Paris, France

## Introduction & Objectives:

Pityriasis lichenoides (PL) includes a spectrum of inflammatory skin diseases from pityriasis lichenoides et varioliformis acuta (PLEVA) and febrile ulceronecrotic Mucha-Habermann disease (FUMHD) to pityriasis lichenoides chronica (PLC). We report a case of a papulosquamous dermatosis, followed by ulceronecrotic lesions, suggestive of PLEVA, after treatment with chimeric antigen receptor T cells (CAR-T cells).

## Materials & Methods:

A 56-year-old patient, after 9 months of complete remission from diffuse large B-cell lymphoma, treated with CAR-T cells, presented a disseminated eruption, gradually developed during 5 months, comprising erythematous papules, some of them covered with fine white scales, spread on the trunk and limbs, predominating on the flexion sites, with 1 necrotic ulceration of the left arm.

Routine laboratory tests showed an increased level of C-reactive protein- 56 mg/l and inflammatory microcytic anemia. Skin pathology of the lesions revealed a superficial and deep infiltrate with small pericapillary lymphocytes and intracorneal pustules. After eliminating the infectious causes and lymphomatoid papulosis, the diagnosis of lichenoid dermatosis was established.

After 6 weeks of treatment (topical steroids and antibiotherapy) the skin lesions disseminated on 60% of his body surface area. The lesions presented different evolutionary stages with new pustules progressing to necrotic ulcerations with thick hemorrhagic crusts, localized on the trunk and limbs, suggesting the diagnosis of PLEVA, without palmoplantar, facial or oral mucosa involvement.

The 2nd biopsy was consistent with PLEVA, reporting focal keratinocytic necrosis. Immunohistochemistry was non-specific and T clonality was negative, against lymphoproliferation or localization of CARTs in the skin. The treatment with oral steroids (prednisolone 20 mg/day), methotrexate 20 mg/week and topical steroids resulted in a progressive regression of PLEVA lesions within 2 weeks, followed by hyperpigmented macules.

## **Results:**

The pathophysiology of PL remains controversial. The hypersensitivity reaction to an infectious agent (EBV, VZV) or related drugs, or underlying lymphoproliferation are the main etiologies. In our case, the etiology could be related to CAR-T therapy, even knowing that skin toxicities are more related to initial cytokine release or T-cell proliferation.

This observation illustrates a diagnosis of PLEVA that should be taken into consideration in case of a prolonged papulosquamous/pustular eruption with necrotic ulcerations. Even though, the initial lesions were consistent with lymphomatoid papulosis, the evolution and the characteristics of the eruption have evoked a PLC and a PLEVA, which often present overlapping features.

#### **Conclusion:**

The diagnosis of PL at an early stage, allows a prompt initiation of an appropriate treatment, that can prevent progression to a severe form of the disease (PLEVA/FUMHD). Also, the patients with PL should be monitored due to the risk of developing lymphoproliferative disorders.



## Extragenital lichen sclerosus presenting as a comedo-like lesion mimicking a mycosis fungoides

Meriem Cherif<sup>1</sup>, Tabka Meriem<sup>1</sup>, Souissi Asmahane<sup>1</sup>, Chelly Ines<sup>2</sup>, Mokni Mourad<sup>2</sup>

<sup>1</sup>La Rabta Hospital, Dermatology, Tunis, <sup>2</sup>La Rabta Hospital, Anatomopathology, Tunis

**Introduction & Objectives:** Lichen sclerosus (LS) is a chronic inflammatory dermatosis of idiopathic origin and a sclerosing course, characterized by the presence of white and atrophic patches. Two forms of LS can be distinguished: genital LS (GLS) and extra-genital LS (EGLS), which may be associated. However, a particular tropism for the genito-anal region has been noted in both sexes in 85% of cases. EGLS often represents a diagnostic challenge as it can have various clinical presentations: pigmented, linear, blashkolinear and annular. Comedo-like openings were only seen under dermoscopy and were considered as a feature with a diagnostic and a prognostic significance. We herein describe an unusual case of "comedonicus" EGLS mimicking a folliculotropic mycosis fongoides (FMF).

## Materials & Methods: Case report

**Results:** A 52-year-old Tunisian female presented to our outpatient dermatology department with asymptomatic lesion on the forehand evolving for five months. She had a history of Hashimoto thyroiditis prior to presentation. She didn't complain of genital pruritus, dyspareunia or vaginal discharge. Physical examination revealed a well-defined ovalar hypopigmented plaque 3\*4cm, slightly elevated, with a wrinkled surface and numerous comedo-like openings, surrounded by an erythematous halo. Polarized light dermoscopy, showed multiple round craters containing yellowish to brownish comedo-like plugs. The examination of the ano-genital area did not reveal any evidence of erythema, white patches or atrophy. Histopathologically, the epidermis had irregular acanthosis and an interface dermatitis with vacuolar alteration of keratinocytes. Compact orthokeratosis and follicular plugging were present. The papillary dermis was fibrotic and immediately beneath the altered papillary dermis, there was a band-like infiltration and perivascular lymphoid infiltrate. The diagnosis of EGLS was made.

**Conclusion:** This case is an original clinical presentation of EGLS which highlights the importance of considering the hallmarks of the disease activity and the variability of the clinical presentation according to the disease stage.



## Extragenital lichen sclerosus: a 10-year retrospective study

Meriem Cherif<sup>1</sup>, Zaouak Anissa<sup>1</sup>, Chamli Amal<sup>1</sup>, Hammami Houda<sup>1</sup>, Fenniche Samy<sup>1</sup>

<sup>1</sup>Habib Thameur Hospital, Dermatology, Tunis

**Introduction & Objectives:** Lichen sclerosus (LS) is a chronic inflammatory dermatosis with a tropism for the genital mucosa; extragenital involvement remains less known and studied.

**Materials & Methods:** Monocentric retrospective descriptive study documenting all cases of extragenital LS (EGLS) in our dermatology department, diagnosed between 2012 and 2022.

**Results:** Among 50 cases of LS, 17 patients (15 women and 2 men) had extragenital involvement. EGLS was associated with genital involvement in 7 cases (14%) and isolated in 10 cases (20%). The mean age at diagnosis was 49 years (20-77 years). The mean duration of evolution was two years. Seven women were menopausal, and one had fibrosing frontal alopecia. Extragenital involvement was asymptomatic in 9 cases, while pruritus was present in 8 cases, judged moderate in 6 and ferocious in 2. The clinical appearance was characterised by hypochromic, shiny macules (11 cases) and slightly raised papules, sometimes depressed in the centre (6 cases), occasionally with an erythematous border (2 cases). Palpation revealed parchment-like atrophy in all cases. Telangiectatic appearance was described in one case. Hyperpigmented areas were associated in 3 cases. In nine cases (53%), lesions were localized. Eight others had diffuse lesions. The trunk was the most affected site (12 cases, 70%). Skin biopsy confirmed the diagnosis in all cases. Treatment was: strong dermocorticoids (DC) allowing reduction of pruritus. Methotrexate was prescribed in three cases, reducing sclerosis in two. However, all our patients retained artophic white scars.

**Conclusion:** Our series highlights the frequency of isolated EGLS. Clinical presentation can be challenging in the early forms because of the lack of sclerosis, the variability of localizations and the absence of anogenital lichen sclerosus.



## Practical attitudes of general practitioners in cases of seborrheic dermatitis

Belharti Kaoutar<sup>1</sup>, Nada Tahri<sup>1</sup>, Zizi Nada<sup>2</sup>, Dikhaye Siham<sup>3</sup>

<sup>1</sup>Department of Dermatology Venereology and Allergology, CHU Mohammed VI, Oujda Morocco., Dermatology , Oujda , Morocco, <sup>2</sup>Department of Dermatology Venereology and Allergology, CHU Mohammed VI, Oujda, Laboratory of Epidemiology, Clinical Research and Public Health, Faculty of Medicine and Pharmacy, Mohammed Premier University, Oujda Morocco Morocco, , Dermatology , Oujda , Morocco, <sup>3</sup>Department of Dermatology Venereology and Allergology, CHU Mohammed VI, Oujda Morocco, Laboratory of Epidemiology, Clinical Research and Public Health, Faculty of Medicine and Pharmacy, Mohammed Premier University, Oujda Morocco, Dermatology , Oujda , Morocco

## Introduction & Objectives:

Seborrheic dermatitis is a common chronic condition that affects 3 to 5% of the general population. It is considered one of the frequent reasons for consultation in general medicine. Our study aims to evaluate the different practical attitudes of general practitioners toward patients suffering from seborrheic dermatitis, and their need for additional training.

## Materials & Methods:

We conducted a descriptive study. A 23-question form was created on Google Forms, that was shared through social networks, assessing attitudes in general practice towards seborrheic dermatitis.

#### **Results:**

One hundred and thirty-eight physicians answered the questionnaire, 78% of whom were women. Seventy-one percent practiced in the public sector, and 29% practiced in the private sector. The average duration of practice was 7±2 years. Seventy-four percent reported treating patients with seborrheic dermatitis, and 26% referred these patients immediately to a dermatologist. Male patients were predominant, representing 54.3% of consultations, and the predominant age group was between 18 and 25 years old (50%), followed by patients aged between 26 and 35 years old (42.9%).

Regarding children, infants accounted for most consultations with 60.5%, followed by children between 2 and 12 years old who accounted for 18.6%, newborns with 14%, and children between 12 and 18 years old with 7% of consultations.

Adult patients had multiple histories, including diabetes in 39 patients, immunodeficiency in 36 patients, alcoholism, aerodigestive tract cancers, and pancreatic diseases in 12, 9, and 6 patients, respectively.

The most common location was the scalp (44.2% in adults, 38.6% in children), followed by the face (27.9% in adults, 29.5% in children), and 25.9% of adults had involvement of two or more sites compared to 18.5% in children.

Moderate involvement was predominant in consultations of all ages, with an association with pityriasis versicolor in 73.9%, and an association with anal inflammation in 33.3% of children, 7% of doctors suggested mycological sampling of the scales, compared to 93% who systematically treated their patients for seborrheic dermatitis.

The first-line treatment for scalp dandruff was mainly a topical imidazole (ketoconazole) in 44.7% and a selenium sulfidebased shampoo in 42.1%. A 0.75% piroctone olamine-based shampoo was recommended by 18.4% of the doctors, and a ciclopirox olamine-based shampoo by 13.2%. Additionally, 23.7% of doctors treated with topical corticosteroids as first-line therapy.

The treatments proposed for erythematous-scaly lesions of the face and trunk were topical imidazoles and dermocorticoids, each with a percentage of 47.5%, followed by zinc pyrithione-based cleansing gels at 40%. And for

children, 97% of doctors proposed treatment with topical imidazoles, and only 3% of doctors proposed dermocorticoids as a first-line treatment.

Eighty percent of the doctors followed the attack phase and maintenance phase, and only 35.2% of the doctors informed their patients about the need for various hygiene measures. In severe, extensive, and topical treatment-resistant forms, 50% of doctors proposed oral ketoconazole, 8.7% proposed oral retinoids, and 30.7% preferred to refer the patient to a specialist.

## **Conclusion:**

A better understanding of common and benign dermatoses in general practice allows for early and appropriate management, highlighting the importance of continuing education.



# Refractory eosinophilic annulare erythema Diagnostic challenge and complete remission with maintenance with dapsone

Igor Kapetanovic<sup>1</sup>, Tijana Orlic<sup>1</sup>, Dubravka Zivanovic<sup>1, 2</sup>, Snezana Minic<sup>1, 2</sup>

<sup>1</sup>Clinic of Dermatology and Venereology, University Clinical Center of Serbia, Belgrade, Serbia., Dermatology, Belgrade, Serbia, <sup>2</sup>University of Belgrade Faculty of Medicine, Department of Dermatology and Venereology, Dermatology, Belgrade, Serbia

#### Introduction & Objectives:

#### Materials & Methods:

#### **Results:**

Eosinophilic annular erythema (EAE) is a rare entity presenting with recurrent, annular, pruritic erythematous and edematous plaques along with tissue eosinophilia. While possibly related to hypersensitivity reaction, its etiology is ultimately unknown. It is considered a figurate erythema but there is controversy whether it is a distinct entity or subvariant of Wells syndrome. Treatment is homogenous, but systemic steroids and antimalarials are considered first line. Multiple other therapeutical options are used but with variable success. Dapsone has been used scarcely in literature for this diagnosis. We present a 60-year-old female presenting with a two year history of chronic and recurrent unilateral left eyelid edema and target-like annular edematous plaque with trailing scaling edge covering the whole left side of her face, nose and right cheek. She concurrently presented with over ten, each 5-20 cm, disseminated annular and arcuate edematous and livid erythematous plaques on her trunk and extremities. Over a two year span, multiple histopathological biopsies were non-specific and she was treated as dermatitis or cutaneous drug reaction. Her regular medication was discontinued and/or substituted but symptoms persisted. Biochemical analysis including tumor markers was within normal limits. Viral titers and Borrelia burgdorferi IgG/IgM were negative. Lesional and perilesional direct immunofluorescence microscopy and lupus band test were negative. Immunological analysis showed ANA-Hep2 nucleoplasm + homogenous (1:80) while ENA screen, C3,C4, dsDNA were within normal limits. For two years she was treated with antihistamines (maximum four-fold dosage increase), systemic methylprednisolone and prednisone with tapering, hydroxychloroquine and topical high-potent corticosteroids with minimal improvement and overall worsening of her symptoms and cutaneous lesions. Skin biopsy was repeated and revealed discrete epidermal spongiosis and exocytosis of regular lymphocytes and infiltrated and perivascular mixed inflammation (predominately eosinophils and less lymphocytes) in the superficial and deep dermis. No flame figures or fibrinoid necrosis were seen. Based on the clinical and histopathological findings, a diagnosis of EAE was made. Given her prior poor response to other modalities, dapsone was initiated with an initial dose of 50 mg, then increased to 100 mg daily. This achieved a complete resolution of the lesions after 8 days, which has been sustained over twenty months of follow-up while on a maintenance dose of dapsone 50 mg daily. EAE is a rare figurate eosinophilic dermatosis with, to the best of our knowledge, only a couple cases treated with dapsone. Our case gives credence that EAE could be a potentially effective treatment modality for quick resolution and remission.

#### **Conclusion:**



## Diagnostic Confusion: When Rosacea Mimics Lupus, a Case Report

Fatima-Ezzahraa Zeroual<sup>1</sup>, Ghita Ghita.Erramli@hotmail.Com<sup>1</sup>, Said Amal<sup>1</sup>

<sup>1</sup>Mohammed VI University Hospital, Department of Dermatology and Venerology, Marrakech, Morocco

## Introduction & Objectives:

Rosacea is a benign and common chronic facial inflammatory dermatosis. Its diagnosis is primarily clinical and can be confused with lupus erythematosus due to the semiological similarities between certain forms of both conditions. We describe here a case of rosacea mistakenly treated as cutaneous lupus for 5 years before the correct diagnosis.

## **Observation:**

We report the case of a 40-year-old patient, followed by a private practice dermatologist for 5 years for cutaneous lupus, treated with hydroxychloroquine and topical corticosteroids with medication abuse, poor adherence, and irregular followup. The initially responsive malar erythema evolved with transient improvement followed by worsening, extension, and the appearance of papulopustular lesions. Additionally, sinus tachycardia secondary to antiphospholipid syndrome (APS) occurred.

Due to the paroxysmal nature of clinical signs, the presence of telangiectasias, and the negativity of immunological tests, the diagnosis of cutaneous lupus was reconsidered, and rosacea was diagnosed according to ROSCO 2017 criteria. The patient is currently undergoing treatment with doxycycline 200mg/day, topical metronidazole, and photoprotection, showing significant clinical improvement.

#### **Discussion:**

Distinguishing between rosacea and acute cutaneous lupus in the case of a facial erythematous eruption is crucial. However, coexistence of both conditions in the same patient has been reported in the literature. The semiological similarities in certain presentations of both conditions can lead to misdiagnosis, emphasizing the importance of a systematic diagnostic approach, primarily clinical. This includes recognizing distinctive elements such as telangiectasias, paroxysmal evolution, and characteristic papulopustular lesions of rosacea, as well as the violaceous coloration of lupus malar erythema, its sharper lateral margins, and possible signs of systematization. In challenging cases, immunological profiling and skin biopsy help to differentiate.

#### **Conclusion:**

The semiological resemblance between rosacea and lupus erythematosus poses diagnostic challenges. The presented case underscores the importance of a vigilant diagnostic and therapeutic approach, especially in early stages, to ensure effective control of the condition while avoiding the burden of unnecessary and heavy treatment.



## Generalized pustular eruption as a presenting sign of COVID-19: a diagnostic challenge

Lucija Marcelić<sup>\*1</sup>, Katarina Dujmovic-Hasanbegovic<sup>1</sup>, Srdan Novak<sup>2</sup>, Tanja Batinac<sup>3</sup>, Sandra Peternel<sup>1</sup>

<sup>1</sup>Clinical Hospital Center Rijeka and Faculty of Medicine University of Rijeka, Department of Dermatovenereology, Rijeka, Croatia, <sup>2</sup>Clinical Hospital Center Rijeka and Faculty of Medicine University of Rijeka, Department of Internal Medicine, Division of Immunology, Rijeka, Croatia, <sup>3</sup>Clinical Hospital Center Rijeka and Faculty of Health Studies, University of Rijeka, Croatia

## Introduction & Objectives:

A variety of cutaneous manifestations have been reported in the setting of COVID-19, including pustular eruptions such as acute generalized exanthematous pustulosis (AGEP) and generalized pustular psoriasis (GPP). Aim of our work is to present a patient with a sudden onset of a generalized pustular eruption occurring as an initial sign of SARS-CoV-2 infection.

## **Results:**

A 69-year-old women with a 10-year history of seronegative arthritis and arterial hypertension, presented with acute onset of a palmoplantar vesicular rash and no other skin or systemic signs or symptoms. She was suspected of having a viral exanthem and prescribed symptomatic therapy with topical corticosteroid cream, however, four days later she developed generalized rash accompanied by fever up to 38 C, malaise and worsening of her arthritis. Upon examination, there were multiple erythematous papules and scattered pustules involving her trunk, face and extremities, as well as scaling of her scalp, palms and feet. Her laboratory tests revealed leukocytosis (12.6×109/L), neutrophilia (10.50×109/L), lymphopenia 1.00x109/L, elevated C-reactive protein (CRP) (191.0 mg/L), and a positive SARS-CoV-2 RT- PCR test. Skin biopsy showed multiple foci of neutrophilic spongiosis and subcorneal pustules, few dyskeratotic cells, papillary dermal edema with dilated capillaries and a perivascular infiltrate of lymphocytes, neutrophils and a small number of eosinophils. Histopathological differential diagnosis included AGEP and GPP, but AGEP was suggested as a preferred diagnosis given the presence of eosinophils. Meanwhile, the patient was started on acitretin 20 mg/day and topical corticosteroid which resulted in significant improvement. At four weeks' follow-up, the patient still had several erythematous and scaly plaques along with severe scaling of her scalp and swollen, painful fingers, so a final diagnosis of psoriasis (and psoriatic arthritis) was made. Due to significant alteration of liver enzymes, and persisting symptoms of arthritis, acitretin was discontinued and methotrexate introduced. In further follow-up, she had almost complete skin clearance but also experienced an additional flare of GPP, triggered by a stressful event.

#### **Conclusion:**

In this patient, making a diagnosis of GPP versus AGEP was a clinico-pathological challenge because of the overlap in the clinical and histopathological findings. Both entities have been described in the setting of an acute COVID-19 disease, however, what makes this case special is that this was new-onset GPP as an initial sign of the infection without the expected respiratory symptoms. The presence of leukocytosis, highly elevated CRP and fever suggested GPP however, these might have been due to COVID-19, and moreover, the finding of dermal eosinophilia was misleading. However, AGEP is a self-limiting condition that usually resolves within 2 weeks. In our patient, a final diagnosis of GPP was made based on the prolonged and relapsing clinical course with persistence of scaly plaques and concomitant arthritis. In clinical practice, there is a great importance in correctly differentiating between the two diagnoses because of different course, prognosis and treatment approach.





## A Rare Case Report of Acquired Reactive Perforating Collagenosis

Caitlin Borowsky<sup>\*1</sup>, Mariam Abu Jubain<sup>1</sup>, Anna Saparamadu<sup>2</sup>, Antonia Barbieri<sup>2</sup>, Pick-Ngor Woo<sup>1</sup>

<sup>1</sup>Northampton General Hospital, Dermatology, United Kingdom, <sup>2</sup>Northampton General Hospital, Histopathology, United Kingdom

## Introduction & Objectives:

Acquired Reactive Perforating Collagenosis (ARPC) is a distinctly rare dermatological condition characterized by the expulsion of deteriorated collagen fibres through the epidermis. ARPC tends to develop in adulthood, and is frequently associated with systemic diseases such as Diabetes Mellitus (DM) or End Stage Chronic Renal Failure (ESCRF). The infrequency of cases of ARPC has resulted in our reliance on knowledge collected from case reports, case series and retrospective analyses to inform our understanding of treatment options for this condition. Here we present a rare case of ARPC, notable for the absence of any systemic disease and successful treatment with Methotrexate.

## Materials & Methods:

Retrospective review of medical records from a patient diagnosed with ARPC.

## **Results:**

We present a sixty-year-old lady who came to our dermatology department as an emergency case with a seven week history of an intensely pruritic rash. Her rash initially manifested on her scalp before disseminating to her neck and arms. Notably, she did not have any systemic disease; she only had a past medical history of depression for which she was taking paroxetine. No new medications had been recently initiated. Upon physical examination numerous erythematous papules and nodules were observed, with central crusting and necrotic centres on some lesions.

Histopathological analysis revealed thick bands of necrotic collagen projecting through the inflammatory exudate within the ulcer cavity. The dermis surrounding the ulcer exhibited necrobiosis of collagen and perivascular inflammatory cell infiltrate, characterized by lymphocytes, histiocytes, eosinophils and occasional neutrophils. These histological findings supported a diagnosis of ARPC.

Various treatments yielded negligible improvement. These included oral and very potent topical steroids, Acyclovir, Cetirizine, high dose Fexofenadine, Hydroxyzine, Acitretin, Itraconazole, Narrow Band UVB phototherapy, Amitriptyline, Doxycycline and the application of emollients. Due to profound pruritus, she experienced sleep disturbances. Subsequently, she received intralesional Triamcinolone injections in private care, resulting in the temporary improvement of the lesions on her chest. However, she continued to manifest new lesions on her lower extremities. In response, we commenced Methotrexate 5mg once weekly, and topical Clobetasol Propionate ointment. She has now exhibited complete clearance of her skin lesions for over six months with no side effects.

#### **Conclusion:**

Our case of ARPC was an initial diagnositic challenge; she presented with the atypical absence of systemic disease. The scarcity of ARPC cases meant that we had minimal understanding on the best treatment strategy for ARPC which resulted in initial ineffectiveness of various treatments. This unique report emphasizes the significance of on-going research and case documentation to advance our understanding and treatment strategies for ARPC and contributes valuable insights into Methotrexate as a successful treatment for ARPC in the absence of systemic disease.





## Role of WW domain-containing oxidoreductase in regulating lipid production in human sebocytes

Whiin Lee<sup>1</sup>, Yun Su Eun<sup>1</sup>, Jae Yun Kim<sup>1</sup>, Euy Hyun Chung<sup>1</sup>, Sung Yul Lee<sup>\*1</sup>, Young Lip Park<sup>2</sup>, Jung Eun Kim<sup>1</sup>

<sup>1</sup>Soonchunhyang University Cheonan Hospital, Dermatology, Cheonan, Korea, Rep. of South, <sup>2</sup>Soonchunhyang University Bucheon Hospital, Dermatology, Bucheon, Korea, Rep. of South

## Introduction & Objectives:

The human WW domain-containing oxidoreductase (WWOX), a tumor suppressor gene located at the common chromosomal fragile site FRA16D at 16q23.3-24.1, has been implicated in various cellular processes, including metabolism, DNA damage response, inflammatory responses, and tumor suppression. Recent studies have also suggested its potential involvement in lipid synthesis, however, as far as we know, no investigation has explored the molecular mechanisms of WWOX in skin lipid metabolism. This study aimed to characterize the role of the WWOX gene in lipid synthesis within sebocytes.

## Materials & Methods:

Immortalized human sebocytes were transduced with a recombinant adenovirus expressing a microRNA targeting WWOX to down-regulate its expression. We analyzed the impact of WWOX knockdown on lipogenic regulators and intracellular signaling in insulin-like growth factor (IGF)-1 induced lipid production using Thin-Layer Chromatography (TLC) and western blot analysis.

## **Results:**

WWOX knockdown suppressed IGF-1-induced lipid production, particularly in squalene synthesis. WWOX knockdown also inhibited levels of lipogenic transcription factors and enzymes, such as peroxisome proliferator-activated receptor (PPAR)γ, sterol regulatory element-binding protein (SREBP)-1, stearoyl-CoA desaturase (SCD), and farnesyl-diphosphate farnesyltransferase-1 (FDFT-1). We revealed that impact of WWOX in lipid production was significantly related to AKT signaling pathways. The role of WWOX as a regulator of lipid production affects the expression of lipogenic regulators and downstream signaling pathways in immortalized human sebocytes.

## **Conclusion:**

These findings hightlight the role of WWOX in lipid metaobolism regulation. Our study provides the therapeutic potential of inhibiting WWOX for treating sebaceous gland-associated inflammatory skin disorders.



## Erythema elevatum diutinum

Serap Öztürkcan\*<sup>1</sup>, Göksu Dalgıç<sup>1</sup>, Sule Yildiz Sağcan Tercan<sup>1</sup>, Gamze Erkılınç<sup>2</sup>

<sup>1</sup>Hafsa Sultan Celal Bayar University Hospital, dermatology, manisa, Türkiye,<sup>2</sup>Hafsa Sultan Celal Bayar University Hospital, pathology, manisa, Türkiye

## Introduction & Objectives:

Erythema elevatum diutinum (EED) is a rare, chronic neutrophilic dermatosis that is characterized by red, violet, brown or yellow papules, plaques or nodules that distributed over the extensor surfaces. Lesions are often asymptomatic, although some patients may experience pain, itching or a burning sensation. Extracutaneous symptoms include arthralgia, fever, oral ulcers, ophthalmologic findings and other systemic symptoms. EED is most commonly seen in third to sixth decade. Erythema elevatum diutinum may be associated with a variety of diseases, such as hematological diseases, autoimmune disorders, connective tissue diseases, malignancies and infections (especially HIV infection).

EED typically resolves within 5 to 10 years. However, in some cases that may last as long as 25 years. Dapsone is used as the first step in the treatment, but relapse is common with cessation of the drug. Niacinamide, colchicine, hydroxychloroquine, tetracyclines may be other treatment options. Potent topical corticosteroids and intralesional corticosteroid injections may be helpful for mild cases, but systemic corticosteroids are generally ineffective. Fibrotic nodules often respond poorly to treatments. Local excision can be used for fibrotic nodules of EED.

#### Materials & Methods:

Here we report, 59-year-old female patient presented with a four-week history of painful erythematous plaques and nodules on the extensor surface of the left pretibial region. A punch biopsy was taken from extensor surface of the left pretibial region with the preliminary diagnosis of Erythema Elevatum Diutinum .

#### **Results:**

Histopathological examination revealed superficial and deep perivascular, interstitial dermatitis; thin lamellar orthokeratosis on the surface; Vascular infiltration in the upper and mid-dermis with predominant neutrophils and fewer eosinophils, plasma cells, histiocytes and nuclear debris were observed. Neutrophil leukocytes and fibrin accumulation were observed in the vessel wall. All these findings were found to be compatible with EED. Our patient's laboratory findings were normal, autoantibody tests were negative and serology for HIV (Human immunodeficiency virus) was non-reactive.

No additional disease was detected in the patient who was consulted by Rheumatology, Hematology and Ophthalmology departments. Partial improvement was observed at the 1-month follow-up of the patient, who was treated with topical corticosteroid and oral doxycycline.

### **Conclusion:**

We wanted to present our case with typical skin and histopatological findings due to rarity of Erythema elevatum diutinum.



Recurrent erythema nodosum after COVID-19 vaccination associated with latent tuberculosis: A case report and literature review

Yun Pei Koh<sup>1</sup>

<sup>1</sup>KK Women's and Children's Hospital, Singapore, Singapore

## Introduction & Objectives:

Cutaneous reactions to COVID-19 vaccination have been widely reported in medical literature. However, cases of erythema nodosum (EN) are rarely implicated. Here, we review the literature and describe a case of recurrent EN in a patient who received two doses of whole inactivated COVID-19 vaccine. His EN was attributable to underlying latent tuberculosis and COVID-19 vaccination, with recurrence of the skin nodules after his second vaccine dose.

#### Materials and methods:

A literature review was performed through PubMed. "Erythema nodosum" and "COVID vaccination" were inserted into the PubMed search box, with 23 results generated. Publication dates ranged from January 2022 to February 2024.

#### **Results:**

A 52-year-old male presented with fever, joint aches and painful leg rashes. He received his first dose of the vero cell inactivated COVID-19 vaccine (Sinovac) 2 days prior to his symptoms. He is a known hepatitis B carrier, with no other medical history. The patient is not on any medications. He has no other localizing symptoms of infection. He had no known history of tuberculosis but resides in an endemic region.

On examination, the patient was febrile and had erythematous palpable discrete tender nodules on bilateral shins. There were no other lumps elsewhere. There was no evidence of joint synovitis.

Laboratory data revealed elevated inflammatory markers with negative autoimmune markers. His COVID-19 PCR test was negative. His tuberculosis T-spot test was reactive; Chest X-ray performed was clear. A skin biopsy was done and histology showed suppurative granulomatous septolobular panniculitis. PAS, GMS, Ziehl-Neelsen stains were negative. Pyogenic, fungal and AFB tissue cultures were also negative. DIF was negative.

A diagnosis of erythema nodosum from latent tuberculosis was made, possibly triggered by his COVID-19 vaccination. The patient was given a course of oral etoricoxib and topical clobetasol ointment. His fever lysed shortly after treatment was initiated, with improvement of the shin nodules. Meanwhile, he was also started on treatment for his latent tuberculosis.

3 weeks after his first presentation, he received his second dose of the inactivated COVID-19 vaccine. 3 days after his vaccination, there was worsening of the shin tender nodules. There was no fever or joint pain. He was restarted on oral etoricoxib and topicals, with the nodules resolving over the next 3 months. Thereafter, he opted to receive the COVID-19 recombinant nanoparticle vaccine as a booster without any recurrence of EN.

#### **Conclusion:**

EN is associated with infections, drugs, malignancy and autoimmune disease. Some cases may be idiopathic. Vaccinerelated EN is rare, with skin manifestation occurring 24-48 hours post vaccination.<sup>1</sup> There are currently 23 reported cases of COVID-19 vaccine-related EN in English literature.

Cases have been reported after COVID-19 mRNA vaccines, adenoviral vector recombinant vaccines and whole inactivated

virus vaccines. In our patient, while his first episode of EN may be attributable to his underlying latent tuberculosis, he had a recurrence following a second dose of inactivated COVID-19 vaccine. He then remained well after receiving an alternative vaccine. Further studies are required to elucidate the pathogenesis of vaccine-related EN.

1. Damevska K, Simeonovski V. Covid-19 vaccine associated erythema nodosum: Factors to consider. Dermatol Ther. 2022;35(5):e15410.↔



# Efficacy of trichloroacetic acid and topical steroids in the treatment of hypertrophic lichen planus: A prospective study of 17 cases.

Anass Abbour<sup>1</sup>, Fatima Zahra Elfatoiki<sup>1</sup>, Cyrine Marmech<sup>1</sup>, Fouzia Hali<sup>1</sup>, Soumiya Chiheb<sup>1</sup>

<sup>1</sup>Ibn Rochd UHC, Dermatology, Casablanca, Morocco

## Introduction & Objectives:

Hypertrophic lichen planus (HPL) is a chronic form of lichen planus with more pronounced epidermal hyperplasia. This variant is particularly pruritic and difficult to treat.

Treatment involves topical and systemic immunosuppressors, with variable efficacy.

The aim of this study is to evaluate the safety and efficacy of trichloroacetic acid (TCA) in combination with topical steroids on hyperkeratosis, inflammation and healing in the treatment of hypertrophic lichen planus.

## Materials & Methods:

A prospective descriptive study was conducted in our dermatology-venerology department, over 1 year from December 2022 to December 2023.

Inclusion criteria were: Biopsy-confirmed, chronically evolving hypertrophic lichen planus, with or without systemic treatment outside the study period. TCA at 50% was applied once a week in the morning and topical steroids (clobetasol propionate) in occlusion every night.

## **Results:**

A total of 17 patients were included with a M/F sex ratio of 1,13. The mean age was 47 years with extremes ranging from 31 to 80 years.

For the therapeutic response onset, 4 patients showed improvement at week 3, 10 patients at week 4, 2 patients at week 7 and 1 patient at week 8. Cessation of severe pruritus was achieved after 6 weeks.

Regarding post-treatment clinical aspects, hyperkeratosis regression was noted in 15 patients, and complete stripping in 2 patients. In terms of pigmentation, post-treatment hyperpigmentation was noted in 11 patients, while depigmentation was observed in 6 patients.

Two patients reported side effects such as burning and infection.

## **Conclusion:**

Hypertrophic lichen planus represents a variant of lichen planus that is often refractory to topical treatment and therefore represents a therapeutic challenge.

Only a few anecdotic cases of the TCA-Topical steroids combination for the treatment of HLP have been reported with satisfying results. Indeed, the exfoliating and immunosuppressive properties of TCA peels improved the absorption and efficacy of topical steroids while reducing the pruritus accompanying hypertrophic lichen, thus diminishing the possibility of Köebner's phenomenon. However, the exact biological mechanisms involved in TCA application remain to be determined, particularly those that regulate inflammation. Note that the use of a topical steroid in combination with a potent exfoliating factor may have a place in the treatment of other skin diseases where hyperkeratosis is an obstacle to

absorption, such as hypertrophic lupus and nodular prurigo.

Knowing that carcinomas can occur on any hypertrophic lesion, early treatment of HLP is mandatory, with TCA and topical steroids combination as a non-invasive and generally well-tolerated treatment.



## Association of trichloroacetic acid with topical steroids in the treatment of verrucous lichen planus.

Salma Zakaryaa<sup>1</sup>, Fouzia Hali<sup>1</sup>, Bouchra Baghad<sup>1</sup>, Soumiya Chiheb<sup>1</sup>

<sup>1</sup>Ibn Rochd university hospital center, Dermatology and venereology, Casablanca

## Introduction & Objectives:

Hypertrophic or verrucous lichen planus (HPL) is a chronic form of lichen planus characterized histologically by more marked epidermal hyperplasia and clinically by symmetrical hyperkeratotic hypertrophic plaques, generally located on the pretibial or peri-malleolar regions, which are extremely pruritic and resistant to topical treatment. However, the combination of trichloroacetic acid (TCA) and topical steroids (very strong class) appears to be an effective local treatment for this type of lichen planus.

## **Observation:**

A 58-year-old patient presented with pruritic, erythematoviolaceous, papulo-nodular lesions on both legs and perimalleolar regions, which had been evolving for 1 year, forming infiltrated patches with a verrucous surface associated with a whitish network in the endo-oral cavity.

Histological examination revealed hyperplastic epidermis with hypergranulosis and lamellar hyperorthokeratosis with focal parakeratosis. The ridges are elongated, with a focal lymphohistiocytic infiltrate with a lichenoid appearance. The mid and deep dermis have a minimal lympho-histiocytic perivascular infiltrate.

The diagnosis was therefore hypertrophic verrucous lichen.

The patient was treated with 50% TCA applied weekly in the morning, combined with a very strong class of topical steroid (Clobetasol propionate) applied daily in occlusion in the evening on the lesions.

Evolution was marked by regression of the hyperkeratosis and subsidence of the lesions at S 15, followed by complete involution at S 24, with persistence of hyperpigmented patches.

## **Conclusion:**

Hypertrophic or verrucous lichen is a chronic inflammatory dermatosis characterised clinically by intense pruritus. This variant is often refractory to topical treatment and therefore represents a therapeutic challenge.

Combining TCA with topical steroids offers a good alternative for treating hypertrophic lichen planus. TCA reduces inflammation and pruritus and facilitates healing. In addition, due to its keratolytic properties, it improves the efficacy of topical steroids by reducing hyperkeratosis.



## Vitamin D Receptor Gene ApaI and FokI Polymorphisms as A Predisposing Factor for Bullous Pemphigoid: A Case-Control study

Parvaneh Hatami<sup>1</sup>, Nafiseh Esmaeli<sup>1, 1</sup>, Hossein Mortazavi<sup>1</sup>, Zeinab Aryanian<sup>1</sup>

<sup>1</sup>TUMS, Iran

## Introduction & Objectives:

Although the role of the VDR gene polymorphisms in several autoimmune and dermatologic disorders has been widely investigated, this association has not been studied before in Bullous pemphigoid (BP). Therefore, we aimed to examine the most frequently studied polymorphisms of the VDR gene in patients with BP in comparison with healthy matched controls.

#### Materials & Methods:

In a comparative case-control study consists of 56 patients with confirmed diagnosis of BP and 51 healthy matched control, TaqI, ApaI and FokI genotyping were performed using restriction fragment length polymorphism (RFLP) PCR.

#### **Results:**

Our results revealed a significant association between the genotypes of TaqI and ApaI polymorphisms of the VDR gene in both groups of participants (patient; p=5.691E-7, controls; p=0.000821). A significant association was found between genotypes of FokI and ApaI polymorphisms only in control group (p=0.040) and the haplotype AAFF was observed more frequently in the controls in comparison with patients group (p=0.03, CI:0.11-0.90; OR:0.31).

P value	Age	P value	Gender	SNP
	>60	<60		Female
0.10	ff	Ff	FF	ff
	4 (11.8)	19 (55.9)	11 (32.4)	1 (4.5)
0.63	Aa	Aa	AA	aa
	9 (26.5)	16 (47.1)	9 (26.5)	5 (22.7)
0.49	Tt	Tt	TT	tt
	4 (11.8)	16 (47.1)	14 (41.2)	1 (4.5)

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#### **Conclusion:**

our results showed the protective role of AAFF haplotype of two known VDR FokI and ApaI polymorphisms in susceptibility to BP. Further studies with larger study populations are suggested.





## Epidemiology, clinical presentation and management of IgA pemphigus: An update from Iran

Kamran Balighi<sup>1</sup>, Parvaneh Hatami<sup>1</sup>, Zeinab Aryanian<sup>1</sup>

<sup>1</sup>TUMS, Iran

## Introduction & Objectives:

IgA pemphigus has been seldom reported in the literature as a few case series to date and its characteristics are still a matter of debate.

Bearing in mind the rarity of this condition, no large or long-term study has been conducted to evaluate the clinical and laboratory details of those affected.

## Materials & Methods:

Here, we assessed demographic, clinical, histopathological and serologic profile of Iranian patients suffered from IgA pemphigus in a retrospective analysis of data retrieved from the clinical records of patients attending the pemphigus clinic in a referral hospital over a period of 10 years (July 2010– July 2020). All individuals with a confirmed diagnosis - defined as individuals with a cell surface IgA in DIF or IIF - were considered eligible, and their data were gathered for the analysis.

#### **Results:**

Among 1683 patients attending the pemphigus clinic during those years, a total of 8 patients, including 1 male and 7 females were identified.

The mean age of onset was 57.4 ± 15.3 years, ranged from 32 to 75 years. All patients had cutaneous lesions and vesicles/bullae were the most common lesions (87.5%), mostly noted on axilla, back and trunk area. Majority of patients did not show characteristic annular or arciform, crusted, or eroded patches with vesicles or pustules at the margins (75% versus 25%). Mucosal involvement was noted in 3 (37.5%) patients which was mostly seen on gingiva and floor of the mouth. Most of patients showed mild disease based on a median PDAI of 6 (range 4–12) and the most comorbidities seen in patients were diabetes mellitus (DM) and hypertension (HTN).

Regarding histopathologic and immunologic findings, intra-epidermal blisters with aggregation of neutrophils, eosinophils or both noted in 62.5%, 12.5% and 25% of tissue samples, respectively. The results of DIF study revealed that the majority of patients had intraepidermal versus subcorneal deposition of IgA (75% versus 25%).IgG and C3 deposition in the intercellular space of the epidermis were found in 3(37.5%) and 6 (75%) patients, respectively

Most of the patients (62.5%) experienced complete remission after 6 months. 1 (12.5%) patient had still lesions on prednisolone and dapsone, which was planned to receive rituximab. Two cases were lost to follow-up.

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#### **Conclusion:**

IgA pemphigus is a rarity that must be considered in the differential diagnosis

of vesiculobullosus lesions. Due to the rarity of this condition, there are insufficient data on the best therapeutic options.

Rituximab may be a beneficial treatment for this disease and although it is still controversial,

it should be considered as a possible thrapeutic option for patients with IgA pemphigus.



## Antioxidant and anti-inflammatory properties of black goji berry extract: an in vitro study

Marija Tasic-Kostov<sup>\*1</sup>, Andjela Dragicevic<sup>1</sup>, Jelena Matejić<sup>1</sup>, Nataša Blažević Kamenov<sup>2</sup>, Dragana Pavlović<sup>1</sup>

<sup>1</sup>University of Niš-Faculty of Medicine, Pharmacy department, Niš, Serbia,<sup>2</sup>Primary health center, Dermatology department, Niš

**Introduction & Objectives:** Beyond their traditional use in China and other Asian countries, goji berries, fruits of the *Lycium* species, have gained popularity as "superfoods" on the international market and attracted great interest for topical use. It was implied that *goji berry's* potential *skin* benefits may include anti-inflammatory, anti-aging and antioxidant properties, as well as enhanced UV protection due to high content of specific bioactive compounds. However, these effects have been fairly investigated and documented in the literature. The aim of the study was to evaluate anti-inflammatory and antioxidant properties of the black goji berry extracts *in vitro* in order to estimate its potential for use in dermatology.

**Materials & Methods:** Extracts were macerated from fresh, crushed black goji berry using two different solvents, purified water and 80% propylene glycol. For evaluation of the antioxidant and anti-inflammatory activity of black goji berry macerates, *in vitro* DPPH (1,1-diphenyl-2-picrylhydrazyl) assay and the protein denaturation assay with an aqueous solution of BSA (bovine serum albumin) were applied.

**Results:** Antioxidant activity of black goji berry water extract, with an IC50 range of 221.48  $\pm$  57.12 µg/ml, was significantly stronger than one obtained by liquid extract made with propilenglycol (142.47  $\pm$  39.19 µl). This extract also showed more than twice stronger anti-inflammatory effect with a percentage inhibition of BSA denaturation of 88.679  $\pm$  0.006, comparing to the propylene glycol extract (35.69  $\pm$  0.029 % of inhibition). The standard value for diclofenac as percentage inhibition in BSA test under the same conditions was 95.6 $\pm$ 0.001%.

**Conclusion:** Oxidative stress results from a prooxidant-antioxidant imbalance, leading to cellular damage. This study shows that black goji berry extracts possesses antioxidant and anti-inflammatory properties, which indicates that they could be used in dermatological conditions connected to inflammation and skin redness such as acne and rosacea. The complex intermolecular interactions between the pharmacologically active components of black goji berry and its numerous health-promoting properties suggest that additional research is needed to fully understand its biological effects. Further studies should be conducted to assess the topical safety and efficacy of black goji berry extracts in inflammatory dermatoses.

**Acknowledgements:** Ministry of Education, Science and Technological Development, Program for financing scientific research work, number 451-03-47/2023-01/200113 and the Faculty of Medicine Internal Scientific Project No. 15



## Ablative CO2 Laser Treatment for Hailey-Hailey Disease: Efficacy, Safety, and Implications

Ines Oteiza Rius<sup>1</sup>, Javier Antoñanzas-Pérez<sup>1</sup>, Ana Morelló-Vicente<sup>1</sup>, Elisa María Gómez González<sup>1</sup>, Ane Carrera Gabilondo<sup>1</sup>, Deiane Marcos Muñagorri<sup>1</sup>, Leire Aguado-Gil<sup>1</sup>

<sup>1</sup>Clinica Universidad de Navarra, Dermatology, Pamplona, Spain

## Introduction & Objectives:

Hailey-Hailey disease (HHD), or familial benign chronic pemphigus, is a rare autosomal dominant blistering disorder impacting skin folds. The chronic condition significantly diminishes patients' quality of life, necessitating multiple therapeutic interventions. Despite extensive research on HHD, treatment options remain limited and often provide only temporary relief. Ablative CO2 laser therapy has emerged as a potential alternative in isolated cases. This study aims to investigate the efficacy and safety of ablative CO2 laser treatment for HHD.

## Materials & Methods:

A prospective, randomized, controlled intra-individual comparative study was conducted on ten patients with refractory HHD at the Clínica Universidad de Navarra, Spain. The study spanned from September 2021 to March 2024, with biopsies confirming the diagnosis. Patients underwent CO2 laser treatment on one side, leaving the contralateral side untreated until 6 months later. Follow-up assessments were conducted over two years, measuring various parameters, including affected area reduction, pain intensity, quality of life, and adverse effects.

## **Results:**

The series included 6 women and 4 men, aged 37 to 81, with a mean disease duration of 20 years. Predominantly affecting the groin (92%), genitals (58%), armpits (42%), neck, and buttocks (25%), all patients had previously undergone various unsuccessful therapies. CO2 laser treatment resulted in a reduction of over 90% in lesion extent at 6, 12, and 24 months, accompanied by improved quality of life. No recurrences were observed in treated areas, and adverse effects were limited to textural and pigment changes.

## **Conclusion:**

The use of CO2 laser treatment in HHD patients proves to be a straightforward option with few side effects, all while maintaining its effectiveness for a minimum of two years.



## Effectiveness and Safety of Biologic Therapies for Necrobiosis Lipoidica: A Systematic Review

Martin Heung<sup>1</sup>, Siddhartha Sood<sup>2</sup>, Abrahim Abduelmula<sup>3</sup>, Jensen Yeung<sup>\*3, 4, 5, 6, 6</sup>

<sup>1</sup>McMaster University, Faculty of Health Sciences, Hamilton, Canada,<sup>2</sup>University of Toronto, Temerty Faculty of Medicine, Toronto, Canada, <sup>3</sup>University of Toronto, Division of Dermatology, Department of Medicine, Toronto, Canada, <sup>4</sup>Sunnybrook Health Sciences Centre, Division of Dermatology, Department of Medicine, Toronto, Canada,<sup>5</sup>Women's College Hospital, Division of Dermatology, Department of Medicine, Toronto, Canada, <sup>6</sup>Probity Medical Research, Waterloo, Canada

## Introduction & Objectives:

Necrobiosis lipoidica (NL) is a granulomatous skin condition, indicated by papules and plaques with active erythematous borders, often presenting with chronic ulceration. Emerging literature has reported on the novel use of biologic agents for refractory cases. This systematic review summarizes reported effectiveness and safety outcomes of biologic treatment effectiveness for this indication.

## Materials & Methods:

Following PRISMA guidelines, Ovid MEDLINE and Embase were searched on October 24, 2023 using variations and synonyms of keywords "necrobiosis lipoidica" and "biologic." Twenty-five articles describing 30 patients with NL and 40 instances of related biologic use were included in our review. A descriptive analysis approach was elected for due to the heterogeneity of the data in reports of NL.

#### **Results:**

The mean age of patients was 43.5 years (range: 14-84 years) with 6 (20%) males and 24 (80%) females. There were 93.3% (28/30) of patients that were refractory to non-biologic treatment options. The biologic agents utilized were: infliximab (30.0%, 12/40), adalimumab (27.5%, 11/40), etanercept (20.0%, 8/40), ustekinumab (10.0%, 4/40), secukinumab (10.0%, 4/40), and rituximab (2.5%, 1/40). The greatest proportion of complete resolution was observed with adalimumab (54.5%, 6/11); followed by infliximab (50.0%, 6/12), etanercept (50.0%, 4/8), ustekinumab (50.0%, 2/4); and secukinumab (25.0%, 1/4). Instances of no resolution were observed after use of adalimumab (27.3%, 3/11) and infliximab (8.3%, 1/12). Disease recurrence was observed in 7.5% (3/40) instances of biologics use, once each with etanercept (12.5%, 1/8), infliximab (8.3%, 1/12), and rituximab (100.0%, 1/1). Twelve (30.0%) treatment-related adverse events (AEs) were observed, most commonly reflecting infections (33.3%, 4/12), including one case each of cellulitis, cytomegalovirus colitis, military tuberculosis, and streptococcal pharyngitis. The biologic agents with the greatest instances of AEs were infliximab (10.0%, 4/40), adalimumab (7.5%, 3/40), and secukinumab (7.5%, 3/40). None were reported for etanercept. One (2.5%) patient discontinued adalimumab (7.5%, 3/40), and secukinumab (7.5%, 3/40).

## **Conclusion:**

This review synthesizes available evidence suggesting biologics have a favourable effectiveness and safety profile for NL. Therapies targeting  $TNF-\alpha$ , such as adalimumab, etanercept, and infliximab led to the highest frequency of resolution. As current evidence is limited to case reports and series, larger-scale studies are recommended to further inform clinical application.



## Total serum Immunoglobulin E levels in chronic inflammatory skin diseases and atopic dermatitis in adults

Ilze Upeniece\*<sup>1</sup>, Andreia Vieira<sup>2</sup>

<sup>1</sup>Riga Stradiņš University, Department of Dermatology and Venereology, Riga, Latvia,<sup>2</sup>Riga Stradiņš University, Riga, Latvia

## Introduction & Objectives:

The role of total serum IgE in inflammatory skin diseases is poorly understood. This study was designed to comprehensively evaluate the levels of Total Serum Immunoglobulin E (IgE) in a variety of chronic inflammatory skin diseases in adult patients, including atopic dermatitis, psoriasis, seborrheic dermatitis, allergic contact dermatitis, irritant contact dermatitis, lichen planus, and pityriasis rosea. The aim of this study was to understand and analyze IgE levels, compare them across different diagnoses, investigate any gender and age-related variations, and assess the correlations between IgE levels and age within each condition.

## Materials & Methods:

This study used a retrospective cohort design in which the medical records of patients aged  $\geq$  18 years who were diagnosed with chronic inflammatory skin diseases between January 2015 and December 2022 were reviewed. The collected data included patient age, gender, diagnosis, and Total Serum IgE levels. Descriptive and inferential statistics were used to analyze the data, including the Kruskal-Wallis H test, Mann-Whitney U test, and Spearman's correlation coefficient test. The significance level for the rejection of the null hypothesis was set at p  $\leq$  0.05.

#### **Results:**

This study included 1118 adults diagnosed with various chronic inflammatory skin diseases. The results of the analysis of overall serum IgE levels across various diagnoses indicated that atopic dermatitis had the highest median IgE level (222 IU/mL), whereas other diagnoses displayed median values below 100 IU/mL. In the case of allergic contact dermatitis, gender-related differences were found to be statistically significant, particularly among middle-aged patients. The median total serum IgE levels for allergic and irritant contact dermatitis were within the normal range, but the upper interquartile range still fell well above the upper normal level. Patients with psoriasis displayed median total serum IgE levels within the normal range but with an upper interquartile range exceeding normal limits. Females demonstrated lower median IgE levels than males, with significant age-related variations among younger and middle-aged groups. Other chronic inflammatory skin diseases, such as seborrheic dermatitis, irritant contact dermatitis, and lichen planus, showed no statistically significant gender or age-related variations in total serum IgE levels. Diagnoses such as parapsoriasis and pityriasis rosea were found to have total serum IgE levels within the normal range, but were excluded from further analysis due to their limited sample size.

#### **Conclusion:**

This study provides valuable insights into the Total Serum IgE levels among patients with chronic inflammatory skin diseases, emphasizing the heterogeneity among diagnostic groups. Atopic dermatitis displayed significantly elevated IgE levels, consistent with the literature; however, the biomarker has not been validated for routine clinical use in diagnosis. Gender-specific differences were noted in atopic dermatitis, allergic contact dermatitis, and psoriasis, suggesting potential variations in disease mechanisms. Despite shedding light on these relationships, limitations, such as exclusion of certain diagnostic groups and single-center nature, highlight the need for cautious interpretation and underscoring areas for further investigation.



## Ear and eye involvement in lichen planus: an association that should not be neglected

Yosra Ben Kraiem<sup>1</sup>, Hyba Taounza<sup>1</sup>, Syrine Hamada<sup>1</sup>, Mariam Meziane<sup>1</sup>, Nadia Ismaili<sup>1</sup>, Laila Benzekri<sup>1</sup>, Karima Senouci<sup>1</sup>

<sup>1</sup>Centre Hospitalier Universitaire Ibn Sina, Département de Dermatologie-Vénérologie, Rabat

#### Introduction & Objectives:

Lichen planus is a chronic autoimmune and inflammatory disease of unknown origin, generally affecting the skin, dandruff and mucous membranes, particularly the buccal and genital areas. Involvement of other mucous membranes has been described but is rare.

We report a case of a patient with lichen planus in whom all three mucous membranes were affected ; oral, auricular and ocular.

## **Observation:**

This is a 50-year-old female patient, with no notable history, who was hospitalized for the management of generalized lichen planus evolving for 6 months. The patient described hypoacusis and lacrimation concomitant with skin involvement.

Examination revealed generalized, pruritic, erythematovioline papules and plaques, and a reticulated network of the oral mucosa. Dermoscopy revealed cutaneous wickham striae. ENT examination revealed bilateral deafness, more severe on the right side on audiogram. The ophthalmological examination revealed moderate dryness with reduced visual acuity in the left eye.

Ear and eye mucosal involvement was considered to be related to lichen planus.

The patient was treated with corticosteroids at a dose of 1 mg/kg/day with dexamethasone+tobramycin eye drops. The evolution was marked by a good improvement in hypoacusis, a reduction in lacrimation and the disappearance of dermoscopic signs of lichen activity on the skin, with a clear improvement in pruritus after 2 months.

#### **Discussion:**

Mucosal involvement in lichen is observed in 75% of patients with cutaneous lesions. The oral mucosa is most frequently affected, followed by the ano-genital mucosa. The conjunctiva, larynx, esophagus, squamous epithelium of the external auditory canal and tympanic membrane may also be affected. In the literature, the prevalence of auricular involvement in lichen planus remains unknown. Involvement of the tympanic membranes in lichen planus was first reported by Warin and col in 1948. The most common symptoms are otorrhea, tinnitus, pruritus and hearing loss. The most common complications are progressive stenosis of the external auditory canal and conductive hearing loss, which may be due to chronic inflammation. Topical treatments can relieve these symptoms. In addition, bone-anchored hearing aids represent an optimal solution for restoring hearing in auricular lichen planus. Systemic treatments based on acitretin and oral corticosteroids are reserved for more severe cases with other extra-auricular involvement.

Ocular lichen planus is a rarely reported entity. Conjunctival involvement was first described in 1928, and eyelid involvement in 1938. Clinically, eyelid involvement, dry eyes, scarring blepharoconjunctivitis, keratitis and damage to the lacrimal drainage system can lead to irreversible stenosis and blindness. Cyclosporin and topical corticosteroids are the first-line treatments in ocular involvement, while the treatment regimen in severe forms needs to be boosted by systemic immunosuppressants such as MMF with long-term maintenance therapy to halt the progression of chronic inflammation of the disease.

# **Conclusion:**

Ear and eye involvement should be sought in patients with known skin involvement, and in the face of any recurrent inflammation of the ear or eye. Early detection and management are essential to prevent progressive and permanent complications that threaten functional prognosis.



# Challenges with misconceptions of alopecia areata and opportunities to drive change: A multi-stakeholder expert perspective

Pedro Herranz Pinto<sup>1</sup>, Jean-Marie Meurant<sup>2</sup>, Bianca Maria Piraccini<sup>3</sup>, Nisha Pillay<sup>4</sup>, Olivia Watkins<sup>4</sup>, Ana Sanz<sup>5</sup>, Gregor Shaefer<sup>6</sup>

<sup>1</sup>La Paz University Hospital, Madrid, Spain, <sup>2</sup>Fédération Française de la Peau, Paris, France, <sup>3</sup>University of Bologna, Division of Dermatology, Department of Experimental, Diagnostic and Specialty Medicine, Bologna, Italy, <sup>4</sup>VML Health, London, United Kingdom, <sup>5</sup>Pfizer, Alcobendas, Spain, <sup>6</sup>Pfizer, Berlin, Germany

## Introduction & Objectives:

Alopecia areata (AA) is a systemic autoimmune disorder that affects nearly 2% of the population at some point in their lifetime.1 Characterised by an infiltration of immune cells, it disrupts the normal hair growth cycle, resulting in non-scarring hair loss.1 Despite its autoimmune nature, AA is often unrecognised and misperceived as a cosmetic issue by the public, healthcare professionals (HCPs), payers, and policy makers, impeding effective diagnosis and treatment, and frequently causing patients to be overlooked. An international multi-stakeholder group of experts were convened as the Global AA Consensus Task Force, with the aim to understand the holistic burden of AA and identify strategies to improve patient care and outcomes.

## Materials & Methods:

From April to July 2023, 45 experts, including psychiatrists, psychologists, dermatologists, dermatology nurses, patients, and patient advocates from 15 countries (Australia, Argentina, Brazil, France, Germany, Hong Kong, Italy, Japan, Malaysia, Singapore, Spain, Taiwan, Turkey, UAE, and the US) were engaged. The initiative employed a mixed-methods approach, including a landscape assessment, a survey collecting both qualitative and quantitative data, and a series of five advisory boards.

## **Results:**

The Task Force identified challenges across the AA care pathway, many associated with a lack of understanding and awareness among different stakeholders. Low public awareness and misconceptions around AA are significant barriers to patients presenting to the healthcare system, delaying diagnosis. Experts agreed that low public awareness may limit understanding and drive stigma [91% (41/45)], which may contribute to a lowered quality of life for patients and caregivers.

Experts agreed that many HCPs, especially non-specialists, lack a comprehensive understanding of AA and its holistic impact on patients [93% (42/45)]. Limited primary care physician (PCP) awareness can delay AA diagnosis, with 96% (43/45) of experts agreeing that the care pathway is fragmented with multiple barriers to diagnosis and treatment.

Failure to recognise AA as a systemic autoimmune disease by payers and policymakers can limit their understanding of the need for treatment, and consequently access to it. Most experts agreed that access to AA treatment is low, driven by payer perception, inadequate coverage, and market and regional disparities [82% (37/45)], with particular treatment reimbursement challenges noted in Latin America and Germany.

Opportunities to engage multiple key stakeholder groups to change the perception of AA were identified, such as targeted education for non-medical professionals and PCPs, developing a standardized psychological assessment tool to quantify the patient and caregiver burden of AA as evidence for payers and policymakers, and launching awareness campaigns for the public, patients, and caregivers.

# **Conclusion:**

Patients and caregivers face challenges across the AA care pathway, often intensified by the widespread misconception that it is merely a cosmetic issue. There is an urgent need to shift the perception of AA from cosmetic to a condition with an underlying immunopathogenesis to enhance effective diagnosis, treatment, and support for those affected by the disease.

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#### a giant pyoderma gangrenosum

Hyba Taounza<sup>1</sup>, Yosra Ben Kraiem<sup>1</sup>, Syrine Hamada<sup>1</sup>, Ismaili Nadia<sup>1</sup>, Laila Benzekri<sup>1</sup>, Karima Senouci<sup>1</sup>, Meziane Mariem<sup>1</sup>

<sup>1</sup>Centre hospitalier Ibn Sina, Dermatologie, Rabat, Morocco

#### Introduction & Objectives:

Pyoderma gangrenosum (PG) is a neutrophilic inflammatory dermatosis.

The diagnosis is often made late in the course of the disease.

We report a case of pyoderma gangrenosum evolving for two years.

#### **Observation:**

This is a 39-year-old female patient, with no notable pathological history, who presented to our training center with two giant ulcerations of the left lower limb that had been evolving for 2 years. The patient reported pain assessed at 7/10 according to the EVA and a very significant impact on quality of life, with a DLQI score of 19.

Clinical examination revealed two ulcers on the dorsum of the left foot measuring 14.5 cm long and on the left leg measuring 22.5 cm long, with a purulent base and an irregular, undermined and overhanging, gunmetal-colored border. Functional impotence of the affected limb, with a knee flessum as a complication, was observed in our patient.

A skin biopsy was performed, and the histological appearance was compatible with pyoderma gangrenosum.

The inflammatory work-up was disturbed, with a CRP increased to 176. There was microcytic hypochromic anemia with Hb=6.8g/dl, and thrombocytosis.

No pathological association was found in our patient.

The patient was put on oral corticosteroids with sessions of LED and motor rehabilitation. Progression was marked by improvement and the onset of healing after 3 weeks.

#### **Discussion:**

Pyoderma gangrenosum is a rare inflammatory dermatosis. Clinically, it presents as painful, rapidly progressive ulcers with erythematous, irregular, violaceous margins. The average size reported in the literature is 53.3 mm.

Our case was unusual in that it was of a large size, so in the face of significant loss of substance, the challenge was above all to correct the factors contributing to good healing, and to restore the functional impact.

The pathophysiological mechanism remains uncertain. Nearly half the cases concern patients with systemic inflammatory or onco-hematological pathologies.

Diagnosis is essentially clinical, and is often made late, with an average delay of 23.94 months, as in the case of our patient.

At present, there is no specific therapy for PG, but the treatment involves immunosuppressive drugs in the first line and oral corticosteroids in high doses of 1 to 2 mg/kg/day.

In our patient, the use of LED light helped to accelerate healing. The pathophysiological mechanism remains unclear.

Nearly half the cases concern patients with systemic inflammatory or onco-hematological pathologies.

At present, there is no specific therapy for PG, but the treatment involves immunosuppressive drugs in the first line and oral corticosteroids in high doses of 1 to 2 mg/kg/day.

In our patient, the use of LED light helped to accelerate healing.

#### **Conclusion:**

The diagnosis of pyoderma gangrenosum should be made in the presence of any ulcer with a chronic course.

Early diagnosis can limit both functional and cosmetic sequelae.



## Genital pyodermatitis-pyostomatitis vegetans in patients with hepatitis C cirrhosis: a rare presentation

Juliano Peruzzo<sup>1</sup>, Renata Alves Sanseverino<sup>1</sup>, Ana Victoria Colognese Gabbardo<sup>1</sup>, Mateus Ceolin Vione<sup>1</sup>, Mariani Magnus Da Luz Andrade<sup>1</sup>, Renan Bonamigo<sup>1</sup>

<sup>1</sup>Hospital de Clínicas de Porto Alegre, Brazil

**Introduction:** Pyodermatitis-pyostomatitis vegetans (PSV) is a rare, chronic inflammatory dermatosis characterized by pustules that coalesce into plaques. It most commonly affects the oral mucosa, but is also described in the genital mucosa, scalp, axillae, and groin. It is classically associated with inflammatory bowel disease (IBD), with some suggesting that it may reflect disease activity. Some authors classify PSV as a variant of pyoderma gangrenosum.

**Case report** A 54-year-old male presented with genital lesions that began 15 days earlier. He reported mild pruritus. He was treated with oral acyclovir and topical miconazol without improvement. He has a previous diagnosis of systemic arterial hypertension, type II diabetes mellitus, and cirrhosis due to hepatitis C, and was treating an infected ulcer and osteomyelitis in the right foot secondary to peripheral arterial obstructive disease. Upon physical examination, confluent pustules over an erythematous plaque on the scrotal, inguinal, preputial and glans regions were observed. There were no previous similar episodes or gastrointestinal alterations. Based on the clinical presentation, PSV was suspected, and a biopsy for histopathology (HP) and cultures, as well as VDRL, were performed to exclude differential diagnoses. HP demonstrated the presence of mixed dermal inflammatory infiltrate including numerous neutrophils and eosinophils with ulceration, spongiosis changes with neutrophilic exocytosis. Immunohistochemistry for *Treponema pallidum* was negative, as were direct examinations and cultures for fungi and bacteria from skin biopsy and lesion scrapings. Fludroxycortide cream 0.125mg/g twice daily was prescribed and resulted in lesion remission.

**Discussion** PSV is a rare neutrophilic dermatosis that is more commonly seen in the oral mucosa, but cases solely affecting the genital region have been reported. Differential diagnoses include infectious diseases such as favus tinea, secondary syphilis, and also vegetative pemphigus. HP findings typically include acanthosis with intraepidermal abscesses containing neutrophils and eosinophils alongside an infiltrate with lymphocytes, eosinophils, and neutrophils. In the present case, the clinical presentation was highly suggestive, and the dramatic therapeutic response to corticosteroids was noted. Although nonspecific, HP findings helped to exclude other dermatoses. Other complementary tests also ruled out the main differential diagnoses. Regarding IBD, despite the strong association, its presence is not mandatory for the diagnosis of PSV. However, it is important for patients with PSV to be evaluated for intestinal symptoms, and subclinical IBD should be investigated with colonoscopy.. We emphasize the presence of comorbidities, including cirrhosis due to HCV, as the virus that can act as a trigger for other dermatoses.



## Breast asymmetry and induration: a multidisciplinary challenge

Vilma Grbas<sup>\*1</sup>, Petra Valkovic Zujic<sup>2</sup>, Sandra Peternel<sup>1</sup>

<sup>1</sup>Clinical Hospital Center Rijeka and Faculty of Medicine University of Rijeka, Department of Dermatovenereology, Rijeka, Croatia, <sup>2</sup>Clinical Hospital Center Rijeka and Faculty of Medicine University of Rijeka, Department of Radiology, Rijeka, Croatia

Introduction & Objectives: to present a diagnostically challenging case of an uncommon skin condition

**Results:** A 36-year-old woman presented to a breast surgeon with a 2-month history of gradual change in the color and shape of the right breast resulting in significant breast asymmetry. She reported initial redness of the breast skin with a gradual change to yellowish-brown discoloration, along with progressive induration and disfiguring shape of the entire breast. The patient has never undergone surgery, radiation therapy or had silicone implants, and denied any preceding trauma to the right breast. There was no personal or family history of breast cancer. Mammography showed extremely dense breast tissue, while magnetic resonance imaging of the breast and nipple showed no area of pathologic imbibition. Ultrasound examination revealed no suspicious solid or cystic lesions either, however, there was evident thickening of the dermis of the right breast, which was most pronounced in the lateral quadrants, with a right-to-left thickness ratio of 3:1. Upon discussion by the multidisciplinary breast team, the patient was referred to a dermatologist for evaluation and skin biopsy. Histological analysis revealed expanded reticular dermis with thickened collagen bundles and abundant perivascular and peri-adnexal lymphoplasmacytic infiltrate extending into the panniculus. This excluded a suspected tumoral pathology and confirmed the clinical diagnosis of morphea. Detailed dermatological examination also revealed a single, 2 cm wide, patch of alopecia areata, but screening for systemic autoimmune diseases was negative. The patient was treated with intralesional triamcinolone, along with topical 0.1% tacrolimus ointment, but with little to no response. Significant improvement was achieved upon a cycle of PUVA phototherapy, manifesting as improvement in the overall shape of the breast, palpably softer skin, and objective decrease in the thickness of the involved dermis as measured by ultrasound.

**Conclusion:** Many conditions can diffusely involve the unilateral breast, ranging from various benign inflammatory breast disorders to breast cancer. Morphea is a localized variant of scleroderma, an autoimmune-mediated inflammation resulting in fibrosis of the dermis and subcutaneous tissue, usually without systemic involvement. Considering being a rare disorder with an incidence of 0.4 to 2.7 per 100,000 people, morphea, when located on the breast, is commonly misdiagnosed as breast cancer or infection. Possible triggers of breast morphea include rather diverse conditions such as radiotherapy, injuries, aesthetic surgeries, and silicone breast implants. In our case, the patient had no obvious triggers for the occurrence of the disease, which made the diagnosis even more challenging, especially given the inconclusive results of the radiological imaging methods. When the clinical history and radiological imaging are not conclusive, early referral to a dermatologist is advocated, especially if ultrasound findings indicate a dermal or subcutaneous process. Moreover, interdisciplinary approach with a radiologist may aid not only in the initial workup of the breast pathology but also in the evaluation of the treatment response.



## Prurigo pigmentosa: A case report during pregnancy

Meryem Khallouki<sup>1</sup>, Maryem Aboudourib<sup>1</sup>, Ouiame Karim<sup>1</sup>, Bendaoud Layla<sup>1</sup>, Ouafa Hocar<sup>1</sup>, Said Amal<sup>1</sup>

<sup>1</sup>Mohammed VI University Hospital of Marrakech, Department of Dermatology and Venereology, Marrakech, Morocco

## Introduction & Objectives:

Prurigo pigmentosa is a rare inflammatory dermatosis of unknown cause distinguished by symmetrical, pruritic, recurring erythematous papules that leave a pigmented scarring network on the trunk. Vesicular or bullous forms have been described infrequently. We provide a case of this syndrome in a pregnant woman.

## **Case report:**

A 33-year-old female presented with a pruritic rash that had been developing in remission-like episodes for a year, with the most recent episode lasting 10 days. The woman was pregnant at 10 weeks of amenorrhea, and the papulovesicular rash was limited to the cleavage, back, and neck, with a pronounced butterfly-wing appearance. The pre-pregnancy flareups occurred in an apyretic environment and resolved spontaneously, resulting in hyperpigmentation. The histological examination revealed spongiform dermatosis with perivascular dermal infiltration of lymphocytes, a few histiocytes, and neutrophilic and eosinophilic leukocytes. Direct immunofluorescence was negative. Prurigo pigmentosa was diagnosed based on the characteristic clinical presentation, a lack of a differential diagnosis, and histological findings. The rash resolved on its own, so no treatment was required.

#### **Discussion:**

Prurigo pigmentosa is a rare inflammatory dermatosis that was initially described by Nagashima in Japan in 1971, although other occurrences have been reported around the world, most notably in France. Prurigo pigmentosa is diagnosed mostly clinically, with a distinctive rash of erythematous macules that progresses to urticarial papules or papulovesicles and eventually to reticulated pigmentation. The histological appearance varies greatly depending on the stage of the eruption; the first stage is characterized by a predominantly neutrophilic perivascular infiltrate of the superficial dermis, followed by spongiosis with a mixed lymphocytic and neutrophilic infiltrate, and finally parakeratosis with a lymphocytic perivascular infiltrate. The cause of prurigo pigmentosa remains unknown. However, variables such as extreme youth, anorexia, and type 1 diabetes.

Several examples of prurigo pigmentosa have been reported during pregnancy, including two cases in patients who presented with severe vomiting: the uniqueness of our case stems from our patient's lack of vomiting and the existence of comparable episodes in previous pregnancies. There are several therapeutic options available, with cyclins being the treatment of choice due to their anti-inflammatory properties, and the prognosis is often favorable, with spontaneous remission likely.

#### **Conclusion:**

Prurigo pigmentosa is an uncommon dermatosis with an unknown etiology; diagnosis might be challenging. A clinical and pathological link is essential for reaching a diagnosis. Prurigo pigmentosa is more likely to occur during pregnancy.



## A Spot of Bother

Lisa Murphy\*<sup>1</sup>, Bairbre Wynne<sup>1</sup>

<sup>1</sup>St James's Hospital, Dublin, Ireland

## A Spot of Bother

## Introduction & Objectives:

## Materials & Methods:

## **Results:**

We present the case of a 34-year-old Caucasian female who first presented to dermatology services 4 years ago with a history of red spots appearing on her face. They did not appear elsewhere on her body and she was otherwise asymptomatic from them. She had no past medical history of note and was not taking any regular medications. On examination, there were a number of erythematous, slightly scaly papules and plaques distributed on her face only. Prior to this review, she had attended a GP with a special interest in dermatology. Her initial differential diagnoses included tumid lupus, granuloma faciale and sarcoidosis. A punch biopsy was taken from her left cheek, and this demonstrated the following: changes suggestive of chronic dermatitis with parakeratosis with serum crust, ovrerall most consistent with chronic spongiotic dermatitis. No fungal forms were seen on PAS staining, and there was no evidence of lupus. She was treated with a number of different agents, including oral and topical steroids, doxycycline, and oral anti-fungal agents, with no improvement seen on any therapy. A further punch biopsy was taken from her right temple when she was seen in our service. Differential diagnoses again included sarcoidosis, as well as possible Jessner lymphocytic infiltrate. Histology was once again deemed to be consistent with chronic spongiotic dermatitis. Given that clinical findings and histology did not correlate, she was discussed at an interdisciplinary dermatology conference. Clinical findings were deemed to be most consistent with a condition called facial discoid dermatosis (FDD). FDD was first described in 2010, and given the rarity with which it presents, it is often misdiagnosed. FDD would appear to typically affect those with darker skin types, of Turkish, Indian, or Chinese descent. Our case is unusual in that she was of Caucasian ethnicity. Given the reported presence of demodex mites on some FDD biopsy samples, it has been hypothesized that there may be value in treating with ivermectin, and so we have commenced our patient on topical ivermectin and await her response to this therapy. It is also hypothesized that FDD is a variant of pityriasis rubra pilaris, and there are some reports of improvements on ustekinumab and acitretin. These may be treatment modalities that are worth exploring further in the future. This case highlights the challenges faced in both diagnosing and treating this rare condition, and may raise awareness of this relatively new cutaneous entity.

#### **Conclusion:**



# Pyoderma gangrenosum in the COVID-19 era

Isabel Toh<sup>1</sup>, Omar Nassar\*<sup>2</sup>, Amal Eissa<sup>2</sup>, Sophie Grabczynska<sup>2</sup>

<sup>1</sup>Warwick Medical School, The University of Warwick, Coventry, United Kingdom,<sup>2</sup>Amersham Hospital, Dermatology, Amersham, United Kingdom

Pyoderma gangrenosum (PG) is a rare, non-infectious neutrophilic dermatosis characterised by painful necrotic ulcers with distinctive violaceous undermined edges. In approximately 50% of patients it is associated with systemic autoimmune and inflammatory disorders, most commonly inflammatory bowel disease and rheumatoid arthritis, but also solid tumours and haematologic disorders including leukaemia.1

However, since the COVID-19 pandemic, multiple Dermatology centres have noticed a potential link between COVID-19 and PG.

We present a report of 29 patients who were diagnosed either clinically and/or histologically with PG since 1st January 2020 vs only 10 patients diagnosed with PG between 1st January 2016 to 31st December 2019. The data shows a significant rise in PG cases during and after the pandemic. Unfortunately, we were unable to find a direct link between either contracting COVID-19 infection or COVID-19 vaccination and PG. Of the 29 patients diagnosed with PG during the pandemic, 8 presented before the vaccine was rolled out and 17 presented between 2 months and 2 years after vaccination. No data about vaccinations was available for 1 patient and 3 patients did not have vaccinations. Further data is needed to show any trends, but to our knowledge we have studied the largest cohort looking at the effects of COVID-19 infection and vaccination on pyoderma gangrenosum.

The pathophysiology of PG is poorly understood, but is thought to be due to dysregulated innate immune function leading to altered neutrophil chemotaxis and autoinflammation.1 Multiple authors speculate that exposure to the COVID-19 spike protein triggers an autoimmune response and activation of pro-inflammatory cytokines such as interleukin-6 (IL-6), IL-8 and tumour necrosis factor- $\alpha$  (TNF- $\alpha$ ), which are also implicated in PG.2,3,4

Furthermore, a 76-year old woman who previously presented with PG before the pandemic treated with only topical clobetasol re-presented to our department 10 months after her third COVID-19 vaccination. However, this time her PG was resistant to doxycycline, mycophenolate mofetil, ciclosporin and prednisolone, and only responded to ustekinumab. Similarly, a previous case report describes a 73-year old woman whose PG had been successfully treated with infliximab, ciclosporin and dapsone, but re-presented to a dermatology department 1 month following her second COVID-19 vaccination with PG refractory to oral prednisone, ciclosporin, and adalimumab.3 Therefore, it is also interesting to note that COVID-19 vaccination may also have an effect on the severity and responsiveness of PG to treatments.

Alongside other types of skin reactions, the rate of PG appears to have increased almost three-fold in relation to the COVID-19 pandemic. Whether this is related to contracting different variants of the infection itself, the different vaccinations provided or some other co-factor is unclear.

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## Pyoderma gangrenosum during Infliximab in severe Hidradenitis Suppurativa: a paradoxical event.

Silvia Giordano<sup>\*1</sup>, Federica Repetto<sup>1</sup>, Gabriele Roccuzzo<sup>1</sup>, Sara Boskovic<sup>1</sup>, Paolo Dapavo<sup>1</sup>, Michela Ortoncelli<sup>1</sup>, Simone Ribero<sup>1</sup>, Pietro Quaglino<sup>1</sup>

<sup>1</sup>University of Turin , Medical Science

**Introduction:** Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis in which innate immune response plays a key role. Usually, it is associated with systemic diseases such as inflammatory bowel diseases (IBD) or hematologic disorders. Moreover several drugs have been shown to be related with PG, leading to the hypothesis of a so called "drug-induced" PG entity. In these specific cases many mechanisms could be involved such as immunomodulated dysregulation, imbalance of cytokines, and epigenetics.

**Case:** We present the case of a 60-year-old man with a long history of hidradenitis suppurativa (HS) previously treated with multiple therapies and then with the monoclonal antibody anti-Tumor Necrosis Factor (TNF)-alpha adalimumab, interrupted due to inefficacy. At the first encounter in our clinic, the patient presented with an Hurley 3 HS, with several fistulas, nodules and abscesses involving the armpits, the inguinal region and the buttocks, accompanied by painful and itchy sensations. Based on the IHS4 Score, the disease severity was 20.

In light of the high disease burden and the previously failed lines of therapy, a treatment with another anti TNF-alpha antibody, Infliximab, was started according to the evidence reported in both retrospective studies and open label trials.

After one month of therapy, a remarkable clinical improvement was observed, with a significant reduction in inflammation and pain.

Unfortunately, four weeks later the patient started developing a highly painful ulcer on the left leg. A PG was suspected and confirmed by an incisional biopsy. In the hypothesis of an infliximab-related adverse event, the therapy was discontinued and oral prednisone was initiated.

After two weeks of steroid therapy the lesion had shrunk dramatically together with the pain. Meanwhile the HS started to get worse, with deterioration of the lesions. A new attempt of reintroducing Infliximab was performed but after 2 weeks the PG reappeared. The likelihood of a drug induced adverse event was confirmed, accordingly to Naranjo algorithm.

**Discussion:** To date few cases of PG after TNF-alpha inhibitors have been described. Such events pose a diagnostic and therapeutic challenge, as anti-TNF-alpha agents have been shown potential benefits in the treatment of PG. In this scenario, it can be hard to distinguish between the natural history of the disease and a paradoxical drug-induced event. As for the other reports, it's noteworthy that in all of these cases the anti TNF was prescribed for diseases which are involved in the spectrum of syndromic PG, such as psoriatic arthritis, rheumatoid arthritis and also hidradenitis suppurativa.

It's well known that TNF-a antagonist inhibits CD4+ T-cell proliferation, lowers IFN-gamma, and reduce the T helper (Th)1 as well as the Th17 effector response. A possible explanation of this paradoxical event could be an imbalance of cytokines: the TNF-alpha inhibition could lead to an over production of IFN-alpha by unopposed regulation of plasmacytoid dendritic cells in predisposed patients. Additionally it has been reported that CD4+ lymphocytosis secondary to decreased apoptotic mechanisms can be induced by TNF-a antagonists. In conclusion, TNF-a antagonists can induce PG in susceptible individuals by offsetting the cytokine balance and downregulating the TNF-a-dependent apoptosis pathway. Dermatologists should be aware of this uncommon yet potentially challenging clinical scenario.



## The impact of rosacea on quality of life

Yousef Almheirat<sup>\*1</sup>, Mounia Benkaraache<sup>1</sup>, Nassiba Zerrouki<sup>1</sup>, Siham Dikhaye<sup>1</sup>, Nada Zizi<sup>1</sup>

<sup>1</sup>Mohammed VI University Hospital of Oujda, Department of Dermatology, Morocco

#### Introduction & Objectives:

Rosacea is a common chronic disease characterized by various skin or ocular manifestations. It is more prevalent in women and individuals aged  $\geq$  30 years. It is a frequent reason for consultation in dermatology practices. The demand for consultation is justified by the aesthetic prejudice and psychological discomfort. Treatment failure depends on several factors, including poor therapeutic compliance.

## Materials & Methods:

This is a descriptive monocentric retrospective study, including all patients monitored for rosacea during the period January 2015 to May 2023. The aim of our work is to detail the impact of this disease on the patients' quality of life.

#### **Results:**

We collected data from 144 patients with rosacea. The mean age of the patients was 45.7 [±13.6] years, with a clear female predominance of 91.6% of the patients. The majority of patients had skin phototype III (62.3%). Sun exposure and heat were the most common predisposing factors, found in 64.3% of cases. The most detected clinical form was the papulopustular form (53%), followed by the erythematotelangiectatic form (43%), and thirdly, rhinophyma (4%). Sixteen percent of the patients had associated ocular involvement. Sixty-eight percent of the patients had moderate to severe impairment of quality of life (QoL) according to the Dermatology Life Quality Index (DLQI) score. In terms of treatment, general measures (avoiding flushes, skin care with gentle products, and sun protection) were recommended for all patients. Azelaic acid and topical metronidazole were used in the erythematotelangiectatic form. In the papulopustular form, we combined oral treatment such as tetracyclines or retinoids and topical treatment such as ivermectin, azelaic acid, or 0.75% topical metronidazole, depending on the severity of the disease. The majority of patients (71.3%) reported clinical improvement, but more importantly, an improvement in their quality of life three months after treatment.

## **Conclusion:**

Our study is consistent with the literature regarding the negative impact of rosacea on quality of life. Good therapeutic compliance is recommended for a lesser impact on the patients' quality of life.



#### **Rosacea and Treatment Adherence**

Yousef Almheirat<sup>1</sup>, Hasna Saddouk<sup>1</sup>, Nassiba Zerrouki<sup>1</sup>, Siham Dikhaye<sup>1</sup>, Nada Zizi<sup>1</sup>

<sup>1</sup>Mohammed VI University Hospital of Oujda, Department of Dermatology, Morocco

#### **Introduction & Objectives:**

Rosacea is a common chronic disease characterized by various cutaneous or ocular manifestations. It is more common in women and people aged  $\geq$  30 years. It is a frequent reason for consultation in dermatology clinics. The demand for consultation is justified by the aesthetic damage and psychological discomfort.1 Treatment failure depends on several factors, including poor therapeutic adherence. **Materials & Methods:** 

This is a retrospective descriptive monocentric study, collecting all patients followed for rosacea during the period January 2015-May 2023. The aim of our work is to evaluate adherence to treatment in patients with rosacea.

#### **Results:**

We collected 144 patients with rosacea. The mean age of the patients was 45.7 [±13.6] years, with a clear female predominance of 91.6% of patients. The majority of patients had phototype III (62.3%). Sun exposure and heat were the most common predisposing factors, accounting for 64.3% of cases. The most detected clinical form was the papulopustular form (53%), followed by the erythematotelangiectatic form (43%) and thirdly rhinophyma (4%). Sixteen percent of patients had associated ocular involvement. Sixty-eight percent of patients had moderate to severe impairment of quality of life (QoL) according to the Dermatology Life Quality Index (DLQI) score. Regarding therapeutic management, general measures (avoiding flushes, skin care with mild products, and sun protection) were indicated in all patients. Azelaic acid and topical metronidazole were used in the erythematotelangiectatic form. In the papulopustular form, we associated oral treatment such as tetracyclines or retinoids and topical treatment such as ivermectin, azelaic acid, or topical metronidazole at 0.75% depending on the severity of the disease. The majority of patients (71.3%) reported clinical improvement but especially an improvement in their quality of life three months after treatment. For therapeutic adherence, poor therapeutic adherence was detected in 33% of patients, 16.3% of whom stopped topical treatment due to the appearance of adverse effects, including facial irritation, 20.6% due to the cost of treatment, 45.7% stopped treatment due to clinical improvement, and 17.4% found that applying several products per day was impossible. It was also noted that adherence to oral treatments is better than that of topical treatments.

#### **Conclusion:**

Our study supports the literature regarding the negative effect of rosacea on quality of life. We recommend good therapeutic adherence for a lesser impact on the quality of life of patients. For this, we emphasize the role of clinicians in educating patients about the nature of rosacea as a chronic disease that requires long-term treatment, as well as the importance of a good skin care routine and avoidance of triggering factors.



## Full blood count monitoring for isotretinoin therapy : are we generating unnecessary expanses?

Romesh Paul Raj Suresh John\*<sup>1</sup>, Francesca Melindo<sup>1</sup>, Isabelle Vandormael<sup>1</sup>, Mabs Chowdhury<sup>1</sup>

 $^{1}$ University Hospital of Wales , Welsh Institute of Dermatology , Cardiff, United Kingdom

#### Introduction & Objectives:

Isotretinoin is a retinoid derivative of vitamin A used to treat severe recalcitrant acne by targeting the production of sebum and keratin. This medication has been associated with several side effects including serum abnormalities. The British National Formulary classifies the risk of anaemia, neutropenia, and thrombocytopenia as common or very common side effects. In comparison, hepatitis is considered a rare or very rare adverse event, whilst hyperlipidaemia is listed as a contraindication. The BNF recommends close monitoring of hepatic function and serum lipids however there is limited guidance on full blood count monitoring.

## Materials & Methods:

This study identifies the prevalence of anaemia and other biochemical abnormalities including neutropenia, thrombocytopenia, hyperlipidaemia, and transaminitis in adult patients taking isotretinoin. Data was collected retrospectively using our clinical portal from March 2018 to April 2023. 72 adult patients (18 male and 54 female) treated with isotretinoin for severe acne were identified. Abnormalities in blood parameters were reviewed with a focus on full blood count (FBC), lipid profile and liver function (LFTs).

#### **Results:**

All patients had pre-isotretinoin blood monitoring. 2.8% (n=2) were found to be anaemic after commencing isotretinoin. However, both patients had a pre-existing history of anaemia secondary to iron deficiency anaemia and B-thalassaemia trait respectively. 4.2% (n=3) of patients developed leucopenia, 1 patient spontaneously resolved during several months of treatment, 1 patient had leucopenia prior to treatment which improved on treatment but still remained leucopenic, and 1 patient had pre-existing leucopenia which worsened on treatment. 1.4% (n=1) of patients developed neutropenia post-treatment.

5.6% (n=4) of patients had a rise in LFTs whilst on treatment. 11.8% (n=8) of patients had hypertriglyceridemia post-treatment. 27.9% (n=19) of patients had new or worsening hypercholesterolemia post-treatment.

## **Conclusion:**

Full blood count costs approximately £6 however this estimation does not include health care professional time, equipment used or travelling cost. On average, patients underwent blood monitoring at least twice during the course of their treatment therefore the total cost of FBC testing was £864 for our cohort. Patients also had urea and electrolytes (U&Es) checked at £6 per test. Both LFTs and lipid profiles are estimated at £23 each. A full panel of blood tests would cost approximately £58 per patient. Our clinical practice should involve careful selection of the blood tests requiring monitoring e.g. hepatic function and lipid profile.

Our study did not identify any significant anaemia or other FBC abnormalities with isotretinoin treatment. We therefore recommend the cessation of FBC monitoring which will also save significant costs across clinical services.



## Linear lichen planus pigmentosis of the face: A rare clinical presentation

Dalel Kemicha<sup>1</sup>, Mouna Korbi<sup>1</sup>, Sboui Karama<sup>1</sup>, Zakhama Abdelfateh<sup>2</sup>, Belhadj Ali Hichem<sup>1</sup>, Zili Jameleddine<sup>1</sup>

<sup>1</sup>Fattouma bourguiba, monastir, dermatology department, <sup>2</sup>Anatomopathology Department

## Introduction & Objectives:

Lichen planus pigmentosus (LPP) is a rare chronic variant of lichen planus. Linear LPP is an uncommon subtype, with only a few cases of facial involvement reported. Herein, we report a rare presentation of LPP with a unilateral linear pattern along the Blascko lines.

## Materials & Methods:

A 58-year-old woman (Fitzpatrick skin type III) with a medical history of diabetes, hypertension and dyslipidemia, presented with a one-year history of slightly pruritic lesion on the chin. Physical examination revealed dark brown macules from the left side of the chin to the underside of the jaw, along the neck, with a linear distribution. Examination of the oral mucosa showed dental amalgams, absence of lacework and erosions. Nails and mucous membranes were not involved. Dermoscopy showed pigmentation with brown dots and erythematous areas. Skin biopsy was consistent with a diagnosis of LPP. Routine biological examination data including blood cell count, biochemistry, and serology for hepatitis C were within normal ranges. Patch testing with dental Screening Series were also negative. The patient was treated with a topical corticosteroid, resulting in the improvement of the lesions after two months.

#### **Results:**

LPP is a rare chronic variant of LP mainly seen in darker skin types, its distribution tends to be symmetrical and is frequently seen on the face, neck, and trunk. Linear LPP is characterized by chronic acquired hyperpigmented, dark brown to grey, macular pigmentation located on sun-exposed areas of the face, neck, and flexural folds. The trunk is the most frequently affected location in blaschkoid and segmental patterns, while the face and limbs are mainly affected in linear LPP. Only a few cases reported a unilateral linear pattern along the Blascko lines, as seen in our case. Clinically, it differs from classical LP by the longer clinical course and rare involvement of the scalp, nail, or mucosal area. The pathophysiology of linear LPP is poorly understood, though it is postulated to be caused by T-lymphocyte autoimmunity against keratinocytes. It has also been proposed to be associated with hepatitis C infection, ultraviolet (UV) radiation, and contact allergens. Histologically, LP and LPP are quite similar, but melanin incontinence is more predominant in LPP. As in other variants of LPP, treatment of linear subtype consists of topical steroids.

## **Conclusion:**

LPP can manifest in diverse atypical patterns. The occurrence of LPP in a blaschkoid distribution is uncommon, emphasizing the importance of considering it in the differential diagnosis of unilateral hyperpigmentation along the Blaschko lines, especially in individuals with darker skin types.



# Comparison of the efficacies of topical rosemary extract and topical 2% ketoconazole lotion in the treatment of seborrheic dermatitis

Maryam Sadat Sadati\*<sup>1</sup>, Fatemeh Alsana<sup>1</sup>, Mohamad Mehdi Parvizi<sup>1</sup>

<sup>1</sup>shiraz university of medical sciences , dermatology , shiraz, Iran

# Introduction & Objectives:

Seborrheic dermatitis, a chronic inflammatory skin disorder, manifests as red, scaly, greasy, and itchy lesions, commonly affecting sebum-rich areas like the scalp and face. Its prevalence is 11.6% in the general population, with various internal and environmental factors contributing to its development.

## Materials & Methods:

This double-blind, randomized intervention included 42 patients with scalp seborrheic dermatitis, divided into rosemary lotion and ketoconazole lotion groups. Both lotions were applied twice daily for two months.

## **Results:**

Ketoconazole demonstrated a more significant reduction in the ASFS score compared to rosemary. Rosemary was more effective in reducing itching during the first and second months. Both groups experienced a significant decrease in the DLQI scale, with no notable difference between them. Adverse effects led two patients in rosemary group to discontinue the study, while one patient in ketoconazole group reported itching.

## **Conclusion:**

Both rosemary and ketoconazole lotions showed efficacy in treating scalp seborrheic dermatitis, with ketoconazole having a more pronounced effect. Rosemary could be considered as an alternative treatment, particularly in cases of azole nonresponse and patients with severe pruritus



## An atypical case of superficial granulomatous pyoderma involving the face

Yasmine Mahdar<sup>\*1</sup>, Fouzia Hali<sup>1</sup>, Bouchra Baghad<sup>1</sup>, Ahmed Aziz Bousfiha<sup>2</sup>, Soumiya Chiheb<sup>1</sup>

<sup>1</sup>Ibn Rochd Hospital Center, University Hassan II, dermatology, casablanca, Morocco,<sup>2</sup>Abderrahim El Harouchi Children Hospital, University Hassan II, pediatric infectious and immunological diseases, casablanca, Morocco

## Introduction:

Pyoderma gangrenosum is a rare ulcerative cutaneous disease classified under the neutrophilic dermatoses group of inflammatory dermatoses. Superficial granulomatous pyoderma is characterized by its indolent clinical course, lack of associated underlying disease, the finding of a granulomatous infiltrate on histology, and better prognosis. While it predominantly affects the lower extremities, it can also manifest in other areas of the skin. However, involvement of the face has rarely been reported.

We report a case of annular ulcerative plaques on the face revealing a superficial pyoderma gangrenosum.

## **Case report:**

A 15-year-old patient, born from a first-degree consanguineous marriage, presented with annular ulcerative plaques with keratotic borders, evolving over 7 months, induced at sites of skin trauma. These plaques were located on the face, neck, and upper limbs, accompanied by pustules and impetiginized crusts. The patient also reported inflammatory arthralgia. Previous treatment with antifungals and antibiotics yielded no improvement. Cultures for bacteria and fungi were negative. Laboratory tests revealed an inflammatory syndrome without evidence of immune deficiency. The autoimmune panel showed no abnormalities. Bacterial and fungal cultures were negative. Histological examination revealed a polymorphic ulcerated granulomatous dermatitis with an inflammatory infiltrate rich in neutrophilic polymorphonuclear cells. Based on these clinical, histopathological, and laboratory findings, a diagnosis of pyoderma gangrenosum was established. The patient was initiated on oral corticosteroid therapy at 0.5 mg/kg/day along with dapsone 50 mg/day, leading to lesions' regression after 1 month of treatment.

## **Conclusion:**

Superficial pyoderma gangrenosum is a rare variant, infrequently reported on the face, with only a few case reports found in the literature. It tends to be more refractory to conservative treatment and responds better to intensive treatment with systemic corticosteroids or immunosuppressants. To avoid disease progression and permanent disfigurement with this apparent condition, it is imperative for physicians to identify both the clinical and histopathological features of superficial pyoderma gangrenosum and to conduct an adapted aggressive medical treatment.



## Association of cutaneous leishmaniasis and discoid chronic lupus : A local immune reaction ?

Bouchra Amine<sup>1</sup>

<sup>1</sup>CHU IBN ROCHD, Dermatology, casablanca, Morocco

#### Introduction & Objectives:

Cutaneous Leishmaniasis (CL) is a protozoan disease caused by Leishmania spp. and is endemic in Morocco. CL could be associated with other diseases, especially immune system disorders representing a diagnostic challenge. Herein, we describe a Moroccan patient with CL associated with Discoid chronic lupus erythematosus on the same location.

## **Case report:**

A 52-year-old patient from Fkih Ben Saleh (an endemic area for leishmaniasis in Morocco), with no medical history, presented with papuloverrucous lesions on the face evolving for 3 years. Clinical examination revealed a violaceous papuloverrucous plaque with an atrophic center on the nose resting on an infiltrated base, annular papular lesions on the mandibular and zygomatic regions of the left cheek, a small verrucous lesion on the right cheek, and finally squamous papular lesions on the concha of the left ear. Dermoscopic examination showed erythema, starry white patterns at the periphery, lupoma grains, hyperkeratosis with a yellowish tear-like appearance, and telangiectasias. PCR identified a *L infantum* species and the patient was treated with intramuscular Glucantime. Given the lack of improvement of lesions of the lesions , a skin biopsy was performed and was consistent with a discoid cutaneous chronic lupus. There was no systemic involvement of lupus .Treatment with hydroxychloroquine resulted in a favorable outcome after 12 months of follow-up. \*\*

#### **Conclusion:**

Our patient illustrate a rare case of CL occurring on the same location of Discoid lupus. This isotopic response suggest a local immune dysregulation involving the activation of type-1 interferon. Dermatologists must be aware of this phenomenon in order to establish an early and appropriate treatment



## Pediatric Multisystem Inflammatory Syndrome associated with SARS-CoV-2 mimicking Kawasaki disease.

Meryame Hammouch<sup>1</sup>, Fatima-Zahra El Fatoiki<sup>1</sup>

<sup>1</sup>CHU ibn rochd, dermatology, casablanca, Morocco

#### **Introduction & Objectives:**

Pediatric Multisystem Inflammatory Syndrome (PIMS) is a complication that can develop in children after infection with SARS-CoV-2. It occurs several weeks after the infection, which may go unnoticed. We report the case of a child with PIMS associated with a documented COVID-19 infection mimicking Kawasaki disease.

## **Case report:**

A 13-year-old child, without any particular medical history, was admitted to the emergency department for a skin rash associated with prolonged fever lasting more than six days, with a history of digestive disturbances including vomiting and diarrhea, and inflammatory arthralgia. The clinical examination revealed an agitated child, with a fever of 40°C, tachycardia, and dyspnea. Bilateral conjunctivitis was observed, and the dermatological examination showed diffuse infiltrated erythematous lesions in pseudo-target-like patterns, with a blistered center in some areas, located on the trunk and limbs, involving the external genital organs, and some purpuric lesions on the lower limbs. There was also mucosal involvement in the form of erosive cheilitis with intraoral erosions. Laboratory tests showed a CRP of 82.9 mg/L, procalcitonin at 18 ng/mL, lymphopenia, thrombocytopenia, and hypochromic microcytic anemia with a ferritin level of 141.4 ng/mL. Hyponatremia was also present. Blood cultures and viral serologies were negative, and the immunological assessment was also negative. A COVID-19 PCR test was performed, which was negative, but IgG serology was positive at 9.83. The diagnosis of PIMS was established after ruling out other etiologies. The patient was treated with antibiotics, symptomatic therapy, and correction of hydroelectrolytic disorders, resulting in significant improvement.

#### **Conclusion:**

The diagnosis of PIMS is rare, so it is essential to consider other, sometimes more common, diagnoses such as infectious etiologies, inflammatory conditions, and drug reactions. Despite the potential severity of the condition, rapid management generally ensures an excellent medium-term prognosis for the vast majority of children. Early recognition of PIMS is therefore crucial, and every physician caring for children should be familiar with the clinical signs that should raise suspicion of this diagnosis. Children with PIMS typically present with poorly tolerated acute fever, gastrointestinal symptoms such as abdominal pain, vomiting, diarrhea, and a skin rash. Less commonly, they may have mucosal involvement and changes in the extremities. Other criteria of Kawasaki disease, such as cervical lymphadenopathy and conjunctival erythema, are less frequent. In some cases, patients develop shock due to myocardial failure within 3 to 5 days after the onset of fever, which represents the severity of the syndrome.

The COVID-19 pandemic has led to many surprises, including the emergence of PIMS, whose clinical and biochemical characteristics are distinct from those of classic Kawasaki disease. The fundamental aspects of PIMS are still largely unknown. Early and appropriate management helps prevent the majority of deaths.



## Annular elastolytic giant cell granuloma resistant to treatment: A case series

Jan Stanič<sup>\*1</sup>, Vanesa Koracin<sup>2</sup>, Vid Bajuk<sup>1</sup>, Barbara Kecelj Žgank<sup>1</sup>, Borut Žgavec<sup>1</sup>

<sup>1</sup>University Clinical Centre Ljubljana, Department of Dermatovenereology, Ljubljana, Slovenia,<sup>2</sup>General hospital Novo mesto, Department of Dermatovenereology, Slovenia

## Introduction & Objectives:

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous disease. Annular plaques appear on photoexposed or covered areas of the skin, likely as an immune response to damaged elastic fibers. Histopathological examination shows granulomatous inflammation, multinucleated giant cells and more than one area of elastophagocytosis, without necrobiosis. Spontaneous regression of the lesions is possible, as well as resistance to treatment. Appropriate sun protection is advised in order to prevent the development of new lesions.

We present two patients with AEGCG, initially diagnosed as granuloma annulare (GA).

## Materials & Methods:

Case 1: 67- year old patient presented with itchy annular erythematous lesions on her back, right shoulder, and temples. Three years back a histological diagnosis of GA was confirmed. She received treatment with topical corticosteroid ointment. After progression of the lesions she was started on therapy with dapsone 50 mg daily, increased to 100mg daily after 4 months. Additionally, she applied topical 0.1% tacrolimus. Macrocytic anemia and further spreading developed, therefore the treatment with dapsone was ceased and she was started on hydroxychloroquine 200 mg daily.

Case 2: 66-year old patient presented with annular lesions on the dorsum of the hands and upper extremities that were present for years. First histopathologic examination suggested GA. Topical therapy (corticosteroid creams, terbinafine and calcipotriol) and later treatment with cryotherapy, doxycycline and PUVA therapy were all unsuccessful. A second biopsy was performed with histopathologic examination revealing elastolytic granulomas, suggesting AEGCG as more appropriate diagnosis.

## **Results:**

Case 1: Due to the poor response to treatment and the distribution of lesions on light-exposed areas of the skin, we reperformed a biopsy. There was a pattern of relatively superficial granulomatous dermatitis and elastophagocytosis, which changed the working diagnosis to AEGCG.

Therapy with hydroxychloroquine was discontinued due to reported side effects. She declined the suggested therapy with isotretinoin or acitretin, and only used indifferent ointment. She was advised regarding sun protection and avoidance measures. Strict sun protection led to a significant improvement.

Case 2: Additional treatment with cryotherapy, doxycycline and local PUVA therapy was unsuccessful. Second biopsy was performed, which suggested AEGCG as more appropriate diagnosis. Strict photoprotection was recommended.

# **Conclusion:**

Clinically, AEGCG and GA cannot be definitely distinguished from each other. Therefore, it is important to perform a biopsy of the lesion, including the erythematous edge and suggest possible diagnosis to the histopathologist.

Both our cases required a second biopsy for the proper diagnosis, since initially clinical suspicion of AEGCG was not

suspected and/or communicated to histopathologist. In both cases significant resistance to various treatment regimens was observed. In patient number 1, improvement occurred following strict sun protection rules and sun avoidance measures, but we could also not rule out the spontaneous regression of skin lesions.



## Rare Form of Cutaneous Sarcoidosis: Annular Sarcoidosis - A Case Report

Bouchra Amine<sup>1</sup>

<sup>1</sup>CHU IBN ROCHD, Dermatology, casablanca, Morocco

#### Introduction & Objectives:

Sarcoidosis is a granulomatous disease involving multiple systems in the body. Skin involvement can occur in up to 25% of patients. Annular sarcoidosis is a rare variant of cutaneous sarcoidosis. Early recognition of this form of sarcoidosis is important because it is usually associated with systemic involvement.

## **Case report:**

A 62-year-old woman presented with itchy lesions on the face for 8 years with notion of dyspnea on exertion. The lesions first started on the right cheek and gradually spread to involve the rest of the face. On skin examination, she was found to have dull coin-shaped erythematous plaques and annular erythematous-squamous plaques ranging from 1 cm × 1 cm to 5 cm × 8 cm scattered over the forehead, cheeks, chin, and scalp. On dermoscopy, a structureless area with linear vessels and yellow-orange areas were observed. A chest CT scan objectified bilateral mediastinal and hilar adenopathies with bilateral pulmonary nodules. Skin biopsy confirmed the diagnosis of cutaneous sarcoidosis.

#### **Conclusion:**

Sarcoidosis is a non-caseating granulomatous disease involving multiple systems in the body. Skin involvement is seen in up to 25% of patients with sarcoidosis and is present with or without systemic involvement. Approximately 25% of the skin lesions in sarcoidosis occurred on the face. Skin involvement in sarcoidosis can be divided into either specific and nonspecific depending on the presence of granulomas in the former, or into acute erythema nodosum and chronic granulomatous form. The skin lesions in sarcoidosis can be of various morphologies, and therefore it has been known as one of the "great imitators". Annular lesions are well known forms of cutaneous sarcoidosis, accounting for approximately 8% of all cutaneous lesions, and are a rare entity that may or may not point to the presence of systemic involvement. Cutaneous involvement not only facilitates early diagnosis due to the presence of non-caseating granulomas, but may also predict the presence of systemic involvement. In addition to skin involvement, our patient also had a pulmonary systemic location.



## Acute generalized exanthematous pustulosis following a spider bite

Billel Merrouche<sup>1</sup>, Houria Sahel<sup>1</sup>

<sup>1</sup>Chu Maillot, Dermatolgy, Algiers, Algeria

## Introduction & Objectives:

Acute generalized exanthematous pustulosis (AGEP) is an uncommon, severe dermatosis characterised by the sudden development of a diffuse febrile erythema dotted with amicrobial pustules. Over 90% of cases are thought to be caused by drugs. Its occurrence after a spider bite is extremely rare. We report a new case.

## Materials & Methods:

A 43-year-old woman treated for hypothyroidism with levothyrox® for several years presented with an acute erythematopustular rash in a febrile setting. On questioning, the patient reported having been bitten by an insect during an outing to a leisure park 24 hours before the onset of symptoms. There had been no infectious episode or recent medication prior to the clinical presentation. The rash, initially localized to the site of the bite (left flank), rapidly spread to the trunk, limbs and folds, and was covered with millimetric, non-follicular pustules. Biological tests showed moderate hepatic cytolysis (transaminases twice normal). The patient was treated with general corticosteroid therapy at a dose of 0.5 mg/kg/d. Clinico-biological evolution was rapidly favorable.

#### **Results:**

PEAG is most often caused by drugs, particularly antibiotics such as  $\beta$ -lactams and macrolides. In our patient, the absence of the causative drug, the fact that she had been bitten by a spider, and the sequence of events, with a 24-hour delay between the bite and the appearance of the rash, which began at the site of the bite before spreading to the entire integument, led us to make the diagnosis of AGEP due to a spider bite. In the literature, a few cases of agep have been published following spider bites, notably of the genus Loxosceles. In our case, the spider involved could not be identified.

#### **Conclusion:**

Spider bites should be considered as a possible cause of AGEP. However, we have yet to understand the pathophysiological mechanisms involved in this type of reaction.



#### Scarring alopecia of the scalp revealing systemic sarcoidosis: a case report

Lamia Chalouli<sup>1, 2</sup>, Slaouti Mounia<sup>2</sup>

<sup>1</sup>Central Military Hospital of Algiers, médecine, algiers, Algeria,<sup>2</sup>Hospital, médecine, algiers, Algeria

## Introduction & Objectives:

The cutaneous manifestations of sarcoidosis are common and often indicative of the disease. Among them, cicatricial alopecia of the scalp remains rare. We report a case observation

## Materials & Methods:

Case :

A 42-year-old female patient, with no significant medical history, presented with three annular erythematous alopecic plaques with atrophic center, located in the frontal region of the scalp, measuring 1 to 2 cm in diameter. Dermoscopic examination revealed yellow-orange areas and multiple telangiectasias. Clinical examination revealed productive cough persisting for 1 year. Dermal infiltration composed of epithelioid and giant cell granulomas without necrosis was found on histological examination of a skin biopsy. Investigations for mycobacterial infection, tuberculosis, leishmaniasis, deep mycosis, and bacterial cultures were negative. Evaluation for systemic manifestations of sarcoidosis revealed elevated serum angiotensin-converting enzyme levels and parenchymal lung involvement without fibrosis, along with cervical lymphadenopathy.

## **Results:**

A diagnosis of sarcoidosis was established, and the patient underwent treatment with systemic corticosteroids (0.5 mg/kg/day of prednisone) and intralesional corticosteroid injections, resulting in partial hair regrowth at 6 months.

Scalp involvement in sarcoidosis, although rare, remains possible and typically results in cicatricial alopecia. Diagnosis relies on the confrontation of clinical and histological elements, as well as the exclusion of other differential diagnoses. In sarcoidosis, an active border often persists, as observed in our patient. Dermoscopy can be of great assistance in diagnosis, frequently revealing grouped translucent orange ovoid structures, linear vessels around the hair follicle orifice, and dystrophic hairs. Scalp involvement is rarely isolated and is often associated with systemic manifestations, particularly pulmonary involvement, especially among African Americans. The often-delayed diagnosis (at the cicatricial stage) partly explains the failures observed with the treatments commonly used in cutaneous sarcoidosis (synthetic antimalarials, local and systemic corticosteroids)

## **Conclusion:**

It is important to be aware of this clinical manifestation in order to enable early diagnosis and appropriate treatment to halt the sometimes severe and irreversible progression of this disease.



## Psoriasis vulgaris and atopic dermatitis: a therapeutic challenge

Daciana-Elena Branisteanu<sup>1</sup>, Elena Andrese Porumb<sup>1</sup>, Mihaela-Paula Toader<sup>2</sup>, Diana Sinigur<sup>3</sup>, Mihaela Cojocaru<sup>3</sup>, Diana-Stefania Dirzu<sup>3</sup>

<sup>1</sup>Department of Dermatology, 'Grigore T. Popa' University of Medicine and Pharmacy, 700115 Iasi, <sup>2</sup>Department of Oral Dermatology, 'Grigore T. Popa' University of Medicine and Pharmacy, 700115 Iasi, <sup>3</sup>Department of Dermatology, Railway Clinical Hospital, 700506 Iasi

## Introduction & Objectives:

Psoriasis (PsO) and atopic dermatitis (AD) are both chronic, inflammatory skin conditions associated with significant morbidity. Patients with overlapping PsO and AD frequently develop hand involvement, respond poorly to topical therapy and may require multiple systemic medications to be properly managed. Since palmoplantar involvement can cause significant functional, social, and psychological impairment, an accurate diagnosis and treatment is crucial.

## Materials & Methods:

Here we report the case of a female patient, known with a 3-year history of severe palmoplantar PsO who underwent topical and systemic treatment according to the therapeutic protocol. She is currently under biologic therapy with anti Il17 agent and continues to have recalcitrant lesions.

#### **Results:**

A 69-year-old patient is known to our clinic with PsO vulgaris and spondylitic and polyarticular psoriatic arthropathy. The onset was in September 2020 with scaly erythematous plaques located on the chest, abdomen, palms and soles and was histopathologically confirmed in 2021.

Initially, the patient followed topical treatments based on topical corticosteroids, vitamin D derivatives, emollients, and systemic treatment with Methotrexate for 6 months, but as a result of the unfavorable evolution, it was decided to initiate biological therapy with Guselkumab. However, the lesions persisted on the palmoplantar level, leading to a plantar biopsy in March 2022, which established the diagnosis of plantar hyperkeratotic eczema. Bacteriological and mycological examinations at the level of palmoplantar fissures have repeatedly detected E. coli, Staphylococcus aureus and Candida albicans, which required antibiotic and antifungal treatments. After 11 months, the ESIF score remained unchanged, leading to a switch to Secukinumab. A period of gradual improvement ensued; nonetheless, the lesions continued to significantly affect the patient's quality of life due to the painful fissures and intense palmoplantar itching. In May 2023, a right sole biopsy revealed plantar eczematous dermatitis and systemic treatment with corticosteroids was instituted, with modest results. In December 2023, following an assessment of the Hanifin and Rajika criteria, the diagnosis of AD was made. She then underwent topical treatment with Pimecrolimus 1% cream b.i.d., leading to complete remission of the palmoplantar lesions after only one month of treatment.

#### **Conclusion:**

We presented a case of a patient with overlapping features of PsO and AD. She had severe involvement of palms and soles. Despite the failure of biologic drugs, including gold-standard therapies for PsO, our patient achieved complete remission of the lesions solely after the addition of calcineurin inhibitors to the medication regimen.

In a patient with known history of PsO with recalcitrant palmoplantar lesions, AD should be considered and the treatment must be adapted bidirectionally so as to target both conditions.

Conventional immunosuppressants are not recommended for long-term use due to safety concerns. All biologics approved for PsO did not show high efficacy in AD trials, while Dupilumab is ineffective on PsO. The use of biologics in combination has not yet been thoroughly studied.

The association of the two diseases is not a novelty, which is why further research would be necessary to identify etiopathogenic similarities at the molecular level in order to develop a suitable treatment that targets the shared pathways.



# **Extragenital Lichen Sclerosus et Atrophicus**

Hind Ouerghi\*<sup>1</sup>, Rym Kort<sup>1</sup>, Olfa Khayat<sup>2</sup>, Achraf Chadli Debbiche1<sup>2</sup>

<sup>1</sup>Tunis El Manar University, Faculty Of Medicine Of Tunis, Dermatology Outpatient Department, Ben Arous, Tunisia,<sup>2</sup>Tunis El Manar University, Faculty Of Medicine Of Tunis, Habib Thameur Hospital, Pathology Department, Tunis, Tunisia

## Introduction & Objectives:

Lichen Sclerosus et Atrophicus (LSA) is an inflammatory dermatosis with a chronic course, predominantly affecting the anogenital area. Isolated cutaneous involvement is uncommon. We report a case of lichen sclerosus in a woman with purely cutaneous involvement, thus highlighting the importance of considering this diagnosis even in the absence of apparent genital involvement.

## Materials & Methods: N/A: Case report.

## **Results:**

An 86-year-old female with a history of hypertension and diabetes presented with diffuse, moderately pruritic, white lesions that had been evolving for four years. The lesions initially affected the lower trunk before extending to left shoulder, and left breast, with later development of diffuse cutaneous sclerosis.

Clinical examination revealed pearly white and atrophic plaques, confluent in large patches in places on the lower trunk, left shoulder, and left breast. The rest of the clinical examination, particularly the cutaneo-mucosal and general examination, was normal. Laboratory studies, including complete blood count, biochemistry, antibodies against the thyroid, antinuclear antibodies, and serology for Borrelia, hepatitis B, and C viruses, were within normal limits or negative.

A punch biopsy was performed on a hypopigmented macule. Histopathology showed focal epidermal atrophy with effacement of the rete ridges and vacuolar degeneration of the basal layer. There was pronounced edema, homogenized sclerosis of the collagen in the upper dermis and a lymphocytic inflammatory infiltrate in the mid-dermis, consistent with LSA. The patient was prescribed a mid-potency topical steroid and hydroxychloroquine. Assessment of the evolution under treatment was not possible as the patient was lost to follow-up.

## **Conclusion:**

Cutaneous involvement in LSA primarily poses an aesthetic issue due to its chronic, leukodermic, and atrophic evolution. Unlike genital lichen sclerosus, extra-genital LSA rarely complicates into malignant transformation.



## Vitamin D and cathelicidin levels in seborrheic dermatitis and acne

Olena Sarian<sup>1</sup>

<sup>1</sup>Kharkiv National Medical University, department of dermatovenereology and surgical dermatology, Kharkiv, Ukraine

## Introduction & Objectives:

Seborrheic dermatitis (SD) and acne vulgaris (AV) are common skin diseases frequently encountered in clinical practice. Dermatoses are chronic diseases of lipid-rich areas of the skin or pilosebaceous unit. Although SD and AV have been recognized clinical conditions for decades, their etiology is far from clear. Numerous skin functions are regulated by vitamin D and/or its receptor. These include inhibition of proliferation, stimulation of differentiation, including the formation of a permeability barrier, stimulation of innate immunity, and others. Activation toll-like receptor 2 leads to the induction of CYP27B1, which in turn induces cathelicidin, leading to the destruction of pathogenic microorganisms.

## Materials & Methods:

A total of 40 patients with SD and AV, 20 healthy controls aged 22-45 years were included in the study. Serum 25hydroxyvitamin D3 (25(OH)D), LL-37 levels were measured in the patients and control groups, and a comparison was made between the two groups regarding these parameters. All patients and control subjects were studied during one season to avoid seasonal variations in vitamin D levels.

#### **Results:**

A total of 40 patients with SD and AV and 20 healthy controls aged 22-45 years were included in the study. Serum 25hydroxyvitamin D3 (25(OH)D), LL-37 levels were measured in the patients and control groups, and a comparison was made between the two groups regarding these parameters. All patients and control subjects were studied during one season to avoid seasonal variations in vitamin D levels.

## **Conclusion:**

Decrease calcidiol serum levels was detected in patients with SD and AV compared to the control group. Serum mean  $\pm$  standard deviation of 25(OH)D levels were significantly lower in SD patients than in AV (20.71  $\pm$  8.16 vs. 15.71  $\pm$  7.78, P = 0.005). The circulating 25(OH)D levels were significantly lower in patients with AV than in SD. Normal 25(OH)D levels was detected in 15% of SD patients and 10% of AV patients, vitamin D insufficiency - in 70% of SD and 55% AV patients, deficiency - in 15% of SD and 35% of AV patients. Insufficiency in 25(OH)D was detected in only 21.5% of the healthy controls. Vitamin D insufficiency prevailed in patients with with moderate and non-inflammatory SD, involvement scalp or face compared with clinical manifestations in patients with vitamin D deficiency (severe SD, inflammatory form, lesions of the scalp, face, trunk). The level of 25(OH)D was inversely associated with the severity of acne, and there was a significant negative correlation with inflammatory lesions.

LL-37 serum levels of patients with SD and AV decreased by 1.45-1.6 times compared to the control group. The level of LL-37 in patients with SD did not differ significantly from the value in patients with AV. The level of cathelicidin decreased in severe forms of the disease compared to that in healthy individuals and was not significantly different from the value in patients with mild and moderate severity of SD and AB.

Serum vitamin D was insufficient and deficient in 85% of cases SD and 90% of cases AV in comparison with 21.5% of controls. Noteworthy is the higher incidence of vitamin D deficiency in AV than in SD (15% and 35%, respectively). Serum vitamin D had an inverse correlation with the severity of acne. LL-37 levels was low in patients with moderate-to-severe

acne in compared with control.

Vitamin D may play an important role in the pathogenesis of SD and AV, since it has an immunomodulatory effect and regulates the synthesis of antimicrobial peptides.



## A Rare Case of a Hypopigmented Variant of Lichen Planoplaris

Tugce Banli\*<sup>1</sup>, Ayse Tuncer<sup>1</sup>, Deren Ozcan<sup>1</sup>

<sup>1</sup>Başkent University Ankara Hospital, Dermatology and venerology, Ankara, Türkiye

## Introduction

Lichen planopilaris (LPP) is an uncommon variant of Lichen Planus characterized clinically by perifollicular erythema, follicular hyperkeratosis, and irreversible hair loss, While several variants of LP exist, including the hyperpigmented type, there have been only one documented cases of hypopigmented LPP to our knowledge. Herein, we present a rare case of LPP manifesting as hypopigmented macules following a hair transplant procedure in the donor area.

## Case

A 38-year-old man presented to our clinic with hypopigmentation on the posterior aspect of his scalp. He reported no accompanying symptoms. The patient had realised the hypopigmentation 2.5 months after the hair transplant procedure. Upon dermatological examination, numerous hypopigmented macules were observed, predominantly located in the donor area on the posterior scalp, with a few additional lesions on the temporal regions. These lesions exhibited a blue-white appearance under Wood's lamp examination. Two 3 mm punch biopsies were performed, one from the hypopigmented occipital area and another from a non-hypopigmented temporal region. Histopathological analysis revealed follicular unit loss, tufted folliculitis, interface vacuolar reaction, perifollicular fibrosis, and sebaceous gland atrophy with a normal level of melanocytes. Based on these findings, the patient was diagnosed with hipopgmented variant of LPP. Treatment was initiated with one sesion of 40 mg methylprednisoloneans topical 0.05% clobetasol-17-propionate applied twice daily to the entire scalp.

## Discussion

Hypopigmented lichen planopilaris (LPP) is an exceedingly rare presentation, with only one documented case reported in literature to our knowledge. In our case, the presence of key LPP histopathological features, including sebaceous atrophy, perifollicular lymphocytic infiltration, perifollicular fibrosis, and tufted hair follicles, led to the diagnosis of LPP. Furthermore, the onset of LPP following a hair transplant procedure adds a notable dimension to our case. While there have been numerous reports of LPP post-hair transplant, often attributed to Koebnerization, a hypopigmented variant has not been previously documented. In summary, we advocate for the consideration of hypopigmented LPP in the evaluation of scalp hypopigmented macules, highlighting the need for broader recognition and understanding of this rare manifestation.



## Pityriasis rubra pilaris (PRP) following COVID-19 -vaccine

Svetlomira Varbanova<sup>1</sup>, Dimitrina Serafimova<sup>1</sup>, Lyubomir Dourmishev<sup>1</sup>, Snejina Vassileva<sup>1</sup>, Kossara Drenovska<sup>1</sup>

<sup>1</sup>Medical University of Sofia, Department of Dermatology and Venereology, Sofia, Bulgaria

**Introduction & Objectives:** Pityriasis Rubra Pilaris (PRP) is a rare inflammatory skin disorder characterized by erythematous scaly plaques, follicular hyperkeratosis, and palmoplantar keratoderma. While the etiology of PRP remains unclear, its occurrence following infections or vaccinations has been reported. During the COVID pandemic numerous post-vaccination skin reactions have been observed, however PRP or PRP-like reactions secondary to COVID vaccines are exceedingly rare with less than twenty cases reported in the literature. We present a case of PRP development shortly after COVID-19 vaccine application.

**Materials & Methods:** A 65-year-old woman presented with widespread red-orange follicular papules and plaques coalescing to an almost erythrodermic condition with small islands of sparing. Additionally, fine scaling on the scalp and face, palmo-plantar hyperkeratosis and nail changes including subungual hyperkeratosis, longitudinal ridging and yellow-brown discoloration were observed. These symptoms of approximately six months duration have appeared within a few weeks following her second dose of Pfizer-BioNTech COVID-19 vaccine. Previous interpretations of the diagnosis have included Sezary syndrome which was further rejected through detailed image and laboratory tests.

**Results:** Routine laboratory investigations were within normal ranges. Histologic examination showed alternating orthokeratosis and parakeratosis in both vertical and horizontal directions ("checkerboard" pattern). Based on the available clinico-laboratory findings the patient was diagnosed with type I, classic adult PRP according to Griffiths' classification. Phototherapy (NB-UVA) and topical emollients resulted in rapid and significant improvement of the diffuse erythema and desquamation of the skin.

**Discussion:** The reported case contributes to the limited body of evidence suggesting a potential link between vaccination and PRP development. The review of the literature found only 18 previously observed PRP or PRP-like reactions following the administration of all currently available COVID-19 vaccines, including Pfizer-BioNTech, Moderna and AstraZeneca. The temporal relation between COVID-19 vaccination and the onset of PRP raises questions about the potential role of immune activation in triggering or exacerbating this dermatologic condition. As vaccination campaigns continue globally, healthcare providers should be vigilant for detecting, reporting and managing unusual postvaccination dermatologic reactions, including PRP.



# Real-World Effectiveness of Upadacitinib on atopic dermatitis severity and hard-to-treat localizations: One-year results from the German observational study UP-TAINED

Stephan Weidinger<sup>1</sup>, Ansgar Weyergraf<sup>2</sup>, Fatima Abousamra<sup>3</sup>, Jenia Neumeister<sup>3</sup>, Kristina Lohmann<sup>3</sup>, Felix Jacobs<sup>4</sup>, Tobias Hagemann<sup>5</sup>, Felix Lauffer<sup>6</sup>

<sup>1</sup>Department of Dermatology and Allergy, University Hospital Schleswig-Holstein, Kiel, Germany,<sup>2</sup>Hautarztpraxis an der Hase, Department of Dermatology and Allergy, Bramsche, Germany, <sup>3</sup>AbbVie Deutschland GmbH & Co. KG, Dermatology, Wiesbaden, Germany, <sup>4</sup>DERMAKULM - Hautarztpraxis Dr. Jacobs & Kollegen, Department of Dermatology and Allergy, Kulmbach, Germany, <sup>5</sup>Hautarztpraxis Bad Sassendorf, Department of Dermatology and Allergy, Bad Sassendorf, Germany, <sup>6</sup>Technical University of Munich, Department of Dermatology and Allergy, München, Germany

## Introduction & Objectives:

Clinical trials demonstrated efficacy and safety of Upadacitinib for the treatment of atopic dermatitis (AD). However, real world (RW) data, especially on hard-to-treat localizations, are still sparse. These are the very first one-year RW results assessing the effectiveness and safety of Upadacitinib in moderate to severe AD patients.

**Objective 1:** to describe the effectiveness of Upadacitinib in Real-World (RW) regarding atopic dermatitis (AD) disease control and disease severity

Objective 2: to describe the RW effectiveness of Upadacitinib on facial and palmar AD lesions

Objective 3: to describe the RW safety of Upadacitinib for up to one year of follow-up

## Materials & Methods:

UP-TAINED is an ongoing German prospective, multicenter, observational study in adolescents and adults with moderate to severe AD (n=351) treated with Upadacitinib 15/30 mg once daily. Outcome measures included ADCT, EASI, DLQI, WP-NRS and adverse events (AEs).

## **Results:**

At week 12, 71% of the patients achieved disease control (ADCT total score <7), this rate was sustained for one year. Among patients with facial AD at baseline (n=260), 67.1% achieved clear or almost clear skin after 4 weeks of Upadacitinib treatment. Of the 216 patients with hand AD at baseline, 71.4% achieved clear or almost clear hands after 4 weeks. Regarding disease severity, 60.6% of the patients achieved an EASI score of  $\leq$ 3 points after 4 weeks, this rate was 68.1% after one year of follow-up. 423 AEs were reported in 163 patients: 59.7% classified as mild; 34.6% as moderate and 5.7% as severe. The most common AEs were worsening of AD, Acne, and COVID-19. No MACE, VTE, or malignancies were reported.

## **Conclusion:**

The results of this study demonstrate the early and sustained effectiveness of Upadacitinib in patients with moderate to severe AD. Especially patients with hard-to-treat localizations profit from Upadacitinib treatment. Moreover, Upadacitinib was well tolerated in clinical practice



## Linear IgA Dermatosis and tangled causes of anemia

Gabriela Turcu<sup>1</sup>, Roxana Ioana Nedelcu<sup>1</sup>, Andra Miu<sup>2</sup>, Brinzea Alice<sup>1</sup>, Andra Dinu<sup>2</sup>, Razvan Theodor Andrei<sup>2</sup>, Dan Mircioi<sup>2</sup>

<sup>1</sup>Carol Davila University of Medicine and Pharmacy, București, Romania, <sup>2</sup>Colentina Hospital, București, Romania

## Introduction & Objectives:

Linear IgA bullous dermatosis (LABD), also known as linear IgA dermatosis or linear IgA disease, is a relatively uncommon subepidermal vesiculobullous disorder that manifests in both adults and children. While children typically present with a distinct clinical pattern termed "chronic bullous disease of childhood," the underlying pathogenesis remains consistent across age groups. However, in adults, consideration of drug-induced causes is essential.

The clinical presentation of LABD in adults varies, posing challenges in differentiation from other autoimmune vesiculobullous conditions like bullous pemphigoid (BP). Symptoms range from scattered, tense bullae on unaffected skin to lesions with a more herpetiform appearance, often accompanied by erythema. These lesions commonly affect the trunk and extremities but can also involve other areas such as the scalp, genital region, or face. In children, a classic presentation includes annular erythematous lesions with vesicle rings, known colloquially as a "crown of jewels" or "string of pearls," typically found on the abdomen, lower back, thighs, groin, and periorbital and perioral regions. However, this hallmark presentation is less common in adults.

Diagnosis of LABD typically relies on detecting linear IgA deposits through direct immunofluorescence microscopy. Treatment options are primarily derived from case reports and series, with dapsone being suggested as a first-line therapy for both children and adults (Grade 2C), often yielding rapid response.

## Materials & Methods:

We present a case of a 45-year-old female with thalassemia minor who presented with pruritic tense bullae involving her face, dorsal upper and lower extremities, chest, and back over 30 days.

A thorough review of medications, including over-the-counter drugs, vitamins, and supplements, revealed no incriminating agent for the patient's clinical manifestations. Differentials considered included dermatitis herpetiformis, bullous pemphigoid, and erythema multiforme.

#### **Results:**

Two punch biopsies were conducted, one from an active lesion for routine hematoxylin and eosin staining and another from perilesional skin for direct immunofluorescence microscopy. Histology and direct immunoflorescence test confirmed the diagnosis of linear IgA bullous dermatosis.

Further blood tests, including a complete blood count, revealed microcytic anemia (initial levels showed a hemoglobin of 9.90 g/dL), a condition known to the patient since childhood due to thalassemia minor.

After confirming the diagnosis of linear IgA dermatosis and detecting normal levels of glucose 6-phosphate dehydrogenase, treatment with low-dose oral dapsone (25 mg/day) was initiated, resulting in rapid improvement of lesions. Subsequent blood tests showed hemoglobin levels of 8.60 g/dL, prompting reduction of dapsone dosage to 25 mg three times per week. After one month, both skin lesions and blood parameters showed improvement, with hemoglobin, hematocrit, and MCV returning to baseline levels.

#### **Conclusion:**

This case represents an atypical presentation of linear IgA disease, notable for the patient's underlying thalassemia minor which posed a unique challenge, with dapsone treatment resulting in significant anemia before reducing the dosage.



## Beyond Skin Deep: Contrasting Clinical Faces with Slightly Similar Pathological Aspects

Gabriela Turcu<sup>1</sup>, Roxana Ioana Nedelcu<sup>1</sup>, Brinzea Alice<sup>1</sup>, Andra Miu<sup>2</sup>, Andra Dinu<sup>2</sup>, Razvan Theodor Andrei<sup>2</sup>, Dan Mircioi<sup>1</sup>

<sup>1</sup>Carol Davila University of Medicine and Pharmacy, București, Romania, <sup>2</sup>Colentina Hospital, București, Romania

#### **Introduction & Objectives:**

Granulomatous dermatoses are a group of clinically and histologically heterogeneous inflammatory skin conditions including granuloma annulare (GA) and interstitial granulomatous dermatitis (IGD).

GA was described in association with diabetes mellitus, chronic infections, malignancies, and thyroid autoimmune diseases. Clinically, GA erupts usually with ring-like erythematous plaques, scattered coalescing papules, or nodules. While the localized GA classically appears on the hands and feet, the generalized form involves extremities and trunk, and the subcutaneous form usually involves the lower extremities in children. On histology, mucin with a palisading or interstitial configuration of granulomatous inflammation represents the typical finding. The palisading pattern appears as dermal palisading histiocytes and lymphocytes surrounding a central zone of necrobiotic collagen in upper and middle dermis.

IGD is described in association with autoimmune diseases like arthritis, systemic lupus erythematous, and autoimmune hepatitis. Clinically, it presents with asymptomatic papules and plaques, varying in colour from skin-coloured to erythematous and frequently located in lateral upper trunk, axillae, and proximal upper limbs. Linear, cord-like nodular lesion called "the rope sign" is considered a characteristic manifestation of IGD, despite the low frequency. Histology shows scattered dermal histiocytes among and around foci of degenerated collagen, often rimming altered collagen fibers which may be "floating" ("floating sign"), while usually no vasculitis or mucin are detected.

#### Materials & Methods:

We present two cases:

Case 1:

A 56-year-old woman with a history of Diabetes Mellitus type 2 presented with a generalized eruption. This eruption consisted of 1-5 cm firm, smooth, shiny ring-like erythematous plaques and papules with central depression. The lesions first appeared 4 years ago on her hands' backs. Over the past month, similar lesions began to emerge on her collarbone area and anterior and posterior trunk.

#### Case 2:

A 68-year-old woman with a history of Rheumatoid Arthritis, treated with sulfasalazine, methotrexate, and corticosteroids since 2015, presented with an eruption. This eruption consisted of multiple skin-colored, elastic, well-defined, round-shaped, asymptomatic soft papule, suggestive for focal areas of flaccid skin. These papules were symmetrically distributed on her neck, arms, trunk, and inner thighs, with an onset one year prior.

While in the first case the clinical aspect was suggestive for a granulomatous dermatoses, in the second one, we considered mainly elastic tissue disorders, including papular elastorrhexis, mid-dermal elastolysis and anetoderma.

#### **Results:**

A punch biopsy from the periphery of a ring-like plaque confirmed Granuloma Annulare in the first case, while in the second case, the punch biopsy of a papule confirmed Interstitial Granulomatous Dermatitis.

# **Conclusion:**

This presentation underscores the complexity that can arise when comparing diseases from both histological and clinical perspectives. While the histopathological features of Granuloma Annulare and Interstitial Granulomatous Dermatitis may bear some resemblance, their clinical manifestations diverge substantially, leading to distinct diagnostic and treatment considerations. Also, it is intriguing to note that this manifestation of IGD is rarely documented in the medical literature.



## An idiopathic case of subcutaneous granuloma annulare

Georgiana Roxana Zaharia<sup>1</sup>, Ana Liliana Butnarus<sup>1</sup>, Diana Savu<sup>1</sup>, Renata Maria Pitulea<sup>2</sup>, Viorel Trifu<sup>1</sup>

<sup>1</sup>Central Military Emergency Hospital "Dr. Carol Davila", Dermatovenereology, Bucharest, Romania,<sup>2</sup>Clinica Domina Sana, Anatomopathology, Bucharest, Romania

## Introduction & Objectives:

Granuloma annulare is an inflammatory granulomatous skin disorder predominantly affecting children and young adults. Its pathophysiology remains poorly elucidated and its etiology remains still unknown. Clinical variants of granuloma annulare include the following subtypes: localized, generalized, perforating, patch and subcutaneous. Subcutaneous granuloma annulare is defined by hard to firm nodules that are often asymptomatic. Given the diverse documented predisposing factors and associated systemic conditions linked to this dermatologic entity, the aim of this case report is to demonstrate that this connection is not mandatory.

## Materials & Methods:

Case report.

## **Results:**

A 39-year-old male presented to our dermatology clinic with an asymptomatic erythematous annular plaque located on the right upper arm, evolving over several months. The patient was not taking any medication at the time of the presentation and no history of trauma at the lesion site was reported. No endocrinologic abnormalities, diabetes, dyslipidemia, or neoplasias were found. To establish a definitive diagnosis and exclude alternative pathologies, a skin biopsy was performed, revealing numerous histiocytes arranged among the collagen fibers in the deep reticular dermis and an accumulation of acidic mucopolysaccharides evidenced by PAS staining. The histopathological findings confirmed the diagnosis of subcutaneous granuloma annulare.

## **Conclusion:**

Subcutaneous granuloma annulare is a complex skin condition with a broad range of differential diagnoses. Typically, granuloma annulare manifests as a nodular lesion, but other presentations may also occur. Even if it appears often as a manifestation of a trigger factor or an underlying disease, this skin disorder can also appear by itself in healthy patients. Further research into its pathophysiology and the potential correlation between lesion onset and subsequent disease development is needed.



## facial solar porokeratosis: a newly discovered variant of porokeratosis

Zineb Mernissi<sup>1</sup>, Saloua Hazmiri<sup>2</sup>, Aboudourib Meriam<sup>3</sup>, Ouafa Hocar<sup>3</sup>, Amal Said<sup>3</sup>

<sup>1</sup>Mohammed VI University Hospital of Marrakech, Department of dermatology venerology a, Marrakech, Morocco, <sup>2</sup>Mohammed VI University Hospital of Marrakech, department of dermatology venerology , Marrakech, Morocco, <sup>3</sup>Mohammed VI University Hospital of Marrakech, Department of dermatology venerology , Marrakech, Morocco

## Facial solar porokeratosis: a newly discovered variant of porokeratosis

## Introduction & Objectives:

Porokeratosis are a set of acquired or hereditary keratinization disorders. They are defined clinically by annular lesions surrounded by a peripheral border and histologically by a parakeratotic column known as cornoid lamella. There are several variants of porokeratosis such as Mibelli porokeratosis and superficially disseminated actinic porokeratosis. A variant manifesting only on the face has been discovered recently and referred to as facial solar porokeratosis. We present our observation of this variant on a case involving two sisters.

## Materials & Methods: Case report

#### **Results:**

Two sisters aged 28 and 30, with no particular pathological history,were admitted for similar hyperkeratotic annular facial lesions that had been developing for 10 years. These lesions were small and gradually increased in size and flattening of the central region. The lesions came with mild itching sensations that worsened with sun exposure. No medication, irradiation or topical application was previously used on the regions. Skin examination showed small rounded lesions measuring 4 to 15 mm with an atrophic and pigmented center surrounded by a palpable and slightly visible thin peripheral border located on the nose and the paranasal region, along with retentional lesions and acne scars. Dermoscopic examination showed a brownish-yellow cornoid lamella surrounding a flat center. Both patients refused to do a skin biopsy. The diagnosis of solar facial porokeratosis was deduced based on the presence of these clinical and dermoscopic signs characteristizing the disorder.

#### **Discussion:**

Porokeratosis is an epidermal disorder caused by the expansion of abnormal epidermal keratinocytes. The two most common types, known as Mibelli porokeratosis and disseminated superficial actinic porokeratosis, are inherited as autosomal dominant traits, and are most often found on the chest and extremities, with rare manifestations on the face. Recently, rare variant known as solar facial porokeratosis has been discovered, characterized by distinct clinical and histopathological attributes.

Facial solar porokeratosis is characterized by single or multiple annular skin-colored lesions measuring between 0.1cm to a few centimeters, surrounded by a keratotic rim appearing most often on the nasal and paranasal regions. Dermoscopic examinations show the yellowish cornoid lamella surrounding a pink-white scarred area at the center. Histopathology of the hyperkeratotic area shows a cornoid lamella specific to porokeratosis.

No specific treatment has been approved for facial solar porokeratosis. Lesions may respond to topical steroids, calcineurin inhibitors, vitamin D3 analogues, cryotherapy, imiquimod or topical 5-fluorouracil, combined with routine photoprotection.

#### **Conclusion:**

Facial solar porokeratosis is a newly discovered type of porokeratosis, defined by distinguishable clinical and histopathological characteristics.



#### fox fordyce disease

Hyba Taounza<sup>1</sup>, Yosra Ben Kraiem<sup>1</sup>, Meziane Meriem<sup>1</sup>, Nadia Ismaili<sup>1</sup>, Benzekri Laila<sup>1</sup>, Senouci Karima<sup>1</sup>, Hamada Syrine<sup>1</sup>

<sup>1</sup>Centre hospitalier Ibn Sina, Dermatologie, Rabat, Morocco

#### Introduction & Objectives:

Fox-Fordyce disease (FFD) is a rare inflammatory disorder of the apocrine sweat glands, little known by physicians and often diagnosed as eczema.

#### **Case report:**

We report a case of a 45-years-old woman who presented with a history of severely itchy persistent lesions of the arm pit and on the breasts for almost 10 years; treated in the past as an eczema with topical steroids with a partial improvement and a relapse on discontinuation of the treatment. we found a skin-colored papules, 2-3 mm sized, firm, dome-shaped, equidistant distribution over the axillae and areolae. She also had decreased hair growth over the axillae region. The surrounding skin was normal. A clinical diagnosis of Fox-Fordyce disease was made, and she was given topical steroids to be applied once daily along with tretinoin cream twice a day, with which, within one month she had remarkable improvement.

#### **Discussion:**

Originally described by George Henry Fox and John Addison Fordyce in 1902, Fox-Fordyce disease (FFD) is a rare inflammatory disorder of the apocrine sweat glands, regarded widely to be "apocrine miliaria", an analogue of eccrine miliaria.

Lesions are classically monomorphic skin colored very itchy papules in the axillary vault smooth, uniform, equidistant, firm, folliculocentric often associated with hair loss.

The pathophysiology consists of the obstruction of the apocrine gland duct due to the insertion of a keratin plug in the hair follicle wall.

The underlying aetiology of FFD remains unclear, although a history of trauma caused by laser hair removal or hormonal factors may be considered as triggers.

In our patient, no evident causes were found.

Most commonly, FFD affects the axillae followed by the anogenital and periareolar areas. However, it also has been reported to involve the lips, umbilicus, sternum, perineum, and upper thighs. Thus, our patient never had a history of lesions in the cited areas.

Histopathologically, an "intraepidermal sweat retention vesicle" is considered the only feature diagnostic of the condition. We didn't take any biopsy sample since the diagnosis is mainly clinic.

The treatment of FFD remains difficult and consist of topical and intralesional glucocorticoids as first-line therapy. Also, topical retinoids for hyperkeratosis have been used with success.

#### **Conclusion:**

FFD is a rare inflammatory dermatosis. Further research is needed to understand its pathophysiology, and hence its

treatment.



#### pustular psoriasis - paradoxical reaction in a bioexperimented patient

Filofteia-Adelina Ghilencea\*<sup>1</sup>, Diana-Andreea Marcu<sup>1</sup>, Marcela Poenaru<sup>1</sup>, Viorel Trifu<sup>1</sup>

<sup>1</sup>Spitalul Universitar de Urgență Militar Central "Dr. Carol Davila", București, Romania

#### Pustular psoriasis - paradoxical reaction in a bioexperimented patient

#### **Introduction & Objectives:**

Psoriasis is a systemic, inflammatory, chronic, multifactorial condition, determined by immunological, genetic and environmental factors. Palmoplantar pustular psoriasis is a localized form of pustular psoriasis, characterized by the presence of sterile pustules arranged on an erythematous, pruritic background, being distributed on the level of the thenar and hypothenar eminences, on the medial part of the plant and in the calcaneus region. Etiological factors involved in the occurrence of pustular psoriasis are smoking, genetic factors, stress, infections and anti-TNF  $\alpha$  inhibitors.

#### Materials & Methods:

We will present the case of a 52-year-old female smoker, from an urban environment, who presented to the clinic for an erythematous plaque, intensely pruritic, with pustules on the surface, well delimited, located on the plantar aspect of the right foot, in evolution for six months. The lesions progressed slowly and had a significant negative impact on the quality of life. From the medical history, I have noted that she is a bioexperimented patient, who in 2012 was diagnosed with Ankylosing Spondylitis, in current treatment with Golimumab (50 mg/month), and since 2019 she also associates with Crohn's Disease, in treatment with Mesalazine (3g/day). The clinical features began 1 and a half years after the administration of the TNF  $\alpha$  inhibitor. The patient had no personal or family history of psoriasis. We performed an incisional biopsy with histopathological examination, with the suspicion of paradoxical pustular psoriasis, because there were reports in the specialized literature of similar adverse reactions to anti-TNF  $\alpha$  agents. Due to the seriousness of the associated pathologies, we did not suspend the medication with anti-TNF  $\alpha$  agents and we added Methotrexate (15 mg sc/week), with a slowly favorable evolution.

#### **Results:**

The histopathological examination revealed parakeratosis, hypogranulosis, Kogoj-Lapiere spongiform pustules, psoriasiform epidermal hyperplasia and mixed perivascular and diffuse infiltrate in the dermis, suggestive of pustular psoriasis.

# **Conclusion:**

The presented case adds to the growing number of patients who have developed adverse reactions during the treatment with TNF  $\alpha$  inhibitors, and understanding the mechanism of action should contribute to improving the treatment of cases.



## Navigating the Dermatological Maze: A Comprehensive Case of PASH Syndrome with Multifactorial Challenges

Anastasia Gheorghiev<sup>\*1, 2</sup>, Tanya Gancheva<sup>1, 2</sup>, Karen Manuelyan<sup>1, 2</sup>, Rosita Lavcheva<sup>1, 2</sup>, Evgenyia Hristakieva<sup>1, 2</sup>

<sup>1</sup>University Hospital "Prof. Dr. Stoyan Kirkovich", Stara Zagora, Bulgaria, Clinic of Dermatology and Venereology, Stara Zagora, Bulgaria, <sup>2</sup>Trakia University, Medical Faculty, Stara Zagora, Bulgaria, Department of Dermatology and Venereology, Stara Zagora, Bulgaria

**Introduction & Objectives:** PASH syndrome, encompassing Pyoderma Gangrenosum, Acne, and Hidradenitis Suppurativa, presents a complex dermatological triad that requires comprehensive understanding and management. This syndrome is particularly challenging from a therapeutic point of view emphasising the need for an early and correct diagnosis. It also prompts a closer examination of the connections between these dermatological and systemic disorders, aiming to enhance our understanding and optimize the care provided to individuals with PASH syndrome and associated comorbidities.

**Materials & Methods:** We present the case of a 44-year-old male patient diagnosed with PASH syndrome alongside a constellation of comorbidities that profoundly affected the choice of treatment strategy and required multidisciplinary diagnostic and therapeutic efforts. The patient's skin complaints started 16 years ago with pus-filled nodules in the inguinal folds, extending subsequently to the axillae, back, and gluteal area. He also developed painful ulcers on the lower legs suggestive of pyoderma gangrenosum. He was first admitted to our dermatology clinic in 2020 after living abroad for several years. His comorbidities included rheumatoid arthritis, essential hypertension, stable angina, dyslipidemia and chronic hepatitis. A number of clinical and paraclinical investigations were performed: complete blood count and biochemical panel, Quantiferon test, T-SPOT TB, serology for HCV, HCV-RNA-PCR test, chest radiography, abdominal ultrasound, colonoscopy and microbiologic tests. Following biological treatment for rheumatoid arthritis he developed latent tuberculosis which was treated with isoniazid. He also underwent knee surgery, hip surgery and colon polypectomy.

**Results:** The patient was treated with multiple courses of antibiotics (metronidazole, meropenem, ertapenem, amikacin). Currently, the patient is on biological treatment with TNF-alpha inhibitor (adalimumab), zinc gluconate, pentoxifylline and silymarin. The maintenance treatment for rheumatoid arthritis is methylprednisolone 8 mg daily. Topical skincare includes chlorhexidine digluconate hygienic wash, polyhexanide and betaine surfactant wound cleansing solution. The therapy effectively reduced the symptoms and signs of HS, PG and rheumatoid arthritis and improved the patient's quality of life.

**Conclusion:** This case highlights the intricate interplay of multiple dermatological and systemic disorders, requiring a holistic and multidisciplinary approach for effective diagnosis and management. The complexity of PASH syndrome is further underscored by the presence of coexisting medical conditions, emphasizing the importance of tailored therapeutic strategies to address the unique needs of this patient population.



## **Topical Management of Pyoderma Gangrenosum: Current Insights and Future Prospects**

Dela Shojaei<sup>1</sup>, Haleh Zabihi<sup>2</sup>, Vincent Piguet<sup>3</sup>, David Croitoru<sup>3</sup>

<sup>1</sup>UBC Faculty of Medicine, Vancouver, Canada,<sup>2</sup>University of Toronto Temerty Faculty of Medicine, Toronto, Canada, <sup>3</sup>Women's College Hospital, Dermatology, Toronto, Canada

## Introduction & Objectives:

Pyoderma Gangrenosum (PG) is a rare, chronic inflammatory dermatosis with neutrophil-dominant dermal infiltration leading to necrosis and ulceration. PG can occur at any cutaneous site with rapid onset and varied morphologic presentations. The management of PG is challenging as there are no FDA-approved treatments. The highest degree of evidence is for systemic corticosteroids, cyclosporine, and calcineurin inhibitors. The use of topical agents however has been less comprehensively evaluated; for mild and well-controlled cases of PG these agents have high utility. We present a comprehensive narrative review of the literature with attention to current and future prospects for topical treatment of PG.

## Materials & Methods:

An initial literature search in the electronic database Ovid MEDLINE was conducted up to 21 November 2023 using the search terms "Pyoderma Gangrenosum" and "pyoderma gangraenosum" in combination with "topical\* or local or transdermal or dermal or cream or ointment or gel or lotion or solution or patch or spray", "roflumilast", "ruxolitinib" "corticosteroid", "Tacrolimus \*", "Cyclosporine", "Pimecrolimus", or "Wound care". Among 936 titles and abstracts screened, 44 were included in the current review. We excluded articles not accessible as full texts and articles not written in English.

#### **Results:**

Key topical treatments for PG include topical wound care, corticosteroids, and calcineurin inhibitors. Level 3A evidence emphasizes the importance of topical interventions for ulcer healing, and the efficacy of ozonated water and 5% potassium permanganate solution are supported by case reports. High-potency corticosteroids, exemplified by clobetasol propionate, are commonly used for mild PG, with up to 43.8% achieving complete healing at 6 months. Tacrolimus, particularly at higher concentrations, effectively reduces wound size. Novel topical therapies include roflumilast, a 2022approved cream with 1b-level evidence, and ruxolitinib, a JAK2 inhibitor, showing success in a polycythemia vera patient and resulting in significant PG lesion healing and pain relief within 10 weeks

#### **Conclusion:**

This comprehensive review provides insights into current and future directions for topical treatments in the challenging landscape of PG management. Key topical treatments, including wound care, corticosteroids, and calcineurin inhibitors, show promise, emphasizing the importance of topical interventions for ulcer healing. Additionally, novel therapies such as roflumilast and ruxolitinib highlight emerging prospects for PG management, offering significant healing and pain relief within a relatively short timeframe.