

**Abstract N°: 8****The Prognostic Role of Reactive Dermatoses in Anti-Interferon- $\gamma$  Autoantibody Patients: A Retrospective Cohort Study**

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**Introduction & Objectives:**

Reactive dermatoses (RD), including neutrophilic dermatoses (ND) and non-ND, are common in adult-onset immunodeficiency (AOID) associated with anti-interferon-gamma (IFN- $\gamma$ ) autoantibodies. Their prognostic significance remains unclear.

To evaluate the prognostic role of RD in AOID patients with anti-IFN- $\gamma$  autoantibodies.

**Materials & Methods:**

A retrospective cohort study was conducted on AOID patients with detectable anti-IFN- $\gamma$  autoantibodies diagnosed at Ramathibodi Hospital, Bangkok, from 2011 to 2024. Clinical and laboratory data were analyzed, comparing active versus stable episodes with and without RD. RD was classified as ND [e.g., Sweet's syndrome (SS), pustular eruptions] or non-ND (e.g., erythema nodosum, vasculitis).

**Results:**

A total of 53 patients with AOID due to anti-IFN- $\gamma$  autoantibodies were identified, with a mean age of onset of  $53.55 \pm 9.69$  years; 62.26% were female. The most commonly involved organs were lymph nodes (94.34%) and skin (83.02%). Disseminated opportunistic infections occurred in 67.92% during active episodes, predominantly Mycobacterium abscessus (59.62%) and Salmonella spp. (36.54%). Higher WBC counts ( $p = 0.002$ ), anti-IFN- $\gamma$  autoantibody levels ( $p = 0.003$ ), and lower hematocrit ( $p = 0.018$ ) were observed in RD-associated active episodes. Cox regression analysis, adjusted for immunosuppressant use, revealed a higher risk of relapse in ND compared to non-ND (HR 6.9, 95% CI: 1.35–35.32,  $p = 0.02$ ) and in SS compared to other RD (HR 3.58, 95% CI: 1.26–10.18,  $p = 0.017$ ).

**Conclusion:**

ND, especially Sweet's syndrome, predicts relapse in AOID. WBC, anti-IFN- $\gamma$  autoantibody levels, and hematocrit are critical biomarkers for monitoring disease activity.

**Table 1. Hazard ratio for time to relapse during stable stages in patients with AOID due to anti-IFN- $\gamma$  autoantibodies.**

Covariates	Hazard ratio (95% CI)	p-value
<b>Overall patients</b>		
Reactive dermatoses	1.19 (0.71-1.98)	0.505
<b>Patients with RD</b>		
Sweet's syndrome	3.58 (1.26-10.18)	0.017*
Neutrophilic dermatoses	6.90 (1.35-35.32)	0.020*

\*A p-value < 0.05 indicates statistical significance.

Cox regression analysis adjusted for the use of immunosuppressant drugs.

Abbreviations: AOID, adult-onset immunodeficiency; anti-IFN- $\gamma$ , anti-interferon - $\gamma$ ; CI, confidence interval; RD, reactive dermatoses.

**EADV Symposium 2025 – PRAGUE**  
**22 MAY - 24 MAY 2025**  
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**Abstract N°: 164****Hypocomplementemic Urticarial Vasculitis: A Rare Manifestation of SLE**Shakira Bou\*<sup>1</sup>, Jose Gonzalez Chavez<sup>2</sup><sup>1</sup>San Juan Bautista School of Medicine, Caguas, Puerto Rico<sup>2</sup>Oficina Dr José González Chávez Dermatólogo, San Juan, Puerto Rico**Introduction & Objectives:**

Urticarial vasculitis (UV) is a rare type of vasculitis that involves the small blood vessels. It is classified as a type III hypersensitivity reaction that presents with the formation of immune complexes and, consequently, the activation of the complement via the classical pathway, leading to an inflammatory cascade. UV is characterized by hives or urticarial lesions that last more than 24 hours and often requires skin biopsy to diagnose. Although the majority of UV cases are idiopathic, some have been associated with other autoimmune disorders such as systemic lupus erythematosus (SLE) and other inflammatory conditions. UV is classified according to the complement protein levels on serum: normocomplementemic UV (NUV) and hypocomplementemic UV (HUV); the latter being more severe. This report highlights a rare type of small-vessel vasculitis; HUVS. By presenting this case, we aim to raise awareness and provide guidance for diagnosing and managing this rare manifestation of lupus, which may be misdiagnosed and progress to systemic organ involvement.

**Materials & Methods:**

A 36-year-old woman presented with a 20-day history of pruritic, warm urticarial lesions with arthralgia. Her medical history included intermittent fevers and arthralgia in 2023, with prior negative laboratory results. In 2024, similar symptoms recurred, and testing revealed positive antinuclear antibodies along with a diagnosis of systemic lupus erythematosus. Further tests showed positive anti-RNP antibodies, low complement proteins (C3 and C4), and elevated serum gamma globulins.

**Results:**

Despite treatment with prednisone (40 mg/day), her urticarial lesions persisted, prompting a referral to dermatology. Physical examination revealed prominent macular rash on face and widespread urticarial eruptions on arms, trunk, legs and face. Punch skin biopsy of her right arm confirmed urticarial vasculitis. She was initiated on indomethacin 75 mg BID, resulting in complete lesion resolution within 48 hours. The patient was diagnosed with hypocomplementemic urticarial vasculitis, a severe form of vasculitis. For long term management, she was placed on low-dose prednisone, indomethacin, and hydroxychloroquine.

**Conclusion:**

Hypocomplementemic urticarial vasculitis is noted as the more severe form of UV, often involving systemic organ complications, such as glomerulonephritis and pulmonary disease. Histologically, it is characterized by a mixed cellular infiltrate with eosinophils, neutrophils and leukocytoclasia. There can also be fibrin deposits in the vascular channels. This case underscores the importance of thorough evaluation and a multidisciplinary approach in patients presenting with refractory urticarial lesions and systemic symptoms to ensure timely diagnosis and optimal management. Further research is essential to better understand this complex condition and improve patient outcomes.



**Abstract N°: 188****From plaques to pustules: a case report on the challenges of pustular psoriasis of pregnancy**Roisin Rynne<sup>\*1</sup>, Roisin Hambly<sup>1</sup>, Cliona Feighery<sup>1</sup><sup>1</sup>Our Lady of Lourdes Hospital, Drogheda, Drogheda, Ireland**Introduction & Objectives:**

Pustular Psoriasis of pregnancy is a rare, potentially life threatening, variant of generalised pustular psoriasis that primarily occurs in the third trimester or postpartum period. It can be complicated by both erythroderma and placental insufficiency. Recognition and management are crucial due to the increased risk of maternal-foetal morbidity and mortality.

**Materials & Methods:**

Single case report, chart reviewed retrospectively, and key learning points identified.

**Results:**

A 37-year-old, gravida 8 para 4+3, at 37 weeks' gestation, was admitted from the antenatal clinic due to a rapid onset pustular eruption on a background of well controlled plaque psoriasis. On examination, she had polycyclic erythematous patches with extensive peripheral non-follicular pustules. This evolved to cover >90% of her body surface area. Her face, palm, plantar and interestingly, intertriginous areas were spared. She remained constitutionally well despite raised inflammatory markers. Initial topical treatment failed and her condition deteriorated. Oral prednisolone was commenced. Due to cardiotocography changes on day 6, she had an emergency caesarean section where meconium stained amniotic fluid was noted. A healthy baby girl was delivered. The patient continued prednisolone postpartum without a significant improvement. Adalimumab was commenced 10 days postpartum whilst initial response was positive, improvement plateaued following discontinuation of oral steroids. She was subsequently started on Ixekizumab with an excellent and sustained response.

**Conclusion:**

Evidence on the treatment of pustular psoriasis of pregnancy is limited. Its management is further complicated by the lack of robust data on the risk to the mother and foetus. This uncertainty compounds the complexity of counselling both the patient and her obstetrician on the necessity of enhanced monitoring and the potential for early delivery. Multidisciplinary care is crucial to optimise outcomes.





**Abstract N°: 222****Bullous and Ulcerative Pyoderma Gangrenosum, Peripheral Ulcerative Keratitis, and Oral Ulcers in the Setting of Inflammatory Bowel Disease**

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**Introduction & Objectives:**

Pyoderma gangrenosum (PG) is a rare, inflammatory, neutrophilic dermatosis presenting with extremely painful and rapidly enlarging ulcers. Peripheral ulcerative keratitis (PUK) is a rare ocular condition characterized by the destruction of the peripheral cornea. Ocular and oral manifestations in association with PG are very rare, with only a few cases reported in literature. We describe a case of severe, bullous and ulcerative, PG associated with PUK and oral ulcers in a 49-year-old woman with IBD.

**Materials & Methods:**

A 49-year-old female with a 2-month history of gradually progressing, necrotic and hemorrhagic ulcers with violaceous, bullous borders on the trunk and extremities, associated with left conjunctival hyperemia, pain, discharge, photophobia, and oral ulcers presented at the emergency department.. Ophthalmologic examination revealed a 1.5 x 10 mm crescent-shaped, epithelial-stromal defect from 3 to 8 o'clock at the inferior perilimbal area. Fecal calprotectin was positive and colonic biopsies showed chronic nonspecific inflammation.

**Results:**

The patient was diagnosed as a case of bullous and ulcerative PG with ocular (PUK) and oral involvement secondary to IBD. Initial treatment consisted of topical and systemic corticosteroids. Due to further enlargement of cutaneous ulcers, infliximab (5 mg/kg) was eventually administered, halting ulcer progression and affording relief of mucosal ulcers.

**Conclusion:**

We report this case to highlight the severe manifestations of PG that can occur due to delayed diagnosis and debridement. We also raise awareness of the rare, but possible, mucosal manifestations of PG occurring in the setting of IBD. Early recognition and treatment of PG are crucial in minimizing the high morbidity associated with this condition.





**Abstract N°: 310**

**Eosinophilic dermatosis of haematological malignancy: unmasking a small lymphocyte lymphoma**

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**Introduction & Objectives:**

Eosinophilic dermatosis of haematological malignancy (EDHM), previously known as insect bite-like reaction, is a rare entity that denotes the appearance of clinically diverse lesions, histopathologically characterized by eosinophilia, in patients with underlying haematological malignancy. Hence, differential diagnoses encompass a broad spectrum, especially when EDHM is the first manifestation of a yet undiagnosed haematological malignancy.

**Materials & Methods:**

We describe a 69-year-old male patient presenting with a four-year history of disseminated pruritic erythematous lesions. The lesions typically appeared in the summer months and tended to subside in the winter. He also confirmed the appearance of night sweats but denied significant weight loss and other systemic symptoms. His medical history was relevant for well-managed diabetes mellitus type 2 and arterial hypertension.

**Results:**

Clinical examination revealed papules and papulovesicles with excoriations and multiple cicatrices affecting the head and neck region, the trunk and the extremities. Palpable, painless, enlarged axillary and inguinal lymph nodes were also noted. A complete blood count showed elevated absolute (up to  $0.9 \times 10^9/\text{ml}$ ) and relative (up to 11%) eosinophil levels, with normal lymphocyte counts. Immunological analyses revealed borderline positive ELISA indices of bullous pemphigoid 230 (1.18 IU/ml) and 180 (1.06 IU/ml) antigens, as well as raised anti-gliadin IgA antibodies (28.9 U/ml) and immunoglobulin E levels (7,660 U/ml). The  $\beta 2$  microglobulin level was 5.1 mg/l, while lactate dehydrogenase values remained within normal limits. Lesional biopsies showed eosinophilic spongiosis with vesiculation, interstitial perivascular eosinophils and lymphocytes, and incipient subepidermal bulla formation. Direct immunofluorescence assays of perilesional biopsies were negative on three separate occasions. A massive retroperitoneal and mesenteric lymphadenopathy, accompanied by enlarged lymph nodes in the soft tissues, were visualized on ultrasonography and confirmed by computerized tomography imaging. Histological and immunohistochemical analysis of the largest axillary lymph node biopsy established the diagnosis of small lymphocyte lymphoma/chronic lymphocytic leukaemia. The patient was referred to a haematology specialist for further investigations and treatment, which is currently underway.

**Conclusion:**

Eosinophilic dermatosis of haematological malignancy is thought to be a manifestation of a paraneoplastic immune dysregulation, leading to an interleukin 5-driven recruitment of eosinophils into the skin. This imbalance may also trigger the secretion of autoantibodies, causing EDHM to mimic autoimmune bullous dermatoses such as bullous pemphigoid and dermatitis herpetiformis, as witnessed in the index case.

The various clinical presentations of EDHM, including pruritic papules, nodules, vesicles or bullae, further complicate the differential diagnosis. Thus, the appearance of pruritic insect bite-like lesions, upon exclusion of primary dermatological disorders and other causes of tissue eosinophilia, warrants further haematological investigation, especially when systemic B symptoms are present.



**Abstract N°: 425****An unusual presentation of pancreatic panniculitis**

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**Introduction & Objectives:**

Pancreatic panniculitis is a rare skin manifestation, affecting 2-3% of patients with pancreatic disease. This eruption is associated with a range of pancreatic pathologies including acute pancreatitis, pancreatic malignancies, pancreatic pseudocysts and pancreatic trauma. The pathophysiology of this disease process is not yet fully understood, however it is thought that the high efflux of pancreatic enzymes causes extra-abdominal fat necrosis including the subcutaneous tissues. Controlling the underlying pancreatic pathology is the mainstay of treatment.

**Materials & Methods:**

A 59-year-old Caucasian woman was referred to the dermatology department with a 3-day history of a progressive, tender, multiple erythematous nodules over both shins and her right forearm. The patient had a past medical history of polycystic kidney disease, orthotopic liver transplant in 2019, now complicated by cirrhosis, type 2 diabetes mellitus and hypertension. The rash started one day after undergoing an elective endoscopic retrograde cholangiopancreatography (ERCP) for a developing anastomotic common bile duct stricture. The patient denied any history of alcohol misuse. The ERCP was complicated by a rising amylase count, reaching a level of 5733 U/L [range 28-100U/L] 24 hours post-procedure. The patient denied any abdominal pain or other gastrointestinal symptoms.

A skin biopsy from the patient's right forearm lesion showed florid active lobular panniculitis with numerous ghost anucleate adipocytes, surrounded by a thin rim of lacy calcific deposition. A dense neutrophilic inflammatory infiltrate was also evident. The patient's history, high serum amylase levels and histology were in keeping with a diagnosis of pancreatic panniculitis.

**Results:**

The eruption was noted to resolve over one week without any treatment, along with the improvement of the patient's amylase reaching a level of 176 U/L.

**Conclusion:**

In conclusion, we present an unusual case of pancreatic panniculitis in a patient with a complex medical history. The patient had no systemic symptoms to suggest an underlying acute pancreatitis and the eruption was deemed to be secondary to her recent ERCP. The exact mechanism by which the patient remained asymptomatic despite such a high amylase count is unknown but this could possibly be secondary to impaired amylase metabolism. Amylase is metabolised by the liver and excreted by the kidneys, our patient was a known case of chronic kidney disease due to her polycystic kidneys and her liver transplant was complicated by cirrhosis.



**Abstract N°: 451****VEXAS syndrome with morbilliform eruption as presenting sign**

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**Introduction & Objectives:**

VEXAS (vacuoles, E1 enzyme, x-linked, autoinflammatory, somatic) syndrome is a recently described systemic autoinflammatory disease with overlapping dermatologic, rheumatologic, pulmonary, and hematologic manifestations, most commonly affecting elderly males. Cutaneous manifestations are frequently the initial presenting sign of VEXAS. Previously described cutaneous manifestations include neutrophilic dermatosis, vasculitis, periorbital edema, and chondritis.

**Materials & Methods:**

We describe a case of VEXAS syndrome with purpuric lesions and morbilliform eruption as presenting manifestations.

**Results:**

A 74-year-old Caucasian man with a past medical history of childhood asthma, hypertension, and steroid-responsive interstitial pneumonitis was examined in the in-patient setting. He was hospitalized for pneumonia and was found to have purpuric lesions and chronic subarachnoid hemorrhage in the setting of suspected amyloid angiopathy. Punch biopsy of purpuric lesions showed mixed inflammatory infiltrate with focally abundant neutrophils, vascular disruption, scattered lymphocytes, eosinophils, and dermal edema, consistent with leukocytoclastic vasculitis. The next month, he was hospitalized for cephalic vein thrombosis and possible steroid psychosis. Bone marrow biopsy revealed findings consistent with myelodysplastic syndrome in absence of supportive genetic findings. He later presented with a diffuse morbilliform eruption. Workup at this time revealed anemia, thrombocytopenia, and elevated inflammatory markers including C-reactive protein, erythrocyte sedimentation rate, ferritin, and IL-2R. Serum protein electrophoresis showed a faint IgG kappa band. Punch biopsy of the morbilliform lesions revealed neutrophil-rich superficial and mid-dermal inflammatory cells infiltrate with rare eosinophils. Bone marrow testing for UBA1 mutation was positive for p.M41V mutation. He was started on a tapering dose of prednisone, colchicine, and tocilizumab with complete control of his skin and systemic symptoms. A diagnosis of VEXAS syndrome was made.

**Conclusion:**

This case underscores the critical role of dermatologists in recognizing cutaneous signs of this rare autoinflammatory syndrome and ordering genetic testing in the appropriate clinical scenario. While neutrophilic dermatoses and vasculitis are commonly reported, morbilliform eruption, as encountered in this case, is rare. The index of suspicion should be high in older males with characteristic cutaneous features, which can be pleomorphic, rheumatologic, pulmonary, and hematological aberrations. Given the unique opportunity to evaluate the visible aspect of the disease, dermatologists can improve the time to diagnosis and potentially minimize associated morbidity and mortality.



**Abstract N°: 487****Chemotherapy and skin: study of nail and hair side effects**

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**Introduction & Objectives:**

Chemotherapies, used in a wide range of cancers, can induce a variety of side effects, including pharyngeal alterations. These effects are often underestimated, but can impact patients' quality of life through pain, functional discomfort or aesthetic appearance. The aim of our work is to describe the dander alterations observed in patients treated with chemotherapy and to assess the impact of nail alterations on patients' quality of life, and possibly study the therapeutic management of these abnormalities.

**Materials & Methods:**

We conducted a retrospective, descriptive study in the Department of Internal Medicine, including patients treated with chemotherapy, over a 1-year period from June 2023 to June 2024.

**Results:**

We enrolled 134 patients, with an average age of 49 +/- 8.3 years, and a sex ratio of 1.45. Among the 134 hospitalized patients, 52% were hospitalized for leukemia (69% for acute myeloid leukemia AML (on Daunorobucine, Cytarabine), 29% for acute lymphocytic leukemia ALL (under GRALL protocol) and 2% for plasma cell leukemia), 28% of patients had a lymphoma, including 17% for Hodgkin lymphoma HL (under ABVD protocol) and 8% for non-Hodgkin lymphoma NHL (under RCHOP protocol). Multiple myeloma (MM) accounted for 20% of patients treated with VCD.

Anagen effluvium was observed in 87.3% (of whom 60% had acute leukemia (L+M), 32% lymphoma (H+NH), and 8% MM), none of whom received preventive treatment. Changes in hair texture (finer, drier, brittle) were reported in 95.5% of patients. Regarding body hair, 80.6% of patients reported depilation.

Nail involvement was present in 66.4% of patients, 76% of whom presented with onycholysis, 57% with onychoclasia, 28% with peri onyxia, 20% with melanonychia). Isolated fingernail involvement was predominant, reported in 68.5% of cases, followed by associated fingernail and toenail involvement in 22.5% of patients, and 9% had isolated toenail involvement.

**Conclusion:**

Our study highlights the prevalence and diversity of phanereal alterations in patients undergoing chemotherapy. These results justify increased attention to the prevention and monitoring of these side effects, aimed at reducing the psychological and aesthetic impact on quality of life.





**Abstract N°: 533****“multicentric reticulohistiocytosis with disfiguring facial involvement: a rare and challenging case”**Sooriya Sekar<sup>\*1</sup>, Vijesh Valsalan<sup>2</sup>, Anuja George<sup>2</sup>, Smitha Varghese<sup>2</sup><sup>1</sup>governement medical college, thiruvananthapuram, dermatology, thiruvananthapuram, India<sup>2</sup>Thiruvananthapuram, dermatology, Thiruvananthapuram, India**Introduction & Objectives:**

Multicentric Reticulohistiocytosis (MRH) is a rare systemic disorder characterized by histiocyte proliferation, typically presenting with skin lesions and arthropathy. Most common in middle-aged women, MRH can also affect men and involve internal organs and can be associated with malignancies. This case explores an unusual MRH presentation in a 38-year-old male with disfiguring keloidal plaques, hypergammaglobulinemia, and lung involvement.

**Materials & Methods:**

A 38-year-old male presented with a two-year history of asymptomatic, disfiguring keloidal lesions around his eyes, initially on the lower eyelids and enlarging to obscure his vision. He also reported left elbow pain for two months. Examination revealed brownish keloidal papules, plaques, and nodules on the face, resembling a “leonine” appearance, and tenderness in the left elbow with restricted movement, suggesting arthropathy. Differential diagnoses included necrobiotic xanthogranuloma, neurofibroma, and lobomycosis. A skin biopsy revealed MRH features, with histiocyte proliferation, foam cells, multinucleated giant cells, and Touton giant cells. Immunocytochemistry for LCA 45 and vimentin was positive, confirming the diagnosis of MRH. Other baseline investigations were done, serum protein electrophoresis indicated hypoalbuminemia and hypergammaglobulinemia. A bone marrow biopsy and lytic lesion biopsy were advised, but the patient declined. An X-ray of the left elbow revealed a lytic lesion in the lower humerus, suggesting bone involvement. A CT scan of the thorax showed subpleural reticulation in both lungs, indicating pulmonary involvement, which occurs in about 20% of MRH cases.

**Results:** MRH is a rare disorder first described by Sir William Osler in 1898, marked by histiocyte proliferation in the skin and joints. The pathogenesis is unclear but may involve autoimmune or inflammatory processes, with abnormal macrophage or dendritic cell activity. Though more common in middle-aged women, MRH can affect men as well. The condition’s hallmark is skin lesions and polyarthritis, often resembling rheumatoid arthritis (RA), in early stages. MRH, however, is distinguished by its characteristic skin lesions and systemic features such as pulmonary involvement, as seen in this case. Approximately 20-25% of MRH cases are linked with malignancies, including carcinomas of the bronchus, stomach, breast, and ovary, as well as lymphomas and melanomas. MRH may be a paraneoplastic syndrome, with the immune system’s response to malignancy activating histiocytes. In this case, the patient’s MRH presented with a lytic humeral lesion and hypergammaglobulinemia, raising suspicion of multiple myeloma or other histiocytosis. Further evaluation was not pursued due to the patient’s refusal.

**Conclusion:** : This case highlights several important points: first, MRH should be considered when patients have skin lesions and joint pain. Second, is the association with malignancy which requires thorough investigation. Lastly, treatment may need a multidisciplinary approach, especially when malignancy is present. The patient underwent excisions and reconstructive surgery to restore his vision. Further research is needed for better management of this rare condition.





**Abstract N°: 540****Lichen myxedematosus: a rare group of cutaneous mucinosis**

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**Introduction & Objectives:**

Scleromyxedema, also referred to as generalized diffuse lichen myxoedematosus, is an infrequent mucinous dermatosis associated with monoclonal gammopathy and often involving multiple extracutaneous organ systems. Although the precise pathogenesis of scleromyxedema has not been completely elucidated, it is hypothesized to result from the stimulation of glycosaminoglycan synthesis.

Clinically, scleromyxedema is characterized by a disseminated eruption of firm, waxy, dome-shaped or flat-topped papules and nodules, typically measuring 2-3 mm, which may coalesce to form plaques on the head, neck, trunk, and extremities. The scalp and mucosal membranes are usually spared. The condition exhibits no significant gender predilection and is most frequently observed in adults within the fifth and sixth

decades of life. The management of scleromyxedema remains challenging due to its rarity and the absence of standardized treatment protocols. Various therapeutic approaches have been explored, including the use of corticosteroids, melphalan, thalidomide,

intravenous immunoglobulin (IVIG), retinoids, extracorporeal photophoresis, and autologous stem cell transplantation. Nevertheless, a definitive treatment algorithm has yet to be established.

**Materials & Methods:**

Here, we report a 59-year-old male patient who presented with a four-year history of multiple 2-3 mm skin-colored, waxy papules on the dorsum of the hands, neck, postauricular areas, face, and arms. A punch biopsy was performed based on the preliminary diagnosis of cutaneous mucinosis.

**Results:**

Histopathological examination revealed a significant amount of mucinous material in the superficial dermis beneath the normal epidermis, and the findings were consistent with cutaneous mucinosis.

The patient was started on acitretin 10 mg treatment. Rheumatology consultation was requested for the patient, whose ANA and anti-Ro52 tests were positive. No rheumatological involvement was detected in the patient. Hematology consultation was also requested for systemic screening. The patient showed IgG lambda monoclonal gammopathy on serum immunofixation electrophoresis and monoclonal gammopathy on protein electrophoresis. Bone marrow aspiration was normal. Hematological malignancy was not suspected in the patient, and he was followed up. The lesions partially regressed with acitretin and narrow-band UVB (nbUVB) treatment.

### **Conclusion:**

Lichen myxedematosus (LM) is an idiopathic cutaneous mucinosis encompassing several subtypes.

For the diagnosis of scleromyxedema, the following criteria must be met: (i) a generalized papular and sclerodermoid eruption; (ii) histological evidence of mucin deposition, fibroblast proliferation, and fibrosis; (iii) monoclonal gammopathy; and (iiii) the absence of thyroid disease.

The exact pathophysiology of scleromyxedema remains incompletely understood.

The most widely accepted hypothesis glycosaminoglycan synthesis and promote fibroblast proliferation. Although monoclonal gammopathy is commonly observed, IgG is the predominant paraprotein in patients with disseminated disease, and the level of paraproteinemia does not correlate with disease severity.

Furthermore, mucin deposition observed in autopsy specimens does not consistently correlate with the clinical manifestations of the disease.

We wanted to present our case with typical skin and histopathological findings due to rarity of Scleromyxedema.



**Abstract N°: 621****Sports Dermatology: Cycling**

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**Introduction & Objectives:**

Cycling has emerged as a globally popular sport and means of transportation, offering substantial health benefits. However, cyclists are uniquely vulnerable to various dermatological conditions due to prolonged mechanical friction, sustained pressure, sweat retention, and extensive environmental exposure. Despite their prevalence, cycling-related dermatological issues are often underdiagnosed and inadequately addressed. This research synthesizes current literature to provide a comprehensive overview of cycling-associated dermatoses, their etiology, management strategies, and preventive measures.

**Materials & Methods:**

A comprehensive literature review was conducted using PubMed to collate relevant studies on cycling-related dermatoses using the search terms “skin diseases” or “dermatitis” or “dermatoses” or “skin disorder” or “cutaneous condition” or “friction dermatitis” or “contact dermatitis” or “saddle” or “pressure ulcer” or “chafing” or “folliculitis” or “abrasion.” The selection criteria included peer-reviewed studies, clinical trials, and review articles focusing on skin disorders linked to cycling. The study synthesizes findings from dermatological, sports medicine, and epidemiological research published in peer-reviewed journals in English.

**Results:**

Cyclists are prone to friction- and pressure-related dermatoses resulting from extended contact with the saddle, tight-fitting gear, and repetitive movement. Genital and perineal conditions are also common due to prolonged saddle pressure, yet remain underdiagnosed due to its associated stigma. Infectious dermatoses, like impetigo, folliculitis, and tinea pedis, frequently arise from sweat accumulation and skin barrier disruption. Outdoor cyclists face heightened risks of sun-induced conditions such as sunburn, actinic keratosis, and melanoma, while cold weather increases susceptibility to frostbite, chilblains, and windburn. Contact dermatitis from cycling gear and equipment is also a significant concern.

**Conclusion:**

Cycling presents distinct dermatological challenges that warrant greater clinical attention. Dermatologists and sports medicine specialists must be aware of these conditions to provide timely diagnosis and management. Protective measures, including proper bike fitting, moisture-wicking and UV-protective clothing, appropriate hygiene practices, and barrier creams can significantly mitigate dermatological risks. Raising awareness among cyclists and healthcare providers can enhance early intervention and improve both performance and quality of life. By integrating preventive dermatological strategies into routine cycling practices and promoting awareness among athletes and clinicians, the burden of cycling-related dermatoses can be substantially reduced. Future research should focus on optimizing protective gear, developing advanced skincare solutions tailored for cyclists, and exploring long-term dermatological outcomes in professional and recreational cyclists.



**Abstract N°: 676****“Photoprotection and lupus: When knowledge does not overshadow practice”**

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**Introduction & Objectives:**

Systemic lupus erythematosus (SLE) is an autoimmune disease ranging from skin issues to severe systemic conditions that can compromise life expectancy. Ultraviolet (UV) radiation is known to worsen SLE symptoms in both adults and children. Sun exposure can trigger or intensify cutaneous and systemic manifestations, making effective sun protection essential for managing the disease and improving patients' quality of life. Managing SLE requires heightened vigilance and a comprehensive understanding of patients' knowledge and behaviors related to the disease and its treatment.

**Materials & Methods:**

A 15-question questionnaire was created on Google Forms and administered to patients being followed for lupus in the dermatology department.

**Results:** A total of 30 patients, aged 11 to 71 years, with a mean age of  $43.8 \pm 15.1$  years, were included in the study. Of these, 80% were female, reflecting the typical lupus patient profile. 73.3% had been diagnosed for more than 5 years, indicating a thorough understanding of the disease. 63.3% had isolated cutaneous lupus, while 36.7% had cutaneous involvement as part of systemic lupus. Additionally, 63.3% had a history of photosensitivity.

Regarding risk awareness, 76.6% recognized the link between sun exposure and worsening lupus symptoms, while 23.3% were unsure or unaware. The most commonly reported symptoms were skin rashes (86.6%) and fatigue (56.6%), followed by joint pain. 80% of patients consider photoprotection to be very important for managing their condition, while 20% consider it moderately important.

Regarding protective measures, 53.3% regularly use sunscreen products (with only 18.75% reapplying every 2 hours), 26.6% use them occasionally, and 20% do not use them at all. Among sunscreen users, 25% choose SPF 30-50, 66.6% use SPF 50+, and 8.3% choose SPF 30 or are unsure. The most common protective behaviors include avoiding exposure between 10 am and 4 pm (86.6%) and wearing wide-brimmed hats (83.3%).

Regarding information sources, 43.3% obtain information mainly from their primary care physician (dermatologist, internist, etc.), and 36.6% rely on online resources or advice from family and friends. 56.6% of patients mentioned a lack of clear guidance, and 60% expressed a desire for more information on photoprotection.

**Conclusion:** The presence of cutaneous signs predicts better photoprotective behavior in lupus patients. While there is general awareness of sun-related risks, the findings highlight the need for improved photoprotection practices and educational support due to the photoinduced exacerbation of lupus. Recommendations include enhanced education, practical support, and improved resources for patients.



**Abstract N°: 735****Paraneoplastic anti-synthetase syndrome – a dangerous mimic**Shihuan Valencia Long<sup>\*1</sup>, Lester Juay<sup>1</sup><sup>1</sup>National University Hospital, Singapore**Introduction & Objectives:**

Antisynthetase syndrome (ASyS) is an uncommon systemic autoimmune disorder characterized by the presence of autoantibodies targeting aminoacyl-transfer RNA (tRNA) synthetase. ASyS is an uncommon presentation of paraneoplastic syndrome, and we report a case of anti EJ-antibody and anti Ro-52 positive ASyS associated with hepatocellular carcinoma.

**Materials & Methods:**

Nil

**Results:**

A 68-year-old gentleman with worsening hand rashes for a year presented with new-onset abdominal tenderness and rib pain. There were fissured hyperkeratotic plaques on his hands and feet with periungual erythema, and scaly violaceous plaques over the lateral aspects of his thighs and forearms. Laboratory investigations revealed positive anti-nuclear antibody, strongly positive anti-EJ and anti-Ro-52 levels. Creatine kinase and aldolase levels were normal. Histopathology of the forearm lesions showed subacute spongiotic dermatitis with focal basal vacuolisation and increased dermal mucin. Further imaging revealed subpleural reticulation, multifocal arterially-enhancing masses in the liver and metastatic osseous deposits in bilateral ribs. He was eventually diagnosed with metastatic hepatocellular carcinoma and paraneoplastic anti-synthetase syndrome (ASS). Unfortunately, he had declined palliative chemotherapy and was referred to a hospice for end-of-life management.

**Conclusion:**

While paraneoplastic ASS is uncommon, its manifestation of mechanical hands needs to be recognised and distinguished from less insidious causes such as hand eczema. Paraneoplastic ASS has been reported to improve upon initiation of treatment for the underlying malignancy.



**Abstract N°: 785****Pure cutaneous form of adult-onset IgA vasculitis: two cases**

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**Introduction:**

IgA vasculitis, previously known as rheumatoid purpura or Henoch-Schönlein syndrome, is a systemic small-vessel vasculitis with immunoglobulin A deposits. Isolated skin involvement is rarely reported in the literature. We report two cases in adults.

**Case presentations:**

Case 1: A 74-year-old woman with a history of type 2 diabetes and atrial fibrillation on Acenocoumarol had been presenting with necrotic-petechial, pruritic vascular purpura of all four limbs, predominantly distally, for 3 months. No triggering factors were identified. There were no clinical or paraclinical signs suggesting extracutaneous involvement. A work-up including immunoelectrophoresis of serum proteins, ANCA testing, antinuclear antibodies, and cryoglobulinemia was unremarkable. Histology showed leukocytoclastic vasculitis with fibrinoid necrosis. Direct immunofluorescence showed vascular deposits of IgA.

Case 2: A 36-year-old man with no with no notable medical history presented with recurrent episodes of petechial purpura with necrotic progression on the abdomen and lower limbs for 18 months. No extracutaneous involvement was found on clinical or paraclinical exams. The work-up revealed no thrombophilia, cryoglobulinaemia or cryofibrinogen, and no autoimmune disease. Skin biopsy revealed leukocytoclastic vasculitis with IgA deposits.

Both patients were diagnosed with pure cutaneous IgA vasculitis. Treatment with colchicine 1 mg/day was started with rapid clinical improvement. Remission was maintained at 1 year (Case 1) and 6 months (Case 2) after the start of treatment.

**Discussion:**

IgA vasculitis is rare in adults. The clinical manifestations are similar to those in children. The disease typically progresses in a chronic manner with a poorer prognosis due to more severe renal involvement. In one-third of cases, nephropathy presents later (up to several months), highlighting the importance of prolonged follow-up.

An exclusive cutaneous presentation without the typical features of rheumatoid purpura is rarely described in the literature. It is sometimes associated with monoclonal IgA gammopathy, although this dysglobulinemia was not found in our patients. The therapeutic management is not clearly defined, and various options are possible: systemic corticosteroids, dapsone, or colchicine. The latter has proven effective in both of our cases.

**Conclusion:**

Adult-onset cutaneous IgA vasculitis is a rare entity that appears to have a good prognosis. However, regular monitoring is still necessary to detect the development of any systemic involvement.





**Abstract N°: 800****McCune-Albright syndrome: When beauty hides an ocular storm**

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**Introduction & Objectives:**

McCune-Albright syndrome is a rare hereditary disorder characterized by the association of fibrous dysplasia of the bone, café-au-lait skin patches and endocrine abnormalities. Ocular manifestations such as retinal vasculitis may also occur, although this is less frequent.

The aim of our work is to draw attention to an extremely rare clinical presentation through a case associating McCune-Albright syndrome and retinal vasculitis.

**Observation:**

This 20-year-old patient, with a history of cerebral aneurysm in 2020, and depression under treatment for two years, presented with an isolated progressive decrease in visual acuity that had been evolving for 10 months. The ophthalmological examination revealed impaired visual acuity on the right (counting fingers at 2 metres), and preserved visual acuity (10/10) on the left. The fundus showed papillary hyperaemia (papilla 1/10), poor macular reflex and exudative occlusive vasculitis with centrolateral haemorrhage and hyalitis on the right. Examination of the left eye was unremarkable. Fluorescein angiography showed papillitis, occlusive venous vasculitis and laserized ischemia with persistent active neo-vessels close to the macula. In addition, dermatological examination revealed congenital café-au-lait spots, segmental with irregular Maine coast margins, blaschko-linear stopping on the midline of the face, "S"-shaped on the thorax, "V"-shaped on the back and linear on the left upper limb. The diagnosis of retinal vasculitis in McCune-Albright syndrome was made in view of the cutaneous involvement, endocrine disorders (precocious puberty) and ocular involvement, with no bone involvement found. The patient was put on a corticosteroid bolus with slight improvement. A panretinal photocoagulation laser was indicated to inactivate the territories of the corticosteroids.

**Discussion:**

McCune-Albright syndrome is a fascinating example of the complexity of genetic diseases. The main features of McCune-Albright syndrome include: bone dysplasia, leading to bone fragility or deformity; endocrine disorders, including precocious puberty, thyroid disorders and other hormonal imbalances; and pigmented café-au-lait skin patches often appearing asymmetrically and well-defined. Retinal vasculitis, although not systematically associated with the syndrome, may be the result of systemic inflammation or an altered immune response. This can lead to symptoms such as visual blur, reduced visual acuity and halos or floating spots. Ocular complications can have a devastating impact on patients' quality of life. Treatment of McCune-Albright syndrome is usually multidisciplinary, involving endocrinologists, ophthalmologists and other specialists. Management of the symptoms of retinal vasculitis may include corticosteroids or other immunosuppressive drugs, depending on the severity of the inflammation. Increased awareness of McCune-Albright syndrome and its manifestations may lead to earlier diagnosis and better symptom management.

**Conclusion:**

McCune-Albright syndrome, and the retinal vasculitis that can be associated with it, illustrate the unique challenges encountered in the management of complex genetic diseases. Thanks to a multidisciplinary approach, ongoing research and increased awareness, it is possible to significantly improve the quality of life of patients affected by this rare condition.



**Abstract N°: 808****Assessing Neutrophil-to-Lymphocyte Ratio as a Key Biomarker in the Differentiation of Deep Vein Thrombosis and Erysipelas**Géza Róbert Nagy<sup>1</sup>, Mohammed Almakrami\*<sup>1</sup><sup>1</sup>University of Szeged, Department of Dermatology and Allergology, Szeged, Hungary**Introduction & Objectives:**

Deep vein thrombosis (DVT) and erysipelas are common medical conditions with overlapping clinical features, such as redness, swelling, and pain in an extremity. In clinical practice, diagnostic tools like Doppler ultrasound, serum D-dimer, and inflammatory markers are used to differentiate between these conditions. However, there are situations where these methods have limitations. Therefore, our aim was to identify new biomarkers to expedite the assessment of patients with DVT or erysipelas. One such biomarker of interest is the Neutrophil-to-Lymphocyte Ratio (NLR), known for its effectiveness in quantifying inflammation in various diseases. Our objective was to evaluate the utility of NLR in distinguishing between DVT and erysipelas.

**Materials & Methods:**

In this retrospective clinical study, we collected data from patients treated at the First Department of Internal Medicine and the Department of Dermatology and Allergology in Szeged from January 2022 to December 2022. A total of 75 patients were identified, and 46 patients met our inclusion criteria.

**Results:**

The 46 study patients were divided into two groups; half had DVT (n=23), and the other half had erysipelas (n=23). The median age was 73 years (range 22-94). We tested NLR correlations with patient age, duration of symptoms, white blood cell count, C-reactive protein, procalcitonin, and D-dimer levels using Spearman rank correlation analysis. We found a correlation between NLR and the last 4 biomarkers ( $p=0.012$ ,  $p=0.001$ ,  $p=0.05$ ,  $p=0.0015$ ) respectively. Next, using the Mann-Whitney U test, we evaluated the differences in these biomarkers between the erysipelas group and the deep vein thrombosis group, and we found a significant association between pretreatment NLR values and erysipelas ( $p=0.0017$ ). Receiver Operating Characteristic (ROC) curve analysis determined an optimal NLR cutoff of 4.91 for predicting erysipelas, with 91% sensitivity and 70% specificity.

**Conclusion:**

Our findings suggest that NLR has promise as a valuable marker for differentiating between DVT and erysipelas, making it a practical tool for routine patient assessment. Furthermore, NLR is cost-effective and readily available, making it easily implementable in everyday clinical practice.





## Abstract N°: 812

### Atypical presentation of adult Still's disease: difficulty and pitfalls, diagnosis and prognostic value: a case report

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#### Introduction & Objectives:

Adult-onset Still's disease (AOSD) is a rare systemic autoinflammatory syndrome with challenging diagnostics. Atypical skin rashes often cause diagnostic delays, sometimes lasting years. The prognosis is variable and can be severe, with life-threatening or functional consequences. Identifying predictive factors for visceral complications is crucial. This case report aims to describe an atypical urticarial presentation of AOSD and its management.

#### Observation:

Our patient is a 54-year-old man with no particular pathological history, hospitalized for a generalized urticarial rash that did not leave a scar, associated with dermographism, inflammatory arthralgia, myalgia and odynophagia with weight loss estimated at 8 kg. The histology of the skin biopsy revealed an appearance of neutrophilic urticarial dermatosis with slight keratinocyte necrosis. Biology noted an inflammatory syndrome with PNN at 21000/ul, a CRP at 70 with an accelerated VS, a normal ferritin, lymphopenia at 970, ALAT at 2 times normal.

Microbiological, neoplastic and immunological explorations came back negative and the rest of the assessment was without abnormality.

The diagnosis of MSA was made based on the Fautrel criterion after eliminating any infectious, neoplastic or immunological disease and general corticosteroid therapy initiated at 1 mg/kg/day associated with colchicine (2 mg/day, later reduced to 1 mg/day) which allowed apyrexia in 2 days, and regression of the inflammation and rash in 2 weeks.

The clinical course was marked by the complete resolution of symptoms, tapering of corticosteroids after three months, and discontinuation of colchicine.

#### Discussion:

AOSD is a very heterogeneous disease. Its typical cutaneous manifestation is a fleeting non-pruritic rash that coincides with fever spikes. In 14% of cases, AOSD can be associated with atypical fixed and pruritic dermatoses. The atypical skin rash is characterized mainly by its papular, pruritic and urticarial appearance, dermographism, residual pigmentation, persistent plaques, a dermatomyositis-like appearance, or a pseudo sweet plaque, to vascular purpura, or even erythroderma. These manifestations may cause isolated organ damage or lead to systemic complications, including multiorgan failure, with potentially life-threatening consequences.

Recognizing atypical skin rashes is of great diagnostic value in adult Still's disease given their frequency. Their coexistence with the disease's cardinal signs should prompt timely diagnosis.

Urticaria with pruritus is the most common manifestation of atypical eruptions, moreover an urticarial eruption with dermographism is predictive of late relapse of the disease, while Polyarthrititis at diagnosis is predictive of erosive progression.

These atypical clinical forms may also serve as markers of disease severity and have been linked to neoplasias occurring up to six years post-diagnosis. Pro-inflammatory cytokine profiles common to AOSD, hematologic malignancies, and breast

cancer (IL-6, IL-18, IL-2) support the hypothesis of a subclinical neoplastic process, necessitating prolonged patient follow-up.

**Conclusion:**

Early clinico-histological diagnosis of AOSD may have prognostic value, especially regarding neoplasias. Its prognosis is complex, with risks of visceral complications and chronic, corticosteroid-dependent disease affecting quality of life.

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**Abstract N°: 866****Celiac Disease and Morphea: Unveiling the Potential Link – A Case Report**Ilhame Jegoual<sup>1</sup>, Bendaoud Layla<sup>1</sup>, Saloua Hazmiri<sup>1</sup>, Mariem Aboudourib<sup>1</sup>, Hocar Ouafa<sup>1</sup>, Amal Said<sup>1</sup><sup>1</sup>dermatology Venereology department, Arrazi hospital mohammed 6, Marrakech, Morocco**Introduction:**

Celiac disease is a chronic autoimmune disorder triggered by the ingestion of gluten, primarily affecting the small intestine in genetically predisposed individuals. Morphea is an inflammatory and fibrosing condition involving the skin and underlying connective tissues, characterized by localized, circumscribed sclerotic plaques, most commonly on the head, neck, trunk, and extremities. We report a case of concurrent celiac disease and localized scleroderma, suggesting a potential association between these two conditions.

**Observation:**

We present the case of a 41-year-old male with type 1 diabetes, managed with insulin therapy, who has been followed in the gastroenterology department for celiac disease, confirmed by biopsy and positive immunological testing on a gluten-free diet, and for collagen enteritis, for which he is receiving salazopyrin 3 g/day. The onset of symptoms occurred 5 months ago, beginning with bilateral lower limb edema, followed by cutaneous sclerosis of the feet and ankle arthralgia, without any other associated signs (notably no Raynaud's phenomenon or digital ulcerations). The disease progressed with the absence of fever or any other general symptoms. On clinical examination, acrosclerosis of the ankles was noted, with a Rodnan score of 4. Laboratory tests revealed microcytic hypochromic anemia (hemoglobin 9 g/dL) with low ferritin levels (8 ng/mL), a C-reactive protein (CRP) of 1 mg/L, and an erythrocyte sedimentation rate (ESR) of 9 mm/h. Skin and fascial biopsies were consistent with localized scleroderma without involvement of the fascia, and MRI of both ankles was unremarkable. The patient was treated with prednisone 30 mg daily and methotrexate 10 mg weekly, resulting in slight clinical improvement.

**Discussion:**

Celiac disease (CD) is a chronic, inflammatory enteropathy mediated by an immunological mechanism, triggered by gluten ingestion, and occurring in genetically predisposed individuals. It is frequently associated with other organ-specific autoimmune disorders, such as thyroiditis, type 1 diabetes, dermatitis herpetiformis, autoimmune myocarditis, Addison's disease, autoimmune hepatitis, and primary biliary cirrhosis. Although rare, it may also coexist with systemic autoimmune diseases, including connective tissue diseases such as systemic sclerosis, as reported in the literature. However, the cause-and-effect relationship remains unclear. While rare, transitions from morphea to systemic sclerosis have been observed, suggesting that this entity may represent a localized form of systemic sclerosis. A potential association with localized cutaneous scleroderma, involving the connective tissue, has been described in the literature, as demonstrated by our patient's case.

**Conclusion:**

It is necessary to conduct research to explore a potential association between morphea and celiac disease, as well as their interactions with other autoimmune disorders, to further understand their respective pathophysiology and improve their management.





**Abstract N°: 920**

**Comparison of the dermatological manifestations between the patients with Chronic Kidney Disease (CKD) on a regular hemodialysis and the patients on a conservative medical treatment**

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**Introduction & Objectives:**

The skin and the kidney constitute two major organ systems of the human body. The skin can serve as an external indicator of underlying renal pathologies, making it a valuable tool for clinicians in diagnosing and monitoring kidney diseases.

Aim is to determine the spectrum of dermatological manifestations in patients with chronic kidney disease (CKD) and to compare these findings between patients on dialysis (hemodialysis group) and patients without (non - dialysis group).

**Materials & Methods:**

This is a one-year duration cross sectional study, involved 200 patients with (CKD), who already or recently diagnosed by nephrologist, they had at least one dermatological finding. Detailed history, examination, estimated glomerular filtration rate (eGFR), and the required investigations were carried out. There were (100) patients on regular hemodialysis at least twice a week for minimum of three months (dialysis group). Another (100) patients were in stage one to stage four (CKD) on conservative medical

treatment (non- dialysis group).

**Results:**

This study included 200 patients with CKD, with the mean age of patients was (49.41 ± 15.49) years and slight male predominant (50.5%). Xerosis (87%), pigmentary alterations (74.5%) mucosal pallor (68%), nail changes (63.5%) and pruritus (55.5%) were the most common dermatological manifestations among patients with (CKD) in order of the frequency. CKD-associated pruritus, ichthyosis, brown hyperpigmentation, sallow -yellow hue, dusky red (fistula related limb), half and half nail, mucosal changes and edema were significantly higher among dialysis patients.

**Conclusion:**

The most common identified dermatological manifestations were xerosis, pigmentary alterations, mucosal pallor, nail changes and pruritus in order of frequency. The prevalence of pigmentations increases with increase dialysis duration and disease severity, while the prevalence of the xerosis increases with increase disease severity only. Dialysis often exacerbate many skin findings and not highly efficient in alleviate skin morbidity.





**Abstract N°: 1016****Beyond Aesthetics: The Complex Therapeutic Dilemma of Parry-Romberg Syndrome with Neurological Sequelae**

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**Introduction & Objectives:**

This case report describes a 19-year-old female with Parry-Romberg syndrome (PRS), a rare disorder characterized by progressive hemifacial atrophy and scleroderma. The patient developed intracranial hypertension (IH) and bilateral optic atrophy, leading to significant visual impairment. The objectives were to document the clinical progression, highlight the multidisciplinary management challenges, and discuss the potential mechanisms linking PRS, IH, and optic atrophy.

**Materials & Methods:**

The patient's medical history, clinical examinations, and diagnostic findings were systematically reviewed. Imaging studies, including MRI and CT scans, were performed to assess intracranial and orbital changes. Laboratory tests, including anti-MOG and anti-AQP4 antibodies, were conducted to rule out autoimmune etiologies. The patient's treatment history, including corticosteroids, methotrexate, and ventriculoperitoneal shunt placement, was also evaluated.

**Results:**

The patient presented with progressive hemifacial atrophy and cutaneous sclerosis since early childhood, managed with oral corticosteroids and injectable methotrexate, which improved cutaneous symptoms. Lipofilling procedures enhanced aesthetic outcomes. In 2024, she developed IH, requiring ventriculoperitoneal shunt placement. In 2025, she experienced visual decline (3/10 in the left eye) and headaches. MRI revealed bilateral optic atrophy, while CT showed collapsed right lateral ventricle due to shunt overdrainage. Anti-MOG and anti-AQP4 antibodies were negative, and immunologic workup was unremarkable. High-dose intravenous methylprednisolone (1 g/day for 5 days) failed to improve vision.

**Conclusion:**

This case highlights the complex progression of PRS, a rare condition with potential systemic and neurologic complications. The development of IH and bilateral optic atrophy in this patient underscores the need for vigilant monitoring and multidisciplinary management. The lack of response to corticosteroids suggests a non-inflammatory mechanism for optic atrophy, possibly related to chronic IH or vascular insufficiency. This report emphasizes the importance of integrating dermatologic, neurologic, and ophthalmologic care in managing PRS and its complications. Further research is needed to elucidate the underlying mechanisms and optimize therapeutic strategies for such challenging cases.



**Abstract N°: 1091****Peripheral T-cell lymphomas with predominant lymph node involvement revealed by skin lesions: A report of 5 cases**

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**Introduction & Objectives:**

Peripheral T-cell lymphomas (PTCL) are aggressive neoplasms originating from T cells, with nodal predominant PTCLs being the most common non-cutaneous subtype, representing 5–10% of cases. These include angioimmunoblastic T-cell lymphoma (AITL), anaplastic large cell lymphoma (ALK-negative and ALK-positive), and PTCL not otherwise specified (NOS). PTCLs typically present with lymphadenopathy, and in 21% of cases, extranodal involvement, including various cutaneous manifestations. This study aims to explore the epidemiological, clinical, and histopathological features of nodal predominant PTCLs presenting with cutaneous involvement.

**Materials & Methods:**

This is a retrospective, descriptive, and analytical study conducted in the Dermatology Department of Mohammed VI University Hospital in Oujda over ten years (December 2014 – July 2024). It includes cases of nodal predominant PTCL presenting with cutaneous involvement. A total of 43 lymphoma cases were collected, including 28 cases of mycosis fungoides, 3 cases of Sézary syndrome, 6 cases of B-cell lymphoma, and 6 cases of nodal predominant PTCL.

**Results:**

Our study identified six cases of peripheral T-cell lymphoma with predominant nodal involvement, accounting for 14% of all lymphomas: three angioimmunoblastic T-cell lymphomas, two peripheral T-cell lymphomas NOS, and one anaplastic large-cell lymphoma.

The mean patient age was 58.33 years (range: 51–65), with a male predominance (M/F ratio: 5:1). Smoking was the most common risk factor (50%). Two patients had occupational exposure, one was treated with methotrexate for chronic inflammatory rheumatism, and one had dermatomyositis. Clinical presentation included pruritic maculopapular rash (33.3%), ulcerative-proliferative lesions (50%), and generalized hyperpigmented plaques (16.6%). All patients had polyadenopathy, and one developed a drug-induced eruption initially diagnosed as DRESS but later reconsidered.

Skin and lymph node biopsies were performed in all cases. In three patients, histology suggested cutaneous lymphoma involvement, with angioimmunoblastic T-cell lymphoma confirmed on lymph node biopsy. Two patients had diffuse dermal infiltration by large CD30+ T lymphoid cells, consistent with peripheral T-cell lymphoma NOS. The last patient had histological features suggestive of pigmented lichen, with lymph node biopsy confirming anaplastic large-cell lymphoma.

Bone marrow biopsy showed reactive changes in all cases. Four patients (66.6%) had systemic symptoms, and three had splenomegaly. According to the Lugano classification, two were stage IIIIE, two IIISE, and two stage IV. Elevated  $\beta$ 2-microglobulin and LDH levels were frequent (n=5). The IPI score indicated low-intermediate risk (n=3), intermediate-high (n=2), and high risk (n=1).

All patients received chemotherapy (CHOEP: n=5, R-CHOP: n=1), with cutaneous improvement in two cases. Three patients died within a year of dermatological involvement (IPI 3–4).

**Conclusion:**

This study highlights the need for clinical suspicion of peripheral T-cell lymphoma in the differential diagnosis of adult patients with cutaneous lesions and underscores the importance of performing a staging workup in cases of cutaneous lymphoma involvement.

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**Abstract N°: 1092****A Case of Reactive Infectious Mucocutaneous Eruption in an Adult Secondary to Influenza B and COVID-19 Infections**

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**Introduction & Objectives:**

We report the case of a 21-year-old man who presented to the respiratory team with a 10-day history of myalgia, fever, and productive cough. Three days prior to admission, he developed painful oral and penile ulcerations, followed by the appearance of a rash on his trunk and limbs 48 hours later. The initial diagnosis of suspected Stevens-Johnson syndrome (SJS) prompted a referral to dermatology.

**Materials & Methods:**

The patient reported severe oral and throat pain, along with excessive salivation. His only prior medical history was asthma, managed with inhalers. He denied using any other medications, including over-the-counter drugs. On examination, he had severe oral mucositis, conjunctivitis, and erosive involvement of the penile shaft and glans. A mild, targetoid rash with a few scattered vesicles on his upper limbs was noted. Based on the clinical findings, a diagnosis of Reactive Infectious Mucocutaneous Eruption (RIME) was made. Microbiological investigations revealed positivity for Influenza B virus and a weakly positive result for SARS-CoV-2.

**Results:**

The patient required inpatient care, including intravenous fluids, pain management, topical corticosteroids, and multidisciplinary input from several specialties. He was discharged after two weeks and made a full recovery.

**Conclusion:**

The term RIME has recently been proposed to describe mucocutaneous manifestations triggered by respiratory infections<sup>1</sup>. It encompasses conditions such as *Mycoplasma pneumoniae*-induced rash and mucositis, first described in 2015<sup>2</sup>. Various infectious agents, predominantly viruses, have been linked to RIME.

The typical clinical presentation includes a preceding respiratory infection, followed 7-10 days later by erosive mucositis involving two or more mucous membrane sites. RIME most commonly affects the oral mucosa but can also involve ocular, urogenital, and anal regions<sup>1</sup>. Cutaneous eruptions occur in approximately two-thirds of cases and typically affect less than 10% of the body surface area. RIME is considered a distinct entity from Severe Acute Drug Reactions such as SJS, with a generally favourable prognosis<sup>2</sup>. However, approximately 20% of patients may develop long-term sequelae.

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**Abstract N°: 1102****Hyperlipidemia not only for internists – a clinical case of a patient with eruptive xanthoma**

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**Introduction & Objectives:**

Eruptive xanthoma is associated with severe hypertriglyceridemia. The disease is clinically manifested by the well-demarcated, yellowish to orange papules occurring mainly on the trunk and inner parts of the thighs. Eruptive xanthoma may be the first symptom of a severe hypertriglyceridemia

**Materials & Methods:**

A 65-year-old patient diagnosed with hypertension, type 2 diabetes and abdominal obesity was admitted to the dermatology department due to diffuse skin lesions in the form of yellow nodules. According to the patient, the lesions appeared about a year before hospital admission and were not accompanied by subjective symptoms. In therapy, he had previously used only topical steroids without clinical effect. The physical examination revealed numerous, diffuse yellow papules and nodules, located mainly on the neck, back, thighs, knees, and extensor parts of the calves. Dermoscopic examination revealed yellow-orange, unstructured areas with telangiectasia and an erythematous-brown halo. Laboratory tests were performed and revealed severe hyperlipidemia with serum triglyceride concentration of 8165 mg/dL (reference range: 30-150 mg/dL), and total cholesterol 884 mg/dL (reference range: 130-200 mg/dL). Blood count was impossible to assess due to high lipemia.

**Results:**

Based on clinical symptoms and laboratory parameters, eruptive xanthoma was suspected. The patient was started on a low-fat diet, intensive insulin therapy, fenofibrate 215 mg/d, pioglitazone 30 mg/d, rosuvastatin 20 mg/d, and  $\Omega$ -3 fatty acids 3 g/d. After six days of therapy, triglyceride level decreased to 1785 mg/dL, total cholesterol to 565 mg/dL, and the lesions partially reduced. The patient was discharged home in good general condition with dietary and therapeutic recommendations. The histopathological examination confirmed the diagnosis of eruptive xanthoma. After one year follow-up the skin symptoms reduced to the hyperpigmented, small macules.

**Conclusion:**

The presented case illustrates the possible significance of skin lesions as an element of the clinical feature of severe hyperlipidemia. Early diagnosis and implementation of hypolipemic therapy reduce the risk of life-threatening conditions such as acute pancreatitis or acute heart failure.



**Abstract N°: 1136****Dupilumab-Induced Alopecia – A Case Report of Rapid-Onset Hair Loss - Case study**Cindy Kok<sup>1</sup>, Tara Sholji<sup>1</sup>, Andrea Tomizawa<sup>1</sup>, Orana Crainic<sup>2</sup><sup>1</sup>The Skin Hospital, Darlinghurst, Australia<sup>2</sup>Douglass Hanly Moir Macquarie Park Lab, Macquarie Park, Australia**Introduction & Objectives:**

Dupilumab is a monoclonal antibody targeting IL-4 and IL-13 cytokines, used for moderate-to-severe atopic dermatitis. Despite well-documented efficacy, emerging reports highlight rare side effects, including drug-induced alopecia. We report a case of rapidly-evolving scalp alopecia following dupilumab initiation.

**Materials & Methods:**

Case Report: A 30-year-old-female presented with erythroderma, secondary to generalised atopic dermatitis (EASI: 57.5, DLQI: 30, PGA: 5). Previous treatment included cyclosporin and oral glucocorticoid therapy. She avoided topical cortisone and calcineurin inhibitors due to steroid phobia and tachyphylaxis. Aside from a family history of thyroid disease, she had no personal or family history of autoimmune diseases. Baseline serology was unremarkable, and dupilumab was initiated. By her third dose of dupilumab 300mg, she reported improved eczematous symptoms, but new-onset scalp hair thinning and shedding.

**Results:**

A scalp biopsy demonstrated follicular miniaturisation and prominent sebaceous gland atrophy. There was no peribulbar inflammation in a “swarm of bees” pattern to suggest alopecia areata. Telogen count was greater than 50% with a majority of follicles shifted into catagen/telogen phase. Histology ruled out other differential diagnoses including psoriatic alopecia or cutaneous lupus. A diagnosis of dupilumab-induced alopecia was made, supported by similar case reports.

Dupilumab was discontinued and after six weeks, hair shedding had ceased and visible regrowth of vellus hair identified on examination.

**Conclusion:**

While dupilumab is being trialled off-label as treatment for alopecia areata, dupilumab-associated alopecia is uncommon. In such cases, hair loss was reported more often in males, occurring months after initiation of dupilumab. This case is notable for its rapid onset in a female patient after just three doses of dupilumab 300mg for atopic dermatitis.

Dermatologists should recognise the potential for dupilumab-induced alopecia, particularly in patients with severe atopic dermatitis. Early recognition can mitigate patient distress, enhance informed consent, and optimise clinical outcomes.





**Abstract N°: 1200****A family case of Neurofibromatosis type 1**

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A family case of Neurofibromatosis type 1

**Introduction & Objectives:**

Neurofibromatosis (NF) encompasses genetic neurocutaneous disorders leading to tumors in the nervous system and skin. NF type 1 and NF type 2 are the most prevalent forms, both inherited in an autosomal dominant manner.

NF1, or von Recklinghausen disease, manifests through neurofibromas, café-au-lait spots, freckling, and optic gliomas. In contrast, NF2 is distinguished by bilateral vestibular schwannomas and meningiomas. Management for both types involves regular clinical monitoring and appropriate medical interventions. Neurofibromatosis type 1 makes up about 96% of all neurofibromatosis cases. Prevalence is 1 in 3000 births.

**Materials & Methods:**

We present a 43-year old female patient that came to our clinic at the beginning of February 2025 with the desire to remove some lesions on her back. She confirms that they are not painful and that she did not notice any other clinical signs. The patient reported that her father suffers from similar symptoms but he had more on his back and they were bigger and larger in size. The patient also reports that her son, who is a teenager, started to notice some brown spots appearing on his trunk.

On examination, on her trunk and extremities there were multiple café-au-lait spots, neurofibromas, and axillar freckling. Patient was sent for a skin biopsy, an ophthalmologic examination, neurologist, and gastroenterologist.

**Results:** The histology from the skin biopsy showed mixed cell types, including: Schwann cells, perineural cells, fibroblasts, mast cells, and a collagenous extracellular matrix. We are still waiting for the other results to arrive. Regular monitoring of the patient and her relatives was proposed.

**Conclusion:**

The diagnosis of NF1 is primarily clinical, adhering to NIH criteria that necessitate at least two of the following: six or more café-au-lait macules, two or more neurofibromas or one plexiform neurofibroma, freckling in the axillary or inguinal regions, an optic pathway glioma, two or more Lisch nodules, distinctive osseous lesions such as sphenoid dysplasia, or a first-degree relative with NF1. Genetic testing is not frequently done.

Treatment for NF1 centers on symptom management and thorough monitoring for tumor progression and related complications. Benign neurofibromas often do not require intervention unless they become symptomatic, at which point surgical excision may be considered. Plexiform neurofibromas have 8% to 13% risk for malignant transformation, thus are addressed through surgical removal and, if necessary, adjunctive chemotherapy. Routine ophthalmologic assessments are essential to detect optic gliomas, which are managed with chemotherapy when visual function is at risk. Pediatric patients should be evaluated for learning and behavioral challenges, with counseling and support services provided to assist both patients and their families.

**Abstract N°: 1204****From a long standing yellowish plaque to a diagnosis of a rare disorder: a case of Erdheim-Chester Disease treated with vemurafenib**

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**Introduction & Objectives:**

Erdheim-Chester disease (ECD) is a rare, systemic, and potentially malignant non-Langerhans cell histiocytosis characterized by the infiltration of foamy histiocytes into multiple organ systems. The diagnosis of ECD is often difficult and time-consuming due to its rarity and heterogeneous presentation. The broad clinical spectrum of ECD varies from asymptomatic bone lesions to life-threatening multisystemic variants including the central nervous system, cardiovascular system, and retroperitoneum. Cutaneous involvement is observed in approximately one-third of cases, usually as xanthomatous plaques on the eyelids. In this report, we present a case of ECD, which was diagnosed after a thorough investigation of the yellow plaque lesion.

**Materials & Methods:**

A 39-year-old female presented with a progressively enlarging orange-colored plaque with a size of 5 x 2 cm on her left periorbital skin over the past two years. Besides, the patient had bilateral lower extremity pain persisting for 1.5 years, significantly affecting her mobility and quality of life. She had multiple sclerosis and a history of stroke. Her familial medical history was unremarkable.\*\*

**Results:**

Histological examination of skin biopsies showed bland appearing histiocytes characterized by abundant foamy (xanthomatous) cytoplasm in dermis, which were positive for CD68, Factor XIIIa and BRAF; negative for CD1a and S100 by immunohistochemistry. Touton giant cells were frequently present.

Immunohistochemical analysis of the bone marrow biopsy showed histiocytic cells positively stained with CD68, Factor XIIIa, and CD14 and negatively for CD1a, Langerin, and S100 supporting the diagnosis of non-Langerhans cell histiocytosis. Positron emission tomography/ computed tomography imaging revealed pathological 18-fluoro-2-deoxyglucose uptake in the bone marrow and widespread sclerotic bone lesions on the bilateral lower extremity. BRAF sequencing determined a BRAFV600E mutation. Depending on clinical and pathological findings, the patient was diagnosed as ECD and vemurafenib, a BRAF inhibitor therapy, was initiated. After three months of treatment, significant improvement was noted in bone pain and a remarkable reduction in the size and color of the orange plaque.

**Conclusion:**

The only diagnostic clue for ECD may be a single yellowish plaque, which needs to be investigated in detail in relation to other patient symptoms. Vemurafenib treatment may provide regression of systemic symptoms and cutaneous yellowish plaques of ECD with BRAF mutation.\*\*

**Abstract N°: 1256****dermatomyositis revealing primary biliary cirrhosis : a case report**

Farouk Elguennouni<sup>1, 2</sup>, Tarik Hanafi<sup>3</sup>, Jawad Azhari<sup>3</sup>, Mohamed El Amraoui<sup>3</sup>, Youssef Zemmez<sup>3</sup>, Rachid Frikh<sup>3</sup>, Naoufal Hjira<sup>3</sup>

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**Introduction & Objectives:**

Primary biliary cirrhosis (PBC) is a chronic autoimmune liver disease marked by bile duct destruction and anti-mitochondrial antibodies (AMA-M2). While PBC frequently coexists with other autoimmune disorders, its association with dermatomyositis (DM) is exceedingly rare. This case report and literature review aim to highlight the diagnostic challenges and clinical implications of DM as an initial presentation of undiagnosed PBC, emphasizing the importance of early autoimmune screening in patients with cholestasis or cytotoxicity.

**Materials & Methods:**

A 65-year-old woman with no prior medical history presented with facial erythema, edema, systemic fatigue, and elbow ulceration. DM was diagnosed based on clinical criteria (shawl sign, heliotrope rash), elevated creatine phosphokinase (CPK), hepatic cytolysis, electromyography-confirmed myogenic syndrome, and histopathological interface dermatitis. Muscle biopsy was inconclusive. After initiating systemic corticosteroids (1 mg/kg/day), she developed severe cholestasis (GGT 27× normal). Further workup excluded viral hepatitis and malignancy. Biliary MRI was normal, but AMA-M2 positivity confirmed PBC. Ursodeoxycholic acid (600 mg/day) was added. A literature review was conducted to contextualize this rare association.

**Results:**

The patient's DM improved transiently with corticosteroids, but cholestasis progressed, necessitating PBC diagnosis. Despite dual therapy, she developed corticosteroid-induced diabetes and succumbed to gastrointestinal hemorrhage three months post-diagnosis. Literature review identified sparse reports of DM-PBC overlap, predominantly in middle-aged women. PBC is associated with Sjögren's syndrome, scleroderma, and rheumatoid arthritis, but DM coexistence remains rare. Diagnostic criteria for PBC include cholestasis, AMA-M2 seropositivity, and histologic cholangitis. While liver biopsy aids staging, it is not mandatory. Early ursodeoxycholic acid improves outcomes, underscoring the need for prompt recognition.

**Conclusion:**

This case illustrates DM as a rare herald of PBC, advocating for autoimmune evaluation in patients with cholestasis and dermatomyositis. Despite timely intervention, the patient's fatal outcome highlights disease severity and complications from immunosuppressive therapies. Clinicians should maintain high suspicion for overlapping autoimmune conditions to guide multidisciplinary management. Early diagnosis of PBC in DM patients enables targeted treatment, potentially mitigating progression. Further research is needed to elucidate mechanisms linking these entities and optimize therapeutic strategies.



**Abstract N°: 1308****Aquatic Dermatoses Part I: Cutaneous Conditions of Pool Water Sports**Carlos Lange Nava<sup>1</sup>, Christina Barrette<sup>1</sup>, Sheila Sharifi<sup>1</sup>, Ryan Scheinkman<sup>1</sup>, Keyvan Nouri<sup>1</sup><sup>1</sup>University of Miami Leonard M. Miller School of Medicine, Miami, United States**Introduction & Objectives:**

The first section of this two-part narrative review explores the dermatologic conditions affecting pool water athletes, including swimming, diving, and water polo, highlighting the sources of a variety of dermatoses. Given the prevalence of participation in aquatic sports, understanding these dermatological conditions, as well as their prevention and management, is crucial for athlete health and performance.

**Materials & Methods:**

A comprehensive search of the PubMed and Google Scholar databases was conducted using the search terms “swimming” or “swimming pool” or “pool water” or “recreational water” or “chlorinated water” or “chlorine exposure” and “dermatoses” or “skin disease” or “skin condition” or “cutaneous disease” or “cutaneous reaction.” For the purposes of our narrative review, articles were included if they directly discussed dermatologic manifestations secondary to pool water exposure, were of the correct article type (e.g., original articles, case reports, case studies, and review articles), and were published in peer-reviewed journals in English.

**Results:**

Our results indicate a conglomerate of bacterial, fungal, viral, and irritant-related dermatoses that may present in poolwater environments. Bacterial infections common to pool water include swimming pool granulomas and Pseudomonas-related infections. Fungal and viral infections, such as tinea pedis and molluscum contagiosum, respectively, may also pose a threat. Environmental exposure dermatoses such as sunburn and actinic keratosis highlight the impact of prolonged sun exposure on these athletes. Additionally, contact dermatoses from wearing swimming gear and exposure to chemically treated water contribute to irritant and allergic reactions. Finally, bikini bottom folliculitis and purpura gogglorum demonstrate the role of repeated friction and pressure in the onset of aquatic sport dermatoses.

**Conclusion:**

Participation in aquatic sports presents a unique exposure to a variety of dermatoses that require specialized strategies for prevention and management. Regular monitoring and maintenance of pool water hygiene are crucial to reducing bacterial and fungal infections. Preventative strategies such as the use of appropriate sun protection, moisture-wicking and UV-resistant clothing, and improved hygiene practices can mitigate many common conditions. Through these efforts, healthcare professionals can better support the dermatological well-being of aquatic sports participants. Future research should explore innovative protective solutions, personalized dermatological interventions, and the epidemiological impact of aquatic dermatoses on long-term athlete health.





Abstract N°: 1353

## When the Kidney Speaks Through the Skin: Case of Nephrogenic Dermopathy, a Rare Cutaneous Manifestation of Renal Disease

Lina Mouline<sup>1</sup>, Meriem Chaouki<sup>1</sup>, Hasnae Zerhouni<sup>1</sup>, Najoua Ammar<sup>1</sup>, Syrine Hamada<sup>1</sup>, Meriam Meziane<sup>1</sup>, Nadia Ismaili<sup>1</sup>, Laila Benzekri<sup>1</sup>

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### Introduction & Objectives:

Nephrogenic dermopathy, first described in 2000 by Camper et al., is a **fibrosing cutaneous disorder** associated with **acute or chronic renal failure**. Initially considered **purely cutaneous**, systemic involvement was later demonstrated. Its pathophysiology remains unclear, with hypotheses suggesting **T-cell activation leading to fibroblast recruitment and excessive collagen deposition**, forming **scar-like lesions**. Hemodialysis and gadolinium exposure have been implicated as triggers, though cases outside these factors challenge this hypothesis.

We report a case of **nephrogenic dermopathy in a newly diagnosed end-stage renal disease (ESRD) patient** emphasizing its **diagnostic challenges, clinical presentation, and therapeutic considerations**

### Materials & Methods:

A **46-year-old woman** with no prior medical history was hospitalized after **ahypertensive crisis**, which led to the incidental discovery of **end-stage renal disease (ESRD)**. Dermatology consultation was requested for **bilateral, symmetrical, post-traumatic scar-like lesions** on the **thighs, legs, and arms**, which had **progressed over four years**, with worsening lesions and the appearance of new ones.

Clinical examination, **dermoscopy, and skin biopsy** were performed. **Systemic involvement was assessed through thoracoabdominopelvic CT and echocardiography**. The impact of skin lesions on **quality of life** was evaluated using the Moroccan-validated **DLQI**.

### Results:

The patient exhibited **bilateral, symmetrical lesions** with two distinct patterns:

- **Lax, wrinkled plaques** on the thighs and arms.
- **Sclerotic, retractile plaques** on the legs.

Dermoscopy revealed **structureless white areas**, mirroring **underlying fibrosis**. **Histopathology confirmed nephrogenic dermopathy**, characterized by **dermal fibrosis, dense fibrous bands, and spindle-shaped fibroblasts**. **Systemic evaluation showed no evidence of visceral fibrosis**.

The **DLQI score was 12**, indicating a **significant impact on quality of life**. The **therapeutic approach** was based on **bi-annual evaluations** of lesion progression following **renal dialysis initiation**, considering reports of **spontaneous regression with treatment of the underlying renal disease**.

### Conclusion:

Nephrogenic dermopathy remains **underdiagnosed** and can **severely impact quality of life**. Its recognition is crucial, as it is **not only a dermatological entity but also a prognostic marker of renal disease**. Given its **association with increased mortality**, early diagnosis and structured follow-up are essential for **optimal patient management**. While

some cases may stabilize or regress with **renal function optimization**, **treatment remains challenging**, with therapies such as **plasmapheresis, phototherapy, cyclophosphamide, corticosteroids, and methotrexate** showing **variable efficacy**. Further research is needed to develop **standardized therapeutic strategies**.

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**Abstract N°: 1364****Parry-Romberg Syndrome: clinical insights from a case report**

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**Introduction & Objectives:**

Parry-Romberg Syndrome (PRS), also known as progressive hemifacial atrophy, is an extremely rare degenerative condition first described by Parry in 1825. This disease is characterized by progressive and severe atrophy of one side of the face, involving subcutaneous tissues, including fat, muscles, and osteo-cartilaginous structures. In this case report, we describe a new instance of PRS.

**Materials & Methods:**

NA

**Results:**

A 25-year-old man with no significant medical history and no known consanguinity presented with facial deformity accompanied by progressive arthralgia in the large joints over the past 5 years. Clinical examination revealed left-sided hemi-facial atrophy, affecting the frontal, periorbital, zygomatic, and mandibular regions, with sclerotic skin in the corresponding areas. He had atrophy of the nasal bridge and left alar, without septal deviation. He also had unilateral temporal cicatricial alopecia. The rest of the physical examination was unremarkable. Anti-nuclear antibodies were positive at 1/180. A facial CT scan revealed soft tissue atrophy on the left side, with no bone involvement. A brain MRI was normal. A diagnosis of PRS was established. Treatment with corticosteroids (0.5 mg/kg/day) and Methotrexate (15 mg/week) was initiated. The outcome was favorable, with stabilization of the atrophy. A liposuction of the face was planned for morphological restoration.

**Conclusion:**

Despite advances in understanding the disease, the etiology of PRS remains largely unknown. Disruption of cerebral lipid metabolism has often been suggested as a primary cause. Clinically, PRS is characterized by progressive hemi-facial atrophy, involving the skin, subcutaneous tissue, muscles, and sometimes osteo-cartilaginous structures. PRS is frequently associated with complications such as trigeminal neuritis, facial paresthesia, severe headaches, seizures, and ocular manifestations such as enophthalmos, eyelid atrophy, uveitis, and retinal vasculitis. Treatment of PRS can be challenging. The aim is to halt the progression of atrophy, alleviate clinical manifestations, and improve the patient's quality of life. Once the disease is stabilized, aesthetic therapies may be considered to correct facial asymmetry.

Further research is needed to better understand the underlying mechanisms of the disease and to develop more targeted interventions.





**Abstract N°: 1370****Systemic scleroderma and psoriasis: Rare association**

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**Introduction & Objectives:**

Systemic scleroderma (SSc) is an autoimmune disorder that affects both connective tissue and skin by damaging vessel architecture leading to fibrosis. Psoriasis (PsO) is a chronic, complex and multifactorial systemic inflammatory disease, mainly expressed on the skin, but can have systemic manifestations as well.

Although rare, the simultaneous presence of psoriasis and systemic scleroderma has been documented. This raises questions about the underlying immunological mechanisms.

**Materials & Methods:**

We report the case of a 29-year-old woman, with history of arthropathic psoriasis since 2021 and treated by topical steroids, injection of methotrexate 15 mg per week for 9 months and folic acid supplementation, with a cumulative dose of 1440 mg. Due to lack of incomes, methotrexate was stopped for 8 months by the patient herself.

Admitted in our department in 2024 for etiological assessment of a cutaneous calcinosis progressing for 2 months within the two coxofemoral joints associated to restriction of mouth opening, sclerodactyly extending beyond the distal interphalangeal joints, telangiectasia of the face, smooth, tense facial skin and tapered nose, with no Raynaud's syndrome.

The diagnosis of systemic scleroderma was retained with an ACR-EULAR score calculated at 11.

Immunological tests showed positive rheumatoid factor > 300 IU/ml, anticentromere antibodies, anti-Scl70 antibodies, anti PM/Scl antibodies and anti-native DNA antibodies were negative.

Chest CT showed diffuse interstitial lung disease, and soft-tissue ultrasound revealed multiple bilateral subcutaneous and fascial calcifications with no soft-tissue infiltration.

Respiratory functional exploration and transthoracic echocardiogram were normal. Our course of action was to start the patient on colchicine with monitoring and follow-up.

**Results:**

The association between arthropathic psoriasis and systemic scleroderma is uncommon, but can present interesting diagnosis and treatment challenges.

In general, patients with an autoimmune disease are at an increased risk of another autoimmune disorder, which can be ascribed to several factors, including dysregulation of immune pathways, shared environmental risk factors, genetic predisposition with specific HLA profiles, and other epigenetic factors.

Numerous hypotheses or mechanisms can be envisioned to explain a supposed link between SSc and PsO. Regarding shared genetic inheritance, associations between MHC-I molecules, such as an increased frequency of HLA-B\*08:01 in SSc and arthropathic PsO have been reported. An important plausible mechanism linking SSc and PsO is the damage to cutaneous target tissues resulting in the exposure of neo-auto-antigens to the immune system, which can trigger local adaptive immune responses.

Moreover, the two diseases share certain immunopathogenic aspects: in PsO, cutaneous inflammation is dominated by CD4+ and CD8+ T lymphocytes; in SSc, CD4+ T lymphocytes.

**Conclusion:**

Despite the fact that cases of this association have been reported, co-occurrence remains rare. This may require further investigations to establish a precise diagnosis.

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**Abstract N°: 1376****Atypical Mycobacterium: A Challenge in the Treatment of Pemphigus Vulgaris ‘The not-so-common blister’**Emily Wales<sup>\*1</sup>, Annabel Scott<sup>1</sup><sup>1</sup>Ashford and St Peter’s Hospitals NHS Foundation Trust, London, United Kingdom**Introduction & Objectives:**

Pemphigus Vulgaris (PV) is an autoimmune acantholytic blistering disease that affects the skin and mucous membranes. It is due to autoantibodies targeted against adhesion molecules desmoglein 1 and 3 in the epidermis. Rituximab is now recommended as a first-line treatment for moderate to severe PV. Here in this case report we present a case of refractory PV complicated by atypical mycobacterium infection with a patient who has developed side effects of long-term high-dose steroid use.

**Materials & Methods:**

In March 2022 a fifty-eight-year-old male presented to his general practitioner with painful blisters in his mouth and on his scalp and body. He had no past medical history of note. He was referred to dermatology with a preliminary diagnosis of PV and was initiated on 1g mycophenolate mofetil BD, prednisolone 60mg and received monthly intravenous immunoglobulins. Unfortunately, after repeated rounds of treatment, the PV was not well controlled and he was later referred to a tertiary centre for Rituximab treatment in 2023. The PV had stabilised on the Rituximab but he had been getting recurrent painful and purulent abscesses on his chest and back. His initial skin swabs grew normal skin flora and moderate pseudomonas and as per microbiology recommendation was treated with repeated courses of Ciprofloxacin. This helped in the initial phase but the abscesses would shortly return after the course had been completed. Further to this, an excision biopsy from an abscess on his back was sent to histology for analysis including mycobacterium culture. The preliminary results from the skin biopsy showed that he had been positive for acid alcohol fast bacteria (AAFB). During this patient’s 2 years of treatment, he developed; steroid-induced diabetes, steroid-induced myopathy and osteoporosis.

**Conclusion:**

Every patient with PV poses a unique challenge in terms of disease control and adverse events. Corticosteroids have been the mainstay of treatment of PV since the time of their approval in the 1950s. Prolonged and high-dose administration of steroids is often needed to control certain autoimmune diseases such as PV. Corticosteroids long-term have severe adverse effects, including hypertension, osteoporosis, atherosclerosis, peptic ulcer disease, aseptic necrosis, diabetes mellitus and increased susceptibility to infections. Recently, rituximab, a chimeric anti-CD20 monoclonal antibody, which causes B-cell depletion, has been shown to improve disease remission rates with faster tapering of steroids compared to the conventional treatment. Rituximab is now recommended as a first-line treatment for moderate to severe PV.

From this case, we have shown that the treatment of PV can be a challenging and complicated journey. Steroids are the first-line therapy for PV, but long-term administration may lead to serious adverse effects and this needs to be addressed when counselling a patient on long-term treatment.





**Abstract N°: 1404**

**Secondary Histiocytic Sarcoma with Involvement of the Skin: Case Report.**

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**Introduction & Objectives:**

Histiocytic sarcoma (HS) a rare malignant neoplasm originating from histiocytic and dendritic cell clones. It can present as a primary malignancy (primary HS) or in association with another haematological neoplasm (secondary HS), particularly those of B-cell origin. Its incidence is 0.17 cases per 1,000,000 individuals, with secondary HS accounting for 20% of cases. The neoplasm affects the lymph nodes, intestinal tract, lungs, skin, and soft tissues, with skin involvement seen in 10–15% of cases. In some cases, skin involvement is the presenting feature. Diagnosis is only possible by histopathology and is confirmed by the immunoreactivity of neoplastic cells for one or more histiocytic markers (CD163, CD68, CD4, CD11c, and lysozyme). HS has a poor prognosis, and the secondary form presents with a more adverse clinical course.

This case report describes the clinical features, diagnosis, and management of a patient with this rare neoplasm.

**Materials & Methods:**

We present a case of a secondary histiocytic sarcoma with involvement of the skin in a 36-year-old male.

**Results:**

A 36-year-old man was diagnosed with acute lymphoblastic leukaemia 2 years ago. The patient had no relevant medical history. He was treated with rituximab, hyperfractionated cyclophosphamide, vincristine, doxorubicin, and dexamethasone (R-HCVAD) therapy.

At the end of R-HCVAD phase IVB, the patient achieved complete morphological remission and was maintained on prednisone, vincristine, methotrexate, and 6-mercaptopurine therapy.

Five months later, the patient developed asymptomatic generalised dermatosis characterised by well-defined, hyperkeratotic, brown-violaceous nodules with an average diameter of 1 cm. The nodules were firm to digital palpation. The patient was admitted for an extensive diagnostic workup.

Excisional biopsy of two lesions on the right forearm and left thigh revealed histiocytic and lymphoid atypical cells spanning the papillary and reticular dermis. Immunohistochemistry showed the neoplastic cells were positive for CD163, CD68, lysozyme, and S100 (focal), with overexpression of cyclin D1. The Ki67 index was 30%, and positive resection margins of neoplasms were observed. Based on these findings, histiocytic sarcoma was diagnosed.

Subsequently, right cervical lymph node enlargement was observed. An excisional biopsy revealed similar findings to the skin biopsy.

During evaluation, pulmonary, gastrointestinal, and other extranodal involvements were ruled out. The patient was treated with ifosfamide, carboplatin, and etoposide therapy and has completed the first treatment cycle.

**Conclusion:**

Due to its rarity, diagnosing HS can be difficult, and there are no established treatment regimens. Most cases of multisite lymphoma are treated with chemotherapy. This case highlights the importance of considering secondary neoplasms in

patients with haematological neoplasms who develop new cutaneous lesions. Reporting the clinical course of this rare neoplasm is crucial, as early identification can significantly impact patient management.

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**Abstract N°: 1412****Title: Aquatic Dermatoses Part II: Cutaneous Conditions of Fresh and Salt Water Sports**

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**Introduction & Objectives:**

The second section of this two-part narrative review examines cutaneous conditions associated with exposure to fresh or saltwater, involved in sports such as scuba diving and sailing, with attention to marine organisms and environmental injuries.

**Materials & Methods:**

A comprehensive search of the PubMed and Google Scholar databases was conducted using the search terms “salt water” or “sea water” or “ocean water” or “marine water” or “fresh water” or “brackish water” and “dermatoses” or “skin disease” or “skin condition” or “cutaneous disease” or “cutaneous reaction.” For the purposes of our narrative review, articles were included if they directly discussed dermatologic manifestations secondary to pool water exposure, were of the correct article type (e.g., original articles, case reports, case studies, and review articles), and were published in peer-reviewed journals in English.

**Results:**

Our results revealed that dermatoses of fresh and saltwater sports are primarily attributed to animal-related injuries and bacterial infections rather than physical trauma and irritation when compared with poolwater dermatoses. Common bacterial infections, such as primary infections and those secondary to injury, include pathogens like *Staphylococcus*, *Streptococcus*, *Vibrio*, and *Aeromonas*. Animals are known to cause both traumatic injury and envenomation. Stingrays, weaverfish, and lionfish, among others, possess potent human toxins, while sharks, piranhas, and sea lions are common culprits for marine bite wounds. Marine invertebrates are responsible for the recognizable stings of true jellyfish, the more toxic box jellyfish, and the Portuguese Man o’ War. However, other species like corals, anemones, sponges, and sea snails may pose significant risks to unaware deep sea divers. Animal-related rashes like Swimmer’s Itch and Seabather’s Eruption may also arise. Additionally, decompression sickness, frostbite, hypothermia, and sun exposure can all present with painful rashes. Sailing, paddlesports, and board sports also put their athletes at risk for unique patterns of traumatic injury.

**Conclusion:**

Open-water athletes are exposed to a diverse set of risk factors for painful and dangerous dermatoses. Fortunately, treatment protocols for many envenomations benefit from similar first aid strategies like venom neutralization and wound debridement, while traumatic wounds can be successfully cared for using antibiotics. However, athletes would benefit from wearing protective clothing and keeping track of what animals may pose a significant risk in their area to reduce exposure. Healthcare professionals working in communities close to the water would also benefit from familiarizing themselves with endemic wildlife that may affect their patients to optimize care. Future research exploring novel sting neutralization strategies, local wildlife education, and protective clothing could greatly improve health outcomes of the open water athlete.





## Abstract N°: 1488

### cannabis arethritis

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### Introduction & Objectives:

Cannabis consumption has increased significantly in recent years, and is accompanied by well-described complications, particularly psychiatric, and less well-known ones such as peripheral arterial disease (1).

Cannabis arteritis manifests itself with clinical and radiological symptoms similar to those of Leo-Buerger's disease with a younger age of onset and very few collateral vessels. (1). It mainly affects men between 18 and 40 years old. We report a case of cannabis-arteritis.

### Materials & Methods:

Patient B.D, 25 years old, history of hypertension, smoker 18 packs/year, drug addict for 10 years (Cannabis, BZD, etc.); who presented progressive and painful bilateral necrotic ulcers of the lower limb evolving for 15 months, with distal pulses that were difficult to perceive and coldness of the extremities. Arteriovenous echo-Doppler without particularities, histology found a polymorphic infiltrate with PNN ++, CT angiography of the lower limbs: Right tibial artery and fibular artery slightly opacified distally, Left posterior tibial artery occluded at 1/3 distal; immunoassay (cryoglobulinemia, ANCA, rheumatoid factor) negative. The patient was put on corticosteroids (1 mg / kg), with local care and morphine for the pain, stopping smoking and cannabis (negative toxic assessment) and anti-platelet aggregation; which led to stabilization of the lesions, then almost total healing of the ulcers, unfortunately the trans-metatarsal amputation of the left foot was inevitable and a reduction in corticosteroids was started. Six months after his discharge, the patient presented with a recurrence of his ulcers, the cannabis dosage was negative but he had resumed smoking.

### Results:

Cannabis arteritis manifests itself with clinical and radiological symptoms similar to those of Leo-Buerger's disease with a younger age of onset and very few collateral vessels. (1). It mainly affects men between 18 and 40 years old

§ Cannabis consumption is often associated with moderate tobacco consumption which constitutes an aggravating factor (2). Cannabis has vasoconstrictor properties 40 times greater than tobacco, due to its major active ingredient delta-9-tetrahydrocannabinol (THC), whose oxidative effect has been demonstrated, causing cellular oxidation, generating free radicals responsible for endothelial lesions and consequently arteriopathy, in addition 9 THC has a Tyramine-like effect on adrenergic nerve endings (3)

§ This arteritis mainly affects the lower limbs bilaterally, in distality +++ and predominates on one limb. It manifests at the beginning by intermittent claudications which result in pain (cramps or tightening) on exertion which quickly worsen and make walking almost impossible. An onset by Raynaud's syndrome, purplish coloring of the extremities associated with small dry necroses are also described. (3).

§ Withdrawal remains the best treatment, definitive cessation of cannabis and tobacco is necessary, and results in a favorable resolution associated with anticoagulant treatment. (5). Without withdrawal, worsening of symptoms is almost inevitable and recourse to amputation is often necessary. (4)

### Conclusion:



In the face of an unusual clinical picture of arteriopathy occurring in a young subject, it would be important to systematically look for cannabis poisoning, aggravated by smoking; in order to propose early withdrawal, thus preserving the functional prognosis of the limb and avoiding amputation.

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**Abstract N°: 1490**

## **Angiokeratoma and Fabry Disease: Diagnostic Challenges in Patients with Darker Skin Tones**

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### **Introduction & Objectives**

Angiokeratomas are benign vascular skin lesions caused by damage to vascular endothelial cells and vessel dilatation in the dermis. They appear as small red/purple raised lesions and can occur in clusters. They may occur in isolation, or be a sign of systemic disease, including Fabry disease (FD), a condition that affects individuals of all ethnicities. FD is an X-linked lysosomal disorder, resulting from a mutation in the GLA gene, which leads to deficiency or reduced activity of the enzyme  $\alpha$  galactosidase A ( $\alpha$ -GalA). This leads to multi-system accumulation of glycosphingolipids, including globotriaosylceramide and globotriaosylsphingosine (lyso-Gb3). We present a unique case of multiple angiokeratomas as the presenting feature of FD in a patient with Fitzpatrick type 5 skin, highlighting the importance of recognising cutaneous manifestations of systemic disease across diverse skin tones.

### **Materials & Methods**

A 36-year-old Asian British Pakistani male with Fitzpatrick type 5 skin presented with a 2-year history of multiple asymptomatic dark papules affecting the groin areas, abdomen, knees, and palms. The lesions frequently bled with minor trauma. Additional symptoms included painful hands and feet (acroparaesthesia), anhidrosis, gastrointestinal discomfort and recurrent blurred vision which was attributed to migrainous attacks. Family history revealed similar symptoms of acroparaesthesia in his mother. A 6mm punch biopsy of a groin lesion was performed, and subsequent genetic testing was conducted given that a diagnosis of FD was suspected from the clinical presentation.

### **Results**

Histological examination showed papillary dermal proliferation with variable sized vascular spaces lined by bland endothelial cells with extension into an acanthotic epidermis. The overlying epidermis was hyperkeratotic, with elongation of rete ridges, encircling the dilated vascular spaces. Findings were consistent with angiokeratoma. Genetic testing confirmed a diagnosis of classical FD (c.154T>C AGAL variant) with low  $\alpha$ -GalA enzyme activity 0.7 nmol/hr/punch (normal range 3.8-20). Serum lyso-Gb3 levels were elevated at >100 nmol/L, further supporting the diagnosis of FD. The patient was commenced on enzyme replacement therapy (agalsidase alfa), delivered by bi-weekly intravenous infusion.

### **Conclusion**

Angiokeratoma in FD occur due to accumulation of Gb3 in dermal endothelial cells, leading to incompetence of the vessel wall and dilatation of vessels in the upper dermis. Angiokeratoma are one of the earliest clinical features of FD, affecting up to two-thirds of men and one-third of women with the condition (1,2). Recognition of the cutaneous manifestations of FD is essential to make a prompt diagnosis and initiate treatment, however manifestations of systemic disease in the skin,

such as angiokeratoma, may be more difficult to identify in patients with darker skin tones. There are few cases in the literature of angiokeratoma as the presenting feature of FD in a patient with Fitzpatrick type 5 skin. The under-representation of darker skin tones in the medical literature may contribute to delays in diagnosis. Addressing this disparity and improving awareness of cutaneous manifestations of FD across diverse skin tones is essential.

References:

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**Abstract N°: 1539**

## **Sneddon syndrome : When skin meets brain**

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### **Introduction & Objectives:**

Sneddon syndrome combines diffuse, non infiltrated livedo racemosa and recurrent strokes. The livedo typically precedes strokes by several years, making early identification crucial to prevent subsequent severe complications. Therefore, dermatologists play a key role in both the diagnosis and follow-up of this rare condition.

### **Materials & Methods:**

### **Results:**

### **Case presentation:**

A 42-year-old female patient consulted our department for marbled skin lesions, evolving for over 5 years. Her medical history includes a spontaneous miscarriage at 12 weeks of gestation in 2006, postpartum hemorrhage in 2007 and 2011, and two episodes of left-sided heaviness that resolved spontaneously in 2021 and 2023. She also reported neurological signs, including chronic, intense, intermittent headaches resistant to conventionnal analgesics, and glove- and sock-like paresthesias. Dermatological examination revealed a branched, erythematous-violaceous livedo with fine, irregular mesh of inhomogeneous caliber, partially closed in some areas and open in others, non-infiltrated, non-purpuric, non-necrotic, painless, and with a relatively diffuse and non-contiguous distribution, asymmetrically affecting both forearms, the lower outer arms, and the inner thighs, with a persistent course, nevertheless worsened by cold and orthostasis. The patient also had acrocyanosis of the forefeet and toes. In view of the above, the diagnosis of Sneddon syndrome, either isolated or associated with antiphospholipid syndrome, was suspected.

The patient underwent an immunological assessment, including testing for anti-nuclear, anti-DNA, anti-cardiolipin and anti-beta1-glycoprotein2 antibodies, as well as for a lupus-type circulating anticoagulant, all of which came back negative. The neurologists' opinion was sought, and based on clinical findings, lumbar puncture results and brain MRI data, the central origin of the symptoms was confirmed, with the vascular episodes labelled as genuine transient ischemic attacks (TIAs). Neuropsychological evaluation revealed cognitive disturbances with a dysexecutive pattern. A cardiac evaluation was also conducted as part of the systematic work-up for young-onset strokes and to screen for potential coronary or valvular pathology, which was not found in this case. Consequently, antiplatelet therapy was initiated, along with the substitution of her estrogen-progestogen contraceptive due to the thromboembolic risk.

### **Conclusion:**

Sneddon syndrome without antiphospholipid antibodies is characterized by a clinically distinctive livedo, usually diffuse, affecting not only all four limbs but also the trunk, with wide, non-infiltrated, non necrotic meshes. Strokes generally have a good short-term prognosis. Migraines and mild hypertension are common. Brain MRI typically reveals infarcts, moderate leukoencephalopathy, and cortico-subcortical atrophy. Echocardiography frequently detects valvular thickening or Libman-Sacks endocarditis. Obstetric complications, such as miscarriages and postpartum hemorrhages, are also commonly reported. Treatment primarily relies on antiplatelets, offering a favorable prognosis. However, a multidisciplinary approach, involving neurologists, cardiologists, and dermatologists, with ongoing surveillance, is essential to prevent stroke recurrence and manage long-term cognitive decline, which can lead to vascular dementia.



**Abstract N°: 1546****Multiple dermatofibromas in a previously healthy young patient: a new association with iron deficiency?**

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**Introduction & Objectives:**

Dermatofibroma is a benign fibrohistiocytic tumor of the skin, characterized by firm, slow-growing nodular lesions. While its exact etiology remains unknown, it has been proposed as a reactive process triggered by trauma, injections, or insect bites. In 0.3% of the cases, these lesions may present as multiple dermatofibromas (MD), described as more than 15 nodules. MD can appear in diffuse or clustered forms. Diffuse MD is further classified into eruptive MD, developing rapidly within months, whereas persistent MD progresses over years. MD has been associated with autoimmune diseases, viral infections such as HIV, hematologic malignancies, and immunosuppressive states. We present the case of a patient with persistent MD, without a known underlying disease, but with severe iron deficiency.

**Materials & Methods:**

A previously healthy 18-year-old woman, presented with a two-year history of more than 15 asymptomatic nodules on her upper and lower limbs and back. She denied prior triggers. Physical examination revealed well-defined, hyperpigmented, violaceous nodules, all with a positive "dimple sign". Dermoscopy showed a peripheral light brown network, central scar-like structures, and homogeneous yellow-brown areas. Additionally, one of these lesions had been previously biopsied, which resulted in a painful keloid lesion in the excision area. The biopsy confirmed the diagnosis of dermatofibroma. Laboratory tests revealed anemia and severe iron deficiency (ferritin 1.7 ng/mL, serum iron 8.08 µg/dL), positive ANA (1:160, cytoplasmic reticular pattern), but normal ENA and complement levels. Lupus and other autoimmune diseases were excluded. HIV and syphilis tests were also negative. The patient was treated with intravenous iron and intralesional corticosteroids for symptomatic relief of the keloid lesion.

**Results:**

Eruptive MD has been associated with immunosuppression, such as lupus and HIV. It has been proposed that MD reflects an immune dysfunction where down-regulatory T-cell inhibition promotes reactive fibroblastic proliferation in response to environmental stimuli.

In our patient, no autoimmune or infectious disease were identified, but severe iron deficiency was present, a finding that hasn't been linked to MD, to date. However, iron deficiency induces immunosuppression by impairing T-cell and macrophage function, which could explain the reactive fibroblastic proliferation in this case.

MD diagnosis is primarily clinical, supported by dermoscopy findings such as a peripheral light brown network and a central scar-like area. However, in cases with atypical features, a biopsy is recommended to exclude other diagnoses.

**Conclusion:**

This is the first reported case suggesting a possible correlation between persistent MD and iron deficiency. Since iron deficiency may induce transient immunosuppression, its role in MD pathogenesis warrants further investigation. Patients with MD should undergo thorough evaluations to rule out underlying autoimmune, infectious, or other immune-altering

conditions.

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**Abstract N°: 1646****Early Detection and Management of Hypocomplementemic Urticarial Vasculitis as Initial Presentation of Systemic Lupus Erythematosus**

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**Introduction & Objectives:** Urticarial vasculitis (UV) is a rare, difficult-to-treat, small-vessel leukocytoclastic vasculitis presenting with recurrent, long-lasting wheals. Hypocomplementemic urticarial vasculitis (HUV), in contrast to normocomplementemic urticarial vasculitis (NUV), is strongly associated with underlying systemic disease, especially systemic lupus erythematosus (SLE).

**Materials & Methods:** A 46-year-old woman was admitted with a 6-month history of recurrent wheals lasting longer than 24 hours. She was initially misdiagnosed as acute and, afterward, chronic urticaria. On admission, she also reported pain in the shoulders and pelvis joints, as well as swelling of fingers and ankles. Histopathological (HP) examination of skin biopsy implicated small-vessel leukocytoclastic vasculitis. Direct immunofluorescent test showed granular deposits of IgM along the basement membrane zone. The sedimentation rate was high (40 mmol/L), and complement levels, including C3 and C4, were notably low (0.4 and <0.03, respectively). Immunological analysis revealed positive speckled ANA-HEp2 (1:640), dsDNA (1:20), and anti-C1q antibodies (>400 IU/ml). The patient was thoroughly examined, minding SLE's neurological, cardiological, pulmological and renal manifestations. No other organ was affected. Based on clinical and immunological criteria, the diagnosis of SLE was made, and HUV was the first manifestation of the disease. Hydroxychloroquine was introduced into therapy, as well as, high-dose intravenous methylprednisolone followed by oral glucocorticoids, and cyclophosphamide pulse therapy. Because of the severity and extent of joint involvement, methotrexate was also initiated.

**Results:** HUV, characterized by recurrent urticarial eruptions, hypocomplementemia, and systemic involvement, adds another layer of diagnostic challenge when it precedes the typical SLE manifestations. Dermatologists play a key role in recognizing the characteristic skin lesions.

**Conclusion:** We present a case of a unique clinical entity within the spectrum of urticarial vasculitis, a hypocomplementemic urticarial vasculitis, which turned out to be the first manifestation of SLE. Early and accurate diagnosis is crucial, as it can prevent delays in treatment and improve patient outcomes. The complexity of HUV as an initial manifestation of SLE requires a multidisciplinary approach, engaging both dermatologists and immunologists to ensure comprehensive patient care.



**Abstract N°: 1904****Tufted hair folliculitis in a patient with Langerhans cell histiocytosis: an unreported association**Alessandra Rallo<sup>1, 2</sup>, Biagio Didona<sup>1</sup>, Dario Didona<sup>1</sup><sup>1</sup>Istituto Dermopatico dell'Immacolata (IDI)-IRCCS, Rare Diseases Unit, Rome, Italy<sup>2</sup>"Sapienza" University of Rome, Dermatology Unit, Department of Clinical Internal Anesthesiological and Cardiovascular Sciences,, Roma, Italy

Langerhans cell histiocytosis (LCH) is a rare neoplastic disorder resulting from the clonal proliferation of myeloid-committed hematopoietic progenitors that differentiate into CD1a+, S100+ and CD207+ (langerin) monocytes. Its incidence is 8.9/1000.000 inhabitants/per year in the pediatric population and 0.07/1000.000 inhabitants/per year in the adult population. LCH is characterized by a heterogeneous clinical presentation, ranging from localized lesions to life-threatening disseminated disease. Skin involvement occurs in up to 10% of adults, showing usually scaly, erythematous rash in intertriginous and genital areas and papules or nodules on the neck, scalp, ears, seborrheic areas of the face, and abdomen. A mutation in mitogen-activated protein kinase pathway (mostly in BRAF V600E) has been reported in up to 85% of cases and is considered as a hallmark of the disease. Treatment of LCH depends on the extent and degree of organ dysfunction, and includes surgery, targeted radiotherapy, intralesional injection of corticosteroids, and chemotherapy. The prognosis for patients without organ dysfunction is generally favorable; however, LCH can be fatal in about 20% of cases. In addition, LCH is associated with severe long-term complication in 50% of patients, including neurological and endocrine dysfunction.

Tufted hair folliculitis (THF) is a clinical manifestation of different forms of scarring alopecia and it has been reported in post-traumatic and post-surgical scarring, as well as folliculitis decalvans, dissecting cellulitis of the scalp, tinea capitis, and pemphigus vulgaris. Furthermore, drug-induced THF has been reported in patients on cyclosporine, lapatinib, and trastuzumab. The clinical hallmark of THF is represented by the presence of 5 to 30 hairs emerging from a single, dilated follicular opening, forming a "tufted" pattern also known as "doll's hair". THF pathogenesis is still unclear, but it has been reported that a dermal fibrosis-driven aggregation of adjacent follicular units, along with the retention of telogen-phase hairs across multiple hair cycles, plays a pivotal role. Here, we describe a 37 years-old man affected by LCH, diagnosed after surgical removal of a pituitary adenoma, which manifested with fatigue, visual impairment, nausea and drowsiness. At the clinical examination, the patient showed papules and plaques involving seborrheic areas of the face and hair follicles, associated with erythema and yellow crusts on face and scalp. In addition, he showed deep, inflamed retroauricular rhagades. Trichoscopy detected tufts of hair emerging from unique dilated follicles surrounded by yellowish tubular scaling, perifollicular and interfollicular scaling, erythema with dilated blood vessels, and white discoloration. Histologically, a dense lymphomonocytic infiltrate with some eosinophils and numerous CD1a+ and S100+ mononuclear elements without any sign of folliculitis was detected. Therefore, a diagnosis of THF in a patient with LCH was done. To the best of our knowledge, no cases of association between LCH and THF have been described yet.



**Abstract N°: 1933****Necrobiosis Lipoidica: A Hospital-Based Study of 12 Cases**

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**Introduction & Objectives:**

Necrobiosis lipoidica is a rare granulomatous dermatosis, frequently associated with diabetes, with a poorly understood pathophysiology. We report a series of 12 cases of necrobiosis lipoidica.

**Materials & Methods:**

This was a single-center retrospective study conducted in our Dermatology Department over a period of 22 years (2002–2024).

**Results:**

The study included 10 women and 2 men. The mean age of the patients was 41.2 years, with ages ranging from 13 to 76 years. The duration of disease progression varied from 3 months to 7 years. The clinical presentation consisted of well-circumscribed, sclerotic, erythematous plaques with a yellowish atrophic center on the anterior aspects of the legs. Unilateral lesions were observed in 4 patients. The most frequently associated conditions were type 2 diabetes (n=7), type 1 diabetes (n=3), and hypertension (n=4). The diagnosis of necrobiosis lipoidica was confirmed histologically in 9 patients. Skin biopsy revealed a dermal granulomatous inflammatory reaction with the presence of epithelioid cells and a few giant cells. This was associated with altered connective tissue stroma. Pigment deposits were found in two cases. Topical corticosteroid therapy was prescribed for 11 patients, while one patient received no treatment. The clinical course was marked by partial improvement of the lesions, and one patient showed significant improvement after two sessions of fractional CO2 laser therapy.

**Conclusion:**

Although rare, necrobiosis lipoidica should be recognized by internists and dermatologists, particularly given its frequent association with diabetes, in order to initiate effective and early management and minimize the risk of unsightly scarring.





**Abstract N°: 2035**

**Exploring the Utility of an Image-Based Flashcard Tool to Support the Development of Visual Pattern Recognition for Pathology on Darker Skin Types**

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**Introduction & Objectives:**

The visual underrepresentation of darker skin types in medical education, rooted in colonial legacies, continues to contribute to healthcare inequality. Efforts are being made to address historical Eurocentric biases in Dermatology, a specialist field reliant on visual skin interpretation. However, Western undergraduate medical curricula continue to perpetuate underdiagnosis and mismanagement of skin conditions in diverse populations.

This project sought to determine whether increased exposure to images of pathology on darker skin types enhances the cognitive associations underpinning pattern recognition, and whether adapting this pedagogical approach could better support clinicians. General Practitioners (GPs) are responsible for diagnosing and managing skin conditions despite often receiving minimal Dermatology training. We designed a novel flashcard tool juxtaposing images of the same pathology on contrasting skin types (e.g. Fitzpatrick Type II and V), which a small sample of volunteer GPs used alongside a single-image version.

Our study explored whether using this dual image-based flashcard tool could enhance clinicians' pattern recognition skills for skin conditions on darker skin types.

**Materials & Methods:**

We conducted a qualitative framework analysis on data from a focus group with three GPs who used the tool, supplemented by questionnaire responses from four additional GPs. Sub-themes were identified using the framework analysis technique.

**Results:**

From the focus group discussion data, ten main sub-themes arose from three overarching themes: *Human Experience of Pattern Recognition and Darker Skin Types*, *Utility of a Flashcard Tool for Developing Pattern Recognition Skills* and *Comparison of Flashcard Designs*. Participants found the tool intuitive and engaging, reporting improved confidence and perceived diagnostic accuracy when assessing pathology on darker skin. Additional potential applications for use in General Practice education, such as in Dermoscopy training and enhancing the design of accessible image-based clinical tools, were also identified.

**Conclusion:**

This study found that limited exposure to pathology on darker skin types contributes to diagnostic uncertainty, which influences clinical behaviours. This study underscores the need for representative and pedagogically designed skin image-based tools in clinical education and practice. The novel, dual image-based flashcard tool is an educational device that requires further investigation but shows promise for improving clinicians' ability to recognise patterns of skin pathology on darker skin types. Expanding the available number of images showing pathology on darker skin types and developing targeted educational interventions are vital steps toward decolonising medical curricula, reducing disparities, and improving patient outcomes.





**Abstract N°: 2058**

**BASCULE Syndrome: A Newly Described and Under-Reported Vasomotor Dermatositis**

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**Title:** BASCULE Syndrome: A Newly Described and Under-Reported Vasomotor Dermatositis

**Introduction & Objectives:**

BASCULE Syndrome is an under-reported vasomotor dermatosis characterised by *Bier Anaemic spots*, *Cyanosis* and an *Urticaria-Like Eruption*. This is a newly described and relatively rare entity with debilitating symptoms. Majority of those diagnosed are between the ages of 12 and 19, with a predominance amongst females. This condition presents with a rapid cyanosis that may evolve to cover the entire surface of the lower limbs. Within two to five minutes, blanched macules and patches resembling Bier anaemic spots appear amongst the cyanosis. This is followed by a reperfusion phase where urticarial-like salmon-red macules and patches arise within the larger blanched spots. Symptoms include heaviness, lower limb aching and pruritus. The sequence of dermatological changes are observed within minutes of standing erect, intensify with time and resolve almost immediately when lying down or with leg elevation. The aetiology and pathophysiology of this condition remains unclear with autonomic dysfunction suggested as a possible underlying mechanism.

**Materials & Methods:**

We describe four young patients diagnosed with BASCULE syndrome.

**Results:**

Our patients were all under 20 years of age at the time of diagnosis and presented with a rapid onset of lower limb cyanosis and pruritus upon standing upright. Within minutes, blanched macules and patches appeared in the cyanotic areas followed by urticarial-like pruritic salmon-coloured patches. One patient showed improvement following an oral brompheniramine maleate/ phenylephrine cough syrup and hence the role of antihistamines should be investigated as a treatment strategy.

**Conclusion:**

We describe four cases diagnosed with BASCULE syndrome, a rare and underdiagnosed entity. Clinical understanding of this vasomotor dermatosis has been impeded by its relatively recent discovery, limited understanding of the condition's aetiology and pathophysiology which in turn has limited the ability of clinicians to diagnose and treat this unusual condition.







**Abstract N°: 2060**

**Confused patient with dysphagia, psychosis, neuropathy, weight loss and a progressing widespread rash unresponsive to steroids - an uncommon case of pellagra**

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**Introduction & Objectives:**

Pellagra is a multi-system disorder caused by a deficiency of Niacin (Vitamin B3). Niacin deficiency affects tissues with high energy demands or cell turnover rate such as skin, bowel and brain. Pellagra is characterized by dermatitis, diarrhea, dementia. Though rare in developed countries due to food fortification, it still occurs in vulnerable populations.

**Materials & Methods:**

We present a patient presented with a scaly erythematous painful rash affecting the face and neck unresponsive to steroids, which was later diagnosed with pellagra. Due to its rarity, Pellagra can often be misdiagnosed and can cause death if left untreated within 4-5 years. There should be a low threshold for considering pellagra as a potential diagnosis in patients presenting with a non-specific rash and cognitive impairment, weight loss or gastro enteric symptoms.

**Results:**

A 50-year-old woman presented with a few months' history of scaly, erythematous and painful rash affecting her face, neck, torso and limbs. She has a history of epilepsy with partial temporal lobectomy, bipolar affective disorder, anxiety, depression, eating disorder and psoriasis. The rash initially looked suspicious for dermatomyositis in a shawl distribution. Skin biopsy was done which highlighted a subtle interface dermatitis without significant dermal inflammation. She also had extensive investigations which ruled out autoimmune, malignancy causes and myositis.

She was prescribed topical mometasone ointment for the body, betamethasone cream for the face, and given 40 mg prednisolone. Her rash however, continued to spread to the dorsum of her feet and she developed oromucosal ulceration, the rash around her neck appeared in keeping with "Casal's necklace". Additionally, she experienced progressive generalized weakness, particularly in her hips, along with new diarrhoea and dysphagia, which necessitated her admission as an inpatient.

It was noted that she was on a a ketogenic regimen for several years on her own accord with hopes to improve her seizure symptoms. This history along with her symptoms and the characteristic "Casal's necklace" rash, pellagra became the primary differential. She was started on nicotinamide 500mg with intravenous infusion of a collection of vitamin B and B12 injections, prednisolone was slowly weaned off and she was reviewed by the dietician. Within a month of treatment, her skin rash resolved completely. Over the following 6 months, she noticed significant improvements in peripheral neuropathy, dysphagia and mental health.

**Conclusion:**

Pellagra presents as dermatitis of sun-exposed areas of skin, beginning as an erythema with pruritus that may lead to vesiculation but more frequently becomes chronic, rough, scaly, and hard with crusts as the result of haemorrhage. Our patient had the characteristic band of dermatitis encircling the neck, called Casal's necklace Gastrointestinal tract may be



involved with glossitis, stomatitis, gastroenteritis with diarrhoea, which is profuse, watery, and sometimes bloody. Our patient had oromucosal ulceration and diarrhoea. Neurocognitive symptoms may present as anxiety, depression, tremor, and reduced or absent tendon reflexes; memory loss with pellagrous encephalopathy may occur in severe cases. Our patient had progressive generalised weakness and peripheral neuropathy. If the deficiency continues, pellagra can lead to death.

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**Abstract N°: 2157****When Skin Speaks: A Severe Case of Paraneoplastic Bullous Pemphigoid**Amy Long<sup>\*1</sup>, Clare Harnett<sup>1</sup><sup>1</sup>Beaumont Hospital, Dublin, Ireland**Introduction & Objectives:**

Herein we present a rare case of paraneoplastic bullous pemphigoid in the context of renal cell carcinoma, clear cell type. To date, only one other case report has documented this association.

**Materials & Methods:**

A 71-year-old male presented with a three-week history of a progressive blistering rash affecting his face, torso, arms, upper legs, and oral and genital mucosae. He was acutely reviewed by dermatology during admission following a radical nephrectomy for a renal tumour, later confirmed as pT3a renal cell carcinoma, clear cell type. His medical history included gout and hypertension, and his regular medications included bisoprolol, perindopril, lercanidipine, and allopurinol. He was systemically well, with no preceding illness or history of herpes labialis. On examination, he had a severe blistering rash with extensive facial involvement, characterised by vesicular lesions with crusting. Scattered bullae were present on his torso and limbs, along with ulcerations on the vermillion lips, buccal mucosa, and glans penis. A punch biopsy of the left thigh revealed subepidermal vesicular dermatitis with predominant eosinophils. Direct immunofluorescence showed linear IgG deposition along the dermo-epidermal junction.

**Results:**

The patient was diagnosed with paraneoplastic bullous pemphigoid and was successfully treated with topical betamethasone valerate for the face, clobetasol propionate for the body, prednisolone mouth rinse, and doxycycline 200 mg once daily.

**Conclusion:**

This case highlights the rare but significant association between renal cell carcinoma and paraneoplastic bullous pemphigoid. Early recognition and appropriate dermatologic intervention are crucial for effective management and improved patient outcomes.





**Abstract N°: 2165**

## **Coexistence of Discoid Lupus Erythematosus and Lupus Panniculitis: A Case Report**

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### **Introduction & Objectives:**

Lupus erythematosus panniculitis (LEP) is a recognized but uncommon manifestation of cutaneous lupus erythematosus. While frequently occurring as an isolated condition, it can also coexist with discoid lupus erythematosus (DLE). LEP is characterized by subcutaneous inflammatory nodules that may lead to profound lipoatrophy, causing cosmetic disfigurement. Although benign, LEP can cause significant morbidity from scarring and atrophy. We report a case of both DLE and LEP, highlighting the need for early recognition and treatment.

### **Materials & Methods:**

A 61-year-old female with no significant medical history presented with progressive facial and scalp lesions over six years, with worsening and severe lipoatrophy in the last two years.

The clinical examination revealed erythematous-squamous plaques on the nasal bridge and left cheek, lipoatrophic erythematous plaques on the right cheek, and an erythematous plaque with cicatricial alopecia at the vertex. The coexistence of atrophic and non-atrophic lesions suggested both DLE and LEP.

Dermoscopy reinforced this diagnosis, revealing DLE features in erythematous-squamous plaques and LEP characteristics in lipoatrophic areas.

Skin biopsies from a non-severely atrophic lesion and a severely atrophic lesion revealed histological features consistent with DLE and LEP, respectively, with a positive lupus band test in both. Immunological analysis showed positive anti-DNA and ANA antibodies, while IgG4 levels were within the normal range, further supporting this diagnosis.

The patient was started on topical calcineurin inhibitors (TCIs) and systemic corticosteroids (0.5 mg/kg/day). Given her contraindication to antimalarial therapy (visual field impairment), methotrexate (10 mg/week) was initiated. The lesions improved significantly during treatment.

### **Results:**

LEP is a known but underrecognized complication of cutaneous lupus, particularly in those without systemic involvement. While it is commonly an isolated entity, its coexistence with DLE is well-documented. Histology is crucial for diagnosis, though sampling bias may limit its ability to confirm LEP in all cases. Persistent lipoatrophy on the shoulders and upper arms is highly characteristic, enabling a retrospective diagnosis.

Although generally benign, LEP can result in significant morbidity due to deformities and atrophy. In our patient, the absence of IgG4 positivity helped exclude a fibrosing disorder that could have mimicked atrophic lupus. Despite the lack of a characteristic lesion distribution, the clinical features, along with histological findings, guided the diagnosis of LEP. Early recognition and intervention are critical to prevent progressive lipoatrophy.

Antimalarials remain the cornerstone of LEP treatment, with systemic corticosteroids for severe or refractory, and flare-ups. TCIs help manage localized forms while minimizing corticosteroid side effects. Low-dose methotrexate is a second-line option when antimalarials are contraindicated. Early recognition and intervention, as seen in our patient, help prevent further lipoatrophy.

**Conclusion:**

This case highlights the importance of recognizing DLE and LEP coexistence in patients with progressive cutaneous lupus. Early diagnosis, proper immunosuppressive therapy and close monitoring are key to preventing irreversible damage. More research is needed to understand the underlying mechanisms and improve treatment strategies.

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**Abstract N°: 2168****Panniculitis Ossificans of Chronic Venous Insufficiency: Ultrasonic and Histopathologic features**Manisha Siriwardene\*<sup>1</sup>, Kurosh Parsi<sup>1</sup><sup>1</sup>St Vincent's Hospital Sydney, Dermatology, Darlinghurst, Australia**Title:** Panniculitis Ossificans of Chronic Venous Insufficiency: Ultrasonic and Histopathologic features.**Introduction & Objectives:**

We describe a 49-year-old female presenting with progressive lower limb aches, pains and cramping on the background of advanced bilateral lower limb chronic venous insufficiency (CVI) and lymphoedema. Symptoms were significant in this patient. On examination, she presented with bilateral lower limb oedema and unilateral lipodermatosclerosis. The anterior surface of the left gaiter region and medial calf were extremely indurated. The aim of the presentation is to describe the ultrasonic and histopathologic features that would differentiate between calcification and ossification.

**Materials & Methods:**

An initial punch biopsy demonstrated subcutaneous calcification. Ultrasound review demonstrated hyperechoic lesions with intense shadow artefact excessive for calcification and more consistent with ossification. A deep incisional biopsy was completed with difficulty. Ossified tissue was extremely challenging to remove. Underlying chronic venous disease was treated with endovenous laser ablation under general anaesthetic.

**Results:**

Histopathology demonstrated heterotopic ossification associated with fat necrosis confirming the clinical and ultrasonic suspected diagnosis. Investigations including blood tests were only remarkable for a microcytic anaemia (Hb 87, MCV 77, Ferritin 9, Iron 2, Transferrin 3.6, Transferrin Saturation 2). ANA, ENA, ANCA and dsDNA returned negative deeming CREST syndrome less likely. Bone scan demonstrated no mild to moderate osteoblastic tracer activity correlating to extensive dense calcification throughout the subcutaneous tissues. Treatment of the underlying CVI helped with symptoms. Further planned management includes endoscopy, colonoscopy, MRI of lower limbs to investigate for myositis ossificans, endocrine, immunology and plastics review.

**Conclusion:**

Heterotopic ossification is a rare phenomenon whereby bone formation occurs in extraskeletal tissue including the subcutaneous fat and muscle. B-mode ultrasound finding of hyperechoic lesions with intense shadow artifact in association with subcutaneous indurated plaques should prompt investigations for panniculitis ossificans. Treatment of the underlying CVI will help with the overall management.



**Abstract N°: 2287****Cutaneous Sarcoidosis Resistant to Therapy**Sofia Tymchuk\*<sup>1</sup>, Irina Smirnova<sup>1</sup><sup>1</sup>Saint-Petersburg State University, Saint-Petersburg, Russian Federation

**Introduction & Objectives:** Sarcoidosis is a multisystem disease that often manifests as cutaneous eruptions, making diagnosis challenging due to the diversity of clinical forms and symptoms. Cutaneous involvement occurs in approximately 25% of cases. This case illustrates the difficulties in selecting effective therapy for a patient with plaque-type cutaneous sarcoidosis and interstitial lung disease.

**Materials & Methods:** The study includes the collection, analysis, and presentation of data such as complaints, medical and life history, findings from physical, laboratory, and instrumental examinations, treatment history, and patient follow-up. A literature review was conducted using original research and review articles from databases such as ScienceDirect, Scopus, PubMed, Elsevier, and others over the past five years.

**Results:** A 37-year-old male patient presented with widespread cutaneous eruptions on the upper extremities, neck, and trunk, accompanied by desquamation. The eruptions first appeared in 2017 after sun exposure, initially affecting the postauricular fold and upper extremities. In 2018, the condition became disseminated, leading the patient to seek medical attention. Treatment with methylprednisolone aceponate for two weeks was ineffective. In 2019, histological examination confirmed the diagnosis of "cutaneous sarcoidosis." At the time of consultation in 2019, the cutaneous process involved the scalp, upper third of the trunk, and extremities. Diascopy revealed a positive "apple jelly" sign, and dermoscopy showed a homogeneous yellow-orange coloration with branching vessels. Chest CT revealed isolated pulmonary nodules in both lungs, confirming interstitial lung disease. Treatment of cutaneous manifestations with hydroxychloroquine was ineffective, prompting the initiation of methotrexate, which showed a positive response. However, after clinical improvement and during the COVID-19 pandemic, treatment was discontinued. In 2024, a relapse of cutaneous eruptions was observed, but chest CT showed no new pulmonary changes. A repeated course of methotrexate at a dose of 10 mg subcutaneously once a week led to hepatotoxicity, necessitating discontinuation of treatment.

**Conclusion:** Difficulties in therapeutic control of cutaneous manifestations, the lack of response to standard treatments such as hydroxychloroquine, and the development of adverse effects from methotrexate highlight the need for further research into effective treatment strategies for cutaneous sarcoidosis and the search for alternative therapeutic approaches.



**Abstract N°: 2417****Polydactylous Onychopapillomas as a BAP1 Tumor Predisposition Syndrome Predictor: A Case Presentation**Lucy Wang<sup>\*1</sup>, Hasret Gunduz<sup>2</sup>, June Moon<sup>3</sup>, Kenneth Shulman<sup>3</sup>, Mehmet Fatih Atak<sup>3</sup>, Banu Farabi<sup>3</sup><sup>1</sup>New York Medical College, Valhalla, United States<sup>2</sup>Northwell Health, Manhasset, United States<sup>3</sup>NYC Health + Hospital/Metropolitan, NYC, United States**Introduction & Objectives:**

BRCA1-associated protein (BAP1) tumor predisposition syndrome (BAP1-TPDS) is an autosomal dominant cancer syndrome associated with an increased risk of cutaneous melanoma, uveal melanoma, malignant mesothelioma, and renal cell carcinoma. BAP1-inactivated melanocytic tumors (BIMTs) are often the first clinical indication of BAP1-TPDS but are often overlooked due to their inconspicuous appearance, highlighting the need for additional diagnostic markers. Onychopapilloma is a benign nail tumor most commonly presenting with longitudinal erythronychia and distal subungual hyperkeratosis. Although onychopapilloma typically affects a single digit, the presence of polydactylous involvement has recently been associated with BAP1-TPDS and may help identify individuals at increased risk of BAP1-TPDS (Lebensohn A, et al. JAMA Dermatol. 2024).

**Materials & Methods:**

A 43-year-old male with multiple congenital nevi presented with a verrucous light to dark-brown papule on the lower back and a dome-shaped, skin-colored papule with asymmetric black pigment on his right shoulder. Additionally, the patient exhibited longitudinal leukonychia, V-shaped onycholysis, and distal fissuring on multiple fingernails. Dermatoscopy revealed localized subungual hyperkeratotic papules, consistent with onychopapilloma, affecting several fingernails and the left first digit toenail. The patient reported nail abnormalities since childhood. A shave biopsy of the lower back lesion revealed BAP1-deficient intradermal melanocytic nevus. Immunohistochemistry (IHC) showed BAP1 expression in small nevus cells but deficient or absent BAP1 expression in the larger epithelioid melanocytes. A combined Ki67 and Melan-A stain showed minimal proliferative activity (~ 2%). A biopsy of the right shoulder papule confirmed a conventional nevus and a Spitz nevus, the latter failing to express BAP1 on IHC. The lower back and right shoulder lesions were completely excised. The patient was referred to the Genetics Department and molecular genetic testing was positive for a germline BAP1 mutation. He was diagnosed with BAP1-TPDS and counseled to undergo regular surveillance for malignancies associated with the syndrome.

**Results:**

Our patient with BAP1-TPDS and polydactylous onychopapilloma since childhood underscores the important consideration of BAP1-TPDS in patients presenting with nail abnormalities affecting multiple digits, especially when accompanied by multiple nevi or atypical melanocytic lesions.

**Conclusion:**

Recognition of polydactylous onychopapilloma as a potential cutaneous marker of BAP1-TPDS may enable earlier detection of BAP1-TPDS and subsequent cancer surveillance in affected individuals and their families.





**Abstract N°: 2429****Leukocytoclastic Vasculitis Secondary to Infection: Case Report**Diana Verónica Romero Escamilla<sup>1</sup><sup>1</sup>ISSSTE, Ciudad de México, Mexico**Leukocytoclastic Vasculitis Secondary to Infection: Case Report**

**Introduction & Objectives:** Leukocytoclastic vasculitis (LCV) refers to a histopathological description of a common small-vessel vasculitis affecting arterioles, capillaries, and postcapillary venules. The inflammatory infiltrate consists of neutrophils with fibrinoid necrosis and nuclear fragmentation ("leukocytoclasia"). The main clinical manifestation is palpable purpura, and diagnosis relies on histopathological examination. Various drugs, infections, and malignancies can trigger LCV. Among systemic diseases, LCV is often associated with ANCA-associated vasculitis, connective tissue diseases, cryoglobulinemic vasculitis, IgA vasculitis (formerly Henoch-Schönlein purpura), and hypocomplementemic urticarial vasculitis. Extensive evaluation is usually required to determine whether the process is confined to the skin or indicative of systemic vasculitis.

**Materials & Methods:** A female patient with a history of dry necrosis of the second right toe due to thrombosis, chronic kidney disease secondary to diabetic nephropathy requiring hemodialysis, chronic heart failure (NYHA II, AHA C, LVEF 72%), moderate mitral and aortic stenosis, small-vessel disease, type 2 diabetes, hypertension, and overweight, presented with cough, unquantified fever, and dyspnea for 10 days. She was initially treated with an unspecified antibiotic without improvement. She later developed petechial lesions on the lower limbs. During a scheduled hemodialysis session, she desaturated to 70% and was referred to the emergency department. A chest CT revealed left apical consolidation. Dermatology evaluation showed a disseminated, bilateral, symmetrical dermatosis affecting all segments of the lower limbs, predominantly legs and feet. The lesions consisted of multiple millimetric petechiae, some coalescing into irregularly shaped purpuric plaques with well-defined, erythematous-violaceous borders, a smooth surface, raised texture, and non-blanching on digital pressure. A biopsy was performed.

**Results:** The patient was started on Cefepime, methylprednisolone pulses for three days, and intravenous diphenhydramine. After completing antibiotic therapy, respiratory and cutaneous symptoms improved. Biopsy results revealed polymorphonuclear inflammation in the small-vessel wall with neutrophil fragmentation, fibrinoid necrosis, and erythrocyte extravasation. Another image showed a blood vessel with a permeable lumen and intact endothelium, but with polymorphonuclear inflammation and fibrinoid necrosis in the vessel wall, confirming leukocytoclastic vasculitis.

**Conclusion:** Leukocytoclastic vasculitis is an autoimmune disorder affecting small vessels, leading to inflammation, destruction, and necrosis. It is frequently underdiagnosed. Its etiology is multifactorial, with a complex pathophysiology, and immunomodulators are crucial for treatment.