EA DV CONGRESS
AMSTERDAM 25-28 SEPTEMBER 2024
EUROPEAN ACADEMY OF DERMATOLOGY & VENEREOLOGY

Abstract N°: 1069

Comparative Analysis of Vitamin D3 Levels and The Impact of Vitamin D3 Supplementation on Disease Severity in Chronic Spontaneous Urticaria Patients: A Randomized Controlled Trial

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Introduction & Objectives: Studies revealed patients with chronic spontaneous urticaria (CSU) had lower vitamin D3 levels. Some studies indicated that high-dose vitamin D3 supplementation for 4-12 weeks may reduce disease activity. However, the results of different trials were inconsistent and there is a paucity of well-designed clinical trials especially from the Indian subcontinent. This necessitates a well-designed randomized controlled trial (RCT) to confirm vitamin D3 supplementation outcomes. The study aimed to compare vitamin D3 levels in CSU patients with healthy controls and assess the correlation between disease severity and vitamin D deficiency. It also evaluated the effect of vitamin D3 supplementation on disease control in CSU patients with low vitamin D3 levels.

Materials & Methods: The study initially recruited 110 CSU patients and 80 healthy controls. Later, 62 CSU patients with low baseline serum vitamin (less than 30 ng/ml) D3 were randomized into two groups using computer-generated random numbers. One group (Group A) received Tab levocetirizine 5 mg once daily, while the other (Group B) received oral Cholecalciferol 60 K weekly for eight weeks in addition to Tab levocetirizine 5 mg once daily. Disease severity and control were measured by USA7 and UCT scores at the baseline, week 4, and week 8. (Figure 1,Figure 2, Figure 3, Figure 4)

Results: The mean serum vitamin D level among CSU patients was 15.83 ± 3.35 ng/ml and that among healthy controls was 32.01 ± 2.15 ng/ml. Compared to healthy controls CSU patients had significantly lower serum vitamin D levels. Among the CSU patients with low baseline serum vitamin D3 who were randomized into 2 groups, both had significant improvement at 8 weeks. However, the group receiving vitamin D supplementation (Group B) had a notably higher decrease in UAS7 (mean UAS7 3.48 ±2.9) at 2 months compared to the group (Group A) receiving only levocetirizine (mean UAS7 21.22 ±8.78), with the differences being statistically significant. CSU patients in group B also had greater disease control in terms of a higher increase in UCT scores (mean UCT 13.80 ±1.89) than group A (mean UCT 6.87 ±2.97) at the end of 8 weeks.

Conclusion: In conclusion, we recommend monitoring vitamin D levels at baseline as supplementation can improve hive symptoms.

Figure 1 (Blue-group A, Red-group B)

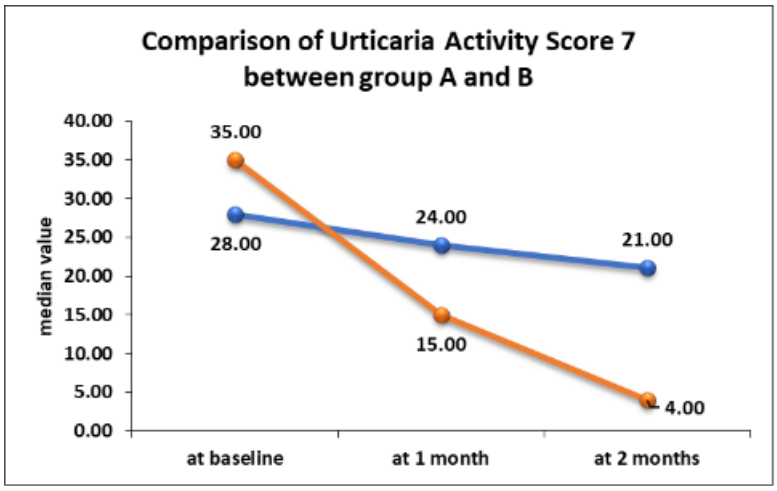


Figure 2

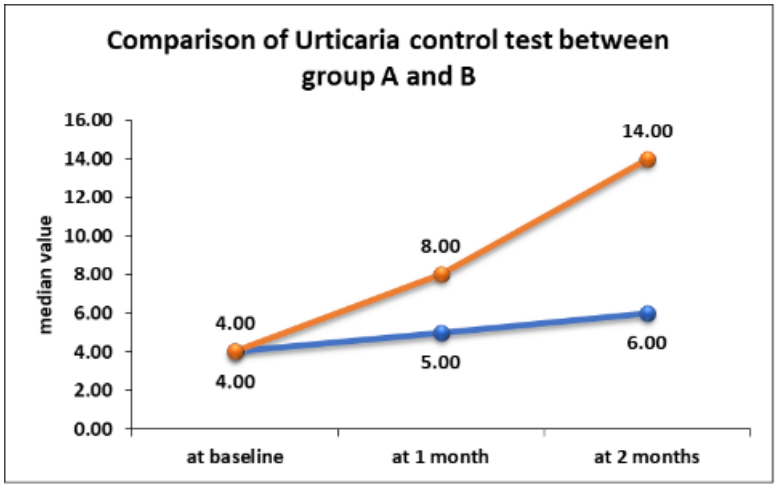


Figure 3

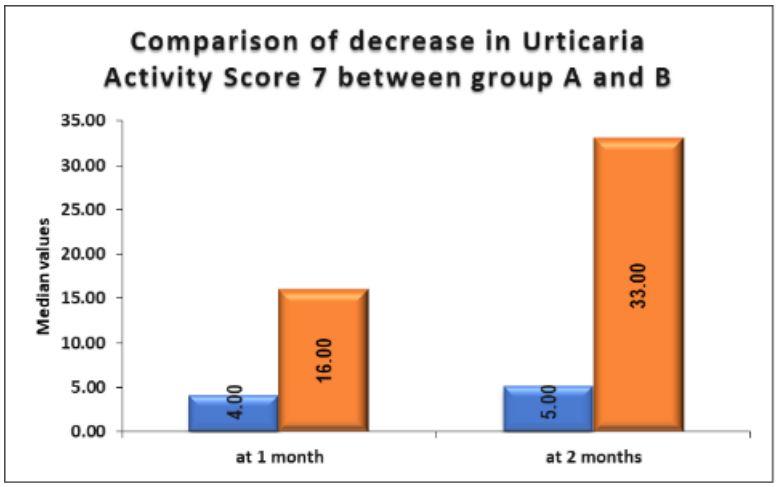
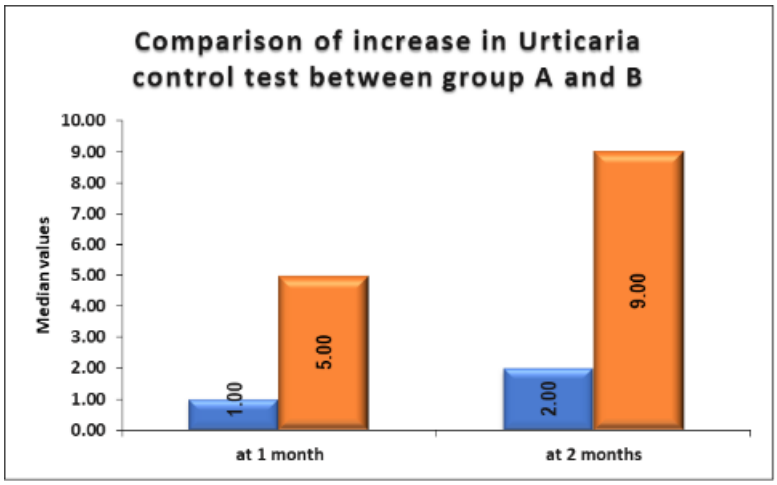


Figure 4


Comparison of increase in Urticaria control test between group A and B
Median values
10.00
9.00
8.00
7.00
6.00
5.00
4.00
3.00
2.00
1.00
0.00
1.00
5.00
2.00
9.00
at 1 month
at 2 months

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Abstract N°: 1767

Assessment of Serum Level of IL-6, D-dimer and CRP in Patients with Urticaria

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Introduction & Objectives:

Urticaria is an allergic disorder. Many studies have highlighted the role of activation of coagulation factors, eosinophils and tissue factor pathway with generation of thrombin potentially contributing to an increased vascular permeability.

The aim of this work was to assess the serum level of interleukin 6(IL-6), D-dimer and c-reactive protein (CRP), complete blood picture (CBC), prothrombin time (PT), partial prothrombin time (PTT), clotting time (CT) and International Normalized ratio (INR) in patients with urticaria as a cheap markers for coagulation.

Materials & Methods:

This case control diagnostic interventional study was carried out on 100 patients (50 patients with acute and 50 patients with chronic urticaria), and 100 normal healthy sex and age matched controls. IL-6, D-dimer ,CRP (were measured by an enzyme-linked immunosorbent assay (ELISA) ,CBC by using (automated haematology analyser) ,CT (by test tube method), PT (by an automated instrument) ,PTT (by citrated plasma, the addition of a platelet substitute , factor 12 activator and CACL2) and INR (=patient PT/ control PT).

Results:

This study showed significant high levels of CRP, IL-6, D-dimer, PT and PTT in both acute & chronic Urticaria and they were significantly higher in acute group comparing chronic groups (P<0.001 ,<0.001,<0.001,<0.006 and <0.042 respectively)

Conclusion:

IL-6, CRP, ESR, D-dimer ,PT,PTT and CT could serve as available cheap biomarkers for detection of the coagulation defect that can be encountered in urticaria and this is supposed to be responsible for expected prognosis and marker for the progression of the treatment response .Ther also can serve as a reliable markers in chronic urticaria to indicate disease chronicity and activity .

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Abstract N°: 2028

Direct Current Electric Field Induce Dendrites Contact of Epidermal Langerhans Cells

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Introduction & Objectives: Langerhans cells (LCs), found in the outermost skin layer, play a crucial role in immune activation and homeostasis. Electric fields can guide immune cell migration, including macrophages, T cells, and B cells. Researchers aim to manipulate immune cell biology using external electric fields. However, understanding LCs' responses to electric fields remains limited, and it's unclear if these cells can modulate their immunological functions. We studied LC behavioral changes after direct current exposure, identifying key proteins involved. Our goal is to develop novel approaches using electric fields to modulate skin immunity.

Materials & Methods: We employed CD207-EGFP mice for live-cell fluorescence tracking of LCs. A continuous 5V direct current electric field was applied to skin of mice ear for 30 minutes. We cultured the skin for 24 hours and performed 3D immunofluorescence imaging lasting 3 hours using laser confocal microscopy to observe and analysis LCs behavior. Additionally, single-cell RNA sequencing of skin detected gene expression changes in LCs following electric stimulation. By utilizing differential gene enrichment analysis and intercellular communication analysis on the data, we aimed to infer key genes potentially associated with LC-LC contact.

Results: Electrically stimulated LCs exhibited significantly higher movement speed compared to the control group in the XY plane (6.16μm/h vs. 1.85μm/h). We also observed dendrites contact among LCs, where two LCs simultaneously extend dendrites toward each other and establish contact, lasting 1-2 hours. The contact rate of LCs in the epidermis after electrical stimulation was significantly higher than control (21.56%-41.36% vs. 5.48%-16.79%). LCs cell division was also captured, providing evidence for LC self-renewal in epidermis. Single-cell RNA sequencing analysis revealed LC cluster from skin samples. Enrichment analysis using GO and KEGG databases highlighted biological pathways related to endocytosis, actin cytoskeleton, and Ras signaling. CellChat analysis suggested that the Sema7a pathway, also named axon guidance pathway, are potential key proteins associated with LC migration and dendrites contact. One of the ligands for Sema7a is Itgb1, which is also one of the differential genes in LCs.

Conclusion: In electric field, LCs exhibits enhanced patrolling within the epidermis, leading to a significant increase in dendrites contact among LC. After skin electrical stimulation, the migration speed of LC in the epidermis markedly increases, manifested by an accelerated movement in the XY-plane. Concurrently, dendrites contact occur during LC patrolling, and the contact rate in the electrically stimulated group is significantly higher than control group, indicating that the electric field promotes interactions and information transfer among LC. Single-cell RNA sequencing of mice skin samples subjected to the electric field reveals that LCs show significant responsiveness in antigen presenting cells. In LC, differential genes are predominantly enriched in endocytosis, cell skeleton, and the Ras pathway. CellChat intercellular communication analysis of LC indicates Sema7a-Itgb1 showing the highest correlation with dendrite elongation and LC-LC contact.

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Abstract N°: 2271

Enhancing allergy patients' health literacy and quality of life through a smartphone application: a randomized controlled trial

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Introduction & Objectives: Increasing instances of allergies and intolerances to medications necessitate personalized pharmacotherapy approaches. A mobile application to help identify appropriate medications by checking drug compatibility and suggesting alternatives could be a new solution to improve health literacy and patient safety.

The aim of the study was to investigate the additional benefits of a smartphone app for allergy patients compared to standard doctor's consultations ("treatment-as-usual"). Questionnaires were used to determine whether the users' health literacy and quality of life had improved over the course of the study period.

Materials & Methods: The prospective randomized controlled trial includes data collection at study entry and after six months using validated and self-developed questionnaires. Patients are eligible for the study if they are of legal age, have certain allergies and/or intolerances to medication, and own a smartphone. All patients were recruited from the allergy departments of two German hospitals. At the beginning of the study, participants are randomized into two groups: the intervention group, which uses the mobile application plus receives standard medical counseling, and the control group, which receives standard counseling alone. The study assesses changes in health literacy, quality of life, and medication management over a 180-day period using validated tools and self-reported patient outcomes.

Results: So far, 141 participants at two study centers have been included in the study. Related to the app, initial evaluations showed that 72,1% of the participants in the intervention group felt more confident in their choice of medication after using the app. In addition, 88,4% of the patients reported that the smartphone application was easy to use, 76,0% also stated that the app could be easily integrated into everyday life. Furthermore, 83,7% of the participants described the app as a helpful tool, and 88,4% indicated that they would recommend it to other patients with similar symptoms. In order to draw final conclusions about the differences in terms of health literacy and quality of life, further evaluations are required once patient recruitment has been completed.

Conclusion: A smartphone app could be a useful addition to the regular medical approach for allergy patients. Especially these days, with almost limitless possibilities for obtaining information, it is more important than ever to provide patients with evidence-based knowledge in an easily understandable form. At this stage of the study, it appears that patients may be willing to use such digital tools as daily support for their allergies and intolerances. The final results of the study will show whether patient-related health literacy and quality of life can be improved through the use of the app.

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Abstract N°: 2574

Efficacy of stapokibart in adults with moderate-to-severe atopic dermatitis and comorbid allergic disease: subgroup analysis from a phase 3 trial

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Introduction & Objectives: Atopic dermatitis (AD) is a chronic inflammatory skin disease that is often associated with atopic comorbidities, with allergic rhinitis (AR) being the most prevalent. Stapokibart, a novel humanized monoclonal antibody against the interleukin-4 receptor alpha (IL-4Rα) subunit, blocks the signal of two essential drivers of type 2-mediated inflammation in various diseases including AD. In a phase 3 trial, stapokibart effectively improved clinical signs and symptoms of AD versus placebo. This subgroup analysis aimed to determine whether the comorbidity of allergic disease affects the efficacy of stapokibart in adults with moderate-to-severe AD.

Materials & Methods: In this phase 3 trial (NCT05265923), patients were randomized 1:1 to subcutaneous stapokibart 300 mg or placebo every two weeks for 16 weeks. Post-hoc endpoints included proportions of patients achieving an Eczema Area and Severity Index ≥75% improvement (EASI-75), an Investigator's Global Assessment (IGA) score 0/1 with a ≥2-point reduction from baseline, and a 4-point reduction in peak pruritus numerical rating scale (PP-NRS) score in subgroups with and without comorbid allergic disease.

Results: At baseline, 109 out of 251 and 116 out of 249 patients in the stapokibart and placebo group reported a history of allergic disease, respectively, of whom, 90 and 101 had a history of AR. Greater proportions of patients with comorbid allergic disease receiving stapokibart versus placebo achieved EASI-75 (73.4% vs. 26.7%, Figure 1A), IGA 0/1 (53.2% vs. 18.1%, Figure 1B), and PP-NRS≥4 reduction (42.2% vs. 12.1%, Figure 1C) at week 16 (all p<0.0001). Specifically in the subgroup with comorbid AR, more stapokibart-treated patients obtained EASI-75 (74.4% vs. 22.8%, Figure 1A), IGA 0/1(51.1% vs. 15.8%, Figure 1B), and PP-NRS≥4 reduction (42.2% vs. 13.9%, Figure 1C) vs. placebo-treated patients at week 16 (all p<0.0001). Patients without allergic disease showed similar results (stapokibart vs. placebo at week 16, EASI-75: 62.0% vs. 25.0% p<0.0001; IGA0/1: 37.3% vs. 14.4% p<0.0001; PP-NRS≥4 reduction: 31.0% vs. 11.4%, p=0.002, Figure 1).

Conclusion: Stapokibart improves AD signs and symptoms in adults with moderate-to-severe AD regardless of the comorbid status of allergic disease.

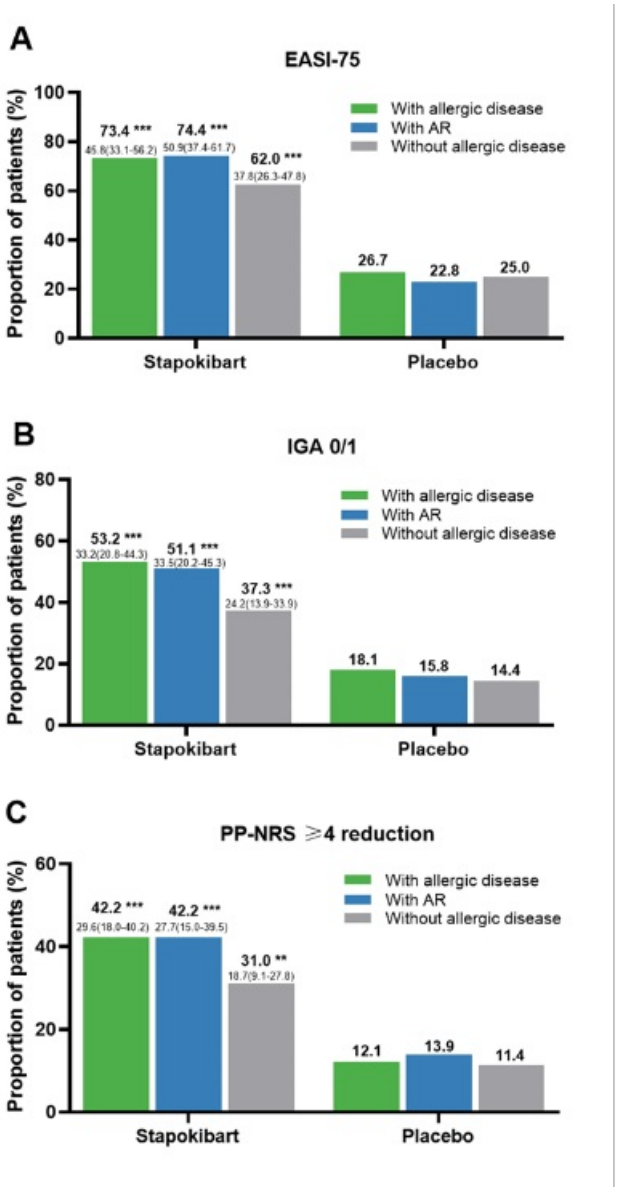

Figure 1. Proportions of patients achieving Eczema Area and Severity Index ≥75% improvement (EASI-75) (A), an Investigator's Global Assessment (IGA) score 0/1 (B), and a 4-point reduction in peak pruritus numerical rating scale (PP-NRS) (C) at week 16. Difference vs. placebo (95% confidence interval (CI)) is shown below percent values on top of bar graphs. ***P < 0.0001, ** P<0.001

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Abstract N°: 3685

Erythematotelangiectatic rosacea and Telangiectasia Macularis Eruptiva Perstans in a Filipino lady: Case Report

Krystel Angela Olano*[1], Johannes Dayrit[1], Luella Joy Escueta-Alcos[1], Gracia Buendia-Teodosio[1]

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Introduction & Objectives: Erythematotelangiectatic rosacea (ETR) and Telangiectasia Macularis Eruptiva Perstans (TMEP) are two distinct dermatological conditions that present with characteristic vascular changes in the skin. ETR is a subtype of rosacea characterized by persistent facial redness, flushing, and telangiectasias. On the other hand, TMEP is a form of cutaneous mastocytosis that manifests as macular or flat reddish-brown patches with prominent telangiectasias. These conditions can significantly impact a patient's quality of life due to their cosmetic appearance and associated symptoms.

We describe a unique case of a Filipino lady who presents with ETR and TMEP, a rare but maybe related condition.

Materials & Methods: A 58-year-old, diabetic and hypertensive, Filipino, female, initially presented with long-standing facial telangiectasias and flushing precipitated by UV exposure since adolescence. Six years prior to presentation, she noted the appearance of multiple telangiectatic erythematous macules and patches on the upper arms, spreading to the upper chest and back with flushing triggered by UV exposure. There were no systemic symptoms and no similar family history.

Dermatologic examination revealed multiple erythematous telangiectasias on the face, and erythematous macules and patches with negative Darier sign on the upper chest, back, and upper arms. Dermoscopy of lesions showed linear vessels, some arranged in vascular polygons on the face, and branching linear vessels and yellowish-orange amorphous areas on the back.

Results: Biopsies were performed to confirm the diagnosis. Histopathological examination of the face revealed solar elastosis, telangiectasia of blood vessels, and a periadnexal inflammatory infiltrate of lymphocytes, histiocytes, and plasma cells. Giemsa and CD 117 stained mast cells in the perivascular infiltrate. Biopsy of the back showed telangiectasia of blood vessels and a mild superficial perivascular inflammatory infiltrate of lymphocytes and mast cells, with more than 10 mast cells per high power field visualized using Giemsa and c-kit (CD117) staining.

The patient was diagnosed with both ETR and TMEP. No systemic involvement was found in the workup.

Management included counseling on avoiding triggering factors such as alcohol, extreme temperatures, exercise, nonsteroidal anti-inflammatory drugs, aspirin, anesthesia, and narcotics. The patient was treated with oral antihistamines, β-CALM complex cream, and broad-spectrum sunscreens, resulting in mild improvement in her lesions. The patient also underwent 595 nm flashlamp-pumped dye laser treatment on the face and trunk to treat the telangiectasias.

Conclusion: This case emphasizes the importance of considering mixed or overlapping dermatological conditions in patients with complex skin presentations to facilitate precise diagnosis and targeted treatment strategies. This report raises awareness of the potential coexistence and relationship of ETR and TMEP in individuals presenting with macules and patches with telangiectasias. Proper diagnosis can guide appropriate management and lead to better patient outcomes. It also highlights the need for thorough examination, detailed history-taking, and the use of diagnostic tools like dermoscopy and biopsy to enhance clinical decision-making and patient care.

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Abstract N°: 3764

Polymorphic Maculopapular Mastocytosis in a Filipino Infant: A Case Report

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Introduction & Objectives: Polymorphic maculopapular cutaneous mastocytosis (pMPCM) is the most common form of cutaneous mastocytosis in children, characterized by brown to red oval lesions, plaques, and nodules of various sizes, typically distributed asymmetrically on the skin. The lesions commonly appear on the head (particularly the forehead and neck) and extremities. The morphology of lesions can change during the course of the disease, and blistering is often seen in infancy but usually resolves by age 2 or 3. Nodular lesions may evolve into plaques in childhood and regress around puberty, potentially flattening over time to mimic anetoderma. Most patients with polymorphic lesions experience a favorable prognosis with spontaneous regression of skin lesions by adolescence, and the condition is generally skin-limited.

We describe a case of pMPCM in a Filipino infant, a rare condition in the population presenting with polymorphic skin lesions.

Materials & Methods: A 1-year-and 9-month-old Filipino male infant presented with widespread skin lesions including macules, plaques, nodules, and bullae. He was born full-term via spontaneous vaginal delivery and was admitted due to sepsis caused by a ruptured amniotic membrane. At 4 months old, brown macules appeared on his upper and lower extremities. By the age of 1, some of the macules progressed into papules and nodules, spreading to the face, trunk, upper and lower extremities. Over time, some of these lesions developed into bullae that ruptured, leaving scars.

The only other significant health event in his history was an episode of acute gastroenteritis with mild dehydration at 7 months old. There is no family history of similar conditions.

Dermatological examination showed multiple brown macules, plaques, nodules, and some with vesicles and bullae, both intact and ruptured, with residual scars on his face, trunk, and limbs. Darier sign was positive. Otherwise, the examination was unremarkable.

Results: Histopathological results showed a very dense, pandermal infiltrate of mast cells, highlighted by CD117. No evidence of systemic involvement was noted on further work up. Treatment included loratadine and mid-potent topical corticosteroids for most lesions, while high-potent topical corticosteroids for the nodules. The patient showed improvement in his lesions. He is being closely followed up and is now 2 years and 5 months old, with observed speech developmental delay.

Conclusion: This case underscores the importance of recognizing the diverse clinical presentations of cutaneous mastocytosis and the need for a high level of suspicion when assessing possible cases. Treatment primarily focuses on relieving symptoms and avoiding factors that can exacerbate the condition. While it can be challenging for both the patient and their family, most cases have a good prognosis. Additionally, this case contributes valuable insights into pediatric mastocytosis within underrepresented populations like the Filipinos, emphasizing the need to enhance diagnostic precision and customize treatment approaches for different demographic groups. This knowledge will improve patient care and outcomes for those living with this condition.

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Abstract N°: 3766

Omalizumab as an emerging treatment for severe refractory cutaneous mastocytosis: A systematic review of the adult and paediatric literature

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Introduction & Objectives:

Mastocytosis is a heterogenous group of diseases characterized by clonal proliferation of mast cells. Severe presentations of cutaneous mastocytosis (CM) can be difficult to treat and have a debilitating impact on quality of life. Omalizumab, a monoclonal anti-IgE antibody, is an emerging off-label therapy for both cutaneous and systemic mastocytosis. In this systematic review of the adult and paediatric literature, we aim to characterize its safety profile and efficacy.

Materials & Methods:

EMBASE, MEDLINE, and CENTRAL were searched using keywords "cutaneous mastocytosis" plus "omalizumab". Adult and paediatric data were analyzed separately. Study quality and bias were assessed using the Grading of Recommendations, Assessment, Development, and Evaluation scale. 78 studies were screened, with eight studies involving 22 adult patients and four studies involving five paediatric patients included in the final analysis.

Results:

All paediatric patients, with an age range of 2-15 years, experienced complete remission within an average of 1.5 months. 20% experienced relapse. Corticosteroid cessation was achieved in 80% of paediatric patients. No major adverse events were reported. Among adult patients, 95% showed improvement in CM, with three patients achieving complete remission within an average of four months. 16% experienced relapse. Two major adverse events occurred in the adult population and required treatment discontinuation.

Conclusion:

Our review provides support for the use of omalizumab as a steroid-sparing treatment for resistant cutaneous mastocytosis in both adult and young paediatric patients. The majority of patients demonstrated clinical improvement, with low rates of major adverse events and relapse.

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Abstract N°: 3800

Serum Immunoglobulin E level and Its relationship with Eosinophil count among patients with Chronic spontaneous urticaria and accompanying disease

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[1]Medical University, Pleven, Bulgaria

Introduction & Objectives:

Urticaria is a frequent, mast cell-driven disease that presents with wheals, angioedema, or both. The role of total serum IgE and Eosinophil count in the pathogenesis of the disease is widely discussed. Our team measured and compared the levels of both biomarkers in patients whit chronic spontaneous urticaria (CSU). Aim of the study is to determine serum immunoglobulin E level and its relationship with eosinophil count among patients with atopy, and insufficient effect of antihistamine/corticosteroid treatment.

Materials & Methods:

It was a cross-sectional study of consecutive patients diagnosed with CSU and hospitalized in allergology department for the period January 4-December 18, 2023. Five milliliters of blood were analyzed for immunoglobulin E estimation using an immunoglobulin E ELISA kit and determination of eosinophil count using pack five hematologic autoanalyzer. Extracted data were analyzed using IBM SPSS version 29.0 software.

Results:

There were 12 1 patients studied comprising 84 (69.4%) females and 36 (30,6%) males.

The range age was from 6 to 84 year old. The average age of all patient was 46.24, (39.61 for men, 49.08 for women). High levels of IgE had 40 of the participants (154.4), and 21 were those with high levels of eosinophil count (2.94). There was no significant correlation between IgE serum level and Eo count in these cases; only 7 of all patients had high levels of both markers.

Patients with atopy and accompanying disease allergic rhinitis were 44 of them; 17 had bronchial asthma; eight had atopic dermatatis. Medical history for allergic contact dermatis and insect allergy had respectively 6 had 8 of the participants. We found a statistically significant difference with increased level of tIgE in patients with CSU and accompanying atopic disease – bronchial asthma ($p = 0,05$) and allergic rhinitis ($p = 0.06$).

Conclusion:

Despite our preliminary expectation for possible correlation between tIgEs and eosinophil count in patients with CSU, there was no significant association. As expected, IgE levels are predominantly elevated in patients with atopy.

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Abstract N°: 4264

clinical and immunological follow-up of trichophytic kerion celsi - a case report

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[1]University Clinical Center of Kosovo, Dermatology, Prishtine, Kosovo

Introduction & Objectives:

Kerion Celsi is a severe, inflammatory form of tinea capitis that predominantly affects children and is caused by fungal infections. This case report details the clinical presentation, diagnosis, and management of a male patient diagnosed with this condition, with a particular focus on the elevated levels of total IgE observed during his treatment. The objectives of this study are to discuss effective diagnostic and therapeutic strategies for managing Kerion Celsi and to explore potential causes behind the high IgE levels, which could suggest an atypical immune response or concurrent allergic conditions [1][2].

Materials & Methods:

The diagnostic approach included a microscopic examination using KOH to detect mycosis, which confirmed a positive result for fungal infection. This was further substantiated by culturing the fungal material, which identified Trichophyton tonsurans as the causative organism. Additional laboratory tests were performed to assess the immune response, notably measuring the levels of total IgE.

Results:

The patient responded positively to the prescribed treatment regimen which included anti-fungal systematic therapy and antibiotics. This combination addressed both the fungal infection and the associated inflammatory symptoms. The follow-up assessments showed a significant reduction in scalp lesions and symptoms, with laboratory tests indicating a normalization of IgE levels.

Conclusion:

This case illustrates the critical need for early diagnosis and comprehensive treatment planning in cases of Kerion Celsi. The successful management of this patient underscores the effectiveness of combining systemic antifungal therapy with local and systemic antibiotic treatments. Future research should consider exploring the role of immune system markers like IgE in predicting treatment outcomes in fungal infections of the scalp.

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Abstract N°: 4499

Durian Hypersensitivity presenting with Acute Angioedema and Urticaria

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Introduction & Objectives:

Durian (*Durio zibethinus L.),* known as the 'king of fruits' is a thorny, pungent tropical fruit grown in Southeast Asian rainforests. Banned on the public transport due to its smell, the demand for durian has grown year-on-year, both locally and globally. There are little known reports of durian hypersensitivity.

Materials & Methods:

We report a case of a 78-year-old woman that was referred to our centre for acute bilateral eyelid angioedema and generalised urticaria without systemic symptoms, one hour after consuming durian of the Mao Shan Wang variety. She was treated at our hospital's emergency department with oral prednisolone and intravenous diphenhydramine with symptom resolution within 24 hours. She was otherwise well, and apart from durian, did not consume any medication nor food prior to this episode. She did not report any symptoms suggestive of viral infection that might account for her cutaneous symptoms.

Results:

Skin prick tests, including both prick-through and prick-to-prick tests to Mao Shan Wang and D13 species of fresh durian pulp were conducted 4 weeks after her initial presentation. The tests yielded positive reactions to both variants of fresh durian pulp (Mao Shan Wang and D13). The same tests were negative on a healthy control, ruling out an irritant reaction.

Conclusion:

This case report highlights the rare occurrence durian hypersensitivity presenting as acute angioedema and urticaria. The positive skin prick tests supports the hypothesis that the patient likely had an IgE mediated hypersensitivity to Mao Shan Wang durian with cross reaction with D13 variety of durian. It underscores the importance of considering food allergies in adults and emphasizes the need for research on tropical food allergies.

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Abstract N°: 4854

The role of IL-31 and its inhibitors in treating pruritus

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Introduction & Objectives:

Pruritus is defined as an unpleasant sensation of varying strength that provokes the desire to scratch, and, depending on its intensity and frequency, can make patients' life truly miserable. One of the clinically relevant pruritogens is the cytokine interleukin-31 (IL-31). The aim of this review is to demonstrate/illustrate the role of IL-31 in the pathogenesis of itch and evaluate the efficacy of IL-31 inhibitors in pruritus treatment.

Materials & Methods:

A systematic review was conducted of the PubMed and Web of Science databases, regarding all articles published until April 20th, 2024. Clinical trials, reviews, as well as prospective and cross-sectional studies were included. A total of 11 studies were included, 2 of them being reviews.

Results:

Three studies on association between IL-31 expression and pruritus in such conditions as psoriasis, allergic rhinitis and epidermolysis bullosa illustrate the correlation between increased levels of IL-31 and presence of itch. Anti-IL-31 antibody, nemolizumab, in four clinical trials with a total of 684 participants, showed significant improvement in pruritus, illustrated by the pruritus visual analogue scale (VAS) demonstrating downward trend, and changes in EASI score for AD patients and PP-NRS for PN patients. The improvement of sleep quality was also observed. The results of two long-term phase III studies (≥ 52 weeks) showed continuous improvement in pruritus, and supported the long-term use of nemolizumab in patients with AD. Moreover, in a different study involving 49 patients with prurigo nodularis (PN), vixarelimab was administered. The results demonstrated a higher rate of patients who received the biologics experienced a reduction of ≥ 4 points in worst itch numerical rating scale (NRS), visual analog scale (VAS), improvement in healing of representative lesions and improvement in

quality of life compared to the placebo group.

Conclusion:

Interleukin-31 appears to play a significant role in pruritis induction, and cell-type-specific contributions of IL-31R to itch, its expression mechanism and the downstream signaling pathway to induce itch should be further investigated. As of now inhibition of IL-31 by targeting the IL-31/IL31RA/OSMRβ axis suggests the improvement in pruritus, sleep quality, and overall, quality of life of patients suffering from pruritic disorders and appears to be a promising treatment option for such illnesses.

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Abstract N°: 5564

When to suspect a Specific Polysaccharide Antibody Deficiency in the adult: A Case Report

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Introduction

Inborn errors of immunity (IEI) include many genetic disorders that affect the innate and adaptative responses. The most common IEI are antibody and complement deficiencies which can affect both adults and children. IEI are underdiagnosed in adults. (1)

Specific polysaccharide antibody deficiency in adults (SPAD) is an impaired antibody response to polysaccharide antigens. It is defined as normal IgG, IgA, and IgG subclass levels, with an isolated impaired response to the 23-valent pneumococcal polysaccharide vaccine (PPV23) and adequate levels of tetanus toxoid (TT) antibodies and/or adequate response to TT vaccine.

Case report:

A 50-year-old man presented with a 4-year history of multiple abscesses. The patient didn't have chronic medical conditions, allergies, or a family history of immunodeficiencies. He referred prior multiple surgeries due to the presence of "migratory" abscesses. The first one was located on his right testicle, and was treated with antibiotics and drainage. One year later he was diagnosed with lumbar spondylodiscitis in L3-L5 which was treated with surgery and systemic antibiotics. At presentation to the dermatology department, the patient had fever. Physical examination revealed multiple scars on his right armpit and groin, and a localized dermatosis on the buttocks with the presence of 3 abscesses with drainage of pus. Testing for hepatitis B and C and HIV were negative. We performed a skin biopsy that showed a neutrophilic and lymphohistiocytic infiltrate, with microabscesses, focal necrosis and isolated eosinophils. PAS, Grocott and Ziehl-Neelsen staining were negative. On the light of these findings and the clinical history, specific laboratory tests were performed with NBT, lymphocyte subpopulations, IgG subclasses and anti-pneumococcal antibodies, in which the last ones were below normal range (0.2 to 0.35 µg/mL), serotype 1 (1):< 0.1, 3(3):< 0.1, 4(4): 0.3, 9 (8): <0.3, 9 (9N): <0.1, 12(12F): < 0.1, 14(14): <0.1, 19 (19F): 0.8, 23 (23F): <0.1, 26(6B): <0.1, 51 (7F): 0.2, 56(18C): 0.1 and 68 (9V):0.1. The diagnosis of SPAD was made. He received PPV23 and presented clinical improvement on followup

Conclusion: SPAD is an IEI that should be considered in patients with multiple, otherwise unexplained, bacterial infections. Dermatologists should be aware of the clinical presentation in adults with recurrent infections.

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Abstract N°: 5644

Vitiligo, alopecia areata as part of an inborn error of immunity due to Haploinsufficiency of A20 as a different clinical phenotype in a Costa Rican pediatric patient

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Introduction & Objectives:

TNAP3 is a protein encodeg by the gene TNFAIP3. It plays a crucial role in the negative regulation of inflammation and immunity. It is ubiquitin-editing enzyme that inhibits molecules involved in the canonical NF-kB signalling pathway1. Zhou published the first report of a human disease caused by haploinsufficiency of A20 (HA20)2. They proposed high-penetrance loss-of-function germline mutations in TNFAIP3/A20. A20-deficient mice (A20-/-) display persistent NFκB activation, spontaneous multiorgan inflammation and early lethality. Aging heterozygous mice(A20+/-) develop autoantibodies resembling human autoimmune conditions3. Patients may present with early-onset systemic inflammation and a Behcet-like disease, or a variety of autoinflammatory and autoimmune features (Debeljak). We report a mutation in the TNFAIP3 gene with a different clinical presentation.

Materials & Methods:

We report a heterozygous pathogenic mutation c.1722dup (p.Ser575Leufs*97) in TNFAIP3 gene using Invitae Inborn Errors of Immunity and Cytopenias Panel®. The patient´s mother gave the informed consent to report the patient.

Results:

We report a 11-year-old patient born from non-consanguineous parents from Quepos, Costa Rica. He has a brother with a diagnosis of early onset Bechet disease, pending genetic studies and immunologic evaluation.

No relevant perinatal history. He lives in a rural area in extreme poverty conditions. He has a complete immunization schedule (including BCG, with no reported complications). His medical history includes 3 hospitalizations due to bronchiolitis before 2 years of age, recurrent otitis media during pre-school years. He has had recurrent fever and arthralgias. He was evaluated by the Immunology Department and hypogammaglobulinemia was documented, so restitution with intravenous immunoglobulin has initiated.

At 4 years of age hypocromic lesions were described in the right inner canthus and an alopecic plaque in the right occipital region of the scalp, as dermatological findings. Throughout the years he presented chronic, intermittent diarrhea and chronic suppurative cough. CT scan revealed lung bronquiectasias. Gastrointestinal biopsies documented absence of plasmatic cells and lymphonodular hyperplasia. He does not have a history of ulcers or uveitis.

Currently, he weights 19.2 kg (under 3 SD for BMI/age: severe malnutrition) and with a height of 122 cm (under under 3 SD for age and sex: severe short stature). His dermatologic findings are an achromic plaque on the nasal bridge. Under dermoscopy there is a darker brownish central hue that could correspond to a previous pigmented mole, with no vascular pattern. He has an alopecic rounded 1.5cm in diameter plaque, which under normal light does not show any hairs, no exclamation marc, cadaveric hairs, but under dermoscopy with woods light added,

shows multiple achromic short hairs. No scaling or erythema present. Perioral hypopigmentation with ill-defined borders.

Conclusion:

We report a patient HA20 syndrome associated with a different clinical phenotype than the usual early onset Behcet disease.

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Abstract N°: 5833

Allergic contact dermatitis to para-phenylenediamine: A series of 10 cases.

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Introduction:

Para-phenylenediamine (PPD) is an amine commonly used as a principal component in hair dyes and henna tattoos. Contact dermatitis is a frequent complication associated with its use. Our objective is to study allergic contact dermatitis to PPD within our institution.

Materials and Methods:

A prospective study conducted over a two-year period (2021 to 2023) within our hospital, aimed at collecting all patients presenting with PPD-induced contact dermatitis confirmed by patch testing.

Results:

Ten patients were included, with an average age of 28 years and a female predominance (60%). Forty percent of patients had a history of black henna application, while 60% had used hair dye. The average onset time was 24 to 48 hours in all cases. An erythematous plaque with vesicles and post-vesicular erosions, associated with itching and a burning sensation, was observed in 100% of cases, with eyelid edema present in 60% of cases. The primary site affected was the face (60% eyebrows, 30% beard), followed by the hands (10%). All patients underwent patch testing, which returned positive for para-phenylenediamine (PPD) in all cases. All patients were treated with topical corticosteroids and advised to avoid hair dyes containing PPD.

Discussion:

Allergic contact dermatitis to para-phenylenediamine (PPD) is a common inflammatory skin reaction resulting from exposure to this chemical found in various hair products such as hair dyes and henna tattoos. This skin allergy can manifest as erythema, vesicles, and intense itching, often localized to areas exposed to products containing PPD. The allergic reaction typically occurs within 24 to 48 hours after exposure and may persist for several days. Due to its high prevalence and characteristic clinical manifestations, PPD-induced contact dermatitis represents a diagnostic challenge and significant concern in allergic dermatology. Hence, there is a need for an educational approach promoting the use of alternatives without this substance to prevent skin conditions.

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Abstract N°: 6112

Contact leucoderma to ammonium persulfate in a hair dye

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Introduction: Contact leucoderma is an acquired form of leukoderma caused by repeated topical or systemic exposure to a variety of chemicals. We herein report a case of contact leukoderma of the scalp due to ammonium persulfate contained in a hair bleaching product.

Observation: A 48-year-old woman developed an asymptomatic depigmented area on her scalp two weeks after the application of a hair bleaching product. At examination, we noted a sharply demarcated depigmented patch at the right frontal scalp region with localized leukotrichia. At wood's lamp examination, there was an accentuation of the depigmented zone. There was no involvement of other skin areas. Patch tests with the European standard series and Hairdressing Series (Chemotechnique diagnostics) were performed. Readings at 48 and 72 hours showed positive reaction to ammonium persulfate (++) and a negative reaction to paraphenylenediamine (PPD). This reaction was relevant given the presence of ammonium persulfate in her hair bleaching product (*Renée Blanche, hair colouring cream*). The clinical diagnosis of contact leukoderma following hair dye application was made and we advised the patient to stop bleaching products containing ammonium persulfate.

Discussion: Contact leukoderma is characterized by a localized loss of skin pigmentation caused by certain chemicals, toxic to melanocytes including derivatives of phenols and catechols. The first cases of depigmentation associated with the application of hair dyes were reported by Taylor et al. Since then, there have been a few similar reports in the literature. A cytotoxic effect of hair color ingredients on melanocytes, have been postulated. This results in loss of melanocytes within the skin and hair follicle, leading to skin and hair depigmentation. PPD, a well-known component of oxidative hair dyes, is the most incriminated agent. Lesions usually develop two weeks to six months after the exposure. The absence of a preceding contact dermatitis, itching or trauma helps to rule out post inflammatory hypopigmentation and kobner-induced vitiligo. In our patient, the history of exposure to hair a bleaching product, the sharply demarcated depigmentation restricted to the area of application, and the presence of leukotrichia are suggestive of contact leukoderma. Our patient had an intense positive reaction to ammonium persulfate, contained in the used hair bleaching product. It is a strong oxidizing agent, commonly used in hair bleaching formulas and one of the main causes of allergic contact dermatitis in hairdressers. Associated respiratory symptoms and anaphylaxis have also been reported. To the best of our knowledge contact leukoderma to ammonium persulfate have not been previously reported.

Conclusion: Contact leukoderma is an under-diagnosed condition that can mimic idiopathic vitiligo. Considering the widespread use of hair bleaching products, clinicians should suspect this diagnosis in cases of scalp depigmentation.

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Abstract N°: 6120

Useful diagnostic parameters of chronic spontaneous urticaria: our research results

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Introduction & Objectives: Considering that in chronic spontaneous urticaria (CSU) the values of serum inflammatory parameters are elevated, the aim of this study was to examine the most important biomarkers of CSU and their relationship in patients with CSU according to the severity of the disease.

Materials & Methods: We included 41 patients with CSU in our study (32 women and 9 men, aged from 23 to 79 years) who filled out questionnaires concerning disease severity and quality of life (the Urticaria Activity Score summed over 7 days, UAS7, the once-daily Urticaria Activity Score, UAS, the Urticaria Control Test, UCT, and the Dermatology Life Quality Index DLQI). Blood samples were taken to measure interleukin 6 (IL-6), complete blood count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), TSH, T3, T4, anti-TPO, anti-TG, D-dimers and vitamin D.

Results: In 44% of patients, the disease lasted from 3 to 6 months, and most of them had moderate CSU (39%) and a small effect of the disease on the dermatological quality of life (42%), 90% of them had uncontrolled disease, 43% had moderate effect of urticaria on life, and only 5 of them have a severe form of the disease. In 22% of patients, an autoimmune basis of the disease was established (determined by an ANA titer equal to or greater than 1:160). According to the obtained results, basopenia appeared to be a good predictor of daily clinical severity of CSU (daily UAS; $p = 0.002$), and was associated with a 6.2 times greater likelihood of moderate or severe CSU (daily UAS; $p = 0.017$). ESR value was the only significant predictor of weekly disease activity (UAS7; $p = 0.043$) and was associated with a 4.9 times higher odds of moderate or severe CSU (UAS7, $p = 0.038$). Serum IL-6 correlated with CRP ($p = 0.016$), with ESR ($p = 0.045$), with D-dimers ($p < 0.001$), with T3 ($p = 0.019$) and with T4 ($p = 0.043$). Serum T4 correlates with the duration of the disease ($p = 0.036$), and T3 with

effect of the disease on the dermatological quality of life (DLQI; p = 0.017).

Conclusion: Of all the serum biomarkers of CSU, our study showed that the most useful biomarkers of CSU were the values of basophils (basopenia), for assessing the daily severity of CSU, then the value of ESR as a significant predictor of weekly disease activity. In addition, serum IL-6 was statistically significantly correlated with the values of CRP, ESR, D-dimers, T3 and T4, while serum value of T4 correlates with the duration of the disease, and T3 with effect of the disease on the dermatological quality of life. Although our results are promising, this study should be conducted with a larger number of CSU patients.

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Abstract N°: 6148

Telangiectasia Macularis Eruptiva Perstans

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Introduction & Objectives:

Telangiectasia macularis eruptiva perstans (TMEP) is a rare subtype of mastocytosis, primarily affecting adults. Early diagnosis is crucial, given its rarity and potential for systemic involvement. We present two cases of TMEP to explore its clinical features and diagnostic complexities.

Materials & Methods/Observations:

An 18-year-old male presented with an asymptomatic, erythematous 10 cm macule on a slightly brownish background in his left pectoral region. Worsening with exercise for a year, without other systemic symptoms.Our 2nd patient, a 53-year-old woman with no prior medical history, presented with red macules covering both breasts persisting for 2 years. Additionally, she had erythematous macules confined to her abdominal stretch marks with no systemic symptoms and/or aggravating factors.

Upon clinical examination, both patients exhibited erythematous macules with telangiectasia on the surface. The lesions were non-blanchable, and Darier's sign was negative.

Dermoscopic examination showed: a vascular pattern. Specifically, in the male patient, linear branching vessels formed a reticular network against a brown-pigmented background. In the female patient, tortuous branching vessels, dotted vessels, and a background of brownish discoloration were observed.

The remainder of the mucocutaneous and systemic examination did not contribute further findings.

Histology showed occasional epidermal atrophy, discrete pigmentation, a mononuclear infiltrate with a notable presence of mast cells surrounding the capillaries, particularly prominent in the upper dermis. , stained by toluidine blue, C-kit proto-oncogene mutation was observed. We explored alternative diagnoses including intravascular B-cell lymphoma and angioma serpiginosum. However, the clinical, dermoscopic, and histopathological findings confirmed telangiectasia macularis eruptiva perstans. Blood count, liver function, and serum tryptase levels, abdominal ultrasound findings were normal in both patients.

Treatment consisted in H1 antagonists along with hypoallergenic diet, avoiding triggers like sunlight, extreme temperatures, alcohol, and specific medications known to exacerbate mast cell degradation. Compliance with these measures led to improved symptoms and reduction in skin lesions.

Results/Discussion:

Mastocytosis involves mast cell proliferation in various organs. Telangiectasia macularis eruptiva perstans (TMEP) is a rare cutaneous variant characterized by telangiectasia macules with a background color ranging from light to dark brown with lesions typically found on the chest and extremities. While usually confined to the skin, systemic involvement can occur. Dermoscopic findings aid diagnosis, with the reticular vascular pattern being characteristic of TMEP. Our patients displayed the typical reticular vascular pattern consisting of linear, tortuous, and branching vessels, corresponding histologically to dilated superficial capillaries surrounded by mast cells around venules and dilated capillaries of the superficial venous plexus of the dermis .Diagnosis relies on clinical and histopathological

assessment, and treatment is symptom-based, lacking a standard approach.

Conclusion:

Prompt identification of TMEP is crucial due to its systemic potential. Dermoscopy is invaluable for diagnosis, especially in challenging cases.

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Abstract N°: 6216

Gut Lachnospiraceae are decreased in patients with chronic spontaneous urticaria

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Introduction & Objectives:

Although the precise etiopathogenesis is largely unknown, chronic spontaneous urticaria (CSU) is considered an immune-mediated inflammatory disease, with an autoimmune etiology in more than 50% of cases. Gut microbiota plays a crucial role in developing and maintaining immune system balance, such as preventing pathogen propagation, controlling metabolism, improving epithelial integrity, and regulating the components of both innate and adaptive host immune systems. One of the most important factors in maintaining immunotolerance are short-chain fatty acids (SCFAs) - produced by gut bacteria through the anaerobic fermentation of indigestible polysaccharides. This study aimed to investigate the association between the abundance of SCFAs – producers in the gut and CSU.

Materials & Methods:

This case-control study included 22 CSU patients and 23 healthy controls. The 16S rRNA gene massive parallel sequencing was performed to analyze the gut microbiota composition from the fecal samples of all participants.

Results:

Beta-diversity showed clustering between the two examined groups, suggesting significant differences of gut microbiota between CSU patients and healthy controls ($p<0,05$). Alpha diversity in the CSU group was significantly decreased according to the Evenness index ($p<0,05$), while other indices did not show significant differences. Linear discriminant analysis effect size (LEfSe) identified a significantly decreased abundance of gut bacteria from the *Lachnospiraceae* family in the CSU group, including *Lachnospira*, *Roseburia, Ruminococcus*, *Coprococcus*, and *Eubacterium eligens group* ($p<0.05$). Furthermore, the receiver operating characteristic curve (ROC) was used to analyze the potential diagnostic value of those bacteria; bacterial genera *Lachnospiraceae NK4A136*, *Eubacterium eligens*, and *Roseburia* in the gut were identified as potential diagnostic biomarkers for CSU.

Conclusion:

Gut microbiota compositions differ significantly between CSU patients and healthy individuals. Our study revealed decreased levels of SCFA-producing bacteria in the gut microbiota of CSU patients. These results suggest that SCFAs could have an impact on immune dysfunction in the pathogenesis of CSU. Furthermore, we showed that some *Lachnospiraceae* members may serve as potential diagnostic biomarkers for CSU. We speculate that modulation of SCFAs could serve as a prospective additional option in the treatment of CSU. Future studies are needed to investigate the potential of SCFAs in the treatment of CSU.

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Abstract N°: 6393

salivary melatonin

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Introduction & Objectives:

In the last twenty years, salivary diagnostics have been developed for the purpose of early detection of certain diseases, measurement of drug concentration, detection of narcotic drugs and alcohol in the organism, as well as hormone concentration in saliva. Saliva sampling is a non-invasive, easier and simpler procedure compared to taking blood samples. Saliva is not an ideal biological material, because the individual composition of saliva can be influenced by numerous factors, such as age, gender, smoking, diet, medications or the pathophysiological status of the oral cavity.

Materials & Methods:

By searching the literature on determining the concentration of salivary melatonin in the last five years, it was most often determined in diseases of the oral cavity, malignant and psychiatric diseases, such as periodontal diseases, caries, oral carcinoma, prostate carcinoma, anxiety disorder, depression, bipolar disorder and post-traumatic stress disorder. Factors that can affect the results of salivary melatonin can be divided into unchangeable and variable factors. Unchangeable factors include genetic factors (the volume of active pineal tissue determines the amount of melatonin secretion), age (with increasing age, melatonin secretion decreases), gender (sex hormones and the use of oral hormonal contraception affect melatonin secretion), while variable factors include they include light (the most important factor that inhibits melatonin secretion), position in which saliva is sampled, and physical activity. Certain diseases of the organs involved in its synthesis and metabolism can affect the activity of melatonin secretion (eg eye diseases, spinal injuries, liver and kidney diseases, chronic periodontitis) as well as the use of certain drugs (beta blockers, benzodiazepines, MAO-inhibitors, antidepressants, oral contraceptives…).

Results:

Due to the simplicity and reasonable price, the most common methods for determining salivary melatonin are the radioimmunoassay and the ELISA test (Enzyme-linked immunosorbent assay). Also, liquid chromatography and mass spectrometry can be used to determine the value of melatonin. The determination of the level of salivary melatonin can be influenced by the circadian rhythm, the speed of saliva flow and the method of measurement.

Conclusion:

By searching the literature, in skin diseases, salivary melatonin was determined only in patients with atopic dermatitis and in patients with CSU. Studies have shown decreased values of salivary melatonin concentration in both mentioned diseases.

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Abstract N°: 6607

Expression profile of innate immunity factors in Sézary syndrome and idiopathic erythroderma

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Introduction & Objectives: Innate immunity represents defense mechanisms that are constitutive against pathogens. Toll-like receptors(TLR) provide the recognition of external agents. Defensins are peptides whose actions are due to direct toxicity to viruses, bacteria and fungi. Cathelicidins are produced by neutrophils and epithelial cells.

Sézary syndrome(SS) is a leukemia erythrodermic form of cutaneous T-cell lymphoma. Infections are the main causes of death for SS patients. Extensive assessments in erythroderma are necessary to achieve an early diagnosis, aiming to reduce mortality, whereas up to 20% of patients may remain undiagnosed, characterizing idiopathic erythroderma(IE). Herein, we focus on SS the innate immunity, mainly related with TLRs cutaneous expression in patients with SS and IE.

Materials & Methods: The expression of TLR2, TLR4, TLR5, TLR9, beta-defensin and cathelicidin were assessed in skin biopsies of SS patients(n=7), IE patients (n=9) and a control healthy donor group (HD, n=18) by immunohistochemistry in the epidermal and dermal layers. The slides were scanned using an Aperio ScanScope. Then, the images were analyzed with Image-Pro Plus, version 4.5.0.29.

Comparisons between the three groups were performed according to Kruskal-Wallis test. The level of significance considered was $p \leq 0.05$. All of the statistical and graphic representations were executed with GraphPad Prism 7 software.

Results: Alterations in the TLR expression at epidermal section were detected mainly in the group of IE compared to HD group(figure 1). As observed as upregulation of TLR2, TLR4, TLR5, TLR9 in IE group. Regarding SS group, showed no upregulation of TLR2 and TLR5 at epidermis, despite the known bacterial skin colonization. Together with IE group, SS samples showed increased expressions of TLR4 and TLR9 compared to HD group.

At dermic section SS group showed upregulation only for TLR9 expression, while IE increased for all TLRs analyzed expression except for decreased TLR2 expression(figure 1). It evidenced that erythroderma, when idiopathic leads for an active innate immunity, while SS was unresponsive, activate only for some TLRs, which may explain their bacterial infections susceptibility.

Beta-defensin and cathelicidin expression was increased at epidermal section in both SS and IE group compared for HD(figure 2). At dermis similar expressions were detected between groups. Moreover, similar expression was observed for cathelecidin in all analyzed groups.

Conclusion: The inadequate activation and expression of TLR in the SS group may contribute to greater susceptibility to bacterial infections and support the higher mortality of people with SS. The data, in a pioneering way, highlight the differential and active response ability of TLR in IE. In addition, it is relevant to expand evaluations related to intracellular signaling mechanisms in the process of recognizing innate immunity in SS and IE.

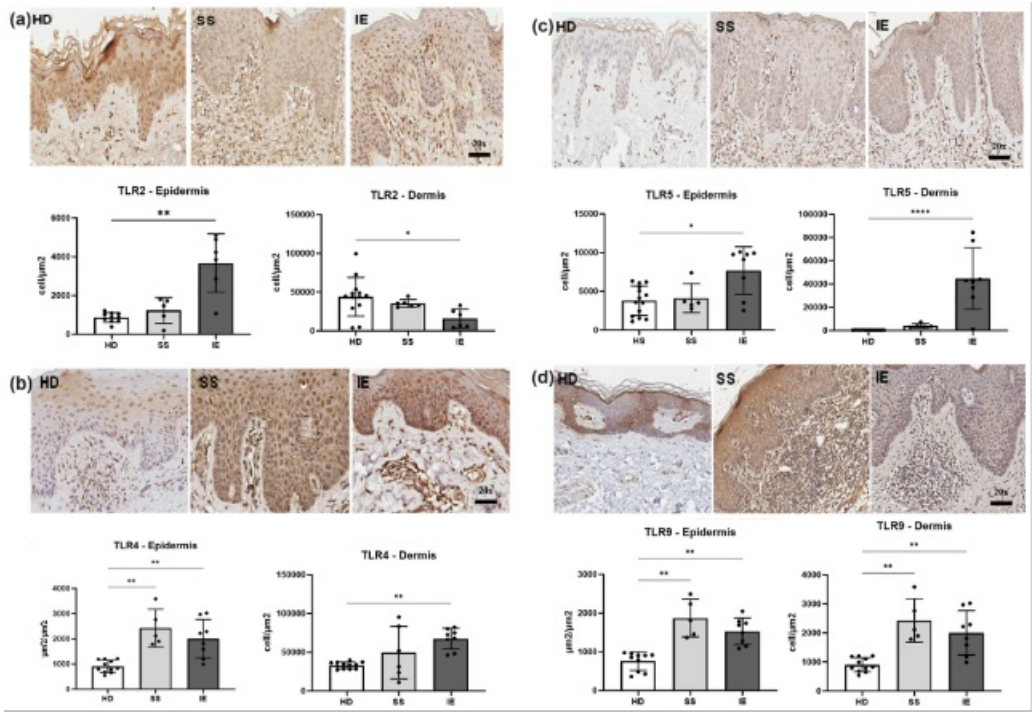

Figure 1 – Altered cutaneous TLRs expression in SS and IE groups. (a) TLR2, (b) TLR4, (c) TLR5, and (d) TLR9 expression was evaluated in biopsies of samples from the SS group in the epidermis (n = 5) and dermis (n = 5-6); from the HD group in the epidermis (n = 10-14) and dermis (n = 11-13); and the IE group in the epidermis (n = 6-8) and dermis (n = 6-8). Markings performed by immunohistochemistry. Analyses were performed separately on the epidermis and dermis. Values are expressed as median and interquartile, *p ≤ 0.05, **p ≤ 0.01.

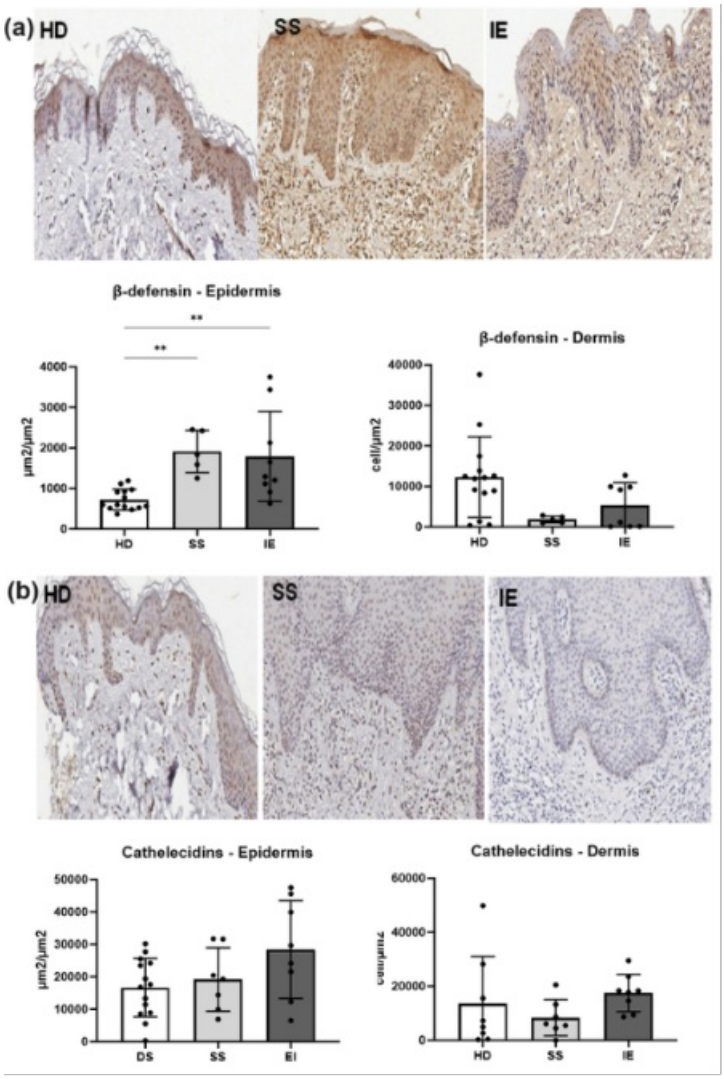

Figure 2 – Expression of antimicrobial peptides. (a) Beta-defensin (b) cathelicidin expressions were evaluated in biopsies of samples from the SS group in the epidermis (n = 5) and dermis (n = 5); from the HD group in the epidermis (n = 14) and dermis (n = 14); and the IE group in the epidermis (n = 8) and dermis (n = 9). Markings performed by immunohistochemistry. Analyses were performed separately on the epidermis and dermis. Values are expressed as median and interquartile, *p ≤ 0.05, **p ≤ 0.01.

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Abstract N°: 6931

Xanthomatous Reaction in Lupus Erythematosus Panniculitis A Clinicohistologic Mimicker of Leprosy in an Endemic Country

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Introduction & Objectives:

Lupus erythematosus panniculitis (LEP) is a rare variant of chronic cutaneous lupus erythematosus that can occur either as a separate disease or concomitantly with systemic or discoid lupus erythematosus. This condition is characterized by firm, painful, and persistent nodules, which heal with scarring and lipoatrophy, leading to significant disfigurement. Leprosy, which is endemic in Indonesia can cause facial swelling and skin nodules, thus mimicking LEP. Our case study illustrates the importance of comprehensive diagnostic evaluation, including clinical presentation and histopathological examination to differentiate between these conditions accurately.

Materials & Methods:

A 40-year-old woman presented with erythematous plaques and nodules on her forehead, cheeks, and chin for the past 4 years. Each time a lump appeared, the patient experienced pain and swelling at the lesion site. Over several months, the skin over the nodules became harder and developed atrophic plaques. The patient had no fever, weight loss, anesthesia, ulceration, nerve thickening, or impaired nerve function. LEP was considered as the diagnosis with leprosy as the differential diagnosis. Acid-fast bacilli test, serology and histopathological examination were performed to confirm the diagnosis.

Results:

The ANA, C3, and C4 examination results indicate normal titers. Histopathological examination revealed atrophic epidermis, perivascular and periadnexal lymphocytic and histiocytic inflammatory infiltrates, along with mucin deposition. Lobular panniculitis with foamy cells was observed in the subcutaneous layer. In some areas, bean-bag cells, which are characteristic features of subcutaneous T-cell lymphoma (SPTCL), were observed. However, immunohistochemical examination showed positive results for CD3, CD4, CD5, CD7, CD8, and CD20; negative for CD30, and low proliferation of Ki67, supporting a non-neoplastic condition. Although a negative acid-fast stain on a slit skin smear and low titers of anti-phenolic glycolipid-1 IgM and IgG initially suggested a non-lepromatous process, the presence of abundant foamy histiocytes on histology indicated otherwise. Fite-Faraco and GMS staining ruled out leprosy by showing no presence of acid-fast bacteria. Foam cell occurrence in cutaneous LE is infrequent. While the underlying mechanism remains elusive, some authors hypothesize that chronic inflammatory processes and trauma leading to epithelial damage and keratinocyte degeneration also trigger the release of lipid material, subsequently degraded by macrophages.

Conclusion:

Distinguishing between LEP and leprosy is crucial, particularly in endemic regions. Although rare, foam cells may be observed in the histopathology of LEP. Microbiological examination, serology, and additional staining for leprosy can assist in establishing the diagnosis.

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Abstract N°: 6938

Association between atopic dermatitis and hidradenitis suppurativa: a series of 3 cases

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Introduction & Objectives: Atopic dermatitis (AD) and hidradenitis suppurativa (HS) are commonly encountered dermatological conditions which show bidirectional association. Although AD is Th2-mediated, elevated Th17 can be observed in its chronic lesions, which is a major player in HS pathogenesis. Both disorders are associated with genetic predisposition and can be deteriorated by mental stress. We aim to present our 3 patients with concomitant AD and HS.

Materials & Methods: case series

Results:

1st case: A 27-year-old female patient suffering from AD since childhood presented to our clinic 9 years ago with Pityriasis amiantacea and eczema on her upper extremities. She also had several small, inflamed nodules and atrophic scars in the inguinal and axillary regions, suggesting a diagnosis of HS. She had been treated topically, but due to further deterioration throughout the following years with generalization of eczema and worsening of HS (moderate severity, IHS4=5), methotrexate was introduced at a dose 10 mg/week. This resulted in only partial improvement of AD, no improvement of HS, and multiple side effects (nausea, weakness, irregular menstrual cycle). At that point, baricitinib in a dose of 4 mg/day was introduced, which led to significant improvement of AD (decrease in SCORAD from 67 to 10), but her HS remained active. After 21 months of therapy, baricitinib was stopped due to secondary ineffectiveness for AD. We decided to switch the patient to upadacitinib (30 mg/day), driven by positive results of the phase 2 clinical trial of upadacitinib in HS. At 3 months of treatment, the patient experienced improvement in both diseases (decrease in SCORAD from 52 to 25,4, less frequent occurrence of inflamed HS nodules).

2nd case: A 32-year-old male patient presented a month ago due to the occurrence of painful nodules and abscesses in the inguinal, gluteal, and axillary regions (HS of moderate severity, IHS4=7). The lesions have been occurring throughout the last 10 years, but worsened in the last few months. In addition, he had a history of AD since adolescence with gradual worsening throughout the last 10 years (current SCORAD 59,6). Initially, doxycycline at a daily dose of 100 mg was introduced for HS, and several persistently inflamed nodules have been surgically excised. Systemic treatment with upadacitinib is planned as soon as complete healing of surgical wounds occurs, with the aim of treating primarily AD, but with anticipated simultaneous beneficial effect on his HS as well.

3rd case: A 42-year-old female patient presented to our clinic one year ago with signs of mild AD which was successfully brought into remission with topical bethametasone, 0,1% tacrolimus and emollients. She also reported repeated occurrence of inflamed nodules with purulent discharge in her inguinal and genital regions, which on examination were in the category of mild HS (IHS4=2). For now, her HS is successfully treated with topical clindamycin and antiseptic.

Conclusion: In our 3 patients, severity of AD and HS seem to correlate, which may have significant clinical implications. Each of these diseases brings substantial burden on the patients, and their simultaneous occurrence has even more profound effect on the patients' quality of life, therefore early intervention is of utmost importance.

AD and HS differ in clinical manifestations and pathogenesis, however, JAK inhibitors (especially upadacitinib) appear to be promising as a single treatment covering both conditions.

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Abstract N°: 7013

Chronic Actinic Dermatitis: positive response to treatment using ultraviolet B phototherapy with topical corticosteroids.

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Introduction & Objectives: Chronic actinic dermatitis (CAD) is a rare photodermatosis with characteristic clinical, histological and photobiological features. Its management involves various therapeutic approaches, including topical and systemic therapies. Efficacy of psoralen ultraviolet therapy (PUVA therapy) was previously demonstrated but only few studies have been conducted on the use of narrowband ultra violet B therapy (NB-UVB) in CAD. We aim to demonstrate the efficacy of ultra violet B therapy (NB-UVB) in case of CAD.

Materials & Methods: We report the case of a female patient with CAD, who responded to NB-UVB phototherapy coupled to topical corticosteroids.

Results: A 65-year-old female patient, without any medical history, presented 7 months ago with erythema on sun-exposed areas, including the face, neck, and upper limbs. As the eruption progressed, she developed very pruritic papules on the upper and lower limbs, followed by eczematous lesions on sun protected sites. Physical examination revealed facial erythema sparing upper eyelids, post auricular areas, and submental chin, associated with lichenified papules and plaques on the neck, upper and lower limbs, as well as eczematous lesions on the trunk. The histopathological examination associated to immunohistochemistry of skin biopsy revealed dermal lymphocytic infiltration, CD3 and CD8 positive. The patient was treated by photoprotection, topical corticosteroids, and NB-UVB phototherapy. After 4 sessions only, we noticed the whitening of trunk lesions and a significant improvement of acral lesions and pruritus.

Conclusion: Chronic actinic dermatitis is an uncommun and severe idiopathic photodermatosis. In the absence of a standardized treatment protocol, its management is difficult and often disappointing. Multiple treatment modalities are possible, but photoprotection remains the central component of the management. NB-UVB has already been proven to be effective and safe in several other photodermatosis, but more studies are necessary to evaluate its effects in the management of CAD. Our protocol of NB-UVB with topical corticosteroids seems to be effective, but a continuous follow-up is necessary to judge its long term efficacity and safety.

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Abstract N°: 7025

Assessment of drug causality in suspected cutaneous adverse drug reactions: A comparison between the Bégaud method and the Naranjo score

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Introduction & Objectives:

Cutaneous adverse drug reactions are serious pathologies that can significantly impact a patient's prognosis. Establishing causality between a medication and the onset of a clinical event is crucial for appropriate medical management. However, this causality assessment can be challenging, especially in routine clinical practice.Several imputability methods are available for assessing drug causality in suspected adverse drug reactions.This study aimed to compare two commonly used methods: the French method developed by Bégaud and the American method developed by Naranjo. It also provided insights into the strengths and limitations of each method, helping clinicians make informed decisions regarding drug causality in cases of suspected adverse drug reactions.

Materials & Methods:

We performed a retrospective analysis of all cases of severe drug eruptions reported to the Pharmacovigilance Center of the Faculty of Medicine and Pharmacy of Casablanca between 2019 and 2023 (DRESS, Acute Generalized Exanthematous Pustulosis (AGEP), Stevens-Johnson Syndrome (SJS), and Toxic Epidermal Necrolysis Syndrome (TEN)). We excluded pathologies induced or exacerbated by drug intake, unidentified medication, and vaccinations.

Results:

Between 2019 and 2023, the pharmacovigilance center reported drug eruptions in 116 patients. The median age was 39 years [1-81], and we observed a female predominance (63%). We identified 58 cases of DRESS (50%), 31 of SJS (26.7%), 15 of AGEP (12.5%), 11 of TEN (9.5%), and one case of SJS-TEN overlap (0.9%). Among 116 patients, 442 drugs were taken. The median number of drugs taken per patient was 3 [1-11]. Antibiotics were predominant (34.5%). According to the Naranjo score, 87.5% of effects were considered probable, 9.7% possible, and 2.7% doubtful. According to the Bégaud method, 42.1% of effects were considered "likely," 35.5% probable, 17.4% possible, and 5% doubtful.The two methods gave the same imputability for 164 observations, around 37% of cases (164 out of 442). No concordance (kappa coefficient = 0.062) was observed between the two methods.

Conclusion:

In our study, we found no concordance between the Begaud and the Naranjo methods, consistent with findings in the literature. The Bégaud method is an algorithmic approach that combines chronological, semiological, and bibliographical criteria, allowing for the exclusion of concomitant drugs with the suspect drug. However, it is more time-consuming than other algorithms. On the other hand, the Naranjo score offers a simplified scoring system that is more accessible for clinicians due to its quick application and international usage. However, its utility is limited when certain drugs have not been stopped or when multiple drugs have been taken. Therefore, we suggest using the Naranjo score in emergencies and in cases where few drugs are taken, and the French method in patients with multiple medications and when certain drugs have not been stopped.

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Abstract N°: 7163

Severe cutaneous reaction to nicotine transdermal therapeutic system misdiagnosed as allergic contact dermatitis

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Introduction & Objectives:

Nicotine transdermal therapeutic systems (TTS) are commonly used for smoking cessation. However, they have been documented to induce both irritant contact dermatitis (ICD) and allergic contact dermatitis (ACD).

Materials & Methods:

Results:

Our patient was a 52-year-old male smoker who had recently started using a nicotine TTS (Nicopatchlib ®). He used two patches per day: a 21mg/24h patch measuring 30 cm2 and a 14 mg/24h patch measuring 20 cm2. He placed them next to each other and alternated the placement site every 24 hours. Five days into the treatment, and twelve hours after applying the patches on his back for the first time, he developed severe erythema, burning, and itching. A similar reaction later appeared when he placed the patches on his chest at a site used two days previously. The reaction made of erythema and vesicles was sharply circumscribed to the transdermal patch area. Resolution occurred 48 hours after the patches were removed. The patient was diagnosed with ACD. He stopped the application of nicotine TTS and he was referred to our Department. Based on clinical presentation, we opted for cutaneous patch tests. The manufacturers disclosed that, in addition to nicotine, the patches contain a copolymer of acrylate and vinyl acetate, a copolymer of methyl methacrylate (MMA) and ethylene glycol dimethacrylate (EGDMA). Patch testing was performed with the inner and outer layers of the nicotine patch, nicotine base (5% aq), and the acrylates-methacrylates included in the dental screening series from Chemotechnique (DS-100). The patch tests came back negative to the tested allergens (including MMA and EGDMA), as well as to both sides of the nicotine patch. There was an initial erythematous reaction to the nicotine base which disappeared on the 72-hour reading. The patient continued to use the patches and adhered to the usage instructions without experiencing cutaneous complications.

Conclusion:

The negativity of the patch tests, the confinement of the reaction, and the absence of recurrence did not support the diagnosis of ACD in our patient. The reaction can be attributed to ICD and the local vasodilatory action of nicotine.3 The latter can explain the initial erythema observed at the nicotine base patch area. Although genuine cases of ACD to one of the components of the nicotine TTS have been reported, ICD is more frequent.1,2,4 Patch testing with the same nicotine TTS, the active principle, the adhesives, and the excipients is required in all cases of reaction onset after application of nicotine patches, as marked localized vasodilatation induced by nicotine and intense ICD may mimic ACD.5 Patients should also be checked for compliance with patch application instructions (i.e., rotation of the application site, gentle cleansing, and careful removal) as those help avoid ICD that when severe can facilitate sensitization and therefore lead to ACD. Using two patches per day increased the contact area of the patches with the skin in our patient. This led to a reduction in the available surface that he could use in the following days without placing the patches on a previously used area. The specific spots within each area should also be alternated.

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Abstract N°: 7169

Atopic dermatitis in children with end-stage kidney disease – a 2 case-series

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Introduction & Objectives:

Atopic dermatitis (AD) is the most common chronic inflammatory skin disease in children, proven to be associated with multiple disorders. Genetic studies found a bidirectional positive association between AD and chronic kidney disease (CKD). We report 2 cases of pediatric patients with end-stage CKD (ES CKD) and severe AD. Our aim is to present the clinical evolution of this particular association and signal the importance of treating the associated skin-disease to obtain a marked improvement in quality of life.

Materials & Methods:

Two patients of 10 and 13 years of age diagnosed with ES CKD and severe AD, were selected. The youngest was receiving peritoneal dialysis, whereas the oldest received a renal transplant and was currently under immunosuppressive therapy. Both patients presented with intense pruritus that impacted sleep quality, symptoms of depression and marked cutaneous xerosis. On examination, the lesions were simetrically distributed all over the body, face and scalp, being most prominent in the axillary, latero-cervical and inguinal folds. The transplanted patient presented a subacute eczema of the intergluteal cleft. The lesions consisted mostly of erythematous plaques with thick scale, multiple excoriations with crusting and lichenification. Multiple atopic stigmata and regional variants of AD were present. The diagnosis was made clinically and overall activity disease was assessed and monitored by using SCORAD and DLQI.

Results:

Both patients presented initially with SCORAD>90 and DLQI>20 and raised levels of immunoglobulin E. Background treatment included bath oils and emollients. The reactive therapy was done by administering high-potency corticosteroid (mometasone) and ointments with variable concentrations of urea depending on the severity and location of the lesion. Topical applications of fusidic acid and betamethasone were added to prevent the infection of the excoriated lesions. Systemic antihistaminic therapy was added for the improvement of itch sensation, allowing the interruption of the itch-scratch cycle. The clinical evolution of the patients was favorable: SCORAD decreased to 60 points after 2 months, with a corresponding reduction of DLQI scores. Proactive therapy was subsequently done by gradually replacing corticosteroids with topical calcineurin inhibitors. In these patients, pruritus was accentuated by uremia. The underlying mechanism for the coexistence of the two diseases is not fully understood, but chronic inflammation in AD with activation of T helper-2 cells and Th-17 may promote renal fibrosis.

Conclusion:

AD is the most common skin disorder in patients with CKD, yet treatment is often overlooked until the disease reaches severe manifestations. Our 2-case-series could raise the importance of treating AD earlier in the development of CKD, markedly improving life quality. If early treatment of AD may impact the progression of CKD is yet to be established, but a multidisciplinary approach is imperative.

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Abstract N°: 7220

A Case of Bullous Fixed Pigmented Erythema with Mucosal Involvement in a Child

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Introduction & Objectives:

Skin reactions in children, in the context of drug administration, are a very common reason for consultation. The hypothesis of a drug eruption is often raised. Here, we present a drug eruption, specifically a bullous fixed pigmented erythema with mucosal involvement, in a child.

Case report:

A 10-year-old child, with no significant medical history, presented with a skin rash evolving for 5 days, following an infectious episode the day before, and had taken amoxicillin-clavulanate and ibuprofen. Upon clinical examination, the child was conscious, afebrile, hemodynamically and respiratorily stable, with erythematous rounded macules with central blistering on the limbs, trunk, and back. The examination also revealed fissured and hemorrhagic cheilitis, along with non-purulent bilateral conjunctivitis with palpebral erosions. Histological analysis showed a normoacanthotic epidermis with liquefactive necrosis of the stratum corneum, associated with keratinocytic necrosis without inflammatory exocytosis. The basal layer was vacuolated without blistering detachment, and the dermis displayed marked edema with inflammatory cells, including lymphocytes and histiocytes. After a pharmacological investigation, ibuprofen was identified as the most likely culprit. The child showed significant improvement after discontinuation of all implicated treatments.

Discussion:

Fixed pigmented erythema is a benign drug eruption, and its clinical characteristics in children are similar to those in adults. It initially presents with one or several well-defined, rounded or oval macules that are erythematous or purplish, occasionally evolving into bullae. The lesions can be singular or multiple, affecting the skin or, more rarely, the mucous membranes. The lesions undergo a cyclical evolution. The prognosis is favorable within a few days, with residual pigmentation. The main drugs implicated are analgesics (pyrazoles, paracetamol, aspirin), antibiotics (sulfamides, tetracyclines), antiepileptics (phenytoin, barbiturates, carbamazepine), and NSAIDs. The distinctive feature in our patient was the blistering aspect of the lesions and mucosal involvement in a child.

Conclusion:

The primary challenge in pediatric drug eruptions lies fundamentally in distinguishing them from infectious diseases, particularly viral infections, which are much more common at this age.

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Abstract N°: 7232

Rare case of photoallergy and leukomelanoderma induced by combined hydrochlorothiazide and losartan therapy

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Introduction & Objectives:

Thiazide diuretics can induce photosensitivity sometimes clinically indistinguishable from chronic actinic dermatitis (CAD). We report a case with an exceptional aspect of leukomelanoderma.

Materials & Methods:

Results:

A 65-year-old man, phototype IV, consulted for a pruritic rash evolving for 6 months, diagnosed as CAD and treated without improvement with topical steroids and antihistamines. He was hypertensive and had been treated for 1 year with a combination of hydrochlorothiazide and losartan potassium. Examination revealed hyperpigmentation covered with eczematous plaques on the face and neck. On the backs of the hands, there were also achromic confetti macules, which had recently appeared, giving the appearance of leukomelanoderma. Hydrochlorothiazide-induced photoallergy was suspected. Histology showed psoriasiform acanthosis, parakeratosis, spongiosis and a dermis rich in eosinophilic polymorphs, consistent with eczema. Photoprotection measures were initiated and the former antihypertensive medication was discontinued. In agreement with the cardiologists, a calcium channel blocker was prescribed. After one month, the eczematous plaques regressed and the pruritus disappeared.

Conclusion:

Thiazides can induce photosensitivity by a photoallergic or phototoxic mechanism. In our patient, a delayed-onset photoallergy was observed. This reaction has been reported with antihypertensives containing an angiotensin receptor antagonist and hydrochlorothiazide. The latter is most frequently incriminated. The potential role of losartan in inducing photosensitivity cannot be ruled out, although it has not been well documented. Leukomelanoderma induced by prolonged photosensitivity is very rare, and its mechanism remains largely unknown. It mainly affects phototype II-IV patients, and no cases have been reported in phototype I subjects. Thiazide-induced dermatitis may mimic CAD, but usually disappears after the drug is discontinued. However, the reaction may persist and develop into full- scale CAD, in which case we speak of persistent photosensitivity. Continued sun protection, even after discontinuation of the causative drug, seems essential. Although rarely associated with the appearance of photodistributed telangiectasia, recently prescribed amlodipine is not known to cause this type of reaction. Thiazide-induced photosensitivity should be suspected in patients with photodistributed leukomelanoderma. Discontinuation of the causative drug is necessary, usually resulting in regression of the lesions within a few weeks. Progression to CAD is possible.

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Abstract N°: 7236

Contact dermatitis secondary to the application of Euphorbia helioscopia, commonly known as “euphorbe réveille-matin.”

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Introduction & Objectives:

The spurges (Euphorbia helioscopia L) are toxic plants that naturally grow in the mid-European regions. The most toxic part is the milky sap (latex), which can induce severe contact dermatitis and conjunctivitis if it comes into contact with the skin or eyes. We report a case of a patient presenting with a case of contact dermatitis associated with secondary conjunctivitis following the local application of Euphorbia helioscopia. There are few documented cases of contact dermatitis in the literature attributed to the plants of Euphorbia helioscopia L.

Case report:

The patient is a 57-year-old woman with no notable medical history, particularly no history of atopy or plant use. She presented with alopecic lesions on the eyebrows and preauricular temporal regions, having directly applied Euphorbia helioscopia through friction to treat the alopecic lesions, with no history of oral ingestion. Eight hours later, she noticed the sudden appearance of a poorly defined erythematous and edematous plaque with intense pruritus. The plaque was covered with crust and exhibited oozing at the site of plant application, accompanied by swelling of the upper eyelids and ocular burning. The condition showed improvement following a ten-day prescription of dermocorticoids, along with the application of a protective and reparative barrier cream, corticosteroid eye drops, and healing eye drops. The toxic agent was avoided during the course of treatment.

Discussion:

Contact dermatitis represents a prevalent group of inflammatory skin disorders, manifesting in two distinct forms: irritant and allergic. In irritant dermatitis, the penetration of the pathogenic agent triggers an inflammatory response due to the release of numerous cytokines and chemokines by various types of innate immune cells. The resultant cutaneous reaction typically remains confined to the area in contact with the irritating agent, as observed in our patient. Conversely, allergic dermatitis stems from an adaptive immune response of delayed hypersensitivity, involving the reactivation of previously sensitized T lymphocytes, which differs from the mechanism in our case.

In our patient, the causative agent was Euphorbia helioscopia, commonly known as “euphorbe réveille-matin.” Euphorbia helioscopia is a toxic plant of variable size (10 to 50 cm) containing major bioactive compounds such as triterpenes, euphorbol, euphorbone, hemolytic saponins, and diterpene esters. Traditionally, it has been applied to the scalp, utilizing latex for hair growth, mirroring the practice observed in our patient.

Conclusion:

The toxic component is the milky sap (latex), primarily containing 12-deoxyphorbol, capable of inducing severe skin irritation and conjunctivitis if it comes into contact with the skin or eyes. Oral contact with the plant or latex-soiled fingers leads to a painful burning sensation in the mouth.

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Abstract N°: 7249

Isolated Histaminergic Angioedema: A Study of Five Patients

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Introduction & Objectives:

Angioedema is a common reason for consultation in emergency departments as well as in dermatology. It represents a deeper form of urticaria. Histaminergic angioedema is often associated with superficial urticaria. Here, we report five cases of isolated histaminergic angioedema.

Case report:

Case 1:A 30-year-old patient, with no personal or family history of atopy, presented with a sudden isolated angioedema of both lips, giving a sausage-like appearance, without prior medication use.

Case 2:A 46-year-old patient, with a history of chronic urticaria in a brother, presented with a sudden isolated angioedema of the left lower eyelid, without prior medication use.

Case 3: An 18-year-old patient, with no personal or family history of atopy, presented with isolated angioedema of the upper lip, with a history of taking NSAIDs for dysmenorrhea.

Case 4: A 20-year-old patient, with a history of chronic urticaria in the mother, presented with recurrent isolated angioedema of the nose and eyelids.

Case 5: A 33-year-old patient, with no personal or family history of atopy, and no history of medication use, presented with recurrent episodes of angioedema affecting the lips, external genitalia, and palmo-plantar regions for two years.

In all five patients, there were no superficial urticarial lesions, no laryngeal or respiratory signs, and no abdominal pain associated with the episodes. The qualitative and quantitative assay of C1 esterase inhibitor was normal. The diagnosis of isolated histaminergic angioedema within the context of chronic spontaneous urticaria was established. Patients were treated with antihistamines (Desloratadine 3 tablets/day). They also received therapeutic education regarding the avoidance of NSAIDs, systemic corticosteroids, and stress reduction. The outcome showed significant improvement.

Discussion:

The particularity of our study lies in the isolated nature of histaminergic angioedema, with its diagnosis established based on improvement under second-generation antihistamines and the normality of the bradykinin-mediated angioedema assessment. Isolated histaminergic angioedema represents 6% of chronic urticaria cases according to a study conducted in our department in 2020.

The treatment for recurrent isolated histaminergic angioedema is the same as for chronic spontaneous urticaria. Antihistamines are the first-line treatment. The intake of aspirin or NSAIDs is a classic triggering factor for angioedema attacks in chronic spontaneous urticaria. Corticosteroids are currently considered exacerbating factors. They can induce angioedema attacks in patients and lead to urticaria resistance to antihistamines.

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Abstract N°: 7254

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

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Introduction & Objectives:

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

Case report:

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

Discussion:

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

Conclusion:

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

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Bare lymphocyte syndrome syndrome type 1 (TAP2 deficiency) mimicking necrobiosis lipoidica

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Introduction & Objectives: Bare lymphocyte syndrome type 1 (BLS-1) is a rare autosomal recessively inherited primary immunodeficiency caused by mutations in the transporter associated with antigen processing 1 (TAP1) or TAP2, resulting in the deficiency of MHC-I molecules on the cell surface. The syndrome may be asymptomatic or present with recurrent bacterial infections, chronic respiratory tract inflammation, and granulomatous skin lesions. Herein, we report a case with TAP2 deficiency presented with bronchiectasis and long-term chronic granulomatous ulcer, which was misdiagnosed as necrobiosis lipoidica for years.

Materials & Methods: A 43-year-old male patient presented with multiple large, foul-smelling draining ulcers on plaques consisting of protruded papules and nodules with irregular edges on both his legs and feet, lasting about 15 years. Besides, he had postinflammatory hyper and hypopigmented patches and subcutaneous irregular nodules palpated on both his arms and upper legs. The patient was histopathologically diagnosed with necrobiosis lipoidica at a tertiary hospital three years ago. He had diabetes mellitus and hypertension and was taking oral metformin hydrochloride and ramipril daily for about three years.**

Results: The histopathological examination revealed ulceration and multiple necrobiotic granulomas in the dermis. The patient had frequent coughing since childhood, and chest tomography showed tubular bronchiectasis, multiple millimetric mediastinal lymph nodes, and interstitial infiltration. Mycobacterial growth was not detected in ulcerative plaques and bronchoalveolar lavage fluid. The interferon-gamma release assay was negative. Immunological evaluation of the patient revealed low HLA-ABC expression (63%). Serum beta-2 microglobulin levels were high. A whole exon sequencing analysis was performed, and a homozygous c.1837C>T mutation in the TAP2 gene was detected. The mutation was confirmed with Sanger sequencing. The segregation analysis in family members demonstrated one asymptomatic homozygous sibling and one asymptomatic heterozygous father and sibling. Systemic antibiotic treatments were administered due to polybacterial overgrowths of the ulcers. The patient was given intravenous immunoglobulin at 400 mg/kg monthly for immunodeficiency. Since no regression of ulcers was observed, 40 mg adalimumab every two weeks was added for ulcerative granulomatous plaques to provide the anti-granulomatous effect of anti-tumor necrosis alpha agent. After six months of combination therapy, most ulcers significantly improved and healed.

Conclusion: Chronic granulomatous ulcers of BSL-1 deficiency may resemble clinically and histopathologically necrobiosis lipoidica.**

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Cetuximab-Induced Papulopustular Exanthema: A Case Report

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Introduction & Objectives:

Epidermal growth factor receptor (EGFR) inhibitors are widely used in the treatment of advanced malignancies. EGFR inhibitors have shown to be effective in the treatment of head and neck cancers as well as brain, breast, kidney, pancreatic, bladder, colon, and lung malignancies. While EGFR is important for proliferation and differentiation of the human epidermis and hair follicles, EGFR inhibitors are associated with prominent cutaneous side effects in approximately two-thirds of patients. The most common skin manifestation is papulopustular exanthema in the seborrhoeic areas.

Materials & Methods:

A 52-year-old women** was admitted to our hospital due to** pruritic, tender, symmetrically distributed erythematous papules and pustules on the scalp, face, and upper trunk. The onset of eruption was within six weeks after starting cetuximab, EGFR inhibitor, due to metastatic colorectal adenocarcinoma.

Results:

Skin biopsy obtained from pustule was processed for routine histology and *stained* with *haematoxylin and eosin*.** Histological examination revealed neutrophilic subcorneal pustules and polymorphous infiltrate of the superficial dermis. This side effect of cetuximab was resolved after topical application of high-potency corticosteroids and oral doxycycline 100 mg bid for six weeks. There was no need for cetuximab dose reduction nor therapy interruption

Conclusion:

During the first few weeks of treatment with EGFR inhibitors, papulopustular exanthema is the earliest and very common cutaneous side effect. The recognition and prompt management of cutaneous adverse effects may prevent severe, extensive symptoms, the need for dose reduction, or antitumor therapy interruption.

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Abstract N°: 7414

Acute Generalized Exanthematous Pustulosis induced by airborne Mimosa pollen : A Case Report

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Introduction & Objectives:

Acute Generalized Exanthematous Pustulosis (AGEP) is a rare and severe cutaneous adverse reaction, often triggered by medications. It manifests as a sudden eruption of erythematous and edematous skin covered with multiple non-follicular pustules, accompanied by fever. Here, we present a novel case with uncommon cause and distribution.

Materials & Methods:

Case- A 26-year-old man, with no significant medical history, presented with a skin rash evolving over 48 hours. Examination revealed a rash composed of small non-follicular pustules on an erythematous base, primarily affecting exposed areas such as the neck, arms, and legs. Mucous membranes were unaffected. He had a fever of 38.3°C. Cytobacteriological examination of pus was negative, but peripheral blood analysis showed leukocytosis with a predominance of neutrophils. Skin biopsy revealed intraepidermal spongiotic pustules associated with papillary edema and dermal inflammatory infiltrate. The patient reported recent occupational exposure to an environment with various trees, days before symptom onset. Spontaneous resolution occurred after 12 days of hospitalization. Recurrence of the rash occurred 48 hours after returning to this environment. Patch tests demonstrated a positive reaction only to a 20% mimosa leaf cream preparation, inducing pustules within two days of application, conducted two months later.

Results (and discussion):

Based on clinical and laboratory findings, a diagnosis of AGEP was made, with a EuroSCAR score of 8, implicating mimosa leaves in the cutaneous reaction. Removal from the occupational setting was advised.

AGEP typically presents with a febrile, erythematous, and pustular eruption, preferentially affecting major folds and the trunk. Drug intake is the most common trigger, while exposure to mercury, allergens, or viral infections can also be implicated. Localized forms have been reported, including rare cases induced by textile dyes. Our case is distinctive in its induction of AGEP by airborne mimosa pollen exposure. AGEP prognosis is generally benign, with lesion regression within two weeks.

Conclusion:

This case underscores the importance of recognizing occupational exposures, such as mimosa pollen, as potential triggers for AGEP, necessitating prompt identification and removal from the offending environment.

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Abstract N°: 7506

Subacute cutaneous lupus erythematosus: characteristics, associations, and outcomes of hospitalized patients in a tertiary care center

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Introduction & Objectives:

Subacute cutaneous lupus erythematosus (SCLE), one of the manifestations of cutaneous lupus, is often provoked by extensive photo exposure, and occasionally related to the use of certain medications. It is associated with a relatively high incidence of mild to moderate systemic disease. Through this retrospective analysis, we aimed to examine the clinical characteristics, etiologic factors, serological findings, applied therapeutic modalities, and outcomes in a series of SCLE patients treated in our center.

Materials & Methods:

Our research encompassed 50 in-patients diagnosed with SCLE between October 2018 and October 2023. Included were only incident, histopathologically confirmed cases. The follow-up period was 69 patient-years.

Results:

The mean age at diagnosis was 61.0 ± 12.1 years and the median duration of changes was 5.5 (IQR 3-12.3) months. Thirty-one (62%) patients were women. Drug-induced SCLE etiology was suspected in 10 (20%) cases. The lesions were photo-distributed in 35 (70%) of patients, and 36 (72%) had annular morphologic findings. The most common accompanying skin lesions were discoid lupus lesions, present in 19 (38%) patients, while malar erythema was seen in 3 (6%) patients. Anti-Ro/SS-A positivity was present in 36/47 tested patients (76.6%), whereas only 4/37 (10.8%) tested positive for anti-La/SS-B. Associated autoimmune connective tissue diseases included Sjögren syndrome (n=9, 18%) and systemic lupus erythematosus (SLE) (n=11, 22%), out of which 4 (8%) had pre-existing SLE, 4 (8%) fulfilled the criteria at the moment of SCLE diagnosis, and 3 (6%) did so in the follow-up period. The most common systemic treatment option were antimalarials, prescribed in 45 (90%) patients, followed by systemic corticosteroids in 36 (72%). A complete response was noted in 40/47 (85.1%) patients with available follow-up data, with a median time to resolution of lesions being 3 months (IQR 1-6 months). On the other hand, 21/44 (47.7%) of followed patients experienced a relapse, requiring hospitalization in half of these cases. The median time to relapse was 10.5 months (IQR 5.3-17 months).

Conclusion:

The results of our study match the literature data for the patients' demographic, clinical, and serological characteristics. More than a third of patients had an associated autoimmune disease or a systemic form of the disease. The majority of patients responded well to the treatment of choice, however, nearly half experienced disease relapse, often requiring rehospitalization.

Our data support the stance that SCLE is often a chronic disease and therefore requires continuous monitoring and frequent reevaluations.

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A Complex Dermatological Puzzle in a Graft-Versus-Host Disease Case

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Introduction & Objectives:

Graft-versus-host disease (GvHD) is a complication that often arises after allogeneic hematopoietic stem cell transplant (HCT), where the immune cells from the donor mistakenly attack the recipient's tissues. In its acute phase, GvHD commonly affects the skin, gastrointestinal tract, and liver, with potential involvement of other organs such as the lungs, kidneys, eyes, and hematopoietic system.

The skin manifestations of acute GvHD usually begin with a maculo-papular rash that appears mainly on the palms, soles, shoulders, and neck. This rash can spread extensively and may progress to severe blistering lesions similar to toxic epidermal necrolysis (TEN).

Materials & Methods:

We report a case of a 35-year-old female diagnosed with interstitial pneumopathy and acute myeloid leukemia one year prior, who underwent a medullary transplant in 2023. The patient presented with a generalized eruption characterized by round, pruritic erythematous-squamous plaques, poorly differentiated, with fine scale involving the face, trunk, and extremities.

A thorough review of medications, including over-the-counter drugs, vitamins, and supplements, revealed no incriminating agent for the patient's clinical manifestations.

Results:

A skin biopsy was performed, and histological examination confirmed the diagnosis of Grade II Host vs Graft Disease, characterized by vacuolization and dyskeratotic bodies.

Subsequent blood investigations, inclusive of a complete blood count, revealed elevated C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), and liver enzyme levels.

The patient was initiated on systemic steroid therapy, specifically methylprednisolone at a dosage of 2 mg/kg/day, with a gradual tapering regimen. After two weeks of treatment, both the skin lesions and laboratory parameters demonstrated notable improvement

Conclusion:

As the transplant patient population expands, medical practitioners are increasingly confronted with diagnostic challenges. Among these challenges, the consideration of graft-versus-host disease (GVHD) as a potential differential diagnosis holds paramount importance.

This case highlights the correlation between disease severity grading and treatment modalities, as well as the challenges in distinguishing between graft-versus-host disease (GVHD) and allergic reactions, particularly when discontinuing essential medications crucial for patient improvement.

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Abstract N°: 7593

Povidone-iodine induced contact dermatitis in a child

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Introduction & Objectives:

Strong iodine solutions are corrosive and can lead to blistering and skin necrosis, commonly known as chemical burns or irritant contact dermatitis. Consequently, iodine has been replaced by substances called iodophores. Povidone iodine is one such iodophore that exhibits low irritant properties despite its preserved antiseptic efficacy, making it advocated as a non-irritating and non-toxic compound. Here, we report a case of irritant contact dermatitis caused by the so-called non-irritating compound.

Materials & Methods:

Results:

A 12-year-old patient with a history of allergic rhinitis, who underwent surgery for femoral dysplasia during which betadine/povidone-iodine was used as the antiseptic. On the first post-operative day, she experienced intense burning sensations and pain at the surgical field. Clinical examination revealed diffuse erythema covered with highly pruritic vesicles and pustules. On systemic examination, all the vitals were within normal range, and there were no clinical signs or symptoms of any systemic diseases. The patient was treated with local care and topical antibiotics for 10 days without improvement. Due to the persistence of the lesions, their pruritic nature, and their localization clearly limited to the area of surgery, the diagnosis of contact dermatitis was suspected. The patient was treated with topical corticosteroids and discontinued the use of povidone iodine, resulting in complete regression after one week.

Conclusion:

Although povidone iodine is classified as a non-irritant antiseptic, it is not entirely devoid of corrosive action. Several cases of post-surgical contact dermatitis due to povidone iodine have been reported. This type of dermatitis can develop in the surgical area or at distant sites exposed to povidone iodine during the surgical intervention. Humidity and occlusion are necessary for contact dermatitis to develop after surgical interventions. Diagnosis is generally based on clinical manifestations, exposure history, lesion site, and patch test results. It is recommended that doctors be aware of the potential for severe irritant contact dermatitis caused by povidone-iodine and similar antiseptic solutions. Proper precautions should be taken to prevent such adverse reactions, especially in individuals with a history of allergic conditions or sensitivities.

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Abstract N°: 7959

Barzolvolimab shows profound efficacy and favorable safety over 52 weeks in patients with chronic spontaneous urticaria

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Introduction & Objectives:

Mast cells (MCs) are key effector cells in chronic spontaneous urticaria (CSU). We previously reported clinically meaningful and statistically significant efficacy data and a favorable safety profile after 12 weeks of treatment with barzolvolimab (anti-KIT monoclonal antibody) in a Phase 2 CSU study (NCT05368285) in patients whose disease was refractory to antihistamines, including patients with prior biologic experience. Here we report efficacy and safety results through Week 52 of the study.

Materials & Methods:

In this double-blind, placebo-controlled trial patients were randomized to receive subcutaneus (SC) barzolvolimab at 75mg Q4W, 150mg Q4W, 300mg Q8W or placebo during a 16-week placebo-controlled treatment phase followed by 36-weeks of active treatment, and 24-weeks of follow-up. Placebo and 75mg Q4W dose groups were re-randomized to 150mg Q4W or 300mg Q8W during active treatment period. Assessments included UAS7, HSS7, ISS7, and safety.

Results:

A total of 208 patients were enrolled and the mean baseline UAS7 score ranged from 30.1 to 31.3 across the treatment groups. For the two groups that received either 150mg or 300mg barzolvolimab throughout the 52W trial, the mean change (SD) in UAS7 from baseline at week 52 was -27.2 (10.6) and -24.9 (12.1) for the 150mg and 300mg groups, respectively. Deepening of response was observed, with complete response rates (UAS7=0) of 71.1% and 52.3% at 52 weeks compared to 51.1% and 37.5% at 12 weeks, in the 150mg and 300mg dose groups, respectively. Patients who initially received 75mg or placebo and were re-randomized to 150mg or 300mg also experienced comparable benefit. A similar pattern of response was observed in the subset of patients who were omalizumab experienced, including those who were omalizumab refractory. Barzolvolimab was well tolerated with 52 weeks of exposure and most adverse events were low grade.

Conclusion:

Barzolvolimab at 150mg Q4W and 300mg Q8W both demonstrated clinically meaningful improvement in UAS7, with a deepening of response over 52 weeks in patients with antihistamine refractory CSU, including patients who had received prior omalizumab. In addition, barzolvolimab continued to be well tolerated with a favorable safety profile. These results support the ongoing Phase 3 studies, EMBARQ CSU1 and EMBARQ CSU2 in biologic naïve and experienced patients with CSU refractory to antihistamines.

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