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Abstract N°: 56

Contribution of Trichoscopy in the diagnosis of discoid lupus: study of 14 cases

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Introduction & Objectives: Chronic lupus erythematosus is a common cause of cicatricial alopecia. Scalp involvement is observed in 34 to 56% of chronic lupus erythematosus according to series and cicatricial alopecia is the initial involvement. in more than half of the patients. It remains isolated in 11 to 20% of patients.

The diagnosis can be difficult. We will see the diagnostic value of trichoscopy.

Materials & Methods: Analytical descriptive study having focused on 14 patients, over a period of 3 years. The diagnosis of lupus discoid was clinical, and confirmed histologically. All the patients benefited from a trichoscopic examination by the Dinolite digital trichoscope, the results are entered on the EPI info, and compared by X^2.

Results: 14 patients, 6 men and 8 women (including a 6-year-old boy and an 8-year-old girl). Average age: 39 years old. Average evolution: 39.37 months. Forms: single patch: 5 cases, multiple patches: 9 cases. The seat: the forehead was the most frequent seat 6 (43%), the temporal region 2 (14%), occipital and fronto-vertical each represented 3 (21%). The trichoscopic examination found: the disappearance of the follicular orifices (cicatricial), and the follicular plugging were the most frequent, they represented respectively, 100%, 87.50% of the cases. Peripilar erythema accounted for 62.50% of cases. The arborizing vessels and the red dot each accounted for 50% of the cases. Tubular scaling and white dot were less frequent, and represented respectively 37.50. 25% of cases.

Comparison with lichen planopilaris found: arborizing vessels ($P<0.001$), follicular plugging ($P=0.0018$) were in favor of discoid lupus of the scalp.

Conclusion (and discussion): the diagnostic contribution of trichoscopy in the diagnosis of cicatricial alopecia is indisputable. The most frequent trichoscopic signs of discoid lupus were: scarring, peripilar erythema, arborizing vessels, and the red dot which corresponded to extravasation of red blood cells in the hair follicle. The results of our study were consistent with those of the literature; the study of Bruna, and Belond et al, who found: arborizing vessels, follicular plugging as pathognomonic signs of discoid lupus. In conclusion, trichoscopy is a non-invasive, promising tool for the diagnosis of cicatricial alopecia; like discoid lupus.

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Abstract N°: 71

Multiple dermatofibromas: to concern or not? A systematic review of 106 cases

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Introduction & Objectives:

Multiple dermatofibromas (DF) are rare benign fibrohistiocytic lesions characterized by asymptomatic hyperpigmented nodules. Multiple DFs have been associated with underlying diseases, especially in patients with altered immune function, including autoimmune diseases like systemic lupus erythematosus (SLE) and human immunodeficiency virus (HIV), immunosuppressive therapies, or both.

In this research, we attempted to systematically review the case reports of multiple DFs (more than one DF lesion) to clearly describe this rare condition. This study investigated several clinical aspects of multiple DFs like associated comorbidities and drugs, histopathology, treatments, and outcomes. Characteristics of different variants of multiple DFs, including multiple eruptive dermatofibromas (MEDFs), multiple clustered dermatofibromas (MCDFs), and giant combined dermatofibroma (GCDF), have also been discussed in detail.

Materials & Methods:

Here, we designed a systematic review by an electronic search of PubMed and EMBASE databases to select case reports of multiple DFs (29 May 2021). From 2145 initially found articles, 96 studies (106 cases) were included. Each eligible article was reviewed by two dermatologists to extract the following variables: age, sex, dermatofibroma type, duration of the disease, characteristics of the lesions, associated comorbidities and drugs, the time interval between associated disease and drug intake and DF presentation, histopathologic findings, immunohistochemistry (IHC) factors, treatment, outcome, recurrence, follow up duration. We classified multiple DFs (>1 DF lesion) into four subtypes and separately investigated clinical characteristics of each one: MEDFs (n=45), multiple (n=41), MCDFs (n=18), and giant combined (n=2).

Results:

The patients' mean age was 38.33± 14.74 years. The majority of them were female (61.3%). The lesions were commonly on the trunk and extremities (36.8%). MEDFs (n=36) has the most rapid disease onset (1.97± 6.62 years). Immunosuppression induced by either HIV (10.3%) and hematologic malignancy (9.4%) or immunosuppressive drugs (23.4%) (prednisolone, either alone or in combination with other immunosuppressive drugs) along with SLE (19.8%) were the most observed associations. Although, 66.7% of the MCDFs patients were healthy individuals. As the management, surgery and follow-up were the preferred options. Most of the cases showed neither resolution nor development of new lesions in follow-up.

Conclusion:

In conclusion, this study has shown that most multiple and MEDFs patients are middle-aged women with hyperpigmented nodules or papules located on the trunk and extremities. MEDFs patients experience a very fast disease presentation. About 83% and 91% of multiple DFs and MEDFs cases suffer from an underlying condition, respectively. HIV and hematologic malignancies are significantly associated with MEDFs. During follow-ups, most of the lesions had neither resolved nor worsened. MCDFs patients are young women with clustered papules and

plaques on the lower extremities. Most of them are otherwise healthy individuals for whom the number of lesions does not increase. The findings of this study suggest that multiple DFs and MEDFs signify immune dysregulation. Therefore, further evaluations should be initiated to identify the cause of these conditions in patients without a known associated disease.

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Abstract N°: 101

Team-based learning (TBL): a novel approach to teach the cutaneous manifestations of systemic disease

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Introduction & Objectives:

Despite skin complaints constituting a relative majority of primary care presentations within the United Kingdom 1, junior doctors often express a low confidence in describing such lesions. Therefore, as medical students receive little exposure to dermatology throughout undergraduate education 2, it is important educators explore the most efficient teaching strategies that may confidently prepare new doctors. This may be achieved by integrating alternative educational approaches into the dermatology and rheumatology undergraduate curriculum, such as team-based learning (TBL); a learner-centred approach to education that promotes problem-solving, discussion and reflection.

The aim of this study was to evaluate the use of TBL within dermatology and rheumatology undergraduate education, achieved through three objectives: [1] use TBL to improve students' confidence in identifying, investigating and managing systemic diseases that may present with cutaneous manifestations; [2] use TBL to improve students' confidence in describing cutaneous lesions; and [3] analyse the acceptability of TBL within dermatology and rheumatology undergraduate education.

Materials & Methods:

Twenty penultimate and final-year medical students completed a questionnaire before and after a TBL session on the cutaneous manifestations of systemic disease. Students' confidence in the subject knowledge and their ability to describe skin lesions was analysed using Likert scales. Open-ended questions helped to determine acceptability.

Results:

Statistically significant positive changes in students' confidence was found in describing skin lesions, and in identifying relevant signs, symptoms, investigations and management options for example systemic diseases presenting with cutaneous manifestations ($p<0.013$).

TBL was perceived to be an effective way to teach dermatology and rheumatology, with many students appreciating an opportunity for open discussion between peers.

Conclusion:

The authors conclude that learner-centred educational interventions provide an efficient and acceptable route to cultivate medical students' knowledge, confidence, soft skills and skin examination approach. Therefore, TBL should be considered a suitable adjunct to traditional teaching methods used within the undergraduate dermatology and rheumatology curriculum.

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Abstract N°: 107

A Case of Pemphigus Foliaceus in a 40-Year-Old FemaleSuccessfully Treated with Doxycycline and Prednisone Combination Therapy

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Abstract

Background: A number of studies confirm the efficacy of doxycycline in conjunction with topical and oral steroids as a first treatment strategy in bullous pemphigoid. However, this treatment regimen and its use with pemphigus foliaceus is less documented.

Observation: Here, we present a 40-year-old female with multiple pruritic occasionally painful vesicles, papules, and plaques in a circinate pattern on seborrheic areas, progressing to erosions and scales. Clinical findings led to the diagnosis of pemphigus foliaceus (PF). Initial treatment with prednisone and clobetasol ointment, however, did not fully suppress blister formation and healing of erosions. Skin punch biopsy revealed a subcorneal split and intracorneal neutrophilic infiltrates, while enzyme-linked immunoassay (ELISA) revealed elevated anti-desmoglein 1 (Dsg1), consistent with PF. Doxycycline was then added to the previous regimen, resulting in remission.

Key Message: This study provides evidence that the use of doxycycline, in conjunction with oral and topical steroids may be effective in the management of PF and adds to the list of medications that may be used as a steroid-sparing agent in PF.

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Abstract N°: 199

Melanotic lupus erythematosus: descriptive series of 3 cases and systematic literature review

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Introduction & Objectives:

Melanotic lupus erythematosus (LE), is a rare, newly described type of chronic cutaneous lupus erythematosus (CCLE), clinically characterized by photodistributed lesions presenting de novo as brownish or greyish macular reticulate pigmentation without atrophy, scarring, or telangiectasia. Only 28 cases have been reported in the literature, to our knowledge. The aim of this review is to synthesize existing data on the epidemiologic, clinical, pathologic, and immunologic features of melanotic LE.

Materials & Methods:

Based on 3 observed cases, we performed a systematic review using PUBMED to identify eligible articles. Eight articles fulfilled the eligibility criteria and were included in the qualitative synthesis.

Results:

Two women aged 33 and 65 years and one man aged 59 years consulted for facial hyperpigmentation. The average duration of evolution was 2 years. All patients were phototype IV. On examination, the lesions appeared as diffuse, poorly limited hyperpigmented patches in 2 cases, and as isolated, well-limited hyperpigmented macules in 1 case. The hyperpigmentation was homogeneous greyish-black in 2 cases and brownish reticulated in 1 case. There was no erythema, scaling or atrophy. Pathology allowed the diagnosis of cutaneous LE in all cases. Antinuclear antibodies (ANA) were positive in 1 case. The systematization workup was negative in the other cases. The patients were put on photoprotection, hydroxychloroquine and topical steroids with good evolution after 6 months of follow-up. A systematic review of the literature allowed us to collect 28 patients with melanotic LE, with a mean age of 57.7 years. A female preponderance was observed (female-to-male ratio of 2.11). The majority of patients were of Indian origin (43.75%). The lesions presented either as solitary and localized, poorly limited, round, or oval patches (50%) or as a more diffuse or generalized, sometimes reticulated, hyperpigmentation (50%). The rate of the patients diagnosed with SLE was 14%, but only 1 patient had severe SLE. The rate of positive ANA in patients was 10.71%. Hydroxychloroquine and topical steroids were the most commonly used treatments. A complete resolution of the lesions was observed in 27.27% of cases. No patient showed atrophy or scarring of the skin.

Conclusion:

Melanotic LE mainly affects darker phototypes. It is characterized by a later age of onset. It can have 2 clinical patterns: a diffuse pigmented patches or a well circumscribed single or multiple macules. Pigmentation varies from brown to slate grey. The diagnosis is based on a clinicopathological correlation. The main differential diagnoses are discoid lupus, from which it differs by the absence of atrophy and scarring, and lichen planus pigmentosus. Melanotic LE has a good prognosis and is rarely associated with systemic involvement. Treatment is based on photoprotection, topical steroids and hydroxychloroquine ensuring a good response in most cases.

Abstract N°: 250

Paraneoplastic dermatomyositis revealing a breast carcinoma

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Paraneoplastic dermatomyositis revealing a breast carcinoma

Introduction & Objectives

Dermatomyositis is a rare disease associating muscle inflammation and a cutaneous syndrome.paraneoplastic origin is founding 6 to 60% of cases.The most frequently associated tumors are ovarian and mammary tumors in women and bronchial, digestive and nasopharyngeal tumors in men. Although breast carsinoma has been reported in different observation,an atypical breast carcinoma has been rarely published.

We report a case of dermatomyositis revealing an atypical breast carcinoma **

Materials & Methods:

70 years old woman with consulted the dermatology department for a cutaneous-muscular syndrome that had been developing for 6 months.The clinical examination revealed heliotropic erythema, erytherma in a “V” shape on the neckline, the limbs, above the metacarpophalangeal joints, a flagellated erythema on the back, peri-ungual erythema, manicure sign was positive. Capillaroscopy showed the presence of megacapillaries. The patient present also a proximal muscular weakness with fatigability.

The biological workup showed elevated muscle enzymes.The ENMG showed a predominantly proximal myogenic syndrome and the diagnosis of dermatomyositis has been made.

The search of neoplasie have yelded to the discover of subcutaneous nodule in left breast, the mammography showed mastitis and an irregular posterior attenuation zone with a malignant appearance classified as 5ACR. Bone scan, thoracic-abdominal-pelvic and cerebral computed tomography (CT) did not reveal any secondary localizations.

Breast biopsy was in favor of infiltrating carcinoma of non-specific type classified as IISBR. Hormone receptor testing : PR+,OR+, HER2 1+ and Ki67 5%

The patient was put under prednisone 1mg/kg/d while waiting for neoadjuvant chemotherapy followed by mastectomy

Results:

Different types of cancer are found in association with dermatomyositis, and practically all locations are possible. However, gynecological cancers seem to be more particularly represented, including breast cancers.

Conclusion:

The association between DM and malignancy is a well-established

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Abstract N°: 446

A clinical conundrum: disfiguring facial plaques and nodules in a man with paraproteinemia

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Introduction & Objectives:

Materials & Methods:

Results:

We present the case of an 88-year-old male with a 4-month history of progressive, non-pruritic violaceous plaques and nodules affecting the head and neck.

An initial biopsy performed by the referring physician demonstrated florid acute suppurative folliculitis with elements of chronic folliculitis and a granulomatous response to follicular rupture. Fucidin cream and flucloxacillin was trialled without improvement.

Examination revealed several large, indurated plaques and nodules on the forehead, cheek, neck and right thigh. There were focal areas of ulceration. Eczematous patches with follicular accentuation were present on the trunk and limbs. The favoured clinical diagnosis was cutaneous lymphoma with differentials including sarcoidosis, extensive (facial and extra-facial) granuloma faciale, atypical erythema elevatum diutinum and progressive nodular histiocytosis.

Skin biopsy was repeated as it was felt the initial histopathology did not correlate with clinical findings. Subsequent histology was similar and favoured a florid insect bite reaction. The possibility of a paraneoplastic bite-like reaction was raised.

Routine full blood count, biochemistry and inflammatory markers were normal. Serum immunoglobulins were raised, with electrophoresis demonstrating an IgG-Kappa paraprotein of 13g/L and evidence of immune paresis. Skin scrapings and tissue culture were negative. A CT chest/abdomen/pelvis did not reveal radiological features suggestive of malignancy. A bone marrow aspirate demonstrated a slight increase in plasma cells, thought to be reactive.

At the time of submission, the lesions are increasing in size and number. He is currently undergoing a trial of prednisone with a plan to transition to dapsone. We present this case as an interesting diagnostic dilemma and review cutaneous associations of paraproteinaemias.

Conclusion:

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Abstract N°: 467

Esophageal evaluation in systemic sclerosis and morphea by clinical, endoscopic, manometric and pH metric features.

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Introduction & Objectives: Systemic sclerosis (SSc) is a generalized disorder of unknown etiology affecting the connective tissue of the body. It affects the skin and various internal organs like gastrointestinal tract, lungs, heart and kidneys. Gastrointestinal tract involvement is seen in almost 90% of the patients. The esophagus is the most frequently affected part of the gastrointestinal tract. Esophageal smooth muscle becomes atrophied and replaced by fibrous tissue leading to severe motility disturbance of distal esophagus. Esophageal motility disturbance classically manifests as a reduced lower esophageal sphincter pressure (LESP) and loss of distal esophageal body peristalsis. Consequently, SSc patients with esophageal involvement have impaired acid clearance which may be complicated by erosive esophagitis and eventually by Barrett's esophagus and esophageal adenocarcinoma.

Morphea, also known as localized scleroderma, is characterized by predominant skin involvement, with occasional involvement of subjacent muscles and usually sparing the internal organs. The involvement of esophagus in morphea has been studied very scarcely. The data regarding esophageal involvement in morphea is meager. The proposed study will investigate the esophageal involvement in the two forms of scleroderma (systemic and localized), compare the same and address any need of upper gastrointestinal evaluation in morphea (localized scleroderma) patients.

The objectives of this research were** to study and compare the clinical, endoscopic, manometric and pH-metric features of esophageal involvement in SSc and morphea.

Materials & Methods: 56 and 31 newly and already diagnosed cases of SSc and morphea respectively were taken up for the study. All the patients were inquired about the dyspeptic symptoms (Heart burn and/or acid regurgitation and/or dysphagia). Upper gastrointestinal endoscopy, esophageal manometry and ambulatory 24-hour pH monitoring were performed in 52, 47 and 41 patients of SSc; and 28, 25 and 20 patients of morphea respectively.

Results: Esophageal symptoms were present in 39 cases (69.6%) of SSc which were mild in 22 (39.3%), moderate in 14 (25%), severe in three (5.3%); and only in four cases (7.1%) of morphea all of which were mild in severity. Reflux esophagitis was seen in 17 cases (32.7%) of SSc and only two cases (7.14%) of morphea. Manometric abnormalities were seen in 32 cases (68.1%) of SSc and none in morphea. Ambulatory 24-hour esophageal pH monitoring documented reflux in 33 cases (80.5%) of SSc and no such abnormality was documented in morphea.

Conclusion: While the esophageal involvement in SSc is frequent, no such motility disorder is seen in morphea. Every patient of SSc needs a meticulous upper gastrointestinal evaluation whether symptomatic or not. However, such a routine evaluation in morphea seems to be unjustified.

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Abstract N°: 482

Cutaneous manifestations and treatment of arsenic toxicity: A Systematic Review.

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Introduction:

Arsenic is a naturally occurring carcinogenic heavy metal, often found in drinking water sources tapped from tube wells. Chronic arsenic toxicity occurs in individuals exposed to lesser quantities over longer periods of time but may be present in developed countries where there may be occupational exposure and medicinal ingestion.

Objective:

To identify all dermatological manifestations, mode of exposure and treatments in adults with suspected arsenic exposure reported in current literature.

Materials & Methods:

A systematic review was conducted following the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines and registered, PROSPERO CRD42020203145. We searched through EMBASE and Medline via OVID. Data were extracted to capture items and a narrative synthesis was used to summarise sample size, recruitment method, study design, types of exposure, country of exposure, cutaneous presentations and treatment options. Risk of bias assessment was done by two independent reviewers using the Joanne-Briggs Institute critical appraisal tool on Covidence.

Results:

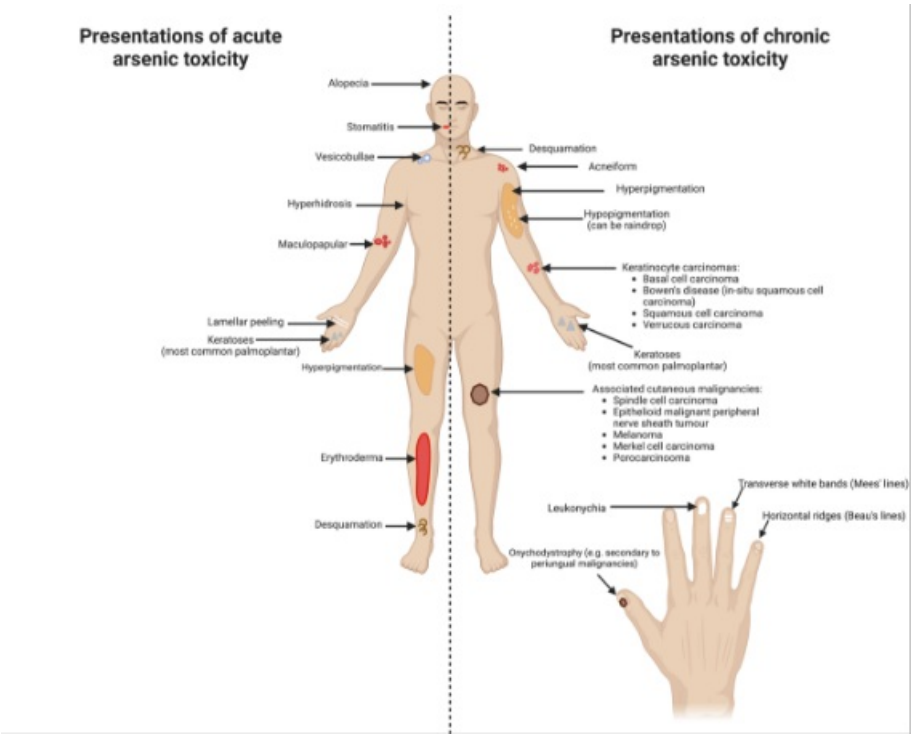
Literature was searched from 1946 to 25th August 2020. We included 72 articles composed of case reports or series (n = 59), case-control studies (n = 2), cross-sectional studies (n = 9) and cohort studies (n = 2). Acute arsenic toxicity may be commonly associated with palmoplantar keratosis and lamellar peeling, involving nonspecific parts of the body. Multiple studies suggest dimercaprol chelation therapy may be used for acute and chronic arsenic toxicity. Meanwhile, chronic arsenic toxicity is associated with keratoses, raindrop pigmented changes and Mees' lines in the nails. Furthermore, there was a wide spectrum of cutaneous carcinomas highlighted in extensive studies consisting of case reports, retrospective and cross-sectional studies of large sample population size. Chronic arsenic toxicity was associated with basal cell carcinoma, squamous cell carcinoma and in-situ squamous cell carcinoma. However, there were some studies that demonstrated associations with rare cancers such as Merkel cell carcinoma and porocarcinoma. Multiple studies of variable level of evidence suggests that standard management of keratinocyte carcinomas may still be effective in those with chronic arsenicism. These included: topical 5-fluorouracil, imiquimod, cryotherapy, Mohs micrographic surgery, and conventional excision with/without skin graft. Figure 1 summarise all findings in selected studies.

Conclusion:

There is a broad spectrum of dermatological findings for acute and chronic arsenic toxicity reported in current literature. Common stigmata of chronic arsenic toxicity including keratoses and pigmentary changes, as well as potential concurrent keratinocyte tumours. Individuals diagnosed in developed nations may have been environmentally exposed in endemic countries at a younger age, but also through ingestion of traditional and

Western medicines, as cutaneous presentations may manifest acutely with other systemic symptoms. However, uncommon manifestations reported in this study are only inferred based on history to be weakly associated with arsenic exposure due to multiple low-level evidence studies identified, and treatment efficacy reported are not supported by studies of higher level of evidence.

Figure 1. Summary of associated acute and delayed cutaneous manifestations of arsenic toxicity in the systematic review.



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Abstract N°: 492

Crystals in the skin: an atypical cutaneous manifestation secondary to hematologic neoplasia

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Introduction & Objectives:

Crystal-storing histiocytosis is a rare manifestation in which intracytoplasmic crystal deposition in histiocytes is observed histologically. These deposits are usually crystallized monoclonal immunoglobulins. It is a secondary process, so its detection must be associated with the screening for an underlying primary disease. We want to report a case of crystal-storing cutaneous histiocytosis secondary to multiple myeloma

Materials & Methods: Clinical case description using patient's medical record

Results:

A 59-year-old man presented with a skin lesion located in the right retroauricular region and another in the frontal region of two years of evolution associated with constitutional syndrome for four months. The patient was being followed by the Hematology service for monoclonal gammopathy of uncertain significance IgG-Kappa of twelve years of evolution. Physical examination revealed a 3 cm diameter mass, skin-colored, polylobulated to the touch, with a hard consistency. At retroauricular level there was a mass similar to the frontal mass. The last hematological control PET/CT scan, performed a week before the consultation, showed bone, subcutaneous and muscular lesions with pathological metabolism, including the dermal lesions described. An incisional biopsy of the frontal lesion was performed due to the suspicion of multiple myeloma infiltration. Histologically, there was a perivascular dermal infiltrate with histiocyte predominance, with abundant multinucleated giant cells, perivascular plasmacytosis with mild atypia and spiculated crystals inside some KAPPA light chain positive histiocytes. The definitive diagnosis was crystal-storing histiocytosis secondary to stage III-A IgG Kappa multiple myeloma. The patient started polychemotherapy with daratumumab, bortezomib, lenalidomide and dexamethasone.

Conclusion:

Crystal-storing histiocytosis is an uncommon condition secondary mainly to multiple myeloma. Although it can affect any organ, its presentation in the skin is less than 5%, being the kidney the most frequently affected organ. Although it has also been described secondary to other hematologic malignancies such as lymphomas and autoimmune diseases such as Sjögren's disease, the most frequent underlying disease is multiple myeloma. Its appearance can precede the development of this neoplasm by months or years, as was our case, so an early diagnosis of this entity could allow early diagnosis of hematologic neoplasm or tighten controls in patients under follow-up for monoclonal gammopathy of uncertain significance.

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Abstract N°: 557

Malignancy-associated Sweet's Syndrome: clinicopathological characterization and different clinical patterns in a cohort of 32 patients

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Introduction & Objectives:

Sweet's syndrome (SS) has been associated with neoplasms in 20-40% of cases. However, the clinical presentation and evolution may vary depending on the associated neoplasia. To date, few data exists regarding the distinction of the different clinical patterns of paraneoplastic-SS. Our objective has been to describe the clinicopathological characteristics of malignancy associated SS associated with neoplasia (MA-SS), to identify the variables associated with MA-SS, and to determine the differences between patients with acute myeloid leukemia (AML) and myelodysplastic syndrome (MDS).

Materials & Methods:

Unicentric retrospective cohort of 93 patients with SS. Patients who fulfilled Curth's postulates for MA-SS were included in this study between 2001-2021.

Results:

From an initial cohort of 93 patients with SS evaluated, 32 patients (34.4%) had a MA-SS. AML was the most frequent neoplasm (16/93), followed by MDS (7/93). The factors that were associated with MA-SS were: male sex ($p=0.006$), fever ($p=0.034$), increased ESR ($p<0.001$), anemia ($p<0.001$) and thrombocytopenia ($p<0.001$). In patients with AML, at the time of SS presentation, 87.5% had de novo AML and 81.3% had active disease. 31% of the MA-SS had ≥ 2 relapses in the evolution. Specifically, 18.8% (3/16) of SS with AML, and 57.1% (4/7) of SS with MDS. SS with MDS required more frequently prolonged corticosteroid courses and/or additional therapies. During follow-up, three MDS patients with SS were diagnosed with VEXAS syndrome.

Conclusion:

The presence of fever, male gender, leukopenia, anemia, and thrombocytopenia should alert to the possibility of SS-AN. An acquired autoinflammatory syndrome should be ruled out in patients with SS or MDS with a recurrent course. A minimum follow-up of 6 to 12 months is recommended to exclude underlying hematologic malignancies.

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Abstract N°: 577

Different faces of cutaneous sarcoidosis in a patient with pulmonary involvement

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Introduction & Objectives:

Sarcoidosis is a multiorgan, granulomatous disease of unknow origin that affects both males and females. It occurs at any age but mostly in between 2nd and 4th decade. Although present in 20-35% of patients with systemic sarcoidosis cutaneous involvement is oftentimes initial clinical manifestation of the disease. Skin lesions are divided into specific and nonspecific. First include macules, papules, nodules, plaques and scar sarcoidosis and they are more often than ulcerative, psoriasiform, verrucous and erythrodermic type lesions. Erythema nodosum as a reactive panniculitis is the most common nonspecific lesion. The clinical differential diagnosis is thus broad taking into consideration various cutaneous manifestations of the disease.

Materials & Methods:

We report a 67-year-old female patient who came to our department after being diagnosed with sarcoidosis involving skin and lungs a year before. Her skin lesions started as a single subcutaneous plaque on the forearm and around the elbow. Upon her first visit at another institution only a cytopuncture of the plaque was done and the diagnosis of granulomatous inflammation was made that along with mediastinal lymphadenopathy confirmed with CT scan was enough to make a diagnosis of sarcoidosis. She started with 40 mg of prednisone with complete regression of pulmonary lesions over time but without any effect on her skin lesions.

Results:

She reported worsening of skin condition in a way that her forearm has gradually become more indurated. When she came over, she had stiff and firm right forearm with some erythematous maculopapules covered with scales. Stiffnes of her forearm was reminiscent of morphea. Around her left elbow she had a firm plaque and proximal part of her right leg was palpatory firm as well. The histological examination of a biopsy from her right forearm, done primarily to exclude the diagnosis of morphea, showed numerous sarcoidal granulomas within papillary, reticular dermis and subcutaneous fat. Epidermis was covered with some parakeratotic scales. We started methotrexate at a dose of 15 mg and folic acid 5 mg weekly along with 2,5 mg of prednisone. After 2 months a slight regression was appreciated regarding the induration of her forearm but in the meantime the patient developed an atrophic plaque on her left leg. A new biopsy showed sarcoid granulomas at the dermosubcutaneous junction. The induration on her right leg, present since the first visit histologically showed signs consistent with the diagnosis of morphea. There were no signs of granulomas within that biopsy.

Conclusion:

Sarcoidosis may cause different cutaneous manifestations within a single patient. Our patient, who can be classified as having polymorphous cutaneous sarcoidosis, most likely initially developed plaques within deep reticular dermis or subcutaneous fat of the forearm that coalesced over time forming stiff, morphea like clinical picture along with maculopapules. Besides, the patient also had a single plaque around her elbow, atrophic plaque on her left leg with sarcoidal granulomas histologically and induration on her right leg that was histologically consistent with the diagnosis of morphea. Whether the patient has two overlapping condition needs to be seen. It is well known that plaque sarcoidosis can be recalcitrant to systemic corticosteroid therapy as is the

case in our patient. Since she is taking only a couple of months methotrexate at the moment its therapeutical efficacy cannot be discussed.

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Abstract N°: 660

an analysis of iris pattern as a risk factor for skin cancer development in renal transplant recipients

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Introduction & Objectives:

Renal transplant recipients (RTR) are at increased risk of keratinocyte skin cancers with a tendency to have multiple, aggressive and difficult to treat tumours. The eye and the skin share the same embryological ectoderm. Iris pattern has recently been reported as a predictive risk factor for skin cancer in non-immunosuppressed Southern European1 and Irish populations2.

Aims

To analyse if an individual's iris pattern is an independent risk factor for the development of KC in RTR.

Materials & Methods

Iris patterns of 110 RTRs were evaluated using the Simionescu visual 3-step technique (Iris periphery, colarette and iris freckling3). Established risk factors for skin cancer in transplant patients were recorded as confounding factors.

Results:

Observational cross sectional study including 110 RTR. Thirty-one participants had skin cancer.

In the skin cancer group, iris periphery was blue/grey in 74.3% ($p = 0.053$, OR 2.5), the colarette was light brown in 57.1% ($P < 0.0043$) and iris freckles were present in 55% ($P=0.044$).

Dark brown and blue colarettes were observed in controls.

Binary Logistic Regression analysis showed light brown colarette is a significant independent risk factor for skin cancer (OR 4.54, P 0.02, CI 1.56–10.57).

Conclusion:

Within this RTR population a blue iris periphery, light brown colarette and presence of freckling confers an independent risk for skin cancer. Iris pattern is a useful tool for identification of transplant patients at risk of skin cancer and an easy to use technique for risk evaluation in this cohort. This is the first study looking at iris pattern and skin cancer risk in RTR.

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Abstract N°: 912

Case of Thymoma-associated multiorgan autoimmunity and literature review

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Introduction & Objectives:

Thymoma is the most frequent neoplasm of the anterior mediastinum. Thymomas are frequently associated with different paraneoplastic syndromes, such as myasthenia gravis or hypogammaglobulinemia. Recently, a condition called Thymoma-Associated Multiorgan Autoimmunity (TAMA) has been described with cutaneous involvement in the form of erythroderma, as well as thyroiditis, hepatitis and colitis. We present a case of TAMA and review the literature in an attempt to clarify the diagnosis and management of this rare entity.

Materials & Methods:

In this paper we present the case of the referred patient, explaining the key diagnostic features of the disease, as well as its management and treatment. In addition, to clarify the management of patients of this type, we performed a literature search in PubMed by entering the terms Thymoma-associated multiorgan autoimmunity and reviewed all the articles published to date.

Results:

We present the case of a 56-year-old patient recently diagnosed with metastatic thymoma, treated with immunotherapy (anti PD-1) who presented with erythematodescamative skin lesions on the trunk. Initially, immunotherapy is suspended with suspicion of toxicoderma. Later, the patient showed frank worsening of the skin lesions, which progressed to erythroderma. In addition, he presented clinical symptoms of bleeding in stool, so colonoscopy with biopsy was performed showing findings compatible with colitis. Laboratory tests showed elevated liver enzymes. A new biopsy of the skin lesions showed a vacuolar interface dermatitis with abundant necrotic keratinocytes and columns of parakeratosis. CD1a staining showed a decrease of Langerhans cells in the epidermis. The histopathological findings and the systemic picture, after review of the literature, lead to suspect TAMA, so oncologic treatment was restarted and systemic corticotherapy was prescribed at high doses, despite which the patient eventually died during hospitalization.

Through a literature search in PubMed, we obtained 28 results about TAMA. Some provided information on the pathophysiology and others on its clinical presentation, diagnosis and treatment.

Conclusion:

TAMA is a rare paraneoplastic syndrome, with poor prognosis, associated with thymoma, resulting from dysregulation of T lymphocytes due to the tumor. It may present as an erythroderma that may be reminiscent of graft-versus-host disease (GVHD), along with colitis, hepatitis and thyroiditis. Histopathologically, similar findings to those of GVHD are observed, with a characteristic absence of Langerhans cells in the epidermis. The therapeutic strategy consists of treatment of the neoplasm, associated with high-dose corticosteroid therapy.

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Abstract N°: 1049

Demodicosis mimetizing GVHD (graft versus host disease), in a bone marrow transplanted man

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Introduction & Objectives:

Materials & Methods:

Results: A 50-year-old white man came to the clinic on the 123rd day after bone marrow transplantation for the treatment of myelofibrosis in the use of :prednison 40 mg every other day, sirolimus , sulfametaxol trimetropin , amoxicilin, pozaconazol , valaciclovir, pentoxifilina, deferasirox , vitamin E, calcium, vitamin D, pyridoxine.

The cutaneous lesion was characterized by an edematous erythematous desquamative plaque with some follicular spicules located on frontal region and nasogenic sulcus, eyelids and cheeks. The lesions itched a little.The hematologists thought that it was GVHD (graft versus host disease), so the immunosuppressive drugs were maintained as well as antibiotics, antiviral and antifungal drugs.

Those follicular spicules looked like pityriasis folliculorum, so a hypothesis of demodicosis in an immunosuppressed man was made. A cutaneous biopsy was taken and the result was:."The skin presents a preserved epidermis with rarefaction of the interpapillary ridges. The follicular infundibulos are elongated by accumulation of keratin and parakeratosis, with specimens of *Demodex folliculorum*. The basal layer is well constituted. In the dermis there is a sparse lymphomononuclear infiltrate"

Topical ivermectin 1% was prescribed once a day, during the night, and oral ivermectin 12 mg / dose, once a week for 2 weeks.

With the pathological anatomy in the hands, the patient returned to the hematologist. The doses of immunosuppressant were tapered and consequently the antiviral, antifungal and antibiotics.

After 2 months the lesions were clearer and the skin was smoother, with very few spicules. The topical Ivermectin1% was prescribed 3 times a week for 3 months because immunosuppressive drugs were being gradually reduced.

Demodicosis should be included in the differential diagnosis of facial lesions in patients after BMT. The mites living in the hair follicles have no prophylaxis and can reproduce freely causing disease.

Conclusion:

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Abstract N°: 1052

A Case Of Gut Tophus Like Calcinosis Cutis

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Introduction & Objectives: Gout is an inflammatory disease characterized by the accumulation of monosodium urate crystals in the joints and soft tissues due to hyperuricemia. Gout arthritis has four stages: asymptomatic hyperuricemia, acute intermittent gout, intercritical gout, and chronic tophaceous gout. Chronic tophi occurs after recurrent polyarticular gout. Lesions usually appear yellow-cream in color and subcutaneous nodules are mobile and firm. Demonstration of strong negative birefringence of intracellular needle-shaped crystals in polarized light microscopy of fluid taken from the joint or tophi is diagnostic for gout.

Materials & Methods: A 78-year-old male patient was admitted to our outpatient clinic with complaints of swelling in the hands and feet. It was learned that the patient with diabetes mellitus, hypertension and chronic renal failure had complaints of swelling in his hands and feet after pain for 8 years. Physical examination revealed swelling in the right and left hand joints, right wrist, ulnar side, and right and left toes. In the rheumatology evaluations of the patient, autoantibodies such as RF, anti CCP negative, anti-JO1, antiSCL70 were negative. It was thought that the findings of the patient, whose calcification was consistent with soft tissue in the hand X-ray and whose PTH level was normal, were due to renal failure. The patient's joint complaints, history of kidney stones and high uric acid (9.3mg/dl) and gout tofu? and calcinosis cutis? The biopsy result obtained with the preliminary diagnosis revealed that there was accumulation of pale basophilic material located in the dermis and many surrounding histiocytes, but it was found to be consistent with gout tophi. After evaluation by rheumatology, the patient was started on allopurinol and called for control.

Results: The incidence of gout tophi was significantly reduced with gout treatment. Xanthomas, rheumatoid nodules and calcinosis cutis are included in the differential diagnosis of gout tophi. Normal serum calcium and PTH levels and negative anti-CCP are important in differentiating calcinosis cutis and rheumatoid arthritis. Complications of long-term untreated gout may include secondary infection, urate nephropathy, kidney stones, neuropathies, and joint fractures with tophi.

Conclusion: In conclusion, it should be kept in mind in clinical practice that tophi, which will occur as a result of delay in treatment in chronic cases, may lead to joint destruction. In our case, the importance of approaching the patient holistically and making a diagnosis with a holistic systematic examination and preventing complications was emphasized.

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Abstract N°: 1053

Juvenile systemic sclerosis : a case report

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Introduction & Objectives:

Systemic sclerosis (SSc) is a chronic disease characterized by skin sclerosis and usually complicated with visceral involvement. This disease generally develops in middle-aged women and is very rare in children. (1) Very few publications have been made in this direction. (2) We report a rare scleroderma case in a pediatric patient.

Materials & Methods:

Results:

Observation: A 13 years-old girl brought to our department for an acrosyndrome evolving since the age of 6 years. On clinical examination, there was a tapered nose, removal of forehead wrinkles, narrowing of the oral orifice with a difficulty in opening her mouth, tense lips, telangiectasias on the nose and cheeks, sclerodactyly of fingers of both hands and Raynaud's phenomenon. All suggesting systemic sclerosis. The patient also presents gastro-oesophageal reflux.

Immunological tests revealed the presence of: anti-nuclear antibodies (ANA) 1/320 positive homogeneous; anti-ro-52 positive antibodies; positive rheumatoid factor.

A peri-ungual capillaroscopy revealed: a reduced capillary density, the morphology showed several ramifications, the presence of megacapillaries and a halo. There were also hemorrhagic spots. This aspect is in favor of early scleroderma.

A chest ct scan showed: an esophageal dilation without obstacle downstream, suggesting gastro intestinal involvement of scleroderma. There were no signs of diffuse interstitial lung disease nor pulmonary arterial hypertension. Respiratory functional explorations and cardiac assessment were normal.

On the basis of these clinical and paraclinical data, the diagnosis of JSSc was retained.

Our patient was put on colchicine as well as hygienic and dietary mesures regarding her digestive involvement.

Conclusion:

Discussion: Juvenile systemic sclerosis (JSSc) is a rare chronic multi-system connective tissue disease characterized by symmetrical thickening and hardening of the skin, associated with fibrous changes in internal organs, as well as vascular and immune system abnormalities in children aged 16 years or younger.(3) It is considered very similar to the adult form of the disease with a better survival rate. (4) As in adults, the visceral manifestations of systemic sclerosis, mainly cardiac, renal and pulmonary with fibrosis, constitutes the main visceral damage involving the vital prognosis. (2) In JSSc, the most frequent systemic involvement followed by the sclerodermal skin symptoms and the Raynaud's phenomenon can be the gastro intestinal tract (Git). All Git locations can be involved, the esophageal dysmotility being the most common.(3), like the case of our patient. Radiological findings may show abnormalities in the absence of any symptoms.(5)

We present our case due to the rare occurrence of systemic scleroderma in children, indicating that it is a collagen

tissue disease with the possibility of acquiring good results with early treatment (3)

Conclusion: Very few cases of juvenile systemic sclerosis (JSSc) have been reported in the literature. The diagnosis is based on the same American Rheumatic Association criteria as for adults. (2) Although the prognosis of JSSc is favorable compared to adult cases, we should keep in mind that there could be patients in whom it takes a fatal course (1). Therefore, early detection of the severity of the disease is essential may play a significant role in establishing the most effective therapeutic regimen.

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Abstract N°: 1054

hemicorporal morphea, parry romberg syndrome and en coup de sabre associated with chronic intestinal pseudo-obstruction : a case report

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Introduction & Objectives:

Localized scleroderma (LoSc), also known as morphea, is a rare fibrosing disease of the skin and underlying tissues. Sclerosis is mostly limited to the skin, but systemic manifestations with visceral abnormalities may occur. (1)

We report the case of a patient with 3 types of LoSc: hemicorporal morphea, Parry Romberg syndrome and En coup de sabre morphea associated with chronic intestinal pseudo-obstruction (CIPO).

Materials & Methods:

Results:

Observation :

A 52-year-old man was referred to our department for a right hemicorporal scleroderma evolving for 15 years with Parry Romberg syndrome of the right hemiface without neurological or ophthalmological involvement, associated with En coup de sabre of the parietal scalp. Laboratory tests including complete blood count, renal and hepatic function were normal. Antinuclear and systemic sclerosis-specific antibodies came back negative. The periungual capillaroscopy did not show any abnormalities. A cutaneous biopsy made on the back returned in favor of morphea.

The diagnosis of LoSc associating 3 types was made. The patient was put on colchicine and corticosteroids for 3 months and then stopped the treatment on his own.

Over the past year, the patient has been admitted four times to the emergency for intestinal obstruction without any mechanical obstacle. He presented symptoms of bloating, abdominal pain and constipation. The diagnosis of chronic intestinal pseudo-obstruction (CIPO) was made. A colon biopsy revealed collagenous colitis (CC). He was put under hygieno-dietetic measures.

Conclusion:

Discussion :

The coexistence of Parry Romberg syndrome and En coup de sabre morphea is known in the literature. (2)

Also, the association of En coup de Sabre morphea and linear morphea has already been described. (3)

However, to our knowledge, the coexistence of 3 forms of LoSc, like the case of our patient, has not been described in the literature.

The risk of developing systemic involvement in patients with LoSc is rare ; gastrointestinal tract involvement appears to be exceptional. It was reported in 1 to 5.1% of patients and consisted mainly of gastroesophageal reflux and dysphagia (1).

Digestive involvement such as CIPO has been described in systemic sclerosis (4)(5)(6), however it has not been described in LoSc to our knowledge.

It is characterized by gastrointestinal motility disorders, having many causes including autoimmune pathologies. (4) It manifests histopathologically by collagenous colitis (CC) whose physiopathological mechanism is based on vascular and collagenic abnormalities, similar to that found in morphea. Asymptomatic in more than half of the cases, it can be very serious due to malnutrition. (4)

Our patient presents the particular association of 3 forms of morphea with an exceptional systemic manifestation which is CIPO.

Conclusion :

We present this case due to the rare coexistence of morphea and systemic involvement such as CIPO, highlighting the need to search and treat it early in order to avoid the resulting complications.

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Abstract N°: 1139

A case of Erdheim-Chester disease-like syndrome exhibiting prominent xanthogranuloma of the eyelid

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Introduction & Objectives:

Erdheim-Chester disease (ECD) is a rare condition characterized by the proliferation of non-Langerhans cell histiocytes, which can cause various symptoms, especially in the bones, central nervous system, cardiovascular system, lungs, kidneys, and skin. The cause of ECD is unknown, but recently it has been considered a neoplastic disease caused by genetic mutations. We experienced a case mimicking ECD, presenting persisting fever, rash, liver dysfunction, and eyelid xanthogranuloma, but lacking bone induration and mutations commonly found in ECD.

Case Report:

The patient is a 37-year-old male who had a history of juvenile idiopathic arthritis at age 16, which was treated with steroids and NSAIDs to achieve a steroid-free remission.

He developed arthralgia, fever, elevated CRP and white blood cell counts, and faint erythema on the neck, trunk, and upper extremities at age 34. During his admission to our Department of Rheumatology, he was diagnosed with adult-onset Still's disease (AOSD) with skin rash, neutrophilia, sore throat, and liver dysfunction. PSL 1 mg/kg/day was started, and later cyclosporine and methotrexate were added, but the patient's systemic symptoms were poorly controlled. Intravenous Tocilizumab (TCZ) was started, and his symptoms tended to improve. Seven months later, swelling of the left eyelid appeared, and the patient was referred to our Department of Dermatology. A skin biopsy was performed, but the findings were nonspecific. TCZ infusion was stopped after six months from the appearance with suspicion of the drug being the cause, but there was no improvement in the left eyelid swelling, but rather a flare-up of arthralgia and fever were observed. The TCZ infusion was restarted two months later, and he showed a tendency toward improvement, but the left upper eyelid swelling gradually worsened, causing vision impairment. The second biopsy, one year after the previous biopsy, showed histology of a xanthogranuloma-like lesion, with perivascular proliferation of CD68, CD163-positive, and S100, CD1a-negative histiocyte-like cells in the full thickness of dermis. However, the clinical presentation was atypical for a localized xanthogranuloma of the eyelid. The patient had recurrent episodes of arthralgia and fever, which led us to consider the possibility of ECD. CT scan showed no characteristic periaortic or perinephric involvement, but pale elevated bone density in the bilateral distal femur, tibia, and fibula was consistent with ECD. However, PET-CT and bone scintigraphy showed no significant accumulation in the bilateral distal femurs, tibias, or fibulas. We searched for mutations in the *BRAF-V600E*, *PIK3CA*, *NRAS*, and *KRAS* hotspots, which are reported to be common in ECD, but no mutations were identified.

Discussion:

Our case, started as AOSD and eventually developed a prominent xanthogranuloma of the eyelid with slightly

elevated CT density in long bones, was suspected to be a systemic non-Langerhans cell histiocytosis. However, the lacks of typical bone/periaortic/perinephric induration in PET-CT and bone scintigraphy were against the diagnosis of ECD. Therefore, we present this patient tentatively as a case of ECD-like syndrome.

Conclusion:

ECD should be considered when histopathological examination of the skin reveals histiocytosis and systemic symptoms are also present. Further accumulation of cases is needed to properly diagnose our case.**

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Abstract N°: 1317

Koebner phenomenon in systemic lupus erythematosus after methotrexate injections.

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Introduction & Objectives:

Koebner phenomenon (KP) is defined as a nonspecific skin stimulus eliciting a disease skin reaction. (1). It was initially described in psoriasis and then in other dermatoses such as lupus erythematosus.

We report a case of KP in systemic lupus erythematosus (SLE) after the injection of Methotrexate (MTX).

Materials & Methods:

Results:

Observation: A 23-year-old woman, with a four year history of SLE, according to the SLICC criteria, with skin (malar rash in butterfly wing, non-cicatricial alopecia), hematological and articular involvement. She was initially put on corticosteroids 0.5mg/kg/day, hydroxchloquine 200mg 2/week, MTX tablets 15mg/week with good tolerance but an insufficient response, hence the switch to the subcutaneous injection of MTX 20mg/week.

After the first administration, the patient presented erythematous and scaly lesions on injection sites, one of which was surmounted by a tight bulla, which persisted for several weeks. The same lesions appeared after intramuscular injections of MTX.

A skin biopsy was performed, revealing: an atrophic epidermis, a junctional and perivascular lymphocyte infiltrate with a slight edema of the papillary dermis, compatible with lupus.

The KP hypothesis following MTX injections was then confirmed by a clinico-histological correlation.

Conclusion:

Discussion: In theory, KP is described in lupus not treated with systemic corticosteroid therapy. That said, our patient presented KP, despite the corticosteroid therapy. This phenomenon is found not only in the skin but also in mucous membranes and internal organs (3).

At the origin of KP in lupus have been incriminated: tattoos, scratches, sun exposure, scars, piercings (2) as well as chemical burns by oil (5).

Cases of KP following subcutaneous injections of MTX have been reported in patients with dermatomyositis (6) and a single case similar to our patient was reported during SLE (7)

The clinical presentation of lesions can be erythematous, papular or plaques with follicular dilatation and plugs (4). Psoriasiform lesions have also been reported, as described in our patient (8). However, the bullous appearance found in our observation was not reported.

In our case, the diagnosis of KP was confirmed by skin biopsy, which found the same histopathological features as the initial dermatosis, thus joining data in the literature (4).

Conclusion: KP, although rare during SLE, should not be neglected in order to prevent the appearance of new

lesions which would aggravate the disease.

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Abstract N°: 1327

Uremic frost in a elderly man with end-stage renal failure: Case report

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Introduction & Objectives:

Uremic Frost is a dermatological condition that manifests in individuals with significant renal insufficiency due to the accumulation of high levels of urea in sweat, leading to the formation of crystalline deposits on the skin, resulting in a frost-like appearance. The incidence of end-stage renal failure has decreased in developed nations since the advent of dialysis.

We present an unusual case of an elderly man with severe uremia and concomitant dermatological manifestation.

Materials & Methods:

A systematic review was conducted on BVS database. The search was carried out using key words related to "Uremic Frost". Of the 19 studies identified, 5 articles were included in this review.

Results:

A 72-year-old male presented to the emergency department with anorexia, nausea, oliguria, and cognitive dysfunction. The patient's medical history includes hypertension and chronic kidney disease, but there is no follow-up information. During his hospitalization, medical examination revealed the presence of small, white, crystalline deposits predominantly on his face, as well as on his scalp, neck, and both arms. The laboratory results for the patient were as follows: blood urea 602.6 mg/dL; serum creatinine 13.3 mg/dL; blood potassium 8.74 mmol/L; and blood phosphorus 14.62 mg/dL. Arterial blood gas analysis was consistent with anion Gap metabolic acidosis. Based on clinical evidence, crystalline deposits indicated uremic frost, suggested end-stage renal illness. The patient had been planned receive hemodialysis, but his primary caregivers refused treatment, resulting in his death.

Conclusion:

Hirschsprung first described uremic frost as a cutaneous manifestation of end-stage renal disease in 1865. In patients with chronic renal failure and uremia, the eccrine sweat glands secrete large quantities of nitrogenous waste products, such as urea, into the sweat. After the evaporation of perspiration, these waste products crystallize on the skin, forming the characteristic tiny, scaly, friable, white, or off-white crystals. The face, neck, scalp, and forearms were the most typical locations for crystal accumulation. In patients who have a diagnosis of end-stage renal disease and the presence of distinctive crystals, uremic frost can be easily diagnosed. Retention hyperkeratosis, eczema, and post-inflammatory desquamation are dermatological conditions that should be considered as possible differential diagnoses. The only effective treatment for uremic frost is the correction of the underlying renal failure.

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Abstract N°: 1384

Incidence and Outcomes of Calciphylaxis in a Large Multicultural Dialysis Unit: A Retrospective Chart Review

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Introduction & Objectives:

Calciphylaxis is a rare skin pathology affecting patients with chronic renal failure. It presents clinically with painful skin lesions and ulcerations, often complicated by superimposed infections. Our tertiary center, which services a large multiethnic population, has diagnosed approximately 30 cases of calciphylaxis over the past decade. Despite limited therapeutic options, the use of intralesional sodium thiosulfate (STS) has shown promise in treating this condition in our center. The objective of this study is to determine the incidence of calciphylaxis and evaluate clinical outcomes in patients treated with STS.

Materials & Methods:

This is a retrospective case series of 30 patients who received dialysis at our centre and had biopsy-proven calciphylaxis treated with STS from January 2018 to January 2023. Patients who are lost to follow-up, refuse, or are unable to receive treatment with STS will be excluded from the study. Data on patient demographics, clinical features, and laboratory values will be collected from patient charts and computerized records. Baseline characteristics will be analyzed. The primary outcome measures include the incidence of calciphylaxis and the effectiveness of STS treatment, which will be measured by pain scores, documentation of wound healing, and evidence for sustained remission beyond cessation of STS therapy.

Results:

Given the location of our study in a major hospital in the Greater Toronto Area, we expect a high degree of patient diversity. We anticipate the incidence of calciphylaxis at our tertiary centre will be similar to that reported in the literature. In addition, it is anticipated that patients who received STS will have improved pain relief, wound healing, and overall survival rates compared to previous treatment options.

Conclusion:

Calciphylaxis is a challenging skin condition to both treat and manage, especially in patients of diverse ethnic backgrounds. The findings of this study will provide valuable information on the incidence of calciphylaxis at a major Canadian hospital and the effectiveness of STS treatment in managing this debilitating condition. The results of this study have the potential to inform the development of guidelines for the management of calciphylaxis and improve patient outcomes.

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Abstract N°: 1519

Paraneoplastic cutaneous manifestations due to nodal marginal zone B-cell lymphoma

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Introduction & Objectives:

Paraneoplastic dermatoses are a group of skin conditions that have strong associations with internal malignancies. Its underlying mechanism is not well understood but they are not directly related to the primary tumour itself or to its metastases. Lack of familiarity with cutaneous clues of internal malignancy may delay diagnosis and treatment of cancer.

Materials & Methods:

Presentation of a case report and review of the literature.

Results:

A 69-year-old man consulted for tense blisters and erythematous-scabby lesions on the scalp and back of the hands. Nine months earlier he had been diagnosed with marginal zone B lymphoma that was not treated due to its low grade and the absence of poor prognostic factors. Urinary porphyrins and anti-epithelial antibodies were determined with negative results. Antinuclear antibodies were positive without meeting criteria for systemic lupus erythematosus. Skin biopsies showed a subepidermal bullous dermatitis with mixed inflammatory and eosinophilic infiltrate, and direct immunofluorescence was negative. Treatment with photoprotection and hydroxychloroquine was started with partial improvement. Two years later, specific treatment for the lymphoma was started with rituximab-bendamustine, obtaining a complete response and disappearance of the skin lesions.

Conclusion:

We present a case of paraneoplastic dermatosis associated with marginal zone B lymphoma. Only one other similar case associated with an indolent variant of mantle B lymphoma had been described in the literature. Interestingly, in both cases, lesions were suggestive of pseudoporphyria, but the biopsy showed a mixed inflammatory infiltrate with eosinophils. In addition, in both cases the specific treatment of the lymphoma managed to resolve the skin lesions.

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Abstract N°: 1527

Design of a phase 2, double-blind, placebo-controlled, global trial of deucravacitinib, an oral, selective, allosteric tyrosine kinase 2 (TYK2) inhibitor, in patients with active discoid and/or subacute cutaneous lupus erythematosus

Victoria P. Werth[1], Joseph F. Merola[2], Joerg Wenzel[3], Nikolay Delev[4], Harini Kothari[4], Richard Meier[5], Shalabh Singhal[6], Malavi Madireddi[4], Shimon Korish[4]

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Introduction & Objectives:

Deucravacitinib is a first-in-class, oral, selective, allosteric tyrosine kinase 2 (TYK2) inhibitor approved in multiple countries for the treatment of adults with plaque psoriasis. In a phase 2 trial in patients with systemic lupus erythematosus (SLE),1 deucravacitinib demonstrated efficacy across multiple outcome measures, including achievement of > 50% reduction in Cutaneous Lupus Erythematosus Disease Area and Severity Index-Activity (CLASI-A) score (CLASI-50) in those with a baseline CLASI-A score ≥ 10, and is now being investigated in 2 phase 3 SLE trials (NCT05617677, NCT05620407). Patients with discoid and/or subacute cutaneous lupus erythematosus (DLE/SCLE) have elevated expression of type 1 interferons (IFNs).2 Deucravacitinib mediates signaling of type 1 IFNs, interleukin (IL)-12, and IL-23 and may be an effective treatment for patients with DLE/SCLE.3

Results of this ongoing phase 2 trial (NCT04857034) will characterize the efficacy and safety of deucravacitinib compared with placebo in patients with active DLE/SCLE with or without SLE.

Materials & Methods:

This phase 2, global, randomized, double-blind, placebo-controlled trial is enrolling adults (aged 18-75 years) with a biopsy-confirmed clinical diagnosis of DLE/SCLE. Key eligibility criteria and study design are depicted below (**Figure**). Eligible patients will be randomized (1:1:1) to treatment with placebo or deucravacitinib (dose 1 or 2) for 16 weeks. At week 16, all patients randomized to placebo will be rerandomized (1:1) to treatment with deucravacitinib dose 1 or 2 until week 52. Patients originally randomized to deucravacitinib will continue treatment until week 52. At week 16, patients will be evaluated for percentage change from baseline in CLASI-A score (primary endpoint) as well as key secondary endpoints, including the percentage of patients who achieve a ≥ 50% reduction in CLASI-A score from baseline (**Table**). This trial will also assess the safety and tolerability of 2 doses of deucravacitinib, exploratory efficacy endpoints, patient-reported outcomes, and pharmacodynamics.

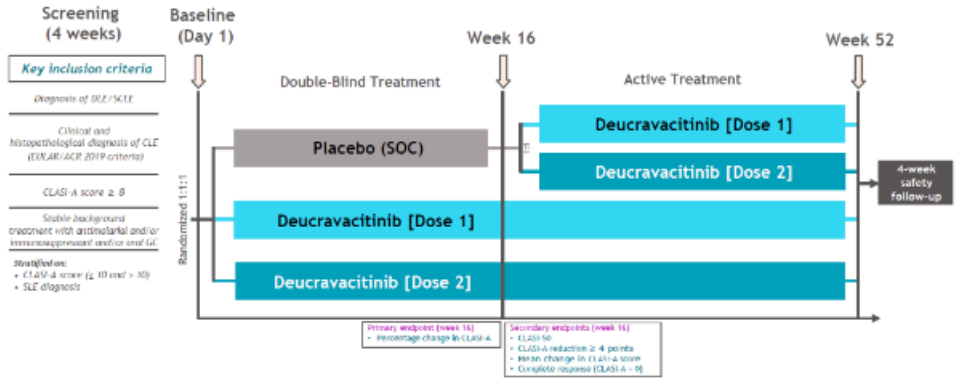
Results:

Total planned enrollment is 75 patients (25 per double-blind treatment group) in North America (US, Mexico) and South America (Argentina, Brazil, Chile, Colombia, Peru); Europe (France, Germany, Poland); and the Asia-Pacific (Australia, Taiwan) regions.

Conclusion:

This phase 2 trial will characterize the efficacy, safety, and tolerability of deucravacitinib in patients with active DLE/SCLE.

Figure. Trial Design



ACR, American College of Rheumatology; CLASI-A, Cutaneous Lupus Erythematosus Disease Area and Severity Index-Activity; CLASI-50 (CLASI-A-50), decrease of ≥ 50% from baseline Cutaneous Lupus Erythematosus Disease Area and Severity Index; CLE, cutaneous lupus erythematosus; DLE, discoid lupus erythematosus; EULAR, European Alliance of Associations for Rheumatology; GC, glucocorticoids; SCLE, subcutaneous lupus erythematosus; SLE, systemic lupus erythematosus; SOC, standard of care.

Table. Primary and secondary endpoints assessed at week 16

Primary endpoint
• Mean percentage change from baseline in CLASI-A score
Secondary endpoints
• Percentage of patients who achieve a ≥ 50% reduction in CLASI-A score (CLASI-50) from baseline • Percentage of patients who achieve a ≥ 4-point improvement in CLASI-A score from baseline • Mean change from baseline in CLASI-A score • Percentage of patients who achieve a complete response (defined as a CLASI-A score of 0)

CLASI-A, Cutaneous Lupus Erythematosus Disease Area and Severity Index-Activity.

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Abstract N°: 1573

Reactive acroangiodermatitis of the arm associated with an arteriovenous fistula

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Introduction & Objectives:

A 54-year-old gentleman presented to Dermatology with a 2-month history of a slow healing ulcer of the left forearm preceded by minor trauma. He had a background history of end-stage kidney disease on hemodialysis via a left brachiocephalic arteriovenous fistula.

Materials & Methods:

On examination, there was a violaceous ulcerated plaque measuring 6 centimetres in length on his left forearm. Histological examination of the skin revealed lobular proliferations of capillaries in the dermis and subcutis thrombosed with red cells. The epidermis was necrotic and the superficial subcutis showed microcystic fat change. Microbiological studies did not identify any organism.

Results:

He was diagnosed with acroangiodermatitis associated with an iatrogenic arteriovenous fistula. Remarkably, the ulcer resolved spontaneously without removal of the arteriovenous fistula.

Stewart–Bluefarb syndrome is characterised by acroangiodermatitis associated with an underlying arteriovenous malformation or iatrogenic arteriovenous fistula. Acroangiodermatitis usually presents as erythematous to violaceous plaques or nodules that may become verrucous or ulcerated. Reactive proliferation of blood vessels in the skin occurs in response to a disturbance in blood circulation. The association with chronic venous insufficiency, known as acroangioedermatitis of Mali, has also been reported. Important differential diagnoses to consider include traumatic ulceration, deep fungal or non-tuberculous mycobacterial infections, Kaposi's sarcoma, and keratinocyte malignancies.

Conclusion:

We would like to highlight this rare entity that can occur in patients on hemodialysis via an arteriovenous fistula. Surgical treatment may be considered to correct the vascular anomaly.

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Abstract N°: 1807

The malignant rash- leukemia cutis as first presentation of acute myeloid leukemia: A case report

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Introduction & Objectives: Acute myeloid leukemia (AML) enocmpasses a heterogeneous group of aggressive blood cell cancers due to clonal expansion of hematopoietic precursor cells in the bone marrow. The reported prevalence of leukemia cutis in AML ranges from 3% and 13%, the clinical presentation may be maculopapular, nodular or patch-like.

Materials & Methods: N/A, Case Report

Results: A 48-year-old otherwise healthy woman has developed a nonpruritic, unspecific rash consisting of disseminated, erythematous brownish macules and papules on her trunk that resembled a paraviral exanthema, drug eruption or secondary syphilis. Diagnostic work-up revealed an elevated blast count in the peripheral blood. Skin biopsy taken from the back showed a dense perivascular malignant infiltrate of pleomorphic blasts with mitotic figures. The immune profile in the skin was consistent with NPM1-mutated AML. The malignant myeloic cells showed strong immune expression of NPM1 and lysozyme and a high proliferative activity with 80% MiB1-positive tumor cells. The malignant cells were negative for CD3 and myeloperoxidase.

Diagnosis of AML subtype “with mutated NPM1 gene” according to the World Health Organisation Classification was subsequently confirmed by bone marrow aspiration.

Under treatment with azacitidine, venetoclax and gilteritinib, the skin lesions resolved within 3-4 weeks, preceding the remission of blasts in the peripheral blood.

Conclusion: This case demonstrates that cutaneous manifestations might be the first presentation of hemato-oncologic malignancies. In particular, clinicians should be aware that AML might present as “unspectacular exanthema”. In our case, leukemia cutis was not only the first presentation of AML but also the first sign of clinical response to treatment.

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Abstract N°: 1843

Necrolytic migratory erythema as a manifestation of metastatic glucagonoma

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Introduction & Objectives:

Necrolytic migratory erythema (NME) is an unusual paraneoplastic dermatosis associated with glucagonoma, an infrequent neuroendocrine tumor of pancreatic origin. NME is a widespread rash characterized by annular erythematous plaques with central bullous or erosive lesions surrounded by hyperpigmentation. It involves the face, mainly the perioral region, the trunk, intertriginous sites, perineum, and extremities. These skin lesions can be the first or the only obvious manifestation in up to 70% of patients before the diagnosis of a glucagonoma in which unfortunately almost half of them have metastatic disease at the time of diagnosis.

Materials & Methods:

38-year-old female presented with a 4-year history of diabetes mellitus, subsequently complained of involuntary weight loss of 12 kg and other nonspecific symptoms such as anorexia and fatigue, accompanied by a pruritic, disseminated cutaneous polymorphic eruption of annular erythematous plaques, some of them coalescent with eroded centers and active scaly borders, with crusts and postinflammatory hyperpigmentation on the periphery. It symmetrically affected the face predominantly on the perioral and centrofacial regions, the antecubital and popliteal fossae, groins, vulvar area, perineum, and distal region of the extremities.

Results:

A skin biopsy was performed, revealing parakeratosis, isolated necrotic keratinocytes on the upper third of the epidermis and perivascular lymphocytic inflammatory infiltrate in the upper dermis. Given the clinical symptoms and findings the suspect of NME leaded us to perform an abdominal computed tomography, which revealed the presence of a pancreatic tail tumor along with liver and spleen metastases. Conclusively, the diagnosis of glucagonoma was confirmed by tumor biopsy and immunohistochemistry.

Conclusion:

NME is an unusual but important paraneoplastic cutaneous manifestation of glucagonoma. Generally, represents a diagnostic challenge due to the low worldwide incidence of this type of tumor, which is estimated to be 1 case in 20 million people. This slow-growing malignant neoplasm is located in the pancreatic tail in up to 87% of cases and more than 50% of these tumors are metastatic at the time of diagnosis. Therefore, dermatologists play a very important role by making an early recognition of this paraneoplastic dermatosis, which gives us the possibility of an opportune diagnosis of the underlaying tumor and a better prognosis for the patient.

Currently, we must not neglect the fact that NME is a specific finding and that it is often the only initial manifestation of glucagonoma. Its presence has been reported in approximately 70-90% of patients, reason why it is considered one of the major criteria for the diagnosis of this tumor. Due to the information given and the findings mentioned in the previous case, we were forced to request complementary imaging studies, which helped us to identify the tumor and metastases of the patient in question, because particularly in this case, the manifestations of glucagonoma were not initially accompanied by skin manifestations, as reported in the minority

of cases, which do not exceed 30%.

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Abstract N°: 1945

The Clinical Efficacy and Tolerance of a Ceramide-Containing Moisturising Cream in the Management of Facial Sensitive Skin

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The Clinical Efficacy and Tolerance of a Ceramide-Containing Moisturising Cream in the Management of Sensitive Facial Skin

Introduction & Objectives:

Patient-perceievd sensitive skin can be the result of several factors, but a leading hypothesis is an imparied epidermal barrier. It is common patients with dry skin or inflammatory conditions, known to be associated with reduced ceramide levels and barrier dysfunction, report experiencing symptoms of sensitive skin. The objective of this study was evaluate the clinical efficacy and tolerance of a cream containing three ceramides (EOP, NP and AP) in the condition and management of the symptoms of sensitive skin on the face.

Materials & Methods:

110 patients with self-perceived sensitive skin on the face were enrolled in this study. The patients applied the test cream twice daily for 28 days. The patients were randomly divided into group A (n=44) and B (n=66). Skin barrier function were evaluated on day 0, 1, 7, 14 and 28 in group A. Self-assessment on sensitive skin, patients' satisfaction, clinical efficacy was scored by patients in group B on different time points. Tolerance was evaluated by dermatologists in all 100 patients.

Results:

After 28 days of usage, the moisture content of stratum corneum in group A increased from (33.90 ± 12.30) C.U to (46.70 ± 10.40) C.U (P<0.05); The skin erythema index EI decreased from 258.30 ± 61.10 to 249.50 ± 54.40 (P<0.05); The sebum content is (65.00 ± 32.00) μ G/cm2 increased to (80.00 ± 35.00) μ G/cm2 (P<0.05); Skin transcutaneous water loss decreased from (17.00 ± 4.60) g/(h · m2) to (15.30 ± 6.40) g/(h · m2) (P<0.05); The skin red area a* value decreased from 14.95 ± 4.31 to 13.59 ± 4.53(P<0.05); 28 days later, the skin irritation and sensitive symptoms of the patients in Group B skin were alleviated; 94% of the subjects agreed the ceramide-containing cream was gentle and comfortable to use; According to the evaluation of dermatologists, the test product was well-tolerated. During the study, the patients had no adverse reactions.

Conclusion:

The test cream containing three skin-identical ceramides (ceramides EOP, NP, and AP) significantly improved skin hydration and barrier function assocaited with the reduction in symptoms of sensitive skin. The test cream was well-tolerated and percieved as gentle by subjects with self-perceived sensitive skin on the face.

C1 - Internal use

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Abstract N°: 1959

Examination of the Relationship between Organ Involvement and Inflammatory Markers in Cutaneous Sarcoidosis

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Title: Examination of the Relationship between Organ Involvement and Inflammatory Markers in Cutaneous Sarcoidosis

Introduction & Objectives:

Sarcoidosis is a chronic granulomatous disease characterized by non-caseating granulomas that can affect many systems. Skin involvement may be the initial presentation of the disease. Systemic immune-inflammatory index, systemic immune response index, neutrophil-lymphocyte ratio, platelet-lymphocyte ratio, and monocyte-lymphocyte ratio have been used for various diseases. This study aims to evaluate the relationship between clinical findings and inflammatory markers in detecting organ involvement that may develop during diagnosis and follow-up.

Materials & Methods:

The data of 58 patients, between 2008 and 2022, were retrospectively analyzed. Four patients with a history of active infection, hepatitis, and malignancy and two whose follow-up data could not be reached were excluded from the study.

Results:

Fifty-two patients were included in the study. 76.9% (n=40) of the patients were female, with a F/M ratio of 3.3. The mean age of the patients at the time of diagnosis was 46.3±9.4 (min: 29, max: 69). Disease duration ranged from 1 month to 20 years (mean 17.9 months). The analysis of the lesions revealed that papules accounted for 61.5% (n=32) of the cases, while plaques and nodules made up 44.2% (n=23) and 48.1% (n=25) respectively. A small percentage of cases were found in lupus pernio (3.8%, n=2), scars (3.8%, n=2) and a single case on a tattoo (1.9%, n=1). The lesions were head and neck in 48.1% (n=25), trunk in 40.4% (n=21), upper extremity in 51.9% (n=27), and in 44.2% (n=23) in the lower extremities. Systemic organ involvement was accompanied by 71.1% (n=37) of the patients. Anergy in 42.3% (n=22) of the patients, an increased serum ACE value in 58.3% (n=28), and an increased sedimentation rate was found in 42.3% (n=22). While lung involvement was the most common (n=37), three patients had an eye, two had bone involvement, and one had kidney involvement. Three patients had more than one organ involvement. During the follow-up period, lung involvement developed in three patients with only skin involvement (n=15) and cardiac involvement in one patient with lung involvement (mean 35.5 months). Significant differences were found in SII, SIRI, NLR, PLR, MLR, lymphocyte count, and sedimentation rate in patients with organ involvement at diagnosis (respectively $p=0.003$; $p=0.016$; $p=0.001$; $p=0.006$; $p=0.006$; $p=0.001$; $p=0.002$). Organ involvement was significantly higher in patients with head and neck involvement and nodular lesions (respectively, $p=0.007$; $p=0.033$). SII>725.8 have a higher risk of organ involvement (sensitivity 62.5%; specificity 80%; $p<0.05$).

Conclusion:

Our study found statistically significant differences in SII, SIRI, NLR, PLR, MLR, and lymphocyte counts in patients with extracutaneous involvement. Sedimentation and these parameters should be evaluated together in determining organ involvement. However, no significant difference was found in the initial parameters of the patients who developed new organ involvement during the follow-up period. Therefore, these inflammatory markers may not be sufficient to predict whether organ involvement will develop. Our study is the first in the literature to examine the relationship between organ involvement and systemic inflammatory markers in patients with cutaneous sarcoidosis.

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Abstract N°: 2022

A case of antisynthetase syndrome with typical mechanic's hands

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Introduction & Objectives:

Antisynthetase syndrome (ASS) is a subset of idiopathic inflammatory myopathies that presents as interstitial lung disease (ILD), fever, polyarthritis, and skin manifestations. Mechanic's hands, known as nonpruritic hyperkeratotic patches on lateral side of fingers, are frequently observed skin lesions in patients with ASS. Although they are usually asymptomatic, mechanic's hands cause decline in quality of life aesthetically.

Materials & Methods & Results:

A 73-year-old female presented with asymptomatic hyperkeratotic, scaly patches on lateral side of digits and fingertips, noticed a year ago. She was undergoing medical evaluation due to progressive dyspnea, arthralgia, and fever. The laboratory test revealed elevated lactate dehydrogenase, creatinine kinase levels and positive anti-Jo-1 antibodies. Chest CT showed an ILD pattern and electromyography confirmed myopathy. Under the diagnosis of ASS, treatment with systemic steroid and mycophenolate mofetil was initiated, resulting in improved systemic symptoms, but the skin lesions remained still.

Conclusion:

Although ASS is a rheumatologic disease, mechanic's hands are characteristic skin manifestation and one of the important diagnostic factors. Therefore, we report a case of ASS to emphasize that dermatologists should be aware of this cutaneous involvement for holistic diagnosis and management of ASS.

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Abstract N°: 2100

clinically amyopathic dermatomyositis: a case report

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Introduction & Objectives:

Clinically amyopathic dermatomyositis (CADM) is an idiopathic inflammatory disease from the group of dermatomyositis (DM) but that does not show clinical or analytical muscle involvement for at least 6 months.

We present a case in which, through the diagnosis of probable CADM, we arrived at the detection of gynecological neoplasia.

Materials & Methods:

A 55-year-old female patient with history of hypertension and non-insulin diabetis, consulted for a 10 months' evolution dermatosis, cephalocaudal onset, pruritic and painful, treated with prednisone 40 mg/day and antihistamines for 1 month.

Brings biopsy report performed 3 months prior reporting vasculitis and actinic keratosis.

On physical examination presented heliotrope erythema and generalized, irregular, poikilodermiform hyperpigmented macules. Also Gottron's papules on both hands, but no signs of muscle weakness. DM was suspected, complementary studies were performed:

Skin biopsy of the dorsal lesion: Cytotoxic pattern with marked increase in interstitial mucins, suggestive of collagen disease versus pharmacoderma

Analytical: normal CPK, aldolase, tgo, tgp and ldh. Non detectable VDRL HIV VHB HCV and FAN and Rheumatoid Factor.

X-ray and tomography of the chest without findings to highlight

Mammary ultrasound without alterations. Gynecological ultrasound: anechoic, heterogeneous image of 64x38mm in the fundus of the uterus.

CT scan of the abdomen and pelvis: heterogeneous adnexal mass and focal peritoneal lesions.

MRI of the pelvis: above the bladder and in front of the uterus, a heterogeneous mass with liquid and solid portions, hematic areas, septa, amorphous, with irregular edges, with enhancement after contrast injection. Peritoneal implants.

AC 125:187

Results:

CADM constitutes between 2 and 18% of all DM cases, with an incidence of 0.1-1 per 100,000 inhabitants/year and a female predominance. Diagnostic criteria include pathognomonic skin changes (Papules-Gottron's sign,

heliotrope erythema, and poikiloderma), compatible skin biopsy, normal muscle enzymes, and clinical absence of proximal motor weakness; criteria met in this case. CADM is considered probable if the criteria are met for at least 6 months and confirmed if they are met for 2 years.

Patients who suffer from it have a higher risk of presenting cancer than patients with PM and both diseases have up to 25% higher incidence of neoplasms than the general population

Conclusion:

As the skin is a visible organ, cutaneous manifestations works as a strong motor for medical consultation and in the skill of dermatologist is the opportunity to diagnose underlying diseases.

In our case, being a second consultation and with almost a year of evolution, we found a locally advanced gynecological neoplasia awaiting surgery for typification and definition of treatment.

Although CAMD has a statistically low incidence, timely and early diagnosis lies the therapeutic opportunity and therefore the patient’s life expectancy, making the difference.

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Abstract N°: 2158

Calciphylaxis of the Breast in a 74-Year-Old Female : A Case Report

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Introduction & Objectives: Calciphylaxis is a rare disease characterized by calcification of small and medium vessels leading to ulcerative-necrotic dermatological manifestations. This disease affects 1-4% of patients in the dialysis stage of renal failure and is fatal in 60 to 80% of cases. It tipically occurs on areas with high fat content and is characterized by the rapid onset of infiltrated, inflamed, purplish, and livedoid skin plaques. These plaques can lead to digging and necrotic ulcerations. We report a case of calciphylaxis that is particular by its seat in the areolar region, in a 74-year-old patient, observed at the Central Military Hospital of Algiers.

Materials & Methods:

Case- A 74-year-old female patient with a history of cholecystectomy 40 years ago, obesity, and diabetes for 30 years on insulin, in the terminal chronic renal failure stage, was hospitalized for exploration of ascites of great abundance evolving for 15 days. The patient presented with painful skin necrosis of the right breast that had been evolving for 1 month. Deep skin biopsy confirmed the diagnosis of calciphylaxis by showing calcification of the walls of the small vessels. Treatment included necrectomy under anesthesia, a modification of dialysis with a low-calcium bath, and sodium thiosulfate infusions.

Results (and discussion)

The examination found a patient in poor general health condition who presented an ulceration covered by a blackish crust in the areolar region of the right breast. The diagnosis of calciphylaxis was made based on the patient's context of renal insufficiency, significant pain, the presence of a hard necrotic plaque surrounded by a livedoid halo, and histological findings. The histology showed an infiltrate rich in neutrophils, calcifications of the connective tissue, and the presence of an organized thrombus in the wall of an arteriole. The breast seat is very rare, and some patients, despite a sometimes-bilateral mastectomy, died as a result of renal failure or secondary infection. A directed healing allowed a clear improvement of the lesions.

Calcifying uraemic arteriolopathy causes tissue necrosis, mainly in the skin and subcutaneous adipose tissue. Deep skin biopsy is required for the diagnosis of calciphylaxis. The breast seat is very rare, and current management of patients with calciphylaxis with sodium thiosulfate infusion seems to improve the prognosis.

Conclusion :

Calciphylaxis is a rare but extremely serious complication of end-stage chronic renal failure requiring long-term dialysis. Early diagnosis allows rapid and appropriate treatment. Extensive debridement with rapid correction of the phosphocalcic balance associated with sodium thiosulphate has made it possible to revolutionize the prognosis of these patients. Further studies are needed to determine the optimal treatment strategy for calciphylaxis.

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Abstract N°: 2193

Hypophosphatemic osteomalacia associated with type I neurofibromatosis: a case report; what is the link ?

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Introduction & Objectives:

Several bone manifestations can be observed during neurofibromatosis. We report an exceptional case of hypophosphatemic osteomalacia associated with type I neurofibromatosis.

Materials & Methods:

We report a case of Hypophosphatemic osteomalacia associated with type I neurofibromatosis

Results:

Patient I.L, 24 years old, with a family history of type I neurofibromatosis, is referred for diffuse bone pain evolving for 6 years with a waddling gait. Dermatological examination found multiple café-au-lait spots, associated with a pigmented and pilling plaque (neurofibroma) on the right leg. The ophthalmological examination notes the presence of iris nodules (Lisch nodules). The bone X-ray shows: Looser Milkmann streaks in the pelvis and both shoulder blades. The presence of bilateral cracks of the neck of the femur, the lower ends of the femur, the upper ends of the tibia, as well as at the level of the upper and lower end of the left radius. A bilateral fracture at the level of the upper and lower third of the fibula. Biology shows alkaline phosphatase at 3 times normal, 316 IU/l, normal serum calcium (2.16 mmol/l) associated with hypophosphatemia (0.69 mmol/l), total vitamin D at 20,22 ng/ml, a normal PTH level at 35.50 and a normal 24-hour calciuria at 3.60 mmol/l. All the clinical, radiological and biological signs are in favor of hypophosphatemic osteomalacia. Osteomalacia is a very rare complication during type I neurofibromatosis. The pathophysiological mechanism is similar to that of mesenchymal tumors with excessive secretion of FGF23 (Fibroblast Growth Factors 23) by the cells constituting the neurofibromas, FGF23, formerly called “phosphaturic factor”. this factor plays a role in the regulation of phosphaturia and in the inhibition of the activity of renal 25(OH) 1 α hydroxylase leading to a defect in the synthesis of calcitriol, resulting in hypophosphatemia and hyperphosphaturia causing osteomalacia. excision of the neurofibroma plaques could lead to the regression of the symptomatology and the normalization of the biological markers, or even a permanent cure

Conclusion:

Conclusion Osteomalacia on neurofibromatosis type I is a very rare condition, however monitoring of the phosphocalcic balance in these patients is recommended in search of an associated osteomalacia.

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Abstract N°: 2196

Anabolic Steroid Induced Cutis Verticis Gyrata

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Introduction & Objectives: Cutis verticis gyrata (CVG) is a benign morphological entity leading to skin hypertrophy of the scalp and the presence of deep convoluted folds and furrows mimicking the sulci and gyri of the cerebrum. CVG can be subdivided into primary forms which include essential or non-essential CVG and secondary forms. Primary essential CVG is an isolated disorder which does not have any associations with other conditions whilst primary non-essential CVG is typically associated with underlying neurological or ophthalmological disease. Secondary CVG is associated with disorders that may modify the trophicity of the skin and is typically due to an underlying systemic condition but in rare cases can be due to medications.

Materials & Methods: A 21-year-old male bodybuilder of Russian descent presented to our dermatology clinic with a year long history of progressively deep cerebriform-like skin folds located on the occipital scalp. He noted that the appearance of his scalp had become extremely prominent in the last month. His medical history included juvenile arthritis for which he was treated with adalimumab by his rheumatologist. He was otherwise well and had no similar family history or noted consanguinity. Further questioning revealed a 20kg increase in weight over the last year from muscular hypertrophy consistent with the use of anabolic steroids and estrogen blockers. Clinical examination was otherwise unremarkable aside from mild folliculitis affecting his trunk, back and upper arms. He had no facial features of acromegaly, no evidence of neurological or ophthalmological abnormalities, no clubbing of the nails and no papular or sclerodermoid bumps. He was started on doxycycline 100mg daily for his folliculitis and told to cease his anabolic steroids and estrogen blockers. **Results:** A 4mm punch biopsy of the posterior scalp was taken for histopathological sampling. The epidermis revealed mild acanthosis. The upper dermis showed mild fibrosis, angioplasia, telangiectasia and perivascular lymphocytic infiltrate. There was increased interstitial mucin deposits in the mid and deep dermis. No evidence of proliferation of melanocytes, fibroblasts, collagen fibres, sebaceous glands, granulomas or folliculitis was seen. His full blood count, thyroid function studies, liver function tests, kidney function tests, fasting lipids, ACE levels and HbA1c were within normal realms. His HIV, ANA, ENA, and syphilis serology were negative.

Conclusion: Although the exact pathogenesis of CVG remains unclear, it is thought that both primary and secondary CVG may have an endocrinological basis. Intriguingly whilst our patient did have evidence of mucin deposition, there was no other cutaneous or histological features suggestive of scleromyxedema for which there has been one case reported in association with the development of CVG. We are uncertain of the significance of the increased mucin, but it raises the possibility of mucinosis related to anabolic steroid use. Association of CVG with misuse of anabolic steroids has been documented in the literature on one occasion previously. Based on the clinical features of our patient, we believe this is the second reported case of CVG induced by anabolic steroid use. Anabolic steroids are known to interfere with the hypothalamic-pituitary-gonadal axis leading to hormonal imbalances. In light of this, our case supports the prevailing scientific consensus that pathogenesis of CVG may be endocrinological in origin.

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Abstract N°: 2203

The Dermoscopic Patterns in Patients with Acanthosis Nigricans and it's Relation to Duration and Control of Diabetes Mellitus

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The Dermoscopic Patterns in Patients with Acanthosis Nigricans and it's Relation to Duration and Control of Diabetes Mellitus

Introduction & Objectives:

Acanthosis nigricans (AN) is associated with endocrine dysfunction, especially insulin resistance and hyperinsulinaemia, as seen in diabetes mellitus (DM). Clinical and dermoscpic finding of AN are associated with control of DM

Aim: The aim of this study is to evaluate the dermoscopic patterns in patients with AN and its relation to duration and control of diabetes mellitus.

Materials & Methods:

This Comparitive analytical cross-sectional study was conducted Prediabetic patients (20 patients) complaining of AN with age ranging from 30 to 52 years old (2 male and 18 female) and Patients of chronic diabetes mellitus of different durations (20 patients), complaining of AN with age ranging from 33 to 60 years old (5 male and 15 female); we investigated HbA1C level and rondom blood glucose, in addition to dermoscopic examination of AN were performed using the dermoscope (DL4).

Results:

Significantly positive correlation between dermoscpic finding of AN and BMI, rondom blood glucose and HbA1C of both groups. Significantly positive corolation between (hyperpigmented dots, hyperpigmented globule and papillary projection) of dermoscpic finding of AN and clinically hyperpigmented patch and Velvety like appearance of both groups. Significantly increase in papillary projection, hyperpigmented dots and hyperpigmented globule in Group II compared to Group I.

Conclusion:

Significantly positive correlation between dermoscpic finding of acanthosis nigricans and BMI, RBG and HbA1C of both groups. Severity of clinical appearance of AN is related mainly to level of blood glucose and IR so it more marked in diabetic group than prediabetic group.

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Abstract N°: 2234

Kikuchi cutaneous disease: Cohort of cases from a tertiary Spanish hospital

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Introduction & Objectives:

Kikuchi disease (KD), or histiocytic necrotizing lymphadenitis, was described in 1972 as a benign syndrome characterized by cervical lymphadenopathy and fever. Of unknown etiology, it has been mainly related to infections and autoimmune processes, mainly systemic lupus erythematosus (SLE). Diagnosis is based on adenopathy histology (paracortical necrosis, karyorrhexis and a mononuclear infiltrate). The 15-40% of the cases present skin lesions, and its histology is similar to that of the lymph nodes. In 2015, Thai et al. described a series of patients with skin lesions with histology compatible with KD but without lymph node involvement, which they called “Kikuchi disease-like inflammatory pattern” (KLIP). Two thirds of this series were finally diagnosed with cutaneous lupus erythematosus (CLE), so it was hypothesized that KLIP could be a new histological pattern of CLE.

Results:

We present 8 patients, all women (4 Spanish, 3 Filipinas, and 1 Peruvian), aged between 22 and 57 years (mean: 42), histologically diagnosed with cutaneous KD between 2009-2022. Three patients were diagnosed with KD by lymph node biopsy, one presented self-resolving palpable lymph nodes before being biopsied, and four did not present lymph nodes, being diagnosed with KLIP. In the three Filipino patients, the diagnosis of SLE was reached (1 prior and 2 concomitantly with the cutaneous KD lesions), while the Peruvian patient later developed chronic CLE. The 3 cases of SLE were severe, requiring ICU admissions due to renal, cardiac, neurological and/or pulmonary involvement. Four patients presented pancytopenia due to a hemophagocytic syndrome. The skin lesions biopsied were polymorphic (annular plaques, papules, nodules, macules, erosions…), but all of them have shown a histological pattern of baseline vacuolization, a dense perivascular, adnexal, and interstitial lymphohistiocytic infiltrate, mucin deposits, karyorrhexis with histiocytes and CD123+ cells. Direct immunofluorescence studies were positive in two patients and negative in three. The patients received multiple treatments, the most used being antimalarials. Three patients have not presented new skin outbreaks, while five have presented new lesions that have responded to antimalarials (2), prednisone (2) and topical corticosteroids (1). All patients are still alive, with a mean follow-up of 7 years.

Conclusion:

Of our series of 8 patients with cutaneous KD, four were diagnosed with KD (one without nodal histological confirmation) and four with KLIP. Three patients achieved SLE diagnosis and one developed chronic CLE lesions. In agreement with the literature, in our series there is an overrepresentation of Asian patients, and the association with lupus is also constant in these patients, which ended up occurring in 50% of the total.

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Abstract N°: 2279

Efficacy of immunoglobulins in corticosteroid-resistant dermatomyositis with dysphagia; about a case

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Introduction & Objectives:

The appearance of dysphagia in dermatomyositis is a sign of severity which conditions the prognosis because it predicts an often more aggressive course and increases the risk of infectious complications (inhalation pneumonia). Intravenous immunoglobulins (IVIG) can currently be offered as a second-line therapy.

Materials & Methods:

We report the efficacy of IVIG in corticosteroid-resistant dermatomyositis with dysphagia.

Results:

An 80-year-old man with a history of diabetes and arterial hypertension has had definite dermatomyositis for two months according to the criteria of Bohan et al. On physical examination found an average general condition with asthenia and weight loss; skin involvement is typical; muscle damage is noisy with deficit of the proximal muscles. Paraclinically, there is an inflammatory syndrome; muscle lysis (10 X normal serum creatine kinase), an electromyogram confirming myogenic damage, and a negative paraneoplastic investigation. Faced with the intensity of the muscle damage, we instituted systemic corticosteroid therapy at a rate of 1.5mg/kg/day with adjuvants and glycemic monitoring. - On day 15 improvement in skin signs with decrease in serum creatine kinase to 1200. - On day 20, appearance of total dysphagia to solids and liquids; that is attached to dermatomyositis after eliminating an infectious or organic cause. The increase in corticosteroid therapy to 2mg/kg/day for several days did not lead to any improvement, so we opted for the addition of IVIG at a rate of 2g/kg/course, leading to a normalization of serum creatine kinase and a resolution of the dysphagia with gain in muscle strength at the end of the 2nd treatment. We begin the reduction of corticosteroids from the 2nd month of the dose of 2mg/kg, with passive physiotherapy. At the fourth treatment, the patient no longer presents any cutaneous signs; he walks normally with muscle testing scored at 5 (against resistance) for the proximal muscles. Apart from the presence of an overlap syndrome or the coexistence of another autoimmune disease (which is not the case in this observation) immunosuppressants have no place in the treatment of classic dermatomyositis . It is therefore IVIG which have an indication in corticosteroid-resistant dermatomyositis as a second intention, by their immunomodulatory action: in monthly cures at a rate of 2g/kg/cure and especially in the face of swallowing disorders linked to progressive myopathies.

Conclusion:

IVIG with immunomodulatory indication find their place in the face of corticosteroid-resistant dermatomyositis with dysphagia as demonstrated in our case.

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Abstract N°: 2396

Avapritinib Improved Skin Findings In Patients With Indolent Systemic Mastocytosis (ISM) In the Registrational, Double-Blind, Placebo Controlled PIONEER Study

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Introduction & Objectives:

ISM, characterized by mast cell (MC) accumulation driven by D816V-mutant KIT, can be associated with debilitating symptoms including skin lesions. Skin findings, including MC burden and immunophenotype, were characterized in patients with ISM treated with avapritinib, a selective KIT D816V inhibitor, vs placebo in PIONEER

Part 2 (NCT03731260).

Materials & Methods:

Objectives included change from baseline to 24 weeks in spot symptom and skin domain on ISM-Symptom Assessment Form (©2018), skin lesions by photography, and MC number and immunophenotype (CD25+/CD30+) in skin biopsies assessed via light microscopy and immunohistochemistry.

Results:

212 patients received best supportive care plus avapritinib 25 mg once-daily (n=141) or placebo (n=71). Mean (SD) change was significantly greater with avapritinib in spot severity at 24 weeks (−1.86 [2.24]) vs placebo (−0.63 [1.48]; p<0.0001), and skin symptom domain (−5.87 [6.49] vs −2.64 [4.11]; p<0.0001). Skin domain and MC-QoL improvements were correlated (R=0.578; p<0.0001). In patients with paired photographs, mean percent reduction (SD) in lesion surface area was −37% (53) with avapritinib vs −2% (14) with placebo in most affected skin region; 86% vs 0% had lightened skin lesion color. Mean (SD) percent change in MC burden decreased at 24 weeks with avapritinib (−22% [106], n=87) but increased with placebo (10% [121], n=49). Avapritinib significantly* decreased CD30+ MC proportion in skin lesions at 24 weeks vs placebo (−14% vs −0.47%; p=0.0015).

Conclusion:

Avapritinib-treated patients experienced marked reductions in skin symptoms, skin lesion area, and MC burden; an organ target that has not been addressed before. These findings suggest avapritinib treatment leads to disease modification.

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Abstract N°: 2435

Core signs associated with the subtypes of cutaneous lupus erythematosus: concept elicitation interviews with dermatologists and rheumatologists

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Introduction & Objectives:

Greater understanding of cutaneous signs and symptoms is needed to comprehensively define and measure cutaneous disease activity in lupus erythematosus. Herein, we interviewed dermatologic and rheumatologic experts: (I) to explore perspectives regarding key signs and symptoms for defining disease activity and damage in cutaneous lupus erythematosus (CLE) subtypes (chronic [primarily discoid], subacute and acute [CCLE [DLE]/SCLE/ACLE]) and (II) to explore potential differences and similarities in clinical signs and symptoms across these CLE subtypes.

Materials & Methods:

This was a** non-interventional, mixed-methods research study, integrating qualitative interviews and latent consensus assessments. Board-certified dermatologists (n=11) and rheumatologists (n=10) based in the United States and Europe with ≥5 years' experience in treating CLE were interviewed remotely, using a semi-structured interview guide (Figure 1). Data were evaluated by thematic analysis.

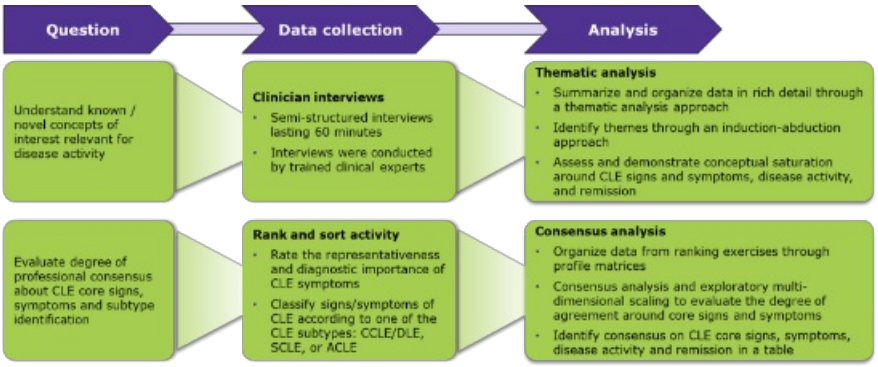
Results:

Skin lesions in CLE show a broad spectrum of clinical manifestations and, therefore, it is important to use validated skin scores and to evaluate and distinguish disease activity and damage. The most frequently mentioned signs of disease activity by dermatologists included erythema (11/11) and infiltration/edema (10/11) followed by alopecia (scarring and non-scarring), malar rash, scaling and dyspigmentation (8/11 for each) (Figure 2A). The rheumatologists also reported erythema (10/10) as the most frequently mentioned signs of disease activity, followed by alopecia (scarring and non-scarring), malar rash, scaling, dyspigmentation and infiltration/edema (Figure 2A). Itch was the most frequently reported symptom by dermatologists (10/11) whereas photosensitivity was the most frequently reported symptom by rheumatologists (8/10; Figure 2B). Erythema and scaling were associated with all subtypes, but infiltration/edema, mucous membrane involvement, alopecia and non-scarring alopecia varied across subtypes (Figure 3). Most clinicians mentioned that CLE progression is often unpredictable, varied across patients and may be influenced by multiple factors including CLE subtype, sun exposure, smoking and medication compliance. The clinical experts characterized remission as the absence of signs and symptoms, with or without medication. CLE remission is negatively influenced by sun exposure and smoking, whereas medication compliance positively impacts duration of remission.

Conclusion:

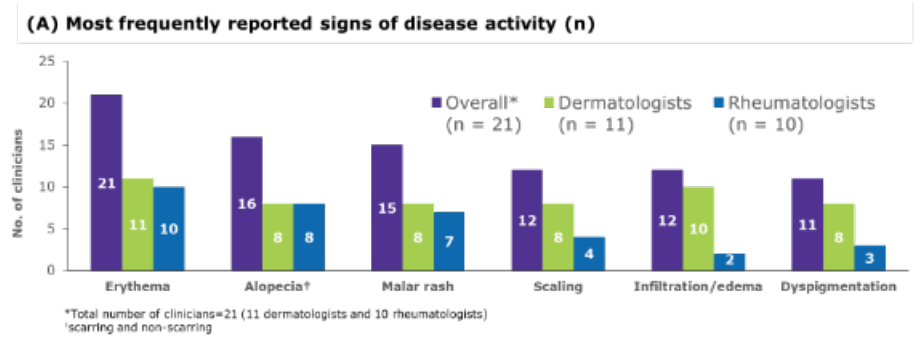
Signs of CLE associated with disease activity may vary across subtypes and must be considered in the definition and measurement of disease activity. Erythema was the most frequently reported sign related to disease activity and was considered relevant across all CLE subtypes.

Figure 1. Methods: Data collection and analysis



The qualitative interviews were analyzed using MAXQDA 2022, whereas the ranking data were analyzed using consensus analysis in UCINET software.
ACLE, acute cutaneous lupus erythematosus; **CLE**, cutaneous lupus erythematosus; **CCLE**, chronic cutaneous lupus erythematosus; **DLE**, discoid lupus erythematosus; **SCLE**, subacute cutaneous lupus erythematosus.

Figure 2. Most frequently reported signs and symptoms of CLE



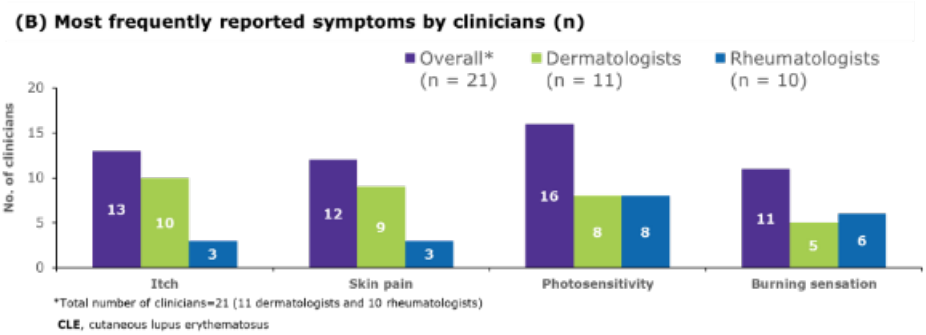
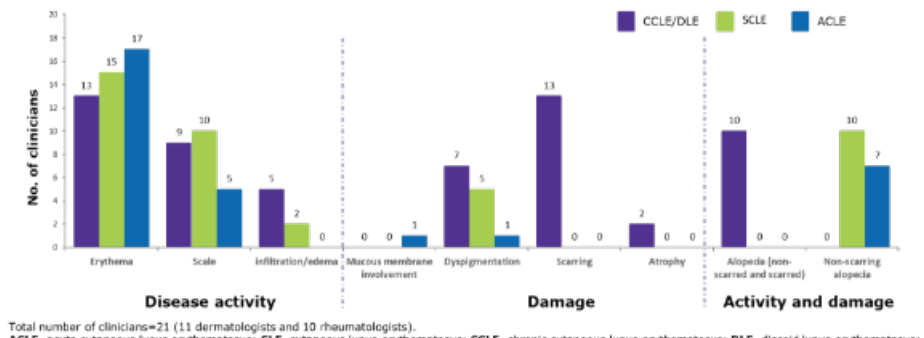


Figure 3. Signs of CLE subtypes as per clinicians



Total number of clinicians=21 (11 dermatologists and 10 rheumatologists).
ACLE, acute cutaneous lupus erythematosus; **CLE**, cutaneous lupus erythematosus; **CCLE**, chronic cutaneous lupus erythematosus; **DLE**, discoid lupus erythematosus; **SCLE**, subacute cutaneous lupus erythematosus.

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Abstract N°: 2587

Scleroderma-like skin lesions in a young patient with phenylketonuria: a case report

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Introduction & Objectives:

Phenylketonuria (PKU) is an autosomal recessive disorder of phenylalanine metabolism. Cutaneous lesions in patients with PKU include pigmentary changes in hair, eye and skin, eczematous or atopic dermatitis and scleroderma-like changes.

Materials & Methods:

A 15-year-old female patient presented to our department with a 6-month history of a hyperpigmented skin rash. Skin examination revealed a generalized skin eruption consisting of multiple well-defined hyperpigmented macules and plaques with a slight induration on the trunk and on the upper and lower extremities. Our patient had a history of phenylketonuria treated with a phenylalanine restricted diet. However, the phenylalanine restricted diet was stopped by our patient, which had as a consequence an elevation of serum phenylalanine concentrations. A few months later the skin lesions were first appeared. The histopathologic examination of a skin lesion revealed a normal epidermis and a papillary dermis slightly homogenous with mild lymphohistiocytic perivascular infiltrate. The collagen tissue in the reticular dermis was slightly densified, with broadened and hypereosinophilic collagen fibers. There has been no clinical or laboratory evidence of systemic scleroderma. Antinuclear antibodies (ANA) were negative.

Results:

The patient's history of phenylketonuria, the fact that the skin lesions appeared with the elevation of serum phenylalanine concentrations, the clinical findings and the histopathologic changes consistent with scleroderma indicated the diagnosis of sclerodema-like skin eruption.

Conclusion:

Phenylketonuria is a rare disease that may be associated with skin disorders, such as hypopigmenatation, including the classical triad of blond hair, blue eyes, and UV light-sensitive pale skin, eczema or atopic dermatitis and rarely with scleroderma-like skin changes. Our patient had a history of phenylketonuria and developed scleroderma-like skin lesions only after discontinuation of the low-phenylalanine diet. We emphasize that, given the fact that some cases of the disease may be missed at birth, children with skin changes suggestive of scleroderma, especially in the presence of mental and motor retardation, should be investigated for the possibility of PKU.

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Abstract N°: 2820

Combination intravenous immunoglobulin, oral prednisone and methotrexate for managing scleromyxedema

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Combination intravenous immunoglobulin, oral prednisone and methotrexate for managing scleromyxedema

Introduction & Objectives:

Scleromyxedema (SMX), the generalized and sclerodermic form of lichen myxedematous (LM), is a chronic mucinosis characterized by cutaneous manifestation and several systemic comorbidities. Data regarding optimal therapy are lacking due to the low number of cases and the absence of randomized trials.

We report a case of SMX with monoclonal gammopathy and a prevalent joint involvement that has been successfully treated with IVIg in combination with oral corticosteroids and methotrexate (MTX).

Materials & Methods:

A 48-year-old man with a background history of hypertension and hepatitis A infection was referred to his primary care physician for smooth papules on his trunk and extremities that have been present for two years. The patient also complained of bilateral joint arthralgia, generalized weakness, and numbness of the hands.

Histopathology from a representative lesion on the hands, showed atrophic skin with circumscribed mucin deposition in the reticular dermis, irregular fibroblast proliferation, and fibrosis, in the absence of inflammatory infiltrate.

Serum protein electrophoresis showed a monoclonal IgG gammopathy with a predominance of kappa light chains. Abnormal investigations included elevated ESR and CRP.

Clinical, laboratory, and pathologic parameters were consistent with a diagnosis of SMX.

Results:

Systemic therapy was carried out with intravenous immunoglobulin (IVIg) at the standard dose (2 g per kg per cycle administrated throughout four days). IVIg administration was carried on at a 4-week interval for 6 months with a significative improvement of the skin manifestations.

Persistence of arthralgia and articular symptoms continued to be reported, therefore, systemic glucocorticoids (prednisone 0,5-1 mg/kg/day) and methotrexate (15 mg subcutaneous injection once a week) with folic acid (one 5 mg tab the day after methotrexate administration) were used together with IVIg to enhance response.

Skin and joint symptoms had a progressive improvement during the 6 months of therapy. Indices of serum inflammation went back within normal limits and the monoclonal gammopathy gradually improved.

No drug reactions or other side effects were reported.

Systemic therapy with IVIg is the most used therapy for SM.X

Even when therapy is successful, relapse commonly occurs upon the discontinuation of treatment, and long-term maintenance infusions every 6-8 weeks are generally required.

Joint symptoms may frequently be resistant to IVIg therapy and, as our ca case report shows, other therapies such as oral corticosteroid and methotrexate with folic acid can be combined to manage both skin disease and extra-cutaneous symptoms. No side effects were reported during the treatment period.

Conclusion:

In conclusion, SM is an unpredictable disease with different and severe systemic manifestations, which can even lead to death. Treating SM still represents a challenging task for dermatologists.

To date, our experience is the first described report on a combination of IVIg, MTX, and oral corticosteroid and represents a successful combined management of both skin disease and extra-cutaneous manifestations.

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Abstract N°: 2948

cutaneous familial lysozyme amyloidosis presenting as bullae - a case report

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Cutaneous Familial Lysozyme Amyloidosis presenting as Bullae - a Case Report

Introduction & Objectives:

Familial lysozyme amyloidosis is an extremely rare autosomal dominant disorder that results in the deposition of abnormal lysozyme proteins in various organs and tissues, leading to organ dysfunction. Cutaneous manifestations of the disease are not commonly reported in the literature. We present an unusual case of a 56-year-old woman with a known history of familial lysozyme amyloidosis who presented with bullous lesions over her skin.

Materials & Methods & Results (Background of case):

A 56-year-old woman with a history of familial lysozyme amyloidosis presented with acute onset blistering and epidermal loss 24 hours post renal transplant. Examination revealed tense linear bullae with straw-coloured fluid under her left breast and sharply angulated erosions of full thickness epidermal loss under her right breast, her arms, abdomen, and groin. These lesions developed after dressings and ECG tab removal from the skin. Five days later, the patient developed new lesions on her neck where adhesive tape that was securing her central line was removed. The patient reported no previous history of skin disease.

Differentials that were considered included staphylococcal-scalded-skin syndrome, cutaneous porphyria, pseudo-porphyria, linear IgA disease, bullous pemphigoid, and cutaneous amyloidosis.

Skin punch biopsy was performed from the lesion under the left breast. Histopathology revealed a subepidermal bulla and further testing with Congo red staining displayed amorphous salmon pink material along the dermal-epidermal junction. When viewed under high-intensity cross-polarized light, apple green birefringence was observed, consistent with the presence of amyloidosis. Based on the patient's clinical history, a diagnosis of cutaneous manifestation of lysozyme amyloidosis was made.

Conclusion:

This case emphasizes the need to recognize cutaneous signs of amyloidosis when evaluating patients with blistering skin lesions, especially if they have a prior history of amyloidosis. Being more familiar with the unusual ways in which amyloidosis can present, including its effects on the skin, can help with prompt identification and treatment.

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Abstract N°: 2977

Colistin induced acquired Bartter-like syndrome in patients of pemphigus vulgaris

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Introduction & Objectives:

Colistin is a polymyxin antibiotic that exerts bactericidal activity against Gram-negative organisms by disrupting the cell membrane. Owing to its nephrotoxicity, colistin sulphate was discontinued in 1970s. However colistimethate (more purified form with relatively lower nephrotoxicity) has now re-emerged as a salvage therapy in multidrug-resistant infections due to limited available alternatives. Systemic use of colistin has been reported to cause acute kidney injury in the form of increasing creatinine, decreasing glomerular filtration rate but little is known about colistin-induced renal tubular damage which maifests with electrolyte and acid base disorders resembling bartter syndrome.

Materials & Methods:

We describe two cases series of acquired Bartter-like syndrome(BLS) due to colistimethate in pemphigus vulgaris patients.

Results:

Case1: A 42 year old female, known case of pemphigus vulgaris was admitted for fever, chills and burning micturition.She was given Inj colistin loading dose of 9 million IU and maintenance of 4.5 million IU twice daily owing to positive urine culture of klebsiella pneumoniae sensitive only to colistin. 6 days after starting therapy, there was hypokalemia, hypocalcaemia, hypophosphataemia, hypomagnesaemia and metaboic alkalosis whereas serum creatinine levels and urine output were normal. Urinary potassium was 22 mEq/L confirming renal loss. Electrolyte imbalances persisted despite multiple corrections. A diagnosis of BLS was made and colistin was stopped on day 12 after resolution of UTI and negative cultures. Electrolytes were restored after 9 days of stopping colistin.

Case 2: A 55 year old female, newly diagnosed case of pemphigus vulgaris was admitted for the management of pemphigus. Blood culture revealed klebsiella pneumoniae with intermediate sensitivity to colistin. Inj colistin 9 million IU followed by 4.5 million IU 12 hourly was initiated. After 8 days of therapy, patient had developed hypokalemia, hypomagnesemia, and metabolic alkalosis. Urinary potassium level was 55 mEq/L, urinary chloride level was 108 mEq/L, and urinary loss of calcium leading to a diagnosis of BLS. After 10 days of stopping therapy with colistin, patient improved spontaneously

Conclusion:

Bartter-like syndrome is a rare complication associated with colistin. A close monitoring of serum and urine electrolytes is thus prudent. Early diagnosis and prompt treatment can avert the risk of fatal complications.

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Abstract N°: 3012

Dermocosmetics in Management of Cancer-Related Skin Toxicities: International Expert Consensus

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Introduction & Objectives: Skin toxicities – one of the most frequent adverse events associated with cancer therapies – can occur with all types of cancer therapeutic interventions. Further, new side effects emerge as new oncology drugs are approved (eg, targeted therapies, immunotherapies), which are associated with a negative impact on quality of life, oncologic treatment dose reductions and/or treatment discontinuation. From a pathophysiologic point of view, skin toxicities during cancer treatment result mostly from alterations in skin barrier function, with altered immune functions inducing inflammation, and phototoxicity. Minimizing alterations in skin barrier function and prescribing a photoprotection facilitate prevention and management of adverse events to optimize treatment outcomes.

Materials & Methods: In partnership with the Association Francophone des Soins Oncologiques de Support (AFSOS) and Multinational Association of Supportive Care in Cancer (MASCC), a group of international experts (onco-dermatologists, oncologist, radiation oncologist, oncology nurse) all involved in management of cancer patients performed a literature review and a consensus meeting to define skin care including dermocosmetics to both to prevent and or manage skin adverse events induced by cancer drugs.

Results: Dermocosmetics, or cosmeceuticals, include a range of products that can have both therapeutic and cosmetic value. These products help support and maintain the epidermal skin barrier with all its functions and in addition the cutaneous microbiome. Some have been formulated for skin that is particularly fragile, and sensitive. For general recommendations,

1- Skin care should be implemented as soon as at initiation of any anticancer drugs associated with skin toxicities, since prevention is a crucial aspect of management.

2- Skin hydration relieves symptoms and reduces exacerbations of xerosis and itching.

3- Cleansers should have a pH close to 5, while basic and neutral pH cleansers should be avoided inducing dysbiosis.

4- Urea, particularly in the case of anti-cancer drugs leading to hyperkeratosis and hand foot syndrome, is important due to its exfoliating and hydrating actions.

Finally, to help to have a global approach of the skin toxicities, the group also developed recommendations for the management of specific skin toxicities depending on their severity.

Conclusion: Emerging evidence is providing support for the beneficial impact of dermocosmetics in management of treatment-related skin toxicities targeting mainly skin barrier and microbiome, together with photoprotection.

These recommendations include as well the need for education of all healthcare providers and patients.

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Abstract N°: 3143

Graft-versus-host disease in patients with hematopoietic stem cell transplantation: a retrospective study with 1,507 patients and its relationship with skin cancer

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Introduction & Objectives:

Graft-versus-host disease (graft-versus-host disease) is a common medical complication in hematopoietic stem cell transplants. It is mainly associated with alogeneic transplantation since, ultimately, it is a complication since the immune cells present in the transplanted tissue recognize the recipient of the transplant (the host) as "foreign." The transplanted immune cells, once activated, attack the recipient's cells, causing the disease.

Graft-versus-host disease can affect any part of the body, with the skin and mucous membranes being a frequently implicated system. Otherwise, classically it has been divided into acute graft-versus-host disease, if it occurs within 100 days of the transplant, or chronic, if it occurs after those 100 days. However, currently there is a tendency to consider that these differences should be made based on the accompanying symptoms, and not on chronological criteria. We lack large series that characterize this entity as well as its possible association with skin cancer.

Materials & Methods:

All patients who underwent hematopoietic stem cell transplantation in a third-level center between June 2007 and June 2020 were retrospectively collected and their baseline characteristics, skin involvement in case of graft-versus-host disease, its extension, and clinical and histological grade and the possible and type of skin cancer if present.

Results:

1,507 patients were collected. 528 patients (35.08%) presented acute GVHD, of which 399 (75.57% of acute GVHD) presented skin involvement. 498 patients (33.09%) presented chronic GVHD, of which 327 (65.67% of the total chronic GVHD) presented cutaneous or mucosal involvement. The majority (>70%) of the patients who experienced acute or chronic GVHD did not present a severe condition, the most frequent being grades 1 and 2, both clinical and histological. There was a 68% correlation between clinical and histological grade.

Regarding skin tumors, an association was found with chronic GVHD, especially with skin or mucosal involvement. The most frequent tumor was basal cell carcinoma (60%) and located on the face or scalp (70%).

Conclusions:

\1. Graft-versus-host disease is a very common condition in patients undergoing hematopoietic stem cell transplantation: its incidence, location, graduation, and treatment are presented.

\2. Skin cancer is increased in patients with injectable chronic host disease, especially if there is skin or mucosal involvement, in elderly patients, and in patients undergoing voriconazole as part of their antifungal prophylaxis.

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Abstract N°: 3205

Prevalence of cutaneous manifestations and its associated factors among patients with chronic kidney disease at Mnazi Mmoja referral hospital, Zanzibar

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Introduction & Objectives:

Cutaneous manifestations are common in all stages of Chronic kidney disease (CKD) particularly towards end stage with a prevalence of 50–100 %. Their management is difficult in low income countries due to high cost of dialysis. The presentation varies based on the stage of the disease.

The aim of the study was to determine the prevalence of cutaneous manifestations and its associated factors among patients with stage III to V chronic kidney disease at Mnazi Mmoja referral Hospital, Zanzibar.

Materials & Methods:

This was a cross-sectional study where a total of 86 patients met the criteria and recruited for the study from September to October 2021. The data were collected using a questionnaire and diagnosis were made by a dermatologist.

Results:

The mean age of the CKD patients was 49.5 (38.7 – 60.0) years. Forty nine (49) of the 86 patients (57%) were on haemodialysis while 37 (37%) were on conservative management. 70 patients (81.4%) had at least one skin problem. The most common skin disorder seen was xerosis 61 (70.9%), followed by pruritus 42 (48.8%) and hyperpigmentation 18 (20.9%). Half and half nail 20 (23.3%) and xerostomia in 37 (43%) was the commonest nail and oral presentation respectively. There was significant correlation between CKD, xerosis and pruritus (P. value <0.001).

Conclusion:

Cutaneous manifestations in CKD are common at our centre, they varies from pruritus, xerosis and hyperpigmentation to perforating dermatosis and scalp alopecia. They were observed more on patients in hemodialysis however there was no significant association. Special consideration should be put in patients with CKD on dialysis to decrease cutaneous discomfort.

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Abstract N°: 3371

Primary cutaneous endometriosis: a rare entity

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Introduction & Objectives:

Materials & Methods:

Results:

Cutaneous endometriosis is a rare subtype of extrapelvic endometriosis, which accounts for less than 5.5% of all endometriosis cases. It is associated with concomitant pelvic disease in about 25% of patients and frequently occurs after an abdominopelvic surgery, typically a caesarean section. When it appears in the absence of prior surgery it is referred to as primary or spontaneous cutaneous endometriosis. Primary cutaneous endometriosis represents less than 30% of cutaneous endometriosis and is usually located in the umbilical region.

A 32-year old nulliparous female patient presented to our department with an 8-month history of an umbilical lesion, with recurrent pain and swelling. Physical examination revealed a firm and painless 0.8 cm-sized dark brownish papule in the umbilicus. The patient had no relevant past medical history and denied prior surgeries. A biopsy was performed and the histopathological examination revealed glandular structures (CK7+, CK20-) without atypia, surrounded by edematous stroma, periglandular and perivascular lymphocytic infiltration, as well as focal hemorrhage and areas of hemosiderin deposition. A diagnosis of cutaneous endometriosis was established and the patient was referred to the gynecological department to exclude pelvic endometriosis and perform surgical excision.

Many theories have been proposed regarding the pathogenesis of primary cutaneous endometriosis. The most widely accepted include hematogenic or lymphatic migration or remnants of embryonic cells in the umbilical fold. This clinical case describes an uncommon presentation of cutaneous endometriosis, in the absence of previous surgery or known pelvic endometriosis. In addition, the relationship between the recurrent symptoms and menstrual cycle was difficult to establish. In fact, a strong clinical suspicion is needed, and cutaneous endometriosis should always be considered in the approach to an umbilical lesion, even in absence of typical cyclical symptoms.

Conclusion:

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Abstract N°: 3380

Botulinum toxin for Raynaud's phenomenon: a decade of real-life experience

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Introduction & Objectives: In the last few years, improvement of Raynaud's phenomenon after the administration of botulinum toxin has been described. However, the number of patients included in the real-life studies and their follow-up period are limited. Only one study analysed possible predictors of therapeutic response. The aim of this study is to evaluate the effectiveness and safety of botulinum toxin for Raynaud's phenomenon in real clinical practice and to analyse potential predictors of better therapeutic response.

Materials & Methods: Single-centre retrospective study of patients treated with botulinum toxin for Raynaud's phenomenon between January 2013 and January 2023. In the long-term study, we included patients with initial response and more than one year of follow-up.

Results: A total of 44 patientes were included. The most frequent diagnosis was systemic sclerosis (59.1%). The rest of the patients presented primary Raynaud's phenomenon (11.4%), lupus erythematosus (4.5%) or other connective tissue disease (13.6%). Twenty-five percent were smokers and 29.5% ex-smokers. In addition, 31.8% had digital ulcers. The total dose was 100 IU in 77.3% and 40-96 IU in the rest.

One month after toxin administration, 70.5% of patients reported global improvement. The mean pain score (Visual Analog Scale, VAS, 0-10) was 7.52 pre-treatment and 3.21 one month later (Wilcoxon $p<0.001$). We analyzed whether treatment response could be influenced by any of the following variables: age, sex, diagnosis, smoking, presence of ulcers, dose 100 IU *vs* <100 IU and baseline pain (VAS). We only found a significant association ($p=0.024$) with baseline pain VAS score. Mean baseline VAS was 8.00 (SD 0.34) in patients with improvement and 6.48 (SD 0.61) in those without.

In the long-term study, 24 patients were included, with a median follow-up of 4.46 years (SD 2.7; range 1-9 years). Of these, 83.3% required to repeat toxin administration after a median of 12 months. Ninety percent of the patients also improved with successive doses. On the other hand, 16.7% did not require repeat infiltrations after the improvement achieved with the first dose.

Eight patients (18.1%) presented any adverse effect: intense pain during (2) or after (3, transitory) infiltration and transitory decrease in manual strength (3). All patients who received several doses reported better or equal tolerance with respect to the first dose, with no new adverse effects.

Conclusion: We present the real-life study of botulinum toxin for Raynaud's phenomenon with the largest number of patients and with the highest mean and maximum long-term follow-up.

Our data support the effectiveness of botulinum toxin administration in most patients, with an adequate adverse effect profile. Most patients require to repeat treatment after approximately one year, with a long-term effectiveness and tolerability similar to the first dose. Baseline pain (VAS score) could influence treatment response.

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Abstract N°: 3446

Assessment of Platelet Function Disorders in Patients Presenting with Hemostatic Disorder-Related Cutaneous Manifestations

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Introduction & Objectives:

Subcutaneous bleeding can manifest as petechiae (small red dots), purpura (larger flat areas), or ecchymosis (very large bruised areas). These symptoms can arise from coagulation disorders or non-accidental injury, highlighting the crucial need for specialists to differentiate between the two. It is worth noting that many of these patients appear normal during the initial laboratory evaluation, leading to missed diagnoses of the underlying cause. The objective of this study was to assess platelet function disorders in patients presenting with cutaneous manifestations related to hemostatic disorders, despite having normal coagulation screens and full blood counts.

Materials & Methods:

A prospective cross-sectional study was conducted on patients referred for subcutaneous bleeding symptoms. Initial screening evaluations, including prothrombin time (PT), activated partial thromboplastin clotting time (aPTT), and complete blood count (CBC), were performed. Subsequently, patients with normal results in the initial screening tests were selected for platelet function assessments. Platelet aggregation was evaluated using a turbidometric method, measuring aggregation in response to Collagen, Adenosine Diphosphate (ADP), Arachidonic Acid, and Ristostin agonists. The results were reported as the maximum amount of transmitted light per percent unit within a 4-minute timeframe.

Results:

The study included a total of 711 patients with a median age of 29 years, ranging from 1 month to 90 years old. The majority of patients were female, accounting for 79.9% (568 individuals). A familial history of bleeding disorders was reported in only 18 patients (2.5%). Among the participants, 81.99% (583 patients) had normal results for prothrombin time (PT), activated partial thromboplastin clotting time (aPTT), and platelet count. Abnormal responsiveness to Collagen, Adenosine Diphosphate (ADP), Arachidonic Acid, and Ristostin agonists was observed in 18.8%, 18%, 17.9%, and 11% of the patients, respectively. Specifically, 47 patients (6.6%) exhibited abnormal PT values ranging from 14.5 to 20.8 seconds while having normal aPTT results. Additionally, 62 patients (8.7%) had abnormal aPTT values but normal PT results. Finally, 20 patients showed abnormalities in both PT and aPTT values.

Conclusion:

The study's findings, based on a substantial cohort of patients with cutaneous bleeding, indicate that the majority of these individuals are initially classified as normal through standard laboratory evaluations, including platelet count, prothrombin time (PT), and activated partial thromboplastin time (aPTT). However, supplementary assessments reveal that platelet function disorders are a frequent observation. Consequently, it is recommended to incorporate platelet aggregation assessment as a diagnostic tool for these patients. By implementing this approach, clinicians can more accurately identify and diagnose platelet function disorders in individuals presenting with cutaneous bleeding symptoms.

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Abstract N°: 3599

Tape Strip Genomic Profiling Identifies Abrocitinib-Mediated Changes in Immune, Barrier, and Itch Biomarkers in the Skin of Patients with Moderate-to-Severe Atopic Dermatitis: A Post Hoc Analysis of the Phase 2a trial JADE MOA

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Introduction & Objectives:

Atopic dermatitis (AD) is a chronic inflammatory skin disease involving several cytokine pathways. Abrocitinib, a Janus kinase 1-selective inhibitor, is approved for the treatment of moderate-to-severe AD. This analysis aimed to characterize the effect of abrocitinib on AD biomarkers using tape strip-mediated sampling, a novel, noninvasive skin sampling approach to evaluating treatment response.

Materials & Methods:

Data were used from adults with moderate-to-severe AD who were randomly assigned 1:1:1 to receive abrocitinib (200 mg or 100 mg) or placebo for 12 weeks in the phase 2a trial JADE MOA (NCT03915496). Tape strips were collected from lesional skin at baseline, and weeks 2, 4, and 12, and from nonlesional skin at baseline and weeks 4 and 12. Differentially expressed genes (DEGs; fold change >1.5 and false discovery rate [FDR] <0.1) were identified using RNA sequencing. Correlations between biomarker expression and disease severity as assessed by Eczema Area and Severity Index (EASI), body surface area (BSA) affected, Investigator's Global Assessment (IGA), and pruritus numerical rating scale (NRS) scores were determined using Spearman correlations.

Results:

Of 46 patients in JADE MOA, 14, 16, and 16 patients received abrocitinib 200 mg, abrocitinib 100 mg, and placebo, respectively. Tape-strip analysis identified 651 DEGs (505 upregulated, 146 downregulated) at baseline between lesional and nonlesional skin across treatment arms. Abrocitinib 200 mg normalized the lesional AD molecular profile toward that of the nonlesional skin, with 67%, 91%, and 109% improvements seen at weeks 2, 4, and 12, respectively; with abrocitinib 100 mg, improvements were 18%, 44%, and 64%. The expression of key AD-associated biomarkers was significantly modulated in both lesional and nonlesional skin in response to treatment with abrocitinib at either dose, with greater improvements seen with abrocitinib 200 mg (**Table**). Decreased expression of immune genes (innate immunity: IL-6; Th1: IFNGR1/2, MX1, STAT1, CXCL10; Th2: IL-4/13/31/4R/7R, CCR4; Th17/22: STAT3, S100A7/8/9, IL-36A/G) and itch-related markers (IL-31, SERPINB1, CTSS, TRPV2, KLK6/7, ORMDL3, NR2F2), and increased expression of epidermal lipid and barrier products (LPL, FADS2, CDH12/19) were observed in lesional and/or nonlesional skin (FDR<0.1 for all). Clinical severity measurements (EASI, BSA, and/or IGA) correlated significantly and positively with markers related to T-cell activation (ICOS), Th2 inflammation (CCL17/CCL22, OX40, IL-7R), Th22/epidermal hyperplasia (S100A7), and Th1 inflammation (IFNGR2) (r>0.4; P<0.05 for all). Itch severity measurements (NRS) correlated significantly with itch-related biomarkers (TRPV2, RAMP1, AHR), and with immune and epidermal barrier biomarkers (FCER1a, OX40, IL-7R, S100A7/8/9, SERPINB1, CCL17/CCL22 (r>0.4; P<0.05 for all).

Conclusion:

Treatment with abrocitinib resulted in significant, dose-dependent, and progressive changes in the expression of AD biomarkers associated with the epidermal barrier, itch, and Th1, Th2, and Th17/Th22-mediated inflammation in AD skin. Improvements in AD biomarker expression correlated with improvements in itch and disease severity. Together, these results suggest that tape strips accurately capture molecular changes with treatment over time in both lesional and nonlesional skin and may be a useful tool to objectively evaluate changes in skin activity in clinical trials.

Table. Fold Changes From Baseline in the Expression of Select AD-Associated Biomarkers in Nonlesional and Lesional Skin by Treatment Group Over Time

		Placebo				Abrocitinib 100 mg				Abrocitinib 200 mg			
Gene Group	Gene	W2 (LS)	W4 (LS)	W12 (LS)	W12 (NL)	W 2 (LS)	W4 (LS)	W12 (LS)	W12 (NL)	W 2 (LS)	W4 (LS)	W12 (LS)	W12 (NL)
Innate	*IL-6*	−1.2	−1.6	−2.4^{+}	−2.4	1.6	−1.6	−2.6^{+}	−1.0	−2.5	−10.9***	−6.6**	−4.3*
Th1-mediated inflammation	*IFNGR1*	1.0	1.3	−1.0	1.1	1.1	−1.1	−1.4	−1.5^{+}	−1.1	−1.2	−2.5***	−1.7^{+}
	IFNGR2	−1.0	1.1	−1.2	−1.4	1.1	−1.2	−1.4	−1.1	−1.3	−1.3	−2.2**	−1.5
	MX1	−1.1	1.2	−1.5	−1.7	−2.6**	−2.9**	−1.8^{+}	−3.0**	−4.8***	−8.6***	−6.0***	−2.2^{+}
	STAT1	−1.1	1.2	−1.3	−1.0	−1.6^{+}	−1.6^{+}	−1.3	−1.5	−2.7**	−1.8^{+}	−4.3***	−1.9^{+}
	CXCL10	−1.5	−1.4	−3.1^{+}	−1.1	−8.8***	−9.1***	−2.6	1.6	−5.8**	−11.9***	−3.4^{+}	−6.8*
Th2-mediated inflammation	*IL-4*	1.2	1.4	−1.3	−1.8	1.4	1.0	1.6	−1.7	−2.0	−2.6*	−1.4	1.4
	IL-13	1.1	−1.6	−2.3	−2.5	1.8	−1.4	1.0	1.2	1.1	−4.4*	−5.4**	−4.6*
	IL-31	1.2	−3.5*	−2.5^{+}	−4.0*	1.8	1.1	1.3	1.5	−2.0	−4.0*	−1.9	−2.0
	IL-4R	−1.2	1.1	−1.3	−1.3	−1.1	−1.3	−1.4	1.0	−1.5	−1.8*	−2.3**	−2.1*
	IL-7R	−1.1	−1.2	−1.7	−2.5*	1.4	1.2	−2.5*	−1.6	−1.1	−3.4**	−4.3**	−2.9*
	CCR4	−1.0	−1.1	−1.4	−2.1	2.2^{+}	2.1	−1.4	−1.7	1.4	−2.9*	−2.2	−2.4
Th17/Th22-mediated inflammation	*STAT3*	−1.3	−1.2	−1.0	1.3	−1.1	−1.3	−1.1	−1.7*	−2.2***	−2.3***	−2.2**	−1.4
	S100A7	−1.1	−2.1	2.0^{+}	1.4	−1.4	−2.3*	−1.3	−1.8	−2.1^{+}	−3.3**	−3.2**	−3.5*
	S100A8	−1.3	−1.4	1.4	1.3	−1.5	−2.6*	−1.4	−1.4	−3.3**	−4.0**	−3.6**	−2.9*
	S100A9	−1.4	−1.6	1.2	1.4	−1.4	−2.3*	−1.3	−1.5	−3.5**	−3.6**	−4.1**	−2.7*
	IL-36A	−2.0	−2.1	−1.0	1.4	−2.2	−2.7^{+}	−1.5	−1.5	−9.1***	−22.5***	−6.9**	−1.1
	IL-36G	−1.4	−2.1	1.3	1.4	−2.3^{+}	−2.9*	−1.6	1.1	−5.4***	−4.8**	−4.1**	−1.8
Epidermal lipid and barrier	*LPL*	1.1	1.3	−1.2	−1.3	1.4	1.2	−2.0	−2.0	2.9*	3.3*	1.9	−2.6
	FADS2	1.2	−1.0	1.0	−1.6	1.6	1.7	2.4*	1.2	−1.0	−1.1	2.4*	1.6
	CDH12	−1.1	−1.2	1.7	1.1	−1.7	−1.0	1.1	1.2	1.8	3.2**	3.0*	1.6
	CDH19	1.4	−1.1	1.4	−1.2	−1.7	1.0	1.9	2.4	−1.1	1.5	5.2**	6.1**
Itch	*IL-31*	1.2	−3.5*	−2.5^{+}	−4.0*	1.8	1.1	1.3	1.5	−2.0	−4.0*	−1.9	−2.0
	SERPINB1	−1.3	−1.0	−1.1	−1.2	1.0	−1.4	−1.6	−1.6	−1.5	−1.7^{+}	−3.6***	−2.2*
	CTSS	−1.3	−1.1	−1.5	−1.4	−1.2	−1.3	−1.9^{+}	−1.5	−1.6	−2.4*	−4.2***	−1.9
	TRPV2	−1.0	1.0	−1.3	−2.1^{+}	1.6	1.4	−1.2	1.2	−1.5	−2.5*	−2.4*	−1.2
	KLK6	−1.6	−2.0	1.4	1.9	−2.4^{+}	−2.2	−1.2	−1.8	−4.7**	−4.9**	−2.9^{+}	−1.7
	KLK7	−1.0	−2.3	1.5	1.5	−1.4	−1.7	1.3	−2.1	−4.3**	−3.4*	−2.6^{+}	−1.5
	ORMDL3	1.1	1.2	1.2	1.2	1.3	1.6^{+}	−1.1	1.2	2.0*	1.7^{+}	1.3	−3.4***
	NR2F2	−1.2	−1.8	1.4	1.7	2.1	2.1	3.4*	−1.9	−1.1	−1.3	2.3	4.0*

AD, atopic dermatitis; BL, baseline; LS, lesional; NL, nonlesional; W, week. $^{+}P<.1$, $^{*}P<.05$, $^{**}P<.01$, and $^{***}P<.001$ (versus baseline).

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Abstract N°: 3693

Treatment optimization of squamouse-hyperkeratotic food mycosis in type 2 diabetis mellitus patients

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Introduction & Objectives: Squamous-hyperkeratotic mycosis of feet is the most prevalent dermatological condition in type 2 diabetes patients. The cutaneous symptoms develop slowly, starting with the formation of painless blistering and hyperkeratosis in areas exposed to pressure. The fungal infection adherence is promoted by poor circulation and traumatisation of the feet skin. The selection of a suitable product containing components with distinct keratolytic and hydrating effects is one part of the problem solution.

The **objective** of the study was to investigate clinical efficacy of a foot balm for callus and hyperkeratosis having antifungal effect

Materials & Methods: We observed 32 patients (17 males and 15 females, aged 43 to 53 years) with mycotic foot damage. The diagnosis of foot fungal affection was confirmed by microscopic examination for pathological fungi in the affected foci. The history of diabetes mellitus ranged from 2 to 5 years in all patients.

Results: Before balm was applied to the foot skin with callus and hyperkeratotic sites, a hygiene treatment had been recommended for all patients. The balm was then applied to the affected sites of dry, clean skin twice a day. All patients experienced dry, atrophic, flaky, pink-purple skin on their toe tips, marginal hyperkeratosis of the entire heels, and symptoms of rubromycosis or epidermophytosis in the in-between toe spaces were present. All patients suffered from itching, pain and discomfort when walking.

The balm contains a large amount of urea, which provides a proteolytic effect in the corneal layer contributing to its loosening, which accelerates the exfoliation of keratinized epidermal cells. Climbazole delivers an antifungal effect. Avocado oil saturates the epidermis with lipids, restoring skin elasticity. Allantoin and lactic acid prevent cell dehydration, soften the skin and stimulate its regeneration. Tea tree essential oil produces an antiseptic effect.

Improvement of the clinical picture was observed after 4 weeks, subjective sensations (itching, pain and discomfort while walking) disappeared in most – 18 (56,2%) patients on day 5, in 10 (31,3%) on day 7 and in 4 (12,5%) on day 10; redness and squamosae after 1 week of treatment reduced in 12 (37,5%) patients, after 2 weeks in 15 (46,9%) and after 3 weeks in 5 (15,6%) patients; blistering and hyperkeratosis disappeared in 14 (43,8%) patients after 3 weeks, in 17 (53,1%) – after 4 weeks of treatment and only in 1 (3,2%) patient after 5 weeks of balm application. To monitor treatment, patients were tested three times for the detection of pathological fungi, following the application of the balm after 4-week treatment with an interval of 5 days. Test results were negative in all patients. After 1.5 months, the foot skin regained its natural colour and the hyperkeratotic sites disappeared.

Conclusion: The data obtained confirm the high efficacy of the balm, which can be recommended as a therapeutic and preventive agent in the care of diabetic feet and to remove calluses and hyperkeratotic sites, including those with mycotic damage.**

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Abstract N°: 3717

A longstanding Langerhans cell histiocytosis with pituitary involvement revealed by skin biopsy

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Introduction & Objectives:

When encountering with scaly or crusted lesions on scalp, dermatologists generally consider common disorders, such as seborrheic dermatitis or scalp psoriasis. However, if the lesion does not improve under conventional therapy, rare diseases should be distinguished.

Materials & Methods:

Results:

A 44-year-old woman was referred for itchy erythematous crusted papules on scalp persisted for several years. She previously treated under the impression of seborrheic dermatitis, but there was partial improvement only. Punch biopsy was done, and the result showed Langerhans cell infiltration in both epidermis and dermis with CD1a and CD68 positive. The patient also had ulcerative lesions on her inguinal area. In addition, she had known pituitary tumor, and being treated with hypopituitarism and hypernatremia. Langerhans cell histiocytosis was diagnosed, and we speculate that her pituitary tumor and accompanied symptoms are manifestations of central nervous system involvement.

Conclusion:

In this case, the patient was under allopathic treatment for hypopituitarism and hypernatremia in individual manner, and her pituitary mass was remaining unidentified for years. Her multidisciplinary symptoms finally elucidated by skin biopsy. Since multisystem LCH often affects skin, it is important for dermatologist to play a pivotal role in diagnosis. We report a case of LCH on scalp with pituitary involvement to highlight the dermatologist's role in multiorgan disease with skin involvement.

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Abstract N°: 3763

Efficacy of intravenous immunoglobulins in lupus panniculitis: about a case

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Introduction & Objectives: Lupus panniculitis is a rare anatomo-clinical form of lupus erythematosus. Its gravity lies in its evolution towards unsightly dystrophic scars. It can be isolated or associated with another type of lupus. Synthetic antimalarials are the first-line treatment, sometimes combined with topical and/or systemic corticosteroids

Materials & Methods:

We report a case of lupus panniculitis resistant to conventional treatments which responded rapidly to treatment with intravenous immunoglobulins.

Results:

A 36-year-old woman, mother of 02 healthy children. In her background, there are two early abortions. For the past 20 months, she has had multiple subcutaneous nodules 2 to 4 cm in diameter, surmounted by edema-colored purple-red skin, spontaneously painful, which sits on the postero-internal surfaces of both arms, on the thighs as well as on the cheeks and at the forehead. Examination of the scalp found two erythematous-scaly patches, well limited, measuring 02 cm in diameter, centered by a whitish atrophy.

A series of additional examinations were requested: NFS: Hypochromic microcytic anemia with hyper reticulocytosis, leucopenia, anti-nucleolar AC > 320, anti-C1q AC > 80 mg/L, C4 < 0.2 mg/L, negative anti-phospholipid AC, the search for proteinuria came back negative . A deep biopsy found lobular lymphocytic panniculitis, a perivascular mononuclear infiltrate associated with very marked diffuse hyperkeratosis of the orthokeratotic type. with at the IFD a negativity of all the structures.

Our patient presents with profound lupus during systemic acute lupus erythematosus (LEAD). She initially received treatment with hydroxychloroquine at a rate of 600 mg/day for 4 months. associated with local and systemic corticosteroid therapy at a rate of 25 mg of cortancyl/day as well as rigorous photoprotection. The evolution was made towards aggravation with the appearance of new similar lesions. This motivated the discontinuation of oral corticosteroid therapy and the introduction of intravenous immunoglobulins at a dose of 2g/kg per course, which led to resorption of all the lesions after two months, giving way to atrophic scars (6 courses in total). Lupus panniculitis is rare. It begins between the ages of 20 and 40, with a female predominance (3F/1M), and affects all ethnic groups. Our patient presents more than 04 diagnostic criteria of the ACR classification in favor of the association of a lupus panniculitis and a LEAD.

The treatment of choice remains synthetic antimalarials, mainly hydroxychloroquine at a dose of 6.5 mg/kg/day, which is considered by some to be a real therapeutic test. Systemic corticosteroid therapy is only used in the presence of visceral LEAD involvement. Rigorous, broad-spectrum is essential.

In our patient, conventional therapies were unable to improve the lesions. The introduction of intravenous immunoglobulins at a rate of 2g/kg per cure was followed by a rapid melting of the nodules from the second cure. No side effects were observed following this treatment.

Conclusion:

Immunoglobulins possess major immunomodulatory activity with excellent safety. They can be a therapeutic alternative for refractory forms of lupus panuculitis.

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Abstract N°: 3771

WHO cares for skin changes in children with severe acute malnutrition? Report from a resource-poor pediatric department in Central Africa.

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Introduction & Objectives:

Severe acute malnutrition (SAM) is a life-threatening stage of undernutrition that is most prevalent among children in poor and disadvantaged populations, characterized by food scarcity and high risk of infections.

The World Health Organization has developed a comprehensive guideline for in-hospital management of SAM, that has been adapted by several countries. There is still room for improvement though, with case fatality rates ranging from 8-16% in controlled settings.

Characteristic skin changes in complicated SAM can be severe and widespread. With the lack of well-documented management options, combined with the documented association with poor outcome, the skin is a relevant focus for improvement of management guidelines.

Research on skin changes is though sparse. Studies are typically driven by external initiatives, and thus driven by a foreign agenda creating a mismatch between recommendations and local needs.

The scope of this study was to investigate what it*means* and what it *takes* to care for skin changes in children with complicated SAM, from a local health care perspective. The goal was to identify local driven and realistic suggestions for future research initiatives.

Materials & Methods:

Through participant observation and semi-structured interviews of the mixed professions (nurses, nutritionists, pediatricians, intern doctors) that make up the health staff at Mwanamugimu Nutrition Unit in Uganda, we condensate their perspectives on challenges in their work and their view on skin to reflect on future research initiatives in dermatological management of patients with SAM complicated with skin changes.

Results:

We found that skin changes in SAM are considered a danger sign that*means* death in the eyes of the staff but observe that treatment often does not follow management guidelines. We learn how these children, in the opinion of the staff, would gain from the care proposed in the guidelines. Furthermore, that treatment *takes* money and currently relies on the economy of the caretaker.

Conclusion:

The resulting recommendation is to test treatment options that are preferably without cost for the caretaker. Furthermore, current treatment recommendations should be evidence-based and must be tested on the patient group to avoid implementing unnecessary expenses to the management guidelines.

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Abstract N°: 3879

A tale of chronic diffuse nodular lesions with acral verrucous keratotic plaques

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Introduction & Objectives: ** Erythema elevatum diutinum (EED) is a rare chronic and recurring form of cutaneous leukocytoclastic vasculitis. EED is characterized by persistent asymptomatic to painful, red-violaceous, red–violet to red–brown papules, plaques, and nodules, symmetrically distributed, that favor acral and periarticular regions, especially the extensor surfaces. Less than 20 cases of EED affecting palms and soles were described. The verrucous or warty aspects seen in our patient were only reported twice in the literature.

Materials & Methods:

We report a case of EED with verrucous keratotic plaques of the soles.

Results:

A 70-year-old man with no past medical history, presented with a 20-year history of asymptomatic nodules and plaques located on the hands, feet, forearms, trunk, and nose. Physical examination revealed symmetrically distributed reddish-brown firm nodules on the extensor surfaces of the hands and the forearms associated with red pedunculated and ulcerated nodules. Firm pigmented round and oval plaques were located on the abdomen and upper limbs. Examination of the soles revealed verrucous red-brown keratotic plaques. Dermoscopic examination of the acral nodular lesions showed yellow-white streaks over a yellowish-red background. A pigment network is visible at the periphery of lesions. Some lesions exhibit a rainbow pattern on polarized Dermoscopy.

Histopathological examination of multiple skin biopsy specimens showed fibrinoid necrosis of vessel walls, subendothelial vacuolization with leukocytoclasia, and dense inflammatory infiltrate including neutrophils, lymphocytes and eosinophils. An increased number of vessels, some of which were vertically oriented, were associated with large collagen bundles in a storiform pattern.

Conclusion:

EED is associated with several diseases including hematological abnormalities, infections such as streptococcal infection and HIV, autoimmune and inflammatory diseases.

Extracutaneous symptoms may involve arthralgia, constitutional symptoms, and ocular abnormalities. In our patient, no culprit triggers including HIV infection, autoimmune diseases, malignant hematologic disorders, or IgA monoclonal gammopathy were found.

Dermoscopic features of EED were only described once in the literature. As in our case, yellow-white streaks, and yellow background were features of late lesions. Early lesions show more telangiectasia. The rainbow pattern was not described in EED lesions before.

The diagnosis is based on clinical and histopathological findings. Early lesions are characterized by leukocytoclastic vasculitis with neutrophilic infiltrate admixed with polymorphonuclear cells including macrophages, histiocytes, and fibrin deposits. In the late stage, capillary walls may have fibrinoid necrosis and show concentric or storiform fibrosis. Vertically oriented capillaries can be noted. At this stage, mixed inflammation contains more histiocytes

and granulation tissue. The proliferation of spindle cells and hypertrophy of vascular endothelial cells can be seen in association with cholesterol deposits in tissue cells and extracellular tissue.

Treatment of the associated disease is essential. Dapsone, in monotherapy, is the most widely used treatment and the most effective before the onset of fibrosis. If no improvement is noted, sulphomanides, niacinamide, steroids, colchicine, and oral steroids can be proposed alone or in association with dapsone.

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Abstract N°: 3940

Parry Romberg syndrome : About 2 cases.

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Introduction & Objectives:

Progressive hemifacial atrophy, also known as Parry-Romberg syndrome, is an uncommon degenerative disease characterized by slow, progressive, usually unilateral atrophy of facial tissues, including muscle, bone, and skin.

We report the case of two young patients with emblematic features of progressive hemifacial atrophy.

Materials & Methods:

Results:

Case 1: A 7-year-old female child, without any notable medical history, presents since the age of 5, a progressive right eyebrow alopecia in band and facial asymmetry.

The clinical examination showed a right facial hemiatrophy with alopecia in band on the right eyebrow, multiple rounded hypochromic macules measuring about 1cm each in the jugal region and a right enophthalmos. The ophthalmological examination revealed enophthalmia with a decreased visual acuity to 8/10, a craniofacial scan showed a thinning of the right frontal bone and an atrophy of the soft tissues of the scalp of the right hemisphere on the cerebral level and an atrophy of the soft tissues of the right hemiface. it also showed an asymmetry of the eyeballs, a thickening of the maxillary sinuses with filling of some ethmoidal cells and a thinning of the ascending branch of the right mandibular bone.

The patient was put on oral corticosteroid therapy at 1mg/kg/day, i.e., 20mg/day, associated with methotrexate injections at 0.3mg/kg/week, i.e., 7.5mg/week.

Case 2: An 8-year-old female child, without any significant medical history, presents since the age of 5, a progressive right eyebrow alopecia in band with progressive evolution.

The clinical examination showed an alopecia in band on the right eyebrow with a right facial hemiatrophy. The ophthalmological examination revealed enophthalmia with a decrease in visual acuity to 9/10. A craniofacial scan showed an atrophy of the soft tissues of the scalp of the right hemisphere on the cerebral level, an atrophy of the soft tissues of the right hemiface and an asymmetry of the eyeballs.

The therapeutic decision was to put the patient on oral corticosteroid therapy at 1mg/kg/day, i.e., 25mg/day, associated with methotrexate injections at 0.3mg/kg/week, i.e. 7.5mg/week.

Conclusion:

Parry Romberg syndrome is a very uncommon disease and its etiopathogenesis remains unclear. Clinically, it is characterized by slow progressive hemifacial atrophy of the skin and soft tissues and even of the bone sometimes. Facial muscles may atrophy and lead to facial bone loss. Ophthalmologic manifestations are common in 10 to 35% of cases, with enophthalmos being the most common, as in our patient’s cases where we found many clinical and radiological similarities

The management is both symptomatic, psychological and surgical (reparative) of sometimes socially “broken” patients.

The value of our study lies in the rarity of Parry Romberg syndrome especially within the pediatric population.

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Abstract N°: 3965

Exclusive cutaneous form of Rosai-Dorfman disease : two case reports

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Introduction & Objectives:

Rosai-Dorfman disease (RDD) is a rare, benign non-Langerhans cell histiocytosis primarily affecting the lymph nodes, practically those in the cervical region. Cutaneous involvement is common and can be an initial indicator of the disease. However, diagnosing RDD based on skin manifestations is challenging and delayed.

Materials & Methods:

We report two cases of RDD with cutaneous involvement as the presenting feature.

Case 1- A 26-year-old male presented with brownish-red papular nodules on the face, which had been initially misdiagnosed as mycosis fungoides 6 months ago. A subsequent histological examination revealed a polymorphic inflammatory dermal infiltrate consisting of foamy histiocytes containing lymphocytes or neutrophils exhibiting emperipolesis. Immunohistochemistry demonstrated positive expression of PS100 and CD68, while CD1a expression was negative. The patient had a pure cutaneous form of the disease. Surgical excision was performed on selected facial lesions, while others were left untreated. The lesions spontaneously subsided after six months of follow-up.

Case 2- A 32-year-old male presented with similar brownish-red papular nodules on the face and lower back, which were misdiagnosed as acne for two years. The condition did not respond to treatment with either Cycline or Isotretinoin. Histological examination revealed a polymorphic inflammatory dermal infiltrate with evidence of emperipolesis, similar to the findings in Case 1. Immunohistochemistry demonstrated positive expression of PS100 and CD68, while CD1a was absent, also resembling the results in Case 1. The patient also had a pure cutaneous form of the disease. Local corticosteroid injections were administered to treat the facial lesions, but no improvement was observed after 12 months of follow-up.

Results:

Both cases presented typical clinical features, such as red-brown papular nodules primarily on the face. Histological examination revealed the presence of characteristic but non-pathognomonic emperipolesis, which prompted further immunohistochemical analysis to confirm the diagnosis of RDD. Considering the potential for serious visceral damage reported in the literature, additional screening examinations were performed, including abdominal ultrasound, lymph node evaluation, bone scintigraphy, brain MRI, and eye fundus examination to rule out any involvement of organs or systems outside of the skin. The results indicated that there was no extra-cutaneous involvement in either of the patients. Although spontaneous involution of the disease's lesions is often observed in RDD, we opted to initiate treatment due to the unattractive appearance of the lesions in our patients

Conclusion:

Diagnosing RDD in its isolated cutaneous form can be challenging, as it often requires an anatomoclinical correlation and additional immunohistochemical studies to confirm the diagnosis.

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Abstract N°: 4028

Pure cutaneous IgA vasculitis in an elderly person

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Introduction:

IgA vasculitis, previously known as rheumatoid purpura or Henoch-Schönlein syndrome, is a systemic small-vessel vasculitis with immunoglobulin A deposits. Isolated skin involvement is rarely reported in the literature. We present a case in an elderly person.

Case presentation:

A 74-year-old woman with a history of type 2 diabetes, atrial fibrillation on acenocoumarol, arterial hypertension and gout presented with necrotic-petechial vascular purpura, accompanied by pruritus, of all four limbs, with a predominance in the distal region. The condition had been evolving in flare-ups for more than 3 months in a context of apyrexia and preservation of the general state. No triggering factor and no clinical or paraclinical signs suggesting extracutaneous involvement were identified.

A workup including: viral serologies, serum protein immunoelectrophoresis, ANCA studies, antinuclear antibodies, cryoglobulinemia, did not reveal any abnormalities. Histological analysis of a skin biopsy revealed leukocytoclastic vasculitis with fibrinoid necrosis. Direct immunofluorescence showed vascular IgA deposits. Serum IgA levels were normal. The diagnosis of IgA vasculitis was made. A treatment with colchicine was started at a dose of 1 mg per day allowing a good improvement. The patient achieved complete remission with a 6-month follow-up.

Discussion:

IgA vasculitis mainly affects children, it is rare in adults and even more so in the elderly.

Its pathophysiology is only partially understood. It classically affects the skin, the joints, the digestive tract and the kidney. Vascular purpura skin involvement is almost constant and precedes the other manifestations of the disease in half of the cases. In adult and elderly patients, the disease often evolves in a chronic mode with a worse prognosis due to more frequent and more severe renal involvement. A longer follow-up is necessary since nephropathy has a late start (up to several months) in one-third of cases.

Elevated serum IgA is present in about 60% of cases, but this does not constitute a definite argument for the diagnosis. Isolated cutaneous presentation without the typical features of rheumatoid purpura, as in our case, is rarely described in the literature. It is sometimes associated with IgA monoclonal gammopathy, with or without myeloma. In our patient, this dysglobulinemia was not found. The

therapeutic management is not clearly defined, different options are possible: general corticosteroid therapy, dapsone or colchicine. The latter was effective in our case.

Conclusion:

We report a new case of IgA vasculitis, strictly cutaneous without systemic involvement, in an elderly person.

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Abstract N°: 4080

Skin manifestations and diagnostic evaluation of "Non-celiac gluten sensitivity": a prospective single centric study

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Introduction & Objectives:

Non celiac gluten sensitivity (NCGS) is characterized by intestinal and extraintestinal symptoms related to gluten ingestion in patients who are not affected by celiac disease or wheat allergy. NCGS is a diagnosis of exclusion since diagnostic hallmarks have not been identified yet.

A high percentage of patients affected by NCGS presents with unspecific skin eruptions, typically improving after a gluten-free diet.

Although a double-blind, placebo-controlled, gluten challenge has been proposed as a diagnostic criterion for NCGS diagnosis, its application remains difficult in daily clinical practice.

The main objective of this study is to describe the dermatological manifestations of NCGS and identify potential biomarkers useful for its diagnosis. Furthermore, we aimed to prepare and validate a patch test containing gluten as a non-invasive diagnostic tool for NCGS.

Materials & Methods:

This is a prospective study including patients who received a diagnosis of NCGS with cutaneous manifestation from 2018 to 2023. We collected clinical data regarding gastroenterological symptoms, associated comorbidities, and cutaneous manifestations. Patients were also investigated by direct immunofluorescence from the perilesional skin to investigate possible skin deposits of immunoglobulins and complement fractions. Finally, all the patients were tested with patch test application, prepared using 3 different concentrations of gluten (2%, 5%, 10%) in petrolatum. Patients have been evaluated for any cutaneous reactivity after 48 and 72 hours.

Results:

We enrolled 30 consecutive patients, 7 males and 23 females, with a median age of 47 years.

Gastrointestinal symptoms included meteorism and abdominal pain (63%), dyspepsia (44%) and headache (38%), diarrhea (38%) and constipation (13%).

Regarding other comorbidities, allergies and autoimmune tiroiditis were reported in 20% of the patients.

Cutaneous involvement was characterized by erythematous scaling, eczematous patches in 9 out of 30 (30%) and psoriasiform lesions in 6 patients (20%). Trunk, upper limbs, and face were the most affected areas. The remaining patients suffered from pruritus without skin lesions (7/30; 23%), unspecific erythematous papular lesions (4/30; 13%), and rosacea (2/30; 7%).

At DIF, the most reported finding was the granular deposition of C3 at the dermal papillae (11/16; 69%).

Fluorescent bodies IgM and of IgA were reported, respectively, in 31% and 25% of cases.

We performed patch tests on 16 patients, but only 1 patient showed a mild positivity at 48 hours, completely disappeared at 72 hours.

Conclusion:

Patients with a cutaneous involvement of NCGS generally presented with unspecific itchy skin eruptions, mostly with eczematous or psoriasiform appearance. The most frequent finding at DIF is the granular deposition of C3 at the BMZ, suggesting it as a potential marker for cutaneous NCGS.

Patch tests seem to be not useful for NGCS diagnosis.

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Abstract N°: 4135

A case of blastic plasmocytoid dendritic cell neoplasm presenting as bruising to the dermatology department

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Introduction & Objectives:

Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN) is an exceptionally rare, aggressive form of haematological malignancy derived from plasmacytoid dendritic cells(PDC's). (1) Its incidence is 0.4 cases per 100,000 individuals. (2)PDC's are a bone marrow derived cell line that are rare or absent in healthy skin. (1) They are characterised by the production of interferon alpha(3). Although PDC's are typically absent in healthy skin, cutaneous manifestations are common presenting signs of BPDCN. Given its aggressive natural history, it is important that dermatologists are acquainted with the polymorphic nature in which it presents in order to facilitate prompt diagnosis. Herein, we present the case of disseminated bruising which culminated in a diagnosis of BPCDN .

Materials & Methods:

A 68 year old female was referred from the emergency department with a three month history of progressive bruising and fatigue. She had no history of bleeding or B symptoms. Full skin examination revealed widespread ecchymoses involving her trunk, limbs and face.

Results:

A skin biopsy demonstrated a dense inflammatory infiltrate composed of convoluted medium sized cells. Corresponding bone marrow aspirate and immunohistochemistry staining confirmed blastic plasmacytoid dendritic cell neoplasm. Although our patient did well initially with aggressive chemotherapy, she ultimately died as a result of her disease

Conclusion:

Although exceeding rare, BPCDN is a disease which can present with cutaneous findings initially. As such, we feel this case is important as it highlights the need for dermatologists to familiarise themselves with this diagnosis and maintain a low threshold to biopsy non-specific bruising.

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Abstract N°: 4142

Monoclonal Gammopathy- Associated Scleredema, a case report.

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Introduction & Objectives:

Scleredema is a rare connective tissue disease characterized by the excessive mucin and collagen deposition in dermis. It mainly affects the upper body, giving as a result a subtle symmetrical, woody-like induration of the skin. Its etiopathogenesis is poorly understood. Very few cases of monoclonal gammopathy- associated scleredema are available in literature so far.

Materials & Methods:

A 61-year-old male under 3 years follow-up for IgG kappa monoclonal gammopathy presented with progressive thickening of the posterior neck and back skin. It was an erythematous, symmetrical, non -pitting cutaneous induration with no movement limitation. He was overall asymptomatic. Serum protein electrophoresis a showed an IgG-k clonal peak (1,77g/dL) and urine protein electrophoresis was anodine. IgG was 2011 mg/ dL. A skin biopsy performed on his neck. It showed mucin deposition in reticular dermis and thickened collagen bundles separated by clear spaces, resulting in fenestration. Epidermis was normal and the skin appendages were unaffected. The patient’s diagnosis was scleredema, associated to its hematological condition. Since he was asymptomatic, and had optimal hematologic control, therapeutic abstention was decided. Up to this date he continues his regular follow up with no evidence of disease progression, scleredema persists with no other cutaneous symptoms.

Results:

It’s been noted its relationship with diabetes mellitus (Type 3 scleredema), streptococcal respiratory tract infections (type 1 scleredema) and monoclonal gammopathy including multiple myeloma (former type 2).

The association of scleredema with monoclonal gammopathies is well known, yet rare. To this day only around 40 cases have been reported. IgG paraprotein is isolated in most of the cases (around 80%), mainly IgG kappa-type as it happens to be in our case. This scleredema subtype normally shows a slowly progressive, non-resolving course.

Up to this day, there is no consensus about which treatment should be performed. Apparently, there is no relationship between the control of the underlying disease and scleredema remission. Among the therapies available, PUVA and UVA 1 phototherapy has shown the most positive results. Systemic corticosteroids and immunosuppressive drugs are also used in unresponsive scleredema and type 2 scleredema. Some paraprotein-associated scleredema have been successfully treated with chemotherapy. Therapeutic abstention is also a habitual practice, this “wait and see” attitude has shown no increase in mortality.

Conclusion:

Scleredema is a rare, chronic disorder that may present in association with monoclonal gammopathy. Dermatologists should bear this in mind when diagnosing this rare skin condition to perform further hematological studies. Giving the non-life-threatening course, the “wait and see” approach is a valid option for asymptomatic patients.

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Abstract N°: 4296

Primary cutaneous diffuse large B-cell lymphoma in adolescent: a case report

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Introduction & Objectives:

Primary cutaneous B-cell lymphomas account for 20–30% of primary cutaneous lymphomas, of which primary cutaneous diffuse large B-cell lymphoma (PCDLBCL) has a poorer prognosis than other types. Herein, we describe a rare case of PCDLBCL in an adolescent.

Materials and Methods:

A case report

Results:

An 18-year-old man presented to our clinic with a 3 months history of an expanding indurated sclerotic violaceous-erythematous plaque without epidermal changes, previously treated in a small hospital. His fingers were cold and purple. He occasionally experienced nausea after meals and bloating at night. Low doses of oral corticosteroids and methotrexate were administered for 3 months following the diagnosis of morphea. A new swollen mass appeared on his neck and his shoulders were strongly bent inward because of the progressive chest lesions. Ultrasound showed an increase in the size of chest muscles. The dose of methotrexate was increased to 15 mg/week, and the oral corticosteroids dose was increased to 0.7 mg/kg/day. A 3 × 4 cm nodule was found in the plaques, which became ulcerated afterwards, and the patient felt exhausted and experienced extreme shoulder pain. In the 7th month, we discontinued the methotrexate, and the oral corticosteroids dose was decreased while awaiting the results of the biopsy performed on the new nodule. Biopsy and immunohistochemical staining (Figure 1&2) revealed PCDLBCL. While waiting for treatment, he had bilateral lower extremity weakness with 2/5 strength, bowel and urinary incontinence, and no peripheral lymph nodes were detected.

Routine blood biochemical tests included lactate dehydrogenase 626 U/L (increase), beta-2 microglobulin 3,480 ug/L (increase), and uric acid 7.65 mg/dL (increase). Computed tomography (CT) of the brain and maxillofacial region with contrast injection revealed no abnormalities. However, chest CT with contrast injection revealed diffuse alveolar lesions in both lungs (suggestive of pneumonia) and bilateral lower lobe collapse with minimal pleural effusion. Abdominopelvic CT with contrast injection showed a low-density lesion on the right side of the liver. Magnetic resonance imaging of the spine with contrast injection revealed scattered spinal cord lesions in the cervical and thoracic spine. Compression fractures of the D5 vertebral body caused compression of the thoracic cord D5; no abnormalities were seen in other regions.

Finally, the patient was diagnosed with non-Hodgkin’s lymphoma and large B-cell lymphoma stage IVB, with two complications: spinal cord compression and tumor lysis syndrome. Chemotherapy was initiated with the R-CHOP regimen (rituximab 375 mg, cyclophosphamide 750 mg, doxorubicin 50 mg, vincristine 1.4 mg, and methylprednisolone 60 mg), then switched to R-ICE regimen (rituximab, ifosfamide, carboplatin, etoposide phosphate) which showed good results.

Conclusion:

PCDLBCL is a rare type of non-Hodgkin's lymphoma involving the skin, especially in young people; thus, it is highly likely to be diagnosed late or misdiagnosed. This case emphasizes the importance of considering neoplastic diseases in younger patients with sclerotic features mimicking other neoplastic and inflammatory conditions.

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Abstract N°: 4369

Sweet syndrome induced by insect bite.

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Introduction & Objectives:

Sweet syndrome (SS) is a rare neutrophilic dermatosis. It is characterized by a constellation of clinical symptoms, biological and histological abnormalities. In recent years, the understanding of its clinical spectrum has broadened but its etiology remains uncertain. Some authors suggest a possible pathergy phenomenon.

Materials & Methods:

We report a new observation of a SS induced by an insect bite.

Results:

A 64-year-old man presented to the clinic with painful and pruritic lesions on the exposed areas (hands, forearms, and neck), which had been evolving for 3 days. The lesions were preceded by insect bites in the same location. The medical history revealed no associated illness, recent medication use, infection, or vaccination. On clinical examination, multiple well-defined, infiltrated, erythematous-violaceous papules and plaques of varying size were noted. Some lesions were centered by a small crust and vesicle or pustule. The patient was apyretic, with a preserved general condition. The biological tests revealed an elevated C reactive protein level (54mg/l) and an accelerated erythrocyte sedimentation rate (1st hour= 76 mm; 2nd hour= 96 mm). The blood count was normal with no leukocytosis or hyper eosinophilia. A skin biopsy was performed, showing an edematous dermis with a dense peri vascular infiltrate containing neutrophils and numerous lymphocytes. Lesions of leucocytoclasis without vasculitis were described. The diagnosis of Sweet's syndrome was confirmed. The patient was treated by topical corticosteroids and non-steroidal anti-inflammatory drugs with complete healing.

Conclusion:

Sudden onset of painful erythematous papular lesions associated with fever should always suggest Sweet's syndrome, even if there is no evidence of an insect bite. If the diagnostic criteria are not fulfilled, Sweet-like reactions due to arthropod bites should be considered.

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Abstract N°: 4398

Acral retiform purpura and heel ulceration as a cutaneous sign of spontaneous cholesterol embolism

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Introduction:

Cholesterol embolism typically occurs in elderly patients with multiple risk factors for atherosclerosis and often after endovascular procedures, chronic anticoagulation, or fibrinolytic therapies. However, it can also occur spontaneously without a clear precipitating factor in about 20% of cases.

Case report:

A 66-year-old female patient with a previous history of hypertension, long-standing dyslipidaemia and a recently diagnosed endometrial carcinoma, consulted for a painful lesion on the left heel of 1 month´s duration, preceded by intermittent claudication since a few months earlier. The patient had not undergone any endovascular procedure and had not started oral anticoagulation recently. Physical examination revealed a circular ulcer on the left heel surrounded by a retiform purpura, as well as cyanosis on the first toe. Autoimmunity screening, including antinuclear antibodies, antineutrophilic cytoplasmic antibodies, antiphospholipid antibodies and cryoglobulins were negative. Complete blood count did not show peripheral eosinophilia. Histopathological examination of a deep skin biopsy showed a non-inflammatory occlusive vasculopathy, as well as the presence of a small biconvex acicular structure occluding the lumen of a small calibre vessel in the serial study. CT angiography revealed thick atheromatous plaques involving the infrarenal aorta, common and external iliac arteries, and the superficial femoral artery. The patient started treatment with acetylsalicylic acid and cilostazol, with a progressive resolution of the ulceration and some improvement in claudication.

Discussion:

From a dermatological point of view, cholesterol embolization is clinically characterized by the presence or sudden onset of livedo racemosa, retiform purpura, acral cyanosis, gangrene, or ulceration, and cutaneous manifestations are present in 35 to 88% of cases. In addition, systemic manifestations such as renal, gastrointestinal, ocular, or even neurological involvement may also occur, depending on the location of the affected arteries. Constitutional symptoms such as fever or myalgias may also be present and analytic abnormalities such as an elevated erythrocyte sedimentation rate or peripheral eosinophilia (in up to 80% of patients) are frequent. From a histopathological perspective, it should be noted that skin biopsy should extend into the subcutaneous fat tissue and that most often serial sections are necessary to identify pathognomonic features. The mortality rate can be as high as 63-81% in some cases. Treatment is not clearly standardised, depends on which organ is affected, and is mostly supportive in an acute setting. While anticoagulant therapy should be stopped if there is the precipitating factor, treatment with corticosteroids, iloprost, hyperbaric oxygen or pentoxifyline, among others, should be cautiously considered. Moreover, antiplatelet agents and HMG-CoA reductase inhibitors are often prescribed to stabilise atherosclerotic plaques.

Conclusion:

Cholesterol embolization syndrome may appear as heel ulceration associated with retiform purpura and may occur

spontaneously. Histopathological examination must be careful and serial sections must be made to locate the cholesterol crystals occluding the blood vessels.

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Abstract N°: 4423

Reactive perforating collagenosis secondary to primary biliary cirrhosis

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Introduction & Objectives:

We present the case of a 68 year-old woman who developed a widespread intensely pruritic nodular rash on her shoulders, abdomen, back, knees and buttocks. She has a past medical history of primary biliary cirrhosis and osteoporosis. On examination, there were extensive crusted violaceous nodules felt not to be typical of a nodular prurigo eruption. Differential diagnoses included verrucous lichen planus, a granulomatous eruption secondary to her biliary cirrhosis, or perforating collagenosis. Blood investigations were unremarkable with the exception of an elevated alkaline phosphatase which was chronic in nature secondary to her PBC.

Materials & Methods:

Initial biopsy was inconclusive indicating scarring and inflammation with focal parakeratosis likely secondary to excoriation. Further punch biopsy demonstrated extensive inflammatory infiltrate and degenerate collagen, in keeping with perforating collagenosis. Treatment with Mometasone was commenced in addition to UVB Therapy.

Results:

Pruritus subsequently improved however the skin lesions remained unchanged. Following 21 sessions of UVB phototherapy, her disease became quiescent with only post-inflammatory hyperpigmentation and scarring.

Conclusion:

This case is unique as there are only a few case reports of perforating collagenosis secondary to primary biliary cirrhosis reported. Our patient had previously been known to Gastroenterology but was lost to follow up. Following her presentation, she was re-referred and was reviewed in their clinic for continued management of her PBC. Acquired reactive perforating collagenosis is a rare skin disease commonly associated with diabetes and end stage renal disease, particularly in patients who are treated with haemodialysis. Histology reveals cup-shaped invaginations with a keratotic plug penetrating the papillary dermis with underlying dermal lymphocytic and histiocytic infiltrate. Differential diagnoses such as prurigo nodularis and arthropod bites should be excluded as both resemble perforating collagenosis.

If driven by an underlying cause, treatment of this can lead to an improvement in symptoms. Our patient was already prescribed ursodeoxycholic acid as treatment for primary biliary cirrhosis; however, as she continued to have active skin lesions, she required further treatments. This includes topical corticosteroids, intralesional corticosteroids, topical retinoids as well as antibiotics. In our patient's case, UVB phototherapy was effective and this approach has shown similar benefits in multiple case reports. Further treatment options include Isotretinoin and Allopurinol which have also proved beneficial in case reports.

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Abstract N°: 4477

Mycosis fungoides following follicular mucinosis and associated with Langerhans cell histiocytosis : Case report

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Introduction & Objectives:

Mycosis fungoides is the most common primary cutaneous epidermotropic lymphoma. The initial histological appearance is nonspecific, and the association with other hematopoietic proliferations can delay the diagnosis.

We report the case of a patient with mycosis fungoides presenting with an initial manifestation of follicular mucinosis and associated with Langerhans cell histiocytosis.

Materials & Methods:

A 76-year-old patient with a history of colonic cancer, which was surgically treated a year ago and complicated by a pulmonary metastasis treated with adjuvant chemotherapy (Capecitabine), presented with generalized pruritic plaques and nodules that had been progressing for 3 years. The patient exhibited well-defined, rounded erythematous plaques primarily located on the back of the thighs and arms. Some of the plaques showed infiltration and were covered with yellowish crusts. An initial biopsy of a plaque on the back indicated follicular mucinosis. Due to the extension of the lesions, multiple biopsies were performed after 6 months, revealing a typical appearance of mycosis fungoides (CD4+, CD8+). In a infiltrated yellowish lesion on the back, evidence of Langerhans cell histiocytosis was observed. The patient underwent 40 sessions of phototherapy, which initially led to improvement but was followed by a relapse. Treatment with methotrexate has been initiated.

Results:

In our case, mycosis fungoides initially presented as follicular mucinosis. The latter is not often a benign self-limiting proliferation, as it can have a tendency for clonal lymphocytic proliferation, leading to mycosis fungoides. The simultaneous occurrence of MF and Langerhans cell histiocytosis is a rare association. The hypothesis of a clonal process affecting a common hematopoietic precursor generating two different neoplastic contingents has been proposed by some authors. Other studies have suggested that histiocytic proliferation is reactive to increased cytokine stimulation by clonal lymphocytes.

Conclusion:

Being able to suspect mycosis fungoides based on a suggestive clinical context and nonspecific or misleading histological features can aid in early diagnosis. The association between mycosis fungoides and Langerhans cell histiocytosis is rare. The nature of the relationship between the two processes is yet to be established.

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Abstract N°: 4515

Facial pyoderma gangrenosum associated with carcinosarcoma

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Facial pyoderma gangrenosum associated with carcinosarcoma

Abstract

Pyoderma gangrenosum(PG) is a neutrophilic dermatosis that is frequently associated with systemic disease.The association between PG and solid organ tumors including gynecologic cancers have been reported. Herein , we report a 61-year-old woman with pyoderma gangrenosum on the posterior auricle associated with an underlying carcinosarcoma, a rare, aggressive uterine carcinoma. The patient was referred to our clinic after multiple unsuccessful works-ups with a painful , necrotic ulcer on the left posterior auricle.The skin biopsy was performed and the pathology results showed pseudoepitheliomatous epidermal hyperplasia, dermal necrosis, extensive abscess formation, mixed acute and chronic inflammation , inflammation of vascular wall which were more in favor of pyoderma gangrenosum.To rule out underlying malignancies an abdominopelvic ultrasonography and MRI were performed . A left adnexal cyst and a nodule were detected. She underwent laparotomy and the cyst and the nodule were excised . The pathology results showed high-grade epithelial neoplasm . IHC results was suggestive of Carcinosarcom.methylprednisolone pulse therapy (1g/day) was administered for three consecutive days, and then treatment continued with prednisolone 60mg, cyclophosphamide 100 mg once daily, and topical silver sulfadiazine ointment. After receiving treatment for 2 weeks, the ulcer’s progression stopped, and both its depth and width gradually shrank. She was referred to an oncologist for further cancer treatment. The oral corticosteroid dose was slowly tapered to 15mg over the course of 4 months while cyclophosphamide was continued at the same dosage. No new lesions were detected, and the ulcer had healed forming a cribriform scar.

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Abstract N°: 4526

Painful plantar nodules

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Introduction & Objectives:

Plantar subcutaneous nodules are infrequent complaint in dermatology. Faced with these lesions, the clinician's fear would be to miss malignant tumors. However, it is important to recognize benign entities that can be easily diagnosed.

Materials & Methods:

We report a case of bilateral plantar painful nodules.

Results:

A 32-year-old male patient, without medical history, consulted for a 2-year history of two subcutaneous nodules of the soles, reaching 2 and 4 cm in diameter and becoming painful after prolonged walking. On examination, they were firm and noninflamed. Ultrasound revealed fibrous and hypoechogenic nodules enclosed in the aponeurosis with discrete hyperemia in Doppler mode. The diagnosis retained was plantar fibromatosis. Appropriate shoes were advised.

Conclusion:

Plantar fibromatosis or Ledderhose disease is a benign aponeurotic fibroblastic proliferation. It is rare and can be associated with other fibromatoses such as palmar Dupuytren's disease and Peyronie's disease of the penis. Among the etiopathogenic factors, we note genetic predisposition, diabetes, alcoholism and prolonged immobilization. In our patient, being a sportsman, the cause would be repeated microtrauma.

The clinical presentation varies from a non-painful firm nodule of the sole to a fibrous lump with a muscle contracture. These lesions may be unilateral or bilateral. Clinical examination may be supplemented by ultrasound or MRI. Differential diagnoses may include leiomyomas, melanomas and synovial sarcomas. In case of doubt, a skin biopsy may be useful.

Conservative treatment options include orthopedic inserts, injection of corticosteroids, collagenase or Verapamil. Tamoxifen and radiation therapy may also be indicated. Surgical treatment is recommended in case of walking impairment or non-response to conservative treatment.

Plantar fibromatosis is a rare and benign proliferation with a typical clinical presentation. In case of uncertainty, a simple ultrasound scan makes the diagnosis. Management is not specified and depends on the impairment resulting from the disease.

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Abstract N°: 4535

A Rare Skin Metastasis of Pancreatic Cancer: Sister Mary Joseph's Nodule

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Introduction & Objectives:

Sister Mary Joseph Dempsey worked as a surgical assistant to William J. Mayo at St. Mary's Hospital in Minnesota between 1890 and 1915. In 1928, she described the Sister Mary Joseph nodule, a skin metastasis located in the umbilicus, which she added to the literature under her own name.

Sister Mary Joseph Nodule is often associated with metastases from gastrointestinal and pelvic malignancies. There are also cases associated with hematologic malignancies in the literature.

The Sister Mary Joseph Nodule is an indicator of malignancy and poor prognosis as it describes skin metastasis located in the umbilicus.

Case:

In our case, a 75-year-old woman presented with a 1-month history of enduring erythematous nodular lesion in the umbilical region. A punch biopsy was taken from the lesion. The biopsy result was adenocarcinoma metastasis. Then, the patient was investigated for malignancy and CT scan imaging revealed pancreatic adenocarcinoma and umbilical skin metastasis.

Results:

Pancreatic cancer is one of the leading causes of cancer-related deaths. It usually presents in the late stage or with jaundice. Rarely, skin metastases in the umbilical region may be the first manifestation of pancreatic cancer. Approximately 50-60% of patients with pancreatic cancer present with distant metastatic disease, 25-30% with regional disease and only 10-15% with local disease.

Skin metastases of adenocarcinomas are rare and Sister Mary Joseph's Nodule is observed as an erythematous, indurated, ulcerated, abdominal skin-fixed nodular lesion in the periumbilical region.

Conclusion:

Our duty as dermatologists should be to suspect internal malignancy metastasis when we see such a lesion and not to consider this lesion as a simple skin lesion, but to evaluate it in terms of sectional and histopathologic examinations for malignancy.

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Abstract N°: 4556

Occupational-related systemic sclerosis : Case report

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Introduction & Objectives:

Systemic scleroderma is a multifactorial autoimmune disease. Although its etiology is still not fully understood, it involves genetic and environmental factors. Among the environmental factors, occupation seems to play a significant role. We report a case of a patient who developed systemic scleroderma as a result of professional exposure to printing products.

Materials & Methods:

A 35-year-old man, with no significant medical history, had been exposed to solvents since 2017 due to his work in a printing company. In 2022, he presented with several dyschromic, indurated, and hyperesthetic plaques on the trunk and upper limbs. Upon questioning, he reported vascular disturbances consistent with Raynaud's syndrome. Examination revealed dyschromic plaques associated with cicatricial alopecia in the temporal areas, limited mouth opening, limited finger mobility with dyschromia. Antinuclear antibodies (ANA) were positive at 1/1280 with a speckled pattern, and anti-Scl 70 antibodies were positive. Skin and scalp biopsies showed histological features compatible with scleroderma. Further investigations revealed esophageal and pulmonary involvement. We concluded a probable occupational origin of systemic scleroderma.

Results:

Several studies have suggested the occupational induction of systemic scleroderma. Among the studied occupational agents, silica and organic solvents have been proven to induce systemic scleroderma. Patients exposed to these agents showed higher rates of diffuse forms of systemic scleroderma with interstitial lung disease and a more frequent association with positive anti-Scl 70 antibodies. A recent study demonstrated that vascular endothelial cell dysfunction can be caused by the internalization of exogenous toxic products, leading to an immune and fibrotic reaction.

Conclusion:

Collaboration between the attending physician and the occupational physician may be justified, especially when investigating the patient's workplace to identify potential exposures associated with the disease for possible legal recognition.

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Abstract N°: 4561

When blood runs cold : disseminated subcutaneous nodules revealing a cold agglutinin syndrome with fatal outcome.

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Introduction:

Cold agglutinin disease is a rare type of autoimmune hemolytic anemia, related to the presence of antibodies that bind to erythrocyte surface antigens at low temperatures called "cold agglutinins". The cutaneous manifestations of this disease are usually triggered or aggravated by cold and may include: acrocyanosis and Raynaud's phenomenon, livedo reticularis, cold urticaria or even digital necrosis. In this case, we describe disseminated subcutaneous nodules that later revealed a cold agglutinin disease in a 30-year-old female patient.

To our knowledge, no similar case has been reported in the literature.

Case report:

A 30-year-old woman, without any documented history of underlying disease was hospitalized for disseminated subcutaneous nodules. The history began 1 month earlier with an infectious episode of diarrhea, vomiting, and cough. Examination on admission revealed several subcutaneous nodules of purplish color on the trunk, abdomen, upper and lower limbs, infiltrated, embedded in the dermis and very sensitive to palpation, the patient was febrile, asthenic with mucocutaneous pallor and splenomegaly without palpable adenopathy.

Biological examinations revealed: an anemia and elevated values of reticulocytes and LDH MCHC, collapsed haptoglobin and hyperbilirubinemia. The coombs test was positive and cold antibodies were detected at a very high rate, the blood smear showed presence of numerous platelet aggregates and anisopoikilocytosis., skin biopsy showed a mixed infiltrate of cells and the presence of congestive vessels with turgid endothelium.

The diagnosis of acute autoimmune hemolytic anemia with cold antibodies was retained, during her hospitalization, the patient presented a frank and brutal anemic syndrome associating dyspnea, tachycardia,"port wine red" urine color and a deglobulization crisis up to 1g/dl justifying her admission in the intensive care unit where a transfusion of phenotyped not warmed blood was initiated but the patient did not survive.

Discussion:

Classically, a distinction can be made between transient acute cold antibody AHAI and chronic cold agglutinin disease. Indeed, the first subtype which is known as cold agglutinin syndrome is most often observed in children and young adults and occurs after a primary infection with EBV, CMV, HCV, HBV, parvovirus B19,VZV or mycoplasma pneumonia and auto-immune diseases or solid malignancy, while the second subtype called CAD is the prerogative of subjects over 50 years, evolves on a chronic background and includes cases with low grade lymphoproliferative disorder.

The clinical presentation of AHAI with cold autoantibodies is very variable, ranging from acute life-threatening intravascular hemolysis to cutaneous manifestations secondary to peripheral hemagglutination such as acrocyanosis, Raynaud's phenomenon, livedo reticularis, cold urticaria and digital necrosis.

In the case of our patient, it was the acute form of cold agglutinin disease, indeed there was the appearance of multiple subcutaneous nodules very sensitive to palpation after an infectious syndrome made of cough, vomiting and diarrhea, then complicated during her hospitalization by a fatal intravascular hemolysis.

Conclusion:

The clinical presentation of our patient has never been associated with autoimmune hemolytic anemia with cold antibodies, its recognition is mandatory because it can put at risk the vital prognosis which makes this case report very original.

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Abstract N°: 4596

Insect bite-like reaction of chronic lymphocytic leukaemia: a lesser-known paraneoplastic inflammatory phenomenon.

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Introduction & Objectives:

Chronic lymphocytic leukaemia (CLL) is the most common hematologic neoplasia in the Western hemisphere, with an incidence rate in Europe of 4.01 cases per 100000 population per year. It mainly affects males with a median age of 70 years. Cutaneous involvement is not an uncommon phenomenon in this disease: up to 8% of patients will present specific lesions with leukemic infiltration; these tumoral cells are usually secondarily found within the infiltrates of other inflammatory processes. Moreover, non-specific inflammatory reactions can take place in up to 45% of patients. They are thought to be the result of the immune dysregulation of lymphoproliferative disease. Insect bite-like reactions are a common clinical picture in this context.

Materials & Methods:

Results:

A 70-year-old man presented with flares of pruritic papules over his back for the previous 6 months. Each individual bout lasted for a week. His medical history was remarkable for stage C CLL, diagnosed 8 years before. He was receiving ibrutinib, but the lesions were already present when the drug was started.

Clinical examination revealed an indurated lumbar plaque and isolated left axillary infiltrated papules.

A biopsy was taken to rule out leukemic infiltration. It showed mild spongiosis, subepidermal oedema and a lymphocytic infiltrate entirely made up of T cells, with occasional eosinophils. A neoplastic B cell-component could not be identified.

Two months later, he consulted a new flare. Multiple infiltrated erythematous papules were noted over his back and flanks. A new biopsy revealed findings similar to the previous one.

Clinicopathological correlation allowed for the diagnosis of CLL-associated insect bite-like reaction (IBLR). Topical mometasone was given for symptomatic relief.

Conclusion:

IBLR has been described associated with CLL, as well as other haematological entities. At first interpreted as exacerbated reactions to actual arthropod assaults, their trigger remains currently unknown in many cases.

The mean time from the diagnosis of CLL to the onset of IBLR is 3.1 years, but skin lesions are known to precede the leukaemia diagnosis in up to 25% of cases. The course of the flares is independent from that of the hematologic disease. The lower limbs, followed by the upper limbs and the back, are more frequently involved.

IBLR is believed to be closely related or even synonym to eosinophilic dermatosis of hematologic malignancy.

Both entities have overlapping clinical manifestations, but the latter term is applied to those cases in which the eosinophilic component is more conspicuous. Non-neoplastic T-cell lymphohistiocytic infiltrates are found in every case of both entities, but eosinophils, previously understood as a histological hallmark of the spectrum, can be lacking in up to 25% of cases.

Treatment usually relies on topical corticosteroids or antihistaminic agents, but other therapies such as phototherapy, dapsone or dupilumab have been used in refractory cases.

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Abstract N°: 4627

The devil is in the details – a case of leukemia cutis in chronic myelomonocytic leukemia diagnosed in a patient hospitalized for cellulitis

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Title: The devil is in the details – a case of leukemia cutis in chronic myelomonocytic leukemia diagnosed in a patient hospitalized for cellulitis

Introduction & Objectives:

Leukemia cutis is a rare specific skin involvement by neoplastic leukocytes. Although any subtype of leukemia can involve the skin, most cases occur in patients with acute myelogenous leukemia. Due to its low incidence, chronic myelomonocytic leukemia (CMML) has been rarely associated with skin involvement.

Materials & Methods:

We describe a case of a 71-year-old male patient, admitted for treatment of a cellulitis of the left arm with intravenous antibiotics, whose complete observation revealed erythematous nodules on the limbs and trunk, and whose etiological study was compatible with a leukemia cutis in the context of a CMML.

Results:

A 71-year-old man was admitted to the hospital with a 1-week history of exuberant inflammatory signs of the medial aspect of the left arm, suggestive of cellulitis, for intravenous treatment with antibiotics. Concomitantly with a pruritic dermatosis, with 1-year history, characterized by well-defined erythematous-descaling plaques and nodules, firm to palpation, on the thighs, back, and upper limbs. He was treated with topical corticosteroids and antihistamines, with no improvement.

The patient was being followed in Haematology consultation for chronic immune thrombocytopenia, currently medicated with eltrombopag. On evaluation, the patient showed normochromic normocytic anemia, persistent monocytosis and myelemia, with a normal white blood cell count after treatment of infectious event. Histology revealed a perivascular and perianexial dense nodular lymphocytic infiltrate in the superficial and deep dermis with a subpopulation of cells in the dermis with large, irregular nuclei and a small cytoplasmic rim, with some atypical mitosis and apoptotic bodies, expressing CD68-PGM1, CD4 and CD43, and negative for myeloperoxidase and CD34, suggestive of leukemia cutis. A myelogram was performed and a diagnosis of CMML was reached. Given the diagnosis of CMML with skin infiltration the patient started treatment with Haematology.

Conclusion:

CMML is a rare clonal disorder of hematopoietic cells with both myeloproliferative and myelodysplastic features. CMML occurs mostly in males between 65-75 years old, and is characterized by monocytosis, cytopenia and blasts. Patients with CMML may present with a variety of cutaneous lesions, being leukemia cutis the involvement of the skin by leukemia cells, although extremely rare. The leukemic infiltrate is a factor of worse prognosis, being imperative the treatment of the underlying disease and surveillance for disease progression. We describe this case given the small number of reported cases regarding leukemia cutis in CMML, and the importance of thorough assessment of patients as a whole in every situation, for the diagnosis and improved prognosis of severe systemic pathologies affecting the skin.

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Abstract N°: 4656

All that blisters is not old: Skin fragility and atrophic scarring in a 47-year-old female

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Introduction & Objectives:

Materials & Methods:

Results:

A 47-year-old female presented with a 2-week history of spontaneous non-pruritic fragile bullae on the upper limbs. There was no mucosal involvement, recorded trauma, recent illness or insect bites. A past medical history of bilateral Perthes disease and bilateral hip replacements in her thirties aroused suspicion. Family history included multiple orthopaedic diagnoses; one sibling had Perthes disease of the hip, a second had childhood Osgood-Schlatter disease, and a third had recurrent knee dislocations. All three were male.

On examination, there was an erythematous patch on the right upper arm at the site of prior blistering, and an ulcer with surrounding erythema on the right wrist. Reticular livedoid change was seen on the lower limbs. Upon prompting, skin hyperextensibility and small joint hypermobility was demonstrated. The right wrist ulceration later healed with formation of a pale atrophic papyraceous scar.

Baseline laboratory investigations including were normal. Antiphospholipid antibodies were negative. Lesional skin biopsy from the wrist demonstrated predominantly post inflammatory findings; the epidermis was acanthotic with a prominent granular cell layer, with underlying dermal fibrosis and a mild superficial perivascular inflammatory infiltrate of small lymphocytes. Direct immunofluorescence of perilesional skin was negative for IgG, IgM, IgA, C3 and fibrin.

There was high clinical suspicion of Ehlers-Danlos syndrome (EDS) in the context of clinical findings and supportive family history. Genetic testing for known mutations responsible for EDS and other connective tissue disorders was negative, and a diagnosis of Hypermobile EDS was favoured.

Hypermobile EDS (hEDS) is the only one of the thirteen recognised subcategories of EDS without a known genetic mutation. hEDS is more common in females, with cutaneous manifestations including soft skin, atrophic or papyraceous scars, piezogenic papules, hyperextensive skin, unexplained striae, and easy bruising. Cutaneous manifestations tend to be more mild in hEDS than other variants of EDS - particularly in comparison to classical EDS, in which most cases have a genetic mutation in COL5A1 or COL5A2. Skin hyperextensibility is less common in hEDS, affecting approximately 50% of those with the diagnosis, in comparison to classical EDS in which it is seen in over 90% of patients. In this case, presentation of blistering may have been related to unrecalled mechanical trauma or topical heat application, which resolved spontaneously and did not recur. The dominant features of atrophic scarring and skin hyperextensibility were notable and in keeping with cutaneous manifestations of hEDS.

Conclusion:

This case illustrates the variety of cutaneous manifestations of hEDS in a young female, with a prominent early orthopaedic history. Skin findings seen in hEDS can be subtle, and the diagnosis should be suspected where other supportive findings in the history and examination are evident. We wish to further highlight to dermatologists the clinical value in taking a targeted personal and family history, in particular in relation to joint problems and

hypermobility, in patients presenting to dermatology services with these findings.

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Abstract N°: 4663

Blastic plasmocytoid dendritic cell neoplasm: experience with a series of cases in a single center in Sao Paulo, Brazil

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Introduction & Objectives:

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare hematologic malignancy derived from plasmacytoid dendritic cells with an aggressive evolution and poor outcome. Skin lesions are the most common manifestation of BPDCN, and prompt recognition of this entity by dermatologists will allow early diagnosis to institute the appropriate treatment for this aggressive disease. Cutaneous manifestation is very typical, presenting as violaceous, bruise-like papules, plaques, and nodules, usually asymptomatic, with or without necrosis, that can be focal or diffuse. Extracutaneous sites of disease include bone marrow, central nervous system, breasts, gallbladder, and paranasal sinuses. BPDCN is a rare condition with few cases published. Therefore, we aim to describe a series of cases with different types of skin lesions and systemic involvement and in different age groups.

Materials & Methods:

A retrospective study was performed by reviewing medical records of patients diagnosed with BPDCN between the years 1990 and 2022 at a reference center in Brazil.

Results: ** Seventeen patients were included in this study with a male predominance of 70.6%. Median age of diagnosis was 59 years old, ranging from 14 to 81 years old. All patients presented violaceous plaques and nodules, 12.5% had focal disease and 87.5% had diffuse skin involvement. Skin biopsies showed diffuse dermal infiltration by CD4+ and CD56+ cells compatible with dendritic plasmacytoid cell phenotype.

Seven of the patients had radiologic investigation for extracutaneous disease at diagnosis. Two (28.5%) of them had extracutaneous disease, with PET-CT evidencing glicolitic metabolism at lymph nodes above and below the diaphragm, bone marrow and breasts in one and at paranasal sinuses and spleen in the other. The other five (71,5%) of them had BPDCN lesions restricted to skin.

At the last evaluation, 4 patients (23.5%) were alive in remission, 7 patients (41.2%) died due to progression of the disease, 1 patient (5.8%) died due to complications of the treatment and 5 patients (29.4%) lost follow-up. Of those patients who are still alive, 75,0% (n=3) were submitted to an allogeneic bone marrow transplantation and chemotherapy and 25% (n=1) does not have treatment information on medical records. In our cohort, median overall survival was 16 months.

Conclusion:

Although very rare, BPDCN is an aggressive medical entity that requires rapid diagnosis and aggressive treatment. As it may first present with cutaneous manifestations, it is important for dermatologists to know its clinical presentation and indicate biopsy to fasten diagnosis and offer more chances of survival for the patients. We presented a series of 17 cases of a tertiary reference center in São Paulo, Brazil.

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Abstract N°: 4665

Calciphylaxis: clinical characteristics and prognostic factors in a cohort of a tertiary hospital

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Introduction & Objectives:

Calciphylaxis is a rare but devastating disorder characterized by calcification and thrombosis of dermal and subcutaneous vessels, with subsequent ischemic necrosis of skin tissue. As a consequence, very painful necrotic ulcers appear, accompanied by significant morbidity and mortality. The aim of this study is to describe the clinical characteristics of patients with calciphylaxis and to identify prognostic factors.

Materials & Methods:

A single-center retrospective study was performed by reviewing the medical records of all patients with a diagnosis of calciphylaxis between November 2008 and April 2023. Demographic, clinical, histological and laboratory data were collected.

Results:

A total of 28 patients were included, 15 females and 13 males, with a mean age at diagnosis of 68 years. Eighteen patients had nephrogenic calciphylaxis and the other 10 had non-nephrogenic calciphylaxis.

The most frequent location of the ulcers was the legs (65%) and one third of the patients had lesions of proximal distribution. Pain was a constant feature and two thirds of the patients had retiform purpura around the ulcer.

Risk factors for developing calciphylaxis included chronic kidney disease and renal replacement therapy, cardiovascular risk factors, use of corticosteroid or vitamin K antagonists, calcium-phosphate metabolism abnormalities, hypoalbuminemia and female sex. No differences were observed in the distribution of risk factors between nephrogenic and non-nephrogenic calciphylaxis, except for calcium-phosphate metabolism abnormalities, which were more frequent in patients with nephrogenic calciphylaxis.

In most patients a skin biopsy was performed and findings compatible with calciphylaxis were found in 89%.

Twelve patients achieved complete healing of the ulcers in a median of 20 months. Treatment was based on pain management, minimizing modifiable risk factors, use of inhibitors of ectopic calcifications (such as intravenous sodium thiosulfate and bisphosphonates) and wound care.

Twenty of the 28 patients died during follow-up. Patients with distal ulcers had a median survival of 80 months, while in those with proximal lesions it was only 2 months ($p < 0.001$, log-rank test). In the multivariable Cox model, proximal location of the lesions (hazard ratio, 9.58; 95% confidence interval, 1.76-52.18; $p = 0.009$), nephrogenic calciphylaxis (HR, 17.67; 95% CI, 1.71-182.21; $p = 0.016$), diabetes mellitus (HR, 11.49; 95% CI, 1.57-84.12; $p = 0.016$) and hypoalbuminemia (HR, 0.88; 95% CI, 0.79-0.98; $p = 0.021$) were identified as independent predictors of mortality.

Conclusion:

Calciphylaxis should be suspected in a patient with risk factors and painful ulcers with associated retiform purpura,

especially in the legs. It is important to initiate treatment early, as mortality rates per year range from 40 to 80%, with a higher risk of death in patients with nephrogenic calciphylaxis, diabetes, hypoalbuminemia and proximal location of the lesions.

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Abstract N°: 4668

Beneath the Surface: Sarcoidosis in Burn Scars

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Introduction & Objectives: Sarcoidosis is a systemic granulomatous disease of unknown etiology, involving skin in up to one-third of cases. Lesions are diverse, often presenting as symmetrical facial maculopapules or extensive infiltrated nodules and plaques. Up to 9% of cases may arise in scars, resulting from surgery, trauma, acne, venipuncture, or tattoos. We report an unusual case revealing its origin in old burn scars after a lengthy evolution.

Materials & Methods (case report): An 85-year-old woman with extensive hot water burns over 15 years ago, presented with acute desquamative plaques and neuropathic pain on the neck and thigh. Initial considerations included eczematous and papulosquamous conditions, or an isotopic response to possible past herpes zoster. Topical and systemic corticosteroids plus analgesia showed poor response. Pandemic disrupted follow-up, and after 2 years, the patient reported continuous use of potent topical steroids due to recurrent skin flare-ups. Examination now revealed similar extent plus considerable skin atrophy and a yellowish hue. Clear involvement of old burn scars previously hidden by inflammation was evident on the neck. Conversely, scars on the affected thigh were imperceptible, despite the patient reporting extensive burns in the area. Clinical findings, dermoscopy, and two biopsies ultimately led to a diagnosis of scar sarcoidosis. To date, no involvement in other systems has been found except for possible small-fiber neuropathy. She is currently being treated with topical tacrolimus, hydroxychloroquine, and multimodal analgesia.

Results (discussion): Infiltration of scars by non-caseating granulomas is a known specific manifestation of sarcoidosis. However, the frequency and clinical characteristics are limited by few reported cases. In existing series, burn scars are underrepresented compared to other types of scars. Lesions may be confined to or extend beyond the scars, as in our case. Greater extension correlates with increased chronicity, severity, and systemic involvement, including pulmonary involvement in up to 40-100% of cases. Our case underscores an unusual presentation with obscured initial scarring due to extensive inflammation, resembling a zosteriform pattern. Moreover, it involves burned areas with no evident residual scarring.

Conclusion: Sarcoidosis mimics various conditions, even when localized to one system. Unusual manifestations emphasize the importance of a thorough medical history. Further studies may reveal the prognosis of distinct disease phenotypes.

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Abstract N°: 4674

Elephantiasis Nostras Verrucosa: A Complication of Portal Hypertension and Obesity

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Introduction & Objectives:

Elephantiasis nostras verrucosa is the progressive disfiguring enlargement of a body part and is characterized by dermal fibrosis with hyperkeratotic, verrucous, and papillomatous lesions that usually occur after chronic lymphoedema. Lymphedema is the increase in the volume of an anatomical region. The infestation was named elephantiasis by Wucheria Bancroft and in 1934 Castellani added nostra to add other causes. It is secondary to superficial lymphatic obstruction, extravasation of lymph, and activation of fibroblasts. It is caused by chronic lymphedema that could be congenital or produced by a non-associated infection (such as tuberculosis, mycotic infection, syphilis), surgery, radiotherapy, trauma, neoplastic obstruction, obesity, portal hypertension, or congestive heart failure. There is no standard treatment for this rare skin disorder.

Materials & Methods:

We describe the case of a 47-year-old man with Portal Hypertension, Obesity, and elephantiasis nostras verrucosa skin lesions on both legs, a history of smoking for 15 years consuming 10 cigarettes a day, and alcoholism since age 17 consuming 120 ml of fermented, with liver disease since 2012, and poor adherence to treatment and a history of grade IV venous insufficiency diagnosed one month before evaluation, at the time of his evaluation he complained of 2 years of worsening edema and verrucous plaques.

Physical examination showed non-pitting edema and impressive mossy plaques and cobblestone-like nodules in both legs and feet, especially on the right, with a positive Stemmer's sign indicative of lymphoedema and a body mass index (31.6kg/m2).

Laboratory evaluation revealed normal results for the complete blood count, renal and liver function tests, from a skin punch biopsy hyperkeratosis of the epidermis, loss of dermal papillae, fibrosis of the dermis and subcutaneous tissues, and widened lymphatic vessels.

Results:

For treatment compression with elastic bandages or stockings has been described to be beneficial. Identification and correction of the cause of the lymphoedema, such as weight reduction, may also help to achieve a better and faster improvement. Topical keratolytics or humectants, such as salicylic acid and urea, may improve hyperkeratosis, as well as topical or oral retinoids. For refractory cases, carbon dioxide laser or surgical intervention may be attempted.

Conclusion:

As the prevalence of hepatic disease and obesity rises, the complications of these diseases will also become more common. This disease can be diagnosed clinically with history and physical examination alone, although other tools are available to assist the clinician.

It is important to rule out susceptibility to cancer (squamous cell carcinoma, lymphoma, melanoma, Kaposi's

sarcoma, Stewart-Treves syndrome) and infections (chronic osteomyelitis and arthritis).

Unfortunately, a disease without effective treatment, and its prognosis is poor. For the dermatologist and internal medicine specialist, this case illustrates the importance of early intervention for patients with obesity and other comorbidities to prevent sequelae such as Elephantiasis nostras verrucosa.

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Abstract N°: 4714

Blue toe syndrome about an idiopathic case

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Introduction & Objectives:

Cholesterol crystal embolism disease or Blue Toe syndrome is a rare and little-known complication of atherosclerosis, the symptomatic forms of which are most often of iatrogenic origin, manifesting as mono-organic or more frequently systemic involvement. The objective of this work is to report our experience through a patient with this disease.

Materials & Methods:

G.L, 74 years old, hypertensive type 2 diabetic under dual therapy was hospitalized in nephrology for acute renal failure evolving for two months, with an altered general condition associated with proteinuria and haematuria. The objective dermatological examination of painful asymmetrical lesions of purpura livedoid with blue appearance of the big toes and two heel necroses; cold extremities with regular, bilateral pulses present. Faced with this subacute picture, multiple diagnoses were eliminated and the diagnosis of cholesterol embolism syndrome was evoked after our consultation (typical skin biopsy confirming the diagnosis). A lesion assessment was requested, namely an ophthalmological and neurological examination, cardiac echodoppler, chest computed tomography (CT) and pancreatic magnetic resonance imaging (MRI).

Results:

Our patient was put on anti-hypertensive, analgesic triple therapy (level I to III); dialysis, statins and systemic corticosteroid therapy at 0.3 mg/kg/day.

Conclusion:

Our case illustrates the rare spontaneous nature of a cholesterol embolism syndrome in a patient at high cardiovascular risk (sex, age, diabetes and arterial hypertension) with multisystem damage (cutaneous, renal, retinal and coronary) and who required a multidisciplinary charge.

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Abstract N°: 4753

Scalp metastasis of Clear Cell Renal Carcinoma in a patient with right radical nephroureterectomy

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Scalp metastasis of Clear Cell Renal Carcinoma in a patient with right radical nephroureterectomy

Introduction

Renal cell carcinoma (RCC) is a genitourinary malignancy of male predominance with an annual incidence of over 73,000 cases in the United States. Clear cell renal cell carcinoma (ccRCC) comprises 75% of these cases, with incidence increasing in recent years. However, only 65% of patients with RCC present with localized disease, while 16% present with metastatic disease. The most frequent metastatic sites are the liver, lung, and bone, while skin metastasis are only 3%. First line treatment, for a complete cure is the resection of localized RCC , but 30% of patients undergoing curative surgery develop metastatic RCC. We present a case of a 64-year-old male man with scalp metastasis of Clear Cell Renal Carcinoma that was treated years previously with right radical nephroureterectomy and needed surgery of the metastatic lesion with immunotherapy.

Results:

A 64-year-old male patient with a history of right radical nephroureterectomy plus bladder wedge and cystoscopy of stage T3NxMx clear cell renal cell carcinoma, was admitted to the clinic due to a 3-month history of scalp lesion in the right hemicranium, without pain, bleeding, or growth.

On physical examination, a circumscribed bright erythematous nodule 20 mm in diameter was observed in the right frontoparietal region of the skull.

He brought the result of a previously taken biopsy. The pathology findings showed skin with rectified epidermis, without alterations. In the dermis, the presence of a lesion occupying the deep superficial dermis is observed, composed of nests of cells with clear cytoplasm, with apparent membranes, arranged in a tree-like pattern, with hyperchromatic nuclei, some of them pleomorphic.

Immunohistochemical markers were performed: strongly positive CKAE1/AE3, EMA, PAX-8 CAIX, CD10, with RCC AND /FOCAL IMENTIN. Other markers, CK7, CK20, D240, S100, HMB45, CD34 CEA were negative. With the microscopic findings, involvement of neoplastic lesion, history and IHC pattern, the metastatic involvement due to clear cell renal tumor was made.

Subsequently, it was treated with surgery, with a pathology report of 11mm in its largest diameter, without renovascular or perineural invasion, with tumor-free section edges. An MRI of the thorax, brain, and abdomen was performed with no signs of secondary metastatic disease. Finally, cancer management was started with immunotherapy with pembrolizumab with good tolerance and response.

Conclusion:

Skin metastases only appear in 0.7-9% of patients with cancer. Clear cell renal cell carcinoma is a tumor that has been shown to metastasize distantly to the skin, especially to areas of scalp, face, and neck. An extension of the disease that implies an increase in the stage of the tumor, should be treated additionally with chemotherapy and immunotherapy.

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Abstract N°: 4786

Retiform Purpura associated with Cold Agglutinin Disease

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Introduction & Objectives:

Retiform purpura (RP) has a specific morphology within the spectrum of reticulate eruptions of vascular origin. In immunocompromised patients, multiple conditions lead to this cutaneous sign, including autoimmune, infectious, or drug-induced microangiopathies. Therefore, determining the etiology of RP can be a diagnostic challenge, even more so, in patients with hematologic disorders. Herein, we present the case of a bone marrow transplant patient with cutaneous vascular occlusion, secondary to cold agglutinin disease (CAD) triggered by a cytomegalovirus (CMV) infection.

Materials & Methods:

A 17-year-old male, with a history of acute lymphocytic leukemia, positive CMV IgG antibodies, and allogeneic stem cell transplantation (ASCT), presented to our clinic six weeks after his ASCT, with purpuric, reticulated patches over the ears, cheekbones, and extremities, that developed 24 hours prior.

Results:

Workup showed red blood cell agglutination on a peripheral blood smear, hemolytic anemia, and a positive Coombs test. Further testing evidenced high titles of cold agglutinins and a viral load for CMV with 1,720 copies/mL. Histopathology revealed vascular occlusion in the superficial and deep dermis. He was promptly diagnosed with an RP associated with CAD, triggered by a CMV infection reactivation. Treatment was established with valganciclovir, achieving complete resolution of his dermatosis after 4 weeks, without signs of recurrence on follow-up.

Conclusion:

CAD is a variant of autoimmune hemolytic anemia, primary (50%) or secondary to lymphoproliferative diseases or viral infections. Although rare (2-6%), an increasing cause of CAD is the reactivation of CMV after an ASCT. It is often reported with acrocyanosis (44%), and Raynaud's phenomenon (33%), however, as Dermatologists, we also should consider RP, as it reflects vascular occlusion or damage. The early recognition of this cutaneous manifestation, especially in immunocompromised patients, is key to a satisfactory resolution and may prevent potentially fatal consequences.

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Abstract N°: 4922

neutrophilic dermatosis of the hands without neutrophiles is it a new entity

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Introduction :

Neutrophilic dermatosis of the hands (NDH) is a rare localized acral variant of Sweet syndrome. NDH predominantly involves the dorsal hands with characteristic dense dermal neutrophilic infiltrate with an upper dermal edema observed on histopathology. We repport 2 unusual cases with palmar involvement and predominant lymphomononuclear dermal infiltration.

Case report :

Case 1:

44 yo female consulted for abrupt onset of redish and violacious palmar plaques evolving since 15 days associated with painful edema of both hands.

Examination revealed multiple erythemato-violaceous plaques topped with bullaes with hemorrhagic and pustular content on both palms associated to a violaceous plaque on the dorsal areas of the right hand and few papules on the feets, otherwise the patient reported signs of respiratory infection 15 days ago, the rest of systemic examination was unremarkable.

Laboratory analysis was notable for leukocytosis 14.340 *10^3µL with neutrophilia (9.340 *10^3µL), elevated erythrocyte sedimentation rate (81 mm/hr) and elevated C–reactive protein level to 64. Skin biopsy was done from a 10 days lesion over palm. Histopathological examination showed perivascular infiltrate comprising predominantly lymphomononuclear cells and plasmocytes; no feature of vasculitis was elicited.

Case 2 :

61 yo female presented with fever and abrupt onset of redish palmar papulo-nodules associated with joint pain. Examination revealed multiple erythemateous papulo-nodules symmetrically distributed over palmar surfaces of both hands, and few papules on the dorsal area of the hands and the feets. The rest of systemic examination objectified joint pain in upper und lower limbs.

Laboratory analysis was notable for leukocytosis 13.820 *10^3µL with neutrophilia (10.870 *10^3µL), elevated erythrocyte sedimentation rate (115 mm/hr) and elevated C–reactive protein level to 124. Skin biopsy was done from a papule over the left palm. Histopathological examination showed marked edema of the papillary dermis with upper and mid-dermal perivascular infiltrate comprising predominantly lymphomononuclear cells and plasmocytes; no feature of vasculitis was elicited.

Discussion:

In 2000, Galaria *et al.* offered a unified designation of “neutrophilic dermatosis of the dorsal hands”, suggesting that it would be a subset of Sweet syndrome. And reported that it has similar clinical features but histology showed neutrophilic infiltration and leukocytoclasis without vasculitis features.

However, as per few previous case reports and palmar involvement as seen in our patients, we excluded the term

“dorsal” from our diagnosis.

In most of the reported cases of NDH, histopathology showed predominantly upper dermal neutrophilic and perivascular infiltrate with or without features of leukocytoclastic vasculitis.

But in our 2 cases both biopsies showed predominantly lymphomononuclear cell infiltrate without features of vasculitis.

NDH is usually responsive to moderate doses of systemic corticosteroids or dapsone,

Both our patients responded well to short-course systemic corticosteroid without any recurrence on 3-month follow-up.

Conclusion:

The purpose of documenting these cases is to highlight the unusual findings of palmar involvement and predominantly lymphomononuclear cell infiltration in NDH, which to the best of our knowledge has not yet been described

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Abstract N°: 4927

:TAMA

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Introduction & Objectives:

Thymoma-associated multiorgan autoimmunity (TAMA) is a rare autoimmune disorder associated with thymoma that causes a pathology similar to graft-versus-host disease (GVHD) targeting the skin, digestive organs, and liver. Herein, we report a case of 53-year-old female with myasthenia gravis (MG) preceded by TAMA.

Materials & Methods:

Results:

She was diagnosed with MG 12 years ago, and thymoma (type3 and type2) was found 3 years later. Thymoma had been resected several times and treated with chemotherapy and radiation therapy. However, it was not completely removed, therefore it had been gradually increasing in size and invaded the pericardial sac. Three months before first visit, she noticed red papules, red macules and erosions on her trunk, subsequently hyperkeratotic papules and erosions spread to the whole body. A biopsy of the macule on the thigh showed hyperkeratosis and parakeratosis, infiltration of mononuclear cells, and interface dermatitis with dyskeratosis, resembling the histological feature of GVHD. Immunohistochemistry revealed that the infiltrating mononuclear cells express mainly CD8(+). Blood examinations showed liver dysfunction (ALT 489 U/L, AST 314 U/L, ALP 302 U/L and γ-GTP 1108 U/L). Gastrointestinal endoscopy revealed numerous erosions and small ulcers in the gastric antrum and sigmoid colon. Finally, she was diagnosed with TAMA and treated with oral prednisolone, intravenous immunoglobulin. Chemotherapy for thymoma was continued by using carboplatin and paclitaxel. The patient was also treated with narrowband-UVB. The skin inflammation and was gradually improved, and the size of thymoma was slightly decreased. The levels of hepatobiliary enzymes were also decreased.

Conclusion:

TAMA is a poor prognostic factor for thymoma patients. It is considered necessary for disease control of TAMA to reduce the volume of thymoma as much as possible by thymectomy and chemotherapy. Narrowband-UVB is known to enhance the regulatory T cell proliferation, and it has been reported that narrowband-UVB is highly effective in acute GVHD skin lesions. Although the pathogenesis of TAMA is not yet clear, we speculate that the modulation and destruction of immune architecture, accompanying regulatory T cell dysfunction, by thymoma may form the pathogenesis of TAMA, which is similar to GVHD.

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Abstract N°: 5029

Secondary erythromelagia induced by diabetic neuropathy

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Introduction & Objectives:

Erythromelalgia (EM) is a rare disorder characterized by episodic erythema and burning pain that commonly involves the extremities. EM can be primary or secondary. Secondary EM can be due to multiple causes, including neuropathic conditions. We present a case of erythromelalgia in a young woman associated with diabetic neuropathy with a spectacular response to aspirin.

Results:

A 39-year-old woman with a history of recently diagnosed type 2 diabetes mellitus presented with complaints of episodic burning pain, redness, and swelling in her feet, which worsened for several weeks. An EMG exam revealed marked diabetic sensory-motor multinevritis of the axon-myelin type in the lower limbs. She received treatment with amitriptyline and clobazam without improvement. Her physical examination revealed circumferential redness and oedema of both feet, with multiple ulcerations of the skin. The extremities were warm to touch. Her symptoms were precipitated and worsened with lying down or exposure to warm temperatures and were abated by cooling measures such as putting the affected limbs in cold water or placing them in front of a cooling fan. She was experiencing multiple daily episodes, each lasting hours. Her symptoms were disabling and included sleeping difficulties. There was no history of drug intake and no family history of similar symptoms. She underwent an extensive evaluation, which included a normal comprehensive metabolic panel and a complete blood count. The autoimmune panel was normal, including negative antinuclear and anti-DNA antibodies. Scanning for internal malignancies with CT imaging was negative. The diagnosis of secondary EM induced by diabetic neuropathy was established and managed with behavioral interventions (limb elevation, cooling techniques with a fan and water) and aspirin (500 mg/day). Two days after starting treatment, her symptoms were controlled, and her sleep was fully restored.

Conclusion:

EM is a relatively unknown disorder. It may occur as a primary or secondary disorder with a similar clinical presentation. EM is characterized by episodes of erythema, increased temperature, and burning pain, mainly involving the extremities. Attacks can last from minutes to days. Triggering factors include heat and exercise. The diagnosis is made by taking a careful history. Relief with ice-cold water immersion is so common and almost pathognomonic of EM. However, it can lead to severe tissue damage, such as skin maceration and ulcerations, as in this case. Secondary causes of EM must be excluded. Screening for a multitude of conditions, including myeloproliferative disorders, connective tissue diseases, infections, and malignancy, was negative in our patient. However, diabetic neuropathy with severe impairment of motor and sensory fibers seems to be the cause of EM in this case. The etiopathogenesis of EM is unknown. Vascular abnormalities, small fiber neuropathy, and arteriovenous shunting are thought to be involved, but the primary abnormality is still unclear.

Avoidance of trigger factors, along with local cooling measures, helps to attenuate symptoms. Managing the underlying condition may be helpful in secondary EM. Pharmacological agents such as anti-epileptics, antidepressants, and calcium channel blockers have varying degrees of success. Aspirin is effective in cases associated with myeloproliferative disorders. However, this case was successfully controlled with aspirin.

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Abstract N°: 5058

Unraveling cutaneous manifestations in Type I cryoglobulinemia with monoclonal gammapathy of undetermined significance

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Introduction & Objectives: Cryoglobulinemia is an entity characterized by the presence of cryoglobulins in the serum that are composed of different immunoglobulins, which has an impact on the clinical presentation and the underlying disease. Cryoglobulinemia is categorized into two subgroups: Type I, which is seen in clonal hematologic diseases, and type II/III, which is called mixed cryoglobulinemia and is seen in HCV infection, hematologic malignancies and connective tissue disorders.

Materials & Methods: We present the case of a 47-year-old woman with a history of cured HBV and seronegative arthritis of 2 years of evolution, with sensory neuropathy in lower limbs, treated with methotrexate and prednisone without improvement. In addition, she presented a monoclonal band in blood IgG Kappa of 7 gr/dl, with a diagnosis of monoclonal gammopathy of uncertain significance (MGUS).

During an admission, the patient developed a condition of racemose livid lesions on lower limbs, which evolved into necrotic plaques with ulceration in some of them.

Two biopsies were taken, one showed fibrin thrombi with polynuclears in an artery, and the other showed superficial and deep leukocytoclastic vasculitis.

With this suspicion, cryoglobulins were requested and were positive for the monoclonal IgGKappa component.

With this finding and the accompanying clinical manifestations, the diagnosis of type I cryoglobulinemia associated with monoclonal gammopathy of undetermined origin was established and treatment with Lenalidomide + Dexamethasone was started, with good response and decrease of IgG Kappa levels in blood.

Results: In type I cryoglobulinemia, the cryoglobulins are monoclonal Igs, typically IgG or IgM,

and it causes hyperviscosity that results from high levels of monoclonal cryoglobulins due to an underlying lymphoproliferative disorder. These Igs precipitate and induce a microthrombotic process within small size vessels. Such aggregates may physically obstruct vessels and rarely mediate inflammatory vasculitis via immune complex deposition, but it may be be found as observed in our biopsy.

Type I cryoglobulinemia classically produces symptoms related to vascular occlusion such as digital ischemia, livedo reticularis, and skin necrosis. Skin manifestations are most commonly observed in these patients, with reports of up to 80%. Extracutaneous disease includes peripheral neuropathy in approximately 20 to 45%, arthralgia in 30 percent, and renal disease in 30%.

These different cryoglobulin isotypes should have an impact on the choice of treatment as the target should be the underlying lymphproliferative disorder. Prognosis depends on the subjacent condition as well.

Conclusion: Cryoglobulinemia type I is a rare condition, which should be suspected in in the setting of protein-secreting monoclonal gammopathies accompanied by livid lesions and necrosis of the lower limbs. The dermatologist can play a fundamental role in the diagnosis if is aware of the disease and knows the main

cutaneous manifestations.

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Abstract N°: 5500

Kyrle disease: navigating the maze towards optimal treatment

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Introduction & Objectives:

Hyperkeratosis follicularis et parafollicularis in cutem penetrans also known as Kyrle`s disease(KD) is a rare controversial entity classified under the generic term of "acquired perforating dermatoses". Classically it is associated with end-stage kidney disease and diabetes mellitus.

Materials & Methods:

We report the case of a Caucasian female presenting with a rare generalised form of KD developed after the initiation of hemodialysis subsequently to diabetic nephropathy.

Results:

A 59-year-old woman was admitted to our dermatology department with umbilicated, round, erythematous and hyperpigmented papules, plaques and nodules with either central crusts or keratotic plugs, along with intense pruritus. The lesions were distributed all over the cutaneous surface except for a well-demarcated area on the central posterior thorax.

A comprehensive anamnesis revealed type 2 diabetes, stage V diabetic nephropathy requiring hemodialysis three times per week, vascular decompensated cirrhosis of mixed etiology (viral-, autoimmune, cardiac), dilatative cardiomyopathy and atrial fibrillation.

The lesions initially appeared three and a half years ago, shortly after the beginning of hemodialysis. The patient was initially diagnosed with prurigo nodularis in another clinic and treated solely with topical corticosteroids and systemic antihistamines. No significant improvement was observed throughout the course of treatment.

Due to the distinctive clinical appearance, a broad differential diagnosis was conducted, which included prurigo nodularis, Kryle disease, multiple keratoacanthomas, hypertrophic lichen planus, reactive perforating collagenosis and perforating folliculitis.

We performed a skin biopsy due to the lack of response to previous treatment along with the worsening of cutaneous lesions in the past. The histopathological result met the Constantine and Carter criteria for KD and along with the clinical aspect, medical history, and dermoscopy examination, allowed us to reach the final diagnosis of Kyrle disease.

Conclusion:

Although considered a rare pathology, KD is not an unusual occurance among patients who have undergone hemodialysis.

The lack of an international guideline for the treatment of KD requires a careful approach. A plethora of treatment options can be found in the literature without being standardized. In our case, the associations of the liver disease impose significant limitations when it comes to systemic treatment.

The distinctive nature of our case begins with the peculiar distribution of the skin lesions and concludes with the treatment challenges. Considering the numerous treatment restrictions imposed by the multitude of systemic comorbidities, renal and liver disease being by far the most significant, the optimal approach for our patient is a combination of topic tacrolimus, retinoids and clindamicyn along with systemic antihistamines and phototherapy.

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Abstract N°: 5502

Association of Behçet's Disease with segmental vitiligo, alopecia areata and poliosis: a case report.

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Introduction & Objectives:

Behçet's disease (BD) is a multisystemic autoimmune disorder, characterized by recurrent oral and genital ulcers, ocular inflammation, and skin lesions. While the exact cause of BD remains unknown, studies suggest that there is a strong association between BD and certain genetic markers, as well as other autoimmune diseases.

Materials & Methods: `

We report the first case in the literature of the co-occurrence of BD, segmental vitiligo, alopecia areata and poliosis in the same patient.

Results:

A 28-year-old woman was admitted to our department for management of segmental vitiligo and alopecia areata. Medical history of the patient included relapsing oral aphthous ulcers for six years, and sudden decrease in visual acuity preceded by analgesic-resistant headaches and vertigo. She did not report other auditory or neurological symptoms.

A physical examination revealed unilateral depigmented patches on the patient's neck, anterior chest, and right arm. Well-defined areas of non-scarring alopecia on scalp. She also had ipsilateral poliosis. On trunk, folliculitis and pseudofolliculitis were noticed. An ophthalmological examination resulted in optic atrophy with sequellar arteriovenous vasculitis.

A pathergy test was negative. Laboratory examination showed serum ferritin: 5,78 ng/ml; Vitamin D: 20,21ng/ml; C-reactive protein: 5.74 mg/L; erythrocyte sedimentation rate: 4mm/h; thyroid stimulating hormone: 3,790 uUI/ml; antithyroglobulin antibody test: <6,4 UI/ml; antithyroid peroxidase antibody test: <0,8 UI/ml. Additionally, laboratory results mentioned above showed an iron deficiency anemia. The immunological assessment was negative. MRI showed small cysts of the choroid plexus in the two posterior ventricular junctions and optic nerve atrophy.

A diagnosis of BD was made according to the International Criteria for Behçets Disease (ICBD). Our patient had 5 points: 2 for oral aphthosis, 2 for ocular manifestation and 1 for skin manifestation. Vitiligo, poliosis and alopecia areata was diagnosed based on prior physical examination.

Therapy was started with oral colchicine in a dose of 1 mg per day along with iron and Vitamin D3 supplementation. And for the alopecia we opted for topical clobetasol in combination with minoxidil.

Conclusion:

Herein, we are presenting a very uncommon case of a patient who has been diagnosed with BD, segmental vitiligo, alopecia areata, and poliosis, all at the same time. This remarkable medical condition provides a unique opportunity to gain a better understanding of the autoimmunity and autoinflammatory process involved in both

diseases by examining the patient’s clinical and laboratory data. By doing so, we hope to identify both shared and distinctive features of these conditions and contribute to the knowledge base of these rare disorders.

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Abstract N°: 5607

Psoriasiform skin lesions in a patient with history of thymoma: Thymoma-Associated Multiorgan Autoimmunity, a rare disease.

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Introduction & Objectives:

Thymoma-Associated Multiorgan Autoimmunity (TAMA) is a multiorgan disease with clinical and histologic similarities to graft-versus-host disease (GVHD), but without a history of hematopoietic transplantation which appears in patients with a history of timoma. To date, only 31 cases of this condition have been described.

Materials & Methods:

A case of TAMA is reported.

Results:

A 49-year-old woman with history of seropositive myasthenia gravis associated with invasive thymoma was admitted to hospital. She consulted for general malaise, febrile peaks, diarrhoea, constitutional syndrome and skin lesions. Physical examination by organs and apparatus was unremarkable, with the exception of minimal ocular muscle fatigue in relation to her underlying pathology and skin lesions. At the cutaneous level, she presented papules and well-demarcated erythematous-squamous plaques, psoriasiform in appearance, located mainly on the neckline and upper back. No mucosal or other type of lesions were observed. Biochemical tests included, altered liver profile with dissociated cholestasis, with the normal values of transaminases and bilirubin. A variety of positive autoimmune markers were found: Antinuclear antibodies + 1/230, Anti-centromere +, Anti-histone +, Anti-nucleosome +, as well as Anti-Acetylcholine Receptor + and Coombs test + without anemia.

Given the absence of etiological diagnosis skin biopsy was performed. It showed a vacuolar interface dermatitis, with apoptotic bodies and lymphocytic exocytosis, as well as a superficial perivascular lymphocytic infiltrate. All these changes were compatible with a graft-versus-host reaction. The diagnosis of TAMA was performed.

Despite treatment with systemic corticosteroids and supportive measures, skin worsened into erythroderma with severe liver disease, multiorgan failure associated with bacteremia and death of the patient.

Conclusion:

The presence of psoriasiform skin lesions or erythroderma in a patient with a history of thymoma, especially if there is evidence of positive autoimmunity and hepatic or digestive involvement, should lead to suspect the possibility of TAMA.

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Abstract N°: 5614

Kaposi's sarcoma aggravated by immune reconstitution syndrome, a rare cause of death in HIV patients.

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Introduction & Objectives: We describe a case of death caused by metastatic Kaposi's sarcoma (KS) after starting antiretroviral therapy in an HIV-infected patient.**

Materials & Methods: Report of a case and bibliographic review.

Results: A 32-year-old male patient, recently diagnosed with HIV with a CD4 T-cell count of 0.02 cells/ml and a viral load of 26,000 copies/ml, was admitted to the infectious diseases department for acute respiratory failure. Bronchioloalveolar lavage PCR excluded *Pneumocystis jirovecii* infection and nasal exudate PCR was positive for SARS-COV2. Ventilatory support therapy and antiretroviral therapy (bictegravir/emtricitabine/tenofovir alafenamide) were established.

On day two of admission, the patient was referred to dermatology due to the discovery of violaceous plaques distributed on the lower limbs, midfacial area and oral mucosa. The lesions were diagnosed as KS, an incisional biopsy was obtained to confirm the diagnosis and a chest CT was requested to rule out visceral involvement.

The CT scan identified lymph node, pleural and pulmonary involvement compatible with KS metastatic infiltrate. A bronchoscopy with sampling (biopsy, cytology) was performed for diagnostic confirmation.

Fourteen days after admission, despite a negative PCR for SARS-COV2, the patient developed a clinical deterioration of his respiratory symptoms. Finally, he died in the Intensive Care Unit. The diagnosis of metastatic KS was confirmed postmortem by histological study of skin and lung samples.

Conclusion: Immune reconstitution syndrome has been associated with KS progression within the first three and six weeks of initiation of antiretroviral therapy. Risk factors identified are: increased extent of Kaposi's sarcoma, high HIV viral load, and antiretroviral therapy instituted without chemotherapy. Progression of Kaposi's sarcoma can be severe and has been associated with death in some patients.

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Abstract N°: 5718

Papules do not always have a fatal prognosis

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Introduction

Degos disease or malignant atrophic papulosis is a rare, multisystem and potentially life-threatening disorder characterized by a diffuse papular skin eruption associated with inner organs involvement, either occurring simultaneously or subsequently. The systemic manifestations can be followed in many cases by serious complications. Most of the lethal courses are due to bowel perforation or central nervous system lesions and their consequences.

Clinical case:

A 30-year-old presented in 2018 with a two-years history of small papules of normal skin color, distributed over the central chest, back, and thighs, some of them umbilicated, which progressively atrophied, leaving a central white spot and small vessels on the periphery. The skin biopsies performed were compatible with Degos disease at different stages of clinical evolution. In addition, laboratory work-up, including coagulation and autoimmunity studies, brain MRI and fecal occult blood test requested were negatives. The patient is currently receiving treatment with acetylsalicylic acid and pentoxifylline, and after five years of follow-up he has not presented gastrointestinal or neurologic symptoms.

Discussion:

Degos disease is a rare thrombo-obliterative vasculopathy characterized by papular skin lesions with central porcelain-white atrophy and surrounding teleangiectatic rim. Systemic manifestations are progressive and can cause serious complications, leading to lethal course in approximately 50% of patients within 2-3 years. There is a minority of patients (approximately 15%) who present a benign course, without systemic involvement and long-term survival. To emphasize this variable clinical picture, some authors advocate terms such as benign cutaneous Degos' disease. However, systemic manifestations can develop years after the appearance of the skin lesions, that is why these patients require continuous monitoring. The probability of a benign course increases with the duration of monosymptomatic cutaneous disease.

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Abstract N°: 5775

Challenges in the differentials of figurate erythemas

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Introduction & Objectives:

Figurate erythemas are a hetergenous group of diseases, characterized by their annular erythematous skin lesions. The diagnosis is made by clinical examination, histological findings and sometimes by exclusion. There are four classic FE: erythema annulare centrifugum, erythema migrans, erythema gyratum repens, and erythema marginatum, the differential diagnosis being numerous and often challenging. Etiologically FE have been linked with underlying conditions including neoplasms, autoimmune diseases, infections or drugs, a lot of cases with no trigger being found.

Case reports:

Case 1. We report the case of a 50 years-old woman who presented with multiple, disseminated, annular lesions, up to 10 cm diameter, with a slightly elevated papulo-vesiculous, infiltrated border, central clearing and trailing scale, localized on the trunk, buttocks, thighs and lower extremities. The lesions appeared 3 years prior, with the spontaneous resolutions of the lesions and the appearance of new ones periodically. The patient was known with chronic hepatitis C treated, since 2010 and no other underlying conditions. The laboratory findings showed a nasal SAH infection. The skin biopsy showed parakeratosis, moderate focal spongiosis in the epidermis and focal and perivascular chronic inflammatory infiltrate in the papillary dermis, suggestive for the superficial form of erythema annulare centrifugum. The therapeutic response was good under systemic and topic corticotherapy.

Case 2. The second case is that of a 46 years-old woman that presented with a six weeks history of a progressing, expanding presternally plaque, with an erythemato-papulous, slightly elevated and infiltrated margin, with a clear center and a diameter of 5/7 cm. The skin biopsy showed slightly acanthosis in the epidermis and moderate to important perivascular lymphocytic inflammatory infiltrate in the dermis. Tests were positive for Borrelia Burgdorferi and systemic therapy with doxycycline was started with a good therapeutic response. There are several clinical variants of erythema migrans, the differential diagnosis being broad and any erythematous patch in an endemic area for Lyme disease must raise the suspicion of the disease.

Discussion, conclusion:

Figurate morphology can appear in many cutaneous diseases, so the differential diagnosis of FE can be challenging and includes infectious diseases (tinea corporis or secondary syphillis), autoimmune diseases like SCLE, psoriasis, neoplastic diseases like MF, allergic or other diseases.

Some authors regard FE as defined clinical reaction patterns to specific stimuli, rather than distinct clinical entities.

In our cases, both of diseases have been triggered by the underlying infectious diseases, a comprehensive history of the patient, the clinical aspect, temporal evolution, histological aspect of the lesion, but also the laboratory results being necessary for the correct diagnosis.

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Abstract N°: 5781

A rare case of NXP-2 and TIF1-γ positive dermatomyositis

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Introduction & Objectives: A rare case of NXP-2 and TIF1-γ positive dermatomyositis

Materials & Methods: Clinical case report

Results: Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterised by progressive and symmetric proximal muscle weakness and skin changes. We presented a 62-year-old man with NXP-2 and TIF1-γ antibodies positive DM.

He presented with a florid rash of two months duration which started on his face which then spread to his torso, hands and nails. He also reported pain on the arm in particular the shoulder area. He is on sulpiride and fluoxetine for schizophrenia. Clinically, he has erythematous scaly patches on his face, V neck of upper chest, periungual erythema with dilated capillaries under dermatoscope. A skin biopsy has shown features suggestive of dermatomyositis. His condition deteriorated rapidly while waiting for further investigation with dysphagia and aspiration pneumonia. Intravenous immunoglobulin and hydrocortisone were given over five days which improve his skin and muscle power. MRI scan of the thighs has shown muscle oedema. Myositis antibody panel has shown positive NXP2 antibody and a weekly positive TIF1-γ antibody. Electromyography and subsequent muscle biopsy have also shown evidence of myositis. Positron emission tomography scan and whole body computed tomography has excluded underlying malignancy. Subcutaneous methotrexate 15mg once weekly was administered. A percutaneous endoscopic gastrostomy feeding tube was inserted due to recurrent aspiration pneumonia. He is currently scheduled for rituximab infusion due to poor respond to methotrexate.

Many autoantibodies subsets are associated with unique clinical phenotypes of dermatomyositis. Anti-NXP-2 autoantibodies in DM are associated with subcutaneous oedema, calcinosis, and a severe muscle phenotype characterized by myalgia, proximal and distal weakness and dysphagia. There is also increased risk of cancer in this group of DM. TIF1-γ antibodies have a strong association with malignancy in patients over the age of 39. Systemic steroid remains the main stay of treatment for DM. Steroid-sparing agent such as azathioprine and methotrexate can be used. For treatment resistant disease, rituximab can be used. While most patients respond to corticosteroids and immunosuppressants, approximately one third of patients do not respond or poorly respond to available therapies and remain debilitated such as this patient.

C**onclusion:**

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Abstract N°: 5802

Association of lichen planus and dysthyroid disease

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Introduction & Objectives:

Lichen planus pigmentosum (LPP) is an uncommon variant of Lichen plan.

This dermatosis has rarely been described in association with other diseases. Thus, through our work, we set out to determine the frequency of dysthroidism among our patients followed for LPP.

Materials & Methods:

We present a single-centre prospective study conducted at our dermatology department. For all patients followed by LPP, a thyroid hormone assay was performed to detect dysthroidism.

Results:

There were 24 histologically confirmed cases of LPP all of them were femal, aged 20-62 years, and the majority (80%) were aged 20-40 years. At presentation, twenty-three (85.2%) of the patients had progressive disease manifested by the appearance of multiple new hyperpigmented macules, involvement of new body areas or an increase in the size of older lesions over the last 3-4 months. Dysthyroidism was found in 7 cases of which 3 had hashimoto's thyroiditis, 3 had hypothroidism, and one had associated hyperthroidism.

Hashimoto's thyroiditis is a chronic autoimmune disease. It is characterised by privileged associations with many autoimmune diseases, including dermatological diseases including the LPP. This association has been described in the literature by case reports. We report 7 cases out of 24 (29%).

A common autoimmune process would probably be the origin of the existence of such an association.

Conclusion

The results of our series show a frequent association of LPP with dysthroidism. However, further studies are needed to better characterise this entity.

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Abstract N°: 5809

A cross sectional study of nail changes in CKD patients on hemodialysis

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Introduction & Objectives: chronic kidney disease patients undergoing hemodialysis presents with a array of mucocutaneous, nail and hair changes. Nail changes are commonly observed and may be directly related to the renal condition, it's complications or due to therapy.

To study the prevalence and characteristics of different nail changes in ckd patients on hemodialysis

Materials & Methods: study compromised of 227 patients of CKD undergoing regular hemodialysis for a minimum of 3 months. After taking informed written consent, a thorough history and physical examination including nail examination was done. Complete blood count, renal function test , serum parathyroid hormone levels were investigate

Results: Nail changes were present in 70.9%(161 patients).Longitudinal ridging was the most common nail change seeing in 20.3% patients, followed by – in descending order half and half nails (19.4%), absent lunula (13.2%), subungual hyperkeratosis (9.7%), melanonychia (6.6%), dystrophic nails (4.8%), clubbing (4.4%). The least prominent changes were muehrcke's lines and pincer nail (0.4% each). Common age group affected was 41 to 60 years. Nail changes were seen more commonly in males (51.4%). Their was no statistically significant correlation between the age of the patient or the duration of hemodialysis with the nail changes. Nail changes were common in patients with low hemoglobin levels.

Conclusion: Patients with chronic kidney disease have higher rates of nail disorders when compared to healthy population.

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Abstract N°: 5812

Leukaemia cutis versus Sweet syndrome – a difficult and fateful differential

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Introduction: Skin involvement in myeloproliferative diseases can occur through direct infiltration of malignant lymphocytes – leukaemia cutis - or as a consequence of reactive inflammatory processes. Neutrophilic dermatosis encompass a group of inflammatory skin disorders characterized by the accumulation of neutrophils in the skin, which often associate with systemic diseases, particularly haematological disorders. Understanding and recognizing these skin manifestations is crucial for accurate diagnosis and appropriate management. As the histopathological representation of these two phenomena can have overlapping features, its differential diagnosis may be challenging.

Case report: We report the case of a 59-year-old male with acute myeloid leukaemia (post chronic myelomonocytic leukaemia) under treatment with venetoclax azacytidine for 2 months. He presented with a painful abdominal skin lesion which appeared 24 hours after his subcutaneous administration of azacytidine, with centrifugal growth for 15 days. He reported episodic fever for 3 weeks. On physical examination, he had an abdominal, oval, erythematous, smooth surfaced, 30 cm plaque, extremely infiltrated, with two concentric peripheral rings and two central flaccid blisters. Blood work was significant for elevated c-reactive protein. Skin histopathology revealed diffuse infiltration of the reticular dermis and epidermis by granulocytes, mainly mature neutrophils, with rare dysplastic granulocytes, but no blasts. Images of karyorrhexis were frequent. Immunophenotyping of skin cells was inconclusive. There was no other evidence of disease progression. Faced with a neutrophilic dermatosis with a "Sweet's syndrome-like" pattern, without blasts, but with evidence of dysplastic cells the diagnostic doubt remained. Concurrent Sweet's syndrome and leukaemia cutis has been reported and can be attributed to two explanations: either the circulating immature myeloid precursor cells are innocent bystanders that have been recruited to the skin as the result of an inflammatory oncotactic phenomenon stimulated by the Sweet's syndrome reactive process, or primary specific leukaemic infiltrates trigger the reactive process. Localized azacytidine induced Sweet syndrome has been rarely reported in the literature. The patient started treatment with prednisolone 0.5 mg/kg/day. The pain improved in 48 hours and only a residual light-purple patch remained after 3 weeks of treatment. The patient kept his treatment with intravenous azacytidine, without recurrence after prednisolone's suspension.

Conclusion: This case illustrates an atypical presentation of neutrophilic dermatosis which responded to systemic corticosteroid therapy, fulfilling diagnostic criteria for Sweet syndrome, even with histopathologic leukaemia cutis characteristics. The clinical and histopathological overlap of the two diagnoses can lead to a hasty presumption of disease progression, altering the therapeutic approach.

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Abstract N°: 5817

Paraneoplastic dermatomyositis revealing an underlying inflammatory breast cancer

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Introduction & Objectives:

Breast cancer is the most common cancer among women in Morocco and worldwide. In contrast, dermatomyositis is a rare idiopathic autoimmune inflammatory disease, classically combining specific cutaneous manifestations and progressive muscular deficiency. It is associated in 18 to 32% of the cases with various types of cancer, thus appearing as a symptom of a paraneoplastic syndrome. However, it is rarely the initial presentation of breast cancer.

We report a case of rapidly aggravating dermatomyositis revealing an underlying inflammatory breast cancer in a menopausal woman, representing a therapeutic challenge for both dermatologists and oncologists.

Materials & Methods:

Results:

Case presentation :

A 50-year-old menopausal patient, with a pathological history of repeated miscarriages, consulted for a pruritic maculo-papular eruption involving the four limbs and the trunk, associated with inflammatory arthralgia and spontaneous myalgia, evolving for one month in a context of decline of the general state.

Our clinical examination showed pathognomonic skin manifestations such as Gottron papules in*** dorsal metacarpophalangeal and interphalangeal joints, along with a facial erythema, a V-shaped erythema of the neck and the upper chest, and a periungual involvement combining telangiectasias and cuticular overgrowth. It also found a mass in the upper-external quadrant of the right breast, resting on an inflammatory skin. The patient was admitted in our department, and developed during her hospitalization a gradually progressive bilateral and symmetric proximal muscle weakness, later complicated by dysphagia. The diagnosis of dermatomyositis was made, based on the aforementioned cutaneous and muscular manifestations, along with increased serum muscle enzymes levels and abnormal findings in electromyogram indicating myopathy. The immunological tests were positive for both Anti-nuclear and Anti-Tif1 antibodies.

In parallel, an echo-mammography exploration followed by a Trucut-needle biopsy were performed on the breast mass before retaining the diagnosis of malignancy.

The initial treatment consisted of an IV bolus of methylprednisolone, then relayed by oral prednisone at a daily dose of 1mg/kg. The patient underwent an assessment of extension by imagery to stage her breast neoplasm before any further specific treatment. She was managed in collaboration with the oncology department where a neoadjuvant chemotherapy line has been proposed.

Conclusion:

Paraneoplastic dermatomyositis can reveal, accompany or complicate any type of cancer in both male and female adults, although the exact physiopathology of cancer-associated myositis remains obscure. Therefore, occult

malignancy should be taken into consideration whenever a patient in the older age group is newly diagnosed with dermatomyositis, and further investigations should be carried out.

Early diagnosis and prompt treatment of malignancy may halt the spontaneous progression of dermatomyositis and prevent its severe functional repercussions. However, the treatment plan remains delicate and depends on both the severity of the dermatomyositis and the presentation of the tumor. Indeed, there is, to this day, no standardized treatment for breast cancer associated with dermatomyositis; hence the interest of a multidisciplinary physician approach and collaboration to ensure a better prognosis for these patients.

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Abstract N°: 5857

A bullous rash in infant: a diagnosis not to be ignored!

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A bullous rash in infant: a diagnosis not to be ignored!

Introduction & Objectives:

Cutaneous mastocytosis mainly affects children from birth to 2 years of age and adolescents over 15 years. We report 3 cases of cutaneous mastocytosis of infants in a bullous form and describe its specificities.

Results:

There were 3 infants (2 girls and 1 boy) with a mean age of 2.5 months. There was no notable pathological history, the pregnancy was well monitored and the mother had no bullous dermatosis. Clinically, we found: extensive bullous lesions evolving for 1 month in a context of apyrexia in the first case, pruritic erythematous plaques on the face, trunk and limbs with bullous lesions with crusts and excoriations on the scalp in the second patient, an erythematopapular placard in the supra-mammary region since birth, surmounted by taut bullae in the third case. Darier's sign was positive in two cases. The haemogram, liver and kidney function tests were normal in all cases. The tryptasemia rate (performed in 2/3) was high. The biopsy in the first two patients was in favour of cutaneous mastocytosis and the diagnosis of diffuse cutaneous mastocytosis in its bullous form was retained (1st and 2nd case) and mastocytosis with bulla (3rd case). Treatment was based on local care and antihistamines with good parental education. The evolution was marked by the recurrence of lesions with no notable systemic involvement.

Conclusion:

Pediatric cutaneous mastocytosis is a rare and benign disease with a varied clinical presentation. The most common form remains urticaria pigmentosum (90% of patients). Bullous mastocytosis includes bullae that appear on skin without pre-existing lesions, whereas bullous mastocytosis occurs on papular or nodular lesions. Bullae may be tense and/or haemorrhagic. The number of lesions generally correlates with the severity of the disease. As in our cases, infants with a bullous lesion as the first manifestation have a poor prognosis. Treatment of bullous mastocytosis depends on the severity of symptoms and may include antihistamines and corticosteroids to reduce symptoms and prevent complications.

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Abstract N°: 5903

Alopecia areata and Hashimoto's thyreoiditis

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Alopecia areata and Hashimoto's thyroiditis

Introduction & Objectives:

Hashimoto's thyroiditis is an autoimmune disease characterized by the presence of TPO and Tg antibodies. The disease may be accompanied by subclinical or clinical hypothyroidism. Alopecia areata is also an autoimmune disease which may affect the scalp, the beard or the whole body. The aim was to present a cohort of 5 patients with alopecia areata who were diagnosed with Hashimoto's thyroiditis.

Materials & Methods:

A cohort of 5 patients, 4 female aged 35 to 56 years, and 1 male patient aged 50 years is described who presented with alopecia areata and Hashimoto's thyroiditis. In all female patients alopecia areata affected the scalp while in the male patient affected both the scalp and the beard. In all patients thyroid function was evaluated.

Results:

All patients were found to have Hashimoto's thyroiditis with positive TPO and Tg antibodies. All patients had hypothyroidism, 4 clinical hypothyroidism and 1 female patient subclinical hypothyroidism. In 3 of the patients vitamin D deficiency was diagnosed. In 1 female patient iron deficiency anemia was also observed. Thyroxine was administered to all patients. Cholecalciferol was administered to the patients with vitamin D deficiency. In the patient with iron deficiency anemia iron supplementation was performed.

Conclusion:

In conclusion, alopecia areata may be accompanied by Hashimoto's thyroiditis and subclinical or clinical hypothyroidism. Vitamin D deficiency may also be observed in this context. Patients should be administered thyroxine to restore euthyroidism. Vitamin D and iron should also be administered as it may have beneficial effects on alopecia. As JAK inhibitors are already tested to be used in the armamentarium against alopecia areata it is important to further investigate the condition as well as its connection with Hashimoto's thyroiditis.

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Abstract N°: 5970

Mycoplasma Pneumoniae Infection and progressive cold agglutinin syndrome in a child: Catastrophic Multiple Cutaneous Necrosis

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Introduction & Objectives:

Cold Agglutinin Syndrome (CAS) is characterized by the high titer of Cold Agglutinins (CAs), which clump Red Blood Cells (RBCs) together at cold temperatures and cause blockage in microcirculation. CAS may occur as a consequence of Mycoplasma Pneumoniae (MP) infection.

MP can manifest in various pulmonary and extrapulmonary symptoms, including dermatological manifestations such as nonspecific rash, urticaria or Stevens-Johnson syndrome, mucositis, and Henoch–Schoenlein purpura. Also, progressive necrosis as a manifestation of CAS due to MP infection is rare in children.

Materials & Methods:

In this report, we describe a 9-year-old girl who presented with acrocyanosis and progressive necrosis of the acral sites after pneumonia associated with a high level of cold agglutinins in MP infection.

Results:

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Conclusion:

Acrocyanosis and gangrene as manifestations of CAS caused by MP infection are rarely seen in children and it may leed to undesirable complications. So, it alert clinicians and needs proper supportive care. In the case we discussed, administration of FFP, plasmapheresis, corticosteroids, and IVIG led to a satisfactory improvement in the patient's condition.

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Abstract N°: 6061

Eruptive xanthomatosis

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Eruptive xanthomatosis

Filipović Nika, Buljan Marija, Šitum Mirna, Filipović Nika

Introduction & Objectives: Eruptive xanthomas are localized lipid deposits in the dermis and an important early clue to severe hypertriglyceridemia. They usually occur as an eruption of multiple erythematous-yellow, dome-shaped papules on the extensor surfaces of the extremities, buttocks, and hands. Early lesions may have an erythematous halo and are associated with pruritus and tenderness. A Koebner reaction may occur. Eruptive xanthomas can occur in people with primary or secondary dyslipidaemias which can be associated with poorly controlled diabetes, cholestatic liver disease, or some medications, such as isotretinoin, estrogens, and cyclosporine.

Materials & Methods: In this report we would like to present a case of a 30-year-old male patient who was referred to our clinic due to a sudden eruption of multiple, densely disseminated, erythematous-yellowish papules on his back, abdomen, upper and lower extremities. The lesions were asymptomatic and occurred on previously clinically unchanged skin 2 months earlier. The patient was otherwise healthy, he never had a serious illness in his life, nor was he taking any medication.

Results: A biopsy of the skin lesion confirmed the histopathological diagnosis of a xanthoma while a subsequent diagnostic test detected marked hypertriglyceridemia (values 50x higher than physiological values), uncontrolled diabetes, and kidney and liver damage. Shortly after, the oral therapy with statins (atorvastatin and rosuvastatin) was initiated. The patient was also referred to the Clinic of Metabolic Diseases for further diagnosis and combined hyperlipidemia therapy.

Conclusion: The present case shows how important is the role of a dermatologist in the process of metabolic disease diagnosis. In the case of our patient further diagnostic tests were done only after dermatologist's medical consultation since the eruption was the first symptom of a lipid abnormality. The early medical treatment has probably prevented a young man from serious health consequences.

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Abstract N°: 6071

a case of pancreatic cancer revealed by pemphigus

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Introduction & Objectives:

Paraneoplastic pemphigus (PPN) is a very rare disease associating erosive stomatitis, polymorphic skin eruption and neoplasia, known or revealed at the time of diagnosis of PPN [1] .

We report a case of PPN revealing pancreatic cancer.

Results:

A 65-year-old female patient with a history of depression and type 2 diabetes ;

presented for 1 year erosive-crusty bullous lesions and urticarial plaques taking on a cocardiform appearance located on the backs of both hands, feet

, the folds, perioral and periocular associated with bucco-genetal erosions, with significant weight loss. The histopathological study has

epidermal bulla associated with a significant lymphocytic dermal inflammatory infiltrate and fibrovascular remodeling, congestive immunofacial

The indirect immunofluorescence did not reveal any anti-cellular substance or anti-basement membrane antibodies.

A cerebro-thoraco-abdomino-pelvic CT scan was performed in front of the

polymorphism and the cachectic state, revealing a process of the head of the pancreas not invading the surrounding tissues, confirmed by a pancreatic MRI.

The rest of the assessment was unremarkable.

The diagnosis of PPN was made in view of the association of clinical and paraclinical

criteria and the revelation of a pancreatic tumor, the patient was put on systemic corticosteroids resulting in a good response on the skin

however the mucosal involvement was resistant.

The patient was presented in a PCR where the decision was to refer her for surgery for the management of her pancreatic tumor.

Conclusion:

In addition to the high rate of association of hemopathies with PNN, solid cancers are also to be looked for, especially since their prognosis is severe.

This is what our observation highlights.

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Abstract N°: 6084

Lupus Panniculitis Complicated By Favus Infection

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Introduction & Objectives:

Lupus panniculitis is a rare form of lupus that primarily affects subcutaneous fat. It may be difficult to diagnose as it resembles other hypodermatitis, particularly if no systemic or other cutaneous lesions are found.

We report a new observation complicated by favus infection.

Case report:

A 42-year-old female patient with a four-year history of cutaneous and hematological lupus with poor adherence to treatment as she stopped all medications after a year. She presented with lesions since seven months.

Clinical examination found multiple nodules confluent in sclerotic and atrophic plaques, ulcerated in some places and covered with thick crusts, located in the lumbar region and at the root of the upper limbs.Scalp examination found a subtotal alopecia, and yellowish crusts. Trichoscopy found large cicatricial alopecia patches and follicle plugs, associated with areas of broken hairs, comma hairs, with corkscrew hairs.

Direct mycological examination showed the presence of mycelia filaments associated with numerous spores and an endothrix-type hair parasitism. Culture was negative.

Skin biopsy of the scalp found multiple spores and mycelial filaments on the surface and in the stratum corneum. Nodule’s biopsy showed an inflammatory infiltrate, dermal fibrosis associated with mixed hypodermatitis.

Systemic assessment found bicytopenia and positive antinuclear antibodies. The diagnosis of lupus panniculitis associated to SLE, LED complicated by Favus was retained.

Therapeutic management consisted of oral Griseofulvin 1 gram a day for 6 weeks, associated with general steroids and hydroxychloroquine.

The following up found a regression of clinical and dermoscopic signs of favus and improvement of skin lesions, but persistence of sclerotic and dyschromic scars.

Discussion:

Lupus panniculitis a rare manifestation of lupus (1 to 3%). It presents as subcutaneous nodules or grouped in patches. Lesions progress generally towards lipodystrophy and subsequently towards depressed, unsightly and definitive scars. Approximately, half of patients with have evidence of systemic lupus.As it was the case in our patient.

Favus is a rare form of tinea capitis that manifests clinically with scutula:a yellowish cup-shaped perifollicular crusts, with a smell of cheese or mouse nest. They are composed of mycelial filaments and epithelial debris.

The cause is Trichophyton schoenleinii. However, scutula-like lesions have been described with Trichophyton

mentagrophytes var. quinckeanum, Trichophyton violaceum and more rarely with microsporum gypseum

A few cases have reported favus infection in lupus patient. Sanusi and al report the case of a 56-year-old lupus patient, presenting with yellowish crusty lesions initially on the scalp with generalization to the trunk, shoulders and buttocks which coalesced into a scutula. Histology showed filaments and spores in the stratum corneum. Mycological culture identified a microsporum gypseum.

Feng et al report the case of a 16-year-old patient with a recent diagnosis of systemic lupus, who presents with rounded yellowish crusty lesions, on the right thigh. Mycological culture identified M.gypsum. In our case the responsible mycotic agent could not be identified.

Conclusion:

Lupus skin lesions can develop a favus infection at any point in evolution and can be favored by immunosuppressive medications. The reasons behind the occurrence of the disseminated forms in lupus patients, however, are still unknown.

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Abstract N°: 6273

A narrative overview of therapeutic uses of homeopathy in selected chronic disorders

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Introduction & Objectives: Homeopathy is a therapeutic method, which is holistic in nature, i.e. treats the organism as a whole. Homeopathic medicines are highly diluted substances, thus free of risks of allergy, drug interactions and teratogenesis. Homeopathic medicines are useful in a wide variety of conditions, including upper respiratory tract disorders, inflammatory dermatoses. There is an increasing interest in the use of complementary and alternative medicine, including homeopathy, by patients, therefore practitioners need to be aware of what homeopathy can do, in order to better help their patients.

This study offers a short review of the available data on the application of homeopathy in some common conditions.

Materials & Methods: The literature from 2 electronic databases, PubMed and Web of Science, were searched using the search terms**: "**homeopathy ", "cancer therapy"," type 2 diabetes ", "complementary and alternative medicine", "COVID-19" and "SARS-CoV-2"

Results:

Many studies have been published on the value of homeopathy in the therapy of diverse diseases including cancer, depression, psoriasis, allergic rhinitis, asthma, otitis, migraine, neu-roses, allergies, joint disease, insomnia, sinusitis, urinary tract infections and acne, to name a few.

Current literature suggests a positive influence of additive classical homeopathy on global health and well-being in different diseases. Homeopathic therapy promises health-enhancing lifestyle changes intended to improve the well-being of both adults and children, but research to assess its cost-effectiveness and clinical efficacy in larger studies is required. Since the findings of greater perceived benefits of homeopathy are consistent with the studies published in the medical literature, healthcare workers and policymakers should take into consideration the integration of homeopathic therapies into current medical practice in order to satisfy patients' needs (e.g., a greater sense of autonomy regarding their health as well as their emotional state).

Conclusion: The results suggest that homeopathy may be associated with symptom improvement for these diseases, but this assertion requires substantiation by more rigorous, randomized, controlled trials.