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Abstract N°: 286

Multiparameter analysis of panniculitis reveals three clusters of patients

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Introduction & Objectives:

Panniculitis are characterized by a triple heterogeneity in (1) clinical presentation with multiple clinical forms,

2. histopathology as lesions may show primarily lobular or septal inflammation with or without vascular involvement and in (3) etiologies including autoimmune, metabolic, infectious, vascular, paraneoplastic and often idiopathic panniculitis. Publications are scarce on this subject with mostly case studies. Therefore, a better classification remains necessary to improve clinical management. The aim of this study is to use a specific epidemiological method, namely that of clusters, in order to characterize a large series of individuals with panniculitis and to describe homogeneous groups of patients.

Materials & Methods:

Patients were selected from the pathological registries and main or secondary diagnoses coded during clinical outpatient visit or hospital stay in three university hospital centers for a period of five years between January 2018-2023.

Results:

299 patients were included. Patients' sociodemographics and past medical history are cited in table 1. Most common clinical presentation was subcutaneous nodules (87.3%) located mainly on lower limbs (67.6%). Histologically, the majority of patients had mixed panniculitis (53.8%) and 51.2% of subjects had a polymorphic infiltrate in the deep dermis and hypodermis. No cause was found in 32.8% of patients considered as idiopathic panniculitis. 17.7% of patients had erythema nodosum, with no cause identified in more than half of the cases. An autoimmune disease was found in 14.2% and an infectious cause in 9% of patients. 72.2% of patients received multiple treatments for anti-inflammatory, anti-infectious or immunomodulatory purposes. We completed this study with a clusterization method using clinical, pathological, biological and imaging parameters and we found that the population could be divided into 3 clusters (tables 2, 3 and 4). The first cluster included half of the patients in this cohort and was characterized by multiple episodes of bilateral

nodular lesions with systemic signs, in young patients (< 50 y.o) without a history of cardiovascular diseases, without vascular involvement on histology, and treated with colchicine and hydroxychloroquine. The other half were distributed between 2 clusters. Cluster 2 patients had unilateral infiltrated erythematous plaques (87%) without systemic signs (97%). Histologically, they had mixed panniculitis (75%) and an inflammatory infiltrate in the hypodermis in almost all cases and vascular involvement in 40% of cases. They were treated with antiinfective treatments (39%) and topical steroids (21%). On the contrary, cluster 3 included elderly patients (65%), with a history of cardiovascular diseases who presented non-painful lesions (39%) limited to the lower limbs (43%) without edema (91%), without systemic signs (90%) and without inflammatory infiltrate in the hypodermis (55%).

Conclusion:

This large series highlights the clinical and histological heterogeneity of panniculitis with approximately 33% of idiopathic cases for which a consensual attitude is lacking. Using clustering analysis, we were able to describe three homogeneous groups of patients based on sociodemographic status, clinical findings, histological features and treatment modalities. This could help building foundations to future prospective studies and therefore a better management of this poorly understood entity.

Patients' characteristics	Total (n=299)
Age, y [median]	51.0 [38.0-62.0]
Female sex	233 (77.9%)
History of cardiovascular diseases	91 (30.4%)
Diabetes	41 (13.7%)
Hypertension	73 (24.4%)
Dyslipidemia	33 (11.0%)
Tobacco	40 (13.4%)
Alcohol consumption	16 (5.4%)
Substance use	8 (2.7%)
Medical or surgical history	
Chronic venous insufficiency	32 (10.7%)
Hematological malignancies	26 (8.7%)
Other dermatological diseases	21 (7.0%)
Solid tumors	14 (4.7%)
Infectious diseases	11 (3.7%)
Autoimmune diseases	11 (3.7%)
Rheumatological diseases	10 (3.3%)
Lupus	10 (3.3%)
Inflammatory bowel diseases	9 (3.0%)
Vascular thrombosis	9 (3.0%)
Renal diseases	8 (2.7%)
Respiratory diseases	8 (2.7%)
Psychiatric diseases	7 (2.3%)
Vasculitis	6 (2.0%)
Pancreatitis	5 (1.7%)
Sarcoidosis	4 (1.3%)
Others	30 (10.03%)
Recent Drug exposure (3 months)	16 (5.4%)
Recent vaccination (3months)	0(0%)

Table 1- Patients' sociodemographic characteristics and past medical history

Variable	Cla/Mod (%)	Mod/Cla (%)	Global (%)	p-value	v.test
Absence of cardiovascular history	71,63	100,00	69,57	<0,001	12,66
Absence of hypertension	65,93	100,00	75,59	<0,001	10,90
Absence of erythematous infiltrated plaque	64,62	91,95	70,90	<0,001	8,24
Absence of diabetes	57,36	99,33	86,29	<0,001	7,10
Absence of dyslipidemia	56,02	100,00	88,96	<0,001	6,72
Age: 26-49 years	68,22	59,06	43,14	<0,001	5,55
Bilateral lesions	58,99	85,91	72,58	<0,001	5,19
More than 5 episodes	78,43	26,85	17,06	<0,001	4,54
Absence of edema	55,74	91,28	81,61	<0,001	4,34
Treatment with colchicine	75,51	24,83	16,39	<0,001	3,95
Absence of vascular involvement on histology	54,58	91,95	83,95	<0,001	3,77
Intact dermis on histology	56,46	79,19	69,90	<0,001	3,48
Skin erythema	55,66	82,55	73,91	<0,001	3,38
Negative deep tissue culture	51,76	98,66	94,98	<0,001	2,92
Absence of anti-infectious treatment	53,97	86,58	79,93	<0,001	2,85
Multiple nodules: > 10 nodules	57,75	55,03	47,49	0,01	2,59
Absence of chronic venous insufficiency	52,43	93,96	89,30	0,01	2,59
Absence of topical steroids treatment	52,24	93,96	89,63	0,02	2,43
Presence of associated systemic symptoms	69,70	15,44	11,04	0,02	2,40
Presence of digestive symptoms	73,91	11,41	7,69	0,02	2,38
Age: 18-25 years	75,00	10,07	6,69	0,02	2,30
Multiple nodules: 5-10 nodules	64,15	22,82	17,73	0,02	2,28
Presence of fever	63,64	23,49	18,39	0,02	2,25
Treatment with hydroxychloroquine	68,97	13,42	9,70	0,03	2,15
Presence of arthralgia	61,54	26,85	21,74	0,03	2,12

Table 2 – Cluster one: list of over-represented variables

Variable	Cla/Mod(%)	Mod/Cla (%)	Global (%)	p- value	v.test
Presence of erythematous infiltrated plaque	62,07	87,10	29,10	<0,001	10,92
Unilateral lesions	52,44	69,35	27,42	<0,001	7,86
Absence of nodules	73,68	45,16	12,71	<0,001	7,62
Presence of edema	61,82	54,84	18,39	<0,001	7,56
Absence of relapse	28,30	96,77	70,90	<0,001	5,64
Absence of cardiovascular history	27,88	93,55	69,57	<0,001	5,01
Presence of vascular involvement on histology	50,00	38,71	16,05	<0,001	4,98
Absence of hypertension	26,55	96,77	75,59	<0,001	4,88
Lesions exceeding lower limbs	26,73	87,10	67,56	<0,001	3,86
Ant infectious treatment	40,00	38,71	20,07	<0,001	3,85
Presence of dermis alteration on histology	34,44	50,00	30,10	<0,001	3,69
Absence of dyslipidemia	23,31	100,00	88,96	<0,001	3,63
Absence of colchicine treatment	24,00	96,77	83,61	<0,001	3,45
Presence of pathogen on deep culture	60,00	14,52	5,02	<0,001	3,34
Mixed panniculitis on histology	27,33	70,97	53,85	<0,001	3,05
Absence of diabetes	23,26	96,77	86,29	<0,001	2,92
Treatment with topical steroids	41,94	20,97	10,37	<0,001	2,82
Pain	24,77	85,48	71,57	<0,001	2,81
Absence of skin erythema	32,05	40,32	26,09	0,01	2,76
Treatment installed	24,54	85,48	72,24	0,01	2,69
Presence of triggering factor	36,59	24,19	13,71	0,01	2,52
Absence of associated systemic symptoms	22,56	96,77	88,96	0,02	2,33
Absence of digestive symptoms	22,10	98,39	92,31	0,03	2,14
Absence of general status alteration	23,28	87,10	77,59	0,04	2,06
Presence of inflammatory infiltrate in the deep dermis and hypodermis	22,02	98,39	92,64	0,04	2,05
Absence of hydroxychloroquine treatment	22,22	96,77	90,30	0,04	2,02

Table 3 – Cluster two: list of over-represented variables

Variable	Cla/Mod(%)	Mod/Cla(%)	Global(%)	p- value	v.test
Presence of cardiovascular history	95,60	98,86	30,43	<0,001	17,60
Presence of hypertension	97,26	80,68	24,41	<0,001	14,79
Presence of dyslipidemia	100,00	37,50	11,04	<0,001	9,27
Presence of diabetes	92,68	43,18	13,71	<0,001	9,20
Age: 50 – 75 years	44,19	64,77	43,14	<0,001	4,84
Age > 75 years	76,19	18,18	7,02	<0,001	4,52
Absence of edema	32,79	90,91	81,61	0,01	2,77
Absence of inflammatory infiltrate in the deep dermis and hypodermis	54,55	13,64	7,36	0,01	2,52
Lesions limited to lower limbs	39,18	43,18	32,44	0,01	2,51
Absence of pain	40,00	38,64	28,43	0,01	2,47
Absence of fever	32,38	89,77	81,61	0,02	2,41
Male sex	40,91	30,68	22,07	0,02	2,25
Absence of treatment installed	38,55	36,36	27,76	0,04	2,10

Table 4 – Cluster three: list of over-represented variables

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Abstract N°: 572

"Cubital gust of wind" appearance is not synonymous of rheumatoid arthritis

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Introduction & Objectives:

Jaccoud's hand is a rare deforming arthropathy, first described in 1869 by Sigismond Jaccoud. Joint deformities are usually reversible, chronic, and non-erosive. Several aetiologies have been suggested.

Materials & Methods:

We report a case of Jaccoud's hand in a patient with systemic lupus erythematosus (SLE).

Results:

A 34-year-old female patient with 20 years' follow-up for SLE with cutaneous, articular, haematological, and renal involvement, was referred to our department for trophic disorders of hands and feet. Physical examination revealed malar erythema, erosive cheilitis, ulcero-necrotic lesions on hands and feet, and "Cubital gust of wind" appearance of both hands. Rheumatological examination revealed swan-neck deformities affecting the second, third, and fourth fingers of both hands, Z-shaped deformities of the thumbs and ulnar deviation of the fifth fingers, which returned to a normal position with a passive maneuver. These reducible deformities were compatible with Jaccoud's arthropathy. The history revealed no Raynaud's phenomenon or pain in the metacarpophalangeal and interphalangeal joints. These changes developed progressively, despite treatment with systemic corticosteroids and synthetic antimalarials.

Conclusion:

The prevalence of Jaccoud's hand in SLE is around 5%. Its non-erosive character distinguishes it from rheumatoid arthritis. Joint deformities result mainly from soft-tissue abnormalities, such as ligament laxity, capsule fibrosis, and muscle weakness. Management relies mainly on muscle-strengthening physiotherapy and the use of orthotics. Orthopedic soft-tissue surgery is also an alternative in cases of significant daily impact.

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Abstract N°: 612

Association of acanthosis nigricans with hypothyroidism - a cross sectional study

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Introduction & Objectives:

Acanthosis nigricans (AN) is often associated with obesity, endocrinopathies and malignancy. However, this has not been studied well and there are only few case reports of AN associated with hypothyroidism. Therefore, we carried out this cross-sectional study with comparative group to find out if there is any association between AN and hypothyroidism.

Materials & Methods: In this cross-sectional study with comparative group, patients were divided into study and comparative group consisting of 81 patients each, after considering various inclusion and exclusion criteria. Study group consisted of cases with AN and comparative group consisted of age and sex matched normal subjects without AN. After detailed history taking and clinical examination to note the clinical features of AN, laboratory estimation of serum T3, T4 & TSH was done in both the groups. Association between the levels of each variable and the disease condition was tested for statistical significance using Pearson's Chi-square test and t-test. Univariate analysis was performed wherein odds associated with each variable in favour of disease were obtained. The statistical significance was tested at 5% level.

Results:

Abnormal T3 value was observed in 7 cases of AN and 1 comparative subject which was statistically significant (Table 1). Abnormal T4 values were seen in 10 cases and 2 comparative subjects, however, this difference was not statistically significant (Table 2). Abnormal TSH level was observed in 11 cases and 8 comparative subjects, which again, was statistically insignificant (Table 3). The mean T3, T4 and TSH levels in both the groups were not significantly different. Hypothyroidism (normal or decreased T4 with increased TSH) was observed in 11 cases and 8 comparative subjects, which was statistically insignificant (Table 4). In AN cases, univariate analysis depicted greater risk of association of AN with hypothyroidism but again it was statistically insignificant.

Conclusion: In this cross-sectional study with comparative group, no association was found between AN and hypothyroidism. Further studies with a larger sample size is needed to firmly establish the true relationship between AN and hypothyroidism.

TABLE NO. 1

T3	Study group(cases)	Comparative group	Total	Statistic	P-value
	No. (%)	No. (%)	No. (%)		
Normal (60 - 200)	74 (91.4)	80 (98.7)	154 (95.1)	3.287*	0.070 (NS)
Abnormal (< 60)	7 (8.6)	1 (1.3)	8 (4.9)		
Abnormal (> 200)	0	0	0		
Total	81	81	162		
Mean ± SD	106.88 ± 39.82	113.05 ± 22.04		1.221**	0.224 (NS)

TABLE NO.2

T4	Study group (cases)	Comparative group	Total	Statistic	P-value
	No. (%)	No. (%)	No. (%)		
Normal (4.5 – 12)	70 (86.4)	77 (95.06)	147 (90.7)	2.497*	0.114 (NS)
Abnormal (< 4.5)	1 (1.2)	2 (2.47)	3 (1.85)		
Abnormal (> 12)	10 (12.3)	2 (2.47)	12 (7.4)		
Total	81	81	162		
Mean ± SD	11.20 ± 14.47	8.48 ± 2.04		-1.678**	0.095 (NS)

TABLE NO. 3

Thyroid	Study group (cases)	Comparative group	Total	Statistic	P-value
	No. (%)	No. (%)	No. (%)		
Normal (TSH 0.3 to 5.5)	70 (86.4)	73 (90.1)	143 (88.3)	0.5366*	0.4638 (NS)
Abnormal (TSH > 5.5)	11 (13.5)	8 (9.8)	19 (11.7)		
Abnormal (TSH< 0.3)	0	0	0		
Total	81	81	162		
Mean ± SD	4.76 ± 16.47	5.36 ± 17.03		3162**	0.693 (NS)
Mean ± SD [Log(TSH)]	0.96 ± 0.73	1.01 ± 0.77		-0.5285†	0.598 (NS)

TABLE NO. 4

Hypothyroidism	Study group (cases)	Comparative group	Total	c2 Statistic	P-value
	No. (%)	No. (%)	No. (%)		
Normal	70 (86.4)	73 (90.1)	143 (88.3)	1.3963	0.4975 (NS)
Clinical hypothyroid	1 (1.2)	2 (2.5)	3 (1.9)		
Subclinical hypothyroid	10 (12.3)	6 (7.4)	16 (9.8)		
Total	81	81	162		

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Abstract N°: 661

Extensive Secondary Cutaneous Involvement in Systemic Diffuse Large B Cell Lymphoma

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Introduction & Objectives:

Materials & Methods:

Results:

Skin involvement can be observed in both Hodgkin's and non-Hodgkin's lymphomas, either as a primary site or as secondary dissemination. Skin manifestations may be the presenting feature of the systemic disease, and a diagnosis may be confirmed on histopathologic examination of the cutaneous lesion. Secondary cutaneous lymphomas predominantly involve the head and neck. Here, we present a case of a unique, disseminated presentation of a secondary B cell lymphoma.

We present a case of a 48-year-old man with a history of diffuse large B cell lymphoma (DLBCL) stage IV-B, with bone marrow involvement. He was diagnosed six months prior and was treated with six cycles of R-CHOP, achieving a complete response. He presented to the dermatology clinic with nodular asymptomatic lesions on his scalp, face, and torso that appeared two weeks prior. Physical examination revealed multiple pink, firm nodules, ranging from 0.5 to 2 cm in diameter, on the scalp, ears, chest, and back. Bilateral inguinal lymphadenopathies were palpable. Histopathology examination of a skin lesion revealed a diffuse dense dermal infiltrate of atypical B-cell lymphocytes with an expression of CD20, BCL-2, BCL-6 (partially) and negative for CD10, CD23, cyclin D1, CD3 and CD5. Ki-67 staining showed high proliferative activity (>80%). A diagnosis of secondary cutaneous DLBCL was made. He was proposed to start treatment with R-ICE. Following two cycles, the skin infiltrates resolved.

DLBCL is the most common lymphoma and accounts for about 25% to 30% of all non-Hodgkin's lymphomas. B-cell lymphoma skin involvement manifests heterogeneously, ranging from few nodular lesions to extensive, fungating, or ulcerated masses. They are typically relatively fast growing, painless and non- pruritic. Secondary cutaneous involvement in DLBCL is infrequent, reported in up to 20% of cases in some series. Compared to primary cutaneous DLBCL, secondary cutaneous involvement in systemic DLBCL often presents with more extensive lesions, advanced stages, and poorer survival outcomes.

In conclusion, we report a unique case of disseminated secondary cutaneous DLBCL, highlighting the importance of timely diagnosis and management in such presentations.

Conclusion:

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Abstract N°: 712

Collateral perforating nodules

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Introduction & Objectives:

Rheumatoid nodulosis is a rare variant of rheumatoid arthritis (RA), characterized by the rapid onset of rheumatoid nodules, often accompanied by oligosymptomatic arthritis and elevated rheumatoid factor titers. Here, we present a case study of a 70-year-old woman with a 20-year history of RA, who developed an unusual presentation of accelerated perforating rheumatoid nodulosis while on methotrexate (MTX) therapy.

Materials & Methods:

A comprehensive examination of the patient's medical history, clinical presentation, and pharmacological treatment was conducted. Diagnostic procedures included culture tests, skin biopsy, and histological examination of the lesion. Additionally, PCR investigations for common pathogens were performed.

Results:

The patient presented with a persistent erythematous-infiltrative plaque on the left medial malleolar region, exhibiting characteristics consistent with accelerated perforating rheumatoid nodulosis. Histological examination revealed palisade granulomas perforating the epidermis with transepidermal elimination of necrobiotic material. Culture tests and PCR investigations were negative for infectious agents.

Conclusion:

This case highlights the importance of recognizing and distinguishing accelerated perforating rheumatoid nodulosis from other dermatological conditions, particularly in patients undergoing immunosuppressive therapy. The perforating variety of the rheumatoid nodule, although rare, has been studied in the literature but, in the context of an accelerated rheumatoid nodulosis, to the best of our knowledge, has never been described.

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Abstract N°: 730

Case series of cutaneous sarcoidosis

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Introduction & Objectives: Sarcoidosis is a chronic inflammatory disorder of obscure etiology with multiple organ involvement. Skin involvement is seen in 20-35 % of the cases and is cited as the second most commonly involved organ. We present a series of 8 biopsy proven cases of sarcoidosis with different types of clinical lesions.

Materials & Methods: We present a series of 8 cases of Cutaneous Sarcoidosis, diagnosed over a period of 2012 to 2022 in Dermatology OPD in Bangalore, India. Diagnosis was suspected based on the clinical morphology and diagnosis was based on histopathology & Serum ACE Levels.

Results: Out of eight patients, females outnumbered males and were middle aged. Most of the patients presented with patches , plaques and nodules. Few of the cases presented with hypopigmented scaly patches, swellings on the nose, voilaceous lesions and ulcerated nodules . Systemic manifestations like breathlessness, dry cough and congestion of eyes were present in two of our patients. All our cases were showing histopathological features of sarcoidosis and other investigations were in favour of sarcoidosis. Patients were treated with hydroxychloroquine and one of them responded well to apremilast.

Conclusion: Sarcoidosis can present with various kinds of clinical manifestations, making diagnosis difficult sometimes. It is known as a “great imitator” in dermatology because lesions can exhibit many different morphologies. It is characterized by the formation of noncaseating epithelioid cell granulomas in several organs or tissues. There are many treatment options available but no one therapy is universally effective.

Our cases series highlights the importance of suspecting Sarcoidosis when there are asymptomatic erythematous papules, nodules and plaques. Our series showed different morphological forms of lesions. Systemic symptoms were seen in only 25% of our cases. Few. Differential diagnosis included mycosis fungoides, Hansen’s disease , lichen planus , discoid lupus erythematosus, sweet syndrome. Most of our patients responded well to hydroxychloroquine and showed complete resolution of symptoms following the treatment. Many cases of Sarcoidosis might be missed as diagnosis is not considered.

There was no conflict of interest.

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Abstract N°: 790

A rare case of erythema gyratum repens in ovarian cancer: a case report

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Introduction & Objectives:

Erythema gyratum repens (EGR) is a rare paraneoplastic dermatosis characterised by an annular erythematous "wood-grain" pattern. The distinctive rash is often associated with an underlying malignancy, most commonly reported in conjunction to lung cancer (1).

Results:

We report a 52-year-old Caucasian female who presented with a three-year history of an atypical rash characterised by recurrent flares of red, pruritic, tender rash overlying the chest and face. On examination, she had a well-demarcated, erythematous patch over her chest with prominent advancing edge and more confluent erythema to face. Patch test and autoimmune screen were negative. At the time of presentation, she had marginally raised CA125 levels although an ultrasound scan of the abdomen and pelvis were unremarkable. Histology revealed patchy zones of parakeratosis, with mild degree of spongiosis and underlying perivascular infiltrate. After extensive review, including in multidisciplinary team, the patient was treated as for eczema with intermittent oral prednisolone for flare ups, phototherapy (UVB), ciclosporin, methotrexate, azathioprine, mycophenolate, and dupilamab without any improvement.

Two years following her initial presentation, she was diagnosed with ovarian cancer with advanced peritoneal involvement. She underwent a total hysterectomy, bilateral salpingo-oophorectomy and omentectomy and treated with carboplatin and taxol with subsequent Olaparib (PARP [poly ADP ribose polymerase] inhibitor). The patient experienced a good partial response to this treatment after 2 months with improved ascites and omental disease which has continued to date. The patient's rash cleared within weeks of chemotherapy and has remained essentially clear over a 12-month period. Review of histology based on additional clinical history and dramatic response to cancer therapy was felt compatible with EGR.

Conclusion:

Textbook description of EGR reports wood-grain widespread rash and perhaps reflects publication bias of classical cases but does not cover the range of presentations of paraneoplastic dermatoses. To the best of our knowledge, our report is the first documented case of EGR as a paraneoplastic eruption of ovarian carcinoma. EGR histology is often non-specific, therefore it is crucial that suspected EGR must be diagnosed in correlation with clinical examination. It is reported that the time between EGR eruption and cancer diagnoses to range from 1 to 72 months, averaging at 9 months (2). This case emphasises the importance of a high index of clinical suspicion to appropriately investigate patients with atypical rashes supportive of a paraneoplastic condition.

References:

\1. Votquenne N, Richert B. Erythema Gyratum Repens. JAMA Dermatology. 2020 June 17;156(8):912.

\2. Boyd AS, Neldner KH, Menter A. Erythema gyratum repens: A paraneoplastic eruption. Journal of the American Academy of Dermatology. 1992 May;26(5):757-62.

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Abstract N°: 798

Nrf2 regulation of the PINK1/Parkin axis mediates keratinocyte abnormalities and intervention in vitiligo

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Introduction & Objectives: This study aims to investigate the effects of Nrf2 regulation of the PINK1/Parkin axis on vitiligo keratinocytes.

Materials & Methods: Immunofluorescence was employed to detect the expression of Nrf2, PINK1, Parkin, and LC3 proteins in depigmented skin tissues of vitiligo patients. The effect of Nrf2 on the promoter activity of the PINK1 gene was analyzed using a dual luciferase gene system. Cell apoptosis was detected using DAPI staining after different treatments, and Western blotting was performed to investigate the expression of mitophagy related proteins. The ROS level was analyzed using the DCFH-DA assay. Mitochondrial morphology was observed using a transmission electron microscope. Human melanocytes were incubated with HaCaT supernatant, and melanocyte apoptosis was detected using Hoechst staining. The effects of keratinocyte-specific high-expression of Nfe2l2, Pink1, and Prkn, as well as the knockdown of Nfe2l2 and/or Lipo (MitoQ), were analyzed in mice with H2O2-induced effects of pigment loss.

Results: The levels of Nrf2, PINK1, Parkin, and LC3 were found to be down-regulated in the skin lesion tissues of vitiligo patients compared to healthy controls. Nrf2 was found to regulate the promoter activity of the PINK1 gene. H2O2 down-regulated Nrf2 expression in HaCaT cells and promoted ROS production. MitoQ alleviated H2O2-induced abnormal mitochondrial morphology, mitophagy, and cell apoptosis through PINK1 and Nrf2. The supernatant of NFE2L2 knockdown HaCaT cells promoted melanocyte apoptosis, while overexpressing PINK1 and PRKN partially increased melanocyte viability. Overexpressing NFE2L2 and PINK1 in keratinocytes, as well as treatment with Lipo (MitoQ), increased the skin thickness and the number of follicles in mice. This also up-regulated the levels of Nrf2, PINK1, Parkin, and Tyrosinase, while down-regulating the level of Caspase3.

Conclusion: These findings indicate that Nrf2 regulates the PINK1/Parkin axis, which in turn affects mitochondrial membrane potential, morphology, mitophagy-related protein expression, and cell apoptosis. MitoQ promotes mitophagy, reduces cell apoptosis, and protects keratinocytes from oxidative damage via the Nrf2/PINK1 axis.

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Abstract N°: 801

Protective effects of liposome-encapsulated MitoQ against oxidative damage in HaCaT cells and depigmentation in mice

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Introduction & Objectives: This study aims to investigate the protective effect of liposome-encapsulated MitoQ against oxidative damage in HaCaT cells and depigmentation in mice.**

Materials & Methods: Lipo(MitoQ) was prepared using the thin film dispersion method. Particle size, polydispersity coefficient, and morphology were assessed using a particle size and surface potential detector, as well as transmission electron microscopy. Nanoliposomes were separated by ultrafiltration centrifugation, and the drug content was determined by UV-vis spectrophotometer to assess the release characteristics of Lipo(MitoQ). HaCaT cells were cultured in high glucose DMEM. The cytotoxicity of empty liposomes was assessed using a CCK-8 assay. Cell proliferation was detected through Ki67 immunofluorescent staining, while ELISA was performed to detect bFGF and SCF cytokines. ROS production was determined using DCFDA fluorescent staining, and mitochondrial membrane potential and apoptosis were detected using Mito-Tracker Red CMXRo staining. Nrf2 nuclear translocation was detected through immunofluorescence staining, and the mRNA expression of HO-1 and NQO1 genes was determined using qRT-PCR. The expression of antioxidant proteins Nrf2, Pink1, Parkin, and LC3 was detected using Western blotting. Cellular autophagy was observed using RFP-GFP-LC3 immunofluorescence staining, and mitochondrial morphology was observed using transmission electron microscopy. The effects of Lipo(MitoQ) on follicle growth, Tyrosinase levels, and melanogenesis were further detected in depigmented mice induced by H2O2.

Results: Cellular experiments were conducted on H2O2 treated HaCaT cells to confirm the effectiveness of Lipo(MitoQ) in extending the release time up to 48 hours. Lipo(MitoQ) not only promoted cell proliferation and increased secretion of SCF and bFGF but also enhanced Nrf2 nuclear translocation and HO-1 expression, improved mitochondrial activity, reduced apoptosis, and demonstrated a more significant protective effect against oxidative stress injury in HaCaT cells compared to MitoQ. Additionally, the efficacy of Lipo(MitoQ) was further validated in depigmented mice induced by H2O2. It was found that Lipo(MitoQ) promoted hair follicle growth, increased Tyrosinase levels, and stimulated melanogenesis.

Conclusion: These findings suggest that Lipo(MitoQ) has the potential to resist oxidative damage in HaCaT cells and depigmentation in mice, thereby offering a new strategy for the treatment of vitiligo.

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Abstract N°: 877

Menopause and the skin: prevalence of skin symptoms in women attending a menopause clinic

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Introduction & Objectives:

The peri and post-menopausal period of a woman's life is driven by declining ovarian function leading to a hypoestrogenic state. The skin contains oestrogen receptors and impairment of the skin barrier is noted in oestrogen deficient skin.1 It is therefore expected that this may affect the normal function of the skin and common skin conditions. We suspect that these symptoms experienced may be underreported and, when they are reported, may not be considered associated to the menopause.

Materials & Methods:

We conducted an observational study exploring the symptoms women experience during the peri-menopausal and post-menopausal time. Patients seen in a community-based specialist menopause clinic were invited to complete a questionnaire.

Results:

32 responses were collected. 77% of respondents were on hormonal treatments, of which oestrogen therapy was most frequently taken. The most reported skin symptoms included dry skin (78%) and itchiness (75%). Dry mouth was the commonest reported oral symptom and 84% of women experienced at least one hair symptom since the onset of their menopause. 91% also reported experiencing at least one vulval symptom. 50% noticed worsening in a previously diagnosed dermatological disorder and 63% of patients reported skin symptoms that they had attempted to manage themselves without consulting their doctor. Dermatology Life Quality Index (DLQI) was also collected for each patient, which showed an average score of 6 and ranged up to 17, indicating a moderate impact on the quality of life of peri and post-menopausal women.

Conclusion:

Our results highlight the significant effect the menopause can have on quality of life and the wide range of dermatological symptoms associated with it. Patients appear reluctant to seek medical support for skin symptoms related to the menopause and consequently may not receive optimal treatment. It is paramount that dermatologists and menopause specialists are aware of the immense impact menopause has on the skin, hair, vulva and mouth. Further research is required to explore management options including hormone replacement therapy for skin symptoms associated with the menopause.

References:

\1. Rzepecki A, Murase J, Juran R et al. Estrogen-deficient skin: The role of topical therapy. Int J Womens Dermatol 2019; 5:85–90.

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Abstract N°: 882

Calciphylaxis in a Young Patient: Navigating Successful Treatment

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Calciphylaxis in a Young Patient: Navigating Successful Treatment

Introduction:

Calciphylaxisis a rare and serious disorder that presents with skin ischemia and necrosis and is characterized histologically by calcification of arterioles and capillaries in the dermis and subcutaneous adipose tissue. It most commonly occurs in patients who have end-stage kidney disease (ESKD) and are on dialysis. We describe a case of calciphylaxis in a young patient which, uncommonly, resulted in a substantially positive prognosis.

Case Report:

A 27-year-old female nursing student with a history of lupus nephritis presented with persistent painful lumps that had progressed to ulceration on her left medial thigh. Despite the tissue viability team's efforts with wound care, the condition prompted dermatological evaluation. Biopsies taken, while comprehensive, were inconclusive for lupus panniculitis, infection, or vasculitis, showing epidermal ulceration, neovascularization, and a mild inflammatory process. The patient's deteriorating condition, marked by excruciating pain exacerbated during dialysis, demanded an expanded diagnostic workup. Deep biopsy results showed calcification in small blood vessels, confirming calciphylaxis. This finding was key to diagnosing this rare condition, especially since the usual signs of calciphylaxis weren't present initially. Following the calciphylaxis diagnosis, the patient's treatment was adjusted significantly. Warfarin was replaced with apixaban, and the dialysis schedule was modified to manage phosphate levels effectively. Treatment with sodium thiosulfate after dialysis, cinacalcet for parathyroid control, targeted antibiotics for infection, and comprehensive pain management markedly improved her symptoms and ulcer healing.

Discussion:

The diagnosis of calciphylaxis, especially in patients lacking the typical presentation, presents a notable clinical challenge. This case highlights the necessity of considering calciphylaxis in the differential diagnosis for patients with chronic kidney disease on hemodialysis, even when hallmark symptoms are absent. The successful management of this case was significantly enhanced by a multidisciplinary approach and early intervention, underlining the importance of prompt action and collaborative care. Factors such as the patient's younger age and the absence of cardiovascular disease, alongside timely diagnosis, played a crucial role in the favorable outcome. Early recognition and comprehensive treatment can markedly improve prognosis in calciphylaxis cases, traditionally associated with high morbidity and mortality.

Conclusion:

This calciphylaxis case highlights the critical importance of early diagnosis and a multidisciplinary treatment strategy, leading to a notably positive outcome. It emphasizes that proactive management can significantly improve the prognosis for patients with complex conditions.

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Abstract N°: 1151

Delayed Toxic epidermal necrolysis like acute graft versus host disease in a patient with acute myeloid leukemia- A diagnosis dilemma.

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Introduction & Objectives: Graft-versus-host disease (GVHD) is a severe complication observed after allogeneic hematopoietic stem cell transplantation (1). The skin is the most affected organ, followed by oral mucosa, liver, eyes, and gastrointestinal tract (2). We describe an atypical case of cutaneous GVHD presenting as Toxic epidermal necrolysis (TEN) after discontinuation of immunosuppressants due to relapse of acute myeloid leukemia. Regarding other cases described in the literature where cutaneous manifestations occur with immunosuppressants at therapeutic doses, in our case, the attenuation of immunosuppression was clearly linked to the onset of skin involvement as GVHD.

Materials & Methods: A 43-year-old woman with a history of acute myeloid leukemia who received haploidentical stem cell transplantation 5 months prior the current presentation, was admitted to the hospital for respiratory symptoms. She exhibited leukocytosis, neutrophilia, and peripheral blood blasts on a complete blood count. Disease relapse was confirmed by bone marrow cytology, flow cytometry, and pathology. She was also diagnosed with pneumonia, so antibiotics were initiated. On day 7 of hospitalization, she developed painless erythematous and purpuric macules and papules, some exhibiting characteristic target lesions on the trunk, lower and upper extremities. There was progression of cutaneous involvement compromising 90% of total body surface area, with blistering and erosions in the oral mucosa, conjunctivae, palms and against host antigen after an allogeneic hematopoietic stem cell transplant and cause local tissue injury and further promotion of an inflammatory response (3). Acute GVHD can affect soles, and genitals with numerous necrotic and purpuric papules. GVHD vs TEN was considered as a differential diagnosis, with antibiotics as the causal medications suspected. Skin biopsy revealed interface dermatitis, most likely acute GVHD. According to the extension in the setting of clinical and laboratory findings severe acute GVHD (aGVHD) diagnosis was made. She received corticosteroids with refractory response, so ruxolitinib and cyclosporine were initiated with a significant improvement of the lesions on the skin and mucosa after three weeks of treatment.

Results: AGVHD is a donor T cell-mediated process in which donor T cells are activated in skin, liver or intestine and is graded based on the severity of symptoms and histologic manifestations into four stages (4). When Stage IV cutaneous aGVHD occurs, it can be difficult to distinguish clinically from TEN. Transplant patients are often on many medications, challenging to differentiate between the two conditions (5).

Conclusion: In this case, the presentation of painless skin lesions, the distribution, and the natural history of the exanthema, along with histological findings, favored the diagnosis of graft-versus-host disease.

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Abstract N°: 1238

Morphea in a patient with clear cell renal cancer and lung metastases

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Introduction & Objectives:

Morphea (localized scleroderma) is an autoimmune fibrotic skin disease with an estimated prevalence of 200 per 100000. Though the pathogenesis remains unknown, the disease is characterized by an initial T-cell-mediated inflammatory phase followed by localized fibrosis of the skin. Most morphea cases in patients with cancer, occur after radiation therapy 65%,chemotherapy 8,5%,immunotherapy-immune checkpoint inhibitors ICIs 5,7%. In 2/3 of morphea in patients with cancer, the mean time onset of morphea is 3,2 years after radiation, 1,7 years after chemotherapy, and 8 months (rapid manifestation of morphea) after ICIs immunotherapy.

Materials & Methods:

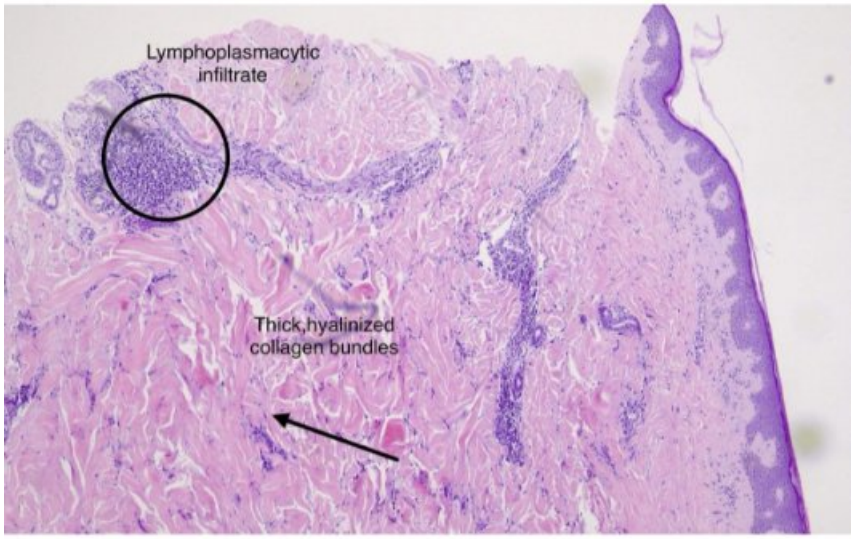
We present a case of a 65-year-old male patient with a history of Clear Cell Renal Carcinoma, who underwent surgical removal of his right kidney 5 years ago. Two years postoperatively, he developed a metastatic nodule in the right upper lobule of his lung. The patient was under treatment with pazopanib ,800 mg/day, for 15 months.Pazopanib is a tyrosine kinase inhibitor that blocks tumor growth and inhibits angiogenesis.The metastatic nodule disappeared but a new one appeared (CT and MRI). While the 2nd metastasis remained stable and the patient was not taking any treatment, he developed the typical skin lesions of morphea , with numerous sclerotic , or atrophic lesions with lilac ring, 2,5 years after the cessation of pasopanib (photo) The diagnosis was confirmed by biopsy (photo) and he is currently under topical treatment with cortisone and calcineurin inhibitors.

Results:

To date several studies have reported sudden development of morphea either before or after a brief interval following the diagnosis of malignancy. Scleroderma is characterized by immunological dysregulation, vasculopathy and hypeproduction of the extracellular matrix by activated fibroblasts. Endothelial inflammatory and mesenchymal cells produce cytokines,and growth factors (IL-1,IL-6) TNF-a connective tissue growth factor, and basic fibroblast growth factor.

Conclusion:

Morphea developed 5 years after the diagnosis of clear renal cell carcinoma and 2.5 years after the cessation of pazopanib - a tyrosine kinase inhibitor , so it should be considered rather a paraneoplasmatic phenomenon than a secondary effect of pazopanib cancer therapy.


Lymphoplasmacytic
infiltrate
Thick,hyalinized
collagen bundles

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Abstract N°: 1245

Sweet Syndrome and Wegener Granulomatosis Association: Case report

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Introduction & Objectives:

Sweet syndrome (SS) is a rare acute febrile neutrophilic dermatosis. Many clinical variations of SS have been described in the last decades, but the precise etiology remains unclear [Vitale a., et al. 2022]. We present a rare clinical case of idiopathic SS with Wegener's granulomatosis (WG). It is known that approximately 10 % WG cases are ANCA negative [McCarthy E, et al. 2017].

Materials & Methods:

A case report.

Results:

A 21-year-old female refered with painful disseminated lesions after treatment of the recurrent febrile respiratory infection with azithromycin. Examination revealed multiple erosions and purulent nodules covered with yellowish crusts on the back and limbs – BSA 18%. In peripheral blood tests elevated neutrophils count and increased erythrocyte sedimentation rate were found. Culture from lesions for bacteriae was negative. Histological skin examination showed dermal oedema with diffuse predominating infiltration of neutrophils together with eosinophils and lymphocytes. SS was confirmed based on 2 major (painful nodules on the skin, in histology neutrophilic dermal infiltrate without vasculitis) and 3 minor criteria (preceded by a nonspecific respiratory tract infection, fever, abnormal laboratory values). Chest CT scan revealed stenosis in the left bronchus. Bronchoscopy demonstrated haemorrhagic endobronchial lesions with granulation and ulceration. Nasal and bronchial biopsies showing necrotising inflammation with fibrin, also vasculitis signs confirmed WG, although ANCA was negative. After oral therapy with methylprednisolone 32 mg/d, Cushing's syndrome appeared. It has been switched to rituximab 1000 mg every 2 weeks (total two cycles). The patient's symptoms improved. The left bronchus stenosis got less in the follow up CT scan of the chest. Skin examination showed no erosions, mainly post-inflammatory hyperpigmentation.

Conclusion:

There are only a few clinical cases reporting the co-occurrence of SS and WG. The treatment of both conditions remains a challenge. Systemic steroids stand out as the most effective treatment for SS. However, forms of neutrophilic dermatoses that are unresponsive and refractory to traditional therapies, especially those associated with autoimmune diseases, could be treated with rituximab.

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Abstract N°: 1260

A rare case of cutaneous siderosis after iron infusion

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Introduction & Objectives:

A 28-year-old male presented to the Dermatology clinic with a pigmented patch over his right arm. A few months prior, he had been hospitalised for iron deficiency anemia and gastrointestinal bleeding. In view of his low hemoglobin levels of 4.8 g/dL, he was administered an intravenous infusion of ferric carboxymaltose via a cannula on his right forearm. After his discharge from the hospital, he noted the gradual development of pigmentation on his right arm which remained persistent.

Materials & Methods:

On examination, there was a 30 by 10 cm grey to hyperpigmented patch over his right forearm extending to his upper arm.

Results:

He was diagnosed with cutaneous siderosis following intravenous iron infusion and offered a trial of laser treatment.

Conclusion:

Ferric carboxymaltose is an intravenous iron supplement used in the treatment of iron deficiency anemia. Rare cases of skin staining due to extravasation of iron into the subcutaneous tissue have been described, with a reported incidence of 0.1% to 6%. Patients typically present with an asymptomatic black, brown, purple, or grey discolouration over the affected area. The onset and persistence of symptoms are variable, and the skin pigmentation may potentially be permanent.

There are no published trials to guide the treatment of cutaneous siderosis secondary to iron extravasation. Topical treatments, massages and lymphatic drainage have yielded minimal benefit. Treatment with Q-switched or Nd:YAG lasers have been reported with varying success.

Measures that may be taken to minimise iron extravasation include checking the integrity of the cannula, flushing the cannula with sodium chloride before and after the infusion and patient education to highlight any pain or discomfort during the infusion.

Although skin staining after intravenous iron infusion is rare, it can cause psychological distress to patients and is extremely difficult to treat. It is thus important for physicians to be aware of and to educate patients about this potential complication.

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Abstract N°: 1313

Acute cutaneous lupus erythematosus induced by terbinafine

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Introduction & Objectives:

The cutaneous involvement in acute lupus erythematosus is characterized by erythema, which may be edematous or scaly. It is most localized to the face, creating the classic "butterfly rash." In diffuse forms, it predominates in sun-exposed areas with the possibility of blistering. Mucosal lesions result in often painful oral erosions. Medications can sometimes induce or exacerbate cutaneous involvement.

Materials & Methods:

We report a case of acute cutaneous lupus erythematosus (ACLE) induced by terbinafine in a lupus patient.

Results:

The patient is a 26-year-old female with a history of "3A" syndrome and systemic lupus erythematosus with renal involvement in remission under hydroxychloroquine, prednisone, and azathioprine. One month after initiating oral terbinafine (for extensive dermatophytosis and onychomycosis), she developed a febrile and pruritic eruption characterized by a malar rash, generalized erythema with targetoid lesions, and cutaneous detachment. Erosive cheilitis and multiple oral ulcers were also present. Laboratory investigations revealed hepatic cytolysis. Histopathological examination showed vacuolar and spongiotic dermatitis, epidermal apoptotic bodies, and a lymphocytic inflammatory infiltrate in the dermis, without eosinophils. Direct immunofluorescence demonstrated granular deposits of IgG and C3 at the dermoepidermal junction. Pharmacovigilance investigation established the causality of terbinafine. Therefore, the diagnosis of ACLE induced by terbinafine was established. Terbinafine discontinuation and increasing prednisone doses resulted in favorable outcomes.

Conclusion:

Several cases of subacute lupus erythematosus induced or exacerbated by terbinafine have been reported. In our patient, it was an isolated cutaneous involvement of acute lupus erythematosus. Histological examination combined with direct immunofluorescence helped exclude the main differential diagnosis: toxic epidermal necrolysis, although the distinction can be challenging. The outcome is generally favorable upon discontinuation of the medication, but recurrence of cutaneous lesions upon terbinafine reintroduction is possible. Therefore, terbinafine should be prescribed cautiously in patients with systemic lupus erythematosus, and azole derivatives can be considered as an alternative in documented fungal infection.

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Abstract N°: 1420

Predictive Factors for Cardiovascular Disease in Dermatomyositis: A Comprehensive Exploration

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Introduction & Objectives:

Dermatomyositis (DM), a rare autoimmune inflammatory myopathy, has been associated with various systemic complications, including cardiovascular diseases (CVDs) such as dilated cardiomyopathy (DCM) and/or congestive heart failure (CHF)1. This study investigates predictive factors contributing to the development of DCM and/or CHF in DM patients.

Materials & Methods:

A retrospective review of records for patients with adult-onset DM was executed at two tertiary care centers in Ontario, Canada, between January 2010-September 2023. The study comprised 114 patients in total, with a median age of 58 years (range= 21-99), and 80% female participants. 16 individuals (14%) were diagnosed with DCM and/or CHF. Statistical analyses showed that the presence of Raynaud's phenomenon, increased disease duration, the presence of anti-Ro 52 antibodies, elevated ESR, and elevated LDH were all predictive factors for the development of DCM/CHF in DM patients.

Results:

In our study, elevated ESR, a marker of systemic inflammation, and elevated LDH, indicative of tissue damage, emerged as significant predictors of DCM/CHF in DM patients.2,3 While current literature has not explored the links between the elevation of these biomarkers and the development of CVD in DM patients exclusively, ESR has generally been recognized as a marker for CVD development in prior studies.4 Additionally, in a 2021 study, LDH was associated with an increased 10-year CVD risk and an independent indicator of CVD.3 These findings raise the question of whether the heightened risk of DCM and/or CHF in DM patients is due to direct myocardial involvement or connected to chronic systemic inflammation.

A study encompassing patients with idiopathic inflammatory myopathies, including DM, revealed that patients with positive anti-Ro52 antibodies exhibited an elevated risk of CVD, including DCM/CHF, aligning with our own findings.5 Contrary to our results, the previous study indicated that individuals without Raynaud's phenomenon were more prone to developing DCM/CHF.5 However, one broad epidemiological study found that Raynaud's phenomenon was associated with CVD (odds ratio 3.51), findings which are more in line with our own .6 Previous literature has revealed that Raynaud's phenomenon is detectable in approximately one-third of all patients with DM.6 As a result of the significant presence of this cutaneous manifestation in individuals with DM, as well as the conflicting relationship between this phenomenon and CVD, further research is warranted.6

Our findings suggest that an extended disease duration may increase susceptibility to CVD in DM patients. However, it is important to note a contrasting 2017 study which found no association between DM disease duration and risk of CVD.7 Despite these discrepancies, literature consistently underscores the critical importance of long-term vigilance and proactive management in DM, given the significant percentage of DM mortality (15-55%) attributed to CVD.8

Conclusion:

In conclusion, this study significantly contributes to our understanding of predictive factors for CVDs in DM patients. The highlighted associations provide valuable insights for clinicians in risk assessment and early intervention planning. Future research may further elucidate the underlying mechanisms and refine our understanding of predictive factors.

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Abstract N°: 1424

Predictive Factors for Interstitial Lung Disease in Dermatomyositis

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Introduction & Objectives:

Dermatomyositis (DM) is a complex systemic autoimmune disease with distinctive cutaneous manifestations. Interstitial lung disease (ILD) in DM is associated with increased morbidity and mortality and poses a significant challenge in the management of DM patients, warranting a comprehensive exploration of predictive factors to enhance early detection and intervention. This study aims to identify key determinants associated with the development of ILD in patients with adult-onset DM.

Materials & Methods:

A retrospective analysis of patient records was conducted at two tertiary care centers and affiliated clinics in Ontario, Canada. The study included a total of 114 patients (80% female, mean age of 59± (SD)17.62 years). A total of 28 patients (25%) were diagnosed with ILD after the onset of DM. Categorical variables were assessed using Chi-square hypothesis tests, while quantitative variables underwent t-tests to compare between individuals with and without ILD. The presence of Raynaud's phenomenon, anti-Ro 52, anti-MDA5 antibodies, anti-Jo1 antibodies, and elevated serum LDH levels were significantly associated with increased risk of developing ILD in DM patients. Age at DM onset, gender, and disease duration were not associated with higher risk of ILD development (Table 1).

Results:

Our study reinforces existing evidence that associates certain MSAs namely anti-MDA5, anti-Jo1, and anti-Ro 52 with development of ILD.1 Previous studies have shown that the anti-Jo-1 antibody is associated with the development of ILD, specifically chronic rather than rapidly progressing ILD.1 Additionally, in a cohort of anti-MDA5 positive DM patients, those who also tested positively for anti-Ro 52 antibodies had a higher likelihood of ILD development, particularly rapidly progressive ILD, as well as a decreased 24-month survival rate.2 Therefore, beyond their diagnostic value, these antibodies serve as prognostic indicators, assisting clinicians in risk assessment and early intervention planning.

In our study, elevated LDH levels emerged as another pivotal predictor of ILD. A retrospective analysis of DM patients with ILD revealed 86.49% had LDH levels above normal.3 This aligns with findings in other studies associating elevated LDH with various inflammatory pulmonary diseases, including idiopathic pulmonary fibrosis.4 As a marker of tissue damage, inflammation, and plausibly the pathological processes associated with ILD, studies suggest elevated LDH is a significant independent predictive factor for the development of ILD in DM patients.3,4 Incorporating LDH into routine assessments can enhance risk stratification, providing clinicians with a comprehensive understanding of disease progression.

Nevertheless, not all our observations aligned with the existing body of literature. While prior studies suggested that the prevalence of Raynaud's phenomenon remains comparable among DM patients with and without ILD, our results revealed a significant increase in the occurrence of Raynaud's phenomenon among DM patients with ILD.5

Conclusion:

In conclusion, our study underscores the importance of specific clinical manifestations, certain MSAs, and LDH levels as predictive factors for ILD in DM. Early identification of at-risk individuals allows for proactive monitoring, early intervention, and a personalized approach for effective DM management.

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Abstract N°: 1427

Blueberry Muffin Presentation in Congenital Langerhans Cell Histiocytosis

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Introduction & Objectives:

"Blueberry muffin baby" is a distinctive neonatal condition characterized by generalized bluish-red, macular, papular, or nodular lesions that resemble blueberries in a muffin. The rash can be caused by multiple factors, such as the rubella virus, cytomegalovirus, herpes simplex virus, syphilis, dermal hematopoiesis, or various neoplasms. Langerhans cell histiocytosis (LCH) is a disease that results from the abnormal proliferation of S-100- and CD1a-positive cells with Birbeck granules. It can present as a single-system disease or as a multisystem disease involving the skin, lungs, liver, lymph nodes, bones, and hematopoietic system. We report two cases of congenital LCH to clarify the significance of the blueberry muffin appearance in diagnosing LCH.

Case Description:

Case 1 involved a 0-day-old male preterm infant born at 36 weeks' gestation who presented with scattered, well-demarcated, bluish to dark erythematous macules of up to 20 mm with mild scaling involving the scalp, neck, trunk, and extremities. Case 2 involved a 0-day-old full-term male infant who presented with generalized eroded dark erythematous lesions of up to 10 mm in diameter. There were no significant findings in the family history, and the pregnancy had progressed smoothly in both cases. Screening of the blueberry muffin baby, including a search for viral and syphilis infection and leukemia, was negative. Dense infiltrates of CD1a-positive Langerhans cells in the upper dermis were observed in the skin biopsies of both patients. Skeletal examination, chest x-ray, abdominal ultrasound, and blood and urine tests showed no abnormal findings in other organs.

Results:

Both cases were diagnosed as congenital LCH and were closely followed up, with the complete resolution of skin lesions noted at day 10 in case 1 and at day 30 in case 2. In case 1, new lesions were found in the lungs and thymus on day 49. Case 2 has shown no relapse or systemic involvement in 11 months of follow-up.

Discussion & Conclusion:

LCH is now recognized as a clinical spectrum ranging from single-system to extensive multisystem disease. Spontaneous resolution sometimes occurs in limited cutaneous involvement at birth; however, as observed in case 1, multisystem involvement may occur at a later stage, requiring prolonged and careful follow-up with imaging studies. The cutaneous findings of LCH are variable, but may include multiple reddish-brown or violaceous papules and nodules, which may be crusted or eroded. These eruptions require differentiation from congenital infections and other neoplasms; however, the blueberry muffin appearance has not received much attention as an initial indicator of LCH. The early detection of congenital LCH may spare children from inappropriate examination and treatment, and the blueberry muffin presentation should be recognized as a key feature.

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Abstract N°: 1486

Mycosis fungoides with an unusual clinicopathological presentation

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Introduction & Objectives:

In primary cutaneous lymphomas (CLs), lymphatic proliferation was primarily confined to the skin which has T-lymphocyte (65%), B-lymphocyte (25%) or NK cells as origins 1 . Among cutaneous T-cell lymphomas (CTCL), mycosis fungoides (MF) represented the most common subtype 2 . A common MF manifestation, initially presented with erythematous scaling lesions in the sun-exposed areas of the skin, was the development of localized or generalized patches, plaques, tumors and erythroderma with a progressive and long-term course 3 . MF patients suffered from a troublesome pruritus, which affected the quality of their life.

Materials & Methods:

Multiple biopsy specimens were obtained from her skin lesions. Findings included hyperkeratosis, parakeratosis, acanthosis, spongiosis and vesicle formation. Further observed was the exocytosis of some atypical lymphocytes into the basal and lower spinous layer. Dermis showed dense patchy infiltrates of mature as well as atypical lymphocytes with irregular nuclear border intermingled with certain eosinophils.

Results:

biopsy showed hyperkeratosis, parakeratosis, acanthosis with multifocal epidermotropism of atypical lymphocytes with enlarged hyperchromatic nuclei and irregular border mainly in the basal layer. Dermis had patchy dense infiltrates of essentially lymphoid cells with enlarged irregular nuclei admixed with some eosinophils. Atypical lymphocytes permeated and destructed sweat glands (Fig. 3,4). Immunohistochemical study was conducted on paraffin-imbedded tissues with antibodies of CD43, CD3, CD4, CD5, CD8, CD7, CD20, CD30, PAX5 and ki67. The majority of neoplastic cells in the epidermis and dermis expressed CD3, CD4 and CD5 (Fig. 7,8). On the other hand, there was a minimal expression of PAX5 and CD8 in the dermis. The neoplastic cells were negative for CD20, CD30, and CD7 (Fig. 8,10), while Ki67 was positive in more than 50% of the lymphoid population. A diagnosis of MF (Stage 2B) with eosinophilia was confirmed for the patient.

Conclusion:

Given the non-specific presentation of MF, particularly at patch/plaque stage, early clinical diagnosis becomes challenging and certain patients are, for years, treated for other differential diagnoses. A typical feature of MF is epidermotropism of small- to medium-sized lymphocytes with cerebriform nuclei. However, other characteristic histology features are not much definitive and cannot deliver a diagnosis without uncertainty (6). Immunophenotyping conduces to the diagnosis of MF through revealing the presence of T-cell antigens such as CD3 and CD5 and the loss of CD7. The similarity of T-cell clones to various biopsy sites further has shown the accuracy of MF diagnosis (7). However, presence of eosinophilic infiltrations is not expected. The unique histopathologic feature in the present case study is the unusual eosinophilic infiltration in her lesions and considerable eosinophilia (1600/ mcl), which has been reported only in very few MF patients so far 8-10. Moreover, only three cases of cutaneous lymphoma in patients with prurigo nodularis have been reported in the literature 11. Confirmed by histopathology and IHC, our case had both of these rare features, underlining the

importance of multiple biopsies in patients with nondiagnostic earlier ones, when there is suspicion of malignancy.

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Abstract N°: 1497

A severe case of trigeminal trophic syndrome

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Introduction & Objectives:

Trigeminal trophic syndrome (TTS) is a rare cause of facial ulceration following damage to the central or peripheral branches of the trigeminal nerve. TTS has been associated with trigeminal nerve ablation or transection and stroke. Less common causes include infection (herpes, syphilis, leprosy) and tumours (astrocytoma, meningioma and hemangioma). We present a severe case of TTS in the context of herpes zoster infection and glioblastoma.

Materials & Methods:

A 38-year-old Caucasian man was referred to the dermatology department with a 6-week history of a non-healing ulcer of the left cheek and destruction of the left nasal alar cartilage. Four months earlier, he had undergone debulking surgery of an unresectable right temporal glioblastoma, followed by palliative radiotherapy and chemotherapy.

He described an intractable cutaneous itch and admitted to repeated self-manipulation of the skin. He was otherwise systemically well.

On examination, there was complete loss of the left nasal alar cartilage. The ulceration extended to involve the left upper cutaneous lip, cheek and pre-auricular skin in a V2 dermatomal distribution. There was no ocular involvement. The contralateral face was normal. His neurological examination was unremarkable.

Results:

A CT head and neck demonstrated a large well-defined cutaneous and cartilaginous defect in the left nasal alar with normal appearances of the facial and nasal bones. There was extensive gliosis within the right temporal lobe. Skin swabs were positive for pan-sensitive staphylococcus aureus and varicella zoster virus. A biopsy from the left cheek demonstrated non-specific ulceration without evidence of vasculitis or neoplasm. Tissue culture was positive for staphylococcus aureus, pseudomonas oryzihabitans and naganishia diffluens. Laboratory tests, including complete blood count, basic chemistry, anti-nuclear antibody, anti-neutrophil cytoplasmic antibody and rheumatoid factor were normal. Hepatitis B, hepatitis C and syphilis serology were negative.

Based on clinicopathological correlation, a diagnosis of trigeminal trophic syndrome was made. He was treated with intravenous aciclovir 10mg/kg TDS, piperacillin-tazobactam 4.5g TDS and teicoplanin 1200mg OD for 3 days, then valacyclovir 1g TDS and flucloxacillin 1g TDS for a further 7 days. For skin-directed care, he applied clobetasone butyrate 0.05% with nystatin and oxytetracyline cream BD and antiseptic wash. For neuropathic pain, carbamazepine 100mg TDS was introduced, but then later changed to amitriptyline 25mg OD due to sedation.

After 3 weeks, his neuropathic pain was well controlled. The left cheek and pre-auricular ulceration had completely healed. The nasal alar cartilaginous loss persisted.

Conclusion:

Our case highlights several important considerations. If left untreated, TTS can progress to involve the cheek and pre-auricular skin. Extensive ulceration may represent secondary infection. Although less common, brain tumours and herpes zoster infection have been reported as associations in TTS. Finally, management of neuropathic pain and behavioural modification is essential.

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Abstract N°: 1576

Improvement in cutaneous manifestations with deucravacitinib in systemic lupus erythematosus: impact of baseline patient demographics and disease activity on CLASI-50 responses in the phase 2 PAISLEY trial

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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 moderate to severe plaque psoriasis. Deucravacitinib inhibits TYK2-mediated signaling of certain cytokines involved in systemic lupus erythematosus (SLE) pathophysiology. Phase 3 trials of deucravacitinib in SLE, Sjögren's disease, and psoriatic arthritis and a phase 2 trial in subacute cutaneous and discoid lupus erythematosus (SCLE/DLE) are ongoing. The global, randomized, double-blind, placebo-controlled, 48-week, phase 2 PAISLEY trial (NCT03252587) in patients with active SLE showed that the deucravacitinib 3-mg twice-daily (BID) dose demonstrated significantly greater efficacy than placebo for the primary endpoint of SLE Responder Index-4 (SRI[4]) at week 32 and all key secondary endpoints at week 48, including ≥ 50% reduction from baseline in Cutaneous Lupus Erythematosus Disease Area and Severity Index activity (CLASI-A) score (CLASI-50 response) in patients with moderate to severe cutaneous involvement at baseline (CLASI-A score ≥ 10). In the PAISLEY trial, the safety and tolerability of deucravacitinib was comparable to those seen in other trials in plaque psoriasis and psoriatic arthritis, with no new safety signals observed in patients with SLE. The objective of this post hoc analysis was to evaluate the effects of baseline demographics and disease activity on CLASI-50 responses in patients treated in the PAISLEY study.

Materials & Methods: Patients with active SLE and a baseline CLASI-A score of ≥ 10 who received placebo (n = 24) or deucravacitinib 3 mg BID (n = 23), 6 mg BID (n = 25), or 12 mg once daily (QD; n = 29) were included in this post hoc analysis. CLASI-50 response rates at week 48 were analyzed by select baseline demographics and disease activity characteristics that could affect these responses. Estimated unadjusted differences between deucravacitinib and placebo groups were calculated with a 95% CI based on the Newcombe (corrected) method. All analyses were descriptive.

Results: At the deucravacitinib 3-mg BID dose, which demonstrated optimal results for the primary endpoint of SRI(4), the CLASI-50 response at week 48 was higher (69.6%) than that seen with placebo (16.7%), with a difference in response of 52.9% (95% CI, 21.7%–72.7%) in the study population. Across all baseline demographic and disease activity subgroups, CLASI-50 responses were greater with deucravacitinib 3 mg BID vs placebo, which is consistent with the observed pattern in the overall study population (**Figure**). Similar trends were seen with the other deucravacitinib doses (6 mg BID and 12 mg QD).

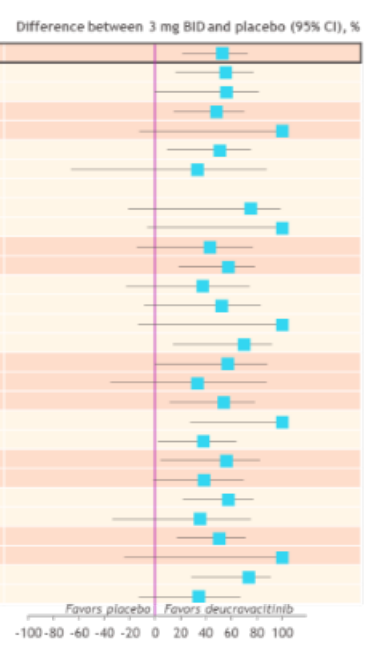
Conclusion: In this post hoc analysis among patients with active SLE, CLASI-50 response rates at week 48 were higher in those treated with deucravacitinib vs placebo across multiple baseline demographic and disease activity characteristics. Although subgroups were small, CLASI-50 responses were generally consistent with those observed

in the overall study population. These findings in subgroups of interest (eg, race, sex, baseline glucocorticoid use at baseline) will be further evaluated in the ongoing phase 3 trials of deucravacitinib in SLE (NCT05620407 and NCT05617677) and the phase 2 trial of deucravacitinib in SCLE/DLE (NCT04857034).

**

Figure. Forest plot of CLASI-50 responses: difference (95% CI) between deucravacitinib 3 mg BID and placebo in baseline subgroups at week 48

	Subgroup	3 mg BID responder, n/N	Placebo responder, n/N	Difference, %
Overall population		16/23	4/24	52.9
Age	18 to < 45 years	10/16	1/14	55.4
	45 to < 65 years	6/7	3/10	55.7
Sex	Female	15/22	4/20	48.2
	Male	1/1	0/4	100
Race	White	9/13	3/16	50.5
	Black	1/3	0/1	33.3
	Native American[a]	0/0	1/3	NE
	Asian	3/4	0/2	75
	Other	3/3	0/2	100
Ethnicity	Hispanic	4/7	1/7	42.9
	Not Hispanic	12/16	3/17	57.4
Region	North America	3/8	0/5	37.5
	Latin America	4/6	1/7	52.4
	Japan	2/2	0/2	100
	ROW	7/7	3/10	70
Time since diagnosis	< 3 years	7/7	3/7	57.1
	3-6 years	1/3	0/4	33.3
	> 6 years	8/13	1/13	53.8
BL SLEDAI-2K score	< 10	6/6	0/5	100
	≥ 10	10/17	4/19	37.8
GC dose at BL	< 10 mg/d	11/13	2/7	56
	≥ 10 mg/d	5/10	2/17	38.2
GC use at BL	Yes	13/18	3/20	57.2
	No	3/5	1/4	35
Antimalarial use at BL	Yes	15/22	4/22	50
	No	1/1	0/2	100
Immunosuppressant use at BL	Yes	11/15	0/10	73.3
	No	5/8	4/14	33.9



Horizontal bars represent 95% CI.
[a]The Native American subgroup comprises American Indian and Alaska Native individuals.
BID, twice daily; BL, baseline; CLASI-50, ≥ 50% reduction from baseline response in Cutaneous Lupus Erythematosus Disease Area and Severity Index activity score; GC, glucocorticoid; NE, not estimable; ROW, rest of world; SLEDAI-2K, Systemic Lupus Erythematosus Disease Activity Index-2000.

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Abstract N°: 1649

Porcelained papules: Malignant atrophic papulosis (Köhlmeier-Degos disease)

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[1]Algeria, algiers, algiers

Introduction :

Malignant atrophic papulosis (MAP), otherwise known as Kohlmeier-Degos disease or Degos disease, is a rare disorder of thrombo-obliterative vasculopathy affecting the skin, gastrointestinal tract, and central nervous system. The characteristic papular skin lesions with central porcelain-white atrophy and a surrounding telangiectatic rim are considered pathognomonic. Two forms of the disease are currently described in the literature: a potentially life-threatening systemic form and a benign monosymptomatic cutaneous form. We report a benign case of Köhlmeier-Degos disease.

Case report :

A-19-year old male was referred for small non-confluent erythematous papules on the trunk and upper extremities. On examination, the lesions were porcelain white atrophic center surrounded by a teleangiectatic rim (Fig. 1). Dermoscopy revealed a central white structureless area surrounded by hairpin vessels (Fig. 2). Histopathological study showed a wedge shaped area of necrosis of the dermis covered by an atrophic granular layer associated with hyperkeratosis. Screening tests of primary hemostasis and coagulation were normal. A systemic Screening did not reveal other anomalies

Discussion:

MAP is a chronic, thrombo-obliterative vasculopathy characterized by papular skin lesions with central porcelain-white atrophy and surrounding teleangiectatic rim. It is rare (Less than 200 cases have been described in the literature). It is generally regarded as a serious vascular disease because it leads to the involvement of multiple organs and results in death within 2-3 years. But in some cases, as in our patient, the disease is limited to the skin and appears benign. The patient was treated with Aspirin 75 mg/d with good initial improvement. He is under regular follow-up.

Conclusion:

The skin lesions of atrophic papulosis are characteristic and easy to recognize. The dermatological diagnosis is rapid. It should lead to early detection of systemic forms, in order to differentiate benign atrophic papulosis from malignant one, which is a rare and serious form. Patients with Degos disease should be followed carefully as there is a continuum from the benign toward the malignant form of the disease. The dermatologist has an important role in the early diagnosis and management of this condition.

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Abstract N°: 1669

Lymphocytic macular arteritis: a rare form of cutaneous vasculitis

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Introduction and Objectives:

Lymphocytic macular arteritis, also known as lymphocytic thrombophilic arteritis, is a rare cutaneous vasculitis affecting medium-caliber vessels. It is characterized clinically by an asymptomatic macular rash and histologically by a predominantly lymphocytic vasculitis. We report the case of macular arteritis in a pregnant woman whose skin lesions have been evolving for 7 years.

Materials and Methods:

A 37-year-old female with no particular pathological history, 15 weeks pregnant , presented livedoid painless, and non-pruritic skin lesions that had been evolving for 7 years on both lower limbs, with no other associated signs, in particular no neurological or osteoarticular signs.

Results:

Examination revealed reticulated erythematous and non-infiltrated macules, localized on both lower limbs, which did not disappear with warming or elevation of the limb; no nodules of the venous tracts or lesions of necrosis or ulceration were noted; neurological and obstetric examinations were normal.

Given this clinical picture, we performed a skin biopsy, which revealed essentially lymphocytic vasculitis lesions involving small- and medium-calibre arteriolar vessels, with an image of intraluminal thrombosis associated with fibrin deposits in the vascular wall.

A complete blood work-up was requested, including immunological tests, all of which came back normal with no systemic involvement found.

According to those results, the diagnosis of lymphocytic macular arteritis was retained.

Conclusion:

Macular arteritis was first described in 2003 by Fein et al. It is a unique entity in terms of its clinical and histological presentation, rarity, and benign course.Its pathophysiology is poorly understood; some authors consider it to be an indolent form of periarteritis nodosa, while others classify it as a separate entity characterized by histological lesions of vasculitis with a predominant lymphocytic infiltrate.

Therapeutic management consists of therapeutic abstention in the majority of cases, while a number of treatments have proved effective: disulone, corticoids, methotrexate, and colchicine.

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Abstract N°: 1674

Blueberry muffin baby: a rare cutaneous manifestation of acute myeloid leukemia.

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Introduction & Objectives: Blueberry muffin baby is a rare manifestation of extramedullary hematopoiesis or neoplasms in infants. This presentation is seen in 25-30% of the cases of acute leukemia, predominantly related to acute myeloid leukemia subtype M4 and M5. We are reporting below the case of a rare presentation of blueberry muffin baby related to acute myeloid leukemia.

Materials & Methods: Records from a patient affected by acute myeloid leukemia were reviewed by clinical dermatology staff and also by the pediatric department in a tertiary hospital.

Results: A 47-day female infant was brought by the parents having a history of violaceous lesions on the trunk and nodules on the head since the 21° day of life with the progressive appearance of new lesions. Obstetric history: dichorionic twin pregnancy with fetal death intrauterine and cesarean operation within 35 weeks of gestation, low weight at birth, maternal serologies in the third semester, with negative results for HIV, syphilis and hepatitis B and C. Physical exam revealed violaceous papules and nodules with different sizes on the trunk, proximal part of the extremities, face, and scalp. Hepatosplenomegaly was present, but lymphadenopathy was not. Laboratory investigation showed anemia, thrombocytopenia, and leukocytosis with predominance of monocytes and blasts in the peripheral blood. Serologies for intrauterine infections were negative. Skin biopsy was performed on a representative nodule and revealed atypical mononuclear cells spread in all the dermis. Immunohistochemical exam: KI-67 positive at around 90% of the cells and CD1A, CD20, CD3, CD34, CD117, S100, myeloperoxidase, and TDT were negative. The immunophenotypic associated with the histopathology findings gave the diagnosis of monocytic leukemia infiltrating the skin. Considering this diagnosis, the patient was admitted in a tertiary pediatric hospital and started receiving chemotherapy. The patient died 10 days after starting treatment due to febrile neutropenia and sepsis.

Conclusion: The phenotypic designation “blueberry muffin baby” is used to describe the presence of multiple dark blue to purple papules and nodules spread in the skin of infants. The lesions are most likely to be found in the head, neck, and trunk. The differential diagnosis includes intrauterine infections, such as rubella and cytomegalovirus, feto-fetal transfusion syndrome and neoplasms. The association of blueberry muffin baby with Langerhans-cell histiocytosis and acute leukemia is even rarer. The variety of differential diagnosis in these cases implies in the necessity of laboratorial investigation and biopsy for histopathological study. The presence of bicytopenia with leukocytosis with a predominance of monocytes in this case report led us to the diagnosis of acute myeloid leukemia and the histopathology with immunohistochemistry reconfirms and establishes the diagnosis of cutaneous infiltration of skin by neoplasm clones.

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Abstract N°: 1780

Necrolytic migratory erythema: A case report

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Introduction & Objectives:

Necrolytic migratory erythema (NME) is a characteristic skin rash most often associated with glucagonoma. The disease can be easily overlooked due to its rarity. Here, we report a case of NME which had been recognised as "eczema".

Materials & Methods:

A 76-year-old woman presented to the dermatology department with a recurrent itchy rash, primarily on her neck and lower legs, which progressed to the arms, buttocks and thighs in 4 months. She had been treated as "eczema" in other clinics, which had poor effects. Physical examination revealed widespread annular or map-like erythematous patches and plaques with remarkable crusts like peeling paint on the damp wall. Biopsy revealed parakeratosis, vacuolation and necrosis of keratinocytes. An enhancing mass in the tail of the pancreas and multiple enhancing masses scattered in the liver were found by contrast-enhanced computed tomography, and the patient's serum glucagon was elevated at 819 (normal range 0–200) ng/L.

Results:

The diagnoses of glucagonoma syndrome and associated necrolytic migratory erythema (NME) were made. The patient rejected treatments for her tumor. Glucocorticoids were applied to relieve itching. Her condition deteriorated rapidly, and the patient died in 3 months.

Conclusion:

The characteristic rash of NEM is frequently unrecognized and the histologic examination is usually nonspecific. Recognition of the specific cutaneous features, followed by careful investigation of the patient's medical history, multiple biopsies and the use of the detailed imaging methods may lead to the diagnosis of this uncommon syndrome. Surgical excision is considered the most definitive treatment to decrease mordality.

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Abstract N°: 1787

Systematic Review on the Skin Manifestations of COVID-19 in association with Disease Severity

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Introduction & Objectives:

The COVID-19 pandemic, caused by SARS-CoV-2, has led to a myriad of clinical presentations. Emerging evidence suggests a potential link between cutaneous manifestations and disease severity, though understanding this relationship remains incomplete. This systematic review aimed to comprehensively explore the prevalence, distinct characteristics, and patterns of cutaneous manifestations in COVID-19 patients. Specific objectives included assessing prevalence, analyzing the temporal correlation of skin lesions with COVID-19 severity, and identifying potential indicators of disease progression.

Materials & Methods:

A systematic search of databases identified 12 studies with a combined sample size of 9547 COVID-19 patients exhibiting skin lesions. Quantitative research methodologies were employed in all selected studies. Methodological quality assessment and data extraction were conducted to synthesize findings.

Results:

The review revealed significant evidence supporting the correlation between cutaneous manifestations and COVID-19 severity. The majority of studies demonstrated that skin manifestations served as early indicators of varying disease severity levels. Specific types of skin lesions correlated with different stages of disease progression. Notably, findings varied among studies, with some pointing to early onset indicators and others emphasizing specific lesion patterns associated with severe cases.

Conclusion:

This systematic review underscores the importance of recognizing cutaneous manifestations as potential markers of COVID-19 severity. Collaboration among healthcare professionals, integration of dermatologic assessments, and standardized research approaches are recommended to enhance understanding and patient care in this evolving field.

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Abstract N°: 1832

A challenging dermatosis in an immunocompromised patient

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Introduction & Objectives: Sweet's syndrome, is a rare dermatological condition with no accurate data on incidence or prevalence. It is more common in females, occuring commonly between 30 and 60 years. It is characterized by sudden onset of well-defined tender plaques or nodules accompanied by fever, arthralgias, ocular inflammation and headaches. Multiple factors are implicated in the pathogenesis. Histopathology is diagnostic with characteristic features of predominant neutrophilic infiltrate in the dermis. Gold standard treatment is topical and systemic steroids. First case of Sweet's syndrome in HIV infected male with TB lymphadenitis is highlighted.

Materials & Methods: A 54 year old male presented with multiple, well defined, large, irregular, erythematous to hyperpigmented infiltrated plaques over forehead, bilateral cheeks, nose, chest, shoulders with overlying hyperpigmented crusts since 10 days. Bilateral periorbital edema was present. He had fever on and off since a month and cough with expectoration since 15 days. He had history of significant weight loss in a span of 4 months. He was a diagnosed case of HIV since 2016, on tab Dolutagravir, Lamivudine & Tenofovir. Sensations were preserved. Deep dermal tenderness was absent.Pathergy test was negative. There was cervical lymphadenopathy. Investigations revealed decreased haemoglobin, raised ESR and CRP, low CD4 count. X ray chest showed mild left ventricular cardiomegaly.USG neck showed enlarged cervical, inguinal and axillary lymph nodes. FNAC of cervical lymph nodes was suggestive of tuberculous lymphadenopathy. HRCT thorax was suggestive of small airway disease. Genexpert was negative for M.TB with positive Interferon gamma release assay. Histopathology was suggestive of neutophilic dermatoses.Diagnosis of Sweets's syndrome in a known case of HIV with TB lymphadenitis was made. Treatment included injectable antibiotics, antipyretic, initation of AKT and continuation of ART.Topical corticosteroid resulted in resolution of skin lesions.**

Results and Conclusion: HIV creates an environment more favourable to pathogenesis of Sweet syndrome through a hypersensitivity response with neutrophil chemotaxis , altered Type 1 and Type 2 helper T-cell response and presence of HIV trans-activating proteins.Treatment remains same as for immunocompetent individual. Association of tuberculosis and Sweet's syndrome is very rare. It should be recognized that M. tuberculosis may be one of the causes of Sweet's syndrome. For unexplained skin lesions in HIV-infected, Sweet's syndrome can be a differential diagnoses. Sweet's syndrome should be included as one of the associated reactionary skin lesions of tuberculosis.

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Abstract N°: 1856

Mask Phenomenon: A Rare Complication Following Bronchoscopy

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Introduction & Objectives:

Purpura is defined an extravasation of erythrocytes into the dermis of the skin. facial purpura can be due to rheumatological, dermatological, infectious, and traumatic causes. However, various benign causes of facial purpura such as forceful coughing, vomiting or Valsalva's maneuver have been reported under the name of "mask phenomenon". The occurrence of mask phenomenon post-bronchoscopy is seldom reported.

Materials & Methods:

We presents a case of a 60-year-old woman who developed a petechial rash on her face and neck, along with subconjunctival hemorrhage, following bronchoscopy.

Results:

Diagnosis of endoscopy-related purpura involves the exclusion of vasculitic, coagulopathic, neoplastic, infectious, and other causes. The main features point to endoscopy- related purpura include absence of vasculitis or coagulopathies in history; normal blood works; superficial lesions such as petechiae; predominantly facial and/or neck distribution during or soon after endoscopic procedure and the spontaneous resolution of rash within 7-10 days .

Conclusion:

The differential diagnosis of facial and neck purpura is extensive, encompassing rheumatologic, dermatologic, infectious, and traumatic causes. It is crucial for physicians to recognize benign causes of facial purpura, such as the mask phenomenon, to avoid unnecessary diagnostic procedures. Moreover, this complication should be considered in the spectrum of bronchoscopy-related adverse events, enhancing the management and counseling of patients undergoing this procedure.

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Abstract N°: 1866

Skin Conditions among Inflammatory Bowel Disease Patients in Tertiary Care Hospital

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Introduction & Objectives:

It is well known that skin disorders may be a frequent extraintestinal manifestation of inflammatory bowel disease (IBD). Through literature analysis, we found an obvious lack of epidemiological data regarding this issue. Therefore, the aim of this study was to establish the prevalence of skin conditions among IBD patients in a tertiary care hospital.

Materials & Methods:

Prospective study included patients with IBD who were managed in a tertiary care hospital. Patients completed questionnaires including the demographic and IBD data and history or present state of skin conditions. Skin conditions were considered as related to IBD if they were diagnosed following the diagnosis of IBD. Skin conditions which were reported before the diagnosis of IBD were considered as not related to IBD.

Results:

162 patients were included in the study, of them 93 (57.4%) were males. Mean age (MA) – 42.5±14.2 years. MA of men – 42.1±14.4, women – 42.9±14.0 years, p>0.05. Ulcerative colitis (UC) was diagnosed in 117 (72.2%) patients, Crohn's disease (CD) – in 45 (27.8%) patients. MA of UC patients was 43.1±14.0, CD – 40.9±14.9, p>0.05.

In total, skin conditions were indicated by 66 (40.7%) subjects. According to our criteria, in 47 (29%; 95% CI: 22.0-36.1%) cases, skin conditions were considered as obviously related to IBD. We further analysed the latter cases.

Among UC patients, 35 (29.9%; 95% CI: 21.5-38.3%) had skin conditions related to IBD, among CD patients – 12 (26.7%; 95% CI: 13.2-40.1%), p>0.05.

Erythema nodosum was reported by 6 (3.7%) patients, pyoderma gangrenosum – 6 (3.7%), acne – 2 (1.2%), psoriasis – 10 (6.2%), vitiligo – 2 (1.2%), epidermolysis bullosa acquisita – 1 (0.6%), haemorrhagic vasculitis – 1 (0.6%), eczema – 11 (6.8%), allergic rash – 5 (3.1%), other (unspecified) – 8 (4.9%).

The detailed comparison of different skin conditions among UC and CD patients is presented in table 1.

Table 1. Prevalence of skin conditions among patients with ulcerative colitis and Crohn's disease

Dermatologic comorbidity	Ulcerative colitis (N=117) n (%)	Crohn's disease (N=45) n (%)	p-value
Erythema nodosum	3 (2.6%)	3 (6.7%)	>0.05
Pyoderma gangrenosum	5 (4.3%)	1 (2.2%)	>0.05
Acne	1 (0.9%)	1 (2.2%)	>0.05
Psoriasis	8 (6.8%)	2 (4.4%)	>0.05
Vitiligo	2 (1.7%)	0 (0%)	>0.05
Epidermolysis bullosa acquisita	1 (0.9%)	0 (0%)	>0.05
Vasculitis	1 (0.9%)	0 (0%)	>0.05
Eczema	9 (7.7%)	2 (4.4%)	>0.05
Allergic rash	4 (3.4%)	1 (2.2%)	>0.05

Among UC patients, skin conditions were reported by 2 out of 17 (11.8%) subjects with proctitis, 7 out of 30 (23.3%) subjects with left-sided colitis and 27 out of 70 (38.6%) subjects with pancolitis, $p<0.05$ between proctitis and pancolitis.

Comparing different types of CD, skin conditions were indicated by 2 out of 17 (11.8%) patients with ileitis, 4 out of 11 (36.4%) patients with colitis and 6 out of 17 (35.3%) patients with ileocolitis; $p>0.05$.

Conclusion:

In our study, the prevalence of skin conditions among IBD patients was 29%. There were no differences in the prevalence of skin conditions between the UC and CD patients. The most common skin conditions in UC were eczema and psoriasis, in CD – erythema nodosum, psoriasis and eczema. We observed the obvious trend of more frequent skin conditions in patients with more extensive UC. There was also the trend for more frequent skin conditions in CD patients with colonic damage.

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Abstract N°: 1879

Dermatological Manifestations Associated with Vitamin B12 Deficiency: A Review

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Introduction & Objectives: Cobalamin, or Vitamin B12, is a water-soluble vitamin implicated in vital cellular processes namely nucleotide synthesis and DNA methylation. The dermatological manifestations of vitamin B12 deficiency are often overlooked despite being readily reversible with adequate treatment. As such, we hereby present a thorough review of the metabolism of vitamin B12, its cellular implications, its association with dermatologic diseases including cutaneous, mucosal, hair and scalp manifestations, the diagnostic approach of suspected deficiency, and its management.

Materials & Methods: A literature review was conducted using the keywords Vitamin B12; deficiency; skin manifestation; dermatology. The search on Pubmed yielded 65 articles out of which 33 met the inclusion criteria and were reviewed more extensively. Screening of the references yielded an additional 19 articles resulting in a total of 52 articles included in this study.

Results: Vitamin B12 deficiency can manifest as cutaneous, mucosal, hair and scalp changes. One of the earliest signs includes hyperpigmentation of the palmar and plantar surfaces, intertriginous areas, oral mucosa, regions exposed to pressure such as the elbows and knees, extensor surfaces of finger and toe joints, and new scars. Other patterns of hyperpigmentation like isolated knuckle, acral and Addisonian hyperpigmentation were also reported. Additionally, nail changes including longitudinal melanonychia and complete 20 nail melanonychia related to vitamin B12 deficiency is documented. Non-specific hair loss, telogen effluvium, androgenetic alopecia, alopecia areata have also been linked to vitamin B12 deficiency, with 20-30% of patients with telogen effluvium having a documented vitamin B12 deficiency. Moreover, prospective studies show B12 deficiency can manifest with mucosal changes like angular cheilitis, glossitis and recurrent aphthous stomatitis. Studies have also established an association between B12 deficiency and dermatological diseases including vitiligo, atopic dermatitis, psoriasis, acne vulgaris, and rosacea. Diagnosis through serum cobalamin assay remains the standard routine diagnostic test. Measuring plasma homocysteine and methylmalonic acid is also indicated in some cases especially when B12 is borderline. Treatment is through supplementation of vitamin B12, possibly in combination with vitamin B6 and folic acid. The decision for route, dose and type of treatment is dependent on the cause of the deficiency, patient presentation and in some cases preference.

Conclusion: In conclusion, vitamin B12 deficiency is associated with different dermatological conditions and manifestations. It is important to test for vitamin B12 deficiency in patients presenting with such dermatological changes. Early recognition and treatment of cobalamin deficiency helps prevent the progression into serious neurological manifestations.

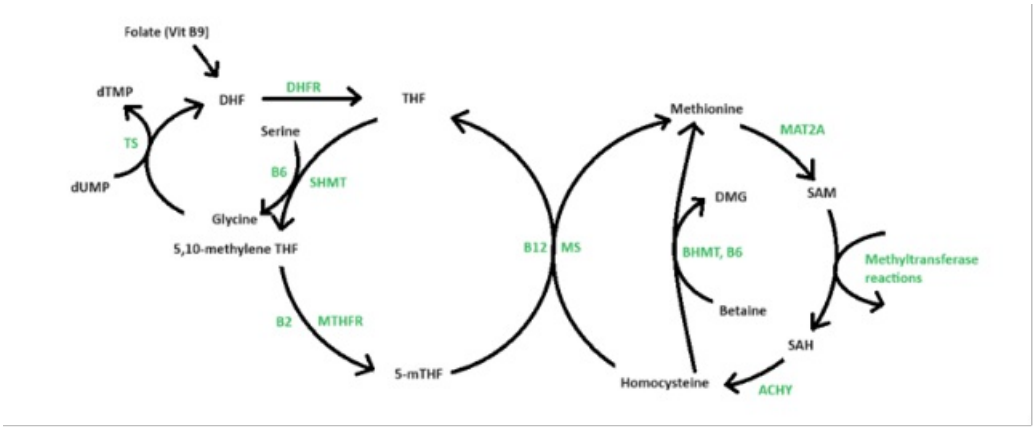

Figure 1: Folate and Methionine Cycles and the role of vitamin B12 and folate in generating and recycling methionine needed for one-carbon metabolism reactions.

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Abstract N°: 1958

Eruptive xanthomatosis - A Case Report

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Introduction & Objectives: Case report describes the case of a 26-year-old man with an exanthema characterized by pruritic, firmer whitish-yellow papules, localized on the buttocks, with gradual progression to the back and arms. Hundreds of papules occurred on his body. The patient had had these symptoms for about 3 weeks and they had occurred for the first time in his life. The patient with this finding was for the first time examined by a doctor at our clinic. Apart from fatigue and increased sweating, the patient did not report any other problems.

Materials & Methods: The patient underwent hematological, biochemical and immunological blood sampling and an exploratory excision was performed. The differential diagnosis was - folliculitis, xanthomatosis,lichen myxedematosus, small-foci sarcoidosis or cutaneous amyloidosis. A very high level of cholesterol (13,7 mmol/l) and triglycerides (32,3 mmol/l) was found during sampling. This established the diagnosis of eruptive xanthomatosis, which was subsequently confirmed by histological examination.

Results: An internist was consulted, and the first recommendation was an absolute prohibition of alcohol consumption. The patient admitted having psychological problems and consuming alcohol daily. The patient stopped drinking alcohol and had not received any other treatment yet. When checked after fourteen days, the lipid levels were almost normal (cholesterol 5,0 mmol/l, triglycerides 1,9 mmol/l) and the cutaneous xanthomas were very flattened. At the control after three weeks, the manifestations of eruptive xanthomatosis were completely healed.

Conclusion: What is particularly interesting about this clinical case is that the only treatment was a ban on drinking alcohol. It also demonstrates the importance of early laboratory blood testing. The poster presentation contains photographs of clinical manifestations of xanthomatosis. Eruptive xanthomatosis mostly reveals a serious disorder of lipid metabolism. An early detection and treatment of this disorder can greatly reduce the patient's cardiovascular risk.

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Abstract N°: 2048

Guselkumab in Chinese patients with scalp and nail psoriasis: subgroup results from a phase 4 study

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Introduction & Objectives:

Psoriasis commonly affects the scalp and nails, in these regions it is typically hard to treat and significantly impacts patients' (pts) quality of life. Guselkumab (GUS) is a human monoclonal interleukin-23 p19 subunit-targeted antibody approved for the treatment of moderate-to-severe plaque psoriasis in multiple countries. A post approval commitment study was conducted to verify the efficacy and safety profile of GUS in Chinese pts. The objective of this subgroup analysis was to analyze the efficacy of GUS in those with scalp and nail psoriasis.

Materials & Methods:

This randomized, double-blind, placebo (PBO)-controlled, multicenter phase 4 study enrolled pts who had moderate-to-severe plaque psoriasis (Investigator's Global Assessment [IGA] score ≥3, Psoriasis Area and Severity Index [PASI] ≥12, and involved body surface area [BSA] ≥10%) and were eligible for systemic therapy or phototherapy. Pts were randomized to receive GUS or PBO-GUS at a 2:1 ratio for a 44-week (wk) treatment period, and safety monitoring was through wk 56. Pts assigned to GUS received GUS 100 mg by subcutaneous injection at wks 0 and 4, and then every 8 wks thereafter. Pts assigned to the PBO-GUS group received PBO at wks 0, 4, and 12, and then switched to GUS at wk 16. Overall psoriasis severity was measured by IGA and PASI at baseline and throughout the 48-wk treatment period, while severity of scalp and nail psoriasis was evaluated by the Scalp Specific IGA (ssIGA) and Nail Psoriasis Area and Severity Index (NAPSI), respectively, at baseline and wks 16, 28, 36, and 48.

Results:

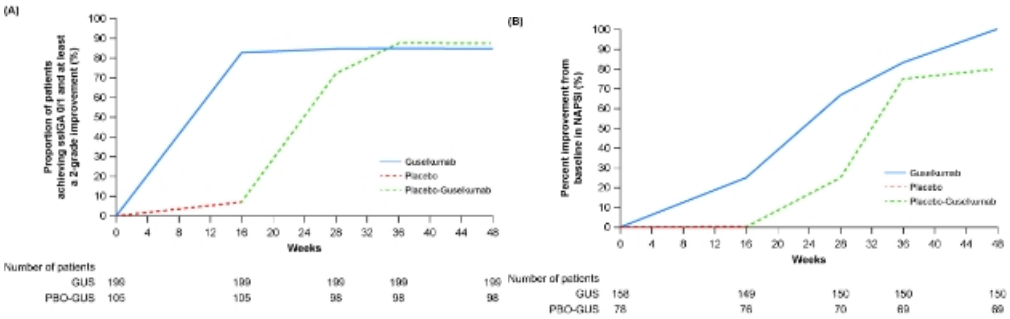
Overall, 327 pts were enrolled, randomized (217 to GUS and 110 to PBO-GUS), and received ≥1 dose of study treatment. Mean (SD) duration of psoriasis was 11.8 (9.4) years. Most pts had an IGA score of 3 (76.4%) or 4 (25.1%), and mean PASI score was 23.9 (10.2). At baseline, 317 pts (96.9%) had scalp psoriasis (ssIGA score ≥1) and 304 (93.0%; 199 in the GUS group and 105 in the PBO group) had a ssIGA score ≥2. Among 236 pts (72.7%; 158 in the GUS group and 78 in the PBO group) with nail psoriasis (NAPSI >0) at baseline, all had an IGA score of 3 (74.5%) or 4 (25.5%) and mean PASI and NAPSI scores were 24.3 (10.6) and 4.2 (2.1), respectively. Among 304 pts with a ssIGA score ≥2 at baseline, a higher proportion receiving GUS achieved ssIGA 0/1 and at least a 2-grade improvement compared with PBO at wk 16 (82.9% vs 7.1%). ssIGA response was maintained in the GUS group (84.8% at wks 28 and 48) and improved for PBO pts after switching to GUS (72.4% at wk 28 and 87.6% at wk 48) (Fig 1A). Among pts with nail psoriasis at baseline, median precent improvement in NAPSI was greater in the GUS group compared with the PBO group at wk 16 (25.0% vs 0.0%); median percent improvements in NAPSI at wks 28 and 48 were 66.7% and 100.0% in the GUS group and 25.0% and 80.0% in the PBO-GUS group, respectively (Fig. 1B). Clearance of scalp and nail psoriasis with continuous GUS treatment was consistent with responses in other body regions as measured by IGA and PASI. GUS was well tolerated in Chinese pts and the

safety findings were consistent with the known safety profile.

Conclusion:

GUS treatment led to meaningful improvements in scalp and nail psoriasis in Chinese pts with moderate-to-severe plaque psoriasis.

Fig 1. Response to treatment. (A) ssIGA curve in patients with a ssIGA score ≥2 at baseline and (B) NAPSI curve in patients with NAPSI >0 at baseline.



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Abstract N°: 2201

A Systematic Review of Clinical Features and Treatment Outcomes of Eruptive Xanthomatosis

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Introduction & Objectives:

Eruptive xanthomas (EX) are cutaneous lesions caused by lipid deposits which can act as an important diagnostic marker of underlying metabolic disorders. This review aims synthesize current research for better clinical practice by systematically reviewing the clinical features and treatment outcomes of EX.

Materials & Methods:

A systematic search of MEDLINE, Embase, and PubMed was performed using 'eruptive xanthoma' and 'eruptive xanthomatosis' as search terms, without restriction on publication date, location or language (CRD42024523873). Article screening was performed independently by three reviewers (LAM, CB, CAC). The inclusion criteria encompassed all clinical studies reporting on patients with EX.

Results:

The search yielded 242 entries. After removing duplicates and full screening, 69 studies met our inclusion criteria, totaling 80 cases of EX. The majority were male (63.8% n=51/80) and the remaining were female (28.8%; n=23/80) (unspecified gender: [7.5%; n=6/80]). The mean age at EX diagnosis was 34.5 years. Associated comorbidities included dyslipidemia (81.3%; n=65/80) and diabetes mellitus (61.3%; n=49/80). The most common presentations were yellow (68.8%; n=55/80) papules (72.5%; n=58/80) surrounded by a halo (16.3%; n=13/80), with the majority affecting the elbow (47.5%; n=38/80) and the knee (38.8%; n=31/80). The complete response rate, indicating clearance of all lesions, was highest with fibrates (69.2%; n=9/13), the most efficacious of which being gemfibrozil (100%; n=4/4). Dietary lipid restrictions led to complete resolution in 46.7% (n=7/15) of cases. Statins led to recovery in only half of patients (50%; n=4/8).

Conclusion:

EX are a significant clinical indicator for serious metabolic comorbidities such as dyslipidemia and diabetes mellitus. Integrating pharmacotherapy and dietary modifications is crucial for effective management. Further studies are needed to explore the potential of EX as markers of disease severity and prognosis.

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Abstract N°: 2286

Predictive factors for concurrent infection in patients with adult-onset immunodeficiency syndrome associated with anti-interferon gamma autoantibodies presenting with reactive dermatoses

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Introduction & Objectives:

Reactive dermatoses are commonly found in patients with adult-onset immunodeficiency syndrome (AOIS) due to anti-interferon gamma autoantibodies (AIGAs). Some reactions might be associated with infections and require antimicrobial treatment along with anti-inflammatory medication. The study aims to investigate predictive clinical features and laboratory findings for concurrent infection in patients with AOIS due to AIGAs who present with reactive dermatoses.

Materials & Methods:

The medical records of patients with AOIS due to AIGAs with skin manifestations during 2013-2022 were retrospectively reviewed. Drug eruptions, infectious dermatoses, and skin manifestation episodes not containing definite diagnoses were excluded. Reactive dermatoses were classified as neutrophilic dermatoses (such as pustular reaction and Sweet's syndrome) and non-neutrophilic dermatoses (such as erythema nodosum and vasculitis). Clinical and laboratory findings in reactive episodes were compared between patients with and without concomitant infection using the Chi-square or Fisher's exact test.

Results:

There were 135 episodes of reactive dermatoses, further categorized into 99 neutrophilic dermatoses and 36 non-neutrophilic dermatoses. The median age of onset was 52.0 years old (IQR: 43.0, 58.0), with a female predominance (73.3% of all episodes). Concomitant infection was identified in 122 episodes, comprising clinical and microbiological confirmation in 93 episodes as well as only clinical diagnosis in 29 episodes. *Mycobacterium abscessus* was the most common organism identified in microbiologically documented infections, found in 65.6% of the cases. Compared to those without concomitant infection, patients with reactive episodes and concomitant infection were significantly associated with certain clinical and laboratory findings including, males (29.5% versus 0.0%, with a p-value of 0.020), anemia (89.3% versus 53.8%, with a p-value of 0.003), mean corpuscular hemoglobin less than 25 pg (54.1% versus 23.1%, with a p-value of 0.033), red cell distribution width more than 15% (72.1% versus 30.8%, with a p-value of 0.004), white blood cell counts more than 11,000/uL (91.8% versus 53.8%, with a p-value of 0.001), absolute neutrophil count more than 7,000/uL (91.8% versus 53.8%, with a p-value of 0.001), and erythrocyte sedimentation rate more than 75 mm/hr (55.8% versus 25.0%, with a p-value of 0.044).

Conclusion:

Various clinical and laboratory results are associated with concurrent infection in patients with AOIS due to AIGAs who have reactive dermatoses. A routine complete blood count panel is an inexpensive and accessible blood test that helps identify patients at risk of concomitant infection in reactive skin episodes. The most common organism

in reactive dermatoses with concomitant infection is *Mycobacterium abscessus.*

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Abstract N°: 2384

What Leads to Acquired Perforating Dermatosis?

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Introduction & Objectives:

Perforating dermatoses (PD) are a rare group of dermatoses characterised by abnormal elimination of dermal material, namely collagen and elastic fibres, "perforating" by extrusion through the epidermis to the surface. The four categories of PD are reactive PD, elastosis perforans serpiginosa, perforating folliculitis, and acquired perforating dermatosis (APD); the latter is associated with a systemic illness.

Materials & Methods:

This case study describes a middle-aged man with pruritic dermatoses in association with chronic kidney disease (CKD) and HIV-1. We conducted bibliographic research using the PubMed database with the search term "acquired perforating dermatosis." After screening, this review contained 15 of the 95 articles.

Results:

A 35-year-old man presented to the dermatology department with a 4-week history of itchy skin lesions on the genital area and both legs. His medical record includes HIV-1 with no history of AIDS-defining illnesses, treated with raltegravir 400 mg and abacavir lamivudine 600/300 mg, and CKD in dialysis.

A dermatological examination revealed several red-brown, hyperpigmented papules with a keratinous plug on an erythematous base on the left testicular area, as well as the front of both the right and left legs. Blood tests: urea 98.1 mg/dL, BUN 46 mg/dL, creatinine 16.72 mg/dL, K 3.97 mg/dL, Ca 8.61 mg/dL, and P 6.63 mg/dL, CD4 714 (46%), and viral load <40 copies/mL.

After a clinical examination, we suspected APD was associated with severe azotemia. We recommended a topical treatment using fluocinolone. In the subsequent appointment 4 weeks later, the dermatosis was not itchy and had lost its erythematous base. A skin biopsy revealed an ulceration with transepidermal elimination of basophilic collagen, and Masson's trichrome stain showed collagen fibres penetrating the epidermis in the dermis, confirming the APD diagnosis. Topical steroids and pirfenidone were the indicated treatments. Four months later, the lesions improved, subsiding residual post-inflammatory hyperpigmentation and some scarring.

Conclusion:

APD, a rare skin disease characterised by the elimination of dermal material through the epidermis, is associated with underlaying diseases like CKD and diabetes and less commonly with solid tumours, HIV, and lymphomas. Experts estimate the incidence at 2.53 cases per 100,000 individuals annually. APD starts as 2–10 mm hyperkeratotic papules that can turn into umbilicated papulonodules with a central keratotic plug on the hands and limbs. Lesions heal, leaving scars and hyperpigmentation. While the exact cause is still unknown, pruritus and recurrent injury from scratching are the main causes of APD, causing the Koebner phenomenon. Pathogenesis includes aberrant keratinization, collagen degradation, genetic vulnerability, and epidermal calcium deposits. Clinical and histological features determine the diagnosis.

While APD is associated with impaired immune systems and higher dermatological susceptibility in HIV patients, our patient's CD4 count was greater than 200, and his viral load was undetectable. The lesions are likely CKD-related due to reduced renal function, significant azotemia, and waste accumulation. It is important for healthcare providers to consider these underlying conditions when managing APD in patients, as treatment options may vary. Future research should investigate APD mechanisms and create targeted therapeutics to enhance patient outcomes.

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Abstract N°: 2538

Spontaneous cholesterol crystal embolism

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Spontaneous cholesterol crystal embolism

Introduction & Objectives:

Cholesterol crystal embolism is a systemic pathology caused by the detachment of atherosclerotic plaque material, such as cholesterol crystals, from large-diameter atherosclerotic arteries causing multiple organs damage.

We report a case of cholesterol crystal embolism with skin involvement.

Case report:

A 71-year-old man with known dyslipidaemia and high blood pressure consulted for an erythematoviolaceous livedo racemosa of the foot soles evolving over the past week associated with right big toe necrosis.

The anamnesis revealed brutal hypoacusis and ischemic stroke incidentally detected on MRI 2 months ago.

General condition was preserved. No digestive symptoms were reported.

Biological assessment of vasculitis, kidneys function and eye fundus were free of anomalies.

Lower limbs arterial echo-doppler revealed extremely distal arteriopathy probably of embolic origin. Angioscanner showed an atheromatous infiltration of the abdominal aorta with very high embolism risk.

Transesophageal echocardiography showed profuse aortic atheroma with voluminous plaques of the descending thoracic aorta.

The skin biopsy revealed significant capillary hyperplasia, gathered in bouquets. Some emboli of cholesterol crystals were found in the dermis and hypodermis.

We proceeded to Iloprost perfusions, necrosis mummification and we intoduced Aspirin and Rosuvastatin which helped halt the disease's progression.

Results:

Cholesterol crystal embolism is a serious pathology, underdiagnosed, with poor prognosis.

It classically occurs in the aftermath of invasive endovascular procedures and thrombolytic therapies especially by the femoral approach. A condition that was not present in our patient.

In the literature, spontaneous cholesterol crystal embolism varies between 12.5et and 20% of cases.

The clinical presentation is heterogeneous and polymorphic. The dermatological signs most often seen, in descending order, are purple or blue toes, livedo, gangrene, cyanosis, ulceration, nodules and purpura.

The diagnosis is based on a combination of clinical, biological, morphological and histological arguments.

The treatment is poorly codified. It aims to relieve symptoms with NSAID or steroids, promote healing, prevent recurrences and attempt to eradicate the emboligenic source.

Any factor that might maintain the embolizing phenomenon should be eliminated. The control of cardiovascular risk factors is essential.

The introduction of statins showed improvement of cutaneous manifestations and renal outcomes.

Eviction of antithrombotic therapy should be considered in the terms of benefit risk balance.

If needed invasive endovascular procedure, the radial approach has shown to have lower risk of cholesterol embolism as compared to a femoral approach.

Conclusion:

Cholesterol crystal embolism is a serious disease with high mortality. Clinicians should be aware of its features to ensure rapid diagnosis and early treatment even in the absence of triggering factors.

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Abstract N°: 2705

Tuberous xanthomas revealing homozygous familial hypercholesterolemia

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Introduction :

Tuberous xanthomas are nodular, firm, painless lesions. Most often linked to a disorder of lipoprotein metabolism. We report the case of a child in whom multiple tuberous xanthomas revealed familial hypercholesterolemia.

: OBSERVATION : OBSERVATION: BN child aged 8 years, from a first degree consanguineous marriage, with a family history of hypercholesterolemia, who consults for papulo-nodular and tumorous lesions, symmetrical, yellowish in color, painless and non-inflammatory, at the elbows, knees, buttocks and backs of the feet evolving for 3 years gradually increasing in size and taking on a tuberous appearance. Dermoscopic examination showed a yellow area without structure. The skin biopsy revealed a nodular lesion made of foamy histiocytic cells containing lipid droplets confirming the diagnosis of xanthoma. The lipid profile showed very high levels of total cholesterol (9.32 g/L), LDL cholesterol (8.80 g/L), HDL cholesterol was reduced (0.34 g/L), triglycerides were within the standards. There were no cardiac or ophthalmological abnormalities. In this child, the very high levels of LDL cholesterol, the presence of cutaneous xanthomas since the age of 5 and the absence of other abnormalities, suggested a homozygous form of familial hypercholesterolemia of type II according to the Fredrickson classification, The patient was managed by hygienic and dietary measures associated with treatment with atorvastatin 10 mg/day. We noted a clear improvement in the lipid profile after 1 and a half months of treatment: Total cholesterol at 06 g/l and LDL at 06 g/l. 5.7 g/l. A screening lipid profile in other members of the family was proposed.

CONCLUSION:Xanthomatoses linked to homozygous familial hypercholesterolemia are very rare. Morbidity and mortality are linked to atherosclerosis. Early and targeted detection allows adequate care and a reduction in early cardiovascular events.

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Abstract N°: 2804

Erythema gyratum repens in a patient with miliary tuberculosis: an infrequent association

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Introduction & Objectives:

Erythema gyratum repens (EGR) is a variant of figurate erythema recognized for its unique clinical and histopathologic characteristics. While its precise pathophysiology remains uncertain, it is linked to an immune-mediated response commonly associated with large tumors or infections. Nonetheless, in several cases, the trigger may remain unidentified.

Materials & Methods:

A 37-year-old male presented with a three-month history of fever, weight loss, dyspnea, abdominal pain, and diarrhea. Within 20 days, he developed a skin eruption. Physical examination revealed erythematous-brown, tender, eroded plaques with a polycyclic configuration and tendency to coalesce, located on the back and extremities.

Laboratory tests showed anemia, lymphopenia, and thrombocytosis. Sputum acid-fast bacillus microscopy was positive. Tomographic findings were consistent with miliary tuberculosis affecting lungs, lymph nodes, and intestines. Biopsy of a cecal granulomatous lesion was positive for Ziehl-Neelsen (ZN) and modified ZN staining. Skin biopsy revealed vacuolar interface reaction pattern and superficial perivascular inflammatory infiltrate. ZN and modified ZN staining were negative.

Results:

A diagnosis of erythema gyratum repens was established. The patient received antituberculous treatment alongside betamethasone cream 0.05% applied every 12 hours. A week later, remission of the lesions was observed.

Conclusion:

We want to highlight that EGR is not exclusively a paraneoplastic syndrome. Nevertheless, it is important to rule out malignancy, as its prompt identification is crucial in reducing morbimortality. A careful evaluation of other etiologies becomes necessary, given that the cutaneous manifestations tend to recede once the underlying issue is addressed. To our knowledge, this could be the first documented case associating EGR with miliary tuberculosis, given that previous descriptions have been related to pulmonary tuberculosis. The causal relationship between both entities was established due to the resolution and absence of recurrence of the skin lesions after antitubercular treatment.

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Abstract N°: 2919

Prurigo pigmentosa: Rationale for Screening for Diabetic Ketoacidosis swiftly

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Introduction & Objectives:

Diabetes mellitus (DM) is one of the most common diseases worldwide and in Saudi Arabia. Various cutaneous manifestations have been historically associated with DM such as bullosis diabeticorum, acanthosis nigricans, necrobiosis lipoidica, and many others. Many of them are linked to longstanding DM and are viewed as cutaneous complications. However, skin changes are rarely the presenting symptom of an undetected type 1 diabetes mellitus, which is usually first detected in patients with symptoms of hyperglycemia with or without ketoacidosis. In the following case report, we present a case, in which clinical dermatological diagnosis of prurigo pigmentosa (PP) led to clinical suspicious of type 1 DM with diabetic ketoacidosis (DKA). As this rare, but potentially life-saving intervention by dermatologists in the detection of type 1 DM is not well reported in literature, we aim to emphasize the importance of correct detection of PP, based on clinical features and provide a rationale for swift testing for DM.

Materials & Methods:

We report an unusual case of 16 years old girl, who presented to the ER with dizziness, body aches and pruritic rash on the chest. The patient's symptoms were dismissed with attention switched onto the skin rash only. On following day, the patient presented to our clinic. Physical examination of the abdomen revealed a reticulated, erythematous patches with numerous, superimposed, excoriated papules. Our clinical assessment was consistent with PP. We ran blood tests, urine examination and obtained a skin biopsy.

Results:

The labs revealed severe hypergycemia, glucosuria, and ketonuria. As result, the patient was admitted as case of DKA secondary to a newly diagnosed Type 1 DM. She was successfully managed in the general ward, without any further complications. Later, the histopathological examination showed features consistent with PP.

Conclusion:

PP is a rare and underreported dermatological condition of unknown etiology first described by Nagashima in Japan. Several associations have been described in the literature including ketosis, ketogenic diet, bariatric surgery. In many cases, however, the triggering factors remain unidentified. Dermatological and general medical literature lacks a clear association and recommendation for immediate testing for DM. In our case report, the clinical suspicion of PP has led to the detection and diagnosis of DKA. DKA is a common, potentially life-threatening complication of DM associated with significant morbidity and mortality. We, therefore, recommend to rapidly screen all patients, that present to dermatologists with a clinical picture suggestive with PP, for DM and DKA. This simple, readily-available, and cost-effective measure can help in avoiding any further delays in the detection of DKA and reduces the risk of DKA-associated complications and mortality.

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Abstract N°: 3208

Liver transplant patients and risk of skin disease: conscious prevention?

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Introduction & Objectives: Liver transplant patients (LT) have high risk of dermatological complications because of the lifelong immunosuppressant therapy. Infection and skin cancer are the most common, but also Graft versus Host Disease can occur.

Aim of the present observational study was to evaluate patient's awareness in regard of the increased risk of skin cancer, and to assess any consequent behavioral modification.

Materials & Methods: LT patients attending the Gastroenterology Unit of the University Hospital of Salerno "San Giovanni di Dio e Ruggi d'Aragona" were enrolled in the study at the moment of the annual dermatological screening, during the past 4 months, form the 1st of December 2023 to the 31st of March 2024. Each patient underwent clinical examination, phenotype and phototype was assessed, and personal and family history, with particular regard to dermatological conditions, was recorded, together with the immunosuppressive therapy scheme. Moreover, patients were asked to answer a questionnaire specifically designed to explore the level of information and the awareness of these subjects toward the increased carcinogenic risk.

Results: During the study period, 75 consecutive patients were enrolled, 49 men e 26 women, mean age 62 ± 10 years (DS 10), and the data recorded were anonymously reported on an Excel spread sheet for analysis. At the enrollment, the mean time after LT was 12 years, and all patients were on one or more immunosuppressive and antiviral drug; 64/75 (85%) had already undergone dermatological consultation, whereas 11 patients of 75 (15%) met a dermatologist for the first time; most of them (10/11) had started the dermatological follow up more than 5 years after the liver transplant. Of 75 patients, 55 (73%) were aware of the skin cancer risk after LT, and concerning their behavior toward the sun, 65 (87%) reported to avoid it after the transplant, whereas 10 (13%) continue to sunbath as before. In regard to photoprotection, 62% of patients (47/75) reported the use of topical sunscreens, whereas the rest (38%) ignored photoprotection. **Conclusion:** Apremilast, methotrexate, and NB-UVB phototherapy are effective and well-tolerated treatments in patients with moderate-to-severe psoriasis. Apremilast shows a better ability to modulate systemic inflammation with a more transversal action due to its both anti-inflammatory and immunoregulatory activity.

Conclusion: Our results emphasize the importance of specific education and multidisciplinary follow up for this group of high-risk patients.

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Abstract N°: 3251

Exploring disease severity and psychosocial factors in severe cases of lichen planus: a prospective monocentric cohort study

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Introduction & Objectives:

Lichen planus (LP) is an inflammatory condition that affects the skin, mucosa, and skin adnexa caused by cytotoxic T cells and antigen-specific mechanisms. It affects around 1.3% of the population globally, more commonly in women. Esophageal LP (ELP), often overlooked, can impair swallowing and cause eating discomfort.

Materials & Methods:

We conducted a prospective cohort study involving multilocular LP patients from 2020 to 2023 at the Department of Dermatology and the Department of Gastroenterology at the University Hospital Freiburg (Germany). We gathered comprehensive data from all LP patients presenting with dysphagia and suspected ELP. Data collected included demographics, weight, smoking, clinical manifestations, endoscopy, histopathology, and questionnaires on quality of life (DLQI), depression (PHQ9, GHQ12), and swallowing problems (dysphagia symptom questionnaire, Eckhard score). The initial cohort had 75 patients, of whom 14 were excluded because there was no confirmed diagnosis of LP and 14 patients had missing data.

Results:

The study enrolled 47 patients with an average age of 62.4 years. 72 % were female and they had an average BMI of 25.3 kg/m^2. The most common manifestations were oral LP (87%), followed by skin (51%), hair and esophageal (both 45%), genital (36%), nail (30%), perianal (13%), and one case of eye involvement. In patients with esophageal involvement, endoscopic findings were stenosis (57 %) and the need for dilation (24 %), trachealization (43 %), mucosal denudation (33 %), hyperkeratosis (33 %).

We aimed to identify key questions to determine a high probability of endoscopic esophageal involvement in this cohort. Three questions showed significance ($p<0.05$): 1) 71 % with ELP had heartburn compared to only 33% without. 2) 52% with ELP had food getting stuck requiring endoscopy, while none without ELP had this issue. 3) 43 % with ELP had swallowing problems every meal, compared to 4 % without.

Patients with skin involvement had the highest mean DLQI of 9.69 (oral LP: DLQI of 7.63). At the same time, 15% of the patients with skin involvement had a unremarkable DLQI (0). In contrast, the involvement of the oesophagus did not allow any conclusions to be drawn in DLQI in 77%. Regarding risk for depression: 62 % of patients had abnormal GHQ12 scores with genital involvement significantly correlated. Patients with pathological GHQ12 were younger (mean 59 versus 68 years), with more oral inflammation and emotional issues. Regarding depression (PHQ9), 44.68% had mild and 38.29% moderate depression, with 3 severe cases, although only 2 patients had a documented history of depression. Those with PHQ9 ≥15 (moderate/severe depression) were more likely to have oral erosions and reported greater eating difficulties and emotional distress.

Conclusion:

In this cohort, ELP occured in 45% of all patients and is thereby among the most common manifestation sites of LP. Dysphagia, heartburn and esophageal bolus obstructions should trigger endoscopy to determine esophageal involvement. ELP presents with endoscopic characteristics such as mucosal denudation and trachealisation and can lead to stenosis and hyperkeratosis.

The DLQI highlighted impacts of cutaneous and oral but not esophageal lesions. Most patients showed mild to moderate depression despite only two documented diagnoses. Oral and genital manifestations significantly affected quality of life and emotional well-being, indicating an unmet need for psychosomatic care.

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Abstract N°: 3261

Calciphylaxis, a diagnostic and therapeutic challenge

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Introduction and Objectives:

Calcyphilaxis, or uremic calciphylaxis, is a rare condition, mainly affecting patients with chronic kidney disease treated with dialysis. It is a rapidly progressive and life-threatening disease that clinically presents with persistently painful, ulcerative, or necrotizing skin lesions in multiple parts of the body. **

Materials and Methods:

We describe the case of a 49-year-old woman with a medical history of hypertension, heart disease, and chronic renal failure who was on vitamin K antagonist and vitamin D supplementation and presented multiple chronic, painful, and necrotic ulcers in her left leg that evolved 3 months prior to her consultation. Complete blood tests, including parathyroid hormone and cryoglobulin dosage, coagulation parameters, and auto-immune antibodies, were performed in addition to an x-ray of the leg and a skin biopsy.

Results:

Laboratory test results revealed blood calcium levels at 109 mg/L, elevated parathyroid hormone at 2484 ng/L, coagulation parameters, and auto-immune tests were normal. X-rays showed the presence of calcifications, and on the skin biopsy, we found epidermal necrosis and ulceration, as well as hypodermal lesions of the intima and media of small- to medium-caliber vessels with mediacalcosis.

Conclusion:

Calciphylaxis is a multifactorial disease; it is a deadly, painful disease with a 1-year mortality rate of up to 50%. It's current diagnosis is mainly based on high-risk factors such as renal failure, calcium and phosphorus metabolism disorders, hyperparathyroidism, characteristic skin lesions such as skin erythema, purpura, livedoreticularis, and ulcers, and features of skin histopathology. Treatment is largely based on observational studies and clinical expertise. Our patient had many risk factors. The therapeutic decision was to stop vitamin D supplementation, add calcium chelators in addition to surgical debridement, optimize dialysis parameters, and propose a parathyroidectomy.

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Abstract N°: 3297

Multicentric reticulohistiocytosis with cutaneous features resembling dermatomyositis

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Introduction

Multicentric reticulohistiocytosis (MRH) is a rare form of histiocytosis characterized by both erosive polyarthritis and the presence of papulonodular skin lesions. Occasionally, the typical cutaneous manifestations may be accompanied by other manifestations reminiscent of those of dermatomyositis (DM), such as Gottron's papules and rashes with a light-exposed distribution, such as the V-neck sign and the shawl sign.

Clinical Case

A 54-year-old woman, with no previous medical history, consulted for skin lesions on the hands, face, trunk and upper extremities of 4 weeks' evolution. The patient also had symptoms of polyarthritis, asthenia and proximal muscle weakness. The lesions were asymptomatic and located on the dorsum of the hands and on the interphalangeal joints and the lateral aspect of the fingers. In the periungual region these papules acquired a distribution in 'coral pearls'. They were shiny, firm to palpation, papules and erythematous-violaceous nodules. Similar lesions were also found on the ala of the nose and the inner canthus of the eyes. She also exhibited a V-shaped maculopapular rash on the anterior and posterior trunk and around the shoulders. In the biopsy, cells with eosinophilic cytoplasm and histiocytic appearance were predominant. Numerous multinucleated giant cells with pale frosted cytoplasm were also seen. Immunohistochemical studies showed positivity for CD-68 and CD-163 and negativity for S-100 protein and CD-1. Based on the histology of this nodule, MRH was diagnosed. Laboratory tests, including muscle enzymes and antinuclear antibody profile, were normal. A body CT scan was ordered, which showed no evidence of associated neoplasia. It was decided to start treatment with non-steroidal anti-inflammatory drugs, alendronate, prednisone and methotrexate 15mg weekly. The patient reported improvement in skin symptoms, joint pain and muscle weakness 8 weeks after starting treatment.

Discussion

MRH is a rare multisystem disorder within the group of non-Langerhans cell histiocytosis. It is slightly more common in females and usually debuts around the fourth decade of life. Arthritis associated with MRH can worsen rapidly and be disabling. The typical skin lesions are firm, shiny, reddish brown papules and nodules. They usually appear on the backs of the hands and fingers and take on a special appearance in the periungual area that is described as "coral pearls". Cases of MRH have been described with an erythematous rash on sun-exposed areas and Gottron papule-like lesions giving a clinical appearance similar to DM. However, there are differences: the rash in MRH consists mainly of papules, whereas in DM the macular erythematous-violaceous rash is more characteristic. Hand papules in MRH are found on the dorsal and lateral sides of the fingers and nail folds. In DM, Gottron's papules are more prominent on the knuckles. In addition, DM is associated with proximal muscle weakness, whereas arthritis is the main feature of MRH. Histopathological examination of these lesions shows a nodular infiltrate composed of histiocytes and multinucleated giant cells with eosinophilic, granular, ground-glass cytoplasm. Approximately one quarter of MRH cases are associated with an underlying malignancy, most commonly breast, haematological or gastric cancer. Treatment options include oral glucocorticoids, immunomodulatory drugs, TNF inhibitors and JAK inhibitors, either in monotherapy or in combination.

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Abstract N°: 3324

Sarcoidosis masquerading as Hodgkin Lymphoma: a remarkable case of the great mimicker in Dermatology

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Introduction & Objectives:

Sarcoidosis is a systemic granulomatous disorder that most commonly affects the lungs, the lymph nodes and the skin. Cutaneous manifestations of sarcoidosis are seen in one-third of patients and may be the first and only clinical sign of the disease. Sarcoidosis is considered one of the great mimickers in Dermatology and its diagnosis is frequently difficult as it may present with a wide array of polymorphous cutaneous manifestations.

Materials & Methods:

We report the case of systemic sarcoidosis masquerading as Hodgkin Lymphoma that imposed a thought-provoking diagnosis.

Results:

A 31-year-old Caucasian female, Fitzpatrick's phototype III, with no relevant comorbidities, presented to our dermatology outpatient department with a one-year history of weight loss, intermittent low-grade vespertine fever, night sweats, anorexia, arthralgia, and gradually growing bilateral neck masses, alongside asymptomatic brownish cutaneous lesions. The patient denied any other symptoms.

On physical examination, there were multiple, non-tender, firm conglomerate lymphadenopathies, on the neck, axillae and inguinal regions, together with erythematous-to-brown, non-tender, firm papules and plaques on the face, upper limbs and trunk. Upon diascopy, the cutaneous lesions had an "apple jelly" appearance. The diagnostic hypotheses of Hodgkin Lymphoma, Sarcoidosis, and Tuberculosis were considered.

Thorough investigation was initiated with the collaboration of the Internal Medicine Department. Laboratory examination revealed leukopenia, thrombocytopenia, hypercalcemia, and elevated angiotensin-converting enzyme, and was otherwise unremarkable. Infectious diseases were excluded, including tuberculosis. Full-body CT scan revealed supra and infradiaphragmatic lymphadenopathy and hepatosplenomegaly. Gallium-67 scintigraphy unraveled a panda pattern and localized cutaneous and hepatosplenic uptake. Cutaneous biopsies showed numerous non-necrotizing epithelioid granulomas, with no surrounding lymphocytic inflammation ("naked" granulomas), on the full-thickness of the dermis and subcutis, and no microorganisms on PAS and Ziehl-Neelsen stainings. These findings were consistent with cutaneous sarcoidosis. Lymph node biopsies were compatible with sarcoidosis and excluded lymphoma and tuberculosis. The patient was treated with methotrexate and prednisolone, with significant improvement.

Conclusion:

Sarcoidosis presents a diagnostic conundrum in Dermatology due to its clinical pleomorphism and potential mimicking of other conditions, such as Hodgkin Lymphoma. Given the diverse and often subtle presentation of sarcoidosis on the skin, dermatologists are at the forefront of recognizing these manifestations and initiating the diagnostic process. This case highlights the importance of a multidisciplinary collaboration in these cases, which is pivotal in achieving accurate diagnosis and optimal management for patients, ultimately improving their quality of

life.

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Abstract N°: 3340

Aquagenic wrinkling of the palms in a patient with cystic fibrosis

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Introduction & Objectives:

Materials & Methods:

Results:

Aquagenic wrinkling of the palms (AWP), also known as aquagenic palmoplantar keratoderma, is a rare dermatological condition characterized by transient edematous white or translucent papules on the palmar surfaces, following brief immersion in water. Although most cases are related to cystic fibrosis (CF), drugs, hyperhidrosis and atopic dermatitis can also be associated.

A 16-year-old female patient, recently submitted to a lung transplant due to CF, presented to our department with asymptomatic palmar lesions, with five months of evolution. She reported palmar hyperhidrosis and worsening of lesions after water exposure. Physical examination revealed whitish and hyperkeratotic papules on both palms. The right hand was immersed in water for 5 minutes, followed by worsening of palmar lesions with evident wrinkling and edematous whitish papules – "hand-in-bucket sign". Thus, a diagnosis of AWP was established and treatment with topical aluminum chloralhydrate was initiated.

AWP association with CF is well established, occurring in 41-80% of CF patients and 25% of CF carriers. Although the pathophysiology of AWP is incompletely understood, the increased tonicity of the sweat and the upregulation of aquaporins in keratinocytes of CF patients are possible explanations. The diagnosis of AWP is mainly clinical, based on the pathognomonic "hand-in-bucket" sign.

Conclusion:

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Abstract N°: 3366

Clinical Efficacy of a New Moisturizer Containing Ceramides and Natural Moisturizing Factors on Subjects with Extra Dry, Itchy Skin

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Introduction & Objectives:

Xerosis is characterized by a diminished capacity of the skin to retain moisture that clinically presents as rough, tight, flaky, and pruritic skin dependent on condition severity. Xerosis is often attributed to a decrease in key stratum corneum lipids, namely ceramides, leading to a decline in skin barrier function as well as a decrease in natural moisturizing factors (NMFs) resulting in reduced water binding capacity. Effectively addressing symptoms, restoring skin barrier function, and preventing the progression of xerosis is important to manage impact to quality of life. This study assesses the efficacy of a moisturizer containing 3 skin identical ceramides and NMFs for clinical reduction of xerosis symptoms and improvement in skin barrier function, hydration, and NMF composition.

Materials & Methods:

A 4-week, single-center clinical study (USA) was conducted on 90 men and women aged 19-75 years with Fitzpatrick skin type I-VI. Subjects presented aggravated, uncomfortable skin with moderate to severe dryness, flakiness/scaling, itching, rough skin texture, and erythema on the global arms, legs, knees, and/or elbows. Subjects who met clinical criteria for leg dryness also required baseline transepidermal water loss (TEWL) >10 g•m-2•h-1 (RG, Cyberderm) and corneometer < 80 units (Dermalab, Cortex Technologies). All subjects applied the ceramide- and NMF- containing cream at least twice daily for four weeks followed by a 3-day regression period when no test product was applied. Clinical scoring for extra dry skin symptoms were conducted at immediate, Day 1, Day 3, Week 1, Week 4, Regression (+3 days) through dermatologist grading on a 5-point ordinal scale with clinical images. Instrumental TEWL and hydration assessments were performed on the legs of subjects throughout the duration of the study. Participant self-perception was assessed at all timepoints.

Results:

There was a statistically significant ($p < 0.001$) improvement in dryness, flakiness/scaling, itching, rough skin texture, and erythema on the global arms, legs, knees, or elbow at all time points. Statistically significant improvement in TEWL and hydration ($p \leq 0.002$) was achieved at immediate, Day 1, Day 3, Week 1, Week 4, and Regression (+3 days) (TEWL: -39%, -37%, -40%, -42%, -45%, and -13% respectively; Hydration: 107%, 105%, 124%, 121%, 129%, and 17% respectively). The test cream was well tolerated. Participants perceived the test product to be gentle and effective.

Conclusion:

The ceramide- and NMF-containing moisturizer provided significant clinical and subject-perceived instant and long lasting relief (Regression +3 days) of xerosis symptoms for all body zones studied. The moisturizer fully repaired skin barrier, provided hydration, and effectively treated xerosis while improving skin's capacity to retain moisture.

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Abstract N°: 3375

Acquired epidermodysplasia verruciformis in a child

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Introduction & Objectives:

Materials & Methods:

Results:

Epidermodysplasia verruciformis (EV) is a rare autosomal recessive genodermatosis characterized by a defect in cell-mediated immunity and increased susceptibility to infections by β-human papillomavirus (HPV) subtypes. In the last decade, an acquired EV-like syndrome was described in immunocompromised patients, including transplantation, hematologic malignancies, systemic lupus erythematosus and HIV.

A 9-year-old female patient with a recent diagnosis of HIV and Hepatitis B infection, presented to our department with an asymptomatic skin eruption with 5 years of evolution. Physical examination revealed multiple hypopigmented flat-topped papules ranging in size from 3 to 5 mm, distributed over the face, upper trunk, arms and dorsal surface of both hands. Histopathological examination revealed focal acanthosis and hyperkeratosis as well as enlarged keratinocytes with bluish-grey cytoplasm and vacuolated nuclei. A diagnosis of acquired EV was established and treatment with isotretinoin 10mg was initiated.

This clinical case describes the typical presentation of acquired EV, with disseminated flat-top hypopigmented papules, resembling pityriasis versicolor, in an HIV-infected child. However, the histopathological findings are essential for the diagnosis, revealing the characteristic enlarged keratinocytes with bluish-grey cytoplasm and vacuolated nuclei. HIV-associated EV is a rare disease, although it appears to be more prevalent in vertically infected children than in adults. The progression of cutaneous lesions is not influenced by the immunological status of the patient and the response to treatment is poor. Therefore, close surveillance for cutaneous malignancy is required.

Conclusion:

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Abstract N°: 3387

Epidemiology, Clinical Characteristics, and Treatment of Moderate to Severe Pyoderma Gangrenosum: A 5-Year Case Series Study

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Introduction & Objectives:

Pyoderma gangrenosum (PG) is a rare, chronic, and painful skin disease classified among neutrophilic dermatoses. PG typically affects women aged between 20-50. Clinically is characterized by the appearance of pustules on an erythematous or violaceous base, evolving into ulcers with necrotic or undermined borders. PG is often associated with systemic diseases, as inflammatory bowel disease, arthritis or hematologic disorders. First-line treatments include systemic corticosteroids and cyclosporine, followed by TNF-α inhibitors. Emerging treatments include IL-1, IL-17, or IL-23 inhibitors, particularly for patients with underlying inflammatory comorbidities.

The objective of this study was to describe the epidemiologic and clinical characteristics, as well as the treatments administered in cases of moderate to severe PG treated over a 5-year period in the Dermatology Department of our hospital.

Materials & Methods:

An observational, descriptive and retrospective study was conducted. Patients diagnosed with moderate-severe PG at the Ramón y Cajal hospital in Madrid in the last 5 years (2018-2024) were reviewed. Patients were selected whether they presented any of the following criteria: single ulcers larger than 3 cms, multiple ulcers, or ulcers allowing visualization of tendon, muscle or bone; as well as a biopsy compatible with PG.

Ten patients were selected. Sex, age at diagnosis, clinical presentation, associated pathologies, location and systemic treatments employed were collected.

Results:

Table 1 shows the results obtained.

Conclusion:

This case series is epidemiologically consistent with what has been published to date. A clear female predominance was found, although the mean age at diagnosis is slightly higher than in other series (57.6). Clinical features are aligned closely with existing findings, mainly classical ulcerative presentation, often on the lower extremities.

Most of the patients exhibited an autoinflammatory context, featuring various conditions like ulcerative colitis and hidradenitis suppurativa. Less common conditions as PAPASH syndrome or aseptic abscess syndrome were also noted. Hematologic disorders and solid tumors were present in one patient. Additional associated diseases included polyarteritis nodosa and autoimmune hepatitis.

In terms of treatments used, most of the cases were refractory to initial treatment with high doses of systemic corticosteroids, requiring the addition of an immunosuppressant agent or biologic treatment in 90% of the cases.

The most commonly immunosuppressant used was cyclosporine (50%). Adalimumab was prescribed in 30% of the cases, but other biologics as infliximab, ustekinumab, secukinumab, rituximab or anakinra were also required. The selection of these treatments was influenced by the specific patients' comorbidities, rendering them suitable candidates for such therapies. All patients were controlled with the prescribed treatment, although the response time was variable.

PATIENT	AGE	SEX	CLINICAL PRESENTATION	ASSOCIATED PATHOLOGIES	LOCALIZATION	SYSTEMIC TREATMENT
1	74	Female	Ulcerative	Obesity	Lower extremity	Prednisone, methylprednisolone
2	29	Female	Ulcerative	Obesity, ulcerative colitis	Lower extremity	Prednisone, methylprednisolone, cyclosporine, mercaptopurine, adalimumab
3	28	Female	Ulcerative (multifocal)	Ulcerative colitis	Lower extremity	Prednisone, cyclosporine, infliximab, adalimumab
4	55	Male	Postsurgical (partial hepatectomy)	Aseptic abscess syndrome, c-ANCA vasculitis	Trunk	Prednisone, methylprednisolone, rituximab
5	58	Female	Ulcerative	Hidradenitis suppurativa, psoriasis	Lower extremity	Prednisone, colchicine, ustekinumab
6	75	Male	Ulcerative	Trycholeukemia, colon adenocarcinoma, cavum lymphoepithelioma, Waldenstrom macroglobulinemia	Acral (hand)	Prednisone, cyclosporine
7	51	Female	Ulcerative (multifocal)	PAPASH syndrome	Trunk & lower extremity	Prednisone, adalimumab, ustekinumab, infliximab, anakinra, secukinumab
8	70	Female	Ulcerative	Obesity	Lower extremity	Methylprednisolone, immunoglobulins, cyclosporine
9	59	Male	Postsurgical (coronary revascularization)	Cutaneous polyarteritis nodosa	Trunk & lower extremity	Prednisone, cyclosporine, infliximab, ustekinumab, mycophenolate mofetil, immunoglobulins.
10	77	Female	Ulcerative	Autoimmune hepatitis	Lower extremity	Prednisone, azathioprine

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Abstract N°: 3395

Erythema nodosum- retrospective study of 79 patients

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Introduction & Objectives:

Ertythema nodosum (EN) is a common form of panniculitis, characterized by mostly symmetrically located painful, inflamed nodules, on the anterior surface of lower extremities. There is a clear female predominance, women being affected three to six times more. The lesions resolve spontaneously in a few weeks, without atrophy or scarring. The exact cause of the disease is unknown; it is considered that EN occurs as type IV delayed hypersensitivity response to numerous antigens. In about 30-70% of cases, no obvious etiology is found and the condition is considered idiopathic.

Materials & Methods:

A retrospective study encompassed 79 hospitalized** patients with Erythema nodosum* in a five-year-period* at our Clinic. For each patient we analyzed demographic data (age and sex), radiological findings (chest X ray), microbiological findings (bacteriological swabs of the throat and nose, coproculture, urine culture), laboratory tests in the majority of patients (antistreptolysin titer (ASL), levels of angiotensin converting enzyme, chitotriosidase and fecal calprotectin; Quantiferon TB Gold test, immunological and serological analyses) as well as the use of certain drugs.

Results:

The average age of patients was 48 years. We found women more commonly affected, with a female to male ratio 5.07: 1 (66 women and 13 men). No obvious cause was recorded in 11 (13.9%) while various infectious agents were found in 44 (55.7%) of patients. Most common infections were bacterial in 37/44, and viral infections in 7/44 patients. Elevated antistreptolysin titer was recorded in 30 (38%). Perihilar limphadenopathy was recorded in 19 (24.1%) of patients, in 16 of them pulmonary sarcoidosis was confirmed with further diagnostic (one female patient diagnosed with Sarcoidosis- Lofgren syndrome). Quantiferon TB Gold test was positive in 22/54 patients. In our study, two patients had co-occurrence of EN and Sweet syndrome (one of them had elevated ASL, the other elevated fecal calprotectin). There were two pregnant women, in their second trimester and the use of oral contraceptives was noticed as the most probable cause of EN in one female patient.

Conclusion:

In the majority of cases, a possible etiology is found, infectious agents being the most frequent cause. Sarcoidosis was recorded in similar percentage as in other studies. We have confirmed the higher incidence of Erythema nodosum in women.

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Abstract N°: 3414

Refractory Polymorphic Cutaneous Symptoms Unveiling Mammary Carcinoma: A Case of Paraneoplastic Dermatosis

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Introduction & Objectives:

The cutaneous presentations of systemic illnesses and malignancies, while significantly polymorphic, encompass a diverse array of manifestations. Typically, Paraneoplastic cutaneous rashes emerge concurrently with the underlying malignancy, although they may precede or succeed its onset. Insufficient recognition of the dermatological indicators of internal malignancies has the potential to impede the timely diagnosis and management of cancer.

Materials & Methods:

We present the clinical case of an 86-year-old female, who was referred to our department for a generalized pruritic cutaneous rash persisting for approximately one year antecedent to her consultation. The rash displayed a polymorphic nature characterized by erythematous papules, some encrusted with blood, distributed across the trunk and scalp, while exhibiting oozing clustered microvesicles on the upper and lower limbs.

Results:

During her visit in the dermatology department, the patient received systemic corticosteroids, antihistamines, and emollients, yet failed to exhibit a satisfactory response.

Given its clustered microvesicular manifestation, dermatitis herpetiformis was initially considered as a potential diagnosis. Acknowledging the established correlation between coeliac disease and dermatitis herpetiformis, an array of laboratory investigations and a biopsy were conducted, ultimately negating the aforementioned diagnosis. Concurrently, during physical examination, a palpable tumoral mass was detected in the right breast region. Subsequently, the patient underwent comprehensive imaging via whole-body computed tomography (CAT SCAN) and assessment by the gynecology department, confirming the diagnosis of mammary carcinoma. Consequently, considering the polymorphous nature and reluctance to treatment, the dermatological diagnosis was revised to reflect a paraneoplastic cutaneous manifestation.

Conclusion:

In summary, this case elucidates the intricate and diverse spectrum of cutaneous presentations associated with systemic illnesses and malignancies. Despite therapeutic interventions, the patient’s rash remained refractory, prompting investigation that unveiled an underlying mammary carcinoma. This underscores the imperative of discerning dermatological cues as putative indicators of internal neoplastic processes. Timely identification and management of paraneoplastic cutaneous manifestations are pivotal for optimizing patient prognoses and expediting the initiation of oncological interventions.

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Abstract N°: 3496

Acquired Pachydermatoglyphia: a Rare Sign Not to be Missed

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Introduction & Objectives:

Acquired Pachydermatoglyphia or Tripe palms, a term coined by Jacqueline Clarke, is a rare cutaneous manifestation often associated with internal malignancies.

Report of a Case:

A 58-year-old man with active smoking habits and a 100 pack year smoking history, presented to the emergency department with a 6-month history of weight loss (10% of previous body mass), asthenia and night sweats. Physical examination showed clinical signs of respiratory distress, significant cachexia, supraclavicular adenopathies and thickened velvety palms with pronounced folds, which the patient reported for the last 2 months.

A thoracoabdominopelvic computed tomography scan revealed an extensive parenchymal consolidation on the right inferior lung lobe lobe accompanied with air bronchogram. Biopsy of the lesion was compatible with acinar adenocarcinoma of the lung. A biopsy from the palm showed a strong compact hyperkeratosis, irregular acanthosis, hypergranulosis and a diffuse lymphocytic infiltrate with increased mast cell numbers papilomatose.

Conclusion:

Tripe palms, originally coined in the literature by Clarke in 1977, is characterized by the enhancement of the epidermal ridges of the palms, which become slightly thickened and acquire a velvety texture, resembling the intestinal villi. In 90% to 95% of cases this condition is associated with the presence of a solid tumor, the most common of which are those of the gastrointestinal tract (30%) and lung (20%). This paraneoplastic syndrome may occur on its own, which is commonly associated with lung carcinoma. When it coexists with malignant acanthosis nigricans, the most frequently associated cancer is gastric carcinoma.

Epidermal growth factor (EGF) has been implicated to mediate the clinical changes seen in tripe palms. FGF is an embryonic growth factor which is important in mammalian development and function. EGF receptors are found in keratinocytes, especially in the basal layer of the epidermis, and the number of receptors is increased in hyperproliferative skin disorders. Autocrine secretion of an EGF from the tumor cells is probably related to the increased plasma levels seen in these patients. A decrease in EGF levels following tumor treatment may be related to the improvement seen in the the skin lesions, suggesting a direct role of EGF in the pathogenesis of triple palms.

This paraneoplastic dermatosis may be found prior to or at the time of diagnosis of the primary malignancy, or rarely, further ahead in time. As such, timely recognition of tripe palms is crucial to ensure opportune neoplastic diagnosis and consequently improve health outcomes.

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Abstract N°: 3552

Red herrings in the examination of the skin: a retrospective cohort study on the diagnostic associations of the Venous Garland of Sahli.

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Introduction & Objectives:

Dermatologists often find themselves at the forefront of diagnosis due to the nature of cutaneous manifestations of systemic disease; however, they must also be aware of possible red herrings in diagnosis. One such example is the Venous Garland of Sahli (VGS), or Sahli-Gefäßgirlande, a relatively common and distinctive pattern of telangiectasias on the thorax. This phenomenon has traditionally been linked to respiratory diseases such as emphysema in German-language medical teaching but, interestingly, is poorly characterized in English-language literature – with only two mentions in translated articles listed by English literature indices. It was first described by Swiss Physician Hermann Sahli, originally in an 1885 letter but formally characterised in his 1905 textbook on examination.

Materials & Methods:

We undertook a retrospective analysis of a cohort of 872 participants in the Australian Centre of Excellence in Melanoma Imaging & Diagnosis (ACEMID) trial that had received total body photography (TBP) at regular intervals over 24 months. Images from all patients were reviewed, and those that had an appearance suggestive for VGS were interviewed.

Results:

Review of these images, alongside participant interviews, revealed presence of VGS in 23 individuals (4 female, age range 33-88 years old), a prevalence of 2.6%. Four anatomical sites of the VGS phenomenon were identified based on anatomical location: parasternal, anterior inferior thoracic aperture, posterior intrascapular, and posterior midback. These anatomical locations are documented with high quality imagery, and persistent across 24 months of TBP. Respiratory disease was seen at a slightly, and non-significantly, higher rate in the VGS cohort compared to the non-VGS participants: 8.7% (n=2) vs 2.2% (n=19); p=0.11. Importantly, no participants with VGS reported a history of chronic obstructive pulmonary disease or emphysema. No other medical comorbidity demonstrated correlation with VGS.

Conclusion:

Contrary to traditional beliefs, our findings suggest a low correlation between VGS and chronic respiratory disease. While we find no association between VGS and systemic disease it is still not unreasonable for a dermatologist, upon encountering the appearance, to suggest a patient follow up with their general practitioner for assessment.

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Abstract N°: 3592

Skin and thyroid: A prospective descriptive study of 131 patients.

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Introduction & Objectives:

The skin is the mirror of the human body, reflecting our lifestyle as well as other internal pathologies that may be tumoral, metabolic, endocrine and more.

What about the thyroid and its link with skin pathologies? What is its mechanism of action? What are its main cutaneous manifestations?

Materials & Methods :

To answer this question, we carried out a prospective study spread over a one-year period, from June 2022 to June 2023, in the dermatology-venereology and endocrinology department at the Mohammed VI University Hospital in Marrakech.

Our study has a descriptive aim, including all patients of all age groups with dysthyroidism and excluding all patients with other endocrinopathies in euthryoidism.

Results:

The total number of patients with dysthroidism was estimated at 131, with a female predominance in the 25-29 age bracket, and hypothyroidism estimated at 61.30%.

Skin involvement was estimated at 61.83% of our sample, or 81 cases, classified into several categories in descending order:

\1. Epidermal involvement (61.70%), including cutaneous xerosis and palmoplantar keratosis.

\2. Association with autoimmune pathologies (39.50%), including lupus, bullous dermatoses, urticaria and chronic pruritus.

\3. Pigment disorders (27.16%), including segmental and extensive vitiligo, as well as hyperpigmentation.

\4. Capillary involvement was estimated at 23.45%, including telogen effluvium, alopecia and early-onset canitis.

\5. Nail involvement was estimated at 2.46%, including cases of onycholysis and onychomadesis.

Discussion :

There are three main mechanisms by which thyroid hormone acts on the skin. The first is the direct action of thyroid hormones on cutaneous and non-cutaneous tissues, and the second is their association with autoimmune diseases.

The first action is mediated by the thyroid hormone receptor, which is usually found in several skin structures, explaining the different types of damage: Epidermal, dermal, phanerian, which differ depending on whether we’re dealing with hypothyroidism or hyperthyroidism.

In hypothyroidism, epidermal damage is mainly characterized by a textural change, with dry, scaly skin. This is explained by the reduction in epidermal sterol synthesis caused by hypothyroidism.

There is also an association between pretibial myxedema and Hashimoto’s thyroiditis due to dermal proliferation of GAGs and hyaluronic acid, as well as an association between hypothyroidism and carotenoderma due to a defect in the conversion of carotene into vitamin A.

Nail disorders are in line with the results of our study, with cases of onycholysis, onychomadesis and the possibility of half-and-half nails.

Our results are also consistent with hair disorders, where there is a correlation between dysthyroidism and telogen effluvium, alopecia, canitis and fibrosing frontal alopecia.

Conclusion :

Given the increasing incidence of dysthyroidism, a dermatological examination should be carried out systematically in any patient with an endocrinopathy.

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Abstract N°: 3686

Efficacy and Safety of Vunakizumab in Moderate-to-Severe Psoriasis Across Different BMI Classifications

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Introduction & Objectives:

The efficacy of biologic therapies for psoriasis, may be affected by Body Mass Index (BMI) increase. Limited data has been reported on IL-17A inhibitors for the moderate-to-severe plaque psoriasis across different BMI classifications. Vunakizumab (SHR-1314), a humanized IgG1 monoclonal antibody targeting IL-17A, has partially disclosed phase III clinical trial data. This study evaluates the efficacy and safety of Vunakizumab in treating moderate-to-severe plaque psoriasis among various BMI categories in China.

Materials & Methods:

This post-hoc subgroup analysis is part of a randomized, double-blind, multicenter, placebo-controlled phase 3 trial (NCT04839016). Patients aged ≥18 years received either Vunakizumab 240 mg or placebo at weeks 0, 2, 4, 8. Since week 12, all patients received Vunakizumab every 4 weeks until week 52. Patients were categorized into three BMI groups: normal weight or below (<24 kg/m^2), overweight (24-27.9 kg/m^2), and obese (≥28 kg/m^2). The primary endpoint was achieving PASI 90 and sPGA 0/1 at week 12. Efficacy and safety through 52 weeks were compared across BMI groups.

Results:

461 patients received Vunakizumab treatment (76.4% male, mean age 41.7 years, mean Body Surface Area 34.45%, mean PASI score 22.23): 179 patients in normal weight or below group, 183 in overweight group, and 99 in obese group. Decrease in response rates to Vunakizumab was observed as BMI increased. During 12 weeks treatment, PASI 75 was reached by 97.2% (95% CI 93.6-99.1), 95.1% (95% CI 90.9-97.7) and 85.9% (95% CI 77.4-92.0) in the normal weight or below, overweight, and obese groups, respectively; PASI 90 by 84.9% (95% CI 78.8-89.8), 77.6% (95% CI 70.9-83.4), and 66.7% (95% CI 56.5-75.8); and PASI 100 by 45.8% (95% CI 38.4-53.4), 39.9% (95% CI 32.7-47.4), and 23.2% (95% CI 15.3-32.8). sPGA 0/1 rates were 83.2% (95% CI 76.9-88.4), 76.0% (95% CI 69.1-82.0), and 61.6% (95% CI 51.3-71.2). The median time to achieve PASI 90 was longest for obese patients (71.5 days, 95% CI 56-57), compared to the overweight (58 days, 95% CI 57-83) and normal groups (57 days, 95% CI 58-85). Compared to normal group, obesity group had lower PASI 75 (difference 11.3%, 95% CI 5.0-19.8; odds ratio [OR] 5.7, 95% CI 2.1-15.8), PASI 90 (difference 18.2%, 95% CI 7.9-29.2; OR 2.8, 95% CI 1.6-5.0), PASI 100 (difference 22.6%, 95% CI 11.0-33.1; OR 2.8, 95% CI 1.6-4.8) and sPGA 0/1 (difference 21.6%, 95% CI 10.8-32.7; OR 3.1, 95% CI 1.8-5.4) response rates (Table). Response rates through 52 weeks showed poorer outcomes in overweight patients (Figure). The incidence of treatment-emergent adverse events was similar across BMI groups.

Conclusion:

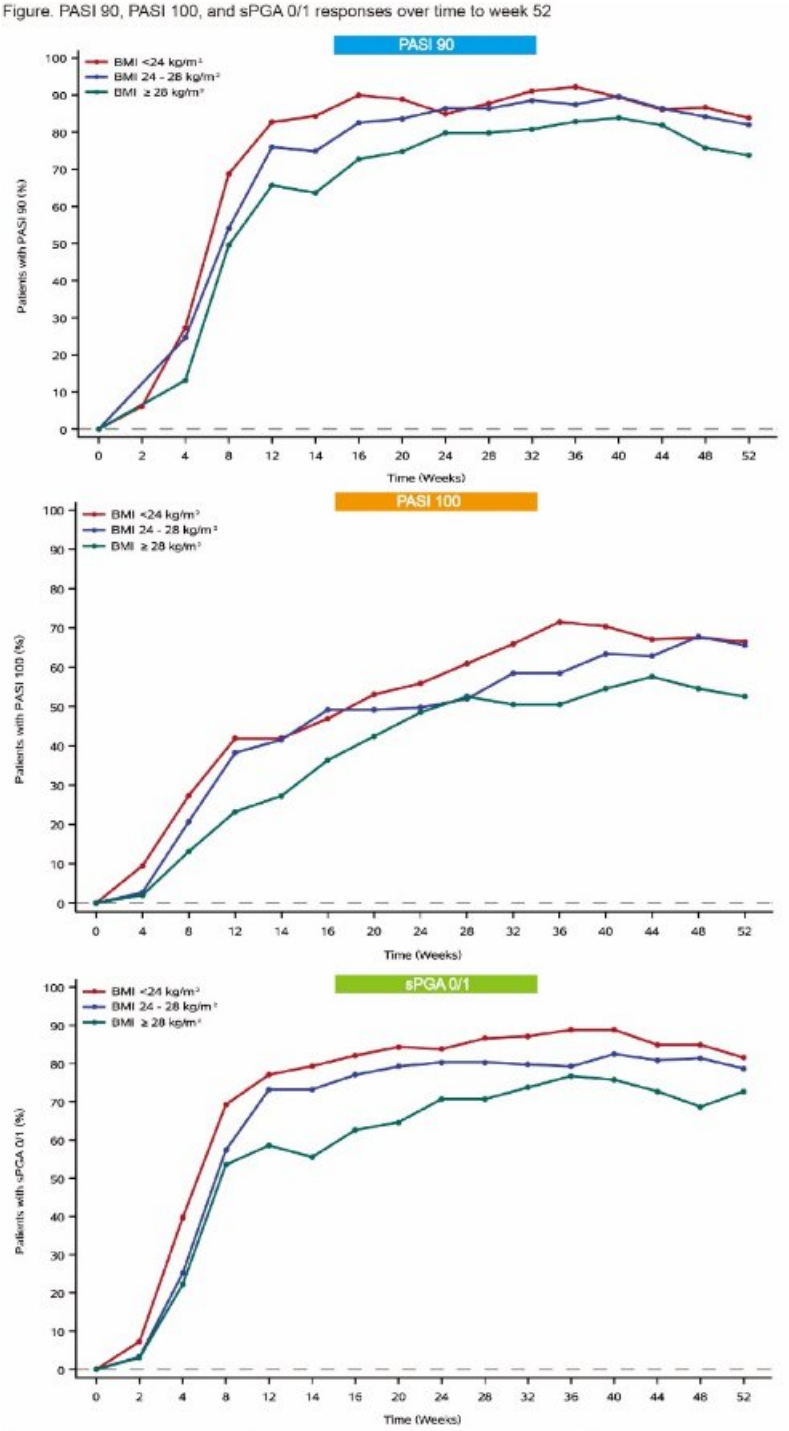
Vunakizumab showed superior efficacy in normal weight or below patients compared to overweight or obese patients over 52 weeks of treatment. Safety profiles were consistent across various BMI classifications, suggesting that higher doses may be needed for effective treatment in overweight and obese patients.

Table: PASI and sPGA Response in 12 Weeks Treatment

Efficacy outcome	Normal weight or below group (N=179)	Overweight group (N=183)	Obese group (N=99)
PASI 90			
Response, n (%) [1]	152 (84.9)	142 (77.6)	66 (66.7)
Response rate, % (95% CI) [2]	84.9 (78.8, 89.8)	77.6 (70.9, 83.4)	66.7 (56.5, 75.8)
Estimate treatment difference, % (95% CI) [3]		7.3 (-0.7, 15.4)	18.2 (7.9, 29.2)
OR (95% CI) [3]		1.6 (1.0, 2.8)	2.8 (1.6, 5.0)
sPGA 0/1			
Response, n (%) [1]	149 (83.2)	139 (76.0)	61 (61.6)
Response rate, % (95% CI) [2]	83.2 (76.9, 88.4)	76.0 (69.1, 82.0)	61.6 (51.3, 71.2)
Estimate treatment difference, % (95% CI) [3]		7.3 (-1.0, 15.6)	21.6 (10.8, 32.7)
OR (95% CI) [3]		1.6 (0.9, 2.6)	3.1 (1.8, 5.4)
PASI 75			
Response, n (%) [1]	174 (97.2)	174 (95.1)	85 (85.9)
Response rate, % (95% CI) [2]	97.2 (93.6, 99.1)	95.1 (90.9, 97.7)	85.9 (77.4, 92.0)
Estimate treatment difference, % (95% CI) [3]		2.1 (-2.1, 6.7)	11.3 (5.0, 19.8)
OR, % (95% CI) [3]		1.8 (0.6, 5.2)	5.7 (2.1, 15.8)
PASI 100			
Response, n (%) [1]	82 (45.8)	73 (39.9)	23 (23.2)
Response rate, % (95% CI) [2]	45.8 (38.4, 53.4)	39.9 (32.7, 47.4)	23.2 (15.3, 32.8)
Estimate treatment difference, % (95% CI) [3]		5.9 (-4.3, 16.0)	22.6 (11.0, 33.1)
OR (95% CI) [3]		1.3 (0.8, 1.9)	2.8 (1.6, 4.8)
Time to achieve PASI 90 within 12 weeks, days			
Median (95% CI) [4]	57 (56, 57)	58 (57, 83)	71.5 (58, 85)

[1] Percentage are calculated using the count of subjects in intention-to-treat set as denominator.
[2] Clopper-Pearson method is used to calculate 95% CI.
[3] Miettinen-Nurminen method is used to estimate treatment difference.
[4] Kaplan-Meier method is used to estimate median time to achieve PASI 90, with the corresponding 95% CI calculated by Brookmeyer-Crowley method.
PASI, psoriasis area and severity index; PASI 75, at least 75% improvement from baseline in PASI score; PASI 90, at least 90% improvement from baseline in PASI score; PASI 100, 100% improvement from baseline in PASI score; sPGA, static Physician's Global Assessment; OR, odds ratio. Normal weight or below included patients with Body Mass Index (BMI) <24 kg/m², overweight subgroup included patients with BMI 24-27.9 kg/m², and obese subgroup included patients with BMI≥28 kg/m².

Figure. PASI 90, PASI 100, and sPGA 0/1 responses over time to week 52



PASI, psoriasis area and severity index; PASI 90, at least 90% improvement from baseline in PASI score; PASI 100, 100% improvement from baseline in PASI score; sPGA, static Physician's Global Assessment.
Percentage are calculated in intention-to-treat set. Missing data were imputed as non-responses.

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Abstract N°: 3776

Zasocitinib (TAK-279), a highly selective oral tyrosine kinase 2 (TYK2) inhibitor, elicits early skin responses and minimal disease activity in patients with active psoriatic arthritis: results from a randomized phase 2b study

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Introduction & Objectives: TAK-279 is a highly selective, oral, allosteric TYK2 inhibitor. In a phase 2b trial in patients with active psoriatic arthritis (PsA; NCT05153148), significantly more patients treated with TAK-279 achieved American College of Rheumatology 20 responses at Week 12 than with placebo (PBO; 15mg [53.3%], 30mg [54.2%], PBO [29.2%]; p=0.002).1 Here, additional data on skin and overall disease activity are presented.

Materials & Methods: This was a randomized, multicentre, double-blind, PBO-controlled study. Eligibility criteria were reported previously.1 If on concurrent PsA treatments (permitted therapies were pre-defined in the study protocol), patients remained on stable doses throughout the study. Patients were randomized (1:1:1:1) to oral TAK-279 (5, 15 or 30mg) or PBO, once daily for 12 weeks. Outcomes included 75/90/100% improvements from baseline (BL) in psoriasis area and severity index (PASI 75/90/100), change from BL in PASI in patients with ≥3% psoriasis body surface area (BSA) at BL, Physician Global Assessment of PsO (PhGA-PsO) response (0/1 and a ≥2-point improvement from BL in patients with a PhGA-PsO score ≥2 at BL) and achievement of minimal disease activity (MDA). Differences were assessed using a Mantel–Haenszel test (binary endpoints) and a mixed model for repeated measures (continuous endpoints); p values were nominal.

Results: Overall, 290 patients were randomized. BL demographics and disease characteristics were similar between groups; 58.6% had psoriasis BSA ≥3% (mean [SD] baseline PASI, 6.2 [5.5]). At BL, 59.7, 63.4, 61.3 and 65.3% of patients were receiving concomitant traditional DMARDs in the PBO and TAK-279 5, 15 and 30mg groups, respectively. In the 30mg group, Week 12 PASI responses were substantially higher than with PBO or lower TAK-279 doses (**Figure 1A**). Reductions in least-squares mean (LSM) change from BL in PASI were greater for TAK-279 30mg than PBO from Week 2 (–1.1 vs PBO; 95% CI: –2.0, –0.2; p=0.023) and maintained through Week 12 (–2.5 vs PBO; –4.3, –0.8; p=0.004, **Figure 1B**). A substantially higher number of patients also achieved a PhGA-PsO response with TAK-279 30mg than with PBO at Week 12 (32.8% vs 15.8%; p=0.034, **Figure 2**). Higher doses of TAK-279 resulted in higher rates of MDA at Week 12 (15mg, 28.0%; 30mg, 29.2%; PBO, 12.5%; both p<0.05; **Figure 3**). TAK-279 was well tolerated with a safety profile consistent with that seen in the phase 2b psoriasis study.2

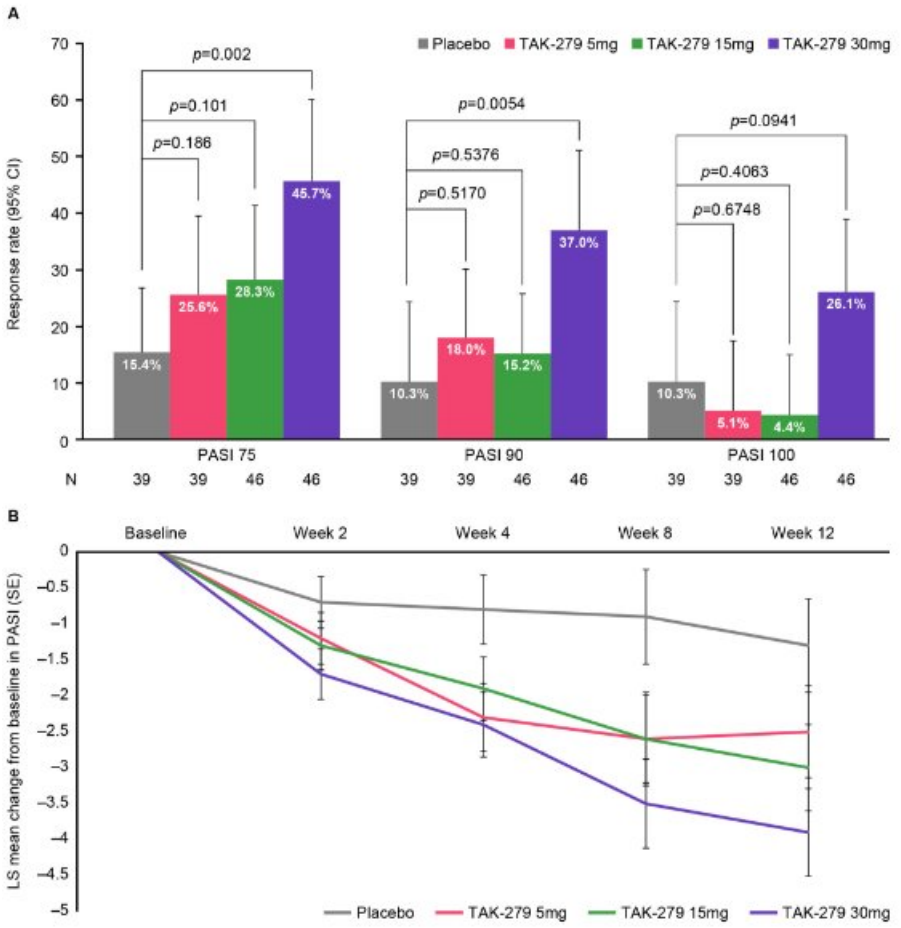
Conclusion: TAK-279 30mg demonstrated favourable safety and robust efficacy across all skin endpoints assessed versus PBO. Skin responses were evident from Week 2 onwards, and more patients treated with TAK-279 30mg achieved total or near-total clearance of psoriasis lesions at Week 12 versus PBO. Higher doses of TAK-279 (15 and 30mg) also achieved higher rates of MDA indicating meaningful improvement in other core PsA domains.

References

\1. Kivitz A, *et al.* Arthritis Rheumatol 2023;75(Suppl 9).

\2. Armstrong A, *et al.* AAD 2023.

Figure 1: PASI responses at Week 12 (A) and LS mean change from baseline in PASI (B) in patients with ≥3% BSA psoriatic involvement at baseline



Difference in LS mean change from baseline in PASI versus placebo (95% CI, *p* value)	Week 2	Week 4	Week 8	Week 12
Placebo				
5mg	−0.5 (−1.5, 0.4; *p*=0.282)	−1.5 (−2.8, −0.3; *p*=0.020)	−1.6 (−3.4, 0.2; *p*=0.080)	−1.2 (−2.9, 0.6; *p*=0.200)
15mg	−0.6 (−1.5, 0.3; *p*=0.203)	−1.1 (−2.4, 0.1; *p*=0.083)	−1.6 (−3.4, 0.1; *p*=0.064)	−1.6 (−3.4, 0.1; *p*=0.060)
30mg	−1.1 (−2.0, −0.2; *p*=0.023)	−1.6 (−2.8, −0.4; *p*=0.012)	−2.5 (−4.3, −0.8; *p*=0.005)	−2.5 (−4.3, −0.8; *p*=0.004)

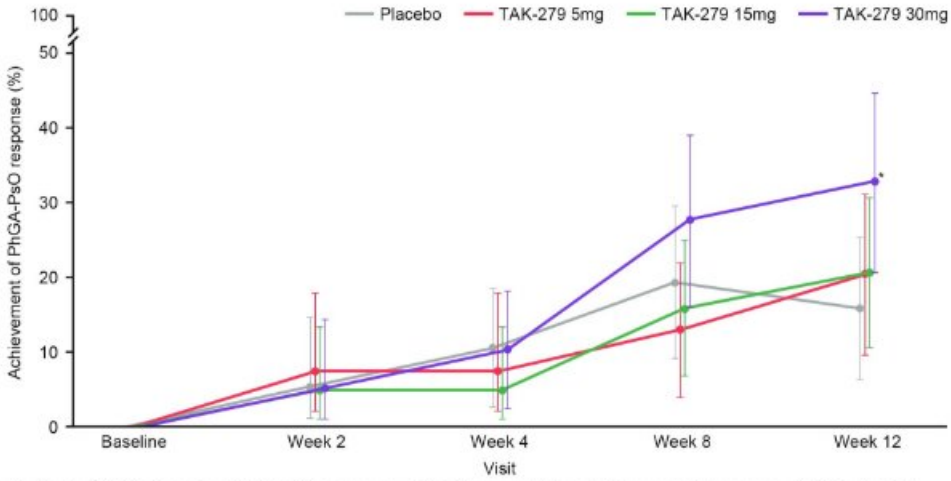
A. During the study, most patients were using concomitant medications, some of which were conventional DMARDs; this may explain the higher than expected PASI response in the PBO group. Comparisons of PASI 75 response rates at Week 12 were made independently between each dose group and the placebo group using two-sided Mantel–Haenszel tests stratified on randomization stratification factors (prior treatment with biologics or non-traditional DMARDs and region). Comparisons of PASI 90/100 responder rates at Week 12 were made independently between each dose group and the PBO group using two-sided Chi-square tests or the Fisher's exact method if the number of responders in any treatment group was <5. *p* values were nominal.

B. Point estimates, 95% CIs and nominal *p* values were obtained from a MMRM model that included fixed effects for treatment group, visit and treatment group-by-visit interaction, with baseline value and the randomization stratification factors (prior treatment with biologics or non-traditional DMARDs and region) as covariates.

PASI 90/100 responses and LS mean change from BL in PASI were analysed *post hoc*.

CI, confidence interval; DMARD, disease-modifying antirheumatic drug; LS, least-squares; MMRM, mixed model repeated measure; PASI, Psoriasis Area and Severity Index; PBO, placebo; SE, standard error.

Figure 2: Physician Global Assessment of PsO (0/1 and ≥2-point improvement from baseline)

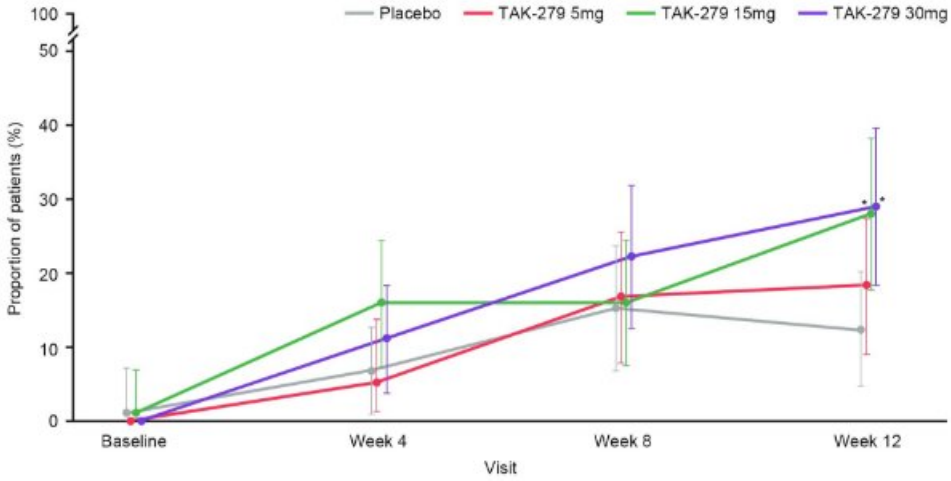


Whiskers show 95% CIs. Comparisons of PhGA-PSO response rates at Week 12 were made independently between each dose group and PBO using two-sided Mantel–Haenszel tests stratified on randomization stratification factors (prior treatment with biologics or non-traditional DMARDs and region).

*p=0.034 versus placebo. All p values were nominal.

CI, confidence interval; DMARD, disease-modifying antirheumatic drug; PBO, placebo; PhGA-PsO, Physician Global Assessment of Psoriasis.

Figure 3: Minimal disease activity responder rate



Patients were classified as achieving MDA when ≥5 of the 7 following criteria were met: tender joint count ≤1; swollen joint count ≤1; PASI score ≤1 or BSA ≤3%; Patient Global Assessment of PsA Pain VAS score ≤15; Patient Global Assessment of PsA VAS score ≤20; Health Assessment Questionnaire-Disability Index score ≤0.5; tender entheseal points, using LEI ≤1. Data are presented as the proportion of patients achieving MDA with 95% CIs.

Comparisons of MDA responder rates at Week 12 were made independently between each dose group and PBO using two-sided Mantel–Haenszel tests stratified on randomization stratification factors (prior treatment with biologics or non-traditional DMARDs and region).

*p<0.05 versus placebo. All p values were nominal.

BSA, body surface area; CI, confidence interval; DMARD, disease-modifying antirheumatic drug; LEI, Leeds Enthesitis Index; MDA, minimal disease activity; PASI, Psoriasis Activity and Severity Index; PBO, placebo; VAS, visual analogue scale.

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Abstract N°: 3779

pretibial myxedema - a case report

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Introduction & Objectives:

Pretibial myxedema (PM) is a rare extrathyroid condition seen in about 0.5 to 4.3 percent of individuals with hyperthyroidism due to Graves' disease, often presenting with associated thyroid orbitopathy.

In most cases, patients with PM have elevated levels of thyroid antibodies, such as thyroid peroxidase (TPO), thyroglobulin, and-most especially-thyroid-stimulating hormone receptor antibodies.

We report a case of pretibial myxedema with no clinical features of Graves' disease in an 63-year-old Pakistani male. The physical examination revealed multiple well defined waxy indurated plaques with prominent hair follicle openings and non-pitting edema disseminated on shins and knees of both legs. Lab workup shows hypothyroidism with very high TSH (>100) and decreased t3/t4 levels. Auto antibodies including anti thyroid peroxidase (TPO) and thyroglobulin antibody counts were elevated.

Histology from an edematous lesion revealed markedly thickened dermis with abundant mucin, especially hyaluronic acid, and the collagen fibers in this portion were splitting up into fibrils.

The case is reported for its rarity and unsual presentation.

Materials & Methods:

A 63-year-old Pakistani male presented with multiple skin swellings on both his legs of 6 months duration. on physical examination multiple well defined waxy indurated plaques with prominent hair follicle openings and non-pitting edema disseminated on shins and knees of both legs were noted.

Examination showed multiple shiny well-circumscribed, non-tender nodules /plaques that were firm to the touch on both lateral shins and knees almost covering the whole leg . There was no clinical hyperhidrosis, acropachy, or thyromegaly.

Lab workup revealed normal routine tests and hypothyroidism with very high TSH (>100) and decreased t3/t4 levels. Auto antibodies including anti thyroid peroxidase (TPO) and thyroglobulin antibody counts were elevated.

Histology from an edematous lesion revealed markedly thickened dermis with abundant mucin deposition and splitting of collagen bundles into widely separated fibers.

Laboratory examination results included a free serum triiodothyronine level of 2.3 pg/mL (normal range: 2.0–4.4 pg/ml), total triiodothyroinine level of 81 ng/dL (normal range: 71–180 ng/dL), free serum thyroxine level of 0.95 ng/dL (normal range: 0.82–1.77 ng/dL), thyroid-stimulating hormone level of 1.670

Results:

Based on these clinico pathological findings, the diagnosis of pre tibial myxedema was made. The patient was given intralesional steroid (kenacort) injection and topical moderate corticosteroids and oral antihistamines under

occlusion and the case is under followup.

Conclusion:

Pretibial myxedema (PM) is a rare thyroid associated condition seen in about 0.5 to 4.3 percent of individuals with hyperthyroidism due to Graves' disease, often presenting with associated thyroid orbitopathy. While it can even be seen in euthyroid rarely and very rarely in hypothyroid state.

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Abstract N°: 3954

Investigation of the Relationship between Seborrheic Keratosis and Insulin Resistance, Metabolic Syndrome and IGF-1 and IGFBP-3 Levels: A Case-Control Study

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Introduction & Objectives:

Seborrheic keratosis is the most common benign epidermal tumor of the skin. Its incidence increases with advancing age; however, the exact pathogenesis has not been fully enlightened. The relationship between seborrheic keratosis and insulin resistance or metabolic syndrome has not yet been investigated. In this study, we aimed to compare the risk of insulin resistance and metabolic syndrome, and serum IGF-1 and IGFBP-3 levels in seborrheic keratosis patients with the control group and to investigate tissue IGF-1 and IGBP-3 expression. We also investigated the possible correlations of dermoscopic features with these clinical and histopathological findings.

Materials & Methods:

The study included 100 patients with at least one seborrheic keratosis and an equal number of age- and sex-matched controls. Fasting insulin, fasting blood sugar (FBS), HbA1c, serum lipids, liver enzymes, serum IGF-1, and IGFBP-3 levels were measured in all subjects, and the Homeostatic Model Assessment of Insulin Resistance(HOMA-IR) was calculated. Anthropometric measurements were performed in all subjects and metabolic syndrome diagnosis was made using National Cholesterol Education Program Adult Treatment Panel III(NCEP ATP III) and International Diabetes Federation(IDF) criteria. One of the lesions from each patient was removed with shave excision after dermoscopic examination and tissue IGF-1 and IGFBP-3 immunohistochemical stainings were performed.

Results:

Fasting insulin levels and HOMA-IR values were significantly higher in patients than in controls, and an increased risk for insulin resistance was found in the patient group ($p<0.05$), but the negative correlation with age seen in the normal population disappeared for serum IGF-1 in the patient group ($p>0.05$). The total number of lesions was positively correlated with serum IGF-1 level ($r=0.250$, $p=0.012$). In immunohistochemical examinations, positive cellular staining for IGF-1 at the granular layer was observed in both lesional and the adjacent normal skin. Focal positivities with IGF-1 in lesion compared to adjacent normal skin were detected in one area in 23 cases (23.2%), in two areas in 19 cases (19.2%), and in three or more areas in 13 cases (13.1%). For IGFBP-3, all cases demonstrated nuclear immunoreactivity of equal intensity and there was no statistically significant difference. In dermoscopic examination, the existence of hairpin vessels was negatively correlated with fasting insulin level ($p=0.038$). The existence of fat fingers was negatively correlated with fasting insulin level, FBS and HOMA-IR ($p<0.01$).

Conclusion:

Patients with seborrheic keratosis have an increased risk of insulin resistance and metabolic syndrome, and investigation of insulin resistance might be useful in these patients. Considering the results we have obtained, IGF-

1 and IGFBP-3 may have a role in the pathogenesis of seborrheic keratosis and further studies are required to establish their exact pathophysiologic mechanism. Population based investigations are needed to validate the association between seborrheic keratosis and metabolic syndrome.

Table 1. Biochemical parameters of the patient group and the control group.

	Patient Group Mean ± SD	**Control Group** Mean ± SD	**Statistical Comparison**
HbA1c (%)	5,63 ± 0,34	5,68 ± 0,33	t=-0,833 / p=0,406
Insulin (fasting) (µIU/mL)	7,9 ± 4,8	6,2 ± 3,4	**z=-2,752 / p=0,006***
Glucose (fasting) (mg/dL)	92,9 ± 13,5	90,4 ± 8,7	z=-1,479 / p=0,139
HDL (mg/dL)	55,31 ± 16,1	57,1 ± 13,5	z=-1,546 / p=0,122
LDL (mg/dL)	136,7 ± 33,1	127,4 ± 32,8	**t=2,003 / p=0,047***
Total cholesterol (mg/dL)	209,5 ± 47,3	199,8 ± 44,8	t=1,491 / p=0,138
Triglyceride (mg/dL)	133,7 ± 90,8	116,7 ± 57,3	z=-1,211 / p=0,226
ALT (U/L)	23,5 ± 10,7	19,9 ± 9,5	**z=-2,567 / p=0,010***
AST (U/L)	22,7 ± 6,6	21,9 ± 6,7	z=-1,066 / p=0,286
ALP (U/L)	71,9 ± 21,6	68,8 ± 18,7	z=-1,025 / p=0,305
GGT (U/L)	29,12 ± 16,2	24,23 ± 21,8	**z=-3,336 / p=0,001***
IGF-1 (ng/mL)	109,12 ± 34,65	102,29 ± 29,84	t=1,493 / p=0,137
IGFBP-3 (µg/mL)	4,64 ± 1,27	4,26 ± 1,16	**t=2,213 / p=0,028***

SD: Standard deviation

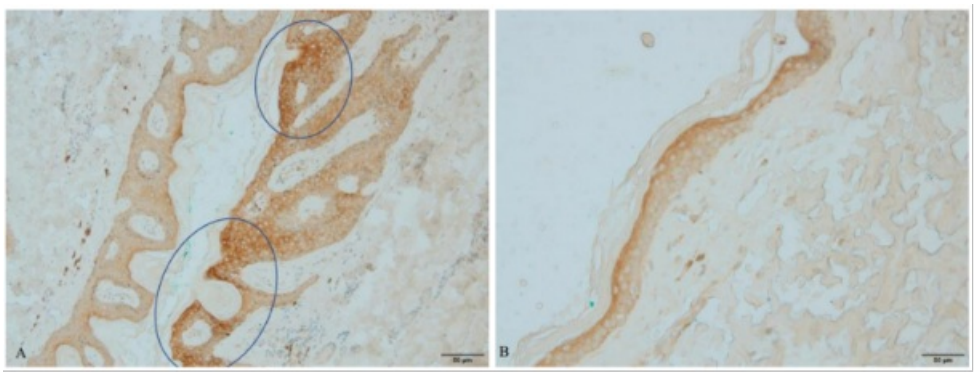

Figure 1. Seborrheic keratosis demonstrating focal positivity with IGF-1 in two areas (IGF-1,x200) (A), Adjacent normal skin showing positive cellular staining for IGF-1 at the granular layer, but no focal positivity with IGF-1 (IGF-1,x200) (B).

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Abstract N°: 3963

Panniculitis as a complication of endoscopic ultrasound-guided fine-needle aspiration biopsy of the pancreas

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Introduction & Objectives:

Pancreatic panniculitis is an uncommon complication of pancreatic disease (affecting 2% of patients with pancreatic disorders), which sometimes can manifest before the detection of the underlying pathology. Clinically, it presents as subcutaneous nodules that often develop on the lower limbs but can also affect the trunk, arms, and scalp. The aim of this poster is to describe a case of post-traumatic pancreatic panniculitis with systemic symptoms recently diagnosed in our dermatology department.

Materials & Methods:

We assessed a 65-year-old female patient who presented with painful erythematous nodules on the posterior aspect of both legs lasting for 4 days. Her medical history included undergoing an endoscopic ultrasound-guided fine-needle aspiration biopsy (EUS-FNA) of the pancreas 10 days prior to symptom onset, as part of the evaluation for suspected recurrence of a gastric GIST tumor with hepatic metastases previously treated with surgery and Imatinib.

Results:

Upon presentation to the Emergency Department, the patient also complained of worsening left hypochondrial pain over several months, leading to laboratory tests revealing elevated lipase levels of 7053 (upper limit of normal: 78) and amylase levels of 5491 (upper limit of normal: 125). Gastroenterology evaluation diagnosed abdominal pain with a probable pancreatic reaction due to manipulation. A punch biopsy of one nodule of the leg was performed, revealing histopathological features consistent with pancreatic panniculitis, including a mixed septal-lobulillar infiltrate, fat necrosis with ghost-like outline of fat cells, associated neutrophil infiltrate, and fine basophilic calcium deposits. Immunohistochemical study ruled out fungal infection.

In the days following the diagnosis, the patient developed ankle monoarthritis and febrile spikes. Medical oncology initiated treatment with Piperacillin-Tazobactam for 14 days and methylprednisolone, leading to resolution of fever and improvement in joint pain.

Currently, relapse of her GIST tumour due to pancreatic involvement has been confirmed, and a decision regarding treatment is pending from the hepatobiliarypancreatic tumor committee.

Conclusion:

The appearance of erythematous cutaneous nodules on the lower limbs may be the first manifestation of pancreatic alteration. These nodules can be associated with visceral fat involvement, and patients may present with fever, abdominal pain, inflammatory polyarthritis, ascites, and pleural effusions. Pancreatic alterations associated with this type of panniculitis include acute and chronic pancreatitis, pancreatic pseudocysts, traumatic pancreatitis

as in our patient’s case, or less commonly, pancreatic carcinoma. It has been demonstrated that elevated pancreatic enzymes (lipase, amylase, and trypsin), even in the absence of pancreatic disease, contribute to the formation of cutaneous lesions. Treatment is based on managing the underlying cause and addressing cutaneous lesions.

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Abstract N°: 4133

Challenging Diagnosis: A Unique Case of Destructive Lupus Panniculitis

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Introduction & Objectives: Lupus panniculitis (LP) is a rare condition that is challenging to diagnose, especially in the absence of concurrent systemic lupus erythematosus (SLE) or discoid lupus. We present a rare and destructive LP revealing SLE.

Materials & Methods: A 32-year-old woman had been presenting deep subcutaneous nodules on her left arm since the age of 13, which were warm, painful, and fistulized, leaving scars. The condition progressed with lesions extending to the back, buttocks, and thighs, accompanied by joint pains. Five skin biopsies were performed, revealing respectively: 1- Inflammatory granulation tissue with granuloma, 2- Necrotizing granulomatous panniculitis, 3- Polymorphic periadnexal inflammatory infiltrate with necrosis and abundant altered neutrophils, 4- Lymphocytic inflammatory infiltrate with perivascular and perisudoral disposition of the dermis with lobular cytophagic necrosis and lipophagic granuloma of the hypodermis, 5- A discreetly lichenoid dermatosis. Lipodystrophy lesions were observed on the upper half of the left arm and buttocks. Additionally, the patient reported photosensitivity. Osteoarticular examination revealed inflammatory arthralgia with morning stiffness lasting over 30 minutes. Another skin biopsy revealed septal panniculitis without vascular involvement. Laboratory tests showed pancytopenia: anemia at 10 g/dl, thrombocytopenia at 125,000/mm3 (normal range: 150,000-400,000), leukopenia at 2,120/mm3 (normal range: 4,000-10,000. Immunological assessment revealed: homogeneous antinuclear antibodies (ANA) at 1/320 (positive for antihistone, anti-native DNA and anti-ribosome). Anticardiolipins were positive with complement consumption. A diagnosis of SLE was established. Optical Coherence Tomography (OCT) showed central eye atrophy in the right eye. Treatment with prednisone 0.5 mg/kg/day along with methotrexate 10 mg/week was initiated.

Results: LP is a rare anatomoclinical form mainly affecting middle-aged women. It often manifests as deep painful nodules progressing to atrophic scars. It can remain perfectly isolated for a long time without any other signs of lupus disease. The most important biological tests are immunological, including the overall identification of ANA and its components, and possibly the investigation of complement system activation. Histological examination requires a broad and deep biopsy to rule out the main differential diagnoses, including Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) and urticarial vasculitis. In this case, the pathological examination mainly suggests LP or LTSCP. These two diagnoses share many common clinical and histopathological characteristics. Clinical manifestations and distribution are similar in both conditions. In lymphomas, general signs are more commonly associated. Lymphadenopathy and/or hepatosplenomegaly may also be present. However, these signs may also be present in individuals with LP, especially in association with SLE. Immunofluorescence can detect a lupus band, suggestive of the disease, but it is inconsistent. Therefore, a negative immunofluorescence does not exclude the diagnosis of LP. Immunohistochemistry may be useful to differentiate between these two conditions.

Conclusion: LP can present diagnostic challenges, especially when mimicking lymphoma. Therefore, pathologists and dermatopathologists need to be fully aware of the significant overlap in this context.

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Abstract N°: 4173

Dermatomyositis revealed during pregnancy: another therapeutic challenge

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Introduction & Objectives: Dermatomyositis [DM] is a rare disease of unknown etiology, with great clinical and evolutionary polymorphism. Gestation is rarely compatible with this condition, due to the negative influence of maternal steroid hormones, which increase the expression of inflammatory myopathy. The onset of dermatomyositis during pregnancy is a rare event, and the prognosis is poor, with fetal death in 50% of cases, but successful therapy will lead to a satisfactory outcome. We report a rare case of DM revealed during pregnancy with a good evolution after treatment.

Materials & Methods: A 32-year-old woman, pregnant at 23 weeks of amenorrhea, with no particular pathological history, was referred to us for a cutaneous-muscular syndrome that appeared at 19 weeks of pregnancy. She complained only of slight muscle pain and fatigue. Clinical examination revealed a "V"-shaped heliotrope erythema on the décolleté, limb extensions, thigh roots and metacarpophalangeal joints. Examination of the fingernails revealed diffuse periungual erythema with a positive manicure sign. Capillaroscopy showed the presence of megacapillaries and glomerular vessels. Neuromuscular examination revealed a slight deficit of the pelvic girdle. Biological workup showed elevated muscle enzymes. ENMG revealed a predominantly proximal myogenic syndrome. On the basis of these results, the diagnosis of DM revealed during pregnancy was retained. The patient was put on prednisone 1mg/kg/d, which effectively controlled the dermatomyositis, and delivered a healthy newborn at term.

Results: Dermatomyositis is a rare disease that complicates pregnancy. Various factors have been considered as triggers for the development of DM during pregnancy, such as the mother's exposure to fetal antigens, changes in maternal hormonal status and the reactivation of certain viruses during pregnancy; maternal complications are dominated by the increased occurrence of pre-eclampsia or eclampsia, but infections are not negligible, notably pulmonary, and linked to the relative state of immunosuppression in these patients. Fetal complications are also linked to placental hypoperfusion. Inflammation of the myometrial muscle fibers explains the blood circulation problems between mother and fetus, as well as the growth deficit in uterine volume. Thus, the evolution of a full-term pregnancy or the birth of a viable newborn remains problematic, as it is fraught with frequent fetal complications. Prematurity may occur, or even fetal death in 50% of cases, or the onset of juvenile diabetes after the post-partum period. In general, optimal pregnancy success can be expected when the disease is in remission. The use of corticosteroids as first-line treatment is indicated in the event of exacerbation of the disease in the mother during pregnancy. In our case, we administered systemic corticoids with good as a first-line treatment.

Conclusion: dermatomyositis in pregnant women remains a potentially serious disease, with sometimes severe maternal-fetal complications. It constitutes a therapeutic emergency. Our case illustrates the theory that the onset of DM during pregnancy has a good outcome after medical treatment, particularly systemic corticosteroids.

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Abstract N°: 4184

A multicentric retrospective study of 108 chronic inflammatory black tattoo reactions

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Introduction & Objectives:

Chronic inflammatory black tattoo reaction (CIBTR) is a recent descriptive term coined by van der Bent et al that covers chronic papules,nodules or complete elevation, strictly limited to black tattoos, irrespective of sarcoidosis or non-sarcoidosis origin, without any clinical of histological signs of infection. In 2016, a Danish series reported 72 patients with such pattern. We report an international multicentric study of 108 patients.

Materials & Methods:

A retrospective analysis of outpatients diagnosed with CIBTR. Patients were either referred at i) the Tattoo Clinic, Dept of Dermatology, Alrijne Hospital, Leiden, the Netherlands, ii) the Dept of Dermatology at Helsinki University Hospital, Finland or iii) the "Tattoo consultation", Dept of Dermatology, Hôpital Bichat-Claude Bernard, Paris, France. We analyzed the demographics, clinical diagnosis, microscopic findings and outcomes.

Results:

108 patients (78 women, 73%, median age 34 years) were included. Permanent make-up were involved in 12% of the cases. Data are summarized in *table 1*. CIBTR presents as permanent or transient papules or nodules occurred within black tattoos in 96% of the cases. Time of onset of symptoms varied from direct up to over 15 years after tattooing. Tattoos were mainly located on the upper limbs (63%). When a biopsy was performed, granulomas were found in 83% of the cases. In most of the cases, laboratory findings and chest X-rays were unremarkable. Sarcoidoisis represented only 16% of all cases and TAGU (Tattoo granulomas with uveitis) only 2%. A past history of sarcoidosis was rather rare. Highly or ultrapotent corticosteroid ointments, topical tacrolimus and oral hydroxychloroquine were the most common treatments.

Conclusion:

We report the largest series of CIBTR. Reaction develops mainly in carbon black tattoos. Other colors (red…) are rarely affected. The clinical cutaneous presentation of CIBTR does not allow to distinguish non-sarcoidosis from sarcoidosis CIBTR. Patients rarely display any features of cutaneous or systemic sarcoidosis. The difficulties to classify a patient with isolated sarcoidal granulomas in tattoos without any other symptoms are well-known. However, sarcoidosis must be ruled out, at least during the initial workup. The completeness of the work-up remains at the discretion of the prescriber according to the patient medical history and clinical findings. Local or intralesional corticosteroids are the first treatment of choice. In persistent cases or extensive cutaneous involvement , hydroxychloroquine or methotrexate are potential options. Long term follow-up studies are necessary to define the exact percentage of sarcoidosis in CIBTR.

Table 1. Characteristics of CBITR

	Alrijne n=68 ***n* (%)**	**Helsinki n=22** ***n* (%)**	**Paris n=18** ***n* (%)**	**Total N=108** **N(%)**
Female	49 (72)	18 (81)	11 (61)	78 (73)
Median age, years	34	31	34	34
Mean age (SD), years	38(11)	37.2 (13.2)	34 .9 (10.4)	37 (11)
PMU PMU in women	9 (13) 9 (18)	2 (9) 2 (11)	2 (11) 2 (18)	13 (12) 13 (17)
Affected colors Black Red Orange Brown Blue Green	 68 (100) 2 (3) 1 (1.5) 0 0 1 (1.5)	 20 (91) 1 (4.5) 0 1 (4.5) 0 1 (4.5)	 16 (89) 2 (17) 0 1 (5.5) 1 (5.5) 0	 104 (96) 5 (5) 1 (1) 2 (2) 1 (1) 2 (2)
Localization Upper limbs Lower limbs Trunk Head & neck Eyebrows Lips	 45 (66) 17 (25) 14 (21) 12 (18) 7 (10) 2 (3)	 14 (64) 2 (9) 3 (14) 2 (9) 1 (5) 1 (5)	 9 (50) 5 (28) 3 (17) 2 (11) 2 (11) 0	 68 (63) 24 (22) 20 (19) 16 (15) 10 (9) 3 (3)
Sarcoidosis	8 (12)	7 (32)	2 (11)	17 (16)
TAGU	2 (3)	0	0	2 (2)
Granulomatous reaction upon biopsies	34 (74)	17 (100)	8 (100)	59 (83)

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Abstract N°: 4204

An Exceptional Case of Overlap Syndrome: Bullous Systemic Lupus Erythematosus and Paraneoplastic Dermatomyositis

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Introduction & Objectives:

Dermatomyositis, an inflammatory myopathy, manifests with muscle weakness and distinctive skin rashes, often linked to autoimmune reactions or underlying malignancies. Bullous systemic lupus erythematosus, a rare form of systemic lupus, presents with autoantibody-mediated subepidermal blisters resembling those in bullous pemphigoid. Overlap syndromes, involving multiple connective tissue diseases, introduce significant diagnostic and therapeutic challenges.

Materials & Methods:

A 57-year-old woman presented with erythroderma of unknown origin. The differential diagnosis considered allergic reactions possibly triggered by recent medication (metformin hydrochloride started two weeks prior) or hair dye exposure. Initial management included intravenous methylprednisolone, transitioning to oral corticosteroids, antihistamines, and topical sodium hyaluronate. Laboratory tests showed mild liver enzyme elevations, hyperglycemia, elevated bilirubin, high IgE (1338.6 kU/l), and CRP (27 mg/l). Discontinuation of glucocorticoids led to a recurrence marked by extensive bullae, skin erosions, subfebrility, and pain, necessitating repeated hospitalizations for further histological examinations and autoantibody testing in a suspected paraneoplastic context. Elevated tumor markers included SHE4, S-ROMA2, CA125*S, and C 15-3.

Results:

Initial histological analyses suggested bullous lupus erythematosus, although autoantibodies were consistently negative. The patient’s condition progressed, marked by skin whitening and stiffening on the hands, upper torso, abdomen, and buttocks, intensifying pain. These symptoms necessitated further biopsies and rheumatological evaluations. The diagnosis of dermatomyositis was primarily based on clinical observations and laboratory results, including positive RNP3RP155 and myositis blot TYF1gama autoantibodies; subsequent histology confirmed this diagnosis. Imaging revealed focal liver involvement indicative of metastasis. Despite unremarkable gastroscopy and colonoscopy results, an MRI showed muscle edema without atrophy. A CT scan identified liver metastases, peritoneal involvement, multiple enlarged lymph nodes, a calcified myoma, and posterior uterine involvement. Ultimately, a liver biopsy confirmed the presence of metastatic adenocarcinoma with an unidentified primary origin.

Conclusion:

This case underscores the diagnostic and therapeutic complexities of managing overlap syndromes like bullous systemic lupus erythematosus coupled with paraneoplastic dermatomyositis. The intertwining of autoimmune disorders with malignancy complicates clinical assessments and decision-making, highlighting the need for a

nuanced approach. The patient's progression to severe systemic involvement indicates a poor prognosis and emphasizes the necessity for a coordinated, multidisciplinary strategy involving rheumatology, dermatology, oncology, and pathology. This case not only reflects the challenges in treating such intricate conditions but also underlines the critical need for advanced research and targeted therapies to better understand and manage overlap syndromes effectively.

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Abstract N°: 4249

Development of a Care practitioner Reported Experience Measure (CREM) for interdisciplinary care in systemic diseases with skin manifestations (skIMIDs)

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Introduction & Objectives:

Immune-mediated inflammatory diseases with skin manifestations (skIMIDs) comprise several complex chronic conditions involving various organ systems. These require potent long-term immunomodulation or -suppression in order to achieve optimal disease control of the different involved organ systems as well as monitoring potential side effects. Thus, interdisciplinary care (IC) is an important approach for the fragmented knowledge and management of skIMIDs. Unfortunately, IC still encounters challenges in various aspects which impacts professional satisfaction, perception of the quality of care, autonomy, collegiality, etc. of health care practitioners (HCPs). This might have an important role in the IC success. Thus, gaining insight on HCP's experience on IC is an essential aspect to assess improvement areas. The objective of this project was to develop a compact and user-friendly questionnaire: a Care practitioner Reported Experience Measure (CREM) to measure HCP's experiences with IC in skIMIDs.

Materials & Methods:

In a PubMed-based literature review, potential relevant items on HCP's experience in IC were identified, which generated an initial longlist. Subsequently, refinement into a shortlist through semi-structured interviews involving HCPs of various specialties with expert knowledge of skIMIDs was conducted. In a focus group of dermatologists and rheumatologists, a final list was created, following a modified Delphi-method. Each item was formulated into a statement, which formed the basis of the CREM questionnaire that ultimately underwent an initial internal validation through email rounds.

Results:

The refining process of the initial longlist of relevant items by 16 interviews and a focus group resulted in 29 items over five domains: communication (e.g. interpersonal dynamics, respect, approachability of colleagues), collaboration (e.g. conflict management, alignment, confidence in expert competencies), organization & structure (e.g. frequency, format, preparation, participant composition, clear tasks and responsibility, participation of meetings and consultations) and care process (e.g. arrangement of patient appointments, standardization, medical records, infrastructure) and personal aspects (feeling of positive contribution to patient care).

Conclusion:

The perspective of HCPs is a relevant fragment in the evaluation of on IC in skIMIDs. Via this novel CREM, experiences of HCPs can be assessed and improvement areas of IC, such as efficacy or work satisfaction can be identified.

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Abstract N°: 4251

Penile calciphylaxis, a clinic to never forget

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Introduction & Objectives: Calciphylaxis is a syndrome characterized by calcification of vessels located in the dermis and adipose tissue mostly seen in patients with renal failure, especially those undergoing dialysis. Penile involvement is an uncommon but severe manifestation, likely due to the rich vascular network in this area. There are less than 50 reported cases of penile calciphylaxis in the literature. The recognition of calciphylaxis often eludes practitioners because of its multiple ambiguous presentations. Clinical differential diagnoses include squamous cell carcinoma and atypical candidiasis. A heightened suspicion for the disorder is therefore required in the case of penile calciphylaxis, given its unconventional location. Its diagnosis is by biopsy and the recommended stain is von Kossa used to identify tissue calcification.

Materials & Methods: We report a case of penile calciphylaxis in a 64-year-old man with multiple comorbidities, terminal chronic kidney disease on interdaily hemodialysis, diabetes mellitus with micro and macrovascular complications. Who consulted the emergency service for a penile ulcer, at the time, one month of evolution, unsuccessfully treated with antibiotics. The patient was referred to the dermatology service and physical examination revealed a 40 mm x 20 mm ulcer lesion at the level of the glans penis, with a white background, dry, well-defined, erythematous borders, non-fetid, and sparing the urinary meatus.

Results: Among the possible diagnoses in an immunosuppressed patient, squamous cell carcinoma and atypical candidiasis were considered, so it was decided to take a biopsy, culture and von Kossa stain, which reported penile calciphylaxis. Given this diagnosis and knowing the poor prognosis that it entails, it was decided together with nephrology (treating specialty) to perform electrolyte and PTH corrections, achieving complete resolution of the lesion, today after almost 2 year of his diagnosis the patient still alive without recurrence or appearance of new lesions.

Conclusion: Penile calciphylaxis is a rare disease with poor prognosis and high mortality rate around 64% within months of diagnosis. Its early recognition is crucial for timely treatment and impact on the patient's prognosis. There is no gold standard in the definitive management of patients with penile calciphylaxis, and multiple approaches have been described. But what is clear is that goals of care should be initiated early and a multidisciplinary team approach is essential, with corrections on parathormone levels y/or electrolytes disorders and wound care to impact the patient's prognosis and quality of life. The importance of this case lies in recognizing this clinical presentation to make a prompt diagnosis, in order to offer rapid treatment and thus impact the patient's prognosis and quality of life

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Abstract N°: 4267

Lupus-associated toxic epidermal necrolysis following anti-PD1 treatment with pembrolizumab for metastatic adenocarcinoma: A diagnostic and therapeutic dilemma

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Introduction & Objectives:

The cutaneous manifestations of lupus are highly polymorphic. Lupus-Lyell, or lupus toxic epidermal necrolysis, is an exceptionally rare entity that closely resembles drug-induced Lyell's syndrome. Here, we present a novel observation of such a case in a patient undergoing pembrolizumab therapy for lung metastatic adenocarcinoma.

Results:

A 57-year-old man with no documented history of dermatological or autoimmune disease presented to our dermatological department following the onset of a maculo-papular rash. His history was significant for stage IV metastatic non-small cell lung cancer (NSCLC), adenocarcinoma subtype, with malignant pericardial effusion and multiple mediastinal and hilar lymphadenopathy. The molecular markers tested positive for 90% PD-L1, but negative for receptor tyrosine kinase (ROS-1), anaplastic lymphoma kinase (ALK) and epidermal growth factor receptor (EGFR).

He was started on a combination of chemotherapy and immunotherapy with carboplatin, alimta, and pembrolizumab. One week after the first cycle of the combination chemotherapy, he developed diffuse non-pruritic erythematous papules coalescing into plaques on his abdomen, back, chest and 4 extremities. Mucous membranes were not involved. The skin biopsies revealed epidermis mild atrophy, vacuolization of epidermal keratinocytes, lymphocytic dermal infiltration, predominantly around adnexal sites, and epidermis leukocyte exocytosis. Direct immunofluorescence assays for IgG, IgA, and C3 were positive consistent with a lupus band. At the time of the rash's presentation, a review of systems was negative and the patient's complete blood count, immunological staining, electrolytes, and renal function tests were normal.

After complete skin resolution, and following a discussion with the patient regarding the risks and benefits of continuing therapy, it was mutually decided to continue the treatment. One week after the second cycle, he developed an extensive, confluent erythema involving the back, chest, abdomen, and extremities with large areas of Nikolsky-positive epidermal detachment on the upper trunk and minimal rubbing of the skin in the genital area. There was no ocular involvement.

He continued to deteriorate with >90 % skin involvement and hemodynamic instability. In addition to lung adenocarcinoma with malignant pericardial effusion, the patient died 5 days later from complications and subsequent multiple organ failure.

Conclusion:

Cutaneous lupus erythematosus is known for its diverse clinical manifestations, with lupus erythematosus necrolysis/Lyell's syndrome being a rare but diagnostically significant expression due to its severity and therapeutic urgency. This broadens the spectrum of reactions associated with PD-1 inhibitor therapy and underlines the importance of correctly recognizing this disease so that patients can be treated appropriately and

at an early stage.

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Abstract N°: 4285

VEXAS syndrome: an unusual histopathological presentation

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Introduction & Objectives:

VEXAS (vacuoles, E1 enzyme, X-linked, autoinflammatory, somatic) syndrome is a late-onset autoinflammatory disorder that results from mutations in the UBA1 gene, which encodes the X-linked, ubiquitin-activating enzyme E1. This syndrome typically affects male patients older than 60 years old and the most prominent clinical manifestations are cutaneous, haematological and rheumatological. Neutrophilic dermatoses represent the most common skin finding. Currently, no formal clinical diagnostic criteria are available for VEXAS syndrome, so the genetic testing is critical to confirm the diagnosis.

Materials & Methods:

We describe a case of VEXAS syndrome with clinical and histopathological manifestations seldom described in this disease.

Results:

A 73-year-old male presented with a 1-year evolution of evanescent erythematous non- pruritic papules on the trunk. The skin biopsy revealed neutrophilic dermatitis without vasculitis. A diagnosis of Sweet syndrome was assumed, and he was treated with oral corticosteroids. Three years later, he was observed again for non-pruriginous erythematous papules, affecting the trunk and upper limbs, as well as yellow plaques and papules on the back and lower lip. Histopathological examination of a skin biopsy from the trunk and lip lesions demonstrated a diffuse infiltrate of histiocytes (CD68 positive, S100 protein negative) in the dermis with abundant foamy macrophages, leukocytoclasia and collagen necrobiosis. At this time, he had myelodysplasia, optic neuritis, and complaints of polyarthralgia. A genetic test was performed and identified a somatic variant c.121A>C on the UBA1 gene. These findings are consistent with VEXAS syndrome diagnosis. The patient has subsequently started on tocilizumab.

Conclusion:

VEXAS syndrome is an autoinflammatory, clinically heterogenous disease affecting various organ systems, caused by mutations in the UBA1 gene. Although neutrophilic dermatitis is the most common finding we describe this patient with xanthomatous lesions and histology revealed an extensive histiocytic infiltrate, probably a reactive infiltrate in the setting of VEXAS syndrome, a few years after the onset of dermatological manifestations. Cutaneous manifestations are a prominent finding and dermatologists play an important role in the diagnosis of this disease. There are currently no recognized clinical diagnostic algorithms for this syndrome, so the molecular diagnostics are required for the correct diagnosis.

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Abstract N°: 4366

Sinus histiocytosis: Extranodal Rosai-Dorfman-Destombes disease

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Introduction & Objectives:

Rosai-Dorfman-Destombes disease (RDD) is a rare non–Langerhans cell histiocytosis (LCH) first described by P. Destombes in 1965 and further deeply researched by J. Rosai and R.F. Dorfman. RDD may be limited to the lymph nodes or have extranodal involvement, which occurs in more than 40% of patients. The most common site of extranodal involvement is the skin. Despite the description of the classical picture of the disease with predominant lymph node involvement, skin lesions may be its only manifestation, which is confirmed by our observation. An important diagnostic criterion in the diagnosis of RDD is immunohistochemical examination. Here, we present a case report of two female patients diagnosed with extranodal Rosai-Dorfman-Destombes disease.

Materials & Methods:

We present two cases of Rosai-Dorfman-Destombes disease with extranodal involvement. The first one is a case of a 23-year-old woman with complaints on skin lesions on the left cheek and left shoulder; skin manifestations included papular and nodular eruptions with intense pink color and a yellowish and brown tint. The second is a case of a 54-year-old woman with the previous history of numerous surgical excisions of nodular lesions on lower extremities but without the proper diagnosis. In both cases, an excisional biopsy with microscopic and immunohistochemical examination of the material was performed to verify the diagnosis. The immunohistochemical examination turned out to be the key to the diagnosis.

Results:

One characteristic morphologic finding of the disorder is emperipolesis. As defined by J.G. Humble et al. (1956) emperipolesis is “the active penetration of one cell by another which remains intact”. Histologic examination typically showed pericapsular fibrosis and dilated sinuses heavily infiltrated with large histiocytes, lymphocytes, and plasma cells. The engulfment of lymphocytes and plasma cells by histiocytes that express S-100 is considered diagnostic of RDD. IHC stains of RDD cells are also positive for CD68, whereas CD1a is typically negative. At the follow-up examination 12 months after surgical removal of the skin lesions, both patients had no new manifestations of the RDD disease on the skin.

Conclusion:

Subsequently it became evident that RDD disease may evolve in extranodal location with no associated lymphadenopathy. Microscopic and immunohistochemical analysis remains the pillar of RDD diagnosis. If performed properly surgical treatment assures excellent prognosis though cases of recurrences were outlined. Because of the wide clinical spectrum of RDD and the consequent variety of specialists evaluating and treating such patients, there is a need for an evidence-based approach to the evaluation and treatment of this condition.

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Abstract N°: 4495

Necrolytic migratory erythema as a cutaneous sign of a metastatic neuroendocrine tumor

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Introduction & Objectives:

Necrolytic migratory erythema (NME) is a rare skin condition associated with an underlying neuroendocrine neoplasm usually as a first sign. We present a 54-year-old male who developed NME 15 years after diagnosis of glucagonoma with following surgical resection and chemotherapy. In 2023, the patient came in our department with two weeks lasting skin manifestation and a month of a consummative syndrome after many years of clinical tumor remission which made us the suspicion of tumor metastases.

Materials & Methods:

A 54-year-old male with erythematous scaling and crusted patches first appeared on the skin of pressure points of the ski boots after a ski trip then rapidly migrated to the trunk, extremities and the perioral area of the face. Some erosions and edema on the skin of the ankles were noticed. Patient reported on a history for a pancreatic tumor diagnosed in 2009 managed with a radical surgical resection, excision of a liver metastasis followed by targeted oncological therapy. The patient was in a poor general condition with a weight loss above 20 kg for the past 4 months, fatigue and fever 37.7 C.

Results:

Following the dermatological presentation, a series of diagnostic evaluations were undertaken. Laboratory investigation showed pancytopenia (HGB 60 g/l, PLC 13 x 10^9), low proteins, high liver enzymes, high blood sugar. Skin biopsy was taken and the histology revealed pale vacuolated keratinocytes in the upper epidermis leading to focal necrosis and a mild perivascular infiltrate of lymphocytes and neutrophils. Diagnosis NME was confirmed. Tumor markers were extremely increased - chromogranin A 492.2 ng/mL (rr.0.0-101.9 ng/mL) and CEA 5.34 ng/mL (rr. 0.0-0.3 ng/mL), while CA19-9 and beta-2-microglobulins were within normal ranges. A CT-scan was performed where new metastases in liver, left kidney, peritoneal and mesenteries lymph nodes were observed.

The patient was managed with a multidisciplinary approach involving dermatologists, oncologists, endocrinologists. Treatment regimens aimed to correct the laboratory abnormalities and skin changes and afterwards changing the targeted therapy was considered. The condition during the therapy worsened and the patient moved to the intensive care unit.

Conclusion:

NME is a rare paraneoplastic dermatological manifestation associated frequently with glucagonoma. This case report demonstrates the critical need for awareness if skin manifestations appeared even after years of clinical and laboratory paraneoplastic remission. The prompt diagnosis and multidisciplinary approach are crucial for optimal patient outcomes, as highlighted by improving the quality of life and prognosis of the patients. Long-term management strategies for patients with underlying neoplastic diseases are needed and appearing of new skin lesions should be under suspicion of a tumor reactivation.

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Abstract N°: 4559

Functional and clinic evaluation in dermatomyositis

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Introduction & Objectives: The objective of this study was to evaluate the clinic and functional signs of dermatomyositis.

Materials & Methods: The patient (woman, 51 years old) was investigated since 2013. The aspect of the skin, biological parameters and the electrophysiological investigations were considered.

The initial stage of the disease was expressed by erythematous placard, muscles fatigue, joints pain and poikiloderma.

Results: The blood lab tests expressed creatininuria 5.8 g/24h, LDH 380 IU/l, CPK 207 IU/l, the red blood cells sedimentation rate 18 mm/h, RF (rheumatoid factor) +, CRP (C reactive protein) +, high level of transaminases and the presence of antinuclear antibodies.

The electrophysiological investigations (bilateral deltoid muscles) aspect revealed normal/low amplitude and duration of unit motor potentials, a high recruitment pattern for right deltoid muscle which means a myogenic type. The systemic and local corticotherapy (Advantan), anti-allergic drugs and vitamin-therapy improved the lab test values and the pattern aspect in EMG.

Conclusion: The variation of biologic parameters and the functional tests aspect are very important for the prognostic of the disease.

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Abstract N°: 4570

Cutaneous Manifestation Predictive of Progression and Poor Prognosis in Non-Hodgkin Lymphoma: A Case Report

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Introduction:

B-cell large cell lymphoma accounts for approximately 25% of all non-Hodgkin lymphomas, making it the most common subtype of lymphoma.

Case report:

In this case report the patient was a 58-year-old woman who reported the appearance of a non-pruritic plaque on her right breast one month ago. She had been undergoing chemotherapy for B-cell large cell lymphoma exclusively involving the lymph nodes, having completed sessions with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone, with the last session less than 6 months ago. Upon examination, she presented with a erythematous and crusted plaque measuring 2.5cm on the right breast. Dermoscopy revealed telangiectasias. A biopsy of the lesion was performed, and the material was sent for Immunohistochemistry, bacteriology, and mycology. No bacterial, mycobacterial, or fungal growth was observed in the sample. The immunohistochemical report showed: “CD3: positive in numerous associated small cells; CD20: positive in most of the dermal lymphoid infiltrate, represented by large cells; CD10: positive in predominantly dendritic morphology cells in the lower dermis; Bcl6: positive, nuclear, in sparse cells; Bcl2: positive in numerous associated small cells; Mum 1: positive, nuclear, in numerous cells of the dermal infiltrate; Ki67%: 90%; CD34: positive in endothelial cells; CD30: negative”. Thus, the histopathological report was “consistent with non-germinal center B-cell lymphoma of the skin.” Based on this data, the patient was referred to Hematology, where therapeutic failure was observed and a new staging of the disease was conducted with tomographic and PET-SCAN exams. It was then decided to restart treatment with dexamethasone, cytarabine, and cisplatin.

Discussion:

Due to the histopathological report and the fact that the last chemotherapy dose was less than 6 months ago, it was concluded that there was disease progression to an extranodal site: the skin. In this context, since the patient initially presented with neoplasia focused only on lymph nodes and, subsequently, dermatological involvement was confirmed through biopsy, it is possible to affirm that there was disease progression. Such disease progression led to a worse prognosis and reduced survival.

Our case is a rare presentation of disease progression, with the main symptom on the skin. This requires initial clinical suspicion so its knowledge is essential for dermatologists. About 25% of non-Hodgkin lymphomas may present at an extranodal site although without systemic involvement. There are several reports in the literature whit skin involvement by lymphoma. Most of these cases however describe the skin as the disease’s primary focus, which differentiates and denotes the rarity of the reported case.

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Abstract N°: 4626

Sodium thiosulfate treatment of calcinosis cutis in patients with systemic sclerosis assessed by novel biomarkers and reflectance confocal microscopy – a study protocol

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Introduction & Objectives:

Systemic sclerosis (SSc) presents a complex challenge in clinical practice. Up to 50% of SSc patients experience calcinosis cutis (CC), painful calcium and phosphate deposits in skin and subcutaneous tissue, significantly impacting quality of life and daily activities. Despite its prevalence and impact, effective treatment options for CC remain scarce. The exact mechanism is unknown but could be related to inflammation and hypoxia. No effect treatment is available, but sodium thiosulfate (STS) has shown promising results. Reflectance confocal microscopy (RCM) offers a non-invasive means to study cellular dynamics in vivo, providing invaluable insights into disease pathology. In this study protocol, we outline our approach to exploring the behavior of CC and investigating the therapeutic potential of STS using a combination of RCM and novel blood and skin biomarkers.

Materials & Methods:

Fifty SSc patients diagnosed with CC will be recruited to participate in this prospective study. Inclusion criteria include the presence of CC lesions on the knee, arm, or hand, with patients receiving intralesional STS treatment at a concentration of 150 mg/ml. Blood samples and skin biopsies will be collected at baseline and throughout a 28-week trial period. These samples will be compared with 25 age- and sex-matched SSc patients without CC. Biomarker analysis will focus on key pathways expected to implicate CC pathogenesis, including collagen remodeling, immune cell modulation, and hypoxia. RCM imaging will be performed before and after STS treatment to assess changes in cellular morphology and architecture.

Results:

This study protocol outlines the methodology for data collection and analysis. We hypothesise that biomarker analyses and RCM reveal dysregulation of structures and inflammatory activities. We anticipate that our comprehensive approach will yield valuable insights into the molecular and cellular mechanisms driving CC progression and response to STS therapy.

Conclusion:

In our collective research project, we explore unresolved complexities surrounding CC.

By looking into the biological and chemical mechanisms, we aim to expand our understanding of the underlying pathways and pathogenesis. These insights hold promise for the development of targeted therapeutic interventions, offering tangible benefits to individuals battling with the debilitating effects of CC on their daily functioning.

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Abstract N°: 4646

Acute graft-versus-host-disease in a male patient without gastrointestinal tract involvement

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Introduction & Objectives:

Graft-versus-host-disease is a common complication following allogenic hematopoietic stem cell transplant. It has multisystemic involvement and manifests as an immune reaction triggered by the transplanted cells from the donor. Graft-versus-host-disease is divided into acute and chronic. The classification was typically made based on the timing from the transplant, using a period of 100 days. Currently, for the acute graft-versus-host-disease, two grading system were proposed, combining the degree of affected body surface area and the degree of involvement of liver and gastrointestinal tract. Cutaneous manifestations of the acute disease include a maculopapular rash involving the extremities, with the possibility of spreading to the whole body. The gastrointestinal tract and liver can be also affected, suggested by the presence of abnormal liver tests and episodes of diarrhea. Herein, we report the case of a male patient who has undergone transplantation for acute lymphoblastic leukemia, exhibiting cutaneous lesions and no involvement of gastrointestinal tract.

Materials & Methods:

A 19-year-old male presented in the Dermatology Department with a generalized macular rash, that occurred 8 months previously. The patient was diagnosed with acute lymphoblastic leukemia in November 2022 and had received allogeneic hematopoietic cell transplantation in July 2023. After one month from the transplant, an erythematous macular eruption had occurred, involving the face and palms. The rash had an intermittent evolution, and local treatment with topical medium potency corticosteroids was applied. There was no history of gastrointestinal or liver malfunction. The physical examination revealed a generalized erythematous rash, consisting of isolated and conflated macules. No other signs and symptoms were present.

Results:

A punch biopsy was performed, and the histologic examination described apoptotic basal keratinocytes, dermal lymphocytic infiltrate, and lymphocytic exocytosis. Liver function tests were normal. An acute graft-versus-host-disease stage 2 diagnosis was established. Treatment with topical high potency corticosteroids was initiated, with the resolution of the cutaneous lesions within 2 weeks.

Conclusion:

Classification of acute graft-versus-host-disease is significant to assess the response to treatment. Liver, along with the upper and lower gastrointestinal tract, are the most frequently targeted organs. The diagnosis of acute graft-versus-host-disease presenting with skin rash and related symptoms to the above-mentioned organs should be considered in any patient treated with allogeneic hematopoietic cell transplantation. Even though clinical and paraclinical evaluations are in parameters, repeated assessments are needed to avoid further complications.

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Abstract N°: 4695

Vulvar ulcers: a rare presentation of a potentially fatal disease

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Introduction & Objectives:

Neutrophilic dermatoses are characterized by cutaneous infiltration of neutrophils without evidence of infection and may sometimes be associated with underlying malignancies. We aim at reporting how vulvar lesions may be the presenting finding of a paraneoplastic neutrophilic dermatosis associated with acute myeloid leukemia.

Materials & Methods:

We report a case of a 57-year-old female that presented with painful vulvar lesions.

Results:

Clinical Case: A 57-year-old female presented to the Emergency Department with a history of fever and painful vulvar lesions with one week of progression and which had not responded to treatment with doxycycline. She also reported nausea and mild arthralgias, but denied other systemic symptoms. She had been previously healthy, with no past medical history besides dyslipidemia treated with atorvastatin.

On physical examination, she was febrile but hemodynamically stable. Multiple ulcerative lesions could be seen on a background of indurated vulvar edema. Most lesions were centimetric and some of them were infiltrated. There were also small oral ulcers on an erythematous base. The serum biochemistry profile revealed elevated C-reactive protein (36,49mg/dL) and D-dimers (71148ng/mL), and the complete blood count revealed significative leukocytosis (40,8 x 109/L), mild normocytic anemia (11 g/dL) and thrombocytopenia (67 x 109/L). Herpes simplex virus DNA was detected in the oral lesions. Serologic tests for syphilis, HCV, HBV and HIV were negative, as well as PCR tests for detection of *N. gonorrhoeae* and *C. trachomatis*.

The patient was admitted to the Dermatology ward for etiologic investigation and treatment – the initial clinical hypotheses were of pemphigus vulgaris and Behçet disease, in coexistence with a herpes simplex virus infection and a possible leukemoid reaction to an underlying systemic process. She was started on meropenem, clindamycin, acyclovir and a low dose of methylprednisolone, with total resolution of the oral lesions but not of the vulvar ulcers. Histopathological analysis of these persistent lesions revealed a neutrophilic infiltrate in both epidermis and dermis, which was consistent with the diagnosis of a neutrophilic dermatosis with features of both Sweet syndrome and pyoderma gangrenosum. HLA-B*51 was negative.

During the hospital stay, the patient developed severe anemia and thrombocytopenia, both requiring transfusion support. She was transferred to the Hematology ward and underwent bone marrow aspiration, which confirmed the diagnosis of acute promyelocytic leukemia (APL). Treatment with all-*trans* retinoic acid was promptly initiated, which resulted in the complete resolution of both vulvar lesions and hematologic abnormalities.

Discussion and conclusion:

Multiple vulvar lesions are most frequently found in the setting of sexually transmitted infections but pose a diagnostic challenge once these are excluded. Histopathological analysis confirmed the diagnosis of a neutrophilic

dermatosis and, in the setting of persistent hematologic abnormalities, prompt investigation for an underlying malignancy was warranted. APL is a subtype of acute myeloid leukemia and mostly presents with pancytopenia or a life-threatening coagulopathy. A paraneoplastic neutrophilic dermatosis consisting of vulvar lesions has not been previously reported in association with APL but was, in this case, an important diagnostic clue that enabled timely treatment.

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Abstract N°: 4746

Study of immunopathogenetic features of psoriasis and acne's course

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Introduction & Objectives: . The most common in dermatological practice are psoriasis and acne, the pathogenesis of which today is considered from the standpoint of immunopathological diseases. The article analyses features of anamnesis, clinical, instrumental and laboratory tests related to chronic dermatitis (acne, psoriasis, arthropathic psoriasis (AP)), considers the relationship of probable mechanisms of disease aggravation and progression.

Objective. The objective of our work was to improve the diagnostics of common chronic dermatoses (acne, psoriasis, AP) taking into account some indicators of the immune system and features of the disease course to specify their role in pathogenesis of these disease.

Materials & Methods: A total of 128 patients with acne and 178 patients with psoriasis, among which 57,4 % women and 42,2 % men were observed have been systematically examined. We have examined patients with psoriasis with varying severity of process development, generalization and the severity of skin, the presence of associated pathology. Additional instrumental studies, determination of biochemical, serological parameters and an assessment of immune system have been conducted in AP patients. The content of trigger cytokines (IL-1β, IL-8, IL-17, IL-22) in blood serum, cellular and humoral immunity condition (CD3 +, CD4 +, CD8 +, CD16 +, CD22 +, IgM and IgG levels) have been studied in patients with acne and psoriasis.

Results: The clinical course of psoriasis and characteristic features of AP instrumental tests are extremely versatile as well as the depth of their present study is insufficient. Regardless of the disease duration period, we have detected in blood serum of psoriasis patients probable changes in concentrations of stress-response mediators (decreased parameters of cellular immunity (CD3+, CD4+, CD8+ of T-lymphocytes, CD22+ fraction of B-lymphocytes and compensatory increased CD16+ of T-cells, cytokines – IL-1β, IL-8, IL- 17, IL-22, immunoglobulins IgM, IgG, and CIC), which indicate tension of their stress-induced mechanisms even despite occasional clinical stabilization of skin and articular process.

Consequently, most of the patients with acne had varying degrees of changes in rates of systemic immunity – the likely reduction in relative and absolute number of total lymphocytes, T-lymphocytes and their subpopulations against the growing number of B lymphocytes and the level of IgM and IgG, which generally indicates the formation in these patients secondary immunodeficiency state of T-link intensified by activation of humoral immunity in response to the development of skin inflammation. The most significant changes in rates of systemic immunity with the depletion of T-cell immunity were found in patients with papular-pustular and pustular acne, and still more significant – in patients with acne conglobate, which justifies differentiated treatment by immunomotropic drugs for these patients.

Conclusion: In patients with acne and psoriasis, changes in systemic immunity indexes that indicate the formation of secondary immunodeficiency state T-cell link, amid an adequate humoral immunity have been found. Relationship between the causes of changes of systemic immunity has been established. The improvement of patients with acne and psoriasis diagnostics taking into account some indicators of the immune-endocrine system and specifics of the disease course, will contribute to improving therapy and mended quality of life of patients.

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Abstract N°: 4768

Unique case report of systemic amyloidosis associated with scleromyxoedema and myeloma multiplex

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Introduction & Objectives:

Cutaneous manifestations often accompany hematologic malignancies, occurring before, after, or at various stages following diagnosis. Conditions like scleromyxedema and amyloidosis may result from paraneoplastic phenomena.

Scleromyxedema, a rare form of cutaneous mucinosis, typically emerges with papules and sclerodermiform skin indurations in middle-aged individuals. Monoclonal gammopathy almost always accompanies this condition.

Amyloidosis encompasses a range of disorders characterized by the accumulation of toxic insoluble beta-sheet fibrillar protein aggregates in tissues. Amyloid deposits can afflict various organ systems, leading to diverse clinical presentations, including cardiomyopathy, hepatomegaly, proteinuria, macroglossia, autonomic dysfunction, neuropathy, renal failure, hypertension, and corneal and vitreous abnormalities. Skin manifestations of systemic amyloidosis include ecchymoses, periorbital purpura, waxy papules and nodules, livedo reticularis, hemorrhagic bullae, scleroderma-like changes, easy bruising, and skin tightness.

Results:

We present a case of 51-year-old Caucasian women with scleromyxedema and systemic amyloidosis associated with myeloma multiplex. Perioral erythema and skin hardening and tightening started two years before admission. In personal history, the patient had a total thyroidectomy; she was otherwise healthy.

Upon clinical examination, several findings were noted: a hoarse voice, macroglossia, perioral erythema, diffuse sclerosis affecting the entire skin with a notable shift of the interscapular and lumbal region, and skin changes indicative of Reynauld phenomenon. Doughy edemas were observed on the lower legs and dorsums of the feet. While muscle strength remained intact, the patient experienced difficulty standing independently from a seated position, with a reduced range of motion detected in nearly all joints.

Skin biopsy revealed fibroblast proliferation, collagen fiber multiplication, and mucin deposition, confirmed by Alcian blue staining. Additionally, amyloid deposition was identified through Congo red and Thioflavin staining. Subsequent biopsy of subcutaneous fatty tissue further confirmed amyloid deposition. Laboratory analyses, including creatinine and urea clearance and proteinuria, were normal, aside from elevated tumor markers (CEA, CA 125, NSE) and NT-proBNP levels. While protein electrophoresis yielded normal findings, urine protein electrophoresis detected paraproteinemia, with immunofixation revealing monoclonal IgG lambda type and Bence Jones proteins. Serum free lambda light chains were notably elevated at 1050 (reference range: 8.3-27), with a low ratio of free kappa/lambda chains. Bone marrow biopsy indicated infiltration with monoclonal plasma cells (lambda+) at 30-40%.

CT examination revealed borderline hepatomegaly (164 mm diameter), adenomyosis of the uterus, and bilateral ovarian cysts. X-ray examination of the calvaria was normal, while echocardiography revealed slow relaxation of the left ventricle alongside other normal findings.

The patient was subsequently referred to hematology for initiation of chemotherapy following the CyBorD protocol.

Conclusion:

To the best of our knowledge, this is the first case of the simultaneous presence of scleromyxedema and systemic amyloidosis associated with myeloma multiplex.

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Abstract N°: 4775

Cutaneous sarcoidosis with atypical histologic features

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Introduction & Objectives: Sarcoidosis is a chronic multisystem inflammatory disorder characterized by the presence of non-caseating granulomas. The most frequently involved organs are the lungs, lymph nodes, eyes and skin. Cutaneous manifestations of sarcoidosis are highly variable and are divided into specific and non-specific lesions based on histopathologic features. Typically, the anatomopathological study of specific lesions shows non-caseating granulomas of aggregated epithelioid histiocytes. However, atypical histological features can make the diagnosis challenging.

Materials & Methods: We report a case of systemic sarcoidosis with cutaneous involvement and unusual histologic features.

Results: A 44-year-old woman with a history of high blood pressure, presented to the dermatology department with 2-year history of asymptomatic skin lesions. She was initially treated with topical corticosteroids with no improvement. Physical examination found non-pruritic, painless, multiple erythematous papules and plaques located on the face, scalp and left shoulder. A skin biopsy was performed, which showed necrotizing granulomas. The chest x-ray showed bilateral hilar adenopathies, whose histologic study showed non-caseating granulomas. Infectious or tumoral conditions were excluded. These findings were compatible with systemic sarcoidosis with cutaneous involvement.

Conclusion: One of the main histological features of sarcoid granulomas is the absence of necrosis. However, several atypical histopathological findings have been described such as the presence of necrosis, foreign material, Grenz zone or perineural granulomas. In these cases, clinicopathologic correlation and laboratory studies are essential in the differential diagnosis with other granulomatous skin diseases.

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Abstract N°: 4787

Development of a Patient Reported Experience Measure (PREM) for interdisciplinary care in systemic diseases with skin manifestations (skIMIDs)

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Development of a Patient Reported Experience Measure (PREM) for interdisciplinary care in systemic diseases with skin manifestations (skIMIDs)

Introduction & Objectives:

Systemic diseases with skin manifestations (skin immune-mediated inflammatory diseases, or skIMIDs, such as connective tissue diseases), require rapid diagnosis and tailored treatment of various organ systems, including long-term immunosuppression. Therefore, management of skIMIDs is provided by different specialties and modern therapies are used to treat several organ systems or comorbidities simultaneously. Thus, skIMIDS require collaboration between different disciplines and interdisciplinary care (IC) is becoming the “new normal”. However, IC encounters challenges, such as fragmented care pathways, insufficient communication, etc. In this context, patient experiences strongly influence adherence and in consequence medical outcomes. Patient reported experience measures (PREMs) are user-friendly questionnaires to monitor the quality of care and identify improvement areas. Objective of this study was to create a concise and user-friendly PREM to evaluate patient experiences during clinical IC interactions.

Materials & Methods:

A literature review on PUBMED identified potential IC-items, from which a longlist was formed. Subsequently, refinement into a shortlist was done through thirteen semi-structured interviews involving patients with different skIMIDs. In two multi-stakeholder focus groups consisting of patients, dermatologists and rheumatologists, a final item list was created, following a modified Delphi-approach. Each item was translated into a statement. The questionnaire underwent an initial internal validation through email rounds.

Results:

The refinement process of the item list by patient interviews and focus groups resulted in 15 statements and one open question covering the most relevant items in four domains: access to information, coordination of care, access to care, patient centeredness and patient satisfaction. Items related to care process were not deemed important enough for inclusion in the final questionnaire. The need for attentive and collaborative doctors, patient participation and understandable information were ranked as most important items. Other significant concerns included emergency contact point, waiting time between appointments with different disciplines and continuity of care.

Conclusion:

The shift in domains from literature search to the final PREM underlines the high importance of information, coordination of care, access to care, patient centeredness and patient satisfaction when treating patients in an interdisciplinary setting with skIMIDs.

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Abstract N°: 4862

severe edematous dermatomyositis: a rare entity of dermatomyositis

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Introduction & Objectives:

Edematous dermatomyositis is a very uncommon entity characterized by localized or generalized subcutaneous edema. It is a severe form of dermatomyositis that requires quick therapeutic treatment . We report one case with difficult therapeutic decisions.

Materials & Methods: Case report

Results:

An 85 year old patient, with history of colon neoplasia in remission, was admitted in hospital for dermatomyositis with typical cutaneous and sever muscular involvement which have been treated by methylprednisolone bolus and prednisone. She then developed severe edema of the left upper limb with aggravation of the cutaneous and muscular symptom. The decision to stop the bolus was taken with therapeutic reinforcement by mycophenolate mofetil and intravenous immunoglobulin.The result was the disappearance of the cutaneous symptoms in particular the edema with partial restitution of the muscular force.

Discussion:

In the literature, only 21 cases of edematous dermatomyositis have been reported .The subcutaneous edema is should not be confused with erythroedema, a classic lesion of DM, or with secondary mucinoses of DM. The edema is white, pitting or not, localized to the limbs or generalized and frequently associated with severe forms of dermatomyositis due to significant muscle damage, including dysphagia. Often refractory to corticosteroid monotherapy and aggravated by the methylprednisolone bolus , edematous dermatomyositis frequently requires the use of immunosuppressants and IV immunoglobulins. The edematous dermatomyositis described in the literature are immediately edematous, with the exception of a single case. The originality of our case comes from the secondary nature of the edematous process, complicating a classic form of treated dermatomyositis .

Conclusion:

Edematous dermatomyositis is a rare form of dermatomyositis .The subcutaneous edema is a potential marker of severity, requiring the rapid initiation of aggressive treatment, including corticosteroids, immunosuppressants and/or IV immunoglobulins.

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Abstract N°: 4864

Tuberous Sclerosis Complex: Bridging the Gap Between Misdiagnosis and Timely Intervention

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Introduction & Objectives:

Tuberous sclerosis complex (TSC) is a neurocutaneous disorder with an autosomal dominant inheritance pattern that is characterized by the appearance of various manifestations in the skin and other organs such as the central nervous system, lungs and kidneys. Currently, the International TSC Consensus Guidelines stipulate the diagnostic criteria for this condition. However, it often goes unnoticed by clinicians at the first level of care and in various medical specialties. This oversight can lead to multiorgan complications and fatal outcomes. For example, multiple renal angiomyolipomas can debut with acute intra-abdominal bleeding, causing hemorrhagic shock and associated complications.

Materials & Methods:

Regarding CNS tumors, subependymal giant cell astrocytomas stand out for their potential to cause hydrocephalus, blindness, and even death if not diagnosed in time. This is associated with multiple neuropsychiatric disorders that encompass various symptoms in the cognitive-behavioral sphere, including autism spectrum disorders and emotional and behavioral state disorders. Additionally, it is important to highlight that chronic pathologies, particularly those associated with epilepsy, can manifest as mild cases or difficult-to-manage epileptic disorders with high morbidity and mortality. At the kidney level, chronic kidney disease and high blood pressure can lead to end-stage renal disease in young patients in the second and third decades of life. Therefore, it is important for clinicians to study and consider the possibility of TSC

Results:

We describe two cases involving adolescents: a 12-year-old and a 15-year-old who were initially diagnosed with acne and received multiple treatments without improvement are presented here. These patients presented significant neuropsychiatric symptoms due to cognitive impairment and learning disorders. In one of the cases, this led to several years of school failure, while in the other, it resulted in analphabetism. These patients experience a range of difficulties in acclimating to their social and family surroundings.

Conclusion:

The role of the dermatologist is crucial, as they, through the anamnesis and physical examination, enable a diagnostic approach to TSC. A timely diagnosis of this condition and multidisciplinary follow-up can positively impact the prognosis and quality of life of patients with TSC, considering the wide spectrum of complications associated with this disorder.

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Abstract N°: 4920

An extremely rare association of large plaque parapsoriasis with advanced prostate adenocarcinoma

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Introduction & Objectives:

Large-plaque parapsoriasis (LPP) is a rare disease, that affects mostly middle-aged male adults, with chronic, round/irregular flat patches or thin plaques distributed on the trunk and extremities, including flexural areas, with a finely wrinkled surface and light red-brown colour. LPP histopathologically bridges chronic dermatitis and mycosis fungoides (MF), the most common form of skin lymphoma, being characterized by a CD4+ T-cell infiltrate in the papillary dermis, with or without a certain clonality. Although asymptomatic or mildly pruritic, lesions of LPP progress to MF at a rate of 10-30%. The therapeutical approach include emollients, topical corticosteroids, phototherapy, laser therapy and topical cytotoxic medication. While the association of LPP with solid cancers (in particular prostate adenocarcinoma) is extremely rare, it may be considered a paraneoplastic syndrome.

Results:

In this case report, we present a 66-year-old male patient diagnosed with prostate adenocarcinoma stage IV, Gleason score 9, with bone metastases, local and distant adenopathies, treated with the transurethral resection of the prostate, radiotherapy for vertebral bone metastases and androgen deprivation therapy with abiraterone and triptorelin. During the first month after the oncologic diagnosis, he developed an asymptomatic generalized eruption, mainly affecting the trunk and the upper limbs, consisting of multiple patches and thin plaques, ranging from 1 to 10 centimetres, light red-brown, round-to-oval, with a slightly scaly surface. Given the clinical features, the main differential diagnosis taken into consideration were LPP and patch/plaque-stage mycosis fungoides. The histopathology and immunohistochemistry examination showed criteria of superficial chronic dermatitis with lymphocytic infiltrate and no evidence of skin lymphoma (absence of epidermotropism, cellular atypia, negative markers). Given the diagnosis of LPP, the patient was treated with topical high-potency corticosteroids, with satisfactory clinical response. Narrowband UVB or PUVA were contraindicated, since the patient was undergoing radiotherapy. Periodic follow-up at 3 months was recommended to observe the dynamic of the dermatologic disease in parallel with the response to oncologic treatment for advanced prostate cancer and to identify the potential progression of LPP to MF.

Conclusion:

To the best of our knowledge, this is the first report of LPP in the context of prostate adenocarcinoma, maybe an underdiagnosed association due to the asymptomatic course of disease and the need for expert training to raise clinical suspicion. It is essential to differentiate LPP from MF by performing multiple skin biopsies on suspicious lesions and to perform follow-up every three to six months for evidence of progression.

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Abstract N°: 5031

An unusual case of bullous Wells' syndrome in a patient with lung cancer

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Introduction & Objectives:

Wells' syndrome is a rare disorder characterized by pruritic oedematous erythematous plaques on the trunk and limbs with occasional fever and peripheral eosinophilia. It is a relapsing-remitting dermatosis, and is associated with haematological malignancies, fungal, viral, parasitic infections, and drugs. Solid organ malignancies are an uncommon cause. The bullous form of Wells' syndrome is a rare form of this condition. We report a case of bullous Wells' syndrome in a patient with lung malignancy.

Materials & Methods:

An 81-year-old female presented with a two-day history of a non-pruritic generalized vesiculobullous eruption. Her past medical history included hypertension and diabetes mellitus. There were no preceding triggers including infection or oral medications. On examination, she had multiple slightly violaceous oedematous plaques, with overlying pustules, vesicles, and blisters on the face, neck, trunk, medial thighs and calves. She had no fevers or joint pains. Initial clinical impression included Sweet's syndrome, Well's syndrome, autoimmune blistering disorders or cutaneous infection such as impetigo, varicella or atypical infections. Her bloodwork revealed normal haematological indices with normal neutrophil and eosinophil count. Renal and liver function were normal. Erythrocyte sendimentary rate was 40, while peripheral blood film was unremarkable. Microbiological cultures from blister fluid were negative for bacterial growth, while polymerase chain reaction testing for varicella and herpes simplex virus were negative. Incisional biopsy from a representative plaque on the right inner thigh showed subcorneal and intracorneal vesicles consisting of neutrophils and eosinophils. A dense inflammatory infiltrate with sheets of eosinophils, some lymphocytes, histiocytes and neutrophils was seen involving the entire dermis extending to the subcutis. There were no features of vasculitis. Direct immunofluoresence from perilesional skin was negative. Tissue cultures were negative for bacterial, fungal and mycobacterial infections. Serologies for BP180, BP230 and indirect immunofluorescence were negative. Quantiferon-TB was negative.

A computer tomography (CT) scan of the thorax, abdomen and pelvis was performed to rule out malignancy and was significant for a 1.6cm x 1.2cm right lower lobe pulmonary spiculated nodule. No intrathoracic lymphadenopathy was observed. A CT-guided lung biopsy revealed the lung nodule to be a lepidic type of adenocarcinoma. Final staging of her malignancy was T1N0M0, and she received radiotherapy.

The patient was started empirically on topical clobetasol propionate 0.05% cream under occlusion once tissue cultures were negative. Her lesions resolved rapidly within 2 weeks with minimal post-inflammatory hyperpigmentation. There were no further recurrences of her rashes at 6-months follow-up.

Results:

We favour the diagnosis of bullous Wells' syndrome given the patient's clinical presentation, skin biopsy findings and diagnosis of lung adenocarcinoma. Her clinical and histological features fulfill the criteria (though unvalidated) by Heelan *et. al.* Our case is unique due to its florid yet rare bullous presentation in combination with an uncommon trigger (solid organ malignancy).

Conclusion:

We present a case of bullous Wells' syndrome in a patient with lung adenocarcinoma.

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Abstract N°: 5085

Investigating The Efficacy and Safety of Anifrolumab In Treating Cutaneous Lupus Erythematosus: An Evidence-Based Review

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Introduction & Objectives: Cutaneous lupus erythematosus (CLE) is a manifestation of lupus erythematosus, a complex autoimmune disease that often involves vital organs. Epidemiological data suggest that 70-80% of patients with systemic lupus erythematosus (SLE) exhibit skin manifestations at some point during their illness. These skin lesions are classified based on their presentation and severity into acute, subacute, and chronic forms. The treatment of autoimmune diseases with skin manifestations has increasingly incorporated biologics, which have shown effectiveness in managing disease activity while maintaining a favorable safety profile. Anifrolumab, a newly approved nterferon type-1 inhibitor, has shown potential in this area. Despite its approval, comprehensive data on its effectiveness and safety in clinical settings, including both controlled trials and real-world applications, are limited. This study aims to systematically review the existing literature to better understand the efficacy and safety profiles of anifrolumab in treating patients with CLE.

Materials & Methods: This systematic review was conducted according to the PRISMA guidelines. The quality of evidence was assessed using the Oxford Centre for Evidence-Based Medicine Levels of Evidence (2011). The review incorporated search terms specific to Anifrolumab and its use in both cutaneous and systemic lupus erythematosus. Screening processes, including the review of titles and abstracts followed by full-text assessments, were performed by two independent reviewers. Any disagreements were adjudicated by a third reviewer. We evaluated treatment efficacy using the cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) for both activity (CLASI-A) and damage (CLASI-D), the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI), and specific outcomes for discoid lupus erythematosus and chilblain. Safety was assessed through the analysis of adverse event reports and their severity within the patient cohort.

Results: The review included 19 studies, consisting of four randomized controlled trials, two retrospective cohorts, three prospective cohorts, four case series, and six case reports, totaling 499 patients, of whom 93.6% were female. The average CLASI-A score before treatment was 17.9, and the CLASI-D score was 12.05. Following treatment with anifrolumab, the average CLASI-A score decreased to 15.16, and the CLASI-D score reduced significantly to 0.86. Improvement in the SLEDAI scores was noted at 61.94% on average. Complete resolution was achieved in 22 patients for underlying SLE and in 11 patients for chilblain outcomes. Partial resolution was observed in 457 additional patients. Adverse events were reported in 391 patients, with 62 (16%) experiencing grade 3 or 4 events. There were 32 treatment discontinuations, including 3 due to adverse event intolerance and one death attributed to treatment-related adverse events. The average duration to achieve these outcomes was 92.6 days, with a mean follow-up period of 6.43 months.

Conclusion: Anifrolumab demonstrates significant efficacy in managing CLE, reducing disease activity across various manifestations. It also maintains a reasonable safety profile. However, the limitations inherent in the study, including the small sample size, the absence of a comparator group, and the heterogeneity of patient data,

necessitate further investigation through larger, more controlled studies.

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Abstract N°: 5181

Skin lesions after hematopoietic stem cell transplantation: graft-versus-host disease versus true dermatomyositis

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Introduction & Objectives:

Chronic graft-versus-host disease (cGVHD) is an alloimmune and autoimmune complication of hematopoietic stem cell transplantation (HSCT). It is well documented that cGVHD may develop in some cases after the allo-HSCT, representing a variety of symptoms closely similar to those in autoimmune diseases. Sometimes true autoimmune diseases can also develop. The occurrence of autoimmune diseases after allogeneic HSCT is infrequent and difficult to interpret due to the reconstitution of the immune system and the multifactorial origin of most of these diseases.

We presented a case of a 45-year-old-woman who developed a cGVHD with identical cutaneous manifestations to dermatomyositis.

Materials & Methods:

A 45-year-old-woman with a history of acute myeloid leukemia was treated with an allogenic HSCT. She developed an acute graft-versus-host disease with cutaneous involvement with good response to steroids and sirolimus.

Six months later she started with cGVHD intestinal symptoms. In addition, scaling plaques in scalp and adjacent to the back of metacarpophalangeal and interphalangeal joints appeared. She had similar scaly plaques at the elbows and on the eyelids.

A cutaneous biopsy was performed showing histological findings compatible with cGVHD (interface dermatitis with epidermal hyperplasia) without specific data of dermatomyositis. This supported the diagnosis of dermatomyositis-like cGVHD. Muscle symptoms were not reported by the patient. CK and aldolase levels were normal. Antinuclear antibody testing was negative with decreased levels of C3. Myositis antibody-specific blot was negative.

One year after transplantation, the patient died of respiratory sepsis with a torpid course.

Results:

Dermatomyositis-like presentations after HSCT are very rarely reported. It can appear between 4 and 52 months after HCST. Skin manifestations could be identical to dermatomyositis. Cases with muscle involvement confirmed by muscle biopsy and severe cases with progressive interstitial lung disease associated with MDA5 antibodies have also bed described.

The distinction between dermatomyositis-like cGVHD and a true dermatomyositis is complex. The isolated finding of complete chimerism of donor lymphocytes in peripheral blood of the receptor does not allow establishing the diagnosis of GVHD due to the persistence of receptor cells with immunogenic potential. It has been suggested that the determination of chimerism in the immune cells of the affected tissue could help to distinguish between

GVHD and an autoimmune disease. FISH analysis on a muscle specimen can determine whether there is a predominance of CD4+ T lymphocytes from the recipient (in favor of an autoimmune process) or CD8+ lymphocytes from the donor (in favor of graft-versus-recipient disease).

Conclusion:

Our patient presented chimerism of 97% of donor lymphocytes in peripheral blood. No specific data of dermatomyositis was observed on skin biopsy. Given the fatal clinical course, no muscle biopsy was performed. Therefore, the diagnosis of dermatomyositis-like cGVHD was performed corresponding to an alloimmune process associated with HSCT.

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Abstract N°: 5222

Association between trichoscopic vascular findings and systemic sclerosis-associated interstitial lung disease in a tertiary hospital in Mexico

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Introduction & Objectives:

Systemic sclerosis (SSc) is a chronic autoimmune disease of the connective tissue with involvement of multiple organs and systems, in which, pulmonary manifestations are present in 50-65% of patients, being interstitial lung disease (ILD) the main cause of mortality in this patients. Although diagnostic tools such as capillaroscopy offer a direct view of microangiopathy in SSc, there is growing interest to develop additional methods that expand our set of diagnostic tools. In this effort, trichoscopy is establishing itself as a non-invasive diagnostic method with great potential, by utilizing the dense vascularization and easy accessibility of the scalp to evaluate vascular changes associated with the disease.

Our main objective was to determine the association between trichoscopic vascular findings and systemic sclerosis-associated interstitial lung disease (SSc-ILD). Our secondary objectives were examine the association of SSc subtypes, Non-Raynaud symptoms and specific antibodies observed in SSc and its assosiation with trichoscopic vascular findings.

Materials & Methods:

We performed an observational, cross-sectional, analytical study examining the relationship between the development of interstitial lung disease and trichoscopic findings in patients with systemic sclerosis. Our study encompassed 40 individuals diagnosed with systemic sclerosis, among whom 23 had SSc-ILD, as verified by computed tomography (CT) scans. For each patient, we captured six high-resolution trichoscopy images at six distinct points along the midline of the scalp, from the frontal to the occipital region, using a DL5 dermatoscope and an iPad. We conducted descriptive statistical analyses, Fisher's exact test and a multiple logistic regression model to achive our research objectives.

Results:

The predominant trichoscopic vascular patterns identified were avascular areas (seen in 70.58% of patients without ILD vs. 91.30% with ILD) and polymorphic vessels (29.42% without ILD vs. 8.70% with ILD). However, the study found no significant correlation between the subtype of systemic sclerosis, antibodies, initial non Raynaud symptom with the most prevalent trichoscopic vascular patterns ($p = 0.09, 0.6774, 0.2093$). Table 1. Additionally, the prevalent vascular patterns did not show a statistically significant connection with the presence of SSc-ILD ($p = 0.18$). Table 2.

Conclusion:

Our investigation did not reveal a significant correlation between trichoscopic vascular findings and the presence of SSc-ILD. Further research with larger sample sizes and advanced diagnostic tools is necessary to confirm these

associations. Nonetheless, this research is pioneering in its efforts to explore such a connection.

Tables and figures

Association of studied variables with tricoscopic vascular findings

Variables	Avascular areas	Polimorfic vessels	
SSc subtype			**P=0.09**
ESD	19 (95%)	1 (5%)	
ESL	14 (70%)	6 (30%)	
Antibodies			**P=0.6774**
Anti-centromere	4	2	
AntiSCL-70	4	1	
Unspecific (ANA)	20	4	
None	5	0	
First Non Raynaud symptom			**P=0.2093**
Arthalgias	11	0	
Esophageal dysmotility	1	0	
Fever and headache	0	1	
None	1	0	
Salt and pepper pigmentation	2	0	
Puffy fingers	14	5	
Digital ulcers	4	1	

Table 1. Association of SSc subtype, antibodies, first Non Raynaud symtom and trichoscopic vascular findings.

Relationship of studied variables with interstitial lung disease Comparison of two logistic models						
Variable	Model with best AIC			Complete model		
	OR	95% CI	p-value	OR	95% CI	p-value
Fitzpatrick Phototype			**0.011**			**0.007**
III	-	-		-	-	
IV	0.17	0.00,2.69		0.00		
V	0.01	0.00,0.24		0.00		
Modified Rodnan score	1.54	1.11,2.57	**<0.001**	2.35	1.13,35.7	**0.006**
Sex			0.11			0.33
Male	-	-		-	-	
Female	0.00			0.00		
SSc subtype			**0.007**			0.73
LSSc	-	-		-	-	
DSSc	16.8	2.06,2.67		2.06	0.02,137	
Time of evolution				1.19	0.70, 2.50	0.53
Antibodies						**0.025**
Anti-centromere				-	-	
Anti-SCL-70				>10	NA	
Most frecuent trichoscopic finding						0.18
Aeas avascular areas				-	-	
Polimorfic vessels				0.03	0.00, 3.44	
AIC		34.3			48	

Table 2. Analysis of the best suggested statistical model and the complete logistic regression model.

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Abstract N°: 5345

An uncommon cause of a persistently pruritic eruption in a young lady with fever

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Introduction & Objectives:

Adult onset Still's disease (AOSD) is a rare systemic inflammatory disorder, mainly affecting young females. It is characterized by spiking fevers, arthralgias, multiorgan involvement and varied cutaneous manifestations. Although the most common cutaneous manifestation of AOSD is an evansescent rash, persistent pruritic eruptions (PPEs) - which include urticarial lesions, generalised erythema, prurigo pigmentosa-like eruption and flagellate erythema may serve as clinical mimics of other inflammatory conditions such as dermatomyositis. This case report describes a case of PPE affecting a young lady with AOSD.

Materials & Methods:

A 29-year-old female presented with 3 weeks of fever, arthralgia and pruritic rashes on her back, anterior chest, upper arms, and thighs. She had no significant medical history and no family history of autoimmune conditions. She denied occurrence of muscle weakness, dysphagia, mouth ulcers, alopecia and sicca symptoms. Physical examination revealed erythematous, edematous papules and plaques on the anterior sternum, upper to mid-back, deltoids, flanks and lateral aspect of proximal thighs. She did not exhibit Gottron's sign, nor had a heliotrope rash. Her palms and soles were uninvolved. There was no palpable lymphadenopathy nor hepatosplenomegaly.

Laboratory analysis of anti-nuclear antibody (ANA), anti double-stranded DNA(ds DNA), ANCA, rheumatoid factor were negative. Complements were normal. Anti-extractable nuclear antigen (ENA) antibodies were negative. Creatine kinase, aldolase levels were normal and myositis panel was negative. Erythrocyte sediment rate (ESR) was elevated (48 mm/hr). Ferritin was elevated (580 ug/L; range 5-204 ug/L).

Results:

Histological examination of the plaque on her upper back revealed epidermal dyskeratosis, with a mixed perivascular infiltrate of neutrophils, lymphocytes and histiocytes within the superficial dermis. There was prominent nuclear dust and red cell extravasation. There was no fibrinoid necrosis of the capillaries. There was increased dermal mucin highlighted by Alcian blue staining. Direct immunofluorescence was negative. Taken together, this patient was diagnosed with persistent and pruritic eruption of AOSD. She was treated with regular non steroidal anti-inflammatory agents with significant resolution of fever and arthralgias.

Conclusion:

The diagnosis of AOSD can be challenging and often delayed due to its protean clinical manifestations. As with its clinical features, a broad histologic spectrum has been reported in AOSD. Of the possible histological manifestations reported in AOSD, the presence of upper epidermal dyskeratosis with apoptotic keratinocytes and a mixed lymphocyte and neutrophil infiltrate in the superficial dermis has been reported to be fairly specific for PPE in AOSD. As the presence of PPEs in AOSD has been associated with the development of a secondary macrophage activating syndrome, conferring a worse prognosis and higher mortality rates, this case is a reminder of the importance of clinopathological correlation and recognition of PPEs in AOSD.

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Abstract N°: 5361

Hirsutism presenting as cutaneous manifestations of an underlying undiagnosed condition : report of 2 cases

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Introduction & Objectives:

Hirsutism is a clinical indicator of androgen excess, and polycystic ovary syndrome (PCOS) is one of the common causes. Other causes include hypothyroidism, insulin resistance, and ovarian and adrenal tumors. Laser hair removal (LHR) is available at beauty salons, and spas done by non-experts at a relatively lesser cost some patients tend to choose them. These centers may not evaluate the underlying cause of hirsutism and sometimes may miss underlying undiagnosed hormonal imbalances and tumors. Here we report 2 such cases.

Case report:

Case 1: A-47-year-old married lady presented with terminal hair growth over her face, chin, chest, and abdomen gradually for 2 to 3 years with irregular menstrual cycle. She had a history of phyllode tumor of the left breast, operated 11 years ago. There was no history of abdominal pain or any other complaints. She underwent 4 sessions of LHR at the local center without any improvement. On further history and examination, she had signs of virilization like hoarseness of voice, increased libido, and clitoral hypertrophy. On investigations, she had markedly raised serum testosterone (612ng/dl, normal range 23-73 ng/dl), and enlarged ovaries on ultrasonography(right ovary volume of 37cc). MRI of abdomen pelvis with well-defined T1 isointense T2 predominantly hypointense lesion in right adnexa size 3.3X3.7X3.2 mm. The right ovary is not seen separately from the lesion. The patient underwent excision of the lesion with total abdominal hysterectomy with bilateral salpingo-oophorectomy. The excised specimen showed a tumor of sex cord origin- Sertoli cell tumor. Post-operatively testosterone levels returned to normal levels. There was clinical improvement in hirsutism, reduction in clitoral hypertrophy, and normal libido. The patient underwent 3 sessions of laser hair removal 3 months after surgery with significant improvement.

Case 2: A 38-year-old unmarried female presented with terminal hair growth over her lower face for 4 to 5 years associated with an irregular menstrual cycle. She was treated with oral contraceptive pills for 3 to 4 months duration multiple times in the past with regularization of the menstrual cycle. For the last 6 months, she was on progesterone pill. She was treated with 11 sessions of diode LHR earlier without any improvement. On inquiry, she gave a history of dysmenorrhea and metrorrhagia but no history of persistent abdominal pain. Her hormonal evaluation was unremarkable with minimally raised HbA1C levels. Her ultrasonography revealed a bilateral ovarian endometriotic cyst and pelvic endometriosis with infiltration of the rectal wall. She underwent bilateral ovarian cystectomy, excision of deep infiltrative endometriosis, and laparoscopic-assisted bowel segment resection and anastomosis.

The patient was started on an injection of leuprolide 3.75 mg once a month post-surgery. LHR started after 2 months of surgery with significant improvement.

Conclusion:

Hisrustim in females is commonly attributed to PCOS. Sometimes perimenopausal patients of hirsutism are not

routinely investigated. Detailed history and examination are very important to suspect the possibility of rare diseases like ovarian tumors and endometriosis as a cause of hirsutism.

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Abstract N°: 5477

Primary systemic amyloidosis: the skin as a guiding sign for diagnosis

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Introduction & Case Report:

We present the case of a 65-year-old woman who attended the Dermatology clinic due to the progressive appearance, over the past four years, of cutaneous lesions, asthenia, and morning paresthesias in both hands. Physical examination revealed ecchymotic and purpuric macules on the upper and lower eyelids, retroauricular region, neck, inframammary folds, and pubis. Macroglossia and residual cutaneous lesions in the form of atrophic brownish macules were also observed.

A skin biopsy was obtained from the cervical region, showing alterations in dermal elastic fibers, without observing amyloid deposits after specific histochemical and phenotypic techniques. Blood tests with protein electrophoresis revealed a significant elevation of lambda light chains in serum and a decreased kappa/lambda ratio. Urinalysis showed no evident abnormalities. Bone marrow biopsy was performed, revealing infiltration of 1.36% by aberrant clonal plasma cells of lambda type, with positivity for the t(11;14) mutation.

Discussion:

The patient was diagnosed with primary systemic amyloidosis associated with primary AL-type plasma cell dyscrasia.

Multiple Myeloma was ruled out since the patient did not meet diagnostic criteria: monoclonal plasma cells did not exceed 10%, and no organ damage secondary to plasma cell proliferation was evident: the blood count showed no abnormalities in the red series, serum calcium was within the normal range, renal function was preserved, and bone damage in the form of osteolytic lesions or osteoporosis was ruled out.

Additional tests were performed to rule out amyloid deposition at other levels, with normal results on echocardiography and whole-body computed tomography.

Our patient is pending a gammagraphy, an abdominal fat and rectal biopsy, and nerve conduction studies for a possible bilateral carpal tunnel syndrome.

Conclusion:

Raccoon eyes and macroglossia are typical dermatological signs of amyloidosis and often appear as the first manifestation of the disease, serving as a guide for diagnosis.

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Abstract N°: 5583

Generalized Dowling-Degos Disease: Case Report

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Introduction & Objectives: Degos disease, or malignant atrophic papulosis, is a very rare but deadly multisystemic vasculopathy, of unspecified etiology, characterized by the presence of papular skin lesions, with an atrophic center, white as porcelain, surrounded by telangiectatic border.

Materials & Methods: We present the case of a 45-year-old patient, addressing the Department of Dermatovenereology for a skin rash, consisting of multiple tender papules with erythematous, elevated edges, having an atrophic, white center, disseminated on the trunk, limbs and genitals. Upon closer examination, some lesions were ulcerated , covered by hematic crusts. The patient had two skin biopsies prior to this current presentation: the first indicated urticaria and the second presented histopathological aspects compatible with the diagnosis of pityriasis lichenoides et varioliformis acute (PLEVA).

Results: In our department, the patient underwent a new skin biopsy which revealed small congestive vessels and agglutinated red cells (thrombi) in the dermis, a vasculopathy, supporting the clinical diagnosis – Degos disease. Considering the associated simptoms: alleging dysphagia, rapid significant weight loss, diffuse muscle pain, and episodes of constipation alternating with diarrhea, our patient was also examined by a multidisciplinary team.

The patient followed systemic treatments with anticoagulants, xanthine derivatives, diuretics, analgesics and antiseptics. Bacitracin zinc powder and neomycin sulfate, as well as sucralfate powder were applied topically, with a slow but favorable evolution of the skin lesions. Unfortunately, the patient died due to cardiovascular complications in the following 6 months.

Conclusion: Degos disease is generally considered a serious vascular disease because it affects multiple organs and can be fatal within 2-3 years (intestinal perforation and secondary peritonitis being the most common cause of death). But in some cases, the disease is limited to the skin and has a favorable prognosis. Diagnosis is based on identifying pathognomonic skin lesions and performing a skin biopsy.

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Abstract N°: 5716

Scleredema diabeticorum – a rare connective tissue disorder associated with diabetes

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Introduction & Objectives:

Scleredema diabeticorum (SD) is an uncommon skin disorder caused by enhancement of collagen synthesis by fibroblasts and a reduced degradation of collagen with consequent reduction of skin elasticity. SD is characterized by severe permanent thickening of the skin of the posterior neck and upper back, what leads to reduced range of motion.* The skin thickening develops over years and presents like peau d'orange. More than 90% of those with this skin condition have diabetes. SD remains a difficult and resistant disease to treat.

Materials & Methods:

We present a case of 58-year-old woman with 13-years history of uncompensated diabetes mellitus type 2 who developed severe scleredema diabeticorum. We also discuss possible therapeutic options.

Results:

A 58-year-old woman was admitted to the Dermatology Department with disseminated skin tightness involving upper back, chest, cheeks and hands. She also demostrated severly decreased range of motion in affected areas. Laboratory tests showed a fasting blood glucose of 133 mg/dl [70-99 mg/dl], a glycated hemoglobin concentration of 9.0% [4.0-6.0]. Ultrasonography described an extensive subcutaneous lesion involving the upper back with homogeneous fibrous lesions. The biopsy confirmed the diagnosis of scleredema. Possible therapeutic options include phototherapy, glucocorticosteroids, immunomodulatory drugs (such as methotrexate), IVIG. Each patient should also be referred to diabetologist and physiotherapist.

Conclusion:

The early recognition of SD is important as it can cause a significant impairment of joint mobility over the years. This rare condition treatment is difficult and tricky, as no specific regimen has been instituted to treat this skin disease.

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Abstract N°: 5775

Acanthosis nigricans and retinitis pigmentosa: An indicator of progressive multi-organ dysfunction due to ciliopathy.

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Introduction & Objectives: Ciliopathies are characterised by rod-cone dystrophy associated with insulin resistance, polyendocrinopathy and progressive cardiac, pulmonary, hepatic and renal dysfunction. Life expectancy is shortened. The age of onset and severity of symptoms is extremely variable, even with the same mutation.

Materials & Methods: Two unrelated young adult males presented with generalised acanthosis nigricans to dermatology at the ages of 18 years and 21 years respectively. Both patients had progressive visual loss due to retinitis pigmentosa. They were both of non-consanguineous parentage and had no similar history in their respective families.

Patient 1 also had bilateral sensorineural hearing loss and retrognathia. On evaluation, he had type 2 diabetes mellitus, dyslipidaemia and hypogonadism.

Patient 2 was found to have type 2 diabetes mellitus, dyslipidaemia, hypogonadism and non-alcoholic steatohepatitis.

A whole exome sequencing was done for both patients.

Results: Pathogenic homozygous mutations in exon 8 of ALMS1 gene were found in both patients, confirming the diagnosis of Alstrom syndrome.

Conclusion: The clinical presentation is similar in both Alstrom syndrome and Bardet-Biedl syndrome. Alstrom syndrome occurs due to mutation in a single gene ALMS1, usually in exons 10,16 or 8. Bardet-Biedl syndrome is associated with mutations in more than 20 genes.

We present this case to highlight the importance of suspecting ciliopathies in patients with early-onset insulin resistance and rod-cone dystrophy. Early management of organ dysfunction can prolong life expectancy.

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Abstract N°: 5788

Clinical and Temporal Characteristics of Carcinoma Erysipeloides: A Systematic Review

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Introduction & Objectives:

Carcinoma erysipeloides (CE) is a rare cutaneous manifestation of metastatic cancer where tumor cells infiltrate dermal lymphatics, mimicking erysipelas. CE has been reported to be associated with advanced-stage breast adenocarcinoma.1 Recognition of CE can often be difficult, but early diagnosis of CE is essential. CE has been described in the literature, but review of its clinical presentation and outcomes is limited. Thus, the goal of this systematic review was to summarize the clinical characteristics and treatment outcomes of CE arising from varied cases of metastatic carcinoma.

Materials & Methods:

Embase, Web of Science and PubMed were screened for CE cases following PRISMA guidelines. Data were pulled by two independent researchers, with any discrepancies settled by a third. Non-English and review articles were excluded.

Results:

72 case reports/series describing 83 total patients were included. Most patients were female (n=51/82, 62.2%), with an average age of 63 years. Most CE cases were diagnosed after the primary malignancy (n=63/79, 79.7%) with a median diagnosis time of 24 months after identification of the primary cancer. The remainder of cases were diagnosed concurrently (n=13/79, 16.5%) or before (n=3/79, 3.8%). The most reported symptoms experienced by patients included tenderness (n=21/69, 30.4%), warmth (n=15/69, 21.7%), pain (n=12/69, 17.4%), itching (n=11/69, 15.9%), and swelling (n=9/69, 13.0%), and 15 patients were asymptomatic (n=21.7%). Lesion morphology was most commonly described as erythematous (n=67/83, 80.7%), plaque-like (n=45/83, 54.2%), indurated (n=27/83, 32.5%), papular (n=14/83, 16.9%), and violaceous (n=8/83, 9.6%). Across patients, CE occurred on the chest/breast (n=46/83, 55.4%), extremities (n=21/83, 25.3%), abdomen (n=18/83, 21.7%), neck (n=14/83, 16.9%), back/buttocks (n=10/83, 12.0%), genitals/pubic region (n=9/83, 10.8%), and head/face (n=3/89, 3.4%).

The most reported primary malignancies were breast cancers (n=35/83, 44.3%), lung adenocarcinoma (n=9/79, 11.4%), gastric cancers (n=6/79, 7.6%), and cutaneous squamous cell carcinoma (n=4/79, 5.1%), with CE being associated with a first cancer (n=52/83, 62.7%) more often than recurrent (n=31/83, 37.3%). Biopsy results described atypical cells within dermal lymphatics the majority of the time (n=60/83, 72.3%), with some reports describing atypical cells invading the dermis (n=41/83, 49.4%) and subcutaneous tissue and vessels (n=11/83, 13.3%).

Treatment of the primary malignancy included combinations of surgery (n=41/78, 52.6%), chemotherapy, (n=48/78, 61.5%), and/or radiotherapy (n=22/78, 28.2%). Of the reports commenting on treatment outcomes, 19/53 patients died within 6 months of treatment initiation (35.8%), 18/53 showed no improvement or spreading of their CE (34.0%), 14/53 showed improvement (26.4%), and 2/53 initially improved then relapsed (3.8%).

Conclusion:

This study emphasizes the importance of early recognition of carcinoma erysipeloides, as it is associated with poor outcomes attributable to underlying malignany. Treatment remains ineffective for many cases of CE, and future studies should review treatment strategies that are most effective at managing this condition.

References:

\1. Finkel L j., Griffiths C e. m. Inflammatory breast carcinoma (carcinoma erysipeloides): an easily overlooked diagnosis. British Journal of Dermatology. 1993;129(3):324–6.

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Abstract N°: 5849

Skin clues of a systemic desease

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Introduction & Objectives:

Sarcoidosis is a multisystem granulomatous disease of unknown etiopathogenesis that frequently affects the skin. The skin manifestations of this entity can be specific, with the formation of non-caseating granulomas, or nonspecific. We present the case of a patient with sarcoidosis whose skin symptoms were closely related to her systemic involvement.

Materials & Methods:

Our patient was a 64-year-old woman diagnosed in 2021 with sarcoidosis with skin and lung involvement. Treatment consisted in systemic corticosteroid therapy. Two years later, an exacerbation of her cutaneous involvement occurred, coinciding with a worsening of her baseline disease.

On examination, the new lesions consisted of violaceous plaques of about two centimeters in diameter, circinate, and with a patchy distribution affecting trunk, upper and lower limbs, and face.

Given the clinical suspicion, the lesions were biopsied. The pathological study showed the presence of non-caseating granulomas, allowing these lesions to be identified as specific for sarcoidosis and confirm the suspected diagnosis.

Results:

Sarcoidosis is a multisystem granulomatous disease of unknown etiopathogenesis. Cutaneous manifestations can precede systemic symptoms in up to 80% of patients, although in 30% of cases it is the only manifestation of the disease. In cutaneous sarcoidosis, we distinguish specific and nonspecific manifestations. The former are characterized by the formation of non-caseating granulomas that are usually asymptomatic. They most commonly consist on papular and nodular forms, although there is a wide range of clinical presentations that include lupus pernio and subcutaneous, hypopigmented or ulcerated forms, among others. Regarding non-specific manifestations, it is worth highlighting erythema nodosum type panniculitis. Treatment must be considered in symptomatic cases, with extensive involvement, ulceration or with important aesthetic repercussions, and therapeutic options include topical, intralesional and/or oral corticosteroids. In refractory cases, methotrexate must be considered.

Conclusion:

In short, skin lesions of sarcoidosis are an accessible clinical manifestation that can sometimes facilitate the diagnosis of this disease, especially when it is the first manifestation of this entity.

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Abstract N°: 5873

Phanerian presentations and dermoscopic findings in Lupus

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Introduction & Objectives:

Scalp changes are prevalent in systemic and cutaneous lupus, often occurring during active disease phases. Conversely, nail changes, despite their variety, have received limited study in this context. Our aim is to examine the clinical and dermoscopic features of phanerian involvement in lupus.

Materials & Methods:

This study prospectively analyzed 30 patients diagnosed with both Cutaneous Lupus Erythematosus (CLE) systemic Lupus Erythematosus (SLE), specifically examining manifestations affecting the nails and scalp.

Results:

We examined a sample comprising 30 lupus patients, noting anomalies on both the scalp and nails. The average age of the patients was 40 years (range 10–61 years), with the majority being women (26 or 86.6%). Among these patients, 17 (56.6%) were diagnosed with Cutaneous Lupus Erythematosus (CLE) and 13 (43%) with Systemic Lupus Erythematosus (SLE). Within specific lupus erythematosus (LE) manifestations. Scarring alopecia was observed in 11 (36.6%) patients, including 10 (33%) cases of discoid lupus erythematosus (DLE), one case of lupus panniculitis.

Moreover, non-scarring alopecia was documented in 13 (43.3%) patients, characterized by patchy non-scarring alopecia in 9 patients and diffuse non-scarring alopecia in 2 cases. Among the nonspecific LE presentations, and one case presented a combination of frontal fibrosing alopecia and LE; one patient exhibited alopecia areata universalis, while 5(16.6%) patients presented solely with Telogen Effluvium (TE), we must note that all of the female patients in our study presented TE, no cases of lupus hair or

lichenoid lupus panniculitis were detected in our sample. The most common trichoscopic findings included fine hair in 46.6% of cases, thick arborizing vessels in 33.3%, and follicular red dots (inverse strawberry pattern) in 26.6%. Wide yellow dots or keratin plugs were found in 13.3% of cases, and honeycomb pigmentation in 30%. Additionally, a perifollicular whitish halo was seen in 10% of cases, while whitish scales were present in 16.6%. Interfollicular erythema areas and whitish regions lacking structures were also observed.

Regarding nail involvement, periungual alterations were the most common, noted in 16 cases (53.3%). Periungual erythema was predominant, occurring in 9 cases (30%), followed by 2 cases (6.6%) each of atrophic erythema with scales, cuticular dystrophy, and paronychia. Accentuation of longitudinal relief was observed in 12 cases (40%), koilonychia in 2 cases (6.6%), and singular occurrences of onychorrhexis and pluridigital melanonychia in a 32-year-old lupus patient. Additionally, one patient exhibited a red lunula. Dermoscopic assessment of the periungual region revealed cuticular microhemorrhages in 3 cases (10%) and flame-shaped hemorrhages in 2 cases (6.6%).

Discussion:

Trichoscopic characteristics in lupus vary based on type and stage, aiding diagnosis of associated alopecia. Nail

involvement, while not exclusive to lupus, requires diagnosis due to potential severe consequences like nail dystrophies. Dermoscopy facilitates examination of periungual microcirculation abnormalities in systemic lupus, aiding diagnosis and treatment evaluation, as improvements in nail involvement often follow lupus treatment**.**

Conclusion:

Dermoscopy is pivotal in diagnosing lupus-related phanerian involvement, aiding in diagnosis, stage assessment, and treatment response evaluation.

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Abstract N°: 5881

Cutaneous manifestation of cryoglobulinemic vasculitis associated with hepatitis C virus: A case report

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Introduction & Objectives:

Mixed cryoglobulinemia syndrome (MCS) is a systemic small-vessel vasculitis, can involve a myriad of organs that includes most commonly skin, joints, peripheral nervous system, and kidneys [1]. We hereby discuss a case of a MCS secondary to HCV presenting with lush cutaneous manifestations.

A 63 years old female with a history of breast cancer reported small and medium joints, additive, symmetric polyarthralgia that began 3 years ago. A year ago she also noted cutaneous lesions in her lower limbs, which made her seek medical care. She was then diagnosed with rheumatoid arthritis and started hydroxychloroquine with resolution of the joint condition, but with worsening of the related injuries on her skin. Confluent hyperchromic spots with a lacy pattern appear on the lower limbs, characterized by livedo racemosa, with painful ulcers.

Materials & Methods:

Serum testing

Antinuclear Antibody negative

Rheumatoid factor 128

Cyclic citrullinated peptide) antibodies negative

C4 1.5

Anti-HCV Positive

PCR HCV 4.495 UI/ml - Log 6,65

Serum cryoglobulin test was positive and histopathological analysis of skin biopsies showed areas of epidermal straightening, discrete perivascular mononuclear inflammatory infiltrate, ulceration crusts, irregular acanthosis, vascular neoformation with thickened walls and fibrosis. In view of these findings, the diagnosis of cryoglobulinemic vasculitis (CV) associated with HCV was established.

Results:

Corticosteroid therapy was prescribed to acute control of the skin lesions. Due to the unavailability of rituximab at the time and significant improvement in skin lesions with corticotherapy, Veltapasvir/Sofosbuvir were started for HCV and the patient was discharged for outpatient follow-up to assess the future need for rituximab.

The disease may manifest as a wide spectrum of symptoms. Necrotizing skin ulcers and purpura are non-healing cutaneous lesions. [1,2] Renal involvement, articular manifestations and neuropathy may also occur.

There are no established diagnostic criteria for MCS. Positive serum cryoglobulins (cryoprecipitate > 1% or 50

mcg/L) associated with clinical indicators of cryoglobulinemic vasculitis or thrombosis with multisystem involvement and decreased C4 support the diagnosis. Positive serum cryoglobulins associated with histological and immunohistochemical is equally enlightening of the diagnosis [3].

Treatment of patients with HCV-positive CV is based on disease activity and severity of symptoms. In patients with mild to moderate manifestations, an anti-HCV direct-acting antiviral (DAA) is often highly effective. Given the pangenotypic effects, sofosbuvir/velpatasvir is recommended in CV patients with HCV of unknown genotype. In those with widespread vasculitis, including renal involvement and severe neuropathy, a combination of DAAs and non-HCV-directed agents is suggested, such as glucocorticoids, plasma exchange, or rituximab [4].

Conclusion:

Cryoglobulinemic vasculitis is a rare disease with variable presentation, often presenting with skin lesions. It is essential that clinicians and dermatologists evaluating these patients have a high degree of diagnostic suspicion.

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Abstract N°: 5942

Case Series of Rare Metastatic Crohn's Disease

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Introduction & Objectives:

Extraintestinal manifestations are common in Crohn's disease [CD], with an incidence ranging from 22% to 44%, and its clinical spectrum is broad. Among these manifestations, metastatic Crohn's disease [MCD] is notably uncommon. It represents a rare granulomatous inflammatory process, sharing similarities with the pathogenic mechanism of CD but occurring at sites discontiguous from the gastrointestinal tract. Herein we report a rare case of MCD involving (a) the face in the form of Postherpetic isotopic response (PHIR) and (b) the breast.

Materials & Methods:

Case 1: A 45-year-old Caucasian woman presented with an eruption of itchy, confluent in places, smooth surfaced erythematous papules, located on the forehead, nasopharyngeal groove, lateral wall of the nose and upper lip, over resolved herpes zoster lesions (HSV1). Two days prior to presentation, the patient experienced HSV1 with V1-V2 distribution, involving the right side of her face. She was on adalimumab for 14 months before the onset of HSV. For the first 8 months of adalimumab treatment, she received the maintenance dose for Crohn's disease, i.e., 40 mg every 2nd week. Subsequently, for the following 6 months (till HSV onset), she received 40 mg weekly due to the appearance of hidradenitis suppurativa type lesions on her thighs, abdomen, and genitalia associated with CD. There was a notable response and lesional regression during the increased dose period.

Case 2: A 35-year-old female with a history of CD presented with a painful, ulcer with a shiny, erythematous base involving the inframammary skin. The rash appeared approximately one month prior and progressively worsened since its onset. An erythematous patch without ulceration was present under the right breast. The patient stated that she had applied topical hydrocortisone under her breasts without relief. She denied any active gastrointestinal symptoms.

Results:

Case 1: There was a great remission of skin lesions after 2 weeks of application topically cream mometasone furoate cream twice daily. Metastatic cutaneous Crohn's disease (MCD) is defined by the presence of cutaneous granulomatous lesions non-contiguous with the gastrointestinal tract or fistulae. Postherpetic isotopic response describes the occurrence of a new, unrelated disease that appears at the same location as the previously healed herpetic infection.

Case 2: Cutaneous metastatic CD, presenting as ulcerated skin plaques, papules, or erythematous nodules, has been reported rarely to be peri-areolar or sub-mammary. In patients with known CD, metastatic granulomatous involvement should be considered in the differential diagnosis of a peri-mammary skin lesions, breast mass, or non-caseating breast granuloma.

Conclusion: MCD, characterized by non-caseating granulomas in extraintestinal sites, with facial or breast involvement is exceptional. Since clinical presentation of MCD varies, it may lead to under-recognition and

misdiagnosis.

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Abstract N°: 6085

Erythema elevatum diutinum and pyoderma gangrenosum associated with chronic myelomonocytic leukaemia: a case report

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Introduction & Objectives:

Neutrophilic dermatoses are often associated with haematological malignancies, but their co-occurrence is rare in the literature, and they are rarely seen before a haematological diagnosis is made. The authors present the case of a 64-year-old female patient who developed erythema elevatum diutinum and pyoderma gangrenosum in association with chronic myelomonocytic leukaemia.

Materials & Methods:

Results:

The female patient initially developed extensive, confluent, infiltrated, erythematous papules on the trunk. In parallel, multiple greyish-brown hyperkeratotic nodules developed on both elbows. Histopathological examination of the trunk showed neutrophilic infiltration and leukocytoclasia with fibrinoid vessel wall necrosis in the dermis. Lesions from the elbow showed marked fibrosis, capillary proliferation and macrophage, plasma cell and lymphocytic infiltration. Based on the clinical and histological picture, a diagnosis of erythema elevatum diutinum was made. The patient's routine laboratory tests showed a significantly elevated white blood cell count and monocytosis. A Jamshidi bone marrow biopsy was performed with the involvement of a haematologist, which described abnormal myeloid cells with MPO and CD68 co-expression both in the bone marrow and periosteal tissue. The bone marrow image confirmed the diagnosis of chronic myelomonocytic leukaemia, for which he was treated in the haematology department. During her hospital stay, she developed septic symptoms for which she was started on combined antibiotic therapy and discharged home after her blood count stabilized. After discharge, she developed a markedly painful ulcer on her right forearm following minor trauma, with a characteristic purplish colour and undermined margins, which tripled in size in 3 days. There was also a small laceration of similar morphology on the left forearm. A pronounced dermal neutrophilic infiltration with mild leukocytoclasia was seen on histological examination of the wound margin. Laboratory tests showed markedly elevated inflammatory parameters. A pathergy skin test on the opposite forearm was positive. Given the presence of major and minor criteria, a diagnosis of pyoderma gangrenosum was made. Due to oedema in the affected limb, a Doppler ultrasound study was performed, confirming a cephalic vein thrombosis, and a therapeutic dose of low-molecular-weight heparin was prescribed. Systemic dapsone was started with marked improvement in her skin symptoms and the forearm ulcer began to slough off.

Conclusion:

In this case, it is noteworthy that dapsone therapy can lead to a significant improvement in both neutrophilic dermatoses. It is also important to draw attention to the recognition and appropriate management of other conditions often associated with the disease, such as upper limb thrombosis as described in this case.

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Abstract N°: 6139

Squamous Cell Carcinoma of the Lips in Chronic Discoid Lupus.

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Introduction & Objectives:

Lip carcinomas are relatively common, comprising nearly 10% of skin cancers. Typically associated with risk factors like sun exposure or smoking, they often arise following a precancerous stage, commonly characterized by actinic or smoker's cheilitis. However, it's considerably rarer for squamous cell carcinoma (SCC) to develop on chronic glandular cheilitis, lichen planus lesions, or chronic lupus. Here, we present a case of squamous cell carcinoma occurring on undiagnosed chronic lupus evolving over a period of 20 years.

Materials and Methods/Observations:

A 62-year-old patient, fair-skinned and former smoker, with a history of multiple squamous cell carcinomas. The first was on the upper lip, operated on 5 years ago, followed by the lower lip operated on 4 months ago. The patient has had erythematous-squamous lesions on sun-exposed areas for 20 years, along with a history of neglected photosensitivity.

Clinical examination revealed erythematous-squamous plaques with fine telangiectasias on the extension surfaces of both hands, featuring a hypopigmented and atrophic center and sharp, slightly hyperpigmented edges. On the face, there were two surgical scars and an erythematous-atrophic plaque on the nasal ridge with central hypopigmentation and peripheral hyperpigmentation accompanied by some scales.

Dermoscopic examination of the lesions showed an erythematous background with telangiectasias, rosettes, pinpoint vessels, peripheral brownish pigmentation, whitish scales, and dyschromia. The remainder of the dermatological and clinical examination was unremarkable.

Histological examination was compatible with chronic lupus, while immunological assessment was negative. Based on the clinical and complementary examinations, we concluded that squamous cell carcinomas developed on lesions of chronic lupus.

Results/Discussion:

Squamous cell carcinomas account for 90% of lip cancers, nearly 10% of skin cancers, and 1.7% of upper aerodigestive tract (UADT) cancers. They predominantly occur on the vermilion of the lower lip, affecting mostly men over 60 years old, with a sex ratio of six to nine. Squamous cell carcinoma of the lips often arises from cutaneous precancerous lesions, with its occurrence on chronic erythematous lupus (CEL) lesions being rare but described on lesions of chronic cheilitis due to CEL. Indeed, chronic inflammatory processes, scarring, atrophy, depigmentation, UV damage, and tobacco use can initiate the precancerous transformation of keratinocytes during CEL.

Conclusion:

CEL skin lesions represent a precancerous condition of squamous cell carcinomas. Though the risk of degeneration is minimal, monitoring these lesions is crucial, particularly in regions with significant sunlight exposure and low health literacy levels.

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Abstract N°: 6150

Masquerading Metastasis: Not just another Pyogenic Granuloma on the Nose

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Masquerading Metastasis: Not just another Pyogenic Granuloma on the Nose

Introduction & Objectives:

Renal cell carcinoma (RCC) is known for its metastatic spread, yet only 7% of patients have cutaneous metastases. For 10-20% of these patients, the cutaneous symptoms manifest as the first presentation of the disease, typically presenting in patients with a history of RCC. Head and neck involvement occurs in only 6%, and involvement of the exterior nasal area is extremely rare, with few documented cases.

Most metastatic lesions arising on the face typically appear as a squamous cell carcinoma, as a result of their underlying origin. However, few cases of cutaneous RCC metastases on the body have presented atypically as vascular lesions suspected to be benign vascular tumours, such as a pyogenic granuloma.

We present two rare cases of nasal lesions mimicking pyogenic granuloma resulted in a new diagnosis of RCC.

Materials & Methods:

Results:

A 56 year-old gentleman presented with a 3-month history of a pedunculated lesion on his nasal bridge, increasing in size and bleeding easily with minimal trauma. Dermoscopy revealed vascular lacunes and white lines, in keeping with a pyogenic granuloma. The lesion was removed with shave biopsy. However, histopathological examination suggested clear cell neoplasm, with positivity for CD10, Vimentin and RCC antigen. A diagnosis of metastatic RCC was made. The second case of a 79 year-old gentleman, with previous oesophageal carcinoma, presented with a vascular lesion affecting the nose, on his right nasal bridge. This was initially diagnosed as a benign haemangioma and remained quiescent for some time. Six months after initial presentation, he was referred again as the lesion increased rapidly in size with an episode of bleeding. Clinically, this had the appearance of pyogenic granuloma, but oesophageal metastasis was considered. The lesion was removed with shave biopsy and histology showed a nodule with trabeculae of cells with clear cytoplasm, cellular atypia and dark ovoid nuclei in a vascular stroma. This was in keeping with metastatic clear cell RCC. This gentleman had recent CT imaging showing bilateral renal cysts. However, both patients were otherwise asymptomatic, without haematuria on urinalysis and with no history of RCC. Both patients were subsequently referred to the Urology multidisciplinary meeting for further management.

Conclusion:

In summary, cutaneous RCC metastasis rarely presents as a vascular nodule on the nose that can mimic pyogenic granuloma. Recognising this rare manifestation is important for appropriate management planning and could serve as the initial indication for diagnosing metastatic renal cell carcinoma. Our cases provide valuable insight and awareness for clinicians to consider this presentation as a differential in relevant cases.

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Abstract N°: 6370

A Case Report of Blastic Plasmacytoid Dendritic Cell Neoplasm

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CIntroduction & Objectives:

Blastic plasmacytoid dendritic cell neoplasm (BPCDN) is a very rare, aggressive malignancy arising from plasmacytoid dendritic cells. BPDCN was formerly called CD4+/CD56+ hematodermic neoplasm and blastic natural-killer cell lymphoma. According to the 2008 WHO classification of myeloid neoplasms and leukemia, it is classified under the title of acute myeloid leukemia and related neoplasms. 2016 WHO classification, it is examined in its own category under the title of acute myeloproliferative neoplasm.

Although BPDCN generally involves bone marrow and asymptomatic skin, lymph node and visceral organ involvement may also be observed. The most commonly involved organ other than the bone marrow is the skin, and skin involvement often occurs before the bone marrow. BPDCN is more common in men between the ages of 53-68. It has been stated that it shows a bimodal incidence pattern, as a result of recent studies showing that it is encountered more frequently than normal in people under the age of 20 and over the age of 60.

Materials & Methods:

A 30-year-old male patient was admitted to us with numerous, asymptomatic, dark erythematous infiltrated papules and nodules on the front of the thorax, back and proximal arms, which had been present for 1 month. A biopsy was performed from the patient with the preliminary diagnosis of mastocytosis, mycosis fungoides, cutaneous metastasis and Langerhans cell histiocytosis.

Results: ** The histopathology result was evaluated as compatible with “blastic plasmacytoid dendritic cell neoplasm”. The patient was referred to hematology, whole body positron emission tomography revealed multiple lymphadenopathy suggestive of malignant infiltration of skin disease, and a bone marrow biopsy was performed.

Conclusion:

We found this case worth reporting because BPDCN is very rare and histopathology is very important in the diagnosis of hematological malignancies.

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Abstract N°: 6676

Diabetic dermopathy as a cutaneous manifestation marker of microangiopathic complications: a cross-sectional study

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Introduction & Objectives

Diabetic dermopathy (DD) is one of the most common cutaneous manifestations in patients with diabetes mellitus.

Main objective: To demonstrate the association between DD and microangiopathic complications of the disease (considering retinopathy, nephropathy, and sensory neuropathy) in diabetic patients (type I and type II) from the Hospital Privado Universitario and Raúl Ferreyra Hospital in Córdoba.

Specific: Establish the prevalence of patients with DD and concomitant retinopathy, sensory neuropathy, and nephropathy. Determine the prevalence of these complications in diabetic patients without DD. Compare the results obtained between patients with and without DD. Establish the prevalence of patients with DD without microangiopathic complications. Establish the prevalence of patients without DD and without microangiopathic complications. Determine if DD affects type II diabetes patients more commonly than type I diabetes patients. Determine if there is a relationship between the value of glycosylated hemoglobin and the frequency of DD. Determine the relationship between the prevalence of DD and the number of chronic complications of the disease in each patient. Determine sensitivity, specificity, positive predictive value, and negative predictive value of DD in relation to the three microangiopathic complications.

Materials & Methods

A prospective, cross-sectional, observational, and correlational study was conducted between May 2014 and February 2016, including 163 patients, of whom 57 belonged to the “cases” group (type I/II diabetics with diabetic dermopathy) and 106 patients were included in the “control” group (type I/II diabetics without diabetic dermopathy), evaluated by the Dermatology and Diabetology departments of the Hospital Privado Universitario. Patients referred from the Raúl Ferreyra Hospital in Córdoba were also included. Each patient underwent: fundus examination (to determine diabetic retinopathy), isolated urine sample, where the presence or absence of diabetic nephropathy was determined through microalbuminuria, and a physical examination, always conducted by the same observer (a specialist in internal medicine), to determine the prevalence of sensory diabetic neuropathy. Statistical data on the prevalence of diabetic retinopathy, neuropathy, and nephropathy were collected for each patient. In a second step, the percentages of these complications in patients exposed to the risk factor DD were compared with those in patients not exposed to it.

Results

In our series, DD was observed to be more frequent in male patients over 50 years of age. The mean disease duration was slightly higher in patients with DD compared to control patients. The association between the presence of DD and the three microangiopathic complications of diabetes: retinopathy, sensory neuropathy, and nephropathy, was significant. It was also demonstrated that the prevalence of DD increases not only with each of the three microangiopathic complications but also in relation to the number of complications in each patient.

Conclusion

DD is a good predictor of each of the three microangiopathic complications of diabetes, as well as the number of patient complications. It is interesting to note that with such an accessible tool as the physical examination of diabetic patients in the office, the systemic complications of this pathology could be presumed.

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Abstract N°: 6743

a case of relapsing polycondritis

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Introduction & Objectives: Relapsing polychondritis (RP) is a systemic, inflammatory, immune-mediated disease characterized by recurrent episodes of inflammation affecting cartilage and proteoglycan-rich tissues, primarily in the upper and lower respiratory tract, ears, and eyes. It may also involve the skin, joints, kidneys, cardiovascular system, and central nervous system. The etiology of TP is currently unknown, with various triggering factors suspected to contribute alongside genetic predisposition.

Materials & Methods: A 49-year-old woman presented to our clinic in January 2024 with a rash in the left auricular region. She complained of severe pain radiating to the nose, mouth, and neck. Further inquiry into her medical history revealed similar symptoms occurring in her right auricle in July 2023. Additionally, she had been experiencing recurrent episodes of widespread erythematous plaques throughout her body over the past year, which spontaneously resolved on the same day without intervention. Erythema, edema, and local temperature elevation were observed in the left auricle, along with mild edema in the nose. Laboratory tests revealed no abnormalities except for mildly elevated sedimentation and CRP levels. No systemic involvement was observed.

Results: Based on McAdam's criteria, a diagnosis of TP was made. Treatment was initiated with methylprednisolone 40 mg, and significant improvement was noted on the second day of treatment.

Conclusion: Unilateral or bilateral auricular chondritis is the predominant initial presentation in the majority of cases (90%) of recurrent polychondritis. A notable characteristic of auricular chondritis is the preservation of the earlobe. Cutaneous manifestations are present in approximately 50% of cases. The prevalent cutaneous manifestations comprise vasculitis and erythema nodosum-like lesions, whereas other observed findings involve wandering superficial thrombophlebitis, oral aphthae, urticaria/angioedema, and livedo reticularis. The diagnostic delay for this condition, frequently misinterpreted as an infection, ranges from 1.9 to 2.9 years. Clinical features and prognosis vary significantly among patients, depending on the degree of organ damage. Prognosis is largely determined by the extent of organ involvement. Treatment typically involves NSAIDs for cases with limited involvement, while systemic corticosteroids, immunosuppressive drugs, and biological agents are reserved for severe cases.

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Abstract N°: 6832

Tuberous xanthomas revealing homozygous familial hypercholesterolemia

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Introduction :

Tuberous xanthomas are nodular, firm, painless lesions. Most often linked to a disorder of lipoprotein metabolism. We report the case of a child in whom multiple tuberous xanthomas revealed familial hypercholesterolemia.

OBSERVATION : OBSERVATION: BN child aged 8 years, from a first degree consanguineous marriage, with a family history of hypercholesterolemia, who consults for papulo-nodular and tumorous lesions, symmetrical, yellowish in color, painless and non-inflammatory, at the elbows, knees, buttocks and backs of the feet evolving for 3 years gradually increasing in size and taking on a tuberous appearance. Dermoscopic examination showed a yellow area without structure. The skin biopsy revealed a nodular lesion made of foamy histiocytic cells containing lipid droplets confirming the diagnosis of xanthoma. The lipid profile showed very high levels of total cholesterol (9.32 g/L), LDL cholesterol (8.80 g/L), HDL cholesterol was reduced (0.34 g/L), triglycerides were within the standards. There were no cardiac or ophthalmological abnormalities. In this child, the very high levels of LDL cholesterol, the presence of cutaneous xanthomas since the age of 5 and the absence of other abnormalities, suggested a homozygous form of familial hypercholesterolemia of type II according to the Fredrickson classification, The patient was managed by hygienic and dietary measures associated with treatment with atorvastatin 10 mg/day. We noted a clear improvement in the lipid profile after 1 and a half months of treatment: Total cholesterol at 06 g/l and LDL at 06 g/l. 5.7 g/l. A screening lipid profile in other members of the family was proposed.

CONCLUSION:Xanthomatoses linked to homozygous familial hypercholesterolemia are very rare. Morbidity and mortality are linked to atherosclerosis. Early and targeted detection allows adequate care and a reduction in early cardiovascular events

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Abstract N°: 6860

Extensive Skin Necrosis In Cutaneous Calciphylaxis : A Rare Case

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Introduction & Objectives: Calciphylaxis, while rare, is a serious condition characterized by ischemia and necrosis. Histologically, it shows calcification of arterioles in the dermis and subcutaneous adipose tissue. Calciphylaxis often affects patients with end-stage renal disease (ESRD) undergoing hemodialysis.

Materials & Methods: A 41-year-old woman with Grade 5 chronic kidney disease (CKD) and type II diabetes mellitus was referred from internal medicine with complaints of black spots on both thighs, groin, middle finger of the left hand, and three toes on the right foot. These spots had developed over three months, initially presenting as dark red bruises before turning black. The patient reported the black spots were hard and very painful. She had been undergoing routine hemodialysis for the past five months. Vital signs were within normal limits. Dermatological examination revealed necrosis in the bilateral thighs and left toes (digits III-V). The right tibialis region showed ulcers with pus, and gangrene was found in the left middle finger (digit III). Histopathological examination confirmed a diagnosis of cutaneous calciphylaxis, with findings of thrombosis, necrosis, and microcalcifications in blood vessels from the superficial to the deep dermis.

Results: Calciphylaxis is a skin complication and a marker of poor prognosis for patients with metabolic disorders, including CKD. While its specific pathogenesis remains unclear, it differs from atherosclerosis, with circular calcium apatite deposits in small and medium-sized arteries. Several proinflammatory cytokines are implicated in its development. Diagnosing calciphylaxis can be challenging due to its atypical clinical presentation of skin necrosis. Skin biopsy and histopathological examination are essential for diagnosis, revealing calcification of small vessels (<100 µm) in the deep dermis and subcutaneous tissue, fibrin thrombi, and sometimes epidermal and dermal necrosis. No single, universally accepted treatment for calciphylaxis exists. Management is multimodal and interdisciplinary, focusing on mitigating risk factors, wound care, optimizing hemodialysis, systemic medical therapy, and pain management (analgesia). The prognosis for calciphylaxis is poor, with a 45-80% mortality rate within one year of onset.

Conclusion: Calciphylaxis is a severe skin complication of metabolic disorders, including CKD. Prompt diagnosis is crucial and relies on history, clinical findings, and histopathological confirmation.

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Abstract N°: 6888

Late-onset Erythropoietic Protoporphyria: two atypical cases arising during with Venetoclax therapy for chronic lymphocytic leukaemia

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Introduction & Objectives:

Erythropoietic protoporphyria (EPP) is a rare cutaneous porphyria which causes the accumulation of excess erythrocyte protoporphyrins due to aberrant ferrochelatase function. EPP, unlike the bullous porphyrias, usually presents with acute pain within minutes following visible light exposure. This study aims to present two atypical cases of EPP, both arising during Venetoclax therapy for chronic lymphocytic leukaemia (CLL), contributing to the sparse literature on acquired EPP associated with CLL and its treatment.

Materials & Methods:

We report on two male patients under Venetoclax treatment for CLL, who exhibited unusual painless bullous eruptions. A detailed clinical and biochemical evaluation was conducted, including porphyrin screens. Secondary causes of red cell porphyrin abnormalities such as lead poisoning and anaemia were considered and excluded.

Results:

Case 1

A 61-year-old gentleman with a background of CLL, Crohn's disease and previous iron deficiency anaemia presented with an acute onset photosensitive bullous eruption amid ongoing treatment with Venetoclax for CLL. He had previously also been treated with Rituximab, although this had been discontinued due to adverse cardiac events. Despite discontinuing naproxen, a cause of pseudo-porphyria, he continued to develop blisters. Unexpectedly, a porphyrin screen revealed marginally elevated total red cell porphyrin levels of 3.2 umol/L (normal range 0.4-1.7 umol/L).

Case 2

A 72-year-old gentleman receiving Venetoclax for CLL presented with an acute bullous eruption localised to his hands which started during the winter. He had a background of cirrhosis secondary to autoimmune hepatic disease. A porphyrin screen demonstrated elevated red cell porphyrin (7 umol/L) and significantly elevated plasma porphyrins.

Conclusion:

In the cases presented, neither had a history of acute onset painful photosensitive reactions as is typically observed in EPP. Nevertheless, biochemical analysis in both patients is suggestive of either an incidental carrier status or that of an acquired EPP. Whilst genetic analysis results are pending, we postulate that this may be a drug-induced phenomenon. Venetoclax inhibits BCL-2, a group of proteins involved in regulating apoptosis. Located on chromosome 18q21.33, the BCL-2 gene is in close proximity to the human ferrochelatase gene (FECH). This close location may cause transcriptional interference, altering FECH function with resultant excess red cell porphyrins.

Although a small number of cases of acquired EPP have been reported with several haematological disorders, to our knowledge there are no reported cases with either CLL or Venetoclax therapy. Whilst rare, this complex interplay presents diagnostic and therapeutic challenges requiring careful consideration of the underlying CLL, liver health and the impact of CLL treatments on porphyrin metabolism.

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Abstract N°: 6963

The Cutaneous Window of Internal Health: A Case Report On Eruptive Xanthomas

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Introduction & Objectives:

Eruptive Xanthomas are rare. In fact, a study by Munoz et al in 2023 reported an incidence only 18 in 100,000 to have it. These eruptive xanthomas present as multiple erythematous to whitish to yellowish dome-shaped papules commonly affecting the extensors and buttocks, which result from severe elevations of triglycerides that permeate into the subcutaneous tissues. Due to the risk of developing atherosclerosis and pancreatitis, it is vital that clinicians are able to detect and diagnose these case promptly.

Materials & Methods:

A 46 year-old Filipino male sought consult for a 7-month history of multiple well- defined asymptomatic whitish to yellowish dome-shaped papules on the nape, trunk, lower back and bilateral extremities. He was initially treated by a private dermatologist as a case of Molluscum Contagiosum using topical 5% potassium hydroxide with no relief of symptoms, prompting current consult. On physical examination, he was seen with multiple 2 to 4 millimeter, well-defined yellowish to whitish papules and nodules, with some forming into clusters of 3 to 4 on the trunk, abdomen, arms and legs. He was Hypertensive, Stage II with a blood pressure of 140/90 and with a BMI of 29.8 (Obese I). Laboratories were also requested.

Results:

The results of his laboratories revealed a notable 16.8 fold elevation of his triglyceride levels. Furthermore, his fasting blood sugar, liver enzymes, low density lipoprotein and very low density lipoprotein are all also elevated. He underwent lifestyle changes and prompt therapy with Tenegliptin 40mg/tab OD, Atorvastatin 10mg/tab OD, Carnitine Orotate + Hepatic Extract Antitoxic Fraction + Adenine HCl + Multivitamins tablet TID, and Amlodipine 10mg/tab OD. This treatment caused a decrease in the number of lesions, increase in hyperpigmentation and decrease in the yellowish discoloration, especially evident on dermoscopy.

Conclusion:

Although uncommon this case is an example of how asymptomatic cutaneous eruptions may be signs of metabolic diseases. This shows the importance of dermatologists in being able to promptly identify, refer and treat these patients.

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Abstract N°: 7063

Eosinophilic fasciitis: a two case report

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Introduction & Objectives:

Eosinophilic fasciitis (EF) is a rare disease characterized by diffuse fasciitis accompanied by eosinophilia, leading to painful swelling and progressive skin thickening, primarily affecting the trunk and limbs. The diagnostic criteria and severity assessment are outlined in Tables 1 and 2, respectively. Key diagnostic features include the *peau d'orange* appearance of affected skin, the "groove sign" - a linear depression along vein courses, eosinophilia, elevated erythrocyte sedimentation rate (ESR), and aldolase levels. Corticosteroid therapy constitutes the primary treatment modality. Through the presentation of two clinical cases, we aim to elucidate key diagnostic considerations and to discuss therapeutic challenges in non-responsive cases.

Table 1. Diagnostic criteria for EF.

Major Criteria	Minor Criteria
Symmetrical plate-like sclerotic lesions present on the four limbs.	1. The histology of a skin biopsy that incorporated the fascia shows fibrosis of the tissue, with thickening of the fascia and cellular infiltration of eosinophils and monocytes.
	2. Thickening of the fascia is seen using imaging tests, such as magnetic resonance imaging (MRI)

Table 2. Severity classification of EF.

Joint Involvement	Points
Joint contracture (upper limbs)	1 point
Joint contracture (lower limbs)	1 point
Limited movement (upper limbs)	1 point
Limited movement (lower limbs)	1 point
Expansion and worsening of skin rash (progression of symptoms)	1 point
A total of 2 or more points is classified as severe.	

Materials & Methods:

Case 1: A 45-year-old male presented with a 7-month history of pain, redness, and swelling in his right lower limb, subsequently spreading symmetrically to involve his legs, thighs, arms, forearms, and trunk. At that time he also

complained of joint pain in hands and elbows.

Case 2: A 62-year-old female reported skin thickening of the lower limbs persisting for 15 months, later extending to the upper limbs and trunk within 2 months. She had been prescribed methotrexate (15 mg weekly) and prednisone (20 mg daily) for a year without improvement, initially diagnosed as scleroderma.

Both patients exhibited eosinophilia (>1000), *peau d'orange* skin appearance, sparing of the face and hands, and in the case of patient 2, the presence of the groove sign and reduced limb mobility. Notably, there were no symptoms of Raynaud's phenomenon or systemic involvement. Both patients underwent fascia biopsy and dermatological ultrasound.

Results:

Patient 1 received subcutaneous methotrexate (25 mg/week) and prednisone (60 mg daily), while patient 2 was prescribed mycophenolate mofetil (1500 mg daily) with an increased prednisone dose (30 mg daily). Phototherapy was not feasible for either patient. After a period on the new regimen, both noted a modest improvement in skin thickness and induration. Given the partial response, both are scheduled for rituximab therapy.

Conclusion:

These cases underscore the diagnostic challenges of eosinophilic fasciitis, often mistaken for scleroderma, delaying appropriate management. Rapid, generalized progression sparing the face and hands, along with the absence of Raynaud's phenomenon, aids in the differential diagnosis. Therapeutically, immunomodulators show promise, with immunobiologicals emerging as viable alternatives in refractory cases. Early recognition and tailored treatment strategies are imperative for optimal patient outcomes.

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Abstract N°: 7065

Idiopathic Hypereosinophilic Syndrome With Cutaneous, Renal And Cardiac Involvement: A Case Report

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Introduction & Objectives:

Hypereosinophilic Syndrome (HES) is a rare disorder characterized by persistent eosinophilia and multi-organ involvement, often presenting with dermatological manifestations.

Herein, we present a case of idiopathic HES in a middle-aged female with cutaneous, renal, and cardiac involvement.

Case description:

A 51-years-old female with history of hypertension, asthma, and chronic smoking presented to our department with a diffuse skin rash persisting for four months. The lesions were pruritic erythematous papules and nodules. There was no indication of a precipitating event, such as an insect bite, or any medications that commonly demonstrate a propensity to cause drug rash with eosinophilia and systemic symptoms syndrome (DRESS).

Initial laboratory studies revealed a prominent eosinophilia of 20,220/l (normal, <500/l). The IgE was 594 IU/ml (Normal range). An extensive diagnostic workup was performed including immunological, stool and imaging, showed no abnormalities except for proteinuria and renal insufficiency. Echocardiography showed considerably decreased left ventricular function with apical obliteration, with no thrombi or restrictive cardiomyopathy detected. Skin and bone marrow biopsy confirmed eosinophilic infiltration without evidence of malignancy. Despite negative findings, the patient was diagnosed with idiopathic hypereosinophilic syndrome with cutaneous, renal and cardiac involvement. High-dose of corticosteroid therapy (1mg/kg/day) lead to clinical, biological and imagining improvement.

Conclusion:

The presented case offers a comprehensive exploration of the diagnostic and therapeutic challenges inherent in the management of idiopathic HES. The intricate interplay between the patient's medical history, clinical presentation, and multi-organ involvement underscores the systemic nature of this rare syndrome. Recognition of HES demands a holistic diagnostic approach, incorporating clinical, laboratory, and histopathological assessments, while therapeutic interventions should be tailored to address both acute symptoms and long-term sequelae. That's why a multidisciplinary collaboration is crucial for optimizing patient care and outcomes in such complex presentations.

Continued research efforts are imperative to elucidate the underlying pathophysiological mechanisms of HES and develop targeted therapies aimed at improving patient outcomes and quality of life.

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Abstract N°: 7150

Acquired Ichthyosis in an Older Patient with Poorly Controlled Diabetes

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Introduction & Objectives:

Acquired ichthyosis (AI) is a rare dermatological disorder characterized by dry, rough, and scaly skin. The condition is associated with various underlying causes, including systemic diseases such as diabetes mellitus (DM). Understanding the underlying mechanisms and potential triggers for AI is crucial for diagnosis and effective management. While AI associated with DM has been reported in younger patients, this case presents AI development in an older patient with poorly controlled DM.

Materials & Methods:

A 67-year-old Hispanic male with uncontrolled DM was evaluated for a five-week history of generalized dryness and itchiness, initially localized to his back and subsequently spreading throughout his body. The patient had a recent hospitalization for diabetic ketoacidosis and had a history of heavy alcohol consumption and significant smoking (50 pack-years). Dermatological examination revealed dry polygonal scales with interscale erythema over multiple body areas. A biopsy was performed of the affected skin on the left shoulder, which confirmed the diagnosis of AI, showing typical epidermal changes consistent with ichthyosis. Laboratory investigations were conducted, revealing elevated fasting plasma glucose (411 mg/dL), dyslipidemia, hyponatremia, hyperkalemia, and chronic kidney disease stage IIIb. Cancer screening, including colonoscopy and prostate-specific antigen, was reported to be up to date.

Results:

Treatment for our patient included a regimen aimed at both managing DM and addressing the symptoms of AI. He was prescribed moisturizers, antihistamines, antifungal shampoo, topical corticosteroids, and tacrolimus for the skin symptoms, alongside an optimized DM management plan. His symptoms improved considerably on this multidisciplinary regimen.

Conclusion:

DM is associated with various cutaneous manifestations, including AI, which typically manifests as generalized scaling and dryness. Unlike congenital forms of ichthyosis, AI often arises later in life secondary to underlying systemic conditions or external factors. AI has been linked with malignancy, autoimmune diseases, chronic infections, and certain medications. Diagnosis is clinical and may be supported by biopsy. Once a diagnosis is made, a systemic workup should be conducted to rule out cancer and other causes of AI. Poorly controlled DM was a significant contributing factor to AI development in our patient, emphasizing the importance of comprehensive assessment in such cases. Early recognition and understanding of this association can help optimize therapeutic approaches and reduce associated morbidity. Further research into underlying mechanisms, whether disease course correlates with hemoglobin A1c, and therapeutic optimization is warranted.

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Abstract N°: 7210

En coup de sabre with Parry-romberg syndrome and Hemimasticatory Spasm

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Introduction & Objectives:

En coup de Sabre is a unique presentation of linear morphea, typically affecting the frontoparietal scalp and/or the paramedian forehead. Parry-Romberg syndrome, a rare degenerative disorder, causes progressive hemifacial atrophy of the skin and subcutaneous tissue. Hemimasticatory spasm is a rare movement disorder characterized by unilateral, involuntary, and paroxysmal contractions of the muscles of mastication.

Materials & Methods:

A 20-year-old man presented to the dermatology department with a 3-year history of linear patches of alopecia over the left parietal scalp and left cheek with a depressed and shiny appearance. Six months later, underlying tissue atrophy was evident over the left cheek, gradually progressing causing facial asymmetry, soon followed by involuntary muscle spasms and firm swelling of muscle tissue. Spasms were associated with chewing and clenching of teeth.

On examination, linear patch of cicatricial alopecia was seen in the left paramedian parietal scalp. An atrophic, hyperpigmented patch was also noted over the left cheek with alopecia, underlying tissue atrophy and adjacent masseter hypertrophy, which was confirmed on radiological investigations. Oral examination revealed left-sided lower lip and tongue atrophy.

Results and Conclusion:

Many authors believe ECDS and PRS to be variants of morphea, sharing a common underlying pathogenesis. HMS is a rare disorder, thought to be due to impaired inhibition of masseter muscle contraction as a result of focal demyelination and ectopic excitation of the trigeminal nerve. In our case, this was possibly caused by nerve compression secondary to atrophic changes in an already confined space.

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Abstract N°: 7238

Nailfold Capillaroscopy to assess the microcirculation in patients of diabetes mellitus, an observational study

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Title - - NAILFOLD CAPILLAROSCOPY TO ASSESS THE MICROCIRCULATION IN PATIENTS OF DIABETES MELLITUS, AN OBSERVATIONAL STUDY

Introduction

- Type 2 diabetes mellitus (T2DM) is a common metabolic disease characterized by chronic hyperglycaemia. It affects the microvasculature causing complications such as retinopathy, nephropathy, and neuropathy. Nail fold capillaroscopy (NFC) is an in vivo study of cutaneous microvascular circulation.

Objectives:

■ Primary Objectives

➢ To study the Nailfold Capillaroscopic changes according to the given parameters in

patients of DM and report any new findings if found in the patients.

■ Secondary Objectives

➢ To assess association between diabetic retinopathy and nailfold capillaroscopic

changes

➢ To assess association between Hba1c levels and NFC changes

➢ To assess association between duration of diabetes and of NFC changes

➢ To evaluate MDAD criteria in patients of Type 2 Diabetes Mellitus

Materials & Methods:

After approval from ethics committee, a single-centre hospital-based cross-sectional study was conducted at outpatient clinic of Department of Dermatology, at a Tertiary care Hospital. 100 T2DM cases and age matched healthy controls were enrolled in the study between the ages of 20-60 years. Baseline Fasting blood sugar (FBS), Hba1c and fundoscopic examination for retinopathy was performed. T2DM patients were further divided in two groups: Those without retinopathy (Group A, n=61) and with retinopathy (Group B, n=39). NFC was performed, qualitative and quantitative parameters along with MDAD (morphology, diameter, architecture and density) criteria were analyzed.

Statistical analysis – All data was entered in excel sheets and statistical analysis was carried out by using GraphPad INSTAT software version 3.06, 32 bit for windows. Categorical and nominal data were expressed in percentage. Categorical data were analyzed by using a chi-square test with the significance threshold of p-value set at <0.05.

Results: Dilated capillaries, Neoangiogenesis, Meandering, Tortuous capillaries, Avascular zones, Visibility of subpapillary venous plexus and receding capillaries were significantly higher in T2DM individuals. Mean capillary density of T2DM individuals was 6.70±0.76 which was significantly lower(p=<0.001) than healthy controls which was 7.01±0.77. Patients with retinopathy had a significantly higher frequency of receding capillaries when compared to individuals without retinopathy.

Conclusion: The NFC changes correlated with microvascular alterations in T2DM patients, possibly helping in non-invasive identification of T2DM individuals. Capillaroscopic alterations were not significant in differentiating between retinopathy and non retinopathy group. The same stands true for effect of duration and glycaemic load in diabetic patients.

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Abstract N°: 7247

Dermatological associations of rheumatoid arthritis: A Case Series

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Introduction & Objectives:

Rheumatoid arthritis (RA) is a chronic progressive disorder characterized by symmetric inflammatory arthritis in association with systemic symptoms. Although considered a "joint disease," RA is associated with involvement of diverse organ systems, including the skin.

Of note are unique cutaneous associations that the dermatologist may encounter, namely pyoderma gangrenosum, psoriasis and bullous pemphigoid, that are highlighted here.

Materials & Methods:

Case 1:

A 65 year old female presented with painful, itchy lesions since two years, which rapidly evolved to form multiple well defined tender ulcers of varying stages over bilateral lower extremities buttocks and abdomen, confirmed as pyoderma gangrenosum.

Case 2:

A 48 year old female presented with itchy, fissured, indurated plaques surmounted with adherent scales over palms and soles since 2 months, diagnosed as palmoplantar psoriasis.

Case 3:

A 54 year old female presented with multiple tense fluid-filled bullae over the body since 1 month, with interspersed erosions and haemorrhagic adherent, foul-smelling crusts, diagnosed as bullous pemphigoid.

The patients were investigated for RA factor and anti-CCP, as a confirmation for rheumatoid arthritis. Dermatological dermatoses were confirmed on histopathology.

Pyoderma gangrenosum	**Psoriasis**	**Bullous pemphigoid**
65Y/F	48Y/F	54Y/F
RA since 4 years	RA since 22 years	RA since 25 years
RA: 431 Anti-CCP: 78.5	RA: 667 Anti- CCP: 64.2	RA:386 Anti-CCP: 57.4

Results:

There appears to be a link in the occurence of autoimmune diseases as they are driven by abnormalities of the innate immune system.

Approximately 50% of cases of PG are associated with a systemic disease, the rest being idiopathic. The most commonly associated diseases are IBD, rheumatoid arthritis and haematological malignancies. In PG lesions, pattern recognition receptors and TLR overexpression may be implicated, like RA.

The incidence of psoriasis in a series of rheumatoid arthritics was found to be between 3 and 4 per cent. Psoriasis is linked with gut dysbiosis and HLA alleles affect disease susceptibility and severity, similar to RA.

In a review of 94 cases of BP, 12 per cent had RA as defined by the diagnostic criteria of the American Rheumatism Association. Unmasking and cross reaction of antigens appears to be the pathophysiology of concurrence in BP and RA.

Conclusion:

Timely and thorough investigations for dermatological disorders should be carried out for patients with rheumatoid arthritis.

This vigilance may lead to the potential understanding of the pathogenesis of these disease processes and could eventually pave the way for more targeted diagnostic and therapeutic approaches.

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Abstract N°: 7268

Mucocutaneous manifestations as a clue for reactive and disabling arthritis

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Introduction & Objectives:

Reactive arthritis is defined as a form of arthritis associated with a coexisting or recent gastrointestinal or genitourinary infection. It is characterized by asymmetric oligoarthritis, enthesitis, dactylitis, and inflammatory back pain. Extramusculoskeletal features include eye involvement, genitourinary symptoms, oral ulcers, and skin manifestations. The diagnosis is mainly clinical, and treatment is symptomatic with non-steroidal anti-inflammatory drugs and/or oral corticosteroids. Typically, reactive arthritis solves spontaneously in 6 to 12 months.

Materials & Methods:

Case report.

Results:

A 50-year-old male patient with history of intravenous drug use, treated HCV infection, and alcohol abuse disorder was admitted to the emergency department due to a 3-month history of heel pain with inability to walk. Aodema and a figurate scaling of the glans penis, a right plantar hyperkeratotic cutaneous lesion and nail abnormalities were observed. The patient also mentioned a three-day episode of red eye with spontaneous improvement. Four weeks prior to the beginning of symptoms he had a gastroenteritis infection that resolved with supportive treatment. The plantar skin findings were previously treated as dermatophyte with superimposed bacterial infections with oral antifungal, amoxicillin-clavulanate, and ciprofloxacin to cover Pseudomonas aeruginosa, but without benefit. ultrasound imaging showed alterations compatible with plantar fasciitis. A clinical picture of keratoderma blennorrhagica, circinate balanitis and nail distrophy with onicomadesis (2nd left and 3rd right fingers) was assumed. The patient was treated with a topical combination of corticosteroid and salicylic acid for the plantar lesion and topical tacrolimus for the balanitis, with partial improvement. No stool cultures were obtained as the gastrointestinal symptoms have subsided completely. First-pass urine sample was negative for Chlamydia trachomatis and gonorrhoea. Skin biopsy showed a cutaneous inflammatory process dominated by neutrophilic infiltrate with no granuloma, favouring the suggested keratoderma blenorrhagica and excluding verrucous tuberculosis, and psoriasis. Microbiology was also negative for Mycobacterium tuberculosis. A diagnosis of arthrits in the third and fourth distal interphalangeal joint of the right hand, and right calcaneal enthesopathy was made by rheumatology and the patient treated firstly with naproxen (500mg bid) and then with oral prednisolone (15mg id).

Conclusion:

This case highlights the importance of skin inspection facing a case of arthritis, for the diagnosis of a reactive disorder. In our case, keratoderma blenorragica, circinate balanitis and nail dystrophy were the cardinal features.

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Abstract N°: 7281

Lupus Panniculitis: About 2 Cases.

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Introduction & Objectives:

Lupus panniculitis, also known as lupus erythematosus profundus, is a rare variant of cutaneous lupus, which can occur in the setting of systemic lupus erythematosus or independently. Clinically, it presents as firm, tender subcutaneous nodules evolving into permanent depressed scars. We report here two cases of lupus panniculitis, one diagnosed in the early stage and the other presenting after years of evolution at the scar stage.

Observations:

Case 1: We report the case of a 37-year-old female patient, followed for discoid lupus since 2018, with strongly positive antinuclear antibodies (ANA), on hydroxychloroquine and corticosteroids, who was admitted to the dermatology department with **firm, tender, erythematous subcutaneous nodules on both cheeks, which have been evolving for 2 months. The diagnosis, suspected in the context of lupus, was confirmed after demonstrating lupus band on direct immunofluorescence (DIF) of a skin biopsy. Methotrexate was introduced in this patient 4 weeks ago.**

Case 2: This concerns a 34-year-old patient, with no particular medical history, presenting with atrophic depressed scars. She reports a history of firm subcutaneous nodular lesions with inflammatory signs evolving for 7 years with remission periods, affecting the bilateral jugal, frontal, right deltoid, and left crural regions. Initially diagnosed in maxillofacial surgery as angiomas with excision, histology post-operatively favored lipoma. The diagnosis, more challenging in this second case, was suspected based on clinical appearance, supported by positive immunological tests (positive ANA and anti-native DNA), and confirmed by correlation with histology compatible with lobular panniculitis. Our patient was treated with hydroxychloroquine without clinical improvement, the patient underwent facial lesion lipofilling with satisfactory results.

Discussion:

Lupus panniculitis is an uncommon variant of lupus erythematosus. It presents as firm, tender, persistent nodules on the face, arms, shoulders, chest, and buttocks with atrophic scarring evolution and sometimes ulceration. In the absence of other signs of lupus erythematosus, diagnosis is challenging as histology may be suggestive but nonspecific, and lupus band testing may be negative without systemic involvement. In addition to diagnostic challenges, this condition poses therapeutic difficulties due to the lack of specific indications and treatment protocols.

Conclusion:

Lupus panniculitis is a rare, poorly understood, and likely underdiagnosed entity. Diagnosis, while easily considered in any lupus patient presenting with subcutaneous panniculitis-like lesions, remains challenging outside the context of chronic or systemic lupus. Regardless of the case, it represents a therapeutic challenge, especially in the presence of atrophic scar lesions.

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Abstract N°: 7287

Diffuse form of systemic sarcoidosis

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Introduction : Sarcoidosis, a systemic granulomatous disease of unknown etiology, is characterized by its great clinical polymorphism and the diversity of its modes of presentation.

We report the observation of a patient with skin lesions;Pulmonary, cardiac and endonasal involvement during systemic sarcoidosis.

observation: :a 39-year-old patient with no notable pathological history, admitted for exploration of a subungual nodular lesion on the big toe of the right foot, an erythematous plaque on the anterior aspect of the right leg, anosmia, nasal obstruction and dyspnea, all entities objectified by clinical examination.

On biology, we noted the presence of a biological inflammatory syndrome with VS 40 (H1)l, polyclonal hypergammaglobulinemia at 24 g/l.An increase in serum angiotensin-converting enzyme activity.Hepatic, renal and phosphocalcic assessments were normal. Chest radiography showed: symmetrical hilar and mediastinal lymphadenopathy, often bulky but not compressive.

The lung parenchyma is normal.Respiratory function tests were normal. The chest CT scan revealed “an interstitial syndrome with predominantly hilar mediastinal lymphadenopathy”. On bronchoscopy there were multiple nodule-type lesions in the nasal cavity as well as in the cavum with histologically “granulomatous inflammation of the nasal mucosa and skin lesions. » Furthermore, the cardiac examination noted a BAV type conduction disorder. Nasal and skin biopsy confirmed granulomatosis.

The diagnosis of systemic sarcoidosis was made. The patient was treated with oral corticosteroid therapy at a dose of 1 mg/kg/day and methrexate . The evolution was favorable with disappearance of nasal obstruction and dyspnea and improvement of the skin lesions.

Conclusion:

The particularity of our case lies in the quadrifocal involvement of the skin, lungs, heart and especially the endonasal region, which is usually rare in sarcoidosis (1-6%). Respiratory problems and nocturnal snoring in sarcoidosis should prompt a systematic ORL examination.

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Abstract N°: 7356

ANCA Negative EGPA with Pulmonary, Cutaneous, Digestive and Neurogical manifestations: A Case Report

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Introduction & Objectives:

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly Churg-Strauss syndrome, is a rare immune-mediated vasculitis associated with anti-neutrophil cytoplasmic antibodies (ANCAs). It characterized by the presence of asthma, hyper-eosinophilia and necrotizing vasculitis with extravascular eosinophilic granulomas.

Herein we report a case of EGPA with Pulmonary, Cutaneous, Digestive and Neurogical manifestations.

Case description:

A 37 years-old female was presented to our department complaining of severe joint ache in her left knee and ankle joints. She had a feeling of numbness in her left leg and hand for the last month. Personal medical history was positive for a diagnosis of asthma for the last three years treated with salbutamol inhaler and anti-histamine medication. The patient also reported a recent weight loss of more than 4 kg and episodes of recurrent respiratory infections treated with unspecified systemic antibiotics. Skin examination showed a painful infiltrated plaque affecting the surface of the left thigh for the last 6 weeks.

The skin biopsy revealed multiple foci of vasculitis, characterized by parietal fibrinoid necrosis and eosinophil infiltration, were present in the mid-dermis and hypodermis, consistent with eosinophilic necrotizing polyangiitis. Laboratory investigations showed a notable peripheral blood eosinophilia, however, cytoplasmic ANCA (C-ANCA) and perinuclear ANCA (P-ANCA) were negative. X-ray of the lungs showed bilateral infiltrations and hilar involvement. A high-resolution CT chest-abdomen showed subtle centrilobular ground-glass nodules along the bronchovascular structures and minimal peritoneal effusion. Electroneurography reported a length-dependent, sensory-motor polyneuropathy. These findings were in consistence with the diagnosis of EGPA.

The patient was started on prednisone 1mg/kg/day and cyclophosphamide pulse 0.75g per month. Neurological symptoms diminished gradually and the number of eosinophils normalized.

Conclusion:

EGPA is a type of ANCA-associated vasculitis that predominantly affects small- and medium-sized vessels of many organs simultaneously.

Dermatologic manifestations are estimated to occur in around 50% of patients, representing a relatively frequent type of involvement.

ANCA is negative in most EGPA patients.

A therapeutic regimen involving corticosteroids and immunosuppressants is effective in preventing this disease from progression and sometimes leading to remission.

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Abstract N°: 7365

Anti- Synthetase Syndrome with Kaposi Sarcoma: A rare association

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Introduction:

Anti-synthetase syndrome (ASS) is an autoimmune disorder characterized by the presence of autoantibodies targeting aminoacyl transfer RNA synthetases (aaRS). Clinical manifestations include interstitial lung disease (ILD), myositis, Raynaud's phenomenon, fever, mechanic's hands, and arthritis. Typically, ASS is not a associated with malignancies in adults. However, there have been two reported cases of ASSD associated with Kaposi sarcoma. Here, we present a rare case of this association involving a 64-year-old woman diagnosed with ASSD

Case report:

A 64-year-old woman with a history of diabetes and ischemic heart disease presented with symptoms of ASS for the past 6 months, including fever, arthritis, polymyositis, and diffuse interstitial lung disease as revealed by a computed tomography scan. anti-Jo1 and anti-R052 antinbodies were positive.

The patient was hospitalized in dermatology department for disseminated skin lesions. Physical examination revealed angiomatous nodules in the left sub-palpebral region and on the toes, along with erythematous-violet macules on the face, trunk, and lower limbs. Purple patches were observed on the chin, ears, palmar-plantar region, and in the intergluteal fold, with Purple papules and nodules on the tongue and the rest of the oral mucosa. Kaposi sarcoma was confirmed on a skin biopsy. HIV 1/2 antibodies were negative. Investigations revealed hematological, neurological, and digestive involvement of the Kaposi sarcoma. A taxane-based chemotherapy was started, but the patient passed away one week after her first session.

Discussion:

Kaposi sarcoma (KS) is a rare angioproliferative tumor that rarely occurs in association with ASS. This latter is not usually associated with malignancy in adults.

The presence of anti-Jo1 antibodies in ASS patients is associated with a higher cancer risk, especially when accompanied by anti-Ro52 antibodies. Our patient presented ASS with both anti jo1 and anti R052 antibodies

The first case of KS occurred in an ASS was reported by Laura B at 2013, after 2 months of treatment

by glucocorticoid Therefore, glucocorticoid-induced KS could not be ruled out. Sellitto et al., reported a case of KS with ASS paraneoplastic syndrome in which ASS was stable during chemotherapy and relapsed after chemotherapy discontinuation. Nan He et al., reported a case ASS complicated with KS 2 months after being diagnosed with ASS. Our patient was diagnosed with KS six months after ASSD onset without prior immunosuppressive treatment. The relation between ASS and KS is not fully understood, therefore more studies are needed to precise this pathophysiological association

Conclusion:

The link between KS and ASS is seldom documented. It is advisable to implement preventive cancer screening and

regular follow-up, including dermatological examinations, for ASS patients, particularly those undergoing immunosuppressive therapy

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Abstract N°: 7370

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

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Introduction & Objectives:

Morphea is a skin condition characterized by excessive deposition of collagen in dermal and/or hypodermic thickening. Lupus erythematosus panniculitis (LEP) is manifested by infiltrated plaques with lesions that ulcerate in 30% of cases. The coexistence of LEP and morphea in the same lesion with a linear and extremely rare distribution, all the more associated with systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APLS)

Herein we report a challenging case of morphea-lupus panniculitis-LES-APLS in a patient

Case description:

A 22-year-old female with deep morphea under strong topical corticosteroids for 3 years was presented to our department for ulcerations on an indurated plaque of the posterior face of the right forearm and the left shoulder. She initially noticed deep subcutaneous nodules which evolve into ulceration. Some of these nodules which spontaneously resolved left depressed lipoatrophic areas.

Standard X-ray of the right upper limb did not show calcium deposits. The biological assessment showed the presence of lymphopenia at 920, haemolytic anemia at 10.3, positive anti-nuclear antibodies (ANA) and anti-ds-DNA antibodies, false syphilitic serology VDRL + 8 and positive anti-phospholipid antibodies. Cutaneous biopsy revealed dense fibrosis of the dermis and hypodermis associated with lymphocytic panniculitis. Direct immunofluorescence does not objectify immunoglobulin or complement deposits. These findings were in consistence with morphea-lupus panniculitis associated with SLE and APL syndrome.

The patient was treated with prednisone (1mg/kg/day), synthetic antimalarials (400mg/day), methotrexate (15mg/week), salicylic acid (75mg/day) and PUVA therapy. These treatments resulted in complete resolution in 6 months and no new lesions had developed.

Conclusion:

Overlap syndromes are defined as disorders that meet the diagnostic criteria for two or more diseases concurrently or consecutively.

The differentiation of LEP occurring without typical skin lesions of LE from deep morphea can be difficult because both diseases may show lipoatrophy, scleroderma-like lesions and deep subcutaneous nodules and/or plaques. They may also share some histopathological findings such as lymphocytic panniculitis, lymphoid nodular structures in the fat, broadening of fibrous septa of fat lobules and lymphocytic vasculitis.

In our case both co-existing of signs of morphea and lupus panniculitis also, positive ANA and anti-ds-DNA and APL, the excellent response to the established treatment strongly suggested the overlap syndromes in our patient

Only cases of morphea-lupus panniculitis overlap have been reported in the literature and are still rare. Umbert and Winkelmann first described the first case in 1978.

We report a historical case of morphea-lupus panniculitis overlap associated with systemic lupus erythematosus

and APL syndrome. To our knowledge, no case is described in the literature associating this triad.

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Abstract N°: 7373

Thoracalgia and facial malignant staphylococcal infection: is there a link?

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Thoracalgia and facial malignant staphylococcal infection: is there a link?

Introduction & Objectives:

Facial malignant staphylococcal infection is a rare but life-threatening condition, characterized by a rapidly progressive cellulitis, usually with *Staphylococcus aureus* bacteriemia, and a high risk of intracranial thrombotic complications.

Materials & Methods:

We report the case of a 26-year-old male with a relevant medical history for schizophrenia who presented to our emergency room with a 3-day history of upper-lip edema and erythema on the right side of his face, that had progressed to the nasal pyramid on the same side. He mentioned he had manipulated a lesion of beard folliculitis 3 days prior. He also mentioned he had constant chest pain since the symptoms had begun. Upon admission, his vitals revealed a fever of 40.0ºC. Besides the edema and erythema, an 8mm erosion with purulent discharge could be seen over his upper lip. Lab results revealed neutrophilic leukocytosis (21.000 leukocytes/uL with 17.840 neutrophils) and CRP 14mg/dL; troponin was negative. Given the chest pain, an EKG was performed, which showed no abnormality. A CT-scan of the face was also performed, which allowed exclusion of deeper complications, such as abscesses or orbital cellulitis. The patient was admitted for intravenous therapy with piperacillin/tazobactam, after pus sample and blood cultures were drawn for microbiological analysis. Two days later, methicillin-sensitive Staphylococcus aureus was isolated in both pus and blood cultures, thus leading to therapy de-escalation to flucloxacillin.

Regardless, the patient maintained constant chest pain, which had pleuritic characteristics and, therefore, warranted a CT-scan. Surprising, the CT showed multiple cavitated pulmonary nodules, suggestive of septic embolization. He then underwent a cervical doppler ultrasound as well as echocardiography, which ruled out ophthalmic vein thrombosis and endocarditis, respectively. He was then put on therapeutic anticoagulation. While on flucloxacillin, he showed favorable clinical evolution course, with total resolution of both facial cellulitis and lung suppurative nodules after six weeks of antibiotic therapy.

Conclusion:

To our knowledge, this is first reported case of septic pulmonary embolization following malignant staphylococcal infection. In cases of facial cellulitis (especially if bacteremia is also present) it is important to rule out any embolic complications, which more frequently occur in intracranial locations, such as the ophthalmic or facial veins. Diabetes mellitus, manipulation of lesions and poor hygiene are major risk factors. Rapid initiation of adequate antibiotic therapy is key to avoiding major complications and, ultimately, reduce death risk.

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Abstract N°: 7380

Cutaneous manifestations of monoclonal gammopathies, the results of a large retrospective study in Greece.

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Introduction & Objectives:

Monoclonal gammopathies (MG) encompass a wide spectrum of hematological diseases characterized by the proliferation of clonal plasma cells that produce monoclonal immunoglobulins, either in their entirety or as fragments. Clinical manifestations range from asymptomatic disease, such as monoclonal gammopathy of undetermined significance (MGUS), to severe disease such as multiple myeloma (MM). The kidneys, peripheral nerves and skin are the organs most often affected by the disease.

Numerous skin diseases have been associated with MG and they have been distinguished into 4 subgroups. The first group of skin diseases is a result of the proliferation of malignant plasma cells or the direct deposition of paraprotein in the skin. Characteristic examples are plasmacytoma and POEMS syndrome. In the second group are skin diseases strongly correlated with the appearance of monoclonal gammopathy such as scleromyxedema and scleroderma. In the third group are classified skin diseases that have been associated with the existence of MG in patients' case series and finally the fourth group includes non-specific skin symptoms related to disease as adverse reactions to treatments, infections etc.

The purpose of the study was to record and classify the skin diseases in a large cohort of patients with monoclonal gammopathy.

Materials & Methods:

Retrospective observational study of patients with MG diagnosed from January 2020 to December 2023 in the national referral center for Plasma Cell Disorders at Alexandra Hospital in Athens, Greece.

During this period, 84 MG patients were referred to the dermato-oncology clinic at "Andreas Sygros" hospital for dermatological assessment and treatment.

Results:

84 subjects were included in the study. 4 cases in group I (2 cutaneous amyloidosis, 1 plasmatocytoma, 1 Cryoglobulinemia), 16 cases in group II (7 cases of vasculitis, 4 cases of scleromyxedema, 3 cases of Sweet's syndrome, 2 xanthomas, 1 case of Schnitzler syndrome). In group III 31 cases were included with the majority of cases being SCCs and BCCs. One case of Kaposi sarcoma and 2 mycosis fungoides were also included. The greater part of the study belongs in group IV with a total of 32 cases. More than half of the cases were drug reactions followed by infections. An interesting finding was a rare case of post kala-azar dermal leishmaniasis. The results of the study are presented on chart 1.

Conclusion:

The treatment of MM in the recent years has shown leaps and bounds with the discovery of new therapeutic agents that increase the survival of patients. However, these advancements often come with significant side effects. Our study highlights the escalating reports of skin drug reaction over time, reflecting a growing concern. Notably, MM patients have an increased risk of developing skin cancer, specifically SCC aligning with existing literature.

A striking finding is that MM patients on immunosuppressive treatments may present with atypical rashes and rare diseases such as the PKDL case or cutaneous lymphomas. Emphasizing proactive patient education to enhance early detection of cutaneous diseases is imperative. Moreover, regular and vigilant monitoring for both disease progression and therapy-related complications is crucial. Appropriate and prompt intervention for toxicities are all important steps toward ensuring longevity of MG treatment.

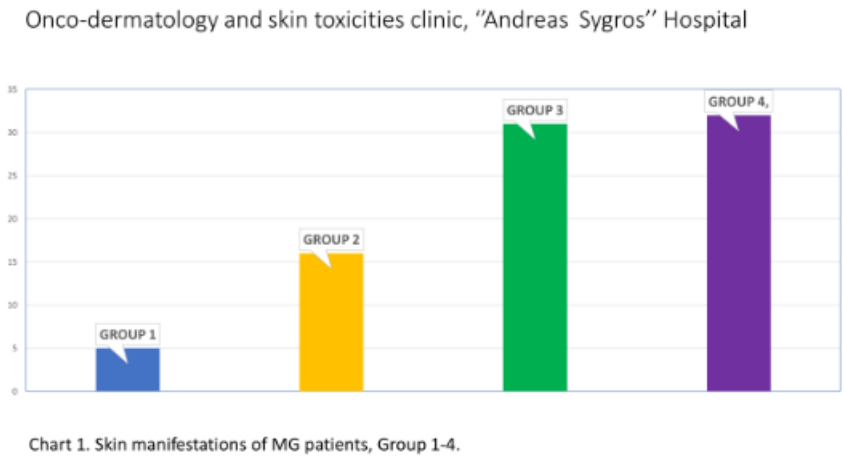

Chart 1. Skin manifestations of MG patients, Group 1-4.

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Abstract N°: 7410

Atrial fibrillation related to dermatomyositis revealed by corticosteroid bolus

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Introduction & Objectives:

Dermatomyositis is a rare auto-immune disease with characteristic cutaneous findings and varying muscular and systemic involvement. Amongst extra-musuclar organs, cardiac participation's prevalence is underestimated but of essential recognition due to potentially fatal outcome.

Patient & Observation:

A 48-year-old patient with asthmatic bronchitis, diabetes and 3 year history of anti-SAE dermatomyositis on corticosteroids and hydroxychloroquine presented with a cutaneomuscular relapse that had been evolving for 2 months. Examination revealed erythema of the face and neckline, gottron papules, a manicure sign, cervical ulcerations and a proximal muscle deficit in the scapular and pelvic girdles. Blood tests were normal, apart from elevated muscle enzymes and anemia. Noting dysphagia, and after a pre-therapeutic assessment with no abnormalities, the patient was started on a bolus of corticosteroids at a dose of 1g/day for 3 days. During the second course, the patient developed palpitations and tremors of the extremities, leading to discontinuation of treatment and performance of a new ionogram, which was normal, and an electrocardiography (ECG) showing atrial fibrillation. The patient had a heart ultrasound that turned out normal and a Holter ECG showing paroxysmal atrial fibrillation, for which she was initiated on amiodarone with good outcome.

Discussion:

Dermatomyositis is an inflammatory disease affecting skin, and inconstantly skeletal muscles and visceral organs. Among its specific autoantibodies, the anti–small ubiquitin-like modifier activating enzyme (SAE) antibody is rare, and anti-SAE positive dermatomyositis (DM) is an uncommon subgroup that presents with characteristic skin features, possibly diffuse rash, dysphagia, and mild muscle weakness.

The originality of our case lies in the fortuitous discovery of a cardiac disorder in an anti-SAE dermatomyositis patient. ### Cardiovascular involvement in dermatomyositis is frequent but often subclinical, and can consist of pericarditis, myocarditis, arrhythmia, or sinus tachycardia. Its recognition is crucial as it is a marker of poor prognosis leading to irreversible dysfunction or even death. As most ECG alterations are non-specific (ST-T abnormalities, ventricular premature complexes, bundle branch block), more advanced diagnostic methods may be required, such as Cardiovascular magnetic resonance (CMR) and Positron emission tomography (PET) imaging. ### Treatment includes heart medication, corticosteroids, and immunosuppressive therapy.

Conclusion:

Cardiac manifestations in dermatomyositis are frequent but underestimated given the diagnostic challenge they pose. Clinical examination, ECG and ultrasound are recommended and management is crucial due to the poor prognosis.

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Abstract N°: 7452

A Shot in the Dark or How to Solve a Gangrene in a Patient with Systemic Sclerosis. A Case Report

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Introduction & Objectives: Digital ulcers (DUs) occurring on the fingers with systemic sclerosis (SSc) are often very challenging to treat. They are associated with considerable pain and may lead to infection, gangrene, thus contributing to functional disability, as well as reduced quality of life. Gangrene is present in around 15- 30% of the patients with SSc and can lead to autoamputation. Effective therapy in these situations remains elusive despite the various treatment alternatives. We present a highly challenging case of a patient with systemic sclerosis and digital gangrene that regained its functionality.

Materials & Methods: A 50-year-old female with a 10-year-old history of systemic sclerosis, including Raynaud phenomenon, interstitial lung disease, pulmonary hypertension, pericarditis, dysphagia and arthralgia presented with a 2-month-old dry gangrene at the tip of her right thumb accompanied by severe pain . The digital manifestations debuted two years before, but any systemic treatment was refused by the patient at the time.

Results: The patient was started on methotrexate 15 mg/week, colchicine 1 mg/day, amlodipine 5 mg/d, pentoxyfilline 1,2 g/d, aspirine 75 mg/d, tramadol 37.5 mg/d, and vitamin E 100 mg/d with local debridement. A creme with nitroglicerine 2% was applied topically. Significant improval of the thumb was noticeable at a six-month follow-up.

Conclusion: DUs in SSc are linked to microvascular impairment and are very frequently a challenge for the clinician. Vasoactive and vasodilatatory approach should always be taken into consideration and initiated as soon as possible. The presented case is particular due to positive outcome of the gangrene despite the long period between the debut of the manifestations and the inition of therapy.

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Abstract N°: 7467

Pediatric necrobiosis lipodica: A case report

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Introduction & Objectives:

Necrobiosis lipodica (NL) is a rare chronic granulomatous dermatosis presenting with red-brown plaques that have sharply demarcated borders and an atrophic center. While its etiology has not been clarified, many studies point out diabetic microangiopathy as the major etiological factor since NL has an association with diabetes mellitus. Mean age of diagnosis is 30 to 40 years, with a female predominance. NL in children is extremely rare. We present a pediatric case of NL with no history of diabetes.

Case report:

An** 11-year-old male patient presented with an asymptomatic, telangiectatic, erythematous, atrophic plaque on the anterior-distal leg, 7x3 cm in size. The lesion had failed to respond to previous mid-potency topical corticosteroid treatment. BMI and physical examination were normal. A review of the systems and patient history was insignificant. The punch biopsy confirmed the diagnosis of NL. Detailed laboratory evaluation revealed a glycosylated hemoglobin (HbA1C) of 6.0% (3.5-5.6%), consistent with prediabetes. The post-prandial blood glucose was 113 mg/dL (70-100 mg/dL). The patient was started with a combination of topical clobetasol propionate and tacrolimus and was referred to the pediatric endocrinology department for further investigation.

Conclusion:

Necrobiosis lipoidica usually presents with lesions in the lower extremities, which are asymptomatic unless ulcerated. Its etiology is unknown, and there is very little data on pediatric cases. The diagnosis is made through clinical and histopathological features. Histopathological findings may vary between diabetic and non-diabetic patients.

Necrobiosis lipoidica, strongly associated with diabetes, has a prevalence of 0.3 to 1.2% among diabetic patients, two-thirds of whom are type I diabetic. In pediatric diabetes, its prevalence is between 0.6% and 1.6%, and up to 2.3% in pediatric type I diabetes. Very few non-diabetic pediatric patients have been reported in the literature. In a recent review of 31 pediatric NL cases, only 6 did not have diabetes mellitus.

Treatment of pediatric NL is challenging with a high rate of therapeutic failure. Potent topical and intralesional corticosteroids are considered as first-line therapy. Topical tacrolimus, local photochemotherapy, systemic corticosteroids, ciclosporin, hydroxychloroquine, pentoxifylline and TNF- α inhibitors have also been used for its treatment.

Due to its low incidence in children, pediatric NL may easily be overlooked, especially in non-diabetic patients. Since our patient had a prediabetic HbA1C level, he was referred to pediatric endocrinology. As dermatologists, although rare, we believe it is important to keep pediatric NL in mind as a possible diagnosis, and to evaluate blood glucose levels and HbA1C in these children.

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Abstract N°: 7518

Cutaneous endometriosis: a fluctuating, bleeding tumor

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Introduction

Endometriosis is defined as the presence of endometrial-like tissue outside of the uterus**1** It is a benign gynecological disorder affecting around 5% of women. The pelvic cavity is the most common location of endometriotic implants. About 12% of the lesions are extragenital** and may involve the skin, the abdominal wall being the most common (0.04% al 5.5%).

Case report

An otherwise healthy 39-year-old woman presented with a 1-year history of an exophytic, purplish, 4.5x2.5x2.5 cm morifform tumor over a previous linear scar due to a cesarean section 7 years prior. She referred spontaneous fluctuations in size, bleeding, and cessation of bleeding without receiving therapy. Red-white background, purple and red lacunae, as well as a papilliform architecture were observed on dermoscopy. The lesion was surgically excised. Islets of columnar, decapitated, secretory cells surrounded by a vascular stroma and a chronic inflammatory infiltrate was noted on histopathology.

Discussion

There are few dermoscopic descriptions in the literature of cutaneous endometriosis. These include homogeneous reddish pigmentation, red globular structures (atolls), amorphous brown areas, polypoid projections of erythematous violaceous color, dark brown globules, white reticular pattern, and areas of active bleeding. Notably, dermoscopic and clinical appearance is subject to change depending on hormonal cycle and skin phototype.

The diagnosis is confirmed on histopathology, where glandular elements and stroma resembling those of the endometrium are observed.

Therapeutic recommendations are limited to case series and descriptive studies. Surgical excision with 1 cm magin is considered the standard of treatment. Involvement of the rectus muscle have been associated with higher recurrence. Oral contraceptives, progesterone, and danazol result only in symptomatic improvement but none can effectively eliminate the endometrioma.

Malignant transformation is rare. Extensive resection is recommended to avoid postoperative recurrence, but due to the lack of evidence and being a rare pathology, the efficacy and long-term complications are unknown. Some reports reveal a postoperative recurrence rate of 4.5% to 11.2%.

Conclusion

This case aims to highlight a rare entity for dermatologists and non-dermatologists, in order to suspect the posibility of an extracutaneous disease when encountering a rapidly enlarging, bleeding, apparently cutaneous mass.

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Abstract N°: 7545

A not so Sweet outcome: highlighting a rare histological variant with prognostic value

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Introduction & Objectives:

Sweet syndrome (SS) or acute febrile neutrophilic dermatosis classically presents as tender erythematous plaques or nodules with sudden onset accompanied by fever, joint pain and ocular inflammation. It may occur in conjunction with several inflammatory and autoimmune diseases, malignancies, infections or drug exposure. When associated with inflammatory bowel disease (IBD), it may be revealing of the diagnosis or signal increased disease activity.

Histiocytoid SS represents a rare histological variant of SS characterized by a dermal infiltrate primarily composed of lymphocytes and histiocytoid mononuclear cells expressing myeloperoxidase (MPO). This particular variant is associated with myelodysplastic syndrome (MDS) in up to one third of patients. As skin lesions may precede the diagnosis of a hematological malignancy, evaluation is compulsory at diagnosis and blood cell count should be monitored for at least 6 months.

Materials & Methods:

We present the case of a 45-year-old female patient admitted to the Infectious Diseases department for high fever of unexplained etiology alongside an eruption consisting of erythematous, edematous papules and plaques, some with targetoid appearance, some with peripheral desquamation, slightly pruritic with an acral distribution (palms, forearms, legs). The symptoms had appeared 2 weeks after initiating Mesalamine therapy for her ulcerative colitis.

Given the time lapse between the introduction of 5-ASA, the morphology and location of the skin lesions an initial diagnosis of drug-induced erythema multiforme-like eruption was presumed. Laboratory tests including various serologies, stool and throat swab cultures did not identify an infectious cause of the persistent fever, nor any gravity markers of a drug eruption such as elevated LFTs or eosinophilia.

Results:

Histopathological examination showed interstitial lympho-histiocytic inflammation with positive MPO staining and mixed, predominantly septal panniculitis, clinically compatible with histiocytoid Sweet syndrome associated with erythema nodosum-like lesions. Pulse therapy with Dexamethasone was initiated resulting in fever remission and attenuation of the skin lesions.

Conclusion:

While an abrupt-onset cutaneous rash along with pyrexia are defining features of SS, which may in turn be associated with IBD, Mesalamine which is a commonly used medication for IBD can generate the same adverse reactions which characterize SS. The challenge in our case was to differentiate between the two entities given the two-sided context. While the histiocytoid variant of SS is rarely associated with IBD, it conversely requires further

hematological follow-up so as not to miss the development of MDS.

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Abstract N°: 7547

Cutaneous leukocytoclastic vasculitis: A case report with onset during pregnancy.

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Introduction & Objectives:

Among the diverse primary vasculitides that may impact young women during their reproductive years are Takayasu arteritis (TAK), polyarteritis nodosa (PAN), ANCA-associated vasculitis (AAV), immune-complex small-vessel vasculitis (IgA vasculitis), and Behçet's disease (BD).

Materials & Methods:

We report a case of cutaneous leukocytoclastic vasculitis that occurred during pregnancy.

Results:

A 27-year-old female patient, 24 weeks pregnant, is being monitored for gestational diabetes. Her medical history includes a miscarriage in the first trimester three years ago. She was admitted to our department for the management of painful diffuse purpuric and necrotic lesions that have been evolving for the past 3 weeks before the consultation, without any other associated signs.

The clinical examination revealed a patient in good general condition. Skin examination identified a necrotizing ulcerative plaques on the posterior of both legs with diffuses purpuric ecchymotic patches. The remainder of the clinical examination, including the obstetrical assessment, did not show any specific abnormalities.

The histology of the lesions suggested leukocytoclastic vasculitis .Direct immunofluorescence revealed weakly positive C3 along the vascular walls.

The biological, immunological infectious and etiological assessments were normal.

The treatment was intravenous methylprednisolone bolus for 5 days, followed by oral corticosteroid therapy with prednisone at a dose of 0.5mg/kg/day.

The progression was marked by a very favorable clinical outcome for both the mother and the child.

Conclusion:

Limited data exist regarding pregnancies in the context of systemic vasculitides. Published cases of vasculitis occurring during pregnancy are scarce, suggesting a minimal role of pregnancy in triggering or exacerbating these conditions.

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Abstract N°: 7596

An unsettled case of (pseudo)porphyria and its curious association with systemic sclerosis

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Introduction & Objectives:

Porphyria cutanea tarda (PCT) is a photosensitive vesiculobullous skin disease characterized by an accumulation of porphyrins mostly in urine, but also in stool and plasma due to an enzymatic deficiency (uroporphyrinogen decarboxylase) in the pathway of heme biosynthesis. Susceptibility factors include alcohol, estrogen use, smoking, viral infections, chronic kidney disease and iron overload.

Pseudoporphyria is a clinically similar entity but typically lacking the abnormal porphyrin levels. The differential diagnosis between PCT and pseudoporphyria can be particularly problematic in the context of renal failure and hemodialysis. The distinction carries therapeutic implications, as PCT is commonly treated with hydroxychloroquine (HCQ) while pseudoporphyria has been reported to respond to N-acetylcysteine. It's hypothesized that HCQ forms high-molecular weight complexes with porphyrins which renders them unfilterable by the dialysis membrane.

Materials & Methods:

We present the case of a 62-year-old female patient with a history of chronic HCV infection and end-stage renal failure under dialysis since the age of 35. She was addressed to the Rheumatology department for a clinical suspicion of CREST syndrome (sclerodactyly with flexion deformity, squared-off telangiectasias, distal digital ulcerations, acro-osteolysis with onychodystrophy, perioral radial furrowing, Raynaud's phenomenon) in the absence of paraclinical criteria (negative autoantibodies for scleroderma and non-specific capillaroscopic findings).

The patient also displayed clinical features of PCT (history of bullae on photo-exposed areas, skin fragility with multiple ecchymoses and hematic crusts, facial hyperpigmentation and hypertrichosis) in a multi-suggestive context of hepatitis C, hyperferritinemia and hemodialysis.

On the other hand the diagnosis of porphyria did not account for the gastrointestinal and pulmonary manifestations, namely oesophageal stasis and interstitial fibrosis.

Results:

Considering the polymorphic manifestations and associated conditions, a diagnosis of limited systemic sclerosis and porphyria cutanea tarda was made. However, pseudoporphyria could not be excluded without determining fecal or blood porphyrin levels which were not obtainable in our unit.

Conclusion:

This case is remarkable from multiple perspectives. Firstly, the clinical picture encompasses features of both PCT and systemic sclerosis. While sclerodermoid changes have been reported in PCT the latter does not explain all the

findings, particularly the esophageal and pulmonary involvement. The association between the two conditions has only been reported in about 10 cases to our knowledge.

Secondly, the patient presents multiple triggering conditions for both PCT and pseudoporphyria, most notably undergoing kidney dialysis. Lastly, the differentiation between the two could not be established given the context of anuria and the unavailability of stool or plasma measurement of porphyrins.