Abstract N°: 35

"Atypical Presentation of Multiple Cherry Hemangiomas Associated with Congenital Vascular Anomaly and Arm Enlargement: A Diagnostic Insight"

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Introduction & Objectives:

Cherry hemangiomas are benign vascular proliferations that commonly occur in adults, which are often small and stable in size and amount. However, rapid growth, increasing amount, or localization with congenital vascular anomalies is unusual. We present a 62-year-old female with multiple, rapidly enlarging cherry hemangiomas on her right arm and breast, associated with a congenital red hyperpigmented birthmark and arm enlargement. This case explores:

1. The underlying pathophysiology of the vascular anomalies.
2. Differential diagnoses, including Klippel-Trénaunay Syndrome (KTS) and POEMS syndrome.
3. The significance of localized arm enlargement in relation to systemic or syndromic associations.

Materials & Methods:

Clinical Evaluation: Detailed history and physical examination focusing on onset, progression of hemangiomas, birthmark characteristics, and arm enlargement.

Imaging Studies: Doppler Ultrasound: To assess vascular malformations and blood flow. MRI with Contrast: To identify soft tissue, vascular, and lymphatic anomalies. CT Angiography: To rule out systemic vascular abnormalities.

Histopathological Analysis: Biopsy of a representative hemangioma was stained with hematoxylin and eosin (H&E) and tested for immunohistochemical markers (CD31, D2-40).

Laboratory Investigations: VEGF, inflammatory markers, and monoclonal protein levels for syndromic evaluation (e.g., POEMS).

Differential Diagnosis Considered: Klippel-Trénaunay Syndrome (KTS), POEMS Syndrome, Parkes Weber Syndrome, Lymphatic malformations with secondary vascular changes.

Results:

Clinical Findings: Multiple cherry hemangiomas on the right arm and breast had progressively increased in size over three months. The birthmark was a well-demarcated red hyperpigmented patch consistent with a capillary malformation. The right arm showed chronic enlargement without lymphadenopathy, ulceration, or systemic symptoms.

Imaging Findings: Low-flow vascular malformations were identified in the arm, with no evidence of arteriovenous shunting or systemic vascular anomalies.

Histopathology:

Hemangioma biopsy revealed dilated capillaries lined by endothelial cells, consistent with benign cherry hemangiomas.

CD31 staining confirmed vascular origin; D2-40 was negative, excluding lymphatic involvement.

Laboratory Results: Normal VEGF and inflammatory markers, with no monoclonal protein detected.

Conclusion:

This case illustrates a rare presentation of multiple rapidly growing cherry hemangiomas associated with localized arm enlargement and a congenital vascular anomaly. While systemic syndromic associations like POEMS syndrome were excluded, the findings support an atypical variant of Klippel-Trénaunay Syndrome. Thorough evaluation using imaging, histopathology, and laboratory workup is critical in differentiating benign vascular anomalies from potential systemic conditions. Further research into the mechanisms linking vascular malformations and hemangioma proliferation is recommended.

Abstract N°: 365

Erythema Elevatum Diutinum in a patient with an unusual polymorphic presentation: A Case Report

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Introduction & Objectives:

Erythema elevatum diutinum (EED) is a rare, chronic inflammatory dermatosis classified within the group of leukocytoclastic vasculitis. The etiology of EED remains unclear; however, it is hypothesized to involve immune complex deposition in the vascular walls, leading to inflammation and vascular damage. Clinically, EED presents as symmetrical, firm, red to purple papules, plaques, or nodules, predominantly located on the extensor surfaces of the limbs, particularly around joints. These lesions may be asymptomatic or associated with pain or burning sensations. EED has been associated with various systemic conditions, including hematological disorders, autoimmune diseases, infections, and malignancies, particularly IgA monoclonal gammopathy and myelodysplastic syndromes. Given its potential to precede or coincide with systemic diseases, early recognition and diagnosis of EED are crucial for appropriate management. This report aims to describe a rare manifestation of EED, emphasizing the clinical course, diagnostic challenges and therapeutic response.

Materials & Methods:

A 64-year-old male truck driver with a history of hidradenitis suppurativa and comorbidities including chronic heart failure, type II diabetes mellitus and obesity, has been admitted to our department. The patient reported a five-day history of fever and painful skin lesions. Cutaneous lesions refractory to prior antibiotic treatment displayed multiple erythematous plaques and bullous changes predominantly on the forearm, hand, and face. Palpable induration is noted beneath erythematosquamous plaques. Laboratory findings revealed elevated inflammatory markers CRP 298 mg/L (range: 0.00-5.00), ASLO 629.0 IU/ml (range: 0.0-200.0), 13,7*10^9/L leukocytes (range: 4-10), 10.30*10^9/L neutrophils/L (range: 1,9-8.00). Antinuclear antibodies (ANA), cytoplasmic anti-neutrophil cytoplasmic antibodies (c-ANCA), and anti-double stranded DNA (anti-dsDNA) antibodies were positive, accompanied by elevated levels of IgA and IgG. Imaging studies identified liver cirrhosis and dilated cardiomyopathy. Histopathology confirmed leukocytoclastic vasculitis, leading to the diagnosis of EED.

Results:

The patient received intravenous corticosteroids, which were later transitioned to oral administration, along with antibiotics according to sensitivity (ceftriaxone and ciprofloxacin), antihistamines and topical antiseptics. The cutaneous manifestations gradually improved with no new lesions developing. The patient was discharged in stable condition under outpatient care.

Conclusion:

EED is a rare cutaneous vasculitis with potential associations to systemic diseases. Early recognition and diagnosis are essential for appropriate management and monitoring of potential underlying conditions. While dapsone remains the mainstay of treatment, other therapeutic options are available and treatment should be tailored to the individual patient, considering any associated systemic diseases.

Abstract N°: 760

Possible haematological indicators for livedoid vasculopathy include platelet count and platelet concentration?.

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Introduction & Objectives: Livedoid vasculopathy is an uncommon vasculopathy that is usually characterized by lesions in both lower limbs. It is thought that thrombus formation in the capillary vasculature is caused by the interaction of endothelial injury, decreased fibrinolytic activity, and increased thrombotic activity. There is yet no recognized pathophysiology for livedoid vasculopathy (LV). Despite the fact that LV is where platelet activation takes place, not much is known about the morphological parameters of LV platelets. Investigating if platelet shape changes in LV and their clinical importance was the aim of this investigation.

Materials & Methods: Included were 30 patients with LV and 30 individuals with cutaneous small vessel vasculitis (CSVV), all of whom were in the active stage. Patients with CSV and LV in both active and stable stages had their platelet parameters compared. Analysis was done on the correlations between these platelet characteristics and the LV composite clinical ratings.

Results: The average age of LV patients was 27.9 years (range: 15–62 years), with women making up 83.4% (25/30) and men making up 16.6% (5/30). Following treatment, LV patients' platelet counts and plateletcrit (PCT) levels were noticeably higher than those of CSVV patients, and active-stage LV patients had higher PCT levels than stable-stage LV patients. Platelet count and PCT levels were favorably connected with LV patient composite clinical scores that represented the severity and activity of the disease.

Conclusion: In LV patients, altered platelet morphology was found. LV activity and relapses can be predicted early with platelet count and PCT, which can also be used to distinguish between LV and CSVV.

Abstract N°: 887

A case report of acroangiodermatitis (pseudo-Kaposi sarcoma) unsuccessfully treated with doxycycline

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Introduction & Objectives:

Acroangiodermatitis (AAD), or pseudo-Kaposi sarcoma, is a rare benign vascular proliferation often associated with chronic venous hypertension (Mali type) or arteriovenous malformations (Stewart-Bluefarb syndrome). Various treatments, including compression therapy, dapsone, and macrolides, have been described, but no standardized therapy exists. This case highlights the failure of doxycycline, an off-label treatment proposed for its anti-inflammatory and anti-angiogenic effects, emphasizing the need for further research on AAD pathogenesis and treatment.

Materials & Methods:

A 73-year-old woman with chronic venous insufficiency and a two-year history of violaceous plaques on the lower legs was diagnosed with Mali-type AAD based on clinical and histopathological findings. Due to anemia and macrolide allergy, first-line therapies were contraindicated. The patient received doxycycline 100 mg twice daily for two months as an off-label treatment. Clinical response was assessed at baseline, one month, and two months, evaluating lesion improvement, symptom control, and adverse effects.

Results:

Initial response included partial improvement of pruritus. However, after three weeks, lesions failed to regress, and symptoms returned to baseline, leading to treatment discontinuation at two months. Compression therapy, initially refused, was later self-initiated by the patient, resulting in significant improvement, suggesting its pivotal role in AAD management.

Conclusion:

Doxycycline, despite its theoretical anti-inflammatory benefits, did not yield clinical improvement in this case, questioning its role as an alternative treatment for AAD. The patient's subsequent response to compression therapy underscores the primary role of venous pressure control in AAD pathogenesis and treatment. Further research is warranted to clarify the disease's mechanisms and establish effective, evidence-based therapeutic strategies.

Abstract N°: 1156

Blue Rubber Bleb Nevus Syndrome: Often Undiagnosed and Underreported

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Introduction & Objectives: Blue Rubber Bleb Nevus Syndrome (BRBNS), also known as Bean syndrome, is a rare congenital disorder characterized by multifocal venous malformations primarily affecting the skin and gastrointestinal tract, but can also involve other visceral organs such as the liver, spleen, and lungs. Approximately 200 cases have been reported in the literature. The condition is marked by the presence of soft, compressible blue or purple nodules, termed “blebs,” which can vary in size and are often found on the trunk and extremities. The pathophysiology of BRBNS is linked to somatic mutations in the TIE2 gene, which encodes a receptor involved in angiogenesis and vascular integrity. These mutations lead to abnormal endothelial cell signaling and the formation of vascular malformations.

Materials & Methods: A 59-year-old patient with a history of arterial hypertension, gallbladder and urinary tract lithiasis presented to the Dermatology Department after detection on the contrast-enhanced CT scan the presence of multiple vascular malformations in the subcutaneous tissue of the upper limbs abdomen, chest and buttocks, with intense contrast uptake with maximum dimensions of 17x10 mm and persistence of the similar lesions on the spleen, liver and ischiorectal fossa. Local clinical examination shows multiple nodular, compressible, purplish, asymptomatic, nodular formations disseminated over the trunk and arms. The onset of the cutaneous lesions was around the age of 20 years, with a slowly progressive increase in size and enlargement until the present day. The patient denies any history of bleeding from the digestive tract. It is worth mentioning that the patient presented a marked anxiety about a possible oncological cause of these lesions. Therefore, a skin biopsy of a lesion was performed and the morphological aspects identified venular ectatic vessels of malformative type.
The biological tests were within normal limits, without the presence of anaemia, unchanged tumour markers, negative fecal occult blood test and colonoscopy did not show any suspicious lesions.

Results: Based on previously reported cases, most patients tend to seek medical attention only at advanced stages of the disease. In most cases, medical intervention is pursued when complications have already developed, or when the cutaneous lesions associated with the condition reach a size that is physically or cosmetically concerning for the patient. Furthermore, some individuals delay seeking care until the lesions are located in areas that cause functional impairment or significant discomfort, such as those interfering with mobility, daily activities, or quality of life.

Conclusion: Based on the clinical case presented, we consider the incidence of Bleb syndrome to be significantly higher than previously estimated. Therefore, this syndrome may remain undiagnosed throughout life if it is not associated with complications and does not pose problems of differential diagnosis. Even in patients who do not present with complications, early diagnosis remains a priority for the monitoring and prevention of these complications.

Abstract N°: 1284

When the Skin Becomes the Mirror of the Vessels: Sneddon's Syndrome

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Introduction & Objectives:

Sneddon's Syndrome, an underrecognized but fascinating rare condition, lies at the crossroads of dermatology and neurology. Imagine a disorder where the skin and brain engage in a strange dance, marked by erythema on the skin network, forming a livedo pattern, and ischemic strokes. Let's explore this vascular enigma through this case, whose diagnostic particularity is the early presentation of livedo and cognitive impairment, without any sensory or motor deficits.

Observation :

We admitted a 42-year-old man with a history of active smoking and previously labeled genetic generalized epilepsy, who presented with dermatological signs of persistent livedo racemosa, non-infiltrative and non-necrotic, extending to all four limbs and the buttocks for several weeks, associated with cognitive impairment. Neurological examination revealed concentration and attention difficulties, with a MMSE score of 19, and no sensory-motor deficits. Cerebral MRI showed multiple territorial cortical-subcortical infarcts, as well as distal ones, with vascular leukoencephalopathy and intracranial stenoses. The initial etiology workup for the infarcts did not reveal any cardioembolic or atherothrombotic causes. Cerebrospinal fluid (CSF) analysis showed no abnormalities, and the autoimmune workup was normal, including antiphospholipid antibody levels and antinuclear factor testing. Given this presentation of neurological/neuropsychological involvement and livedo reticularis, we concluded a diagnosis of Sneddon's syndrome in its antiphospholipid-negative form as a rare cause of ischemic strokes. The patient was started on antiplatelet and antiepileptic treatment with favorable progress.

Conclusion:

Sneddon's syndrome, although rare, represents a cause of stroke in young individuals. The fact that early signs can present solely with skin findings like livedo, without any obvious sensory or motor deficits, makes the diagnosis even more challenging. Therefore, it should be considered and quickly identified in the presence of livedo to prevent vascular complications and improve patients' quality of life.

Abstract N°: 1302

Facial kaposi sarcoma in a non-HIV patient

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Facial kaposi sarcoma in a non-HIV patient

Introduction & Objectives:

Kaposi's sarcoma (KS) is a vascular tumor that originates from endothelial cells and is caused by the human herpesvirus 8 (HHV-8). It is characterized by the appearance of violaceous macules, plaques, or angiomatous nodules on the skin or mucous, and can also affect internal organs. While KS most frequently affects the extremities, the facial involvement is considered rare. The disease can appear in different forms, including the epidemic form (associated with VIH), the classic form, the endemic form (African), and the iatrogenic form. In the classic form, KS usually touches elderly men of Mediterranean descent, with lesions typically localized to the extremities. The African form is more aggressive, often presenting with infiltrative lesions that may progress to involve internal organs. Iatrogenic KS is seen in patients with immunosuppressive conditions. VIH-associated KS, often involves the face, but also extremities, and mucous tissues.

The objective of this case is to highlight the importance of considering Kaposi's sarcoma in the differential diagnosis of vascular skin lesions on the face in immunocompetent individuals.

Materials & Methods:

We present a case study of a 67-year-old male patient with no significant medical history, who presented two progressively enlarging ipsilateral nodules on his face for the past month.

During the skin examination, two vascular-appearing nodules were observed: One nodule measured 0.5 cm in diameter and was located on the alar crease of the nose, the second nodule measured 1 cm in diameter, located at the base of the alar rim of the nose, ulcerated, and covered with a yellowish crust.

The dermoscopic analysis showed a rainbow pattern on the periphery, scales, and a central ulcer covered by a yellowish crust on the second lesion.

Further clinical examination revealed no additional skin or mucous lesions.

A skin biopsy of both lesions was performed.

Results:

The patient's diagnosis of Kaposi's sarcoma was confirmed by biopsy with immunohistochemistry showing HHV-8 positivity. Despite the facial lesions, which are atypical for the classic form of KS, the HIV serology was negative, ruling out VIH-associated KS. The patient had no history of immunosuppression or recent transplantation, and blood tests showed no underlying abnormalities.

The patient underwent a complete workup and excisional surgery of both nodules, with a good outcome.

Conclusion:

Kaposi's sarcoma is commonly found on the extremities, particularly the lower limbs, in most forms of the disease,

including the classic and iatrogenic types. Facial involvement is atypical for KS, especially in non-immunocompromised patients, but it is more frequently observed in VIH-associated KS. Although this patient did not present typical risk factors such as HIV infection or immunosuppression, the case highlights the need to consider KS in the differential diagnosis of vascular lesions, even in unusual locations like the face. Further monitoring and follow-up are essential to assess disease progression.

Abstract N°: 1436

Occult neuroblastoma in an infant with a segmental lumbosacral hemangioma

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Occult neuroblastoma in an infant with a segmental lumbosacral hemangioma

Introduction & Objectives:

Infantile Hemangiomas (IH) are common vascular tumors in newborns affecting an estimated 2.8% of infants. They are benign tumors that form from aberrant proliferation of endothelial cells that typically slowly involute by age 10. Treatment depends on the size, location, and associated conditions. Propranolol is routinely used to treat IH by inhibiting angiogenesis and inducing vasoconstriction and apoptosis. However, new treatment options such as mTOR inhibitors and laser therapy are emerging for more complex IHs. Segmental IHs in the lumbosacral region may suggest a rare condition, LUMBAR syndrome, characterized by Lower body hemangiomas accompanied by Urogenital anomalies, IH Ulceration, spinal cord Malformations, Bony defects of the spine and lower extremity, Anorectal malformations, Arterial anomalies and/or Renal anomalies. In LUMBAR syndrome, propranolol should be used with caution as occult arterial anomalies may be present.

Materials & Methods:

This case report was compiled from physician documentation in the patient's electronic medical record.

Results:

We present a case of an occult neuroblastoma in association with a segmental lumbosacral IH in a 13-month-old female infant. Prior to evaluation by dermatology, she was treated by pediatric cardiology with propranolol. Due to minimal regression of her IH, dermatology was consulted to assess the potential benefit of Pulsed Dye Laser therapy. Despite normal developmental milestones and no history of musculoskeletal abnormalities or urinary tract infections, imaging was ordered to screen for occult abnormalities which may be observed in patients with segmental lumbosacral IHs. Ultrasound subsequently identified a solid-cystic retroperitoneal mass, concerning for malignancy. Stat MRI confirmed findings consistent with neuroblastoma. However, the lesion was non-MIBG avid on MIBG scanning, indicating an absence of tracer uptake, which can suggest atypical neuroblastoma characteristics, potentially influencing staging, treatment, and prognosis. Surgical excision was completed within three weeks of the MRI results. The tumor encased the renal artery and inferior vena cava and abutted the abdominal aorta. Meticulous dissection allowed for the complete extirpation of the tumor. At the most recent follow-up, the patient had recovered, but required surgery for a SBO, likely due to adhesions from the neuroblastoma resection.

Conclusion:

While no known association exists between LUMBAR syndrome and neuroblastoma, this case highlights the need for a multidisciplinary approach in managing complex segmental IH presentations as these lesions can coincide with unexpected pathologies. Further research is needed to determine potential links between segmental hemangiomas and malignancies and to refine management strategies for infants with clinical features indicative of LUMBAR syndrome.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1606

Klippel-Trenaunay Syndrome : A Rare Diagnosis

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Introduction & Objectives:

The Klippel-Trenaunay syndrome (KTS) is a rare and complex congenital vascular disorder characterized by a classic triad: capillary malformations, venous anomalies, and hypertrophy of soft tissues and/or bones, typically affecting a limb. The pathogenesis of KTS remains not fully understood but is thought to involve somatic mutations in genes regulating vascular development. This case report highlights the clinical presentation and diagnostic challenges of KTS.

Materials & Methods:

Results:

Case presentation:

An 8-year-old girl, with no notable medical history, presented to our department with a patchy erythematous skin lesion on the left lower limb, present since birth. Clinical examination revealed a port-wine stain with irregular borders, varying in color from pink to reddish-purple, extending from the lumbosacral region to the anterolateral surface of the left lower limb, along with varicose veins following the course of the great saphenous vein in the same limb. Palpation and auscultation of the vascular axes did not reveal any thrills. Comparative measurements of both lower limbs showed a 2 cm difference in length and diameter compared to the contralateral limb. Doppler ultrasound revealed incompetence of the left saphenous vein, along with a vascular formation on the posterior side of the left thigh. CT angiography of the lower extremity revealed moderate dilation of the superficial portion of the great saphenous vein. The patient was prescribed elastic compression and scheduled for regular clinical follow-ups.

Conclusion:

Although Klippel-Trenaunay syndrome (KTS) is commonly defined by a the classic triad described above, this triad is present in only about 30% of cases. The exact etiology of KTS remains unknown, and no sex predilection has been identified. Deep venous abnormalities are often associated with the condition and may complicate its clinical presentation. Diagnostic evaluation typically involves non-invasive imaging techniques, such as Doppler ultrasound, standard radiography, and magnetic resonance imaging (MRI), to assess vascular and skeletal involvement. Given the complexity of KTS, early and multidisciplinary management is crucial, including venous compression therapy and orthopedic interventions for limb length discrepancies, to optimize functional outcomes and improve the patient’s quality of life.

EADV SYMPOSIUM PRAGUE 22-24 MAY 2025

Abstract N°: 1743

Classic Kaposi's Sarcoma

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Introduction & Objectives:

Kaposi's Sarcoma (KS) is a multifocal neoplastic disease primarily caused by infection with Human Herpesvirus 8 (HHV-8). The classic form is the most common. The aim of this work is to describe the epidemiological, clinical and treatment outcomes of classic KS.

Materials & Methods:

This is a retrospective study including cases of classic KS followed in our department from January 2014 to March 2024. Diagnosis was confirmed by histology with immunohistochemical study of HHV-8. The classic form was retained after excluding iatrogenic causes or HIV infection.

Results:

A total of 42 patients were included, with a mean age of 74.32 years. The sex ratio (M/F) was 0.69. Medical history included: diabetes (12 cases), hypertension (13 cases), cardiopathy (7 cases) and erysipelas (3 cases). No patient was under immunosuppressive treatment. Skin lesions were angiomatous, appearing as macules, nodules or plaques. Lesion sites included: lower limbs (32 cases), upper limbs (12 cases). Lymphedema was associated in 76% of cases. No visceral involvement was reported in our series. Management included: therapeutic abstention (6 cases), cryotherapy (22 cases), intramuscular bleomycin (12 cases), and radiotherapy (2 cases). Fifty percent of patients treated with cryotherapy had stable lesions, 20% showed partial improvement, 20% experienced lesion extension, while 10% could not tolerate the pain. For bleomycin, 60% of patients showed partial lesion improvement after an average of 6 cycles, 25% had stable disease, and 15% were cured, with an average duration of 9 months.

Conclusion:

Our findings align with literature regarding late onset and the predominance of lesions on the lower limbs. It is indeed an indolent disease requiring simple monitoring. In case of extensive lesions, systemic treatment is often necessary. In developing countries like ours, therapeutic options are limited. In our series, monotherapy with bleomycin stabilized the disease in most cases.

Abstract N°: 1851

Purpura like dermatitis at lower extremities induced after laser hair removal . A case report.

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Introduction & Objectives:

Laser hair removal is a well-established method for permanent hair removal with good efficacy and safety profile. Adverse events that may occur are mild and temporary and subside shortly after treatment. We would like to present a case report of a rare skin reaction after laser hair removal.

Materials & Methods:

A 32-year-old woman of Caucasian origin with skin phototype III presented in our clinic with an acute skin reaction over her bilateral lower extremities one day after undergoing a full body hair removal treatment. The clinical examination revealed a gradually deteriorating eruption of multiple red-brownish round, non-blanching, minor itching papules on both of her calves. She didn't refer any systemic symptoms. Recent infections and medications were denied. Regarding medical and family history no significant incidences were mentioned. The patient had no previous experience with laser epilation treatments. The treatment was performed with a laser 750nm Alexandrite with a dynamic cooling device (DCD). Following parameters were used: spot size 18mm, Pulse duration 3ms, Fluence 10J/cm2, Frequency 2Hz.

Results:

Besides the clinical examination we performed dermoscopy which showed wide, homogeneous, structureless purpuric areas with perifollicular halos and red-purple network patches at the background. The laboratory screening was unremarkable, however was performed to exclude thrombocytopenia, rheumatologic diseases and viral infections. The patient denied the biopsy. We suggested a short course of orally administered antihistamines and corticosteroids to accelerate clinical resolution. Moisturizing emollients and elastic garment were also recommended. At the follow-up, 2 weeks later the symptoms have subsided thoroughly.

Conclusion:

Laser hair removal induced purpura is a rare adverse event, which affects mostly women and appears at the lower extremities. To our knowledge, there are two similar cases reported in bibliography. Further studies are required to clarify the pathogenesis as well as the trigger factors in order to improve the therapeutic approach.

Abstract N°: 1945

A Rare Case of ADA-2 Deficiency (DADA-2) Presenting with Cutaneous Polyarteritis Nodosa-Like Findings

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Introduction & Objectives:

Adenosine deaminase-2 (ADA-2) deficiency, presenting with findings similar to cutaneous polyarteritis nodosa (PAN), is defined as a systemic inflammatory vasculopathy caused by mutations in the CECR1 gene. These mutations result in the loss of function of the gene encoding the ADA-2 enzyme. This syndrome primarily manifests in childhood, though it can rarely present later. In this case, we present a patient with a late-onset ADA-2 deficiency.

Materials & Methods:

We describe the diagnostic process of a 21-year-old male with leg numbness, livedo reticularis, and ulcerative lesions.The patient's family history, clinical findings, pathology results, imaging studies, and a multidisciplinary diagnostic approach were evaluated.

Results:

The patient initially had presented to an external center with severe muscle and joint pain, joint swelling, morning stiffness, and mild erosive lesions in the testicular area. Due to increasing muscle pain, a muscle biopsy had been performed, revealing findings consistent with vasculitis. Given the presence of arthritis, myalgia, and ulcerative lesions, the patient had been diagnosed with cutaneous PAN and started on methotrexate (15 mg/week) and systemic steroid therapy.

His medical history included frequent upper respiratory infections and a six-pack-year smoking history; there was no alcohol or substance use. In his family history, his mother and two uncles had been followed for an unspecified rheumatologic disease. One of his uncles had a history of recurrent febrile episodes and spontaneously healing ulcerative lesions in the pretibial region. A cousin developed ulcerative lesions in the pretibial and scrotal regions at a similar age. His mother had two transient ischemic attacks at the age of 18.

One year after his initial cutaneous PAN diagnosis, the patient presented to our center with high fever, severe muscle pain, a draining ulcerative lesion on the ventral penis, and livedo reticularis with necrotic ulcers up to 3×5 cm on both lower extremities and dorsum of the feet. Additionally, he had papulopustular steroid acne and striae due to long-term oral steroid use.

Systemic evaluation revealed no additional organ involvement; however, arthritis was detected in the right knee and ankle. No infectious cause was found for his fever. It was learned that he had discontinued methotrexate and had been taking 16 mg of prednisolone for 7-8 months. Laboratory tests showed ANA positivity (1/80, granular pattern), while ANCA, C3-C4 complement levels, and RF were normal. His IgG level was low (5.6 g/L; lower limit: 7.67 g/L). Acute-phase reactants, such as CRP and ESR, were elevated. Lupus anticoagulant screening and cryoglobulin tests were negative.

A venous Doppler ultrasound performed one year earlier had shown signs of prior thrombosis. However, current CT and venous Doppler examinations revealed no new thrombotic changes other than widespread myofasciitis and osteochondral alterations.

Due to the family history, genetic analysis was performed, revealing a mutation in the CECR1 gene. After a prolonged diagnostic process, the patient was diagnosed with late-onset ADA-2 deficiency mimicking cutaneous PAN.

Conclusion:

This case highlights the importance of family history and aims to raise awareness of genetic diseases, particularly DADA-2, which can mimic cutaneous PAN. This case underscores the critical role of family screening and genetic evaluation in diagnosis and management.

Abstract N°: 2014

Henoch-Schönlein purpura in adults: A series of 8 cases

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Introduction & Objectives:

Henoch-Schönlein purpura (HSP) is an IgA-mediated vasculitis affecting small-caliber vessels. HSP is more common in children than in adults. Through this study, we aim to analyze the specific characteristics of this disease in the adult population.

Materials & Methods:

We conducted a retrospective, single-center, descriptive study including all adult patients diagnosed with HSP between January 2014 and May 2024 in our dermatology department.

Results:

Eight patients were included in the study, comprising five men (62.5%) and three women (37.5%), with a male-to-female ratio of 1.6. The mean age was 53 years (range: 20–74 years). The majority of patients (87.5%) were over 40 years of age, and 37.5% were older than 60. A preceding upper respiratory or otorhinolaryngological infection was documented in two cases (25%). Clinically, all patients exhibited palpable purpura, which was bullous in three cases (37.5%), ecchymotic in two cases (25%), and necrotic in four cases (50%). In one case, purpuric lesions were associated with pustules. The predominant anatomical distribution was acral involvement, observed in all patients (100%), followed by buttock involvement in two cases (25%) and truncal involvement in one case (12.5%). Systemic symptoms, including fever and general health deterioration, were reported in two cases (25%). Histopathological examination of skin biopsies from all patients revealed leukocytoclastic vasculitis characterized by neutrophilic infiltration and erythrocyte extravasation. Direct immunofluorescence demonstrated granular deposits of anti-IgA antibodies along the vascular walls in all cases, with concomitant granular deposits of anti-C3 antibodies in four cases (50%). Articular involvement was observed in six patients (75%) and manifested as inflammatory polyarthralgia affecting the knees and ankles. Gastrointestinal involvement was present in five cases (62.5%), with abdominal pain in three cases (37.5%) and vomiting in two cases (25%). One patient (12.5%) developed peritoneal effusion, confirmed via abdominal computed tomography. Renal involvement was documented in six patients (75%), with peripheral edema in three cases (37.5%). Acute kidney injury was present in three patients (37.5%), and proteinuria was detected in four cases (50%). Laboratory investigations revealed systemic inflammation in five cases (62.5%) and hypoalbuminemia in three cases (37.5%).

Therapeutic management included systemic corticosteroids in six cases (75%) and colchicine in two cases (25%). The clinical course was favorable in all patients.

Conclusion:

HSP is a small-vessel vasculitis characterized in adults by a high prevalence of bullous and necrotic cutaneous lesions, as well as a notably increased frequency of gastrointestinal and renal involvement, as observed in our cohort. Short-term prognosis is primarily influenced by the severity of gastrointestinal complications, whereas long-term outcomes are dictated by the extent of renal involvement. Longitudinal studies have reported that approximately one-third of adult patients progress to end-stage renal disease. However, in our series, all patients exhibited a favorable clinical course without long-term renal impairment.

Abstract N°: 2025

An Incidental Diagnosis: A Case of Nevus Oligemicus

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Introduction & Objectives:

Nevus oligemicus is a rare, localized cutaneous disorder characterized by selective vasoconstriction of the deep dermal vascular plexus relative to the superficial vascular plexus. This case report aims to highlight the clinical features of nevus oligemicus, which was identified incidentally in an asymptomatic patient.

Materials & Methods:

A 61-year-old overweight male patient accompanied his wife, who had complaints of pruritus, to the hospital. The accompanying visitor himself reported mild pruritus. During the examination of the accompanying visitor dermatological evaluation incidentally revealed an irregularly bordered, blanchable erythematous-violaceous and telangiectatic patch in the periumbilical region. The lesion exhibited a cooler temperature compared to the surrounding healthy skin.

The patient's medical history was notable for chronic alcohol consumption. Additionally, he had undergone percutaneous coronary intervention and stent implantation five months prior due to myocardial infarction. His current medications included clopidogrel and acetylsalicylic acid.

Results:

Laboratory tests were within reference ranges. A skin biopsy was obtained from the livid erythematous plaque to further evaluate the lesion. Histopathological analysis revealed mild orthokeratosis and acanthosis in the epidermis, along with mild perivascular lymphocytic infiltration in the dermis. The presence of characteristic localized temperature reduction and blanchable erythematous-violaceous patch, along with the absence of distinctive histopathological findings, led to the clinical evaluation of the case as nevus oligemicus.

Conclusion:

Nevus oligemicus is classified as a functional nevus and is characterized by hypothermia and violaceous-erythematous patch lesions with an uncertain pathogenesis. It is hypothesized that alterations in adrenergic activity may lead to increased sympathetic tone, resulting in vasoconstriction of the deep dermal vascular plexus, which in turn causes localized hypothermia within the lesion. As a compensatory mechanism, secondary vasodilation occurs in the superficial dermal vascular plexus, leading to the formation of an erythematous telangiectatic patch.

Clinically, nevus oligemicus is typically asymptomatic and manifests as an acquired, stable, irregularly bordered, blanchable erythematous-violaceous patch. In some cases, cyanotic, pale, and telangiectatic areas may also be observed. Ischemic changes are absent in the adjacent skin.

One of the defining features of nevus oligemicus is a localized decrease in temperature, with the affected area measuring at least 2°C lower than the surrounding healthy skin. This can be objectively confirmed using a contact thermometer.

The diagnosis of nevus oligemicus is primarily clinical. Histopathological examination typically demonstrates dilated capillary vessels in the papillary dermis, without a significant alteration in overall vessel density. Importantly, skin biopsy is not considered essential for diagnosis, as the most reliable diagnostic marker remains temperature measurement of the lesion.

Currently, an established treatment for nevus oligemicus is lacking. Longitudinal follow-up of previously documented cases has shown that the lesions remain stable over time, without any progression in morphology or size and without the development of new lesions.

Abstract N°: 2237

Clinical and therapeutic experience of Klippel-Trenaunay syndrome: Series of 6 patients

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Introduction & Objectives:

Klippel-Trenaunay syndrome (KTS) is a rare complex congenital vascular malformation, the diagnosis of which is based on at least two features of the triad associating limb plane angioma, venous malformations and soft tissue and/or bone hypertrophy, with or without lymphatic malformation.

The aim of this study is to identify the main demographic features of Klippel-Trenaunay syndrome, its clinical manifestations and therapeutic management.

Materials & Methods:

This is a retrospective, descriptive study carried out in the dermatology department over a period of 9 years, including all patients with Klippel-Trenaunay syndrome. Data were collected from medical records and quality of life was assessed using the DLQI score.

Results:

A total of 6 patients were included. The mean age of patients at diagnosis was 22.3 +/- 16 years, with extremes ranging from 3 to 42 years. Females predominated, with an F/H sex ratio of 2.

A history of iron-deficiency anemia was noted in 3 patients, congenital Raynaud's syndrome of the homolateral upper and lower limb, recurrent hematuria, several episodes of infectious dermo-hypodermatitis, a history of surgically drained septic arthritis and a history of thrombophlebitis of the lower limb in one patient each.

All cases were congenital and progressive. The reason for consultation was pain or discomfort in all patients, edema in 5 patients, functional impotence in 2 patients, varicose veins, bleeding, ulcer and finally pruritus in one patient each.

In terms of clinical presentation, 5 patients presented with the classic SKT triad. Another patient presented with a combination of venous malformation, lymphatic malformation and limb hypertrophy.

Involvement was unilateral in 5 patients, mainly involving the MID in 3 cases. Bilateral involvement of the MID associated with involvement of the trunk, upper limb and neck was noted in a single patient.

The patients' mean DLQI score was 16.6 before the start of treatment, reflecting a significant effect on quality of life, compared with 9.7 after treatment in treated patients, reflecting a moderate effect on quality of life.

Pain was the most frequently observed complication (83.3%), followed by recurrent dermohypodermatitis (50%), bleeding (16.6%), thrombophlebitis (16.6%), venous ulcer (16.6%) and lameness (16.6%).

In terms of management, two patients received compression bandaging of the affected lower limb. Treatment with Everolimus was initiated in 3 patients (50%), who showed a marked improvement in symptoms, with a reduction in edema and an improvement in discomfort. One patient was offered treatment with Sirolimus, but declined due to lack of funds. Two patients were lost to follow-up.

Conclusion:

Klippel-Trenaunay syndrome is a rare disease with little research available. Studies show divergent results on its epidemiology, clinical aspects and treatments. Management of symptoms and associated complications is crucial, and long-term follow-up is often necessary to adapt treatments to individual patient needs.

More studies are therefore imperative to improve the quality of life of patients with this rare disorder.

Abstract N°: 2251

Phakomatosis Pigmentovascularis Type II: A Rare Case of an Infant with Extensive Dermal Melanocytosis

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Introduction & Objectives:

Phakomatosis pigmentovascularis (PPV) is a rare disorder characterized by vascular and pigmentary anomalies. It is classified into five subtypes, with Type IIa involving only cutaneous findings and Type IIb including systemic manifestations. While GNA11 and GNAQ mutations have been implicated in its pathogenesis, systemic involvement remains variable. Early recognition is crucial due to associations with Sturge-Weber syndrome (SWS) and other neurovascular conditions. We present an infant with extensive dermal melanocytosis, nevus anemicus, and a port wine stain, raising concern for PPV Type IIa and highlighting the importance of early diagnosis and multidisciplinary care.

Materials & Methods:

A full-term female neonate presented at 21 days old with slate-gray to blue patches on the back, left arm, buttocks, and thigh, consistent with dermal melanocytosis. Additional hypopigmented patches (nevus anemicus) were noted on the back and left cheek, alongside a pink capillary malformation (port wine stain) on the occipital scalp and upper back.

A comprehensive workup was initiated to confirm the diagnosis and assess for systemic involvement. Genetic testing for GNA11 and GNAQ mutations was performed to evaluate potential molecular contributions to the disease. An ophthalmologic assessment was conducted to screen for glaucoma and ocular melanocytosis, while a neurologic evaluation assessed for seizures and developmental abnormalities. Lysosomal enzyme screening and a skeletal survey were completed to rule out underlying metabolic or structural abnormalities. The patient was followed through serial evaluations to monitor disease progression, systemic involvement, and potential treatment interventions.

Results:

Genetic testing for PPV-associated mutations was negative, though skin biopsy was deferred. Ophthalmologic and neurologic exams were normal, with no seizures, glaucoma, or developmental delays at the three-month follow-up. Given the lack of systemic involvement, a diagnosis of PPV Type IIa was made.

At follow-up, the patient remained stable, meeting all developmental milestones, and no systemic abnormalities emerged. Laser therapy for vascular and pigmentary lesions was planned for early childhood to minimize psychosocial impact and treatment burden.

Conclusion:

This case highlights the importance of early recognition and thorough evaluation of suspected PPV. While PPV Type IIa typically follows a benign course, ongoing surveillance is essential as systemic manifestations may emerge over time. The presence of a port wine stain necessitates monitoring for SWS-related complications.

Dermatologists play a key role in recognizing PPV, ensuring early referrals for systemic evaluation, and guiding treatment strategies. Laser therapy initiated in childhood can optimize cosmetic outcomes and quality of life. Further research into genetic underpinnings and long-term outcomes is needed to refine diagnostic and management approaches.

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Abstract N°: 2263

Lymphangioma circumscriptum successfully treated with radiofrequency ablation and pulse dye laser on both halves: A case report

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Introduction & Objectives:

Lymphangioma circumscriptum (LC) is the most common type of lymphatic malformation consisting of enlarged lymphatic channels surrounded by lymphatic endothelium. They tend to involve proximal extremities, trunk and axilla. They are usually superficial lesions, but they can also extend into the subcutaneous soft tissue.

Typical clinical findings are multiple grouped or scattered, translucent or slightly hemorrhagic vesicles that can form plaques and resemble frog spawn.

Intermittent swelling, hemorrhage, leakage of lymphatic fluid, pain and tendency for infection are potential complications.

Various treatment options for LC include surgical excision, sclerotherapy, ablative lasers, pulsed dye laser (PDL), cryotherapy and radiofrequency (RF) ablation. However, LC tends to recur after the treatment. Clinicians should evaluate lasers and ablative methods in appropriate patients before considering surgical treatment because of its poor cosmetic outcomes.

Here, we present a successful split-lesion treatment of an LC on the right thigh with RF ablation and 585 nm PDL and has not recurred on both halves in five years follow-up.

Materials & Methods:

A 15-year-old, otherwise healthy female patient, presented to our dermatology outpatient clinic with a congenitally present group of erythematous- violaceous papules and translucent -hemorrhagic vesicles on the right thigh. The lesion was 15 x 15 cm in diameter, extending from the anterior aspect of the limb medially to the posterior.

MRI was performed to detect the deep tissue involvement. The MRI revealed a lesion located within the subcutaneous adipose tissue, exhibiting T1- hypointense and T2-hyperintense signal characteristics, consistent with a hemangioma.

Subsequently, a punch biopsy was performed to confirm the diagnosis. Histopathological examination in correlation with the clinical findings led us to the diagnosis of LC. RF ablation was applied to one half of the lesion in cut and coagulate mode with wire loop electrode, while a 585 nm wavelength PDL (7 J/cm2) was performed on the other half. 2 sessions of RF ablation and PDL combination at 4-week intervals led to the regression of the lesions. The results of the RF and PDL treatment halves of the lesion were similar.

Five years later, the patient presented to our department again with recurrence. The third session of the same procedure - RF ablation and PDL combination-was performed in July 2019. Lesions were regressed completely in the first month follow-up. No recurrence has occurred since 2019, and the patient is still under surveillance.

Results:

In our patient, PDL and RF were equally safe and effective and have remained recurrence-free to date.

Conclusion:

Treatment of LC is challenging because of the persistent nature of the disease. PDLs cause selective vascular damage without harming the surrounding tissues and used for treating vascular lesions. They can be also used for treating lymphangioma circumscriptum, especially for superficial lymphatic malformations that contain blood. RF ablation is another nonsurgical treatment option which also has safe and effective results. It provides almost complete clinical ablation with coagulation of lesional and perilesional skin causing fibrosis of perivascular lymphatics.

Non-surgical treatment methods such as PDL and RF ablation in cases of LC can be applied alone or consecutively.

Abstract N°: 2406

A rare case of Schamberg disease in dark phototype

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Introduction & Objectives:

Schamberg disease is the prototype of pigmented purpuric dermatoses. The etiology of this rare condition is still unclear. It's a chronic, progressive and relapsing condition that appears in the 4th-5th decade, mainly in the lower extremities. The diagnosis relies on the clinical and dermoscopic examination with a histopathological confirmation.

The objective of this case is to describe the clinical and the dermoscopic features of Schamberg disease and to discuss this case through a review of the literature.

Materials & Methods:

We report an uncommon case of a 65 year old woman presenting a Schamberg disease of the lower extremities with histopathological confirmation.

Results:

A 65-year-old female patient with no previous medication, no contact allergy, and no previous skin infection, developed two years ago asymptomatic lesions of the right leg initially that progressed 2 months later to the left leg. The lesions were evolving by relapses and partial remissions. Physical examination revealed red pinhead-sized petechiae with orange-brown macules on the lower extremities. We didn't find any sign of chronic venous insufficiency, such as swelling, varicose veins, or venous ulcers. Dermoscopy showed comma-like and linear vessels, red dots, globules, and circles with a coppery brown background. Histopathological examination found a superficial perivascular lymphocytic and histiocytic infiltration with marked hemosiderin deposition, dilated superficial vessels. Doppler ultrasound of the lower limbs was normal. We proposed UVB phototherapy and topical steroids, the treatment is still going on.

Conclusion:

Schamberg disease is an acquired chronic disease with an unkown etiology and a non-standardized treatment. More studies need to be done in order to understand this rare condition.

Abstract N°: 2422

Preoperative Ultrasound Preventing Misdiagnosis: A Saphenous Vein Aneurysm Masquerading as a Lipoma

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Introduction & Objectives:

Lipomas are common benign soft tissue tumors, often diagnosed clinically without imaging. However, in atypical presentations, clinical examination alone may lead to misdiagnosis and unnecessary surgical intervention. This case highlights the importance of preoperative ultrasound in differentiating vascular anomalies, such as a saphenous vein aneurysm, from lipomas, thereby preventing potential surgical complications.

Materials & Methods:

A 52-year-old patient presented with a slow-growing, painless subcutaneous mass in the lower limb, clinically resembling a lipoma. To ensure accurate diagnosis, an ultrasound evaluation was performed, followed by Doppler imaging for further vascular assessment.

Results:

Ultrasound revealed a well-defined, compressible, hypoechoic lesion with vascular flow, raising suspicion of a vascular anomaly rather than a lipoma. Doppler ultrasound confirmed the diagnosis of a saphenous vein aneurysm. As a result, surgical excision was avoided, preventing potential complications such as hemorrhage or thrombosis.

Conclusion:

This case underscores the critical role of preoperative ultrasound in evaluating soft tissue masses, particularly in atypical presentations. Routine ultrasound assessment before presumed lipoma excision can help prevent misdiagnosis and unnecessary surgical procedures, especially when vascular anomalies mimic lipomatous structures.

Abstract N°: 2437

Angiolymphoid hyperplasia with eosinophilia resistant to treatment: case report

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Introduction & Objectives:

Angiolymphoid hyperplasia with eosinophilia is a rare, benign, chronic vasoproliferative disease that affects individuals between 20 and 50 years of age with a slight predisposition in the female sex. It is characterized by skin lesions of the type of nodules or papules that can be single or multiple and whose color varies between red or brown to a violet appearance. The characteristic site of appearance is the head and neck area, with special involvement of the periauricular area and the scalp, although there are also reports of extracutaneous locations. Its pathophysiology involves an interaction between angiogenesis, immune deregulation and both genetic and environmental factors.

Materials & Methods:

We present the case of a 51-year-old male patient with no history of chronic degenerative diseases, who presented with a dermatosis in the left retroauricular area characterized by three nodular and erythematous neoformations of 5 mm to 2.5 cm in diameter. He reported that his current condition began 6 months prior to his consultation with the appearance of a slow-growing, slightly painful and non-pruritic tumor, so he went to a physician who provided treatment with an unspecified emollient and after the lack of improvement, an external biopsy was taken which reported a process compatible with Kaposi's sarcoma in its nodular phase, so he was sent to our Dermatology service for evaluation.

During his approach it was concluded that it was an angioproliferative process, however, complementary studies were requested which did not present alterations and the viral panel was non-reactive. A new punch biopsy was scheduled, which concluded that the lesion was angiolymphoid hyperplasia with eosinophilia without evidence of malignancy, and the histopathological appearance was not related to Kaposi's sarcoma. A simple and contrast-enhanced CT scan of the skull and neck identified the lesion with a diameter of 41x17 mm in its major axes with subcutaneous nodules that did not show any increase or change in density with the contrast medium.

The patient was treated with 5 sessions of intralesional corticosteroid and 5 sessions of cryotherapy without obtaining improvement or reduction of the lesion, so he is awaiting surgical excision due to failure of conservative treatment.

Results:

Conclusion:

The treatment of choice for angiolymphoid hyperplasia with eosinophilia is surgical excision with free margins. However, there are case reports that have shown improvement and even remission of lesions with the combined use or monotherapy of intralesional corticosteroids and cryotherapy. In the case presented, this was not the case despite dual therapy, which is explained by the fact that it is a chronic and recurrent pathology even with surgical excision in 30-50% of cases.