

Abstract N°: 162**Atypical presentation of angiolymphoid hyperplasia with eosinophilia - a rare entity**Rajkiran Takharya^{*1}¹Manipal Tata Medical College Road, Dermatology, Jamshedpur, India

Introduction: Angiolymphoid hyperplasia with eosinophilia (ALHE), an uncommon benign vascular proliferation, was initially thought to be a late stage of Kimura's disease but is now considered a separate entity. ALHE may coexist with Kimura's disease. We report a case of ALHE with anemia (Hemoglobin = 10.6gm%) and diabetes mellitus (Blood Sugar = 178mg%). These associations have not been reported previously. Our patient also had lymphadenopathy, a feature of Kimura's disease.

Case Report: A 34-year-old male woodcutter by profession presented with itchy multiple skin colored to reddish raised lesions present over lower lip and chin area for 1.5 years. Examination revealed multiple soft to firm, smooth and shiny, erythematous papules and nodules, few are hemorrhagic bleed on touch, varying in size from 0.5cm to 1.5cm present on lower lip and chin with few excoriations. There was an associated regional lymphadenopathy. Known case of Diabetes Mellitus for 6 years. Laboratory reports: Hemoglobin=10.6gm%, Eosinophil count - 17.3%, Blood Sugar 178 mg% , HBA1c 6.2%, ESR 132 mm/Hr, HCV, HbsAg, VDRL all came Negative. PAS stain negative. Histopathology revealed increased thick-walled capillaries and venules in upper dermis surrounded by an infiltrate of lymphocytes and eosinophils with mild edema and proliferating fibroblasts. Patient is admitted under plastic surgery for surgical excision of the lesions.

Discussion: ALHE was first described in 1969 by Wells and Whimster and it was considered that ALHE to be a late stage of Kimura's disease. It is now generally accepted that these are two separate entities. It is an uncommon benign but potentially disfiguring vascular proliferation. It has a particular predilection for head and neck area, especially for the ears. The condition presents with erythematous or skin-colored dome-shaped dermal papules or nodules, often associated with spontaneous bleeding, pain, pulsation, pruritus, and growth. The frequent presence of mural damage or rupture in intralesional large vessels of ALHE has suggested a role of trauma or arteriovenous shunting in its pathogenesis. Histologically, it appears as an angiomatous lesion with abundant proliferating blood vessels lined by prominent endothelial cells with a "histiocytoid" or "epitheloid" appearance and vacuolated cytoplasm. There is diffuse infiltration by lymphocytes and eosinophils. Treatment of ALHE is dictated in part by the number, location, and size of the lesions. Rare instances of spontaneous regression have been reported. Patients with solitary or a few small lesions may benefit from excision or Moh's surgery. About one-third of lesions recur after excision. Other treatment modalities used successfully include systemic and intralesional steroid administration, interferon therapy, cryotherapy, laser therapy, and topical application of tacrolimus.

Conclusion: Angiolymphoid Hyperplasia with Eosinophilia rare entity with uncontrolled sugar levels and anemia, along with features overlapping with Kimura's disease has not been reported previously.

Abstract N°: 313**Marginal Zone B Cell Lymphoma and Non-HCV Associated Type II Cryoglobulinemic Vasculitis: A Rare Presentation**

Ayris Öztürk¹, Batuhan Tural¹, Irmak Caprak¹, Banu Yaman², Nilay Duman², Can Ceylan¹

¹Ege University Medical Faculty Hospital, Dermatology and Venerology, Izmir, Türkiye, ²Ege University Medical Faculty Hospital, Pathology, Izmir, Türkiye

Introduction & Objectives:

Cryoglobulins are immunoglobulins (Ig) or antibodies composed of Ig and complement components that precipitate below 37°C and dissolve upon rewarming. Cryoglobulinemia (CG) usually leads to a systemic inflammatory syndrome characterized by malaise, arthralgia, vasculitis, ulcers, neuropathy, and/or glomerulonephritis. In this case report, we present a 72-year-old HCV-seronegative 72-year-old woman with Marginal Zone B Cell Lymphoma and Type II Cryoglobulinemic Vasculitis (CV).

Materials & Methods:

The patient presented to our clinic with complaints of abdominal pain, fatigue, weight loss for 2 years, and painful sores that started on the legs 4 months ago and spread to both lower extremities. Comorbid hypertension, type 2 diabetes mellitus, polyneuropathy, and hypothyroidism were present. She was taking atorvastatin, methylprednisolone (MP) 15 mg, levothyroxine, perindopril arginine, indapamide, amlodipine, vildagliptin, metformin, insulin, colchicine, pregabalin, alpha lipoic acid. Physical examination revealed infected ulcers with necrotic crusts on both lower extremities.

Results:

Laboratory tests revealed elevated C reactive protein, sedimentation, cryoglobulin, rheumatoid factor, low complement 4, and normal complement 3. Normochrome normocytic anemia, lymphopenia, and thrombocytosis were present. Viral serology was compatible with previous hepatitis B (HBV). Hepatitis C virus (HCV) and human immunodeficiency virus antibodies were negative. Antinuclear antibody, anti-histone, anti-polymyositis-scleroderma antibody, and cold agglutinin were positive. IgM was high, IgA, and IgG was normal. Histopathology was consistent with CV (Figure 1).

The patient was diagnosed as type II CG due to a history of HBV, laboratory and HP findings, concomitant sensorial neuropathy, and leg ulcers and was started on MP 500 mg for 3 days and then 60 mg daily. Biopsy of suspicious lymph nodes found in malignancy screening was compatible with nodal marginal zone lymphoma (NMZL) with intense plasmacytic differentiation and bone marrow aspiration biopsy was consistent with B-cell lymphoma infiltration. No pathology was detected in other malignancy scans. A rituximab-bendamustine regimen for NMZL was planned. Entecavir treatment was started for prophylaxis of hepatitis B reactivation. Significant clinical response was obtained with treatment.

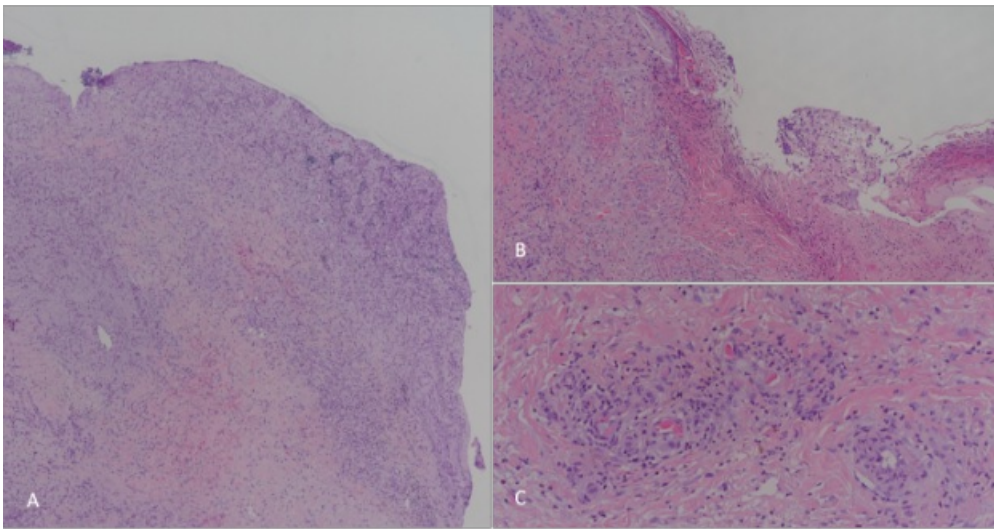


Figure 1: Ulcer, inflammatory granulation tissue, mixed inflammatory cell infiltration (H&E, x40, x100, x200, respectively)

Conclusion:

Type I CG consists of monoclonal IgM or IgG; Type II consists of monoclonal IgM/polyclonal IgG immune complexes; Type III consists of polyclonal IgM and IgG. In Type II and III Mixed Cryoglobulinemia (MC), the IgM fraction has RF activity. Type I CG is usually associated with monoclonal gammopathies, whereas MC is associated with infections (HCV, HBV), autoimmune diseases, B-cell lymphomas, or solid neoplasms.

Typical laboratory findings of MC, which usually presents with the triad of purpura, malaise, and arthralgia, are elevated RF, hypocomplementemia, and hypergammaglobulinemia. CV is difficult to manage due to severe organ involvement and high mortality. Treatment of the underlying malignancy, glucocorticoids, cyclophosphamide, rituximab, and plasmapheresis are among the treatment options. This case report emphasizes this rare association between HCV-seronegative B-cell lymphoma and CV and aims to raise clinician awareness.



Abstract N°: 584**A Case of Superficial Spreading Capillary Hemangioma**Hyunjoon Chang*¹, Yoon Seob Kim¹, Kyung Ho Lee¹, Chul Jong Park¹¹Bucheon St. Mary's Hospital, Department of Dermatology, Bucheon-si, Korea, Rep. of South**Introduction & Objectives:**

Superficial spreading capillary hemangioma is a benign skin tumor that is characterized by lobular proliferation of capillaries confined to the papillary dermis. Mihara et al. first described this rare histological subtype of acquired hemangioma in 1986. It typically presents as asymptomatic red papules mainly on the plantar surface of the feet, and only a few cases have been reported in the dermatology literature. Our objective is to report a case of superficial spreading capillary hemangioma on the plantar surface of the left foot and highlight the clinical presentation of this rare disease.

Materials & Methods:

We conducted a retrospective analysis of the medical records and clinical photographs of a patient who presented with superficial spreading capillary hemangioma on the plantar surface of the left foot, confirmed by histopathologic examination.

Results:

A 33-year-old male patient presented with multiple asymptomatic reddish patches on the left sole, which had occurred without trauma one month prior. Histopathological examination revealed hyperkeratosis, acanthosis, and thin-walled, dilated capillary vessels grouped in small lobules confined to the papillary dermis, separated by extended epidermal ridges. Immunostaining for CD31 showed positive endothelial cells. The diagnosis of superficial spreading capillary hemangioma was made based on these findings. Although vascular laser treatment was recommended, the patient declined.

Conclusion:

Herein, we presented a case of superficial diffuse capillary hemangioma on the plantar surface of the foot, adding to the limited literature available on this rare disease. The case also highlights that superficial spreading capillary hemangioma can present as patch lesions, underscoring the importance of considering this possibility in the differential diagnosis of reddish patches on the feet.



Abstract N°: 1070**thrombosed cutaneous cavernous hemangioma in a pregnant woman**

Naomi Carrara Matsuura¹, Cibele Toledo², André Cesar Antiori Freire Pessanha¹

¹UMC - Universidade de Mogi das Cruzes, Dermatology, Brazil,²UMC - Universidade de Mogi das Cruzes, Brazil

Introduction & Objectives: Cavernous hemangiomas are benign congenital proliferations of blood vessels, less frequent than other hemangiomas, differentiated by the formation of large vascular channels involving deep structures. Mainly affect the skin and mucous membranes. It is more common to appear in childhood, but may also occur in adulthood. Small hemangiomas may appear during pregnancy, and pre-existing ones may increase dramatically and eventually regress after birth. It is believed that estrogen and progesterone levels are related to its formation, since they are increased during pregnancy, contributing to its rapid proliferation.

We report a case of thrombosed cavernous hemangioma of unusual clinical form in a pregnant patient, which presented accelerated growth during pregnancy.

Materials & Methods: Case Report

Results: A 27-year-old female patient, 28 weeks pregnant, complains of a lesion on the anterior region of the left thigh that started 1 year ago and has grown rapidly in the last 2 months. The initial lesion was a reddish papule with a smooth surface that eventually bled spontaneously. During the third trimester of pregnancy the lesion grew rapidly, evolving into a purplish tumor with crusts on its surface. The patient reported pain throughout the limb and spontaneous bleeding from the lesion.

The patient had no comorbidities, was taking ferrous sulfate and folic acid due to pregnancy, and had no personal or family history of oncology. The diagnostic hypotheses of pyogenic granuloma, amelanotic melanoma and squamous cell carcinoma were made.

Excisional biopsy of the lesion was performed, the histopathological result was compatible with the diagnosis of ulcerated cavernous hemangioma with extensive thrombosis.

Conclusion: During pregnancy many vascular changes occur in response to estrogen and other growth factors. Varicosities affect more than 40% of pregnant women, being more common in the legs and anal region, due to increased blood volume and venous pressure in the femoral and pelvic vessels.

In the reported case there was an accelerated vascular proliferation of a hemangioma during the second trimester of pregnancy, occurring besides its expansion, thrombosis, ulceration and symptoms such as pain and bleeding. The exuberant clinical presentation and the way it evolved make us think of several diagnostic possibilities, including malignancy, which justifies the urgency to remove the lesion.

Cavernous hemangiomas are usually oligosymptomatic, but can present severe symptoms due to the possible occurrence of intravascular thrombosis or rupture of channels leading to intralesional bleeding.

Although the appearance or increase in size of the vascular lesions caused by hormonal changes during pregnancy is expected, and even knowing that they may spontaneously regress in the postpartum period, observation and attention should be maintained for the possible complications that may occur.



Abstract N°: 1071**Severe thromboangiitis obliterans in a 35-year-old young man: tobacco is not the only culprit**

Mohand Ouali Ait Si Ali¹, Lamia Chaloul¹, Assya Djeridane¹

¹Dermatology, Algiers, Algeria

Introduction & Objectives:

Thromboangiitis obliterans (Léo Buerger's disease) is an inflammatory disease of the small and medium vessels affecting the young subject. Regular cannabis use is as responsible as active smoking for the development and aggravation of lesions.

We report a case of severe thromboangiitis obliterans in a young cannabis user.

Materials & Methods:

A 35-year-old patient with no particular history, smoker (40 packs/year) and cannabis user (5 to 6 joints/week) for 10 years, was reported to us for necrosis of the toes with notion of intermittent claudication. Clinically, the necrosis affected the second toe of both feet and the pulp of the left big toe. The popliteal hems were weakly perceived with the abolition of the pedal hems. A blood pressure asymmetry is found with left carotid murmur.

Results:

Biologically no abnormalities apart from a modified lipid balance, namely a triglyceridemia at 2.39g/l and a total cholesterol at 2.50g/l.

A distal occlusion with a decrease in the flow of the anterior and pedal tibial arteries is objectified on arterial echo-Doppler of the lower limbs, confirmed by CT angiography with an amputation of the distal third of the anterior tibial and a bilaterally small aspect of the posterior tibial arteries.

The patient had received antiplatelet drugs, vasodilators and statins. In view of the non-improvement and the appearance of intense pain resistant to analgesics, the amputation of the toes was inevitable.

Conclusion:

Faced with severe arteritis in a young subject, cannabis consumption should be systematically investigated. In most cases, stopping toxic substances (tobacco, cannabis) and medical treatment (antiplatelet agents and vasodilators) make it possible to obtain healing of the lesions without considering amputation. In our patient an amputation was inevitable.



Abstract N°: 1092**iatrogenic arterio venous fistula in the forearm in a seven month old infant**

Aïcha Salhi¹, Yamoun Mohamed², Foughali Mohamed³, Djeridane Assya², Enjolras Odile⁴, Aitbenamar Abdelhamid²

¹University of Algiers, dermatology, Alger Centre, Algeria, ²University of Algiers, Alger Centre, Algeria, ³University of Algiers, cardiology, Alger Centre, Algeria, ⁴Lariboisière Hospital AP-HP, radiology, Paris, France

Introduction: Pediatric arterio venous fistula AVF can be congenital or acquired. These last can be induced by trauma but mostly they are iatrogenic complications of vascular access. Incidence of iatrogenic AVF after cardiac catheterization in children is 0.3 per cent. Clinical manifestations associated with AVF are non specific, thus these cases can constitute a diagnostic challenge. Doppler ultrasound is the diagnostic tool of choice; it can as well evaluate the severity by calculating the shunt volume. When AVF are deeply situated or associated with other vascular complications, CT, MR or angiography can be required. Surgery is the treatment of choice. We report the case of a 7 month old infant with iatrogenic AVF in the forearm.

Observation: A male infant 7 months old presented to our department for swelling of his right forearm. He has been hospitalized as he was newborn for prematurity and low birth weight which required multiple veni punctures due to blood sampling. After a one month stay in the incubator, the weight normalized and the infant was discharged. The mother noticed from the third month a swelling and color change of the right hand and forearm. Physical examination was normal concerning weight, tall, head circumference and psychomotor development. The distal two thirds of the right forearm were oedematous with violaceous discoloration. Carefull examination revealed numerous dilated capillaries on the wrist territory. Palpation was not painful. Comparison of the humeral pulses showed an intense pulse by the right side but there was no thrill and no audible bruit. Doppler ultrasound DUS revealed an arterio venous fistula AVF in the right cubital fossa due to a communication between the humeral artery and the humeral vein. The flow rate of the AVF was evaluated at 400ml/mn. Cardiac examinations with electrocardiogram and echocardiogram ultrasound were normal. The patient was sent to the vascular surgery department for surgical repair of the AVF. As the AVF was due to a very narrow communication, there was no operative indication. The AVF was treated with ultrasound guided compression. Doppler ultrasound control one year later was normal. A seven years follow up showed no relapse at DUS but the right forearm remained slightly larger with no functional impairment.

Conclusion: Reports of iatrogenic AVF in children are rare and they are mostly due to vascular access chiefly repeated vascular access. Scalp AVF have been reported in hemophiliac children because of the easy access of scalp veins. AVFs have been reported in antecubital and inguinal levels as well. Large adult series have shown that 1/3 of AVFs close spontaneously within one year from diagnosis. The treatment of pediatric AVFs is usually surgical because stent can lead to thrombosis and thus to chronic ischemia and extremity length discrepancy. In our case we show that compression is a safe and effective nonsurgical alternative technique to treat AVF complicating arterial punctures.



Abstract N°: 1140

Clinico-pathological and Immunofluorescence study of cutaneous small vasculitis : a hospital based cross-sectional study.

Ayushi Khandelwal¹

¹M S Skin centre, Bengaluru, India

Introduction & Objectives:

Cutaneous vasculitis represents a group of disorders characterized by inflammation of blood vessel wall and it may involve any organ system. Skin biopsy is considered gold standard for the diagnosis and direct immunofluorescence (DIF) aids in categorization. There are very few studies from India which correlates clinical, histopathological as well as DIF findings in patients with cutaneous small vessel vasculitis (CSVV).

Materials & Methods:

A hospital based cross-sectional study was designed. All clinically diagnosed cases of CSVV were included and relevant history taken. Complete hemogram, urine microscopy and two skin biopsy specimens for histopathology and DIF were taken from all patients. Clinical, histopathological and immunofluorescence findings were analyzed.

Results:

Out of 52 patients, 36 (70%) were male (M:F= 4:1). Adolescents and adults were the common sufferers (n= 40,91%). Upper respiratory tract infection (n=11, 20.4%) was the commonest precipitating factor followed by NSAID (n=8,15.3%). Palpable purpura was the commonest cutaneous manifestation (n=52 ,100%) and extracutaneous involvement was noted in 36(70%) patients i.e., joint pain, abdominal pain and hematuria. Joint pain was the commonest systemic complaint (n= 25,52.2%). On histopathology, the commonest pattern seen was leukocytoclasia and extravasation of RBCs. DIF showed overall positivity (n=49, 94.2%) with IgA (n=30, 50.8%), C3 (n=49, 94.2%), and two patients had both IgA and IgM positivity. DIF findings were suggestive of Henoch Schoenlein purpura (HSP).

Conclusion:

Majority of the patients had HSP as per DIF study. In contradiction to the current belief adolescents and adults were the common sufferers in this series.



Abstract N°: 1415**A giant vulvar lymphangiomatous tumor after radiotherapy**

Lucian G. Scurtu^{*1}, Mariana Costache², Denisa Vladulescu¹, Olga Simionescu¹

¹Carol Davila University of Medicine and Pharmacy, Colentina Clinical Hospital, Dermatology I Department, București, Romania, ²Carol Davila University of Medicine and Pharmacy, Bucharest Emergency University Hospital, Pathology Department, București, Romania

Introduction & Objectives:

Radiotherapy may generate a broad spectrum of cutaneous vascular proliferations, varying from benign tumors to malignant, high-grade angiosarcomas. Benign lymphangiomatous papules and plaques (BLAP) after radiotherapy represents a rare variant of benign tumor, most frequently encountered after breast cancer external radiation therapy. The lesions usually arise as asymptomatic vesicles or papules that may coalesce into plaques and have a median latency interval of 6 years following breast irradiation.

Materials & Methods:

A 63-year-old patient presented with firm, asymptomatic, erythematous-violaceous papules on the vulva, evolving over the last three years. The lesions tended to coalesce into a large plaque and displayed a cauliflower-like tumoral aspect. The lymph nodes were not palpable. The patient underwent an incomplete tumoral electrocauterization one year before the presentation, but it recurred and rapidly increased, particularly over the last three months. She had a history of cervical cancer twenty-three years ago and underwent a total hysterectomy with bilateral adnexectomy, completed with postoperative radiotherapy. Her medical history also included a transurethral resection of a benign bladder tumor, two months prior to presentation, stage III hydronephrosis, and type II diabetes mellitus. With the initial clinical impression of a Buschke-Löwenstein tumor, angiosarcoma, or a giant Bowenoid papulosis, an 8-mm punch biopsy was performed.

Results:

Histological examination revealed dermal fibrosis and a proliferation of lymphatic vessels, some with an enlarged lumen localized in the papillary dermis, with prominent endothelial cells, hyperaemic capillaries, and a scarce lymphohistiocytic inflammatory infiltrate. Immunohistochemistry was strongly positive for CD-34 and CD-31, together with D2-40, a marker distinctive to lymphatic vessels, and was negative for human herpes virus 8 and Ki67. Based on these findings and on the clinical appearance, she was diagnosed with an infrequent variant of vulvar BLAP after radiotherapy. The patient was referred to the plastic surgery department for surgical excision of the lesions.

Conclusion:

In 1999, Diaz-Cascajo *et al* proposed the term "benign lymphangiomatous papules following radiotherapy" for benign tumors with a lymphatic origin, that arise after radiotherapy. The term "benign lymphangiomatous papules or plaques following radiotherapy" was subsequently proposed, and it better suits this clinical variant. This rare vulvar presentation posed several challenges, including the unusual localization, the clinical evolution, and the long latency time following radiotherapy. Standardized classifications of these lymphangiomatous tumors after radiotherapy are warranted, given the continuously increasing number of radiotherapeutic procedures worldwide.

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Abstract N°: 1518**Giant facial AVM controlled by embolotherapy made feasible thanks to thalidomide pretreatment**

Aïcha Salhi¹, Odile Enjolras², Abdenour Cheiffa^{1, 3}, Audrey Lentin⁴, Alexander Gerdom⁴, Laurence Boon⁴, Mountassir Ourrad³, Assya Djeridane³, Abdelhamid Aitbenamar³

¹medicine university of algiers, dermatology, Algiers, Algeria, ²Lariboisière Hospital AP-HP, Paris, France,

³University of Algiers, Alger Centre, Algeria, ⁴Cliniques universitaires Saint-Luc (UCLouvain), Bruxelles, Belgium

Introduction: Arterio venous malformation AVM is a vascular malformation due to direct communication between dysplastic arteries and veins creating a nidus which is made of arterial feeders and enlarged draining veins connecting through arterio venous fistulas AVFs. AVMs can be congenital, some are discovered at puberty or in adulthood. Superficial AVMs usually appear as warm angiomatic blumps with pulsations, thrill and bruit at auscultation. Head and neck is most common location. Ultrasonography with color doppler UCD is the best diagnosis tool showing low resistance high velocity arterial flow, CT angiography, magnetic resonance angiography MRA and arteriography, describe the nidus and the involved vessels. Complications can occur: pain, ulceration, bleeding, and cardiac failure with a fatal evolution. Resorting to partial excision, ligation, or endovascular occlusion of the feeding artery aggravate AVMs. Therefore embolization with various agents followed by wide surgical resection and reconstruction is recommended to manage AVMs. Molecules with anti angiogenic properties were recently used in difficult cases. We report the case of a giant facial AVM that could only be embolized after thalidomide treatment.**

Observation: A 68 years old man with functional renal failure presented an enormous angiomatic mass of the cheek and upper lip by the left side. The skin covering the masse was red with tortuous draining veins all around the mass. He suffered from pain and repeated bleeding from mucosa. The lesion expands with increased red color, warmth, evident bruit, pulsation and thrill after a facial trauma that happened ten years ago. UCD showed high velocity of the left, facial and internal maxillary arteries with aneurysm. The draining veins were three times larger than contralateral ones. Angiogram revealed 2 AVFs, in the upper lip and in the left cheek. Treatment with rapamycin was tried for 3 months without any improvement. The patient was transferred to St Luc hospital in Brussels where an embolization attempt failed due to the very high flow of the AVF. To reduce the AVM flow, thalidomide started at 100mg/d for 2 months then reduced at 50 mg/d for 1 month. The second attempt of embolization with coils and ethanol was successful. Three embolotherapy sessions resulted in a complete control of the AVM. Surgery was ruled out because of renal failure. A follow up for 7 years found a markedly reduced mass with no more bleeding.

Conclusion: The management of complex bleeding AVMs is very challenging. The failure of endovascular therapy necessitates the implementation of medical management. The use of thalidomide for the treatment of AVMs is novel but can lead to life threatening adverse events. Generally, the response is achieved with a higher dose of thalidomide up to 600 mg/d, for several months. In our patient because of renal failure the dosage was 50mg/d. Despite this, thalidomide was able to decrease the AVM flow enabling embolization with success.**



Abstract N°: 1536**Purpura fulminans: a case report with literature review**

Isabel Gainza Apraiz¹, Irene Arevalo Ortega¹, Laura Blanch Rius¹, Angela Gainza Apraiz², Frederic Samson³, Ana Sanchez Diez¹, Rosa María Izu Belloso¹

¹Basurto University Hospital, Dermatology Department, Bilbao, Spain, ²Basurto University Hospital, Plastic Surgery Department, Bilbao, Spain, ³Basurto University Hospital, Paediatric Department, Bilbao, Spain

Introduction & Objectives:

Purpura fulminans (PF) is an acute rash characterized by rapidly progressive purpuric lesions and skin necrosis due to coagulation of the microvasculature that leads to disseminated intravascular coagulopathy (DIC). It may appear in three different settings: newborns with a homozygous congenital deficiency of proteins S or C (neonatal PF), in the context of acute severe infections (infectious PF) or due to a transient autoimmune deficiency of protein S (idiopathic PF). The latter usually affects children and is often preceded by an infection (90% of idiopathic PF are postinfectious). Prognosis varies depending on the evolution of the cutaneous lesions and the extent of systemic thromboembolic phenomena.

Materials & Methods:

A case report and literature review.

Results:

A previously healthy 3-year-old girl presented to the emergency room due to the rapid onset of well-defined extensive and painful violaceous plaques in both shins within less than 24 hours. Hyperemia and non-exudative tonsillar hypertrophy were observed on physical examination, without associated fever nor systemic symptoms. Blood tests showed decreased fibrinogen levels, an elevated D-dimer and prolongation of aPTT and INR. PF was suspected, and further studies revealed a severe deficiency of protein S and elevated anti-protein S antibodies. Anti-streptolysin O (ASLO) antibodies were detected in blood samples and throat swabs revealed a positive result in the strep test. PCR for Metapneumovirus was also positive. Treatment with empiric antibiotic therapy, enoxaparin, fresh frozen plasma and immunoglobulins was initiated, with eventual normalization of protein S levels and the coagulation profile. Subsequently the patient underwent surgery and partial skin grafts were placed over both shins to repair the necrotic sequelae.

Conclusion:

Idiopathic PF must be suspected in patients (mostly children) who following an upper respiratory tract infection (1-2 weeks prior to presentation) develop rapidly progressive purpuric plaques in the lower limbs, usually in the absence of shock or systemic symptoms. It is a true dermatological emergency in which early diagnosis and a proper management are needed in order to prevent irreversible complications such as amputation or shock.

Abstract N°: 1996

Eosinophil extracellular trap cell death in the affected skin of eosinophilic granulomatosis with polyangiitis

Tamihiro Kawakami*¹

¹Tohoku Medical and Pharmaceutical University, Division of Dermatology, Sendai, Japan

Introduction & Objectives:

Eosinophilic granulomatosis with polyangiitis (EGPA) is associated with eosinophilic infiltration around the vessels, nerve fibers, and adjacent tissues of the affected skin. Recent findings revealed that eosinophil cytolysis represents a process of active cell death, referred to as eosinophil extracellular trap cell death (EETosis), which is characterized by the release of filamentous chromatin structures called eosinophil extracellular traps (EETs). EETosis is associated with the crystallization of small punctate galectin-10 to form Charcot-Leyden crystals, slender bipyramidal hexagonal crystals, as a hallmark of eosinophilic inflammation.

Materials & Methods:

We recruited patients with EGPA (n=4) from those who visited our department between October 2019 and November 2021. The patients were diagnosed according to the ACR criteria and Chapel-Hill definition. Skin biopsy samples from two patients were obtained after immunosuppressive treatment. We investigated EETosis in the affected skin of four patients with EGPA using an immunofluorescence staining method. Immunofluorescence staining for MBP, galectin-10, and DNA revealed various degree of EETosis and Charcot-Leyden crystals in skin tissue, suggesting the different degree of eosinophil activation status.

Results:

In Patient 1, we found eosinophils markedly infiltrated in the dermis including nerve fibers. Many ETotic eosinophils, detected based on MBP single positive staining, were found in collagen fibers and the perineurium of nerve fibers in the dermis. The ETotic eosinophils were formed partly in the perineurium and around the inflamed nerve fibers. In addition, Charcot-Leyden crystals, acidophilic bipyramidal structures in H&E staining that stained with galectin-10, were present proximity of the ETotic eosinophils. The number of ETotic eosinophils in Patient 1 was higher than that in other patients. In Patient 2, eosinophils moderately infiltrated the dermis using H&E staining. However, significant ETotic eosinophils were noticed using immunostaining. In Patient 3, few eosinophils were detected in the skin tissue. In Patient 4, a few intact eosinophils were detected, but MBP was markedly deposited in collagen fibers of the dermis.

Conclusion:

We found eosinophils markedly infiltrated in the dermis and many eosinophils induced EETosis in Patient 1, who had not undergone any treatment and had accompanying severe conditions such as cardiac involvement. The number of ETotic eosinophils in Patient 1 was much higher than that in Patient 2, who also had not undergone any treatment and had only moderate accompanying conditions. In contrast, there was a low frequency of ETotic eosinophils detected in Patient 3 and Patient 4, who had undergone systemic corticosteroid therapy and treatment with additional immunosuppressive agents. In addition, we found EETosis-associated Charcot-Leyden crystals in the affected skin of Patient 1. The histopathological characteristic features may help physicians establish an earlier diagnosis of intractable eosinophilic related disease including EGPA. The ETotic eosinophils were formed partly in the perineurium of the nerve fibers in the dermis of Patient 1, who had mononeuritis multiplex as a sign of peripheral neurologic involvement prior to systemic treatment. ETotic eosinophil infiltration in perineurium of

skin tissue might play a primary role in peripheral neuropathy of this disorder.

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Abstract N°: 2098**Diffuse Capillary Malformation With Overgrowth (DCMO): A Case Report and Literature Review**

Omar Safran Alhothali*¹, Sarah M. Fageeh¹, Sara F. Alharbi¹, Ethar T. Alsaedi¹, Rola R. Alsulami¹, Ahmad N. Alharbi², Ghadeer E. Alamri¹, Khalid Al Hawsawi³

¹Umm Alqura university, ²Taibah University, ³King Abdulaziz Hospital · makkah, SAU

Introduction & Objectives:

Diffuse capillary malformation with overgrowth (DCMO) was proposed in 2013 as an independent entity that is characterized by diffuse capillary malformation (CM) of the skin accompanied by proportional, nonprogressive enlargement of soft and/or bony tissue.

As with other vascular malformations, the cause is unknown; however, GNA11 mutation has been reported in some patients. The CM component of DCMO is characterized by widespread reticulated erythematous patches that may be confluent in some areas or appears block-like in others, often with a sharp midline demarcation on the trunk. Patients with DCMO might have prominent subcutaneous veins that are not considered true venous malformation. The associated overgrowth is proportionate, not progressive, and does not correlate with the location, intensity, or morphology of the CM. Other extra-cutaneous features of DCMO include syndactyly, sandal-gap, or macrodactyly in 30% of patients. Here, we report a unique case of DCMO. As the condition is rare, the patient's presentation made his disease a diagnostic challenge.

Case Presentation

A one-year-old male child with no past medical history presented with skin lesions persistent since birth, not associated with any symptoms. He was born after a non-eventual full-term pregnancy and with a spontaneous vaginal delivery. There was no history of prenatal, perinatal, or postnatal complications. The systemic reviews were unremarkable. The patient was seen by a pediatric neurologist and pediatric orthopedics. There were no neurological symptoms and no family history of similar cases. Examination revealed normal head circumference. Skin examination revealed widespread non-scaly reticulated erythematous patches all over his body. There were homogenous erythematous patches on his abdominal wall too. Musculoskeletal examination revealed that the circumference of the right calf and mid-thigh was 13 cm and 20 cm, respectively, whereas the circumference of the left calf and mid-thigh was 11 cm and 18 cm, respectively. The length of both lower extremities was similar. There was also syndactyly of the right 2nd and 3rd toes. Differential diagnoses include cutis marmorata telangiectatica congenita (CMTC), DCMO, and macrocephaly-CM (M-CM) syndrome. Based on clinical features, the patient was diagnosed with DCMO. The parent was reassured and put under follow-up by pediatric orthopedics for periodic monitoring of growth asymmetry.

Conclusion:

DCMO is a newly introduced clinical diagnosis denoting a vascular anomaly, which is characterized by reticulated patches and nonprogressive soft tissue hypertrophy. It is a rare diagnosis and the presentation of the patient made his disease a diagnostic challenge. Dermatologists should be aware of its presentation as such patients require multidisciplinary management including pediatric orthopedics, neurology, and dermatology. Patients with DCMO need periodic follow-ups to monitor for leg length discrepancies.



Abstract N°: 2388**Eccrine Angiomatous Hamartoma Associated with Lower Extremity Venous Insufficiency: a Rare Case Report**Isabela Dorado Caycedo*¹, Eduardo Rivera Perdomo², Juan Carlos López Takegami³, Claudia González Díaz⁴¹Sanitas, Dermatology, Bogotá, Colombia, ²Fundación Universitaria Sanitas, Department of Dermatology, Bogotá, Colombia, ³Sanitas, Department of Dermatopathology, Bogotá, Colombia, ⁴Ultrasonido Altamente Especializado, Radiology, Bogotá, Colombia**Introduction:**

Eccrine angiomatous hamartoma (EAH) is a benign tumor characterized by the proliferation of eccrine glands and vascular structures. It can be congenital or appear during childhood, with rare cases arising in adults. EAH typically presents as a purplish plaque or nodule on the inferior extremities that may be associated with hyperhidrosis or hypertrichosis. It is usually solitary, but a multifocal presentation in a blaschkoid or segmental distribution has been described. Histopathological hallmarks include mature eccrine gland proliferation in a close relationship with benign vascular proliferation and a variable presence of pilar, lipomatous, mucinous, or lymphatic structures.

We report the case of an extensive EAH following a vascular distribution arising in an adult diagnosed with venous insufficiency at a young age.

Results:

A 35 year-old-male presented with a history of bilateral lower limb pain that began at age 16. He was diagnosed with venous insufficiency; at 19 years old, he had a left saphenectomy. A few months later, the patient started to present multiple violaceous reticular plaques following a vascular pattern associated with hyperhidrosis and mild pain. The lesions have spread from the legs to the thighs and have darkened over time. The symptoms worsen when he is standing up.

Histopathology revealed hyperkeratosis, undulating acanthosis with hyperpigmentation of basal keratinocytes. In the dermis, an increase in the number of small and medium-sized blood vessels with muscle walls of uneven thickness and proliferation of mature eccrine glands and ductal structures intermingled with mature adipose tissue. These findings were entirely consistent with EAH.

Venous Doppler examination showed a right saphenous and perforator vein insufficiency and a left leg perforator vein insufficiency with epifascial collateral proliferation. Arterial Doppler was normal.

High-resolution dermatological ultrasound showed an increase in the diffuse hyperechogenicity of the epidermis with an increase in its thickness. The dermis had multiple tubular, serpentine structures corresponding to vascular dilatations of variable caliber. Most were venous and compressible on Doppler exploration, and some were low-resistance arterial vessels. These dermal vessels followed a disorganized, irregular pattern and came into contact with larger varices that diffusely compromised the subcutaneous cellular tissue. Superficial variceal dilatations were compressible with no signs of thrombosis and included the territories of the greater and lesser saphenous veins in both legs. In the dermis, the hypertrophy of the sweat glands with hyperechoic, oval, prominent nests became evident.

Conclusion:

Eccrine angiomatous hamartoma is a rare tumor of unknown etiopathogenesis. Some authors have proposed that

it results from failed interaction between epidermal and mesenchymal components. Another theory states that it is derived from repeated microtrauma. We report the case of a patient with an extensive EAH where venous insufficiency could have induced hamartomatous eccrine and vascular proliferation. High-resolution dermatological ultrasound proved to be a valuable tool for studying this case.

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Abstract N°: 2767**Giant chancre in an octagenarian: remember not to forget syphilis.**Mauro Ramos*¹, Andressa Goldman Ruwe²¹Faculdade de Ciências da Saúde Moinhos de Vento, Porto Alegre, Brazil, ²University of Vale do Rio dos Sinos Campus São Leopoldo, Brazil

Introduction & Objectives: Syphilis has turned into an epidemic over the last two decades in high-income countries, but low and medium-income countries are disproportionately hit. It is a polymorphous and unpredictable course, can be asymptomatic or alternate clinical silence and presence of manifestations. The primary lesion typically consists of an erosion or shallow ulcer that occurs over an indurated base.

Methods: Case presentation

Results: An 85-year-old Caucasian male was seen at a dermatology clinic complaining of a painless penile lesion which started two weeks previously. He reported having had an oral sexual relationship with a female sex worker six weeks before the beginning of the lesion. He reported no other health problems, allergies or treatments. He denied having syphilis or other STI in the past. Because of partial cognitive deficit, he was accompanied by a relative. Both consented to the publication of the case and image. Physical exam: three to four-centimeter-wide ulcer that reached the subcutaneous tissue down to the Buck's fascia. The ulcer presented a fleshy clean floor, a punched-out edge, and an infiltrated border. Palpation of the inguinal regions revealed bilateral, multiple, non-tender, elastic, and movable lymph nodes, and one of them presented was unequivocally larger ("pléiade inguinale de Ricord").

The on-site rapid immunochromatographic test for syphilis was positive. HIV, hepatitis B and C were negative. Considering the available epidemiological, clinical and laboratory data, performing a biopsy was ethically inadmissible. A direct test for *T. pallidum* was not available. VDRL reagent up to a dilution of 1:16. Immediate treatment: injection of 2.400.000 UI of benzathine-penicillin. Topical treatment: gentle cleansing, no topical treatment was recommended. The lesion disappeared after treatment, but the patient did not come back for serological follow up.

Conclusion: The morphology of the hard chancre varies greatly. Large and deep lesions are not common, and they may be referred to as giant chancre when larger than two centimeters. It is associated with immunodepression states, such as aging, AIDS, malnutrition, alcohol, or other drug related problems. STI and other infectious diseases must be considered in the differential diagnosis, especially chancroid, herpes simples and donovanosis. Long-lasting lesions demand diagnostic exclusion of neoplastic processes, especially in the elderly.

Our case reinforces the inclusion of STI in the differential diagnosis of genital ulcers with disregard to the age of patients. The false belief that old age, and even the presence of cognitive impairment, exclude sexual activity and exposure to sexually transmitted agents must be abandoned.



Abstract N°: 2875

Vascular malformations of the lip in the pediatric population: a series of six cases

Mouhsine Zineb¹, Fatim Zahra El Fatoiki¹, Hali Fouzia¹, Kamelia Chban², Soumia Chiheb¹

¹UCH Ibn Rochd , Department of Dermatology and Venereology, , casablanca,²Harouchi Children's Hospital, Pediatric radiology department, casablanca

Introduction:

Vascular malformations (VVMs) are abnormalities of vessel development during intrauterine life. The 1996 International Society for the Study of Vascular Anomalies (ISSVA) classification, updated in 2018, distinguishes vascular malformations into high-flow VVMs, low-flow VVMs and combined VVMs. Labial localization remains rare.

Objectives:

The aim of our study is to share our experience in the diagnosis and management of children with labial vascular malformations.

Materials and Method:

Prospective descriptive study from March 2021 to March 2022, including all children presenting to the pediatric dermatology consultation. During this period, six cases of labial vascular malformations were collected.

Results:

There were 3 girls and 3 boys, with an average age of 6 years. The average time to diagnosis was 8 months. The lesions were present from birth in 4 cases and at the age of 7 years following a trauma in 2 cases. Clinically, 4 children initially presented with a red, warm, pulsatile labial macule, evolving a few months later to a hypertrophic deformity in 3 cases and ulceration and spontaneous bleeding with localized hypertrichosis in 1 case. The two remaining children had a bluish mass that was compressible to palpation, resulting in major aesthetic and functional sequelae. In the majority of cases, the diagnosis was based on the history and clinical examination, but additional biological and radiological examinations (ultrasound coupled with Doppler, angio-MRI, and CT Scan) were requested before any treatment. Four cases of arteriovenous malformations and two cases of venous malformations were found. Treatment by embolization was performed in four cases; only one case was treated by sclerotherapy. Abstention with monitoring was recommended in only one case.

Discussion:

Vascular malformations are pathologies of variable frequency and severity. Labial localization remains exceptional. Most of them are sporadic, but some are genetically transmitted. They are present from birth but can be triggered by an infection, a trauma, or during puberty, and persist for life. Diagnosis is most often clinical, but further investigations are often necessary and useful in determining the type of malformation and guiding the choice of treatment. The management of this pathology calls upon the skills of multiple practitioners from different specialties. The development of lasers, interventional radiology, and surgical techniques has improved the quality of life of patients.



Abstract N°: 2944

Clinico-immunological analysis of vasculitis: A case series of 16 patients.

Bushra Muna*¹

¹Mahatma gandhi medical college and Research Institute, Dermatology, Puducherry, India

Introduction & Objectives:

Cutaneous findings are often a window to underlying systemic disorders. Similarly, cutaneous vasculitis can be a marker alluding to an inapparent more sinister systemic involvement. Also, in cutaneous vasculitis; skin tissue can be easily sampled for diagnostic tests like histopathology and immunofluorescence studies. Cutaneous small vessel vasculitis is a type of leukocytoclastic vasculitis which is usually confined to skin with rare extracutaneous manifestations in less than 30% of the cases.

Objectives:

To study the clinical and immunopathological features and their correlation in cutaneous small vessel vasculitis.

Materials & Methods:

It is a prospective observational study where patients of all age and gender with a clinical diagnosis of vasculitis were included in the study in period of three months were included and those who did not consent to participate in the study were excluded. After getting informed consent, clinical examination findings were documented. Complete hemogram, urine microscopy, renal and liver function tests, CRP, ESR, ASO titre, recording of vital signs were done in all patients. Under local anaesthesia and aseptic precautions, skin biopsy of the suspected vasculitic lesions was performed using 4 mm disposable skin punch. One specimen was formalin fixed, paraffin embedded and stained by hematoxylin and eosin stain for light microscopic examination and another specimen was examined by direct immunofluorescence and results were recorded. ANA, ANCA, serum complement level, USG abdomen, CT imaging, serology for hepatitis and HIV were done in patients suspected of systemic involvement.

Results:

Slightly greater than half of the patients were female (10/16, 62.5%). The most common extracutaneous involvement was musculoskeletal (joints) which was present in ten patients. History of COVID-19 vaccination in recent past (within 2 weeks) was present in two patients and one patient turned out to be HCV positive with negative cryoglobulin test.

The most common skin lesion was palpable purpura (13/16) and the most common site involved was leg (14/16). Systemic involvement was seen in more than half of the cases (10/16). On radiological imaging (CT-abdomen) two patients had features suggestive of mesenteric inflammation and terminal ileitis and both patients also had presence of occult blood in stools. ANCA was negative in all patients while ANA was positive in only one patient. On histopathology examination, most patients showed features of leukocytoclastic vasculitis with the most common inflammatory infiltrate being mixed (neutrophilic and lymphocytic). Interface dermatitis was seen in one patient who also had positive ANA titre. In direct immunofluorescence, the most common immune deposit seen was IgA deposits.

Clinical diagnosis of cutaneous small vessel vasculitis was made in 8 patients, HSP/IgA vasculitis in five patients, urticarial vasculitis in one patient, lupus vasculitis in one patient, erythema elevatum diutinum in one patient.

Conclusion:

The patients with raised neutrophil to lymphocyte ratio, eosinophilia, extensive skin involvement and raised ESR/CRP had systemic involvement. Cutaneous vasculitis needs more studies to understand the enigmatic condition so that guidelines can be formed for appropriate lab workup so that unnecessary cost for investigations can be reduced and to provide proper management. Our study is limited by the limited number of participants.

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Abstract N°: 2966

Macular lymphocytic arteritis: a rare form of cutaneous vasculitis

Hajar Douma¹, Maryem Aboudourib¹, Ouafae Hocar¹, Said Amal¹

¹chu med VI, dermatology, Morocco

Introduction & Objectives:

Lymphocytic macular arteritis (LMA) is a rare cutaneous vasculitis. It is characterized by erythematous and pigmented macules of the lower limbs, of chronic and indolent evolution, which anatomopathological examination reveals a lymphocytic arteritis of variable intensity. We present a new case of this pathology.

Materials & Methods:

This is a case report

A 37 year old patient, without any particular pathological history, pregnant at 15 weeks of amenorrhea, presented with livedoid skin lesions that had been evolving for 7 years, located on both lower limbs, painless and non-pruritic, without any other associated signs, notably no neurological or osteoarticular signs.

On examination, they were reticulated erythematous macules, livedoid, non-infiltrated, located on the two lower limbs, not disappearing when the limbs were warmed up or raised, there were no nodules on examination of the venous tracts, no necrosis or ulceration lesions, and the neurological and obstetrical examination were normal.

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

A complete blood workup was requested including immunological workup, all came back normal, no systemic involvement was found.

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

The patient was put on colchicine. After 7 months, the skin condition was stable with persistent pigmented scars.

Results:

Macular lymphocytic arteritis is a cutaneous vasculitis of medium-caliber vessels, singular in its clinical presentation as hyperpigmented macules without extracutaneous signs, and its histologic appearance of lymphocytic arteritis with an unruptured internal elastic boundary and a ring of fibrinoid necrosis with a parietal hyaline appearance. Clinically, the skin lesions in MLA are indolent and non-infiltrative, as opposed to the palpable and classically painful lesions seen in vasculitis. There are no associated systemic signs. The histological feature is the presence of a lymphocytic infiltrate within the arterial wall, associated with a ring of eosinophilic fibrinoid necrosis of hyaline appearance and a variable degree of luminal thrombosis close to poyarteritis nodosa . Preservation of the internal elastic boundary would be the main criteria for differentiating macular lymphocytic arteritis and periarteritis nodosa. The evolution is benign, but the lesions respond poorly to conventional treatments.

Conclusion:

Lymphocytic macular arteritis is a newly individualized anatomoclinic entity characterized clinically by a macular elemental lesion and histologically by an image of lymphocytic arteritis.

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Abstract N°: 3011**BASCULE syndrome after COVID -19 vaccination**

Raquel Carrascosa¹, Elisa Hernández de la Torre Ruiz¹, Cristina Mauleón Fernández¹, Inmaculada Balaguer¹, Kevin Díez Madueño¹, Ana Simón Gozalbo¹, Maria Gamo¹, Pablo De la Cueva Dobao¹

¹Hospital Universitario Infanta Leonor, Dermatology

Introduction & Objectives: Bier anemic spots, cyanosis and urticaria-like eruption (BASCULE) syndrome is an underreported paediatric vascular disorder from the group of acrosyndromes. Most cases are adolescents between 12 and 19 years of age or newborns in the first few months of life. The skin lesions usually appear in the lower limbs after prolonged standing and resolve in a few minutes by lying down or walking. The course is unpredictable and the symptoms usually affect the patients' quality of life.

Materials & Methods: A 14 year-old woman presented with a painful and pruritic erythematous and cyanotic rash, mainly located on her lower limbs. The lesions had appeared 8 months earlier, after receiving her second dose of Pfizer COVID-19 vaccine a few weeks ago. Past medical history included hereditary coagulation factor VII deficiency (normal hemostasis range). The lesions typically appeared on the lower extremities acutely after prolonged standing, resolving rapidly by lying down or walking. Upper extremities and hands were affected occasionally. Physical examination, after a few minutes in a standing position, showed multiple hypochromic macules and urticarial papules with a surrounding erythrocyanotic background on the bilateral legs and thighs.

Results: Laboratory tests, including a complete blood count, basic metabolic, hepatic and renal panel, C-reactive protein, thyroid studies, antinuclear antibody and IgA antitransglutaminase antibody, were unremarkable. The patient was referred to paediatric cardiology for an active standing test in order to exclude postural orthostatic tachycardia syndrome and other forms of orthostatic intolerance. Heart rate, blood pressure, electrocardiogram and echocardiogram were normal. Bilastine 20 mg twice daily was started and she achieved nearly complete resolution of pruritus while taking the drug, but recurred when the dose was halved. Additional preventive measures were recommended, such as physical exercise, healthy diet and elevating the legs when possible.

Conclusion: The pathogenesis of BASCULE syndrome remains unknown. An exaggerated response to decreased arterial oxygen saturation due to venous pooling induced by orthostatism appears to be the trigger of erythrocyanosis and Bier anemic spots, while urticaria-like eruption may result from mast-cell degranulation in response to hypoxia. SARS-COV2 infection has been associated with a variety of skin lesions, including pernio-like changes ("COVID-toes") and peripheral acrocyanosis. The pathophysiological mechanisms are not clearly elucidated, but it is well known that COVID-19 virus and COVID-19 vaccines promote a proinflammatory state that facilitates microvascular thrombosis and intussusceptive angiogenesis. Both may trigger Toll-like receptors 7/8 inducing type 1 interferons production and others proinflammatory cytokines. We present a case of acute-onset BASCULE syndrome following prior COVID -19 vaccination. Berrebi D et al published a case of acute-onset BASCULE syndrome following prior asymptomatic SARS-COV-2 infection. Although they assumed that this finding may very well be coincidental, this case uncovers a plausible biological mechanism by which post-COVID endothelial dysfunction and exaggerated arteriolar constriction trigger or exacerbate BASCULE syndrome in individuals with pre-existing vascular hypersensitivity. The same explanation may be attributed to our case.

Abstract N°: 3685**Polyarteritis nodosa in a kidney transplant patient: A case report**Mohand Ouali Ait Si Ali¹, Ratiba Halladj¹, Imad Eddine Rabia¹, Assya Djeridane¹¹Hopital Central de l'Armée, Dermatology, Algiers , Algeria**Introduction & Objectives:**

Polyarteritis nodosa is a medium vessel vasculitis that affects multiple organ systems. Characterized clinically by general signs, peripheral neuropathy, arthralgia, myalgia and sometimes-visceral damage. Renal involvement is related to renal ischemia with infarction, but there is no glomerulonephritis.

We report a particular case of polyarteritis nodosa by its occurrence in a kidney transplant recipient

Materials & Methods:

A 45-year-old patient with history of kidney transplant after hypertensive nephropathy eight years ago. He is received immunosuppressant treatment (ciclosporin and mycophenolate mofetil). On examination he presented an erythematous nodular lesions with painful on palpation, associated with arthralgia affecting the peripheral joints and weight loss of 16 kg.

Results:

Her investigations revealed a normal renal assessment, an inflammatory syndrome, a negative immunological assessment and a negative viral serology (hepatitis B and C). Skin biopsy report objectified an image of leukocytoclastic vasculitis. The graft ultrasound was without abnormality. In front of clinical, biological and histological elements, the diagnosis of PAN was retained. The management included corticosteroid boluses with increased doses of prednisone 70 mg/day, ciclosporin 80 mg/day and substitution of mycophenolate mofetil by azathioprine 150 mg/day. There was a good improvement in the skin lesions after one month of treatment.

Conclusion:

Polyarteritis nodosa is a well-recognized but rare cause of vasculitis affecting the peripheral nerves, skin, kidney, and gastrointestinal tract. Signs and symptoms of polyarteritis nodosa are primarily attributable to diffuse vascular inflammation and ischemia of affected organs. Renal hypertension may occur due to multiple renal infarcts. In our patient, the origin of the renal failure, which led to the transplant, can be attributed to an unrecognized polyarteritis nodosa that recurred eight years after the transplant.



Abstract N°: 3719

A case report of IgA vasculitis in an adult patient: a diagnostic and therapeutic challenge

Maria Ionescu*¹, Mihai Catalina¹, Iancu Isabela¹, Popescu Mihaela^{1, 2}

¹Spitalul Victor Babeş, Dermatovenerologie, Bucureşti, Romania, ²Carol Davila University of Medicine and Pharmacy, Bucureşti, Romania

Introduction & Objectives:

Immunoglobulin A vasculitis (IgAV) is predominantly affecting children and is less prevalent among adults. Given the low incidence, adult demographic often encounters misdiagnosis and undertreatment, despite being prone to more severe forms of IgAV and long term complications.

Our intention is to heighten awareness of IgAV within adult population and to enhance the diagnostic and therapeutic approach.

Materials & Methods:

A 39-year-old male, without significant medical history, presented with an ill-defined erythematous plaque on his lower left thigh, exhibiting the classical signs of inflammation described by Celsus: edema, tenderness, hyperemia and increased local temperature. In addition, the patient showed a symmetrical, bilateral eruption of erythematous-violaceous papules on the lower extremities that developed 4 days apart from the initial plaque. The patient reported frequent exposure to insect bites due to his gardening activities.

Laboratory blood tests revealed neutrophilic leukocytosis and elevated markers of inflammation. The remaining blood analyses and urinalysis were within the normal range. Based on these findings, we established the diagnosis of cutaneous necrotizing vasculitis caused by infection and we initiated a course of intravenous antibiotic therapy combined with topical corticosteroids.

The disease showed periods of remission and exacerbation. Within 72 hours, the erythematous plaque resolved and the intensity of the purpuric lesion diminished. However, after the next 5 days, the patient reported the onset of joint pain and edema. We administered oral corticosteroids and his symptoms ameliorated. Despite his clinical improvement, inflammatory markers remained elevated and over the following days, the purpuric lesions extended proximally to his thighs, gluteal region, trunk and arms.

The patient was further investigated and the immunologic analysis revealed elevated IgA seric levels, confirming the diagnosis of IgA vasculitis. The patient received intravenous corticosteroid therapy that led to overall improvement regarding the joint pain and cutaneous lesions.

Results:

We were unable to establish the diagnosis of IgAV based on the clinical classification criteria described by EULAR/PRINTO/PRES in 2010 during the first week following the onset of the disease, as there was no evidence of systemic involvement.

Immunologic investigations proved useful in guiding the diagnosis.

Conclusion:

The course of IgAV in adults may be atypical, have a fluctuating pattern, remission can be incomplete followed by

relapse. Therefore a thorough investigation is necessary and monitoring the patient for an extended period of time may prove useful in discovering early signs of complications.

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Abstract N°: 4018**Kasabach-Merritt phenomenon: a single center experience**

Sophia Abdelilah¹, Bochra Bennour¹, Imane Hakim¹, Maryem Aboudourib¹, Said Amal¹, Ouafa Hocar¹

¹Marrakesh, Department of Dermatology,, Marrakesh, Morocco

Introduction & Objectives:

Kasabach merritt phenomenon (KMP) is a rare life-threatening condition, defined by the association of thrombocytopenia and consumptive coagulopathy to specific vascular tumors: tufted angioma (TA) or kaposiform hemangioendothelioma (KHE).

TA also called angioblastoma and KHE belong to the spectrum of benign vascular tumors; these tumors of a challenging diagnosis, are characterized by their clinical and histological presentation and appear mostly during infancy.

Despite of their benign character, they can be life-threatening especially when associated with the KMP.

Materials & Methods:

We report a case series of KMP seen in the paediatric emergency room of Mohammed the VI university hospital over a period of 10 years from January 2013 to January 2023.

Our serie included 6 cases of the Kasabach-Merritt phenomenon, complicating FOUR Tufed angioma and TWO kaposiform hemangioendothelioma . The sex ratio was 5/1, the age varied from a few hours to 3 months old and the tumor locations were : face (n=3), trunk(n=2), thighs(n=1), and in one case an infant presented with a cutaneous and mediastinal located tufted angioma .

Results:

All of our patients had severe thrombocytopenia with a prolonged prothrombin time and partial thromboplastin time, hypofibrinogenemia, and increased D-dimer levels .Four of them were successfully treated with mTOR inhibitors such as sirolimus or everolimus associated to prednisolone, acetylsalicylic acid and ticlopidine . In fact within 4 weeks of starting mTORS inhibitors, they had significant improvement in clinical and biological parameters, the tumor decreased by half of its initial size and the platelet count drastically increased .

Unfortunately the two other children died as a result of severe sepsis before starting mtor inhibitors.

Conclusion:

Kasabach-Merritt phenomenon was first described in 1940, in a child presenting with a giant hemangioma associated to thrombocytopenia , and was considered to be a rare complication of infantile hemangiomas, until 1990 when Enjolras and al reported the histopathological characteristics of the tumors linked to this phenomenon describing tufted angioma and kaposiform hemangioendothelioma .

This phenomenon pathophysiology is still unknown making it a real diagnostic and therapeutic challenge.

Inhibitors of the mammalian target of rapamycin (mTOR), Sirolimus and Everolimus , have been used since 2010 and are a promising therapy in the management of KMP, these medications can inhibit lymphangiogenesis and angiogenesis and have demonstrated their efficacy and safety .

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Abstract N°: 4182**Alpelisib For PIK3CA-Related Vascular Anomaly**Yaron Gu^{*1, 2}, Simon King³, Deshan Sebaratnam^{1, 2}¹University of New South Wales, South West Sydney Clinical Campuses, Sydney, Australia, ²Liverpool Hospital, Dermatology Department, Liverpool, Australia, ³University of Newcastle, School of Medicine, Newcastle, Australia**Introduction & Objectives:**

PIK3CA-related overgrowth spectrum (PROS) is an umbrella term relating to somatic mutations in PIK3CA which encodes for a catalytic subunit of phosphatidylinositol 3-kinase (PI3K). PROS is associated with hyperactivation of the PI3K signalling pathway and characterised by vascular malformations and abnormal overgrowth of various tissues. Current and emerging targeted agents for PIK3CA-related disorders include sirolimus, miransertib and alpelisib. We report a rare case of PIK3CA-related vascular anomaly and one of the first successful treatments with alpelisib in Australia.

Materials & Methods:

A 25-year-old female presented for review of a vascular birthmark. She reported increasing pain, bleeding, and soft tissue overgrowth within a congenital plaque on her right flank. She reported temporary symptomatic relief with cautery and debulking surgery performed years prior to presentation. Her past medical history was remarkable for cross-fused renal ectopia and endometriosis. The birthmark caused significant pain for which she had previously trialled multiple analgesics with minimal improvement. Examination revealed a geometric maroon patch measuring 35cm by 30cm. It was studded with dark vesicles, characteristic of microcystic lymphatic malformations in association with soft tissue swelling. Incisional biopsy demonstrated dilated, thin-walled vessels containing blood and proteinaceous fluid within the dermis and underlying subcutis. No endothelial atypia or complex architectural vascular formations were identified. Inflammation was inconspicuous. CT revealed a poorly defined serpiginous lesion within the subcutaneous tissue, with large tortuous veins, no intra-abdominal extension, or features suggestive of high-flow malformation. Repeat biopsy of lesional tissue demonstrated a PIK3CA c.1624G>A variant; a mosaic missense substitution in which glutamic acid is replaced by lysine, previously identified as a hotspot mutation. Her presentation was in keeping with a PIK3CA-related vascular anomaly composed of capillary, lymphatic and venous malformations. Given the morbidity associated with her birthmark, she was commenced on targeted treatment with a PI3K inhibitor, alpelisib 250mg once daily. At two-month follow-up, there was significant improvement with lightening of the capillary malformation and decrease in the number of microcystic lymphatic malformations.

Results & Conclusion:

The study of vascular birthmarks is a rapidly evolving area of medicine as our knowledge of the genetic mechanisms underpinning these lesions advances. We present one of the first patients in Australia to be treated with alpelisib and highlight the importance of multidisciplinary care foregrounding the roles of dermatology, pathology, and genetics in the management of this case.

Abstract N°: 4512

Familial Cutaneous Glomuvenous Malformation

Chadia Naji¹, Oumayma Handi¹, Maryem Aboudourib¹, Said Amal¹, Ouafa Hocar¹

¹CENTRE HOSPITALO-UNIVERSITAIRE MOHAMMED VI MARRAKECH, dermatology and venerology, Marrakesh, Morocco

Introduction & Objectives:

Glomuvenous malformations (GVMs) present as asymptomatic multiple pink-to-blue nodules or plaques. Familial cases are caused by mutations in the glomulin gene. We present a case of a 29-year old male who presented clinically with multiple slowly enlarging masses of the left upper limb revealing a familial Glomangiomas.

Materials & Methods: this is a case report

Results:

A 29-year-old male presented with complaints of appearance of bluish-to-dusky red-coloured nodules over left upper limb since childhood. On examination, there were 5 dusky red-to-bluish firm nodules ranging in size from 1 to 3 cm, one on the left forearm, the others grouped forming a plaque on the back of the left hand. They were non-compressible and mildly tender on palpation. There was no associated bruit or thrill. His father and his brother had similar lesions but in a limited distribution.

Keeping the possibilities of GVM, blue rubber bleb nevus and venous malformation, a skin biopsy was taken from the nodule. Microscopic examination of the biopsy showed histological features consistent with the diagnosis of GVM. On ultrasound, there was a tubuliform formation consistent with a venous ectasia type vascular malformation, colour Doppler was normal. Our patient is scheduled for sclerotherapy next month.

Discussion:

A glomuvenous malformation (GVM) is a type of vascular malformation formed by an abnormal growth of blood vessels and the presence of glomus cells in the wall of the malformation. GVMs can occur as sporadic and inherited lesions, the pattern of inheritance for multiple GVMs is autosomal dominant with incomplete penetrance and variable expressivity involves mutations in the glomulin gene.

Multiple GVMs are uncommon and account for <10% of all reported cases. GVMs present with multiple lesions, most often seen in children or adolescence. Our patient had multiple bluish nodules and pink-to-blue plaques in a disseminated pattern on the left upper limb, other variants are the disseminated type, which consists of multiple lesions distributed over the body and the congenital plaque-like glomus tumors. In addition, our case was a familial case of GVM with similar lesions in the father and brother. The prevalence of inherited GVMs varies from 38% to 68% of the cases in various studies. Internal organ involvement with GVM is rare, however, there appears no specific association with multiple cutaneous lesions and internal organ involvement. Our patient did not have any systemic complaint.

The treatment modalities reported for treatment of multiple glomangiomas are surgical resection, sclerotherapy, sirolimus, ethanol in the form of a gel for injection, [pulsed dye laser](#), electron-beam radiation. The prognosis for patients with glomangiomas is excellent, and most patients never experience any related medical problem.

Conclusion:

This case is important because, although rare, glomangiomas are a cause of multiple lesions in the subcutaneous tissues that can mimic the findings of lesions that may present as multiple nodules.

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Abstract N°: 4923**Current approaches in the management of cutaneous vasculitis**Beatrice Balaceanu^{*1}, Mara Mihai^{1, 2}, Silvia Farcas², Alexandra Timofte¹, Calin Giurcaneanu^{1, 2}¹Elias University Emergency Hospital, Dermato-oncology, Bucharest, Romania, ²Carol Davila University of Medicine and Pharmacy, Bucharest, Romania**Introduction & Objectives:**

Vasculitis is a heterogeneous group of afflictions characterized by inflammation of the vascular wall, affecting any organ and system. Vasculitis is divided according to the size of the affected blood vessels into four broad categories: small vessel, small and medium vessel, medium vessel and large vessel vasculitis. Cutaneous vasculitis includes any type of vasculitis whose manifestations involve skin damage. The correct, up-to-date management of patients who develop cutaneous vasculitis is essential, demanding an early diagnosis, as well as the involvement of a multidisciplinary medical team.

The objectives of our study consisted of the analysis of personal, clinical and paraclinical data of patients with cutaneous vasculitis admitted to the hospital, followed by a descriptive analysis of the population according to demographic data, the assessment of the type of cutaneous vasculitis, diagnostic and treatment options.

Materials & Methods:

We performed an observational, retrospective study, on a group of 84 patients, hospitalized between January 2012 and October 2020. The patients' data were obtained from the electronic archive of the hospital. The observation forms and paraclinical investigations were analyzed, including the results of the histopathological examinations that were performed during this period. The aim of our study was to carry out a detailed analysis of the types of cutaneous vasculitis.

Results:

The most common type of cutaneous vasculitis found in our study was ulcero-necrotic vasculitis (29 patients - 34.52%). As other research studies prior confirmed, the main clinical skin manifestations of vasculitis were ulcerations and palpable purpura, predominantly localized on the lower limbs. The most useful paraclinical diagnostic methods were blood count, inflammatory markers, specific and non-specific antibody dosage, histopathological examination of skin lesions to confirm clinical suspicion and also bacteriological examination of skin lesions in certain cases. The most common systemic therapy recommended was corticosteroid treatment, followed by synthetic antimalarials, antihistamines and immunosuppressants. The main exacerbating factors encountered were certain treatment regimens, associated infections and smoking. As a result, patient education may reduce the number of exacerbations of the existing pathology and provide a better outcome.

Conclusion:

In conclusion, the results of our study support the importance of a proper management of cutaneous vasculitis, with an early and correct diagnosis, in order to control and improve the disease's manifestations and, accordingly, to increase the patient's quality of life.

Abstract N°: 5015**Vulvar Pyogenic Granuloma in a pregnant women: A rare association**Zakia Douhi¹, Meryem Soughi¹, Sara Elloudi¹, Hanane Baybay¹, Fatima Zahra Mernissi¹¹University Hospital Hassan II , dermatology, fez, Morocco**Introduction**

Pyogenic granuloma is a relatively common benign vascular lesion whose exact cause is unknown. The lesion usually occurs in children and young adults on the head, neck, extremities, and upper trunk. The localization of the vulva is a relatively rare finding. In this case report, we present for the first time a woman at the second trimester of pregnancy with pyogenic granuloma of the vulva.

Observation :

A 26-year-old multiparous woman at 34 weeks of gestation with history of gestational diabetes; who presented with a complaint of a growing and occasionally bleeding mass on the right labia majora that she had for half month. According to the patient, it bled occasionally on contact with her underwear. However, she did not recall any trauma or injury before she noticed the papule other than a history of itching for one month. She visited our dermatology department due to the malignant appearance of the lesion and the pain that she occasionally experienced.

On examination: a reddish papule pedunculated was observed on the right labia majora, the surface of the lesion, which measured about 15-20 mm, was ulcerated. The dermoscopic examination revealed : A reddish homogeneous area with whitish zones,** a white lines like a double rail,** and an** ulceration which are frequently observed in pyogenic granuloma

The lesion was excised under local anesthesia. Histopathology revealed an epidermis ulcerated which is replaced by a fibrino-leukocytic coating. In the dermis there was a vascular proliferation of congestive vessels bordered by turgid endothelial cells with inflammatory cell infiltration. The picture was suggestive of pyogenic granuloma.

Discussion : **

Pyogenic granuloma (lobular capillary hemangioma) is an acquired vascular tumor lesion of the skin and mucous membranes. This hypervascularized lesion grows rapidly in a couple of weeks or months and usually presents as an erythematous, pedunculated, exophytic, or sessile lesion, with a smooth or lobulated surface. It results from a reactive inflammatory process and is filled with proliferating vascular channels, immature fibroblastic connective tissue, and scattered inflammatory cells, the surface is usually ulcerated and the lesion exhibits a lobular architecture.

The etiopathogenesis of pyogenic granuloma remains unclear. Minor traumas or underlying cutaneous diseases could cause an excessive local production of angiogenic growth factors or cytokines, which could be an important factor of the pathogenesis of the tumor. In our case, a history of itching with is common and often due to hormonal changes observed at pregnancy, also, a [vaginal yeast infection](#), whose risk is increased in diabetes is a condition that causes itching and, as a result a chronic irritation hasten the growth of lesions.

Pyogenic granuloma in pregnant women may resolve spontaneously after delivery, and conservative therapy is sometimes the best strategy in those cases. Laser surgery can also be performed but it has not been proven to be superior. Patrice et al report that full-thickness skin excision appears to yield the lowest chance of recurrence.

Conclusion :

Vulvar pyogenic granuloma associated with pregnancy is a very rare clinical condition. We should keep in mind the diagnosis of PG in cases of rapidly growing, lobulated, erythematous, bleeding lesions of the vulva. This case was presented to help physicians become aware that lobular capillary hemangiomas may occur at this site.

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Abstract N°: 5110**An excellent evolution of a multiple pyogenic granuloma with corticotherapy and Beta-blockers.**

Asmaa Lahrougui¹, Maryem Aboudourib¹, Ouafa Hocar¹, Said Amal¹

¹CHU Marrakech, Dermatology-venerology, Marrakech , Morocco

Introduction & Objectives:

Pyogenic granuloma or Botryomycoma is a rare benign vascular tumor, its etiopathogenesis remains unresolved, causing aesthetic damage especially for conspicuous localizations.

We report a case of multiple pyogenic granuloma in a 47 year old patient following an AVP.

Materials & Methods: Case report

This is about a 47-year-old patient, victim of a trauma due to a car crash that caused a deep facial wound, abdominal dermabrasion, and an ankle fracture for which he was operated 1 month ago.

The patient consulted for diffuse skin swellings, evolving 20 days after his trauma, causing aesthetic discomfort.

The clinical examination revealed the presence of angiomatous, budding, diffuse, painless and non-pruritic nodules of different sizes with irregular edges, located at the site of the facial wound and abdominal dermabrasion, all evolving in a context of apyrexia and conservation of the general state.

The patient was treated with oral corticosteroids at a dose of 1mg/kg/D with a 10mg degression every 10 days, as well as with systemic propanolol.

The clinical evolution was spectacular, with a clear clinical improvement from the first week, as well as a near-disappearance of the lesions over 3 weeks.

Discussion :

Pyogenic granuloma (PG) is a benign tumor belonging to the lobulated capillary angiomas, first described by Poncet and Dor in 1897.

Often the prerogative of children and young adults, rare clinical forms have been described in the elderly, typically manifesting as a small fleshy mass, 0.5 cm to 2 cm, soft and red, whose slightest touch leads to bleeding, with a preferential localization on the extremities.

Pyogenic granuloma is mostly solitary, but may in rare cases be multiple, eruptive or recurrent, and may appear spontaneously or following various triggers such as trauma, wounds or burns.

The diagnosis is usually clinical, but is confirmed histologically.

Various treatments have proven effective, including electrocoagulation, cryotherapy, silver nitrate, laser, and topical or systemic corticosteroid therapy and beta-blockers.

However, recent publications have proven the local efficacy of table salt in pyogenic granuloma.

The evolution is generally good, nevertheless the risk of recurrence remains considerable

Conclusion :

The particularity of our observation lies on the atypical clinical aspect (large size, multiple, atypical and multilobular localization), the age of the patient, as well as by its spectacular and rapid evolution after treatment.

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Abstract N°: 5111**Syndrome de Sneddon (Environ douze observations)**

Souad Choukri¹, Zakia Douhi¹, Nora Kalmi¹, Hanane Baybay¹, Sara Elloudi¹, Meryem Soughi¹, Fatima Zahra Mernissi¹

¹CHU Hassan II, Fez; Morocco, Service de dermatologie , Fes, Morocco

Introduction & Objectives:

Sneddon syndrome (SS) is a rare disorder, characterised by the association in the young person of a reticular livedo and recurrent cerebrovascular accidents (CVA), the annual incidence is four cases per million habitants. It usually affects women between the ages of 20 and 42 .

Our Objective was to study the epidemiological, clinical, paraclinical, and therapeutical features of this disorder through 12 observations.

Methods:

A retrospective study of the cases of 12 patients, collected in the dermatology and neurology departments at the Hassan II University Hospital in Fez, over the period from 2016 to 2022. All patients had a complete dermatological and somatic check-up, and an etiological work-up including cerebral imagery, a cardiovascular exploration, a skin biopsy, and an immunological assessment.

Results:

They were 12 women with age ranging from 30-50 years. The previous history was dominated by an ischemic vascular accident (2 cases), an abortion (1 case), a foetal death in utero (1 case).the clinic was polymorphic: dyspnea (1 case), arterial hypertension (3 cases), one of which was fortuitously discovered during hospitalization; isolated hemiplegia (6 cases) and one associated with facial paralysis and dysarthria , hemihypoesthesia (1 case). All the patients presented a non infiltrated and non-necrotic branching livedo, extended to the limbs, persisting in the heat, without other cutaneous or mucous signs. Imaging showed AVCI in 10 patients and was normal in two others. The remaining work-up revealed valvulopathy (2 cases), and anti-phospholipid antibodies (1 case), and skin biopsy showed Sneddon's syndrome in 3 patients, while it was not significant in the others.

Anticoagulation was started in one patient, ten others were put on aspirin for secondary prevention, and clinical monitoring was indicated in one patient.

Discussion:

Sneddon syndrome is an autoimmune disease more frequent in women; characterized by a multisystem involvement. Its physiopathology is not completely elucidated, but the presence of antiphospholipid antibodies in 50% of patients, as well as the findings of skin and brain biopsies, suggested a progressive disorder affecting the small and medium arteries of the skin and the brain. In addition, many other organs may also be involved in this disorder.

The clinical manifestations combine strokes, valvulopathy, and reticular livedo, preceding cerebral infarcts by many years, hence the interest of early detection as soon as the skin signs appear.

Hypertension is found in about 50% of patients.

The prevention of the cerebral infarction recurrence is based on antiplatelet drugs. Anticoagulants are suggested in case of association of Sneddon's syndrome with antiphospholipid syndrome .

Conclusion:

In the lack of any treatment, the Sneddon syndrome progresses to vascular complications, especially cerebral ones, which imposes a rapid diagnosis for a best therapeutic management.

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Abstract N°: 5178**Maladie d'Osler : le rôle du dermatologue**

Souad Choukri¹, Zakia Douhi¹, Nora Kalmi¹, Hanane Baybay¹, Sara Elloudi¹, Meryem Soughi¹, Fatima Zahra Mernissi¹

¹CHU Hassan II, Fez; Morocco, Service de dermatologie , Fes, Morocco

Introduction :

“Osler-Weber-Rendu” is a rare autosomal dominant genetic disorder characterized by muco-cutaneous and visceral telangiectasias due to abnormal vascular structures.

Despite the recent progress in the understanding of the genetics of the disorder and the determination of diagnostic criteria, the diagnosis is most often delayed, at the stage of complications

Observation :

A 23-year-old patient consulted for palmoplantar red lesions evolving since the childhood associated with an intermittent effort dyspnea and recurrent epistaxis, as well as a similar unexplored symptomatology in his father.

Clinical examination revealed a stage I dyspnea, with palmoplantar telangiectasias.

A thoracic angioscanner was realized, which showed a pulmonary arterio-venous malformation (AVM).

Therefore, the diagnosis of “Osler-Weber-Rendu” disease was retained, and the patient benefited from embolization of the pulmonary AVM, and a systematization check-up that did not show any additional visceral involvement.

Discussion:

“Osler-Weber-Rendu” disease, also called “Hereditary Hemorrhagic Telangiectasia” is due to a disturbance in the regulation of the Angiogenesis, which is the cause of multiple cutaneous-mucosal telangiectasias and visceral arterio-venous malformations mainly affecting the lungs, liver, digestive tract and the brain. (1)

The diagnosis of this condition is based on the clinical criteria of spontaneous recurrent epistaxis, cutaneous-mucosal telangiectasia, and visceral vascular malformations, as well as family history. The presence of three out of four criteria confirms the diagnosis (1-2). Our patient had enough criteria to retain the diagnosis.

Extra-cutaneous involvement can lead to severe complications. In particular, embolic strokes and transitory ischemic accidents that often occur in patients who have clinically silent pulmonary or cerebral AVMs, indicating the necessity of an early diagnosis and treatment (2).

Conclusion:

Even though the skin manifestations of this pathology are regarded as benign, the dermatologists' role is still indispensable in early identification of the pathology in order to allow timely multidisciplinary, preventive and curative management.

Abstract N°: 5591**Vulvar angiokeratoma after radical hysterectomy and radiotherapy: a case report**Hafsa Hamraoui¹, Sara Elloudi¹, Hanane Baybay¹, Douhi Zakia¹, Meryem Soughi¹, Fatima Zahra Mernissi¹, Layla Tahiri²¹CHU Hassan II, dermatology, Fez, Morocco, ²CHU Hassan II, anathomopathology, Fez, Morocco

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Introduction & Objectives:

Angiokeratomas are rare vascular malformations whose surface is keratotic. Their localization at the vulvar level called angiokeratoma of Fordyce. Appear primarily on the labia majora and more rarely on the clitoris they may appear as solitary or multiple, unilateral or bilateral lesions, however most are multiple, unilateral, and located on the left side of the vulva, especially in premenopausal women. These benign vascular tumors may have a smooth, papular, nodular, lobulated, or verrucous morphology, with colors varying from violet, red, brown, blue-black, and gray. Dermoscopic signs suggestive of angiokeratoma are not very specific in isolation but include the presence of red to purplish ovoid lacunae, a white veil partially covering the lesion. Histologically, these are dilated subepidermal vessels, associated with epidermal hyperplasia such as acanthosis or hyperkeratosis. Conditions presumed to be associated with angiokeratomas include pregnancy, post-hysterectomy, chronic inflammation, radiation therapy.

We report the case of a vulvar angiokeratoma after hysterectomy and radiotherapy, we illustrate also the value of dermoscopy coupled with histology in the diagnosis of this pathology

Materials & Methods:

This is a clinical case of whose diagnosis was based on clinical and dermoscopic with histological confirmation.

Results:

This is a 54-year-old patient with a history of squamous cell carcinoma of the cervix 12 years ago, treated by hysterectomy with bilateral iliac dissection, radiotherapy and chemotherapy, 2 years ago she reported the appearance of lesions pruriginous at the genital level. Clinical examination revealed a polylobed erythematous plaque made of small confluent translucent papules on two labia majora, initially we thought of a vulvar localization of a carcinoma, Buschke-Löwenstein tumor or condyloma but the dermoscopic examination showed red lacunae, a white veil, and hypopyon aspect, which rectified the diagnosis to an angiokeratoma or microcystic lymphangioma. Anatomopathological examination of the lesion revealed a papillomatous epidermis surmounted by orthokeratotic hyperkeratosis with numerous dilated vessels in the papillary dermis concluding in an angiokeratoma then the patient received CO₂ laser sessions with good improvement.

Conclusion:

Vulvar angiokeratoma is often misdiagnosed as genital warts or a malignant vascular tumor. Women who have had radiation therapy to the area may be at increased risk of developing such a lesion, dermoscopy orients the diagnosis, which requires histological confirmation.



Abstract N°: 5598**Angiolymphoid hyperplasia with eosinophilia in an unusual location?- difficult diagnosis.**

Jowita Sroka-Tomaszewska*¹, Jakub Żółkiewicz¹, Monika Sikorska¹

¹Medical University of Gdańsk, Department of Dermatology, Venereology and Allergology, Gdańsk

Angiolymphoid hyperplasia with eosinophilia in an unusual location?- difficult diagnosis.

Introduction & Objectives:

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a benign neoplastic growth of blood vessels. Lesions are usually located head and neck region, however, they may appear in other areas or affect extracutaneous tissues. It usually presents with solitary or multiple papules or nodules involving mostly the head and neck region and rarely encompasses other parts of the body. Histological examination shows clusters of proliferating capillaries with plump, swollen endothelial cells and cellular infiltrate composed mainly of lymphocytes and large numbers of eosinophils seen around the blood vessels. Despite ALHE belonging to the spectrum of benign vascular hyperplasias, lesions tend to be recurrent. Early age of onset, long disease duration, multiple lesions, and symptomatic disease seem to have a higher recurrence rate. Spontaneous regression can occur occasionally. Herein, we present a rare case of the patient, who was diagnosed with ALHE in unusual localization.

A 50-year-old woman presented in the Dermatology Outpatient Clinic with an odorous, ulcerated hyperkeratotic tumour of the right big toe. The patient reported that the lesion appeared several years ago and was gradually increasing. The lesion has not been previously diagnosed. Due to the non-healing nature, ulceration and neoplastic growth, including angiosarcoma, lymphoma or squamous cell carcinoma, were primarily taken in the differential diagnosis. To establish the diagnosis, a skin biopsy was performed. Histopathological examination ruled out the neoplasm. Based on the clinical and microscopic picture, angiolymphoid hyperplasia with eosinophilia was diagnosed. The patient was referred to a surgical oncologist and eventually, the toe was amputated. However, in the last histopathological examination of the entire lesion, the diagnosis was not confirmed, and consultation of the histopathological block was requested.

Materials & Methods:

A retrospective analysis of a patient's medical records diagnosed with a non-healing tumour of the right big toe was performed.

Results:

Based on the clinical and microscopic picture, the diagnosis of angiolymphoid hyperplasia with eosinophilia is most likely.

Conclusion:

Angiolymphoid hyperplasia with eosinophilia (ALHE), also known as epithelioid hemangioma, is an inflamed vascular tumefaction of unknown cause, however, reactions to trauma and insect bite are postulated to contribute to the disease development. In the course of the disease lesions usually appears in the head and region of skin. Sometimes cases of the disease are reported in other locations. Not only diagnosis but also choosing the right treatment modality is challenging in this condition. The treatment of the first choice is surgical resection. Regardless of the procedure, relapses are very common. Regardless of the treatment regimen, relapses are very common. In the case of non-healing wounds, underlying neoplasm should be always excluded.

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Abstract N°: 5683**Leg ulcer: A case report of an atypical presentation of Buerger's disease**

Hajar Boumechdi¹, Yassine Zerouali¹, Ouafa Hocar¹, Said Amal¹, Maryem Aboudourib¹

¹mohamed VI university hospital, dermatology and venereology, marrakech, Morocco

Introduction & objectives:

The leg ulcer is defined as a loss of substance that involves the epidermis and the papillary layer of the dermis that can extend into the subcutaneous tissue and always appears in the presence of pathological underlying tissues.

The etiologies are multiple, namely vascular, immunological, infectious, neoplastic and paraneoplastic.

The etiology of the ulcer will define the therapeutic management. Complementary codified examinations generally make it possible to establish the diagnosis.

We report the case of a patient with a leg ulcer revealing Buerger's disease.

Case descriptions:

We report the case of a 50-year-old patient, chronic smoker with 40 PA, not known to be diabetic or cardiac, who presented with a leg ulcer that had been evolving for 2 months following a scarification. The clinical examination revealed a painful ulceration measuring 7 cm in length, with a small circular ulceration on the knee. A 3-part biopsy showed an aspect in favor of Pyoderma Gangrenosum . An exhaustive paraclinical workup was requested (biological, immunological, serologies, tumor markers, thrombophilia) without anomalies. However, arterial doppler ultrasound and angiography scan showed a total thrombosis of the subrenal abdominal aorta extended to the iliac bifurcation and of the two internal iliac arteries, bilateral deep venous thrombosis, all of which suggested Buerger's disease. In addition to smoking cessation, the patient was put on double antiplatelet aggregation and ACE inhibitors then benefited from an aortoiliac stent. The evolution was marked by the necrosis of the foot then the leg, and the patient underwent an amputation of the leg.

Discussion:

Buerger's disease or thromboangiitis obliterans is an inflammatory, segmental and occlusive disease affecting the small and medium caliber arteries and veins of the extremities of the limbs. This pathology mainly affects young men under 50 years of age and smokers. Its etiology remains unknown and smoking seems to play a major aggravating role. Immunological factors could be involved and recently an alteration of the endothelium-dependent vasodilatation has been highlighted. The diagnosis remains a diagnosis of exclusion and is based on the history of the disease, clinical abnormalities, and arteriographic appearance, although none of these aspects is specific. Raynaud's phenomenon, digital arteriopathy, and migrating superficial thromboses are arguments in favor of the diagnosis. Smoking cessation remains the most important factor in improving the prognosis of this disease, for which there is currently no effective long-term pharmacological treatment.

Conclusion:

The etiology of Buerger's disease is unknown, but the role of smoking is preponderant in the onset and evolution of the disease. Our observation underlines the interest of vascular imaging for any leg ulcer.

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Abstract N°: 5688**is topical timolol effective and safe in the therapy of infantile hemangioma and nevus simplex ?**Inaya Mahmoud^{*1, 2}, Nikolai Tsankov^{1, 2}, Mariela Hitova¹, Jana Kazandjieva^{1, 3}¹EuroDerma Clinic, Dermatology and Venereology , Sofia, Bulgaria,²Acibadem City Clinic Tokuda University Hospital, ³Medical University of Sofia**Introduction & Objectives:**

Infantile hemangiomas (IH) result from benign proliferation of endothelial cells and can impair vital or sensory functions. IH are the most common vascular tumors in infancy and can even cause disfigurement. Nevus simplex is a benign capillary malformation, present in approximately 40% of newborns. The safety and efficacy of topical timolol cream in the therapy of infantile hemangioma and nevus simplex have not been evaluated in a clinical study. Our goal was to assess topical timolol's effectiveness, safety and document reported adverse events if present.

Materials & Methods:

A clinical study of 30 infants, aged between 5 weeks and 3 months, with small superficial hemangioma with confirmed growth in the last 2 weeks, not > 4 cm in size, localized in body areas excluding the central parts of the face and ears, the chest, mid lumbo-sacral region and 10 infants with nevus simplex were treated with topical timolol 1% cream twice daily for the time period of two months.

Results:

Topical timolol 1% cream was well tolerated in the thirty infants with superficial hemangioma and in the ten infants with nevus simplex. There were no side effects observed. It demonstrated best response to small superficial infantile hemangioma and proved to be a safe treatment option with good effectiveness.

Conclusion:

We present a clinical study that assessed the effectiveness and safety of timolol 1 % cream in the treatment of infantile haemangioma and nevus simplex. It is difficult to say whether the effect of topical timolol doesn't coincide with the natural figurative involution of nevus simplex. However, we had good and fast results.



Abstract N°: 5702**Successful treatment of propranolol resistant infantile hemangioma with atenolol: a case report**Insaf Moubine*¹, Fouzia Hal¹, Soumia Chiheb¹¹CHU Ibn Rochd, Dermatology, CASABLANCA**Introduction & Objectives:**

Infantile hemangioma (IH) is the most common pediatric vascular tumor. Since the first case report in 2008 of an IH responding to propranolol, it has become the first-line treatment when systemic therapy is needed, but failure may occur. We describe a case of IH that was refractory to therapy with propranolol and subsequently responded to atenolol.

Materials & Methods:**Results:**

A 3-month-old child with no prior medical history presented with an angiomatous swelling on the face that had been evolving since the age of 15 days. Clinical examination found a bilateral jugal swelling with multiple angiomatous lesions, along with an irregular swelling of the lower lip with a central fibrous ulceration scar. The patient was diagnosed with infantile hemangioma and started on propranolol at a dose of 3 mg/kg/day. There was no improvement after 4 months of treatment. Thus, sirolimus at a dose of 0.8 mg/m² twice a day was added. After 15 days of sirolimus treatment, the patient developed febrile respiratory failure with diarrhea and vomiting. A thoracic CT scan was performed, revealing interstitial pneumonia, which led to the discontinuation of treatment. Propranolol was maintained, but the patient continued to experience episodes of wheezing, requiring repeated discontinuation of the drug. It was therefore decided to replace propranolol with atenolol at a dose of 2.5 mg/kg/day, resulting in a marked regression of the subcutaneous component after 3 months of treatment.

Conclusion:

The precise mechanism by which betablockers induce accelerated involution of IH remains to be fully elucidated. Proposed mechanisms of action include vasoconstriction, inhibition of angiogenesis, and induction of apoptosis in proliferating endothelial cells. Globally, propranolol, a non-selective adrenergic receptor blocker, is the first-line treatment for problematic proliferating IH but is associated with fairly common adverse effects. Many of these adverse effects are attributable to its non-selective action and its interaction with two receptors. Other therapeutic alternatives include other systemic blockers, such as selective 1-blockers such as atenolol. Several studies have shown similar response rates and overall efficacy between propranolol and atenolol for the treatment of IH. 1 mg of atenolol equates to approximately 1.6mg of propranolol, and due to its longer half-life, atenolol offers the benefit of once-daily dosing.** Atenolol remains a therapeutic alternative to consider in cases of major side effects such as bronchial hyperactivity, hypoglycemia, and agitation, which may lead to poor adherence to treatment and therefore a lack of response.

Abstract N°: 5749

Successful Treatment of Angiolymphoid Hyperplasia with Eosinophilia by Propranolol

Kaoutar Elmachichi¹, Handi Oumaima², Amal Said², Hocar Ouafae³, Maryem Aboudourib²

¹Chu Mohamed Vi Marrakesh , DERMATOLOGY, Marrakech, Morocco,²Chu Mohamed Vi Marrakesh , DERMATOLOGY, ³Chu Mohamed Vi Marrakesh , dermatology

Successful Treatment of Angiolymphoid Hyperplasia with Eosinophilia by Propranolol.

Introduction & Objectives:

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare idiopathic condition, usually seen in adults and characterized by the presence of isolated or grouped papules, plaques or nodules in the skin of the head and neck region, specifically on the preauricular region. In the view of management, majority of related reports of therapeutic methods comprise of surgical excision, intralesional/topical/systemic corticosteroid...

In some cases, the oral propranolol was demonstrated as an alternative treatment of ALHE. Here we reported a case of a 30-year-old woman with ALHE successfully treated with propranolol.

Materials & Methods:

A 30-year-old woman, without past medical history was admitted with multiple painful papules, appeared 7 months ago, spontaneous bleeding in the auricle and the preauricular area

Physical examination revealed multiple red-brown firm dome-shape papules in the helix and antihelix, without any other cutaneous lesions. There was no local or regional lymphadenopathy, and other system examination were unremarkable and the complete blood count was normal.

Surgical removal of one of the lesions was performed and pathological examination confirmed the histological diagnosis of ALHE.

The patient was started on oral propranolol 40 mg once daily. Within 2 months, several of the lesions had decreased. Propranolol was subsequently stopped within 8 months of initiating treatment after a complete disappearance of all lesions and without any adverse effects of the medication.

Results:

Angiolymphoid hyperplasia with eosinophilia is a benign, locally proliferating disease but the pathogenesis is not clearly understood whether it consists of a vascular neoplasm or a lymphoproliferative process.

ALHE commonly present with some symptoms, like pruritus, spontaneous bleeding and pain. This is consistent with our patient that had intense pain and bleeding. It tends to arise in the skin of the head and neck, particularly in the scalp and the periauricular area. Other features include regional lymphadenopathy and peripheral eosinophilia which is an inconstant feature as in our case.

Histologically, it is characterized by proliferation of vascular channels with inflammatory infiltrate composed of lymphocytes and eosinophils.

However, from previous case reports, surgical excision is considered as the most effective. Moreover, some reported cases showed the interesting successful treatment by oral propranolol. Although the definite mechanism

remains unclear, the hypothesis has been proposed that beta-blocker-induced localized vasoconstriction, inhibition of angiogenesis and apoptosis of capillary endothelial cells.

Conclusion:

Our case, shows that oral propranolol is one of the good therapeutic options in the treatment of ALHE with an excellent improvement in clinical presentations and symptoms and without any adverse effect.

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Abstract N°: 5842**Orbital cavernous hemangioma in a child: a rare pediatric case**Hajar Moata¹, Fouzia Hal¹,¹, Soumia Chiheb¹¹Ibn Rochd university hospital, DERMATOLOGY AND VENEROLOGY, Morocco**Introduction :**

Orbital cavernous hemangiomas are benign, congenital vascular tumors. They are the most common tumors of the posterior cone of the **orbit** in adults . In the opposit ,they are rare in children. We report an unusual pediatric case of cavernous hemangioma .

Observation :

A 4-year-old girl presented for an exophthalmos of the right eye that had been evolving for 2 years without any notion of decreased visual acuity. The examination revealed painless unilateral non reducible exophthalmos of the right eye, with Swelling of the right cheek without any inflammatory signs. Cerebroorbital MRI shows a right orbital vascular malformation suggestive of a right orbital cavernous hemangioma with bilateral temporal cortical atrophy. A surgical treatment in double team (ophthalmologist and neurosurgeons) was proposed.

Conclusion:

Cavernous hemangiomas are benign tumors of the orbit in young adults.They are exceptional in children where vascular tumors are dominated by capillary hemangiomas. These tumors are most often unilateral, of variable location and volume. Treatment is surgical for symptomatic forms, but the topography of the lesion and its contact with the intraconal vascular and nervous elements are the main difficulty



Abstract N°: 5952

Successful reduction of intramuscular cavernous hemangioma following corticosteroid injections: a case report

Fatima Ezzahraa Chekairi¹, Fouzia Hal¹, Soumiya Chiheb¹

¹Ibn Rochd University Hospital Center, Dermatology and venereology, Casablanca, Morocco

Introduction & Objectives:

Intramuscular hemangioma (IMH) is a rare and benign vascular tumor that represents 1% of all hemangiomas, it usually appears in the skeletal muscles of the limbs or trunk.

Complete surgical resection is the reference approach; however, it remains challenging due to the location of the IMH. Various non-surgical treatments have been described in the literature but results remain inconclusive and controversial.

We herein report a case of cavernous hemangioma in the right trapezius muscle successfully reduced after corticosteroid injections.

Materials & Methods:

Results:

A 22-year-old male patient with no particular history, presented a discomforting mass in the right side of this back especially during muscle contraction at work, evolving since childhood with a progressive size increasing.

Clinical examination showed an enlarged mass in the right side of this back with a bluish facing skin. On palpation, the tumor's consistency was relatively firm and soft, compressible and mobile, measuring 10 cm on its long axis x 5 cm. No pulsation was found nor thrill on auscultation.

Echo-Doppler revealed the vascular nature of the mass and histopathological study of a muscular biopsy confirmed the diagnosis of intramuscular cavernous hemangioma.

Magnetic resonance imaging (MRI) showed a well-encapsulated oblong heterogeneous mass composed of large vessels lined by flattened endothelial cells within the right trapezius muscle.

The patient received one intralesional injection of corticosteroids every month with significant decreasing in size and infiltration of the tumor measuring 7 x 4 cm after 3 months of treatment versus 10 x 5 cm initially, with no side effects nor complications during injections.

Conclusion:

Our present case of successful reduction of intramuscular cavernous hemangioma suggests that intralesional corticosteroid injections could be an interesting and very useful preoperative approach making the curative resection easier by reducing the large size of an IMH.

Therefore, preventing unnecessary and severe complications of an invasive surgical procedure that requires a wide excision, and insure complete and definitive IMH removal.

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Abstract N°: 6287**Unraveling the Similarities: Cutaneous Polyarteritis Nodosa Masquerading as Calciphylaxis**Inês Pereira Amaral^{*1}, Ivânia Soares¹, Madalena Pupo Correia¹, Paulo Filipe¹¹Centro Hospitalar Universitário Lisboa Norte, Dermatology, Lisbon, Portugal**Title:** Unraveling the Similarities: Cutaneous Polyarteritis Nodosa Masquerading as Calciphylaxis**Introduction & Objectives:**

Cutaneous polyarteritis nodosa (cPAN) is a rare form of vasculitis primarily affecting small-to-medium-sized arteries of the skin, of unknown etiology. It presents as a diagnostic challenge due to its resemblance to various dermatological conditions, including calciphylaxis.

Materials & Methods:

We describe a case of a 75-year-old woman, with end-stage renal disease, who presented with a three-month history of painful skin lesions on her lower extremities. After extensive evaluation, the diagnosis of cutaneous polyarteritis nodosa was made.

Results:

A 75-year-old woman was admitted to the hospital with a three-month history of multiple subcutaneous and erythematous nodules, with a plaque of necrosis that slowly progressed to form well-defined ulcers, on both thighs. The patient was complex, with multiple comorbidities, such as heart failure due to valvular and hypertensive heart disease, with a mechanical aortic valve prosthesis, and an end-stage renal disease in a single kidney patient. The clinical presentation raised concerns of calciphylaxis, given its association with chronic kidney disease. However, a thorough evaluation including laboratory investigations and imaging studies was normal, thereby prompting further investigation. An incisional biopsy was performed, demonstrating the presence of necrotizing vasculitis involving small-to-medium-sized arteries in the dermis, consistent with a diagnosis of cutaneous polyarteritis nodosa. The patient's clinical features, including the absence of systemic involvement, supported the classification of limited cPAN.

The patient initiated oral prednisolone with slow dose tapering. The ulcers showed gradual healing with the treatment and the patient experienced significant symptomatic relief.

Conclusion:

Cutaneous polyarteritis nodosa is a rare variant of polyarteritis nodosa primarily affecting the skin. Its clinical presentation can mimic other dermatological conditions, posing a diagnostic challenge. Calciphylaxis, characterized by ischemic skin necrosis due to microvascular calcification, shares similarities with cPAN, particularly in patients with chronic kidney disease. However, certain clinical and laboratory findings can help differentiate between the two conditions. Accurate diagnosis through incisional biopsy and histopathological examination is essential for appropriate treatment selection and avoidance of unnecessary interventions. This case highlights the importance of considering cutaneous polyarteritis nodosa as a potential diagnosis in patients presenting with cutaneous erythematous nodules with necrosis, particularly when other mimickers are suspected. Improved awareness and understanding of cPAN will facilitate timely diagnosis and optimized management strategies, ultimately improving patient outcomes.

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