

Unveiling Granulomatosis with Polyangiitis: Uncommon Onset as an Umbilical Pyoderma Gangrenosum-like Ulcer

Špela Šuler Baglama¹

¹University Medical Centre Ljubljana, Clinic of Dermatovenereology, Ljubljana, Slovenia

Unveiling Granulomatosis with Polyangiitis: Uncommon Onset as an Umbilical Pyoderma Gangrenosum-like Ulcer

Introduction & Objectives:

The abstract uncovers the rare scenario of granulomatosis with polyangiitis (GPA) starting as an umbilical pyoderma gangrenosum-like ulcer. It stresses the vital role of dermatologists in recognizing and diagnosing such unusual cases, urging for a broad approach to differential diagnosis in chronic ulcers to avoid diagnostic delays.

Materials & Methods:

Results:

This abstract presents a challenging case of a 57-year-old patient with an 8-month persistent chronic umbilical ulcer resistant to conventional treatments. The condition escalated into a multifaceted clinical scenario a month before referral, marked by painful hypertrophic gums, epistaxis, and inflammation of the left lower eyelid. The examination revealed a 4.5 x 3 cm undermined ulcer in the umbilical area, characterized by a violaceous border, reaching a depth of 3 cm, with minimal granulation and 80% fibrin coverage (Figure 1). The left lower eyelid displayed granulation tissue with ulceration and purulent discharge (Figure 2), while gums exhibited strawberry-like hypertrophy (Figure 3). Additional otorhinolaryngological examination revealed necrosis and distinct granulations in the right middle concha and lower alveolar ridge. Diagnostic investigations, including biopsies and positive proteinase 3-antineutrophil cytoplasmic antibody (PR3-ANCA), identified GPA. Treatment involved 1 mg/kg prednisolone equivalent/day orally and topical measures for ulcer care. Subsequent referral to rheumatology for rituximab treatment was initiated. Regrettably, feedback on the efficacy of this treatment is unavailable.

Conclusion:

GPA, a rare pauci-immune ANCA-associated systemic vasculitis, typically affects small- to medium-sized vessels and often causes prodromal symptoms preceding specific manifestations (1). Although cutaneous involvement is common, the initial presentation with pyoderma gangrenosum-like ulcer, as in our case, is rare. ANCA positivity and distinct histological features serve as crucial diagnostic markers between GPA and pyoderma gangrenosum. It is essential to further differentiate GPA ulcerations from infections, connective tissue diseases, extraintestinal Crohn's disease and certain cutaneous lymphomas (2). Other possible cutaneous manifestations of GPA are palpable purpura, mucocutaneous ulcers, subcutaneous nodules, digital necrosis, papulonecrotic lesions, and livedo reticularis (3).

To sum up, recognition of atypical GPA, including cutaneous presentations, is vital for prompt rheumatologist referral. Collaborative care is essential for thorough diagnostics and tailoring systemic treatment based on GPA severity.



Chronic Scrotal and Penile Ulcers: A Diagnostic Challenge Culminating in Juvenile Gangrenous Vasculitis

Hetav Pandya*^{1, 2}, Krishnakant Pandya²

¹Care multispeciality hospital, Dermatology - Venereology, Pune, India,²The Rejuvene Clinic (Dr. K. B. Pandya), Dermatology - Venereology, Rajkot, India

Chronic Scrotal and Penile Ulcers: A Diagnostic Challenge Culminating in Juvenile Gangrenous Vasculitis

Introduction & Objectives:

Juvenile Gangrenous Vasculitis of the Scrotum (JGVS) is a rare vasculitis that presents with acute, often painful, scrotal ulcerations in young males. It is frequently misdiagnosed as sexually transmitted infections (STIs) due to the genital involvement, leading to a potential delay in appropriate treatment. The objective of this detailed report is to provide an indepth clinical characterization of JGVS, outline the diagnostic approach, and advocate for increased awareness among GPs and specialists to facilitate timely and accurate diagnosis and management.

Materials & Methods:

A 19-year-old male patient presented with a 4-year history of recurrent scrotal ulcers, characterized by intermittent episodes of inflammation, pain, and partial healing. The lesions varied in appearance, with some presenting as erythematous bases with irregular, raised borders, and others as deep, necrotic centres with black eschars, indicative of underlying vasculitis and tissue necrosis. A comprehensive evaluation was undertaken, including detailed STI screening panels, complete blood counts, biochemistry profiles, autoantibody screenings, histopathological examination of the ulcer biopsy, and scrotal ultrasonography. Special attention was given to the patient's sexual history and potential exposure to risk factors for STIs.

Results:

The patient's serological tests for common STIs, including HIV, syphilis, herpes simplex virus, and chlamydia, were negative. Additionally, the patient's sexual history was non-contributory, as he reported no recent sexual activity or high-risk behaviours. The elevated SGPT level hinted at systemic involvement, but liver ultrasonography revealed no abnormalities. The biopsy displayed classic hallmarks of JGVS, with epidermal necrosis, swollen vessel walls in the dermis, and infiltration by neutrophils and eosinophils, without evidence of granuloma or microbial pathogens. The scrotal ultrasound showed increased vascularity and oedematous changes without signs of deeper tissue involvement. A conservative treatment approach was adopted, comprising of systemic anti-inflammatory therapy with prednisolone, pain management with a combination of tramadol and acetaminophen, and topical antibiotic treatment with Fusidic acid to prevent secondary bacterial colonization. The patient was advised on regular wound care with non-adherent dressings. Follow-up over a sixweek period demonstrated significant regression of the ulcers and symptomatic relief.

Conclusion:

This case illustrates the clinical complexity and the diagnostic challenge posed by JGVS, especially in differentiating it from STIs and other causes of genital ulcers. The findings underscore the critical need for GPs and specialists to be cognizant of JGVS in the differential diagnosis of persistent genital ulcers. Through detailed patient history, careful exclusion of STIs, and corroborative histopathological examination, an accurate diagnosis can be established, allowing for suitable management and avoidance of unnecessary surgical interventions. Moreover, this case highlights the importance of considering a broader clinical spectrum for JGVS and the potential for more chronic and relapsing courses, advocating for ongoing research and education in this domain.



From Paper to Pixels: Digitalisation of Patient Information Leaflets in Vascular Dermatology

Jo-Yve Wong^{*1}, Myranda Attard¹, Rebecca Mckillen¹, Pavithren Sellathurai¹, Susannah Hoey¹

¹Royal Victoria Hospital, Belfast, United Kingdom

Introduction & Objectives:

Patient information leaflets are widely used in vascular dermatology clinics, playing a pivotal role in providing information to patients on their diagnoses and treatment options available. However, conventional leaflets are often lengthy and printed onto several pages. In line with the growing shift towards environmental sustainability and greener healthcare, our quality improvement initiative aimed to streamline and reduce the number of pages given, with a secondary aim of improving patient satisfaction and accessibility of information.

Materials & Methods:

Data on the number of printed pages of patient information leaflets distributed at paediatric and adult dermatology clinics, as well as vascular anomaly multi-disciplinary team clinics, was prospectively collected from a single tertiary centre between October 2023 to January 2024. Patient questionnaires were also distributed, including questions assessing how accessible it was for patients to use QR (quick-response) codes, how satisfied patients were with the information received, preferences between using paper or QR codes, barriers to using QR codes if any, and other suggestions for improvement. Interventions for change were rolled out in stages, including collating leaflets into a single page of QR codes (Figure 1), reducing print sizes from A4 to A5, and updating sclerotherapy leaflets with plain reader feedback. The QR code page included links to leaflets on adult sclerotherapy, paediatric sclerotherapy, vascular malformations, lymphatic malformations, infantile haemangiomas, propranolol for haemangioma of infancy, vascular birthmarks, port wine stain, and topical timolol.

Results:

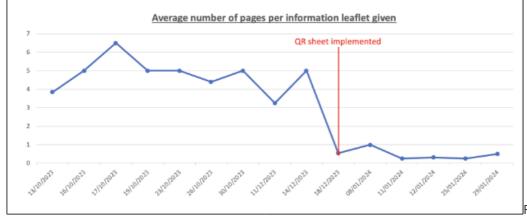
Initial average pages per leaflets given was 3.83, this reduced by 87% to 0.50 following interventions (Figure 2). Baseline patient satisfaction was 8.25 (on a scale from 0 to 10, with 10 indicating most satisfied), increasing by 19% to 9.8 following interventions. Patients found QR codes accessible, rating it on average 9.1 (on a scale from 0 to 10, with 10 indicating most accessible). Overall, 76% of patients preferred using QR codes over paper handouts to receive information. Feedback commended the ease of accessibility to leaflets using the QR sheet, although suggested that potential barriers to use may include those without access to a smartphone, internet, or those with difficulties using technology.

Conclusion:

In conclusion, digitalising patient information leaflets into QR code format is an effective method of reducing paper printing and encouraging sustainability, while maintaining patient satisfaction and accessibility of information. The shift towards greener practices highlights the importance of leveraging technology in healthcare. Moving forwards, addressing potential barriers to QR code adoption will be essential for ensuring equitable access to information for all patients.

VASCULAR DERMATOLOGY LEAFLETS SCAN THE QR CODE BELOW WITH YOUR CAMERA TO GAIN A BETTER UNDERSTANDING Adult sclerotherapy Vascular malformat Paediatric sclerotherapy Propranolol for haemangioma of infancy Lymphatic malformation Infantile haemangioma Vascular birthmarks Port Wine Stain cal timolo

Figure 1 (Vascular Dermatology QR sheet):



H

Figure 2 (Run chart of average

number of pages per information leaflet given):



Ulcerated infantile haemangiomas and intense pulsed light

Salma Zakaryaa¹, Fouzia Hali¹, Bouchra Baghad¹, Soumiya Chiheb¹

¹Ibn Rochd university hospital center, Dermatology and venereology, Casablanca

Introduction & Objectives:

Infantile hemangiomas are the most common benign tumors in childhood. It is diagnosed clinically; the standard treatment is propranolol, and ulceration is one of its most worrying complications.

Lasers and intense pulsed light (IPL) have multiple indications in dermatology for the treatment of vascular lesions, particularly in the case of ulcerated infantile hemangiomas.

Materials & Methods:

Descriptive study collecting patients who presented with ulcerated infantile hemangiomas over a period of 6 years from January 2017 to December 2023 at the dermatology and venereology department of Ibn Rochd University hospital in Casablanca.

Results:

Forty cases of infantile hemangiomas that had benefited from vascular lasers were included, of which 30 cases (75%) were female and 10 cases (25%) male, with a sex ratio of 3:1.

The mean age was 5 months, ranging from 1 month to 24 months.

Hemangiomas were located on the face in 15 cases (37.5%), on the perineal region in 15 cases (37.5%) and on the limbs in 10 cases (25%).

The average ulceration size was 2 cm and the average number of IPL sessions was four.

Pain subsided on average after 7 days from the first IPL session and healing was complete after an average of 4 weeks.

Conclusion:

Any infantile hemangioma can ulcerate and compromise the infant's aesthetic and functional prognosis, becoming a therapeutic emergency.

In fact, these ulcerations may cause intense pain, secondary infections and bleeding, with feeding difficulties when they are located on the labia, thus representing the only true "urgent" indication for dermatological lasers.

Pulsed dye laser treatment has a role to play in the management of ulcerated infantile hemangiomas.

Our study confirms, in the absence of PDL, that the IPL source (with good parameter management) is an effective alternative in the treatment of ulcerated infantile hemangiomas.



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

Saliha Jebbouje¹, Fouzia Hali¹, Bouchra Baghad¹

¹chu Ibn Rochd Casablanca , dermatology

Introduction & Objectives:

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

Case report:

A 37 years-old women presented severe joint ache in her left knee and ankle joints. She had a feeling of numbness in her left leg and hand for the last month. Personal medical history was positive for a diagnosis of asthma for the last three years treated with salbutamol inhaler and anti-histamine medication. The patient also reported a recent weight loss of more than 4 kg and episodes of recurrent respiratory infections treated with unspecified systemic antibiotics. Skin examination showed a painful infiltrated plaque affecting the surface of the left thigh for the last 6 weeks. The skin biopsy revealed multiple foci of vasculitis, characterized by parietal fibrinoid necrosis and eosinophil infiltration, were present in the mid-dermis and hypodermis, consistent with eosinophilic necrotizing polyangiitis. Laboratory investigations showed a notable peripheral blood eosinophilia, however, cytoplasmic ANCA (C-ANCA) and perinuclear ANCA (P-ANCA) were negative. X-ray of the lungs showed bilateral infiltrations and hilar involvement. A high-resolution CT chest-abdomen showed subtle centrilobular ground-glass nodules along the bronchovascular structures and minimal peritoneal effusion. Electroneurography reported a length-dependent, sensory-motor polyneuropathy. These findings were in consistence with the diagnosis of EGPA. The patient was started on prednisone 1mg/kg/day and cyclophosphamide pulse 0.75g per month. Neurological symptoms diminished gradually and the number of eosinophils normalized.

Conclusion:

Our patient illustrated a EGPA with multisystemic involvement . Indeed, EGPA is a type of ANCA-associated vasculitis that predominantly affects small- and medium-sized vessels of many organs simultaneously. Dermatologic manifestations are estimated to occur in around 50% of patients, representing a relatively frequent type of involvement. ANCA is negative in most EGPA patients. A therapeutic regimen involving corticosteroids and immunosuppressants is effective in preventing this disease from progression and sometimes leading to remission.



Leriche syndrome revealed by unilateral cutaneous necrotic ulcerations

Meryem Khallouki¹, Bendaoud Layla¹, Sarah Zemrani¹, Maryem Aboudourib¹, Ouafa Hocar¹, Said Amal¹

¹Mohammed VI University Hospital of Marrakech, Department of Dermatology and Venereology, Marrakech, Morocco

Introduction & Objectives:

Leriche syndrome (LS), also commonly known as aortoiliac occlusive disease (AIOD), is a product of atherosclerosis affecting the distal abdominal aorta, iliac arteries, and femoropopliteal vessels. LS, first described by Leriche and Morel in 1924, presents itself more frequently as chronic claudication, erectile dysfunction, and absent femoral pulses. We present a patient with an atypical presentation of painful unilateral necrotic ulcerations of the lower limb, revealing LS and ischemic cardiopathy.

Case report:

A 70-year-old male patient presented with a 2-month history of deep, painful, non-healing ulcerations on the right lower limb. The patient had a history of smoking for the past 30 years, non-treated hypertension, unrecognized chest pain, intermittent claudication, and paresthesia of the lower limbs. Physical examination revealed multiple stepped necrotic ulcerations of the right lower limb, livedo reticularis, and local purpuric lesions. The arterial pulsations of the right tibial arteries were reduced; no pulsations were found over the right femoral artery. Arterial echo-Doppler of the lower limbs showed diffuse obliterative arteriopathy with multiple calcified atheromatous plaques of the aorto-iliac axis and the arteries of both lower limbs. The CT angiography showed Leriche syndrome with total occlusion of the abdominal aorta associated with bilateral occlusion of the iliac arteries. Coronary angiography showed tri-truncular lesions with stenosis of the middle anterior interventricular artery, middle right coronary artery, and circumflex artery. Revascularization was indicated, but the patient died of cardiogenic shock.

Discussion:

LS is a rare and critical complication of peripheral arterial disease (PAD), caused by atherosclerosis. The prevalence of LS is unknown given that many cases are asymptomatic, but PAD, the etiological agent of Leriche syndrome, has a prevalence of approximately 115 million worldwide, with 70,000 deaths in 2019. Dyslipidemia, male gender, smoking, diabetes mellitus, and hypertension are the main risk factors. Cutaneous manifestations varied from nonspecific inflammatory lesions through ulcers to gangrene of low extremities, and the diagnosis might be difficult. Patients report severe intermittent claudication, ischemic rest pain, or burning and pain in the cutaneous lesions; they may initially have no symptoms. In some cases, non-specific symptoms, such as stiffness, paresthesia, reduced sensitivity or pain, and claudication, appear. A case with similar clinical features to our patient reported multiple deep and painful ulcers with necrotic borders along the lower extremities, revealing Leriche syndrome. Another case involving a woman with perineal ulcers has recently been reported in association with AIOD. Patients with critical limb ischemia had a significantly higher relative risk for myocardial infarction, major amputation, cardiovascular mortality, major adverse cardiac events, and allcause mortality. Unfortunately, our patient died from a myocardial infarction.

Conclusion:

This case illustrates the importance of proper history-taking and physical examination with careful examination of peripheral pulses of both lower limbs in patients presenting with multiple non healing cutaneous ulcerations in order to avoid misdiagnosis of AIOD.





Atypical presentation of spontaneous Idiopathic Acroangiodermatitis of Mali in a young adult.

Pichano Khuvung¹

¹Thanjavur Medical College & Hospital, Dermatology, Venereology & Leprosy, Thanjavur, India

Introduction & Objectives:

Acroangiodermatitis otherwise called pseudokaposi's sarcoma is a long-standing condition where new blood vessels are organized from pre-existing vessels. Most cases of Acroangiodermatitis of Mali have associations like venous inadequacy or anomalies while few spontaneous cases are also reported. This benign condition presents clinically as violaceous macules, progressing into indurated plaques, nodules, or ulcers on the extensor surfaces of lower extremities. Here, we detail a case of a young adult who presented with a non-healing nodule over the left lower leg and the diagnosis was confirmed by histopathology.

Case report:

An 18-year-old male presented to our opd with complaints of recurrent episodes of chronic non-healing nodule over the left lower leg which burst to form an ulcer for 18 months and was associated with pain. On examination, a single wellcircumscribed hyperpigmented slightly tender nodules of size 3*3 cm with crusting in the centre were present over the middle 1/3 on the medial aspect of the left lower leg with hyperpigmentation & xerosis of the surrounding skin. The patient also had pitting oedema on the left lower leg and other general examination were within normal limit. No significant personal and family history. Bleeding time, clotting time, and the prothrombin time were within normal limits. Colour Doppler study of the bilateral leg was normal, ELISA for HIV non-reactive, and Mantoux test negative. Histopathology section [H & E, 10x & 40x] from the edge of the ulcer shows stratified squamous epithelium with elongated rete ridges, subepithelium shows lobular arrangement with capillary sized blood vessels surrounded by pericytes in the dermis. This patient was started on oral doxycycline & topical corticosteroids and is on regular follow-up. The patient was also advised to keep affected limb elevated whenever possible.

Conclusion:

Acroangiodermatitis is a rare entity that needs to be differentiated from aggressive malignant Kaposi sarcoma, especially in the present HIV era where morbidity and mortality are high. Therefore, non-healing ulcers irrespective of age must undergo a histopathogical examination along with imaging studies so that a precise diagnosis will be reached and treated accordingly. The approach to treating Acroangiodermatitis of Mali typically involves a comprehensive strategy aimed at managing the underlying vascular condition and easing symptoms linked to the skin.



Venous malformation presenting a diagnostic problem

Sakhri Maroua¹, Houria Sahel¹

¹Chu Maillot, Algiers, Algeria

Introduction & Objectives: Vascular malformations encompass high-flow and low-flow categories. Among these, venous malformations (VM) are the most common. We report a challenging case of VM highlighting the complexities encountered in its diagnosis.

Materials & Methods: A 2 year old male presented with a congenital swelling on the right shoulder labelled on ultrasound as a cystic hemolymphangioma, which progressively increased in size with painful inflammatory episodes. Haut du formulaireOn clinical examination, there was a subcutaneous mass, soft, depressible, non-inflammatory, painless and non-pulsatile, located on the right shoulder. Laboratory findings: D-dimer level of 5703 ng/ml (n <500). X-rays of the right shoulder showed calcifications, while ultrasound of the soft tissue suggested phleboliths. Soft tissue echo-doppler suggested a VM, possibly related to a cystic hemangioma or hemo-lymphangioma. The CT scan: a large cystic lymphangioma of the right shoulder. Soft tissue MRI: a deep intramuscular soft tissue formation in the right shoulder, suggestive of an hemangioma. A second soft-tissue Doppler ultrasound: a VM of the right shoulder, mainly involving the deltoid muscle. A review of the MRI:a deep VM of the right shoulder, mainly involving the deltoid muscle. Appropriate elastic restraint was introduced.

Results: VM are the most common congenital vascular malformations. They appear from birth and can become symptomatic during adolescence. These anomalies exhibit variability in both size and location, and may invade adjacent structures, as observed in our patient's case (intramuscular location). While generally asymptomatic, VM maybe associated with pain. Phleboliths, considered pathognomonic of VM, are sometimes palpated. It is reported in only 20% of cases .They do not regress spontaneously, and most tend to increase in size over time, particularly those on the trunk and limbs. Haut du formulaireClinical manifestations vary depending on the anatomical site; VM located in the skin and subcutaneous tissues is more likely to be noticed at birth as a bluish, palpable and compressible mass, whereas intramuscular or visceral VM may become evident later in life as a discrete mass or pain. Localized intravascular coagulopathy (LIC) may be observed, leading to the formation of phleboliths, increased fibrinolysis, and an increased risk of bleeding due to the consumption of coagulation factors. large lesions or those of substantial size and the presence of phleboliths are risk factors for LIC.From a biological point of view, the most frequent abnormality in CIL is an elevated D-dimer level. Low platelet and fibrinogen levels and the presence of soluble fibrin complexes may be observed in more severe cases. Imaging plays an important role not only in confirming the diagnosis, but also in assessing the extent of VM. Ultrasound can reveal phleboliths, which are pathognomonic for VM, but MRI remains the gold standard. Although standard X-rays can show phleboliths, their sensitivity is very low, with phleboliths present in less than 30% of cases. Compression remains the most conservative treatment of choice for VM, depending on its location.

Conclusion: In this patient, the diagnosis of VM was based essentially on a combination of clinical and biological factors, in particular the D-dimer level, as well as imaging by visualising phleboliths and ultrasound.



Treatment of PHACE syndrome with Propranolol: A case report

Sakhri Maroua¹, Houria Sahel¹

¹Chu Maillot, Dermatolgy, Dermatology, Algeria

Introduction & Objectives:

Infantile hemangioma (IH) is the most common tumor in infants. Propranolol is the first-line treatment. Segmental IH of the face is at risk of PHACE syndrome. The prescription of Propranolol in this case must be cautious, as there is a greater risk of stroke if there are associated cardiac or vascular anomalies.

Materials & Methods:

A 40-day-old female infant from a pregnancy complicated by hypothyroidism in the mother and delivered by caesarean section for foetal distress. She was admitted to hospital with IH of the S1, S2 and S4 segments of the right hemiface, which had been progressing for 5 days and was complicated by bleeding, ulceration and ocular obstruction. The ophthalmological examination was normal. Cerebral Angio-MRI revealed an orbital-facial haemangioma, a reduced-calibre permeable intracranial carotid artery and a non-expansive arahnoid cyst in the posterior cerebral fossa. The diagnosis of PHACE syndrome was made, and propranolol was started at a dose of 0.5 mg/kg/day over 6 doses, then increased in increments of 0.5 mg/kg/week with good tolerability and improvement in IH.

Results:

The acronym PHACES covers anomalies of the posterior fossa, facial hemangioma, intra- and extracranial arterial anomalies, congenital cardiac anomalies and aortic coarctation, ocular anomalies, sternal and ventral anomalies, etc. Children with a large segmental facial HI are at risk of PHACE syndrome. The results of initial screening tests should guide therapeutic management and monitoring. Initial imaging of the cervical and cerebral arteries can help classify patients in one of the following categories: low, intermediate or high risk of stroke.

Daily doses lower than the standard 3mg/kg/d or a gradual, slower increase should be considered, with closer monitoring. The total daily dose should be divided into 3 doses and may be increased as clinically indicated. Acute ischemic stroke was reported in 2 PHACE patients on Proporanolol. Both were also treated with oral corticosteroids and had severe arteriopathy. In another series of 32 PHACE with arterial anomalies, the majority of patients tolerated the treatment well.

Conclusion:

We report a new PHACE case successfully treated with Proporanolol. PHACE is not therefore an absolute contraindication to the use of beta-blockers, but the benefits and risks must be carefully weighed up.



Propranolol as a magistral preparation in the management of infantile hemangiomas

Sakhri Maroua¹, Hamchaoui Farida¹, Houria Sahel¹

¹Chu Maillot, Dermatolgy, Algiers, Algeria

Introduction & Objectives:

Infantile hemangioma (IH) is the most common tumor in infants. Propranolol is the first-line treatment. When it is not available, magistral preparations are used. The aim of this study was to assess their efficacy and safety.

Materials & Methods:

We conducted a descriptive, cross-sectional study comprising a retrospective part (December 2019-March 2023) and a prospective part (April-June 2023). Patients included were those hospitalized for the management of IH and treated exclusively with propranolol compounded in capsules.

Results:

We enrolled 32 patients (24 for the retrospective part and 8 for the prospective part). The mean age was 7.21 months (range: 2 to 20 months). The 0-5 months age group was the most affected (47% of cases). The sex ratio was 0.14. Among the cases , 56.3% were mixed IH, 28.1% tuberous and 15.6% subcutaneous IH. 81.3% were localized IH, 1 segmental and 2 multifocal. 56.2% were located on the head, 21.8% on the lower limb, 18.7% on the trunk, 15.6% on the genitals and 9.4% on the upper limb. therapeutic indication was established for functional impairment in 47% of cases, a complication in 47% of cases (ulceration in 37.5% of cases), and aesthetic concerns in 2 cases. 03 patients were lost to follow-up before treatment was initiation , and 04 before the introduction of the 3 mg/kg/d dose. The mean duration of treatment was 4.2 months, with an average duration of 6 days for the 1 mg/kg/d dose, 17 days for the 2 mg/kg/d dose and 3.2 months for the 3 mg/kg/d dose, with a maximum duration of 07 days, 6 months and 9 months for these doses respectively. Progression was favorable in 87.5% of patients. One adverse event, bronchial obstruction, occurred one day after treatment, leading to its discontinuation. No relapses were recorded.

Conclusion:

We report on the efficacy and tolerability of propranolol in magistral preparation for the treatment of IH. It serves as a viable and cost-effective alternative, especially in countries where appropriate formulations may not always be available.



Sturge-Weber syndrome: two cases and review of the literature

Sakhri Maroua¹, Houria Sahel¹

¹Chu Maillot, Dermatolgy, Algiers, Algeria

Introduction & Objectives:

Sturge-Weber syndrome (SWS) is a neurocutaneous syndrome defined by the presence of leptomeningeal capillary or capillary-venous malformations. Glaucoma is present in half of patients. Epilepsy, encephalopathy and hemiplegia are also frequently associated with SWS. We report two cases of SWS.

Materials & Methods:

Case 1: A 5-year-old female child was was hospitalized for SWS. At birth ,an erythematous spot on the right hemiface was noted. Echocardiography, ophthalmological examination and brain CT scan were normal.Local propranolol with 3 sessions of laser therapy led to discrete improvement. At the age of 5, she presented a convulsive seizure, asthenia and tingling sensations in both feet. Clinical examination revealed a flat angioma in the V1 territory of the right hemiface and the medio-occipital region. . Cardiac ultrasound showed no anomalies. Brain- MRI was consistent with Sturge-Weber encephalo-trigeminal angiomatosis, revealing cortical calcification with temporo-occipital right pial angioma. The EEG showed an asymmetric, poorly organized pattern consistent with right cerebral damage and a right occipital epileptogenic focus compatible with partial epilepsy. Monitoring in neurology, dermatology and ophthalmology was recommended.

Case 2: At the age of one week, a male child presented with a flat angioma of the left cheek which increased in size to encompass the entire left hemiface. At the age of 12, macrocheilitis of the lower lip was noted. During a clinical examination at the age of 14, a flat angioma of the V2 and V3 territories of the left hemiface was found, slightly warm on palpation, non-pulsatile and painless. Aditionally, another flat angioma of 03 cm long axis of the V3 territory of the right hemiface was found. Cerebral MRI revealed extensive supratentorial demyelinating lesions, maxillary sinusitis and anterior sphenoid sinusitis. The patient was referred to maxillofacial surgery for treatment of his macrocheilitis with neurological, dermatological and ophthalmological monitoring.

Results:

SWS is a sporadic, congenital neurocutaneous syndrome involving the skin, brain and eyes. It is caused by a somatic mutation in the GNAQ gene located on chromosome 9q21, affecting neural crest cells.Bilateral planar angiomas or those extending from the forehead to the cheek have a higher risk of SWS, with the forehead remains the best predictor of this risk. However, SWS can also occur in the absence of associated facial vascular anomalies. The identification of a facial plane angioma at risk, particularly those involving the forehead, should therefore prompt an ophthalmological examination to look for congenital glaucoma. Early diagnosis of SWS is recommended, including screening of asymptomatic patients with "high-risk" facial hair malformation by brain MRI, in order to reduce neurological morbidity, enabling the early introduction of laser treatments, primarily using pulsed dye laser (PDL) as the first-line treatment which can improve treatment results. The factors that have been shown to have prognostic value are the extent of brain damage and the early onset of severe seizures that are difficult to control in the first few months of life (before the age of 9 months).

Conclusion: This case highlights the importance of a multidisciplinary approach to the management of SWS and the factors that need to be taken into account, particularly the psychosocial impact of the disease and the family's financial situation.



Treatment of infantile hemangiomas complicated with ulceration using telemedicine methods and topical betablockers under the condition of limited access to medical care in terms of war, pandemics, or natural disasters

Olga Bogomolets^{*1}, Anna Bogomolets-Lanina², Pavelko Mariia²

¹Ukrainian Military Medical Academy, Dermatology, Kyiv, Ukraine, ²Dr. Bogomolets' Institute of Dermatology and Cosmetology, Kyiv, Ukraine

Introduction & Objectives:

In regions affected by war, pandemics, or natural disasters, access to medical care is often severely restricted, leading to challenges in managing infantile hemangiomas (IH), particularly when complicated by ulceration. This study aimed to evaluate the efficacy and safety of telemedicine methods combined with topical beta-blockers in treating ulcerated IH under conditions of limited medical access. We assessed treatment outcomes, complications, and parental experiences to provide insights for optimizing care delivery in resource-constrained settings.

Materials & Methods:

A retrospective analysis was conducted on 497 patients with IH treated between 2022 and 2023. Data on patient demographics, treatment modalities, and outcomes were collected. Telemedical consultations were initiated for patients unable to access in-person care due to conflict-related or logistical barriers. In the case of telemedical or online treatment, without the possibility of a medical examination of children to exclude contraindications for the systemic usage of beta-blockers - only local beta-blockers were used in the form of sterile timolol maleate gel 0.25%, 0.5%, 1%, which was applied to the surface of the hemangioma depending on its location and the weight of the child - every 3–6 hours in the form lubrication or the form of lubrication under a gel bandage.

Results:

Of the 497 patients, 356 (72%) received telemedical or online treatment due to restricted access to medical facilities. Ulceration was diagnosed in 37 cases (7.4%), with a notable increase from 2.9% in 2022 to 9.2% in 2023. Telemedical interventions led to complete healing of ulcers in all cases without reported side effects. Additionally, 70% of ulcerated IH cases achieved complete resorption following beta-blocker treatment. The duration of ulcer healing ranged from 1 to 7 months, with regression of IH observed within 4–12 months. In 7 (19%) cases, the treatment with the pulse dye laser was used 3–6 treatments with a 4-6 week interval. The patient's parents noted the impossibility of starting timely treatment for hemangioma due to confusion - 31 (84%), lack of access to medical facilities 24 (64%), and lack of information about the possibility of online consultation and treatment 34 (94%).

Conclusion:

In conflict zones or emergencies, the incidence of ulcerated IH increases due to delayed treatment initiation. Parental confusion, lack of access to medical facilities, and limited awareness of telemedical options contribute to treatment delays. Telemedical approaches, combined with topical beta-blockers, offer an effective and safe alternative for managing ulcerated IH in resource-constrained settings. Utilizing sterile gel bandages for wound care empowers parents to participate in treatment, facilitating child care. This study underscores the importance of telemedicine in expanding access to care and optimizing outcomes for vulnerable populations amidst crises.



Livedoid Vasculopathy Undiagnosed for 8 Years in a 40-year-old Man

Liliya Tivcheva*¹, Zdravka Demerdzhieva¹

¹Acibadem City Clinic Tokuda Hospital, Sofia, Bulgaria

Introduction & Objectives:

Materials & Methods:

Results:

A 40-year-old male Caucasian patient presented with an 8-year history of undiagnosed recurrent painful and pruritic "wounds" on the lower legs. Over the years the patient had been treated with various topical medications (not specified) without significant improvement. He reported that his condition had worsened in the past two months with the appearance of swelling, redness and blisters with serous liquid secretion. He consulted a dermatologist who applied two injections of depo corticosteroid i.m. with some improvement in the dermatologic status.

The clinical examination revealed numerous livid polycyclic cicatrices on the lower legs bilaterally, in places with porcelain white atrophy and excoriations. In the right perimalleolar region a single sharply-demarcated ulcerative lesion was observed.

The patient was admitted to the Dermatology ward for diagnostic procedures and further treatment. A punch biopsy was performed to establish a precise diagnosis. The histologic result corresponded to a segmental hyalinizing vasculitis (a.k.a. livedoid vasculopathy) - thickened and hyalinized vessel walls in the whole dermis, some of them to the point of obliteration, as well as scarce perivascular lymphocytic infiltrate. An ANA screening was conducted which was negative, but the anti-beta-2-glycoprotein antibodies and anti-cardiolipin antibodies were positive. Therapy was started with pentoxyfilline 400mg/d. i.v., dipyridamole 25mg tabl. 3x2t./d.; clobetasol propionate 0.05% cr. 2t.d. loc.; oxytetracycline hydrochloride/hydrocortisone spray 2t.d. loc.; wet dressings with KMnO4 2t.d. loc. There was a significant improvement in the patient's condition during his hospital stay, and he was prescribed long-term therapy with regular check-ups with a dermatologist.

Livedoid vasculopathy is a rare disease that is more prevalent among women (3:1 ratio) and is usually characterized by a long delay between presentation of symptoms and diagnosis. The disorder can have chronic complications if not treated timely, such as scarring, hyperpigmentation, and mononeuropathy multiplex, among others. This is a rare case of a 40-year-old male with livedoid vasculopathy, who had gone undiagnosed for 8 years before finally receiving a histopathological and clinical diagnosis.

Conclusion:



Pseudo-Kaposi verrucous: A aase report of chronic venous insufficiency mimicking Kaposi's sarcoma

Mohamed Bennanii¹, Malek Ben Slimane¹, Dorra Mdhaffar¹, Faten Rabhi¹, Kahena Jaber¹, Mohamed Abderraouf Dhaoui¹

¹The Military Hospital of Tunis, dermatology, Tunis, Tunisia

Pseudo-Kaposi verrucous: A Case Report of Chronic Venous Insufficiency Mimicking Kaposi's Sarcoma

Bennani Mohamed , Ben Slimane Malek, Dorra mdhaffar, Rabhi Faten, Jaber Kahena, Mohamed Raouf Dhaoui

The Military Hospital of Tunis, dermatology, Tunis, Tunisia

Introduction & Objectives:

Pseudo-Kaposi sarcoma (PKS) is a rare vascular phenomenon associated with congenital vascular malformations or acquired venous insufficiency. It is characterised by purpuric macules, papules or plaques on the dorsum of the feet and the malleolus. Here we report a case of atypical PKS.

Materials & Methods:

A case report of a 72-year-old man presented with hyperkeratotic verrucous plaques on the lateral aspect of both feet and the plantar aspect of the hallux. He also had varicose veins. Biopsy revealed features consistent with pseudo-Kaposi verrucous. histologically we had The epidermis is acanthotic, papillomatous, spongy and hyperorthokeratotic. Keratinocytes do not show cytonuclear atypia. Absence of koilocyte-like cells. The underlying dermis is replaced by a polymorphic inflammatory infiltrate of moderate density with exocytosis in the epidermis. Skin biopsy showing pseudo epitheliomatous hyperplasia of the epidermis in favor of pseudo kaposi verrucous. Further evaluation with Doppler ultrasound confirmed chronic venous insufficiency. The patient was treated with keratolytic ointment and support stockings with symptomatic improvement.

Results:

Acquired PKS, also known as Mali type or vascular dermatitis of the extremities, results from chronic venous insufficiency. Lesions typically affect the distal part of the lower leg and foot, concentrating around the dorsal and back of the toes, especially the 1st and 2nd toes. They appear as reddish-purple patches, papules, or plaques that usually develop slowly. Rarely it can become verrucous, as seen in our patient. The differential diagnosis includes warts, verrucous lichen planus, Kaposi's sarcoma, and squamous cell carcinoma. However, careful histological examination revealed features consistent with PKV, highlighting the importance of accurate diagnosis by biopsy in distinguishing benign conditions from malignancy. The association of PKV with chronic venous insufficiency (CVI) is noteworthy. The presence of varicose veins in our patient prompted further investigation with Doppler ultrasound, confirming the diagnosis of CVI. This finding suggests a possible pathophysiological link between PKV and CVI, emphasizing the importance of considering underlying vascular etiologies in patients presenting with PKV-like lesions, particularly in the lower extremities. The management of PKV involves addressing both the dermatological manifestations and the underlying vascular pathology. Keratolytic agents treat hyperkeratosis and reduce the thickness of verrucous plaques, improving symptoms and cosmetic appearance. Additionally, venotonics and compression stockings manage CVI, aiming to reduce venous hypertension and prevent disease progression. This combined approach was effective in our patient, resulting in symptomatic improvement and resolution of hyperkeratotic lesions.

Conclusion:

The verrucous type of PKS is uncommon and timely diagnosis and appropriate management, including both

dermatological and vascular interventions, are essential for optimal patient outcomes.



Leriche syndrome revealed by unilateral cutaneous necrotic ulcerations

Meryem Khallouki¹, Bendaoud Layla¹, Sarah Zemrani¹, Maryem Aboudourib¹, Ouafa Hocar¹, Said Amal¹

¹Mohammed VI University Hospital of Marrakech, Department of Dermatology and Venereology, Marrakech, Morocco

Introduction & Objectives:

Leriche syndrome (LS), also commonly known as aortoiliac occlusive disease (AIOD), is a product of atherosclerosis affecting the distal abdominal aorta, iliac arteries, and femoropopliteal vessels. LS, first described by Leriche and Morel in 1924, presents itself more frequently as chronic claudication, erectile dysfunction, and absent femoral pulses. We present a patient with an atypical presentation of painful unilateral necrotic ulcerations of the lower limb, revealing LS and ischemic cardiopathy.

Materials & Methods: case report

Results:

A 70-year-old male patient presented with a 2-month history of deep, painful, non-healing ulcerations on the right lower limb. The patient had a history of smoking for the past 30 years, non-treated hypertension, unrecognized chest pain, intermittent claudication, and paresthesia of the lower limbs. Physical examination revealed multiple stepped necrotic ulcerations of the right lower limb, livedo reticularis, and local purpuric lesions. The arterial pulsations of the right tibial arteries were reduced; no pulsations were found over the right femoral artery. Arterial echo-Doppler of the lower limbs showed diffuse obliterative arteriopathy with multiple calcified atheromatous plaques of the aorto-iliac axis and the arteries of both lower limbs. The CT angiography showed Leriche syndrome with total occlusion of the abdominal aorta associated with bilateral occlusion of the iliac arteries. Coronary angiography showed tri-truncular lesions with stenosis of the middle anterior interventricular artery, middle right coronary artery, and circumflex artery. Revascularization was indicated, but the patient died of cardiogenic shock.

Discussion:

LS is a rare and critical complication of peripheral arterial disease (PAD), caused by atherosclerosis. The prevalence of LS is unknown given that many cases are asymptomatic, but PAD, the etiological agent of Leriche syndrome, has a prevalence of approximately 115 million worldwide, with 70,000 deaths in 2019. Dyslipidemia, male gender, smoking, diabetes mellitus, and hypertension are the main risk factors. Cutaneous manifestations varied from nonspecific inflammatory lesions through ulcers to gangrene of low extremities, and the diagnosis might be difficult. Patients report severe intermittent claudication, ischemic rest pain, or burning and pain in the cutaneous lesions; they may initially have no symptoms. In some cases, non-specific symptoms, such as stiffness, paresthesia, reduced sensitivity or pain, and claudication, appear. A case with similar clinical features to our patient reported multiple deep and painful ulcers with necrotic borders along the lower extremities, revealing Leriche syndrome. Another case involving a woman with perineal ulcers has recently been reported in association with AIOD. Patients with critical limb ischemia had a significantly higher relative risk for myocardial infarction, major amputation, cardiovascular mortality, major adverse cardiac events, and allcause mortality. Unfortunately, our patient died from a myocardial infarction.

Conclusion:

This case illustrates the importance of proper history-taking and physical examination with careful examination of peripheral pulses of both lower limbs in patients presenting with multiple non healing cutaneous ulcerations in order to avoid misdiagnosis of AIOD.



Leukocytoclastic vasculitis affecting the gallbladder and skin as a paraneoplastic syndrome in pancreatic carcinoma

Suzana Nikolovska¹, Maja Dimova¹, Ivana Dohcheva Karajovanov¹, Hristina Breshkovska¹, Silvija Duma¹, Anita Najdova¹

¹University Clinic of Dermatology, Skopje, North Macedonia

Introduction & Objectives:

Although rare, leukocytoclastic vasculitis (LCV) secondary to solid organ malignancy has been documented. We herein describe a case of LCV of gallbladder and skin presenting as a paraneoplastic syndrome associated with pancreatic carcinoma which remarkably improved in parallel to the clinical course of the malignant tumor.

Case report:

55-years-old female patient with the most prominent manifestation of upper abdominal pain in the setting of normal liver tests was presented to University Clinic of Abdominal Surgery. An abdominal ultrasound demonstrated a thickened gallbladder wall, without gallstones. Diagnose of acute acalculous cholecystitis was made and laparoscopic cholecystectomy was performed. The patient was referred to the angiology outpatient clinic of the University Clinic of Dermatology one week after cholecystectomy with palpable purpura, hemorrhagic bullae and necrosis on the lower extremities. Both biopsies, from gallbladder and skin were conclusive with diagnosis of LV. Interestingly, the deeper dermal structure were involved with the presence of histiocytes and plasma cells in the infiltrate. This finding raises the suspicion that it is a case of paraneoplastic LV. The patient was treated with oral corticosteroids (1mg/kg). Further examinations revealed diagnosis of pancreatic carcinoma. After surgical removal of the tumor, regression of the skin lesions occurred.

Duscussion:

Acute acalculous cholecystitis presents 2-15% of all cases of acute cholecystitis. LCV of the digestive tract is rare, especially isolated LV of the gallbladder. It is recommended that all patients with signs of acalculous vasculitis without peritonism should be given corticosteroids and not immediately treated surgically.

Suspicion that LCV is a paraneoplastic phenomenon should be raised in case: (1) LCV is associated with symptomatology from other systems and/or organs; (2) Laboratory studies indicate a lesion of another organ; (3) In the histopathological image, involvement of the deep dermis is registered and there is a different cellular "milieu" (plasma cells and histiocytes).

Conclusion:

LCV as a paraneoplastic phenomenon is more often associated with myeloproliferative diseases, and less often with cancers of solid organs. This is the first described case of LCV of gallbladder and skin associated with pancreatic adenocarcinoma.



Angiolymphoid Hyperplasia with Eosinophilia: A Rare Entity and a Therapeutic Challenge

Maria-Alexandra Visan^{*1, 2}, Ana Maria Rosca¹, Diana Savu¹, Ana-Liliana Butnarus¹, Monica Darmanescu¹, Viorel Trifu¹

¹Dr. Carol Davila Central Military Emergency University Hospital,²Carol Davila University of Medicine and Pharmacy, Physiology Department, Bucharest, Romania

Introduction & Objectives:

Angiolymphoid hyperplasia with eosinophilia (ALHE) is considered to be a rare benign disorder of a yet unknown etiopathogenesis, consisting of solitary or multiple erythematous-violaceous papules or nodules most frequently localised in the head and neck region, particularly in periauricular sites. As the name suggests it, ALHE has a vasoproliferative nature, with lymphocytic and eosinophilic infiltration, however current literature challenges its significance on whether it represents a vascular neoplasm, a reactive process to different stimuli or even a T-cell lymphoproliferative condition of a benign or low-risk malignant type. Treatment proves difficult, with no standardised regime, as well as unsatisfactory results and high recurrence rates.

Materials & Methods:

We herein introduce the case of a 45-year-old Caucasian male patient, with no relevant comorbidities. He was admitted for the occurrence of asymptomatic red-to-brown papules of a 0.5-2 cm diameter, irregularly delineated and localised on the forehead, nose, as well as bilateral temporal and auricular regions. Topical corticosteroids, imiquimod, and oral propranolol had been previously attempted as therapeutic strategies, with little to no improvement. Laboratory analysis revealed leukocytosis with neutrophilia and lymphocytosis, slightly elevated erythrocyte sedimentation rate and gammaglutamyl transpeptidase, however no peripheral eosinophilia or elevated immunoglobulin E (IgE) levels were depicted. Histological examination highlighted dermal expansion through multiple lymphangiectasia and a polymorphic inflammatory infiltrate with a dense perivascular disposition, occasional leukocytoclasia and follicular plugging. Periodic acid-Schiff (PAS) staining revealed proliferating blood vessels lined by large turgescent endothelial cells protruding to the lumen, accompanied by a focally aggregated inflammatory infiltrate consisting of lymphocytes and polymorphonuclear leukocytes, including frequent eosinophils. Surprisingly, ultrasound examination identified cervical and axillary lymphadenopathies, which may be more frequently linked to Kimura's disease, the primordial differential diagnosis of AHLE. However, it was excluded based on epidemiologic profile, biopsy results and IgE levels, along with other diagnoses like Kaposi sarcomas, granuloma faciale, sarcoidosis or angiosarcoma lesions.

Results:

Although surgical excision has reported the lowest recurrence levels across literature, the multiplicity of lesions led to a first therapeutic attempt of intralesional corticosteroids (six sessions at three-week intervals), with almost complete remission, taking into consideration future targeting of remaining lesions through vascular laser treatment.

Conclusion:

Although benign, AHLE remains an enigma, due to uncertainties in its etiopathogenesis, poor treatment response and frequent recurrence, along with a low prevalence which renders its diagnostic and treatment personalisation challenging.