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**Abstract N°: 333**

# Successful treatment of multiple glomovenous malformations / glomangiomatosis with combined 585nm Pulsed Dye Laser (PDL) and 1064nm Long-Pulsed Neodymium Yttrium Aluminium Garnet (Nd:YAG) Laser.

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**Introduction & Objectives:**

Glomuvenous malformations [GVMs] are rare benign vascular neoplasms that may present at birth or appear later in life. Typically due to an abnormal growth of blood vessels with accumulation of glomus cells. Although GVMs may be sporadic, they have been linked to a glomulin gene defect and can be present at birth or appear later in life. They present as pink, red or blue localised or segmental nodules or coalescing plaques that may be associated with pain. Patients usually receive treatment for esthetic reasons or to relieve the pain and improve functionality. Current treatment modalities depend on location and size of GVMs. They include surgery, scleropathy or laser. In the case of multiple or large GVMs, treatment options are challenging, as surgery may be impractical and leading to scarring. Other treatment options have side effects such as skin ulceration and hyperpigmentation.

**Materials & Methods:**

We present a case of glomangiomatosis treated with combined 585-nm PDL and 1064-nm Nd:YAG laser achieving an excellent clinical outcome with resolution of the pain associated with the lesions

**Results:**

A 29 year old woman presented to dermatology outpatients with a more than 10 year history of multiple blueish/purple lesions on her arms, legs and trunk. She described no family history. The Lesions were very tender and interfered with her work as a primary school teacher as they were causing pain when cuddling or picking up young children. Multiple glomuvenous malformations were suspected and a punch biopsy was taken for confirmation. Histology revealed dilated vessels and a prominent population of glomus cells consistent with a glomangioma.

We treated her with 1 session of long-pulsed Nd:YAG laser alone (1064nm, 10ms pulse width, 60-100J/cm^2 fluence, 7mmspot) and 5 sessions of combined Pulsed Dye Laser (585nm, 0.5-10ms pulse width, 7-8J/cm^2 fluence) and Long-pulsed Nd:YAG laser (1064nm, 15-20ms pulse width, 30-60J/cm^2 fluence) using Multiplex settings and a 10mm spot. She achieved complete or partial remission of all lesions and resolution of her painful symptoms.

**Conclusion:**

Treatment of GVMs, especially when multiple or extensive, is challenging with a high recurrence rate and is associated with several side effects. With surgery being impractical in the multiple variant, laser as an approach can be a promising treatment option. The dual administration of PDL and Nd:YAG laser can target different depths improving the likelihood of a better outcome while reducing the possible side effects of Nd:YAG such as ulceration and scarring. Although PDL is superficial and can only help flatten superficial lesions and relieve pain, PDL increases the absorption of Nd:YAG allowing the use of lower fluencies thus reducing the possible side effect and allow a better outcome with thicker lesions which may be more challenging to treat. We present a case study

of safe, effective and well tolerated combined PDL/Nd:YAG laser treatment for multiple GVM.

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**Abstract N°: 775**

# Tufted angioma treated with low dose aspirin in a 1-year-old Filipino boy: a case report

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**Introduction:**

Tufted angioma is a rare, benign, vascular neoplasm of the skin. Diagnosis of this condition is infrequent due to its rare occurrence. Only 158 cases have been described as of 2015. Treatment reported in literature is very limited with no clear guidelines on its management. Currently, there are no reported cases in the Philippines of tufted angioma treated with aspirin.

**Case Report:**

This is a case of a one-year-old Filipino boy presenting with multiple dusky red papules and plaques on the left side of the cheek, pre-and post-auricular areas, parieto-occipital areas, chest, and upper back. His lesions started at 2 months of age, noted to increase in size, number, and thickness over time. Dermoscopy revealed homogenous erythematous background with perifollicular lacunae separated by thin septa. Histopathology revealed dilated vessels in the papillary dermis with proliferation of endothelial cells in lobules, surrounded by dilated crescent shaped vascular channels in the mid to deep dermis, consistent with tufted angioma. Patient was treated with low dose aspirin (5 mg/kg/day) once a day for 1 month and was noted to have decrease in size and thickness with more hyperpigmented patches and plaques on the affected areas. After 4 months, no new lesions, no increase in size, nor symptoms were noted.

**Conclusion:**

Low dose aspirin is an effective and safe option for monotherapy of tufted angioma in pediatric patients.

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**Abstract N°: 1255**

# Purpuric Lesions: Practical Clinical Algorithm Based on Physiopathological Mechanisms for the Initial Approach

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## Introduction & Objectives:

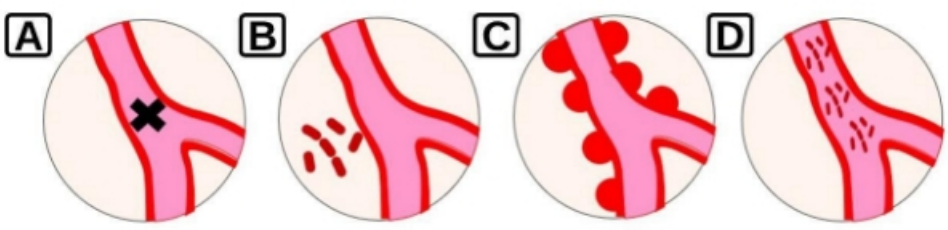
Purpuric lesions are caused by the extravasation of red blood cells from the cutaneous blood vessels. They pose a diagnostic challenge for physicians due to their association with various medical conditions. Some purpuric lesions can be potentially fatal because of their relationship with systemic disorders. The clinical characteristics of purpuric skin lesions (size, color, and distribution) can point to the underlying specific pathophysiological mechanism. There are multiple pathophysiological underlying mechanisms associated with purpuric lesions. Coupled with the patient interview, exploration, and laboratory tests, it is possible to determine the diagnosis and provide the patient the proper medical treatment.

## Materials & Methods:

Design:** Review of the physiopathological mechanisms of purpuric lesions. Data extraction included review articles from various medical conditions associated with purpuric lesions. Creation of an algorithm based on the pathophysiological mechanisms of purpuric lesions.

## Results:

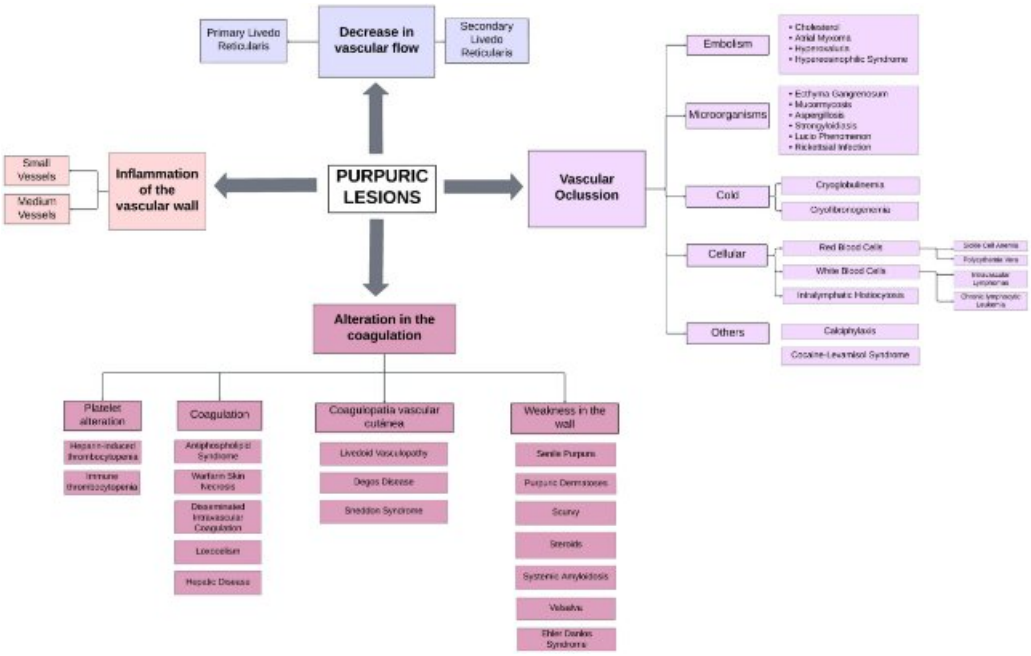
There are different mechanisms by which erythrocytes extravasate and cause purpuric lesions. The first mechanism occurs due to vascular occlusion that leads not only to ischemia of the adjacent tissue but also of the vascular wall that culminates in the rupture of the vessel and extravasation of red blood cells (Figure 1A). The second mechanism occurs due to the leakage of erythrocytes due to an alteration in the coagulation cascade or due to weakness of the vascular wall (Figure 1B). The third mechanism occurs due to inflammation of the vascular wall (vasculitis) secondary to a reactive inflammatory process (Figure 1C). Finally, the fourth mechanism is explained by a decrease in vascular flow that can be caused by hyperviscosity or vasoconstriction (Figure 1D).




Each mechanism of vascular damage manifests itself in a different way, and, although clinical lesions can overlap, they guide the clinician regarding their origin. Vascular occlusion can manifest with branching violaceous lesions that reflect the architecture of the vascular network of the skin. When the obstruction of blood vessels is partial, it is called livedo racemose and when complete it is known as retiform purpura. The leakage of erythrocytes due to

alterations in coagulation or weakness of the vascular wall manifests itself with erythematous-violaceous macular lesions called petechiae when they measure less than four millimeters, macular purpura when they measure between four and nine millimeters, and ecchymosis when they measure more than one centimeter. Inflammation of the wall, called vasculitis, manifests itself in many ways but palpable purpura is the most common. The decrease in blood flow can occur due to vasoconstriction or hyperviscosity due to an increase in components in the blood, this manifests as livedo reticularis.

Figure 2 schematizes the possible diagnoses that manifest with purpuric lesions according to their pathophysiological mechanism.




**Conclusion:**

In conclusion, this article seeks to provide an approach to purpuric cutaneous lesions, analyzing them through their pathophysiological mechanism, with the aim of offering a comprehensive framework for diagnosis. Taking a complete history, along with laboratory tests and especially skin biopsy, are essential tools in the diagnostic process, to ensure the most appropriate treatment for the patient and identify causes that threaten the patient's life.

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**Abstract N°: 1425**

**Burn and infantile haemangiomas, clinical and histopathological similarities**

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**Laser assissted transepidermal drug delivery in Dermatology**

**Introduction & Objectives:** Absorption of topical products through the epidermis is limited by the skin's barrier function. Laser-assisted drug delivery enhances the penetration of topically applied treatments, leading to greater delivery and bioavailability. It has been used to imrove the effect of many drugs used to treat various skin disorders.

**Materials & Methods:** We discuss the therapeutic application of laser-assisted drug delivery in clinical practice in, vitiligo, melasma, and alopecia areata. The studies including 34 patients with stable vitilgo treated with Tacrolimus ointment and ER:YAG, 25 patients with melasma treated with Kojic acid and Er:glass laser and 35 patients with Alopecia Areata traeated with Minoxidil and Er:glass laser.

**Results:** showed that the application of laser-assisted drug delivery enhances topical agent efficacy, potentially reducing the agent concentration and duration of topical treatment required in all three diseases.

**Conclusion:** Laser-assisted delivery of various drugs used, is a safe and effective method for the treatment of the enrolled patients with vitiligo, melasma and Alopecia areata.

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**Abstract N°: 1578**

# Therapeutic management of infantile hemangiomas with beta-blockers: experience of the department of dermatology, CHU Tlemcen

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**Introduction & Objectives:** Infantile hemangioma (IH) is the most common tumor in children. Because of their natural involution in a few years without major sequelae, in the majority of cases, therapeutic abstention is the rule. Only complicated forms are treated. The discovery of the spectacular action of β-blockers on infantile hemangiomas has transformed the prognosis of hemangiomas at risk.

The aim of our study is to investigate the response and tolerance of different beta-blockers and the different factors influencing the therapeutic response

**Materials & Methods:** Prospective study from January 01, 2015 to June 30, 2018 at the level of the Service of Dermatology CHU Tlemcen, involving 166 children with infantile hemangioma having an indication for treatment with Betablocker

**Results:** : Over a period of 42 months, we collected 166 patients. The mean age was 3.81±1.33 months, 78% of the cases were female. In our sample the notion of a free interval was almost constant. The mean time to change the premonitory lesion was 27.98 days, the tuberous form was found in 47.76% of cases, 4.97% had a subcutaneous form. The mixed form involved 81 infants (40.29%). The lesions were multiple in 15.7% of the patients.the therapeutic indications were ulceration in 37.34% of the cases, the presence of a functional risk in 50% of the cases, the presence of an aesthetic risk in 33.13% of the cases and a vital risk in 4.21% of the cases. We found a rate of 28.6% of side effects which were mostly minor. After six months of treatment, the response was favorable in 145 infants, i.e. 87.34% of the study population. The therapeutic response was complete in 17.5% and almost complete in 69.9% of cases. An analyticunivariate study showed that the factors of good therapeutic response were cervico-thoracic location (p = 0.001), segmental distribution (p = 0.001), peri-orificial location; hemangiomas of size ˃ 3cm(p = 0.001).

**Conclusion:** Our study corroborates previous publications (regression rate between 50 and 96%) and shows a significant correlation of treatment response with large hemangiomas, with cervicocephalic localization compared to other localizations and with segmental hemangiomas compared to localized configuration and per-orificial localization at cervicocephalic level. Beta-blokers allow a rapid decrease in the size of hemangiomas with few effects .a duration of more than 6 months are necessary

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**Abstract N°: 1579**

# A case of hyperkalaemia secondary to the application of topical timolol:A paediatric case report

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**Introduction & Objectives:**

Infantile haemangioma is the most common benign tumour in children. At present, propranolol is the treatment of choice. Its use by the systemic route could expose the patient to hyperkalaemia.

We report an observationof hyperkalaemia in an infant treated with timolol (Geltim 1mg/gr) for an ulcerated infantile haemangioma.

**Materials & Methods:**

Our observation concerns a female infant aged 6 weeks who presented with an infantile hemangioma ulcerated since birth. Clinical examination revealed a tuberous hemangioma of sacral location extending to the intergluteal fold and respecting the anal margin.Treatment with timolol (Geltim 1mg/gr) at a dose of 0.1 mg/kg was started, along with hydrocolloid dressings. On the fourth day of treatment, the girl developed a disturbed ionogram: Na+135mEq/L Kà 7.5 mEq /l confirmed on two blood samples. After elimination of other differential diagnoses responsible for the hyperkalaemia and urgent correction of the ionogram, the diagnosis of iatrogenic hyperkalaemia induced by timolol (Geltim1mg/gr) was made.

**Results:** Timolol ophthalmic preparations are widely used in the elderly, and in the absence of experimental data in children, the gel is not recommended for these patients. However, several studies have shown the absence of systemic effects of transcutaneous timolol due to the very small dose received (max: 0.1mg/kg).

Topical timolol appeared to be a promising treatment for superficial IH, particularly for small IH and those with a contraindication to the oral route.a few observations of hyperkalaemia have been reported with atenolol and Avlocardyl by the general route in other indications.to our knowledge, no case of hyperkalaemia has been reported with the oral form, to date.

**Conclusion:** Local-acting BB blockers may be associated with the same systemic effects as systemic BBs

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**Abstract N°: 1650**

**PHACES syndrome: many faces of PHACE, Four cases report**

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[1]Algeria, algiers, algiers

**Introduction :**

PHACE syndrome (**p**osterior fossa anomalies, **h**eman- gioma, **a**rterial lesions, **c**ardiac abnormalities/coarctation of the aorta, **e**ye anomalies and **s**ternal clefting or **s**upraumbilical raphe) is characterized by large infantile hemangiomas (IH) of the face, neck, and/or scalp that are associated with developmental defects. We report two cases of PHACES syndrome.

**Case report :**

**Case 1**: A 4 and a half month old infant presented with a mixed infantile hemangioma of the left hemiface (S2, S3) extending to the lips and left ear. (Fig.1). Angio-MRI of the brain showed a vascular anomaly of the arteries in the cerebral region with an anomaly of the posterior cerebral fossa such as partial lower vermian hypoplasia. The echocardiogram was without abnormalities.

**Case 2**: A 4-month-old infant presented with a segmental hemangioma of the face, occupying the S2, S3 and S4 segments with extension to the nasal and oral mucosa. Clinical examination also revealed a hernia of the supraumbilical linea alba. (Fig.2). Head MRI revealed an associated parotid hemangioma, abdominal ultrasound showed a hernia of the supraumbilical white line with uncomplicated digestive contents, nasofibroscopy, echocardiography and supra-aortic trunks were without abnormalities. Given the association of segmental facial hemangioma of the face and white line abnormality, the diagnosis of PHACE syndrome was retained.

**Case 3**: A 1-month-old infant with a history of maternal-foetal infection presented with an infantile hemangioma of the left side of the face (S1, S2 and S3) (Fig .3). MRI of the brain showed hypoplasia of the left vermian and cerebellum and a dysplastic appearance of the left internal carotid artery. Ophthalmological examination revealed ptosis and strabismus of the left eye. The echocardiogram was without abnormalities.

**Case 4**: A 4-month-old infant presented with an infantile hemangioma of the left hemiface (S2, S3, S4). (Fig.4). Cervico-cerebral angio-MRI showed a completely aberrant course of the internal carotid artery, a slender appearance of the left external carotid artery and an anomaly of the aortic arch. The echocardiogram was without abnormalities.

The 4 patients were treated by Propranolol at a dose of 0.5 mg/kg, gradually increased to 3 mg/kg/d with good progress.

**Discussion:**

Extensive facial or cervical hemangiomas may be associated with one or more systemic malformations grouped under the term PHACES syndrome. Its prevalence is unknown, but it has a clear female predominance. Neurological and vascular malformations, sometimes with serious functional consequences, are the most common. This is why it is important to know how to recognize PHACES syndrome.

**Conclusion:**

The interest in presenting these 4 clinical cases is to raise awareness among dermatologists so that they can quickly recognize newborns with a segmental hemangioma, and requiring an assessment for PHACE syndrome and to quickly initiate treatment by Propranolol as well as close monitoring.

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**Abstract N°: 1726**

**A Rare Case of Vascular Eccrine Spiradenoma in the forearm.**

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**Introduction & Objectives:**

Cutaneous adnexal tumors are a group of skin tumors that are differentiated towards one of the four primary adnexal structures, which are hair follicles, sebaceous glands, apocrine glands and eccrine glands that can be found on our normal skin. Spiradenoma is a rare, benign, slow-growing skin tumor that is thought to arise from eccrine sweat glands first discovered back in 1956 by Kersting and Helwig. It normally occurs between the ages of 15 to 35 years, it is most seen on the head, neck and trunk. It presents as a single, grey-to-pink nodule that develop on the skin. Spiradenoma when it is richly vascular called Vascular Eccrine Spiradenoma (VES). Excisional biopsy is required for definite diagnosis. Given its non-specific presentations, it is important to rule out other clinically worrying dermatological conditions that could be misdiagnosed.

**Materials & Methods:**

This is a 64-year-old female who presented to her GP due to recent changes of her left forearm lesion. Patient has the nodule since she was a teenager. It remains painless and does not bleed. The nodule turned bigger and darker recently which prompt the GP consultation. No history of skin malignancy. She is not on any regular medications.

On gross skin examination, a 2cm hyperpigmented (black/dark blue) nodule on her left forearm was noted. No palpable cervical and axillary lymph nodes. No associated signs of bruising, telangiectasia or lymphedema.

Given that it is a hyperpigmented skin lesion, we wanted to exclude malignant melanoma however it only scored 2 points for change in size out of the weighted 7-point checklist. Beside malignant melanoma, it was vital to rule out vascular origin lesion including angiosarcoma which can start as a small nodule, grow rapidly and tend to ulcerate. Other features such as lymph node metastasis, bruising, telangiectasia and lymphedema were not found on our patient which make it unlikely to be angiosacroma.

**Results:**

Patient subsequently received an ultrasound scan which revealed a 12x18x18mm heterogeneous soft tissue mass on the left forearm. It demonstrated mild vascularity on Doppler scanning. Excision of the skin lesion performed, and the gross examination shows** a dark tan ellipse of skin measuring 40x18x15mm in depth bearing a raised area with haemorrhagic and cystic cut surface measuring up to 20mm. Histological examination revealed a well circumscribed dermal tumour. Within the center there is large and widely dilated and congested vascular channels surrounded by tumour cells arranging into anastomosing cords and bands. The tumor cells express CK7, S100, EMA (ductal differentiation) and show negative staining for Melan A. There is no atypia or malignancy. The lesion appears to be completely excised.

The diagnosis of vascular eccrine spiradenoma was made. Given it is a benign skin lesion, referral to 2-week-wait soft tissue sarcoma wasn’t required. During post excision review, the wound on the left forearm was healed completely. There was no residual lesion and no worrying signs under dermatoscope. Patient was discharged back

to her GP with regular monitoring and a safety netting advice.

**Conclusion:**

The rarity of VES poses diagnostic challenges, and histopathological examination remains essential for accurate diagnosis. Further research is needed to better understand the epidemiology, pathogenesis, and optimal management strategies for this rare tumor.

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**Abstract N°: 1824**

# Long-term efficacy and safety of JAKi in refractory livedoid vasculopathy: two cases and scoping review

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Long-term efficacy and safety of JAKi in refractory livedoid vasculopathy: two cases and scoping review

**Introduction & Objectives:**

Livedoid vasculopathy (LV) is a rare chronic ulcerative disease severely affecting patients' quality of life. To the authors' knowledge, there is no established therapeutic recommendation for LV, and anticoagulants and anti-inflammatory medication were both prescribed commonly. For such relapsing ulcerations, novel treatment is warranted, especially for treatment-resistant cases. Recently, Janus kinase inhibitors (JAKi) showed efficacy in dealing with LV, while the long-term follow-up profiles are lacking.

**Materials & Methods:**

This study reported two Chinese females with refractory LV treated with JAKi for 75 and 94 weeks, respectively. Also, we did a scoping review of the efficacy and safety of JAKi in patients with LV.

**Results:**

In both cases, we noted that the ulcers and pain were relieved significantly in the progressive stage, while recurrences occurred due to the medication self-withdrawal or COVID-19 infection. One patient reported nausea and dizziness during the treatment of tofacitinib, which resulted in drug withdrawal, and she and another patient denied any adverse event in the therapeutic session of baricitinib.

**Conclusion:**

In conclusion, our observations supplemented the long-term efficacy and safety of JAKi in LV, indicating that JAKi could be a potent alternative therapeutic option for refractory livedoid vasculopathy. In the future, long-term, large-scale clinical trials are expected.

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**Abstract N°: 2174**

# Unique presentation of Schamberg's disease: a clinical case

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**Introduction & Objectives:**

Pigmented purpuric dermatoses (PPD) represent a diverse group of rare inflammatory skin conditions characterized by a symmetrical eruption of petechial and pigmentary macules primarily affecting the lower limbs. Classified into five distinct types based on rash morphology, location, distribution, and accompanying symptoms, Schamberg's disease emerges as the most prevalent subtype.

**Materials & Methods:**

**Results:**

We report the case of a 46-year-old male presenting with a one-month history of symmetric purpuric and brown macules and patches localized to the penile glans and foreskin, gluteal, inguinal, and axillary creases, as well as the lower limbs. Dermoscopy showed coppery-red pigmentation in the background, red-brown dots and globules, as well as linear vessels. Bloodwork was unremarkable and systemic symptoms were absent. The diagnosis of Schamberg's disease with an uncommon topography was proposed based on physical examination and dermoscopy findings. Subsequent confirmation was achieved through cutaneous biopsy and histopathological examination, revealing a superficial, perivascular, interstitial lymphohistiocytic inflammatory infiltrate, along with evidence of extravasation of red blood cells and hemosiderophages (Pearls +), without any epidermal alterations.

**Conclusion:**

This case underscores an unique presentation of un uncommon condition, with only one case reported to date.

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**Abstract N°: 2179**

# A case report of generalized essential telangiectasia with an atypical presentation

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**Introduction & Objectives:**

Generalized essential telangiectasia (GET) is a rare, clinically benign condition of unknown etiology, characterized by widespread telangiectasias without systemic manifestations. Typically, lesions develop on lower limbs, gradually extending upwards to the trunk and upper arms.

**Materials & Methods:**

**Results:**

We report the case of a 76-year-old man with a long-standing history of telangiectatic patches on his abdomen, progressively extending to his upper and lower extremities. Remarkably, he had no familial or personal history of recurrent hemorrhage or telangiectasis, and his overall health remained unaffected. Additionally, his bloodwork results were within normal ranges. Diagnosis of generalized essential telangiectasia was established through clinical presentation, dermoscopy and histopathological examination.

**Conclusion:**

While GET may pose cosmetic concerns, it is not associated with other complications or diseases. This case underscores the rarity of this disease, which herein had a striking presentation. As such, we raise awareness for an overlooked condition.

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**Abstract N°: 2806**

# Infantile hemangiomas of the gluteal and perianal region: case series

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**Introduction & Objectives:**

Infantile hemangiomas (IHs) of the gluteal/perianal region are classified as high-risk for the development of ulcerations. Additionally, IHs in these regions can be associated with various congenital malformations of the lower half of the body, within the so-called LUMBAR syndrome.

**Materials & Methods:**

Over a 12-year period (January 1, 2012, to December 31, 2023), we identified all the IHs located in the gluteal/perianal region among infants who were treated and followed up at our university tertiary-level clinic.

**Results:**

During the study period, we found a total of 9 infantile hemangiomas (IHs) in the gluteal/perianal region. Among the affected infants, there were 5 boys and 4 girls. Six infants were born preterm, with 5 cases being very preterm (less than 32 gestational weeks). All presented infants had only one IH in the gluteal/perianal region, which was solitary in all but two cases, where an additional IH was found on another region (one on the trunk, one on the thigh). There were no additional congenital anomalies or internal organ IHs in any of the presented infants. In 3 cases, IH was complicated by ulceration and treated with oral propranolol. Among infants with ulcerated IHs, there were 2 girls and one boy, all born very preterm. In two cases, the maximum size of the IH was 6 cm, and in one case, it was 4 cm. All other non-ulcerated IHs were sizes up to 3 cm.

**Conclusion:**

Although comprising only nine cases, our case series supports the observation that IHs in the gluteal/perianal region are high-risk for ulceration, especially in infants born with low gestational age and birth weight, and in IHs larger than 3 cm.

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**Abstract N°: 2807**

**Octreotide treatment in a Hennekam syndrome case : Follow up of 9 years**

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**Introduction:** Hennekam syndrome HS is an autosomal recessive disorder resulting from malformation of the lymphatic system. It is very rare, appears in any ethnic group (only 100 cases reported). HS can be caused by bi-allelic variants in CCBE1 (18q21.32), FAT4(4q28.1)or ADAMTS3(4q13.3). The characteristic signs of HS are lymphedema, lymphangiectasia, facial anomalies and mental retardation. Lymphedema concerns mainly lower limbs and genitalia but can be seen elsewhere like ascite. Lymphangiectasias mostly described in intestines leading to protein-losing enteropathy can also occur in lungs, pericardium, thyroid and kidneys. Facial features are characterized by a flattened appearance of the face, a broad depressed nasal bridge, hypertelorism, epicanthal folds and a small mouth. The collagen and calcium-binding EGF domain-containing protein 1 (CCBE1) gene encodes a putative extracellular matrix protein crucial for lymphangiogenesis. Ccbe1 mutant mice lack lymphatic vessels and develop edema. There is no specific treatment for this syndrome. We report the case of child with HS in whom supportive therapy and several vasculotrop molecules were not efficient resolving lymphoedema. Finaly octreotid gave a better response.

**Observation :** An infant 5 months old born to consanguineous parents was received in our consultation. Because of hydramnios, labor was induced at 32 nd week. She was born with generalized lymphedema and chylous ascite. Her sister born one year before would have died 3 days after birth in a similar clinical presentation. During hospitalization our patient presented hypoalbuminemia, urinary infection. She received antibiotics, albumin, peritoneal draining and aggressive diuresis (furosemide) with significante improvement. We could notice then a dysmorphic face with flat midface, hypertelorism, depressed nasal bridge, a bulbous nasal tip, and epicanthal folds. HS diagnosis was retained. Despite this treatment with adapted diet comprising fat (50% composed of medium-chain triglycerides) and liposoluble vitamins, follow up during 4 years showed relapsed chylous ascite requiring punctures. We then tried sildénafil during 6 months without improvement. Rapamycin was given as well with no results. As she was 9 years old she presented a severe pleural and pericardial effusion with recurrent ascite and hypoalbuminemia. We began then octreotide 200µg twice daily by intravenous route. A month later albumin normalized, peritoneal,pleural and pericardial effusions decreased but peripheral lymphedema persisted. After 2 months treatment patient discharged in good general condition . In this patient genetic testing lacked.

**Conclusion:** We report one child who was diagnosed as having HS on the basis of clinical diagnosis, showing protein loosing enteropathy due to primary intestinal lymphangiectaia associated to peripheral lymphatic vessels impairment causing mixed edema. Using occtreotide normalized serum albumine what improved pleural, peritoneal and pericardial effusions but not limbs lymphoedema. This is likely to be related to generalized maldevelopment of the lymphatic system seen in HS and the possibility that lymphatic function is not altered by octreotide in HS.

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**Abstract N°: 2816**

# Infantile Hemangiomas of the Face in Preterm Infants

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## Introduction & Objectives:

The aim of our report was to emphasize the significance and approach to infantile hemangiomas (IHs) of the face, given their cosmetically and potentially functionally sensitive nature, particularly in preterm infants.

## Materials & Methods:

We collected data on IHs in preterm infants over a 12-year period (2012–2023) who were treated and followed up at our university tertiary-care pediatric clinic. We selected infants with solitary or multiple IHs, with at least one located on the facial region (forehead, eyelids and periocular region, nose, cheeks, perioral region), with a maximum size of at least 10mm. We chose the 10mm cut-off for IH size considering the significant risk of complications posed by these IHs in infants younger than 3 months.

## Results:

We identified 11 preterm infants with one or more IHs on the facial region, with sizes ≥ 10 mm, comprising 6 girls and 5 boys. Four of these infants were born extremely preterm (with less than 28 gestational weeks). There were a total of 11 IHs on the face: in 9 infants, there was one solitary focal IH, in one infant there was one focal IH on the face and another on the trunk, while one patient had an IH with minimal or arrested growth. The IHs were located in 3 cases on the forehead, 2 on the nose, 3 on the cheek, and 3 on the eyelid. Therapy was administered in 9 cases – 7 infants were treated with oral propranolol, and 2 with topical timolol. Propranolol was used in all cases of potential complications such as obstruction of the visual axis or nasal patency, exulceration, or permanent cosmetic disorders. Only one IH underwent exulceration, which occurred in an extremely preterm infant in whom propranolol therapy had to be discontinued due to adverse side effects. In two untreated cases, IH therapy was proposed, but the parents did not opt for it.

## Conclusion:

IHs on the face in preterm infants represent a very delicate condition. Therapy should be proposed for every IH of this region with a size ≥ 10 mm due to the high risk of serious functional and/or cosmetic impairment.

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**Abstract N°: 3049**

**arrested growth hemangioma (IH-MAG) :DO NOT CONFLUENCE WITH capillary malformation**

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**Introduction & Objectives:**

Infantile haemangioma (IH) is the most common tumour in infants, usually absent at birth. It appears in the first few weeks of life and undergoes a phase of rapid growth, followed by a period of slow involution with possible residual lesions (1).

Abortifacient haemangiomas, a little-known entity, are distinguished by the absence of the proliferative phase and may be confused with certain vascular malformations. Diagnosis can be difficult and constitutes a real diagnostic challenge in the first few days of life.

We report the case of an infant who responded favourably to topical beta-blockers Translated with DeepL.com (free version)

**Materials & Methods:**

Our observation concerns a one-month-old girl, A.C., referred to our centre for treatment of a left frontal capillary malformation that may be part of a syndromic pattern. On clinical examination, there was a pink component with a vascular appearance, very reticulated, with warm telangiectasia on palpation (photo 1); with small proliferative activity at the periphery of the lesion, which appeared a few days after the initial consultation . The diagnosis of S1 infantile abortifacient haemangioma was accepted. Investigation for PHACES syndrome was negative.

The patient was put on timolol with a very good response after one month of treatment and disappearance of the superficial component which was observed to respond very well to treatment there was no rebound phenomenon when treatment was stopped .

**discussion**

Positive diagnosis of infantile haemangioma is generally easy.In difficult cases, confirmation by positive GUT 1 labelling is necessary.Plane angiomas belong more to the spectrum of vascular malformations, being present from birth and manifesting as pinkish to purplish spots commonly known as “wine spots” without relief and whose borders may be crumbled or well defined.

Our patient had an abortifacient infantile haemangioma with very minimal proliferative activity, which led to a misdiagnosis. In fact, minimal or arrested growth hemangioma (IH-MAG) is a recent entity defined as having a proliferative component equal to less than 25% of their total surface area. Clinically, it resembles the premonitory lesions that precede infantile haemangiomas or capillary malformations.

Abortifacient haemangiomas are often present at birth, which makes diagnosis even more difficult. In the case of segmental haemangiomas, they are often associated with PHACEs syndrome.

IH-MAGs are initially difficult to differentiate from capillary malformations, but the reticulated appearance, the presence of telangiectasias and above all the warm character on palpation indicate underlying haemodynamic activity in favour of a vascular tumour. In the event of clinical doubt, colour Doppler ultrasound can guide the

diagnosis.

Capillary malformations associated with RASA1 mutations share the same characteristics as abortifacient hemangiomas; however, they are generally multifocal and randomly distributed, which may help to distinguish them from IH-MAGs

**Conclusion:**

The diagnosis of IH-MAG is sometimes difficult because it is often confused with a capillary-type vascular malformation. This misdiagnosis can be medically serious, particularly in segmental IH-MAG involving the upper face and forehead, leading to unnecessary concern and work-up for Sturge-Weber syndrome, while ignoring the potential for PHACEs syndrome.

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**Abstract N°: 3296**

**Effect analysis of lauromacrogol foam injection under ultrasonic guidance for pediatric superficial lymphatic malformation**

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**Effect analysis of lauromacrogol foam injection under ultrasonic guidance for pediatric superficial lymphatic malformation**

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**Introduction & Objectives:**

To evaluate the efficacy and safety of ultrasound guided lauromacrogol foam sclerotherapy in the treatment of children's superficial lymphatic malformation.

**Materials & Methods:**

Retrospectively analyzing 64 pediatric patients (36 male, 28 female), aged 3 months to 14 years, who underwent lauromacrogol foam sclerotherapy with ultrasonic guidance for superficial lymphatic malformation at Hunan Children's Hospital from March 2018 to November 2022, the study grouped patients into 18 macrocystic, 40 microcystic, and 6 mixed types. Pre-sclerotherapy evaluations included local ultrasound, blood routine, liver and kidney function, and coagulation tests. The surface anesthetic lidocaine cream was applied 30 minutes prior, followed by ultrasound-guided lymphatic malformation area localization, cyst fluid extraction, and lauromacrogol foam injection (lauromacrogol: air = 1:3; single dose < 20 ml) to fill the cyst cavity, with retention of the lauromacrogol stock solution (< 2 ml). Assessing drug dispersion in the cyst cavity via ultrasound and confirming high-echo gas filling, the follow-up period ranged from 3 to 12 months, with evaluations on clinical manifestations, imaging data, treatment efficacy, and complications.

**Results:**

The children received treatment 2-6 times, with an average of 4 treatments per case. A total of thirty-three cases(51.56%) achieved complete remission, while twenty-eight(43.75%) cases showed significant improvement. Three cases failed to respond and subsequently underwent surgical resection. The overall efficacy rate was determined to be 95.31%. Early edema occurred in five cases, localized skin pigmentation in one case, thrombotic superficial vein inflammation in two cases, and transient cough in one case. None of the children experienced serious complications such as allergic reactions, deep vein thrombosis or pulmonary embolism, skin necrosis or discoloration, chest tightness or other cardiopulmonary issues.

**Conclusion:**

The utilization of ultrasound-guided foam sclerotherapy with lauromacrogol represents a secure and efficacious approach for managing lymphatic malformations in pediatric patients.

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**Abstract N°: 3461**

# Kasabach Meritt phenomenon : a 4 patient case series of a potentially lethal entity

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**Introduction & Objectives:**

Kasabach-Merritt phenomenon (KMP) is a rare, life-threatening vascular condition in infants, involving platelet entrapment in vascular tumors particularly tufted angioma and kaposiform hemangioendothelioma (KHE). Prognosis factors and long-term follow-up data are lacking, and optimal treatment is still under discussion.

**Materials & Methods:**

We conducted a retrospective descriptive study of the clinical, biological and therapeutic aspects of KMP patients in our department.

**Results:**

Four children (1F/3M) had congenital (n=3) or early-onset tumors (n=1) with KMP.

KMP onset averaged 45 days post-tumor detection (0-5 months). Features included a rapidly enlarging plaque with telangiectasias (n=2), and superficial papules (n=1), with an average tumor size of 10 cm.

Thrombocytopenia (average: 31,000/mm3) with elevated D-dimers lasted 26.2 days on average post-treatment initiation.

Imaging (n=3) revealed tumor infiltration involving muscles, with bone extension without lysis in one case.

Initial treatment with systemic corticosteroids (SC) (2 to 5 mg/kg/day) and antiplatelet agents (APA) showed improvement, but two relapsing cases required sirolimus or vincristine.

One patient had persistent tumor enlargement, hypoparathyroidism, hypercalcemia with fatal complications.

All our cases were maintained on APA, with one relapse after discontinuation, improving upon reintroduction.

Two patients experienced late-onset muscular pain (7 years), alleviated with short-term SC, APA and physical therapy.

**Conclusion:**

The severe prognosis of KMP contrasts with the favorable course observed in our series (3/4).

KHE, the most common implicated tumor, is locally aggressive and potentially infiltrative, as seen in our lethal case.

The intensity of KMP varies depending on the tumor’s ability to trap platelets, which is dependent on tumor mass and infiltration level, constituting a prognostic element for KMP severity. This is illustrated by fatality related to the largest tumor in our series.

Cases of KMP associated with consumptive hypothyroidism are extremely rare. Only one case has been found in

the literature of hypothyroidism normalizing after surgical excision. The diagnostic hypothesis of a paraneoplastic phenomenon remains likely. We report the first case of hypoparathyroidism associated with a KMP.

Large series showing permanent coagulopathy (elevated D-dimer) even after resolution of KMP suggest the presence of chronic low-grade platelet trapping, with possible sudden worsening, and raises the possibility of prophylactic anti-platelet therapy as shown efficient in our case series. The recognition of minimal coagulopathy in a susceptible tumor may indicate the need for prophylactic antiplatelet drug treatment.

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**Abstract N°: 3581**

# Two rare familial cases of capillary-arteriovenous malformations (CMAVM).

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**Introduction & Objectives:**

CMAVM is an acronym for multiple cutaneous capillary malformations (CMs) and a risk of having one or many malformations and arteriovenous fistulae that may be present in different sizes and locations.

We describe two rare cases of CMAVM in two sisters aged 3 and 6 respectively.

**Materials & Methods :**

This is the case of two sisters aged 3 and 6, from a non-consanguineous marriage, who presented a few months after birth with a similar symptomatology consisting of non-pruritic erythematous macules with anarchic and diffuse extension all over the body.

The dermoscopy revealed a similar appearance in both patients, with a mixed vascular pattern of arborescent telangiectasias associated with a homogeneous, brownish, non-networked pigmented pattern known as café-au-lait staining.

We performed a skin biopsy with immunohistochemistry, including staining for CD 31, 34 and 117, enabling us to rule out mastocytosis and to hold CMAVM as the most likely diagnosis.

Following this, we indicated the performance of cerebral-medullary angio-MRI under sedation, as well as karyotyping and molecular sequencing in both patients, that are still in progress.

Finally, the decision was made to abstain from treatment, respecting the choice of the parents, who had refused the laser sessions.

**Results:**

CM-AVM a rare, autosomal dominant genetic disease discovered in 2003, as a rasopathy.

In 2017, two CM-AVM subtypes based on pathogenic variants of two different genes: RASA1 and EPHB4 were identified, with a similar clinical appearance and an onset from birth or early childhood.

Preferential localization remains the face, trunk and extremities, with the following elementary lesions: multifocal vascular macules ranging in size from 1 to 15 cm.

Dermoscopically, the café-au-lait sign has been described as highly suggestive of CMAVM, and is common to neurofibromatosis type 1, which is also a rasopathy.

Positive diagnosis is based on clinical and genetic studies, while histology and immunohistochemistry can be useful remain non-specific.

Numerous studies have also highlighted the importance of cerebral imaging, given the significant association of arteriovenous malformations and fistulas with CMAVM.

As far as long-term evolution is concerned, clinical and radiological monitoring is of prime importance, given the dynamic nature of CMAVMs and the fact that normal angio-MRI cannot eliminate the risk of developing vascular anomalies in the long term.

Treatment is essentially based on vascular lasers, although other molecules targeting the RAS signaling pathway are still under investigation.

**Conclusion:**

It’s important not to completely reassure parents in the front of normal brain imaging, and not to hesitate to repeat it in case of any warning sign.

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**Abstract N°: 3590**

**Histopathological and ultrastucural study for treating Port wine stains by Pulsed dye laser.**

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**Introduction & Objectives:**

Port-wine Stain (PWS) is usually present at birth affecting 0.1 % to 2 % of population , usually pale pink or deep red or purple in color, varies in size from few mms to large cms in diameter which do not change with age, and usually starts to be darken, raised, thickened slowly and progressively by age.

Trauma may lead to PWS Stain formation in older ages, with an identical clinical picture to the classic PWS.

(PDL) is considered one of the best treatment choices for PWS, however some lesions doesn't respond completely.

To evaluate the efficacy of pulsed dye laser (PDL) in the treatment of port wine stain (PWS) by histopathological and ultrastructural assessment.

**Materials & Methods:**

201 patients with PWS, 196 with congenital PWS and 5 with Aquired PWS was treated with PDL 585 and 595nm wavelengths, 450 msec pulse duration for a bi-monthly laser session.

Selected patients used Oxygen mask to increase their oxy-haemoglobin oxygen tension and monitored by Pulse Oxymeter

Clinical response and histopathological examination before and after treatment using punch skin biopsies for H&E, Orcein and Masson trichrome special staining was done.

Ultrastructure assessment by Transmission Electron Microscopy before and after PDL treatment.

**Results:**

16.9% of cases showed excellent response, 33.1% showed very good result, 41.9% showed good response, 8.1% showed no – poor response with an average of 10.1 sessions,

H & E examination showed decrease in the size of the ectatic blood vessels with a maximal effect on superficial blood vessels than deeper vessels,

Partial degeneration was observed by EM of deeper vessels, the endothelial surface facing laser beam showed a maximal damage effect, while the opposite side showed a minimal one, this effect is due to the shielding effect.

Perivascular mast cells, elastic fiber, oesinophil were seen around blood vessels.

while 5 patients with acquired PWS were treated by PDL 33.3% of patients had excellent result, and 66.6% patients had very good results with an average 4.8 sessions.

201 patients with PWS was treated, 148 patients ended this study. Females had significantly higher v. good and good prognosis than males ($P < 0.0005$)

71.1% were females, 53.2% of them were skin type III, 35.3% were between 20 – 30 years, only 8.1% of patients only showed no response, the rest showed different types of good and excellent response.

**Conclusion:**

PDL which depends upon the principle of selective photothermolysis revolutionized the treatment of PWS.

However some PWS are resistant to PDL treatment due to deeper blood vessels as proved by histopathological and by E/M study and may need more longer wave length to be combined for better deeper penetration.

Although laser treatment is not perfect, it is extremely safe and allows for improvement or complete clearance of previously untreatable abnormalities.

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**Abstract N°: 3921**

# Exploring the Etiological Spectrum of Cutaneous Vasculitis: Findings from a Hospital-Based Retrospective study

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**Introduction & Objectives:**

Cutaneous vasculitis (CV) represents a spectrum of disorders characterized by inflammatory processes affecting vessel walls, leading to diverse clinical presentations. Understanding the etiological factors contributing to CV is crucial for tailored management strategies.

This study aims to provide a comprehensive overview of the clinical, etiological, histological, and biological characteristics exhibited by patients diagnosed with CV.

**Materials & Methods:**

This retrospective study, conducted from January 2021 to December 2023, involved all histologically confirmed CV cases hospitalized in our Dermatology Department. Data, including sociodemographic, clinical, and examination results, were collected from hospital records using a standardized form.

**Results:**

During the study period, forty-three cases of CV were identified with a sex ratio of 0.53 (M/F). The mean age of patients was 53 years, ranging from 18 to 85 years. Personal medical histories were documented in 55.8% of subjects, among which hypertension and diabetes were associated with CV in 23.3% of cases. The median duration of symptom evolution was 3 weeks, ranging from 1 day to 4 years. Cutaneous lesions were predominantly purpuric (60.5%) and necrotic in 18.6% of cases. Other cutaneous lesions observed included urticarial papules (7 cases), blisters (5 cases), pustules (4 cases), and ulcerations (3 cases). Cutaneous lesions were localized on the lower limbs in 86% of cases, upper limbs in 46.5% of cases, trunk in 34.9%, and face in 11.6%. Systemic manifestations were observed in 11.6% of patients, notably arthralgia (9.3%) and digestive involvement (4.7%). Only 7 patients (16.3%) presented with biological inflammatory syndrome. Leukocytoclastic vasculitis (LCV) was the predominant histological type, found in 53.5% of skin biopsies. The etiological assessment concluded idiopathic cutaneous vasculitis in 48.8% of cases. CV was drug-induced in 20.9% of cases and infectious (2 cases). Pharmacovigilance investigation implicated 6 drugs, including allopurinol (2 cases), amoxicillin, an SSRI, and an antihypertensive. Responsible infectious agents were viral (1 case) and parasitological (1 case). Systemic vasculitides accounted for 16.3% of CV cases. These included rheumatoid purpura in 6 cases (13.9%).

**Conclusion:**

Our results corroborate literature data regarding female predominance and the diverse etiologies of CV. This study highlights the frequency of iatrogenic forms and underscores the necessity of a multidisciplinary approach for CV management, in line with current recommendations.

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**Abstract N°: 3946**

# The hidden etiology beneath the skin: a case study of Sneddon's syndrom

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**Introduction & Objectives:**

Sneddon's Syndrome is a rare condition primarily affecting young women. Its estimated incidence is 4 per 1 million per year in the general population, and it is characterized by the presence of livedo racemosa and central nervous system disorders.

**Materials & Methods:**

We report the case of a 46-year-old woman with a history of head trauma 17 years ago. She was admitted to the neurology department for the management of progressive left hemiparesis over the past year. Neurological examination revealed left hemiparesis, cognitive impairment, aphasia with a tetrapyramidal syndrome, and spastic hypertonia.

Dermatological examination revealed fixed livedo racemosa on the lower limbs, with thick incomplete meshwork that did not disappear upon warming.

**Results:**

Neurological exploration by brain MRI revealed significant cerebral atrophy with multiple leukoaraiosis. Cerebrospinal fluid examination was strictly normal. The patient did not have thrombocytopenia, and immunological tests for antiphospholipid antibodies were negative.

Skin biopsy showed pseudo-intimal hyperplasia without signs of associated vasculitis.

Given the combination of livedo racemosa, neurological symptoms, white matter changes on brain MRI, negative immunological assessment, and histological findings, a diagnosis of Sneddon's syndrome was established.

**Conclusion:**

Sneddon's syndrome is a rare, non-inflammatory occlusive vasculopathy. The most common non-neurological manifestation is livedo racemosa, characterized by a wide-meshed net-like pattern, either generalized or localized, without infiltration. Currently, there is no specific criterion for Sneddon's syndrome, but the presence of multiple criteria is suggestive. Therefore, the occurrence of a stroke or neurological symptoms in a young individual with livedo, in the absence of vascular causes, embolic cardiopathy, or blood disorders, supports the diagnosis of Sneddon's syndrome.

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**Abstract N°: 3969**

# Complete disappearance of all rashes of purpura annularis telangiectodes (Majocchi disease) at the peak of fever due to COVID-19: A Case Report

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**Introduction & Objectives:**

Purpura annularis telangiectodes (Majocchi disease) is a pigmented purpuric dermatosis, first described by Domenico Majocchi in 1896, manifested by annular, symmetrical, purpuric, telangiectatic patches of different diameters. Linear or arcuate patches may also be present. The most common site of damage is the lower extremities. Spread to the upper limbs and trunk may occur. Skin lesions of purpura annularis telangiectodes are usually asymptomatic but sometimes itching. The course is chronic, characterized by relapses and remissions.

In this article, I present an interesting case of the rapid disappearance of all lesions of purpura annularis telangiectodes in a 21-year-old man at the peak of fever due to COVID-19.

**Materials & Methods:**

This abstract presents an interesting case of a 21-year-old man diagnosed with purpura annularis telangiectodes. In mid-December 2023, the patient developed the first elements of a rash on the lower limbs, accompanied by slight itching and several patches on the trunk, which were not bothersome. The patient could not associate the initiation of this condition with anything; he denied all known causes, such as significant physical exertion, infectious processes, alcohol consumption, chemical ingestion, drugs, and vaccines for at least the last three months. The patient has not been diagnosed with systemic diseases, and additional examination revealed no abnormalities in essential laboratory examinations.

**Results:**

At the end of December, the patient was prescribed topical corticosteroids. At the end of January 2024, the patient applied for a repeat examination; the rash had significantly spread to the trunk and upper limbs, and he was bothered by minor itching and aesthetic appearance. Treatment with narrowband ultraviolet B (NBUVB) phototherapy was prescribed. The patient attended one session and fell ill with COVID-19, due to which the course of phototherapy was temporarily stopped. The course of the coronavirus disease was severe, and at the peak of the fever of about 40 degrees, the gradual disappearance of the rash became noticeable. When the patient's temperature normalized, the purpura annularis telangiectodes rash, which existed for about two months, completely disappeared and did not appear again.

**Conclusion:**

This interesting case raises many questions about the pathogenetic connection between purpura annularis telangiectodes and coronavirus disease. It's common knowledge that fever can be beneficial because many viruses multiply less well at elevated temperatures, so that fever can reduce viral load. But what exactly played a role in the recovery from purpura annularis telangiectodes remains a mystery. The value of this case lies in its paradoxical nature, which provides food for thought for physician-scientists and clinicians.

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**Abstract N°: 4042**

# Ciprofloxacin-induced leukocytoclastic vasculitis in elderly man.

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**Introduction & Objectives:**

Leukocytoclastic vasculitis is a cutaneous small vessel vasculitis of the dermal capillaries and venules. Many classes of antibiotics, nonsteroidal anti-inflammatory agents, antiepileptic drugs, propylthiouracil, and omeprazole can cause leukocytoclastic vasculitis. There are several cases described in the medical literature of ciprofloxacin - induced leukocytoclastic vasculitis. We report a case of a patient with a palpable purpura who developed leukocytoclastic vasculitis.

**Materials & Methods:**

A 82-year-old white man presented with purpuric palpable rash distributed predominantly over the lower legs. Laboratory investigations revealed elevated erythrocyte sedimentation rate. Histopathological examination demonstrated a mild superficial perivascular lymphoid cell infiltrate with admixed neutrophils and apoptotic debris and abundant extravasated erythrocytes, consistent with leukocytoclastic vasculitis. Patient developed complete resolution of the lesions on ciprofloxacin withdrawal.

**Results:**

The development of a purpuric rash is uncommon and it presents a diagnostic challenge to physicians. Most common clinical manifestation of leukocytoclastic vasculitis is palpable purpura. The disease is usually limited to skin, pain or burning may be present, and systemic involvement is uncommon. Most cases resolve within months but it can be life threatening if it affects lungs or brain.

Therapeutic options for systemic involvement include corticosteroids, colchicine, dapsone, or other immunosuppressive agents. No other specific therapy is usually necessary for patients with skin-limited disease.

**Conclusion:**

This case further emphasizes that caution should be used when prescribing ciprofloxacin to the elderly patients. It highlights an association between ciprofloxacin and leukocytoclastic vasculitis. Clinicians should be aware of this rare but serious cutaneous adverse effect.

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**Abstract N°: 4108**

**Atypical Cobb syndrome treated with sirolimus**

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**Introduction & Objectives:**

Cutaneomeningospinal angiomatosis, also known as Cobb syndrome, describes the presence of spinal arteriovenous malformation (AVM) and a vascular skin lesion affecting the corresponding dermatome. We present here a new case with an unusual clinical presentation that showed significant improvement with sirolimus treatment.

**Observation:**

A 47-year-old female patient, with a history of consanguinity, presented with a vegetating, dark violaceous papulo-nodular angiomatous lesion, infiltrated and occasionally ulcerated, extending from the right hypochondrium to the dorsal region corresponding to D10-D11. There was no thrill on auscultation. This lesion was associated with paraplegia due to established spinal cord compression 6 years ago. History revealed a dorsal metameric blue macule at the same level, along with congenital scoliosis. The remainder of the clinical examination was unremarkable.Angio-MRI confirmed the presence of a spinal arteriovenous malformation. A diagnosis of Cobb syndrome was established. The patient was successfully treated with sirolimus.

**Conclusion:**

Cobb’s syndrome is a rare, non-inherited disorder which represents the concurrent findings of a spinal AVM in the same metamere as the cutaneous lesion, which is explained by the embryological origin of blood supply to the vertebrae and spinal cord from segmental dorsal arteries. This syndrome typically manifests most often in late childhood but can occur at any age.Cutaneous manifestations, including macular port-wine stains and various types of papular or nodular vascular lesions such as angiomas, angiokeratomas, angiolipomas, and lymphangioma circumscriptum, do not tend to regress spontaneously. Diagnosis is often prompted by the appearance of cutaneous symptoms such as bleeding with trauma, or neurological symptoms ranging from muscle weakness to paraplegia or even tetraplegia. These symptoms are not solely due to compression by the spinal AVM; other factors such as venous hypertension and spinal cord ischemia are also considered potential mechanisms of myelopathy. Bladder and bowel dysfunction typically occur later in the disease course. Treatment usually involves embolization to address the spinal arteriovenous malformation.

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**Abstract N°: 4209**

# Assessing Risks and Benefits of Beta-Blocker Therapy in PHACE Syndrome with Arteriopathy

Maroua Sakhri*[1], Nabila Damou[1], Sara Benrejdal[1], Aicha Salhi[2], Houria Sahel[1]

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**Introduction & Objectives:** Infantile hemangioma (IH) stands as the most prevalent tumor in infants, with propranolol emerging as the primary treatment option. Notably, segmental facial IH, prone to complications, frequently exhibit a remarkable response to propranolol. However, these hemangiomas also entail the risk of PHACE syndrome. Here, we present a case of PHACE syndrome successfully managed with propranolol.

**Materials & Methods:** A female infant, 40 days old, was born from a pregnancy complicated by hypothyroidism in the mother and delivered by cesarean section due to fetal distress. She was admitted to the hospital due to a segmental infantile hemangioma affecting segments S1, S2, and S4 of the right hemiface, which began evolving at the age of 5 days. This condition was further complicated by bleeding, ulceration, and ocular obstruction. Additionally, a small superficial infantile hemangioma was noted at the occiput. Ophthalmological examination yielded normal results. Electrocardiogram, cardiac assessment, and abdominopelvic ultrasound showed no abnormalities. Cerebral angio-MRI revealed a hemangioma orbitofacial, a reduced caliber intracranial carotid artery (patent), and a non-expanding arachnoid cyst of the posterior cranial fossa. The diagnosis of PHACES syndrome was established, and the child was started on propranolol at a dosage of 0.5 mg/kg/day, divided into 6 doses, with subsequent weekly increases of 0.5 mg/kg. The treatment was well tolerated, and initial clinical improvement was observed.

**Results:** The acronym PHACES encompasses anomalies of the posterior fossa, facial hemangioma, intra and extracranial arterial anomalies, congenital cardiac anomalies and aortic coarctation, ocular anomalies, sternal and ventral anomalies. It's widely recognized that children with large segmental facial hemangiomas are at risk for PHACE. The results of initial screening examinations should guide therapeutic management and monitoring. Initial imaging of the cervical and cerebral arteries can aid in categorizing patients into low, intermediate, or high risk of stroke. Propranolol is considered the first-line treatment in patients with infantile hemangioma (IH) requiring systemic treatment. However, its use in patients with PHACE and arterial abnormalities with a risk of stroke is controversial. Lower doses or a slower gradual increase should be considered, and patients should be closely monitored. The total daily dose should be divided into 3 doses, and adjustments can be made based on clinical assessment

**Conclusion:** While PHACE syndrome doesn't necessarily prohibit the use of beta-blockers outright, it's crucial to carefully assess the benefits and risks. It's recommended that infants with substantial facial hemangiomas undergo thorough examination for PHACE syndrome, particularly focusing on the presence of arteriopathy.

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**Abstract N°: 4219**

# Infantile Hemangiomas Profile: A Study of 60 Hospitalized Cases

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**Introduction & Objectives:** Infantile hemangioma (IH) is the most common tumor in infants. The aim of this study was to establish the epidemiological and clinical profile of IH cases requiring hospitalization

**Materials & Methods**: We conducted a single-center descriptive cross-sectional study comprising a retrospective part (December 2019 to March 2023) and a prospective part (April 2023 to March 2024). Included patients were those hospitalized for the management of IH. The variables studied were epidemiological and clinical.

**Results:** Sixty patients were enrolled in this study, with 24 during the retrospective period and 36 during the prospective period. The mean age was 7.18 months, ranging from 1 to 24 months. The most affected group was 0-5 months aged patients (46.7%). The sex ratio was 0.2. Notably, 80% of patients experienced symptom onset before the 15th day of life. Among them, only one patient had low birth weight, while three were born prematurely, and two were from complicated pregnancies. Regarding the characteristics of IHs, 68.3% of patients had a mixed IH, 23.3% had a tuberous IH, and 8.3% had a subcutaneous IH. Distribution analysis revealed that 95% of cases were localized IHs, with only 5% (3 cases) being segmental. IHs were predominantly located on the head (50%), followed by the upper limb (6 patients), lower limb (10 patients), trunk (7 patients), and genital organs (12 patients). Additionally, rare cases of PHACES syndrome (2 cases) and SACRAL syndrome (1 case) were identified.

**Conclusion:** We provide an epidemiological overview of hospitalized children with IH in our department, showing findings in line with existing literature.

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**Abstract N°: 4257**

# Use of Propranolol in the Management of PHACE Syndrome: About 2 Cases

Maroua Sakhri*[1], Nabila Damou[1], Sara Benrejdal[1], Aicha Salhi[2], Houria Sahel[1]

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**Introduction & Objectives:** Segmental infantile hemangiomas (IH) located on the face pose an elevated risk of PHACE syndrome. Prescribing propranolol in such cases requires caution due to a heightened risk of stroke. We present two cases of PHACE syndrome effectively managed with propranolol treatment

**Materials & Methods: CASE 01:** A female child, 40 days old, born from a pregnancy complicated by fetal distress, was hospitalized due to an IH affecting the S1, S2, and S4 segments of the right hemiface. The IH had been evolving since the age of 5 days and was complicated by ulcerations and ocular obstruction. Ophthalmological examination, electrocardiogram, cardiac and abdominopelvic ultrasound showed no abnormalities. Brain angio-MRI revealed the presence of an orbito-facial hemangioma, a reduced intracranial carotid artery, and a non-expanding arachnoid cyst in the posterior cerebral fossa. The diagnosis of PHACE syndrome was established. Propranolol was initiated at a dose of 0.5 mg/kg/day, divided into 6 doses, and then gradually increased by 0.5 mg/kg/week. The treatment was well-tolerated, and there was improvement in the IH. **CASE N02:** A 3 months female child, born from a pregnancy complicated by chorioamnionitis and respiratory distress, was hospitalized due to two roughly symmetrical bilateral subcutaneous hemangiomas located at the level of the S3 segment, which had been present since birth. The right hemangioma measured 5 cm in length, while the left 4 cm. Cutaneous aplasia over the sternum was also observed. During this hospitalization, the infant presented several episodes of respiratory distress. ENT examination revealed a hemangioma of the lower lip extending to the gum, and nasofibroscopy showed laryngomalacia. Ophthalmological examination yielded normal results. Head and neck MRI confirmed the diagnosis, while brain MRI ruled out any anomalies. Cardiac ultrasound showed an exclusive left-right shunt with a restrictive profile. The diagnosis of PHACE syndrome was established. Propranolol treatment was initiated, following the elimination of any contraindications, at a dose of 0.5 mg/kg/day, gradually increased with good tolerance. The absence of respiratory distress attacks was noted since the introduction of treatment.

**Results:** The acronym PHACES encompasses anomalies of the posterior fossa, a facial hemangioma, intra and extracranial arterial anomalies, congenital cardiac anomalies and aortic coarctation, ocular anomalies, and sternal and ventral anomalies. Children with large segmental facial IH are at risk for PHACE syndrome. Initial screening examinations should guide therapeutic management and monitoring. The use of propranolol is contentious in cases of PHACE with arterial abnormalities due to the risk of stroke. Consideration should be given to lower than standard daily doses or a gradual, slower increase, with closer monitoring. The total daily dose should be divided into 3 doses, with adjustments based on clinical response. In a series of 32 PHACE cases with arterial abnormalities treated with propranolol, the majority of patients tolerated the treatment well.

**Conclusion:** PHACE syndrome does not represent an absolute contraindication to the use of beta-blockers. However, the benefits and risks should be carefully assessed and weighed.

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**Abstract N°: 4275**

**Experience with rapamycin in bean syndrome**

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**Introduction & Objectives:** Blue rubber bleb nevus syndrome (BRBNS) or Bean syndrome is a rare entity included in the classification of the International Society for the Study of Vascular Abnormalities (ISSVA). It is characterized by venous malformations that affect the skin and visceral organs due to spontaneous de novo somatic mutations that affect the TEK gene which encodes TIE2 that activates the PI3K/AKT/mTOR signaling pathway.

The lesions appear as violaceous subcutaneous tumors (type I lesions), small, painful, bluish or purplish nodules that are depressible, shaped like a rubber nipple (type II) and/or bluish macules (type III). Their size and number increase directly proportional to age. Malformations that affect internal organs can cause hemorrhages that can lead to death.

Previously, treatment was limited to local management of cutaneous vascular malformations with vascular laser or surgical resections, control of anemia secondary to bleeding resulting from gastrointestinal malformations through photocoagulation, laser and sclerotherapy. Nowadays, recent knowledge of the pathogenesis has allowed the treatment of VMs with molecular inhibitors such as Sirolimus, a drug that inhibits the m-TOR pathway involved in angiogenesis, demonstrating excellent results in these patients by reducing the size of the malformations, morbidity and increase survival.

**Materials & Methods:** A 25-year-old female patient with a diagnosis of BRBN, with skin involvement due to venous malformations (type I and II), right upper limb amputation due to a large venous malformation that limited her functionality at the age of 4 years and venous vascular malformations on intestinal tract with active bleeding that led her to multiple hospitalizations with transfusion requirements given the severe iron deficiency anemia she presented (Hemoglobin (Hb) 4 gr).

**Results:** Taking into account the clinical repercussion, it was decided to start systemic therapy with Sirolimus at a dose of 1 mg/day initially for 1 year, with excellent development but after treatment suspension she presented new episodes of bleeding from the digestive tract that required hospitalizations and transfusions. Therefore, Sirolimus was restarted, readjusting the dose to 2 mg every 24 hours, with a favorable response, without subsequent bleeding, a decrease in the size of the skin and digestive malformations, reaching a Hb value of 14 g/dl. After 6 months of continuous treatment, due to the progressive decrease in cutaneous vascular tumors, intervention with Nd-YAG vascular laser was possible.

**Conclusion:** BRBNS syndrome is a rare entity and a diagnostic and therapeutic challenge for the dermatologist. Knowledge of the pathogenesis of the disease and the availability of new drugs such as Sirolimus has made it possible to address the management of patients in a comprehensive and multidisciplinary manner. Sirolimus, is a drug that dermatologists must know and handle in order to intervene effectively in this type of entities.

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**Abstract N°: 4277**

# Atypical Cobb syndrome treated with sirolimus

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**Introduction & Objectives:**

Cutaneomeningospinal angiomatosis, also known as Cobb syndrome, describes the presence of spinal arteriovenous malformation (AVM) and a vascular skin lesion affecting the corresponding dermatome. We present here a new case with an unusual clinical presentation that showed significant improvement with sirolimus treatment.

**Observation:**

A 47-year-old female patient, with a history of consanguinity, presented with a vegetating, dark violaceous papulo-nodular angiomatous lesion, infiltrated and occasionally ulcerated, extending from the right hypochondrium to the dorsal region corresponding to D10-D11. There was no thrill on auscultation. This lesion was associated with paraplegia due to established spinal cord compression 6 years ago. History revealed a dorsal metameric blue macule at the same level, along with congenital scoliosis. The remainder of the clinical examination was unremarkable.Angio-MRI confirmed the presence of a spinal arteriovenous malformation. A diagnosis of Cobb syndrome was established. The patient was successfully treated with sirolimus.

**Conclusion:**

Cobb’s syndrome is a rare, non-inherited disorder which represents the concurrent findings of a spinal AVM in the same metamere as the cutaneous lesion, which is explained by the embryological origin of blood supply to the vertebrae and spinal cord from segmental dorsal arteries. This syndrome typically manifests most often in late childhood but can occur at any age.Cutaneous manifestations, including macular port-wine stains and various types of papular or nodular vascular lesions such as angiomas, angiokeratomas, angiolipomas, and lymphangioma circumscriptum, do not tend to regress spontaneously. Diagnosis is often prompted by the appearance of cutaneous symptoms such as bleeding with trauma, or neurological symptoms ranging from muscle weakness to paraplegia or even tetraplegia. These symptoms are not solely due to compression by the spinal AVM; other factors such as venous hypertension and spinal cord ischemia are also considered potential mechanisms of myelopathy. Bladder and bowel dysfunction typically occur later in the disease course. Treatment usually involves embolization to address the spinal arteriovenous malformation.

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**Abstract N°: 4288**

**A Rare Case of Sturge-Weber Syndrome Associated with Macrocheilitis**

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**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 a case of SWS

**Materials & Methods:** 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.

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**Abstract N°: 4300**

**Molecular Characterization of Laser-Resistant Port Wine Birthmark using Patient-derived Induced Pluripotent Stem Cells and their Differentiated Endothelial Cells**

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**Molecular Characterization of Laser-Resistant Port Wine Birthmark using Patient-derived Induced Pluripotent Stem Cells and their Differentiated Endothelial Cells**

**Introduction & Objectives:**

Port wine birthmark (PWB) is a congenital vascular malformation resulting from developmentally defective endothelial cells (ECs). Pulsed dye laser (PDL) is the treatment of choice for PWB. Unfortunately, complete removal of PWB occurs in less than 10% of patients. Approximately about 20% of lesions do not respond to laser treatment. The molecular mechanisms underlying laser-resistant phenotypes remain incompletely understood. The objective of this study is to develop clinically relevant disease cell models for PWB and explore whether these cell models can recapitulate PWB pathological phenotypes including laser resistance.

**Materials & Methods:** PWB iPSCs were generated by reprogramming lesional dermal fibroblasts and differentiated into ECs. The functional phenotypes of iPSC-derived ECs were characterized by capillary-like structure (CLS) formation *in vitro.* Indocyanine green (**ICG**)-mediated photodynamic therapy (**PDT**) was performed to evaluate cell response to laser-based treatment. Bulk RNA-seq was performed to identify differentially expressed genes (DEGs) and gene set enriched analysis (GSEA) in laser-resistant lines versus sensitive lines.

**Results:** Human PWB and control iPSC lines, termed as PWB_iPSC and Ctl_iPSC, were generated through reprogramming of dermal fibroblasts by introducing the "Yamanaka factors" (*Oct3/4, Sox2, Klf4, c-Myc)* into them; the iPSCs were successfully differentiated into ECs. In total, iPSC lines were generated from one normal subject (Ctl_iPSC_52521, 3 cell lines) and two PWB patients (PWB_iPSC_4221, 2 lines; and PWB_iPSC_3921, 3 lines), respectively. These iPSCs and their derived ECs were validated by expression of a series of stem cell and EC biomarkers, respectively. PWB iPSC-derived ECs (4221 and 3921) showed impaired CLS *in vitro* with larger perimeters and thicker branches* as compared to control iPSC-derived ECs (52521). Interestingly, PWB_iPSC_3921 lines and their derived ECs were resistant to PDT; while PWB_iPSC_4221 lines and Ctl_iPSC_52521 lines and their derived ECs were sensitive to PDT. Bulk RNA-seq and GSEA showed that the convergent enriched dysregulated pathways were cell junction, oxidative stress, hypoxia, and metabolic impairments in both PWB_iPSC_3921 and their differentiated ECs as compared to PWB_iPSC_4221, Ctl_iPSC_52521, and their derived ECs, respectively.

**Conclusion:** PWB iPSCs and their derived ECs render novel and clinically relevant disease models by retaining pathological phenotypes. The distinct laser-resistant and sensitive phenotypes in PWB iPSCs and ECs reflect differences among patient subpopulations in response to laser treatments. Our study will provide the targeting pathways for the sensitization of laser-resistant lesions, which will be the focus of our future studies.

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**Abstract N°: 4317**

# Treatment of infantile hemangiomas with Atenolol :About two cases

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**Introduction & Objectives**: Propranolol is the first-line treatment for complicated infantile hemangioma (IH). We report the use of Atenolol as an alternative to Propranolol.

**Materials & Methods:** Case 1: Female child, 11 months old, asthmatic. She presented with a large cervical IH. Corticosteroid therapy at 0.5 mg/kg/d resulted in clinical improvement but with signs of cortisone impregnation. Atenolol at a dose of 1mg/kg/d allowed the discontinuation of corticosteroid therapy, with a clear regression of the IH. Case 2: Female child, aged 13 months. She presented with an ulcerated IH of the left cheek. Propranolol showed good improvement, but was discontinued due to the onset of asthma. Atenolol at a dose of 1mg/kg/day reduced the size of the IH, with good tolerability, especially in terms of respiratory function.

**Results:** Several studies have demonstrated the efficacy and safety of Atenolol in the management of IH. Indeed, it appears to offer comparable efficacy to Propranolol with fewer side effects. As a selective beta-blocker, it can be employed in cases of obstructive bronchial disease, where the use of Propranolol is contraindicated (asthma in our two patients). Its method of administration and adapted doses contribute to better compliance and adherence to treatment.

**Conclusion:** Atenolol is a good alternative for the treatment of IH when Propranolol is contraindicated.

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**Abstract N°: 4332**

**Venous malformation: presenting  a diagnostic problem**

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**Introduction & Objectives:** Vascular malformations, congenital anomalies in blood vessel development, encompass high-flow and low-flow categories. Among these, venous malformations (VM) are the most common, affecting the skin, subcutaneous tissue, muscles, and occasionally bones, tendons, or joints. We report a challenging case of venous malformation, highlighting the complexities encountered in its diagnosis.

**Materials & Methods:** A 10 year old boy with a history of neonatal infection associated with moderate intra-cerebral hemorrhage and a family history of vascular malformations 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 formulaire

On clinical examination, there was a subcutaneous mass approximately 7.5 cm long, soft, depressible, non-inflammatory, painless and non-pulsatile, located on the right shoulder, with a genu-valgum. Laboratory findings indicated anemia (10.8 g/dl) and a D-dimer level of 5703 ng/ml (n <500). Thrombophilia tests were normal. X-rays of the right shoulder showed multiple round calcifications, while ultrasound of the soft tissue suggested phleboliths. Soft tissue echo-doppler suggested a vascular formation, possibly related to a cystic hemangioma or hemo-lymphangioma. The CT scan suggested a large cystic lymphangioma of the right shoulder. Soft tissue MRI revealed a deep intramuscular soft tissue formation in the right shoulder, suggestive of a hemangioma. A second soft-tissue Doppler ultrasound showed a venous malformation of the right shoulder, mainly involving the deltoid muscle. A review of the MRI revealed a deep venous malformation of the right shoulder, mainly involving the deltoid muscle. Appropriate elastic restraint was introduced.

**Results:** VM are the most common congenital vascular malformations. There is a rare autosomal dominant familial form linked to a mutation in TIE2. Our patient was not tested for this mutation.

They appear from birth and can become symptomatic during adolescence and pregnancy. 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, venous malformations maybe associated with pain.Clinical 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, pathognomonic of VM.The most frequent abnormality in CIL is an elevated D-dimer level.Imaging plays an important role not only in confirming the diagnosis, but also in assessing the extent of VM. Ultrasound can reveal phleboliths 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.

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**Abstract N°: 4341**

# Propranolol as a Compounded Preparation in Managing Infantile Hemangiomas

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**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 single-center, 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**.**

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**Abstract N°: 4363**

# Angiolymphoid hyperplasia with eosinophilia presenting as a solitary painful nodule mimicking cutaneous abscess

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**(Note: This is a report of a rare case; thus, subsections of methods and results were not available.)**

**Introduction & Objectives:**

Angiolymphoid hyperplasia with eosinophilia (ALHE), or epithelioid hemangioma, is a rare, benign vasoproliferative disorder of unknown etiology. It is characterized by solitary or multiple, pink-red/brown papules or nodules on the head and neck region. ALHE can mimic other vascular, lymphoid proliferations or infections involving the skin and it is usually persistent. Pain is not one of the leading symptoms. Herein, we report a case of ALHE in a patient presenting with an extremely painful solitary nodule located in the preauricular area which was initially misdiagnosed as an abscess and attempted to be treated with various antibiotherapy regimens. Successful treatment was achieved through excision.

**Case report:**

A 41-year-old female, otherwise healthy patient presented with complaints of painful nodule in left preauricular area for the past 6 months. The patient stated that she previously used multiple courses of antibiotics, with partial improvement. Dermatological examination showed a solitary, reddish-brown colored inflammatory nodule. The lesion was extremely tender during palpation. There was no regional lymphadenopathy. Laboratory parameters were normal. Histopathological examination confirmed the diagnosis of ALHE with the presence of thick-walled vascular proliferation lined by vacuolated large endothelial cells and inflammatory infiltration of lymphocytes, plasma cells and eosinophils in the dermis. Despite the absence of growth in the swab culture, tissue culture specimen yielded growth of *S. epidermidis*, which may explain the partial response observed to previous antibiotic therapies. The patient was first treated with trimethoprim-sulfamethoxazole, which was designated as a sensitive agent, and then the lesion was successfully excised with clean margins. No relapse was observed during the follow-up period.

**Conclusion:**

ALHE is an uncommon condition. It can also manifest as a solitary nodule and when accompanied by symptoms such as pain and tenderness or secondary infections, as observed in our case, the clinical diagnosis can be challenging. The diagnosis is made by histopathology. Surgical excision is the treatment of choice and when completely excised it rarely recurs. This report emphasizes the need for a high clinical suspicion of this rare disorder, which potentially greatly impairs the quality of life of affected individuals.

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**Abstract N°: 4969**

# Propylthiouracil-induced antineutrophil cytoplasmic antibody-associated vasculitis in a patient with hyperthyroidism: a case report

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**Introduction & Objectives:**

Propylthiouracil (PTU), a drug of choice in the treatment of hyperthyroidism, has as an adverse reaction the development of antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV). The latter may affect the skin only or also involve internal organs (kidneys and lungs). We report a patient with PTU-induced ANCA-vasculitis, who took PTU as a treatment for hyperthyroidism.

**Materials & Methods:**

A 63-year-old female presented to our clinic on the occasion of a 5-day appearance of well-defined brownish-red and purple hemorrhagic macules on the arms, as well as three lesions on the left leg. In the center of some lesions, hemorrhagic blisters were formed. In the next days, the margins of the lesions grew and the patient reported feeling mild pain and tension. Seven days before the appearance of the rash she reported having fever and angina, and was treated with paracetamol 500 mg three times daily for 5 consecutive days. On the day of the admission, she tested negative for COVID-19 infection. The concomitant conditions of the patient included arterial hypertension treated with methylprolol succinat 50 mg daily, hyperthyroidism treated with propylthiouracil 50 mg two tablets two times per day (she had the treatment for two years), diabetes type two treated with metformin hydrochloride 500 mg, two tablets per day. She reports no environmental, professional, or lifestyle risk factors or any family history.

**Results:**

The laboratory results from the whole blood count showed elevated thrombocytes: 501.0 10^9/l [range: 140-400], leukocytosis: 13.55 10^9/l [range: 3.5-10.5], and from the differential blood count neutrophil granulocytosis 72.3% [range: 42-70], lymphopenia 19.3% [range: 22-48]; elevated ESR (Westergreen) 38 mm/h [range: 2-30]; elevated glucose serum levels: 7.45 mmol/L [range: 2.8-6.1]; elevated C-reactive protein: 58.4 mg/L [range: 0-10]. The immunology results showed deviation in ANCA-MPO (IgG, M, A) – ELISA: 91.1 AU/ml [range 0-20] and ANCA-PR3 (IgG, M, A) – ELISA: 10.4 U/mL [range: 0-5].

Skin biopsy taken out of the rim of the lesions was compatible with leucocytoclastic vasculitis/necrotizing vasculitis with the involvement of small and medium-sized vessels. No direct immunofluorescence was done. A clinical diagnosis of PTU-induced AAV was made.

The patient was treated with intravenous dexamethasone, 12 mg for the initial 6 days and 16 mg for the consecutive 7 days, esomeprazole 40 mg once daily for 6 days, and local clobetasol propionate cream. During this period, the lesions started to regress but did not resolve completely. She was consulted with an endocrinologist, who advised the total removal of the thyroid gland and the cessation of the propylthiouracil intake.

**Conclusion:**

The physicians should be aware of the possibility for the development of ANCA-associated vasculitis as an adverse effect of propylthiouracil treatment for hyperthyroidism.

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**Abstract N°: 5252**

# A case of ulcerative cutaneous polyarteritis nodosa

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**Introduction & Objectives:**

Cutaneous polyarteritis nodosa (CPAN) is a rare form of cutaneous vasculitis involving small and medium sized arteries of the dermis and subcutaneous tissue with recurrent chronic benign course and without systemic involvement. Although the cause is often unknown, it may be associated with medications, infections, mostly streptococcal or underlying disease.

**Materials & Methods:**

We present the case of a 62-year-old female who reported the appearance of erythema and ulcerations on lower legs associated with myalgias. Fever and other systemic manifestations such as weigth loss, hematuria, abdominal or chest pain, shortness of breath, bloody stools and paresthesias were abscent. The use of new drugs or supplements was denied. Her past medical history included hypertension, asthma and diabetes mellitus.

**Results:**

On clinical examination, irregularly shaped ulcerations covered with eschar, fibrin deposits and necrotic crusts were present predominantly in perimalleolar region as well as on the pretibial aspect of lower legs and feet associated with erythema and edema. The diagnosis was made based on the clinical presentation and histopathological findings which revealed characteristic pathologic feature of leukocytoclastic vasculitis in the small to medium-sized arterioles of the dermis. Complete blood count showed leukocytosis and neutrophilia, increased sedimentation rate and CRP, while complete biochemical panel, urinalysis and FOB tests were normal. Serological and immunologic tests (Hepatitis B and C, HIV; antinuclear antibodies, ANCA, cryoglobulins), throat and urine culture were also negative. Although she had no medical history of streptococcal infection, antistreptolysin O titer was elevated 450 IU/ml. Malignancy was excluded based on tumor marker tests, serum and urine protein electrophoresis, serum immunofixation electrophoresis, chest X ray, abdominal and soft tissue ultrasound which were within normal limits. She was initially treated with prednisone (1 mg/kg/day) with a tapering course and intensive local debridman. After two weeks, the patient showed partial remission. The further course was complicated by cellulitis of the right lower leg, therefore the treatment was continued with systemic antibiotic therapy. Complete and rapid control of disease activity was established after adding azathioprine (100mg/day) in therapy. At this moment, the patient is treated with reduced dose of 50 mg/day. Combined prednisone/azathioprine threatment led to marked and quick improvement in 3 months with remaining atrophic scars. Now, she is periodically monitored at our department and shows no signs of new skin lesions or any other systemic symptoms.

**Conclusion:**

Cutaneous PAN is considered a rare disease. The diagnosis is based on clinical presentation, chronic disease course and characteristic histopathological features. Early diagnosis and treatment are necessary to prevent serious complications. We present satisfactory results after combined treatment in our patient.

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**Abstract N°: 6049**

# Urticaria Vasculitis with atypical presentation

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**Introduction & Objectives:**

Urticarial vasculitis is a rare and underdiagnosed, clinical-anatomopathological disease characterized by the presence of persistent urticarial lesions with histopathological findings of leukocytoclastic vasculitis. Clinically, it manifests as erythematous and indurated wheals, with or without angioedema, located on the trunk and the proximal region of the extremities. It is distinguished from chronic urticaria because each lesion persists for more than 24 hours, is associated with burning, pain or itching, and frequently resolves with pigmentation. Cases that present with systemic symptoms and hypocomplementemia are frequently associated with underlying autoimmune disease.

**Materials & Methods:**

We present the case of a 13-year-old female patient diagnosed with urticaria vasculitis.

**Results:**

13-year-old female patient, with no pathological history, consulted for asymptomatic dermatosis of one week's duration, located on the roots of the limbs, associated with intermittent arthralgias. She reported an episode 7 months prior with a duration of 3 months and spontaneous resolution.

Physical examination: erythematous urticarial plaques with petechiae on their surface. Negative vitropressure. Located at the root of the lower limbs and anterior surface of the left arm.

Pathological anatomy: Perivascular mixed inflammatory infiltrate in vascular walls with erythrocyte extravasation in the superficial dermis. Compatible with urticarial vasculitis.

Complementary clinical examinations: no particularities. Normal complementemia.

Treatment: Oral corticosteroids and antihistamines.

Evolution: Complete resolution of lesions after 15 days of treatment without subsequent regrowth.

**Conclusion:**

Urticarial vasculitis is an underdiagnosed disease. It presents with exclusive skin involvement or associated with systemic diseases, mainly in hypocomplementemic forms. Evolution in outbreaks. Complex treatment in some cases, depending on the severity of the symptoms and associated systemic disease, the first therapeutic option being antihistamines and oral corticosteroids. We present a pediatric case with associated systemic symptoms and normal complementemia. We highlight the importance of studying a possible underlying autoimmune disease and thus making a timely diagnosis and treatment.

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**Abstract N°: 6299**

# efficacy and safety of mammalian target of rapamycin inhibitors in pediatric vascular anomalies: a single center experience.

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**Introduction & Objectives:**

Vascular anomalies represent a large and diversified group of disorders. the International Society for the Study of Vascular Anomalies distinguishes clearly two groups : vascular tumors (characterized by vascular cell hyperplasia ) and vascular malformations (due to defective embryologic vasculogenesis).

This classification is constantly updated due to the identification of mutations in important signaling pathways, including the PI3K( phosphoinositide 3-kinase)/AKT (protein kinase B)/mTOR(mammalian target of rapamycin) .

These finding have led to the evaluation of tailored strategies with preexisting cancer drugs that interfere with these signaling pathways: especially the mTOR pathway such as sirolimus.

In this study we aimed to assess the efficacy, safety and tolerance of mTOR inhibitors in children with slow flow vascular malformations and complicated vascular tumors in Moroccan population.

**Materials & Methods:**

In this retrospective study, data were obtained from patients who were treated with Rapamycin at the pediatric dermatology department between May 2017 and September 2023.

The indications for treatment included 4 tufted angioma and one case of kaposiform hemangioendothelioma complicated by Kasabach-Merritt and regarding vascular malformation, there were 12 cases of microcystic lymphangioma of the head and neck and 3 cases of Klippel Trenauny syndrom that failed sclerotherapy and surgery.

All of our patients had clinical symptoms : CIVD in patients with Kasabach Merritt phenomenon, and local complications in patients with low-flow vascular malformations that included pain, functional impairment, thrombophlebitis, bleeding, infection, ulceration and leakage.

In total, 17 patients were treated with low dose sirolimus and 3 patients everolimus. Initially, all children received the sirolimus at a dose of 0.1 mg/kg/d orally divided in two daily doses.

**Results:**

A total of 20 patients were available for evaluation ( 8 girls and 12 boys), In all patients, rapamycin led to significant volume regression in the malformation group within a few weeks, and significant positive effects on pain, bleeding, leakage, functional impairment and quality of life.

The mean duration of sirolimus therapy was 8,4 months ( 3-18 months). Six patients are still on medication. For 13 patients, treatment was discontinued but had to be restarted in 2 cases due to a relapse, Sirolimus was stopped gradually in all these cases.

In terms of safety of mTOR inhibitors, 1 patient experienced an opportunist infection, 2 patients experienced diarrhea. Regarding biological side-effects, there were no increased lipid or liver enzyme activity.

**Conclusion:**

The study shows that sirolimus has significantly decreased the volume of both vascular tumors and low flow malformations (especially in lymphatic malformations), the treatment has also improved symptoms such as : pain, bleeding, oozing, feeding difficulties f, decreased function of limb.

Regarding the Kasabach-Merritt phenomenon which involved vital distress, all our patients normalized their blood count parameters and reduce the tumor’s volume.

Vascular tumors are proliferative, and malformations enlarge through expansion of a developmental anomaly.

Up to now, treatment options have been limited, Rapamycin is considered an ideal therapy for this type of patients who experience sever chronic pain, and poor quality of life, for its capacity to target cellular pathways including PI3K/AKT /mTOR.

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**Abstract N°: 6327**

# A Case of Macrocephaly-capillary Malformation with Phakomatosis Pigmentovascularis

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**Introduction & Objectives:**

Macrocephaly-capillary malformation(M-CM) and phakomatosis pigmentovascularis (PPV) are collectively referred to as port-wine stains related syndromes. The M-CM is characterized with megalencephaly, capillary malformation of skin with or without distal limb anomalies. PPV is common in port-wine stain related syndromes and it is presented with capillary malformation and extensive dermal melanosis.

**Materials & Methods:**

A 3-year-old female presented with head overgrowth, multiple telangiectasia of head, neck, trunk, arms and legs, blue and grey pigmentation of trunk and arms. She had an open anterior fontanelle and right-side dominant facial asymmetry as well as mild truncal asymmetry with right-sided hypertrophy. While she didn't have syndactyly nor polydactyly. She showed developmental language disorder and mental disability. The cerebral magnetic resonance imaging (MRI) showed left transverse sinus narrowed, partial right transverse sinus undisplayed, brain volume loss(especially white matter), extracerebral space widened, both lateral, third ventricles and midbrain aqueduct dilated, small vessels in brain increased, and no significant abnormalities in cerebral great vessels.

**Results:**

Based on the clinical features and neuroradiological findings, she was diagnosed with M-CM and PPV.

**Conclusion:**

The dermatologist advised a photodynamic therapy for the multiple port-wine stain on the skin. The neurosurgeon advised regular follow-up and rehabilitation therapy.

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**Abstract N°: 6335**

# Successful therapy with apremilast in immunodeficient patient with pyoderma gangrenosum

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**Introduction & Objectives:**

Pyoderma gangrenosum is a relatively rare disease whose etiopathogenesis is not fully understood. Immune system dysfunction plays a role. It is characterized by the occurrence of small red papules and nodules that rapidly disintegrate into very painful irregularly shaped ulcerations with necrotic base., infiltrates can also be observed in internal organs (lungs, liver, bones).

The histological picture is not entirely typical. The aim of this case report is to present the course of the disease and the effect of apremilast therapy in a polymorbid immunosuppressed patient.

**Materials & Methods:**

Female, 61 years old, is treated with type 2 diabetes mellitus, severe pulmonary hypertension, corticoid osteoporosis, has proven hypogamaglobulinemia type IgA . Since 2021 abscess deposits with spontaneous perforation and secretion of hemorrhagic-serous fluid have formed on the extremities.

Systemic corticotherapy with prednisone at an initial dose of 60 mg/day in combination with antiulcerative and antibiotic treatment was indicated due to suspicion of pyoderma gangrenosum. In four weeks from the setting of treatment, there was a significant regression of the lesions. When the dose of prednisone was reduced below 30 mg/day, rapid recurrence and formation of disintegrating defects occurred. Due to high doses of corticosteroids, steroid type 2 diabetes mellitus with the need for PAD compensation, the development of Cushing's syndrome in the face, pathological fractures of the Th11, L1, L3-4 vertebrae gradually occurred.

Considering the existing complications and the little effect of the current high-dose corticosteroid treatment, continuation of corticosteroid monotherapy was contraindicated. On 07/2023, apremilast was administered in gradual titration up to 30 mg twice daily in combination with prednisolone 10 mg/day and intensive local treatment using the moist healing method. From 01/2024 monotherapy with apremilast was indicated.

**Results:**

Complete healing of ulcerations and remission occurred on apremilast therapy 04/2024

**Conclusion:**

Based on this case report, the conclusions can be generalized in the levels we describe below.

Pyoderma gangrenosum is associatid with idiopathic intestinal inflammation, haematological diseases (myelodysplastic syndrome, myelofibrosis, paraproteinaemia), rheumatoid arthritis or tumours can be observed in up to 50%. Liver diseases (hepatitis, primary biliary cirrhosis), autoimmune diseases (vasculitis, lupus erythematosus and Sjogren's syndrome) and surgical interventions (around the stoma, surgical scars. Associated diseases and paraneoplasia should always be excluded. Oral or intravenous corticosteroids are most commonly used in therapy. Other immunosuppressive therapies include apremilast, azathioprine, cyclophosphamide,

mycophenolate mofetil, methotrexate and dapsone. In recent years, biological therapy, especially TNF alpha inhibitors (adalimumab, infliximab), has also been used.

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**Abstract N°: 6595**

**A Multilayered Necrotizing Fasciitis Primary Vasculitis-Like**

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**Introduction & Objectives:** Necrotizing fasciitis is a severe, rapidly progressive, and often fatal infection affecting the subcutaneous tissue and deep fascia. It is a rare, mutilating condition that can jeopardize life through septic complications, with a high mortality rate ranging between 30 to 76%. This condition can present clinically confusing manifestations, occasionally mimicking primary vascular diseases. This case details an instance where necrotizing fasciitis simulated a primary vasculitis, emphasizing the diagnostic complexity in such pathologies.

**Materials & Methods: none**

**Results:**

A 64-year-old chronic smoker presented with extensive painful ulcerations evolving over 15 days before hospitalization. Initially, these ulcers were observed on the right thigh and both antero-external and internal aspects of the right leg and ankle, accompanied by a deterioration in general health and the absence of digestive, respiratory, or joint symptoms. Clinical examination revealed ecchymotic purpura in sheets on the right thigh, inner aspect of the right ankle, and infiltrated left leg.

Linear, well-defined ulcers with necrotic bases were noted on the anterior aspect of the right thigh, as well as on the internal and anterior aspects of the right leg. A large ulceration, measuring 8 cm in diameter, with detached edges and a fibrinous hemorrhagic base was also present. Hypoesthesia was observed. The initial diagnosis considered was a primary vasculitis resembling polyarteritis nodosa, given the general health deterioration, purpura, necrosis, ulcers, and sensory disturbances.

Immunological assessments were negative, while arterial ultrasound revealed diffuse atheromatous plaques. Bacteriological examination of a skin fragment identified beta-hemolytic Streptococcus A. Magnetic resonance imaging (MRI) and skin biopsy confirmed the diagnosis of necrotizing fasciitis.

The patient underwent emergency surgical debridement, coupled with intravenous triple antibiotic therapy, followed by transfer to the intensive care unit due to worsening local conditions and electrolyte disturbances. Unfortunately, the patient’s condition deteriorated, leading to death within the context of septicemia.

**Conclusion:**

The staged nature of the ulcerations and the diverse lesion morphology initially perplexed the diagnostic process, prompting an initial suspicion of primary vasculitis. However, this clinical enigma evolved into a distinctive presentation of necrotizing fasciitis, exhibiting features of vasculitis likely stemming from arterial ulcerations. Particularly in the context of chronic smoking and intermittent claudication, along with the discovery of

atheromatous plaques on arterial ultrasound, there is a significant suggestion of a link between cutaneous ulcers and underlying arterial disturbances.

This complexity underscores the importance of a comprehensive symptom analysis and the need for rapid, multidisciplinary management in such scenarios, aiming to optimize therapeutic interventions and enhance clinical outcomes.

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**Abstract N°: 6856**

# Case series of urticarial vasculitis in three females following in vitro fertilization therapy

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**Introduction & Objectives:**

Urticarial vasculitis (UV) is a type of immune mediated vasculitis, of either normocomplementemic or hypocomplementemic type. We present three cases of UV that developed following in vitro fertilization (IVF) treatment, done for assisted reproduction for infertility. Two of the females were of normocomplementemic variety whereas one had low complement levels. The development of vasculitis in IVF is rare, and UV being a diagnosis of exclusion needs thorough clinical and laboratory work-up for management, and further decision on infertility treatment.

**Materials & Methods:**

All three patients presented with generalized painful, erythematous urticarial plaques over the body, associated with joint swelling over multiple small and large joints of hands and feet. They had been on multiple cycles of IVF treatment for primary and secondary infertility. Two were previously diagnosed by treating physician as generalized urticaria as possible adverse drug reaction to treatment but symptoms worsened and remained recalcitrant to treatment. All three had elevated acute phase reactants (ESR, CRP) and one was hypocomplementemic (Low C3, C4). Skin biopsy was done in two, suggestive of small vessel vasculitis and leukocytoclastic changes.

**Results:** Management consisted of completely stopping the assisted reproductive treatment, and start of cyclosporine at 3.5mg/kg body weight. Careful monitoring of blood pressure and renal function was done, along with symptomatic, supportive treatment. The lesions and systemic involvement were well controlled around the 6–8-week mark, and only evidenced by post inflammatory pigmentary changes. At the 12-month follow-up, two of the patients voluntarily opted out of further cycles of IVF therapy, while one had re-started, but with a different regimen, with no development of similar symptoms.

**Conclusion:** There is a possible role of immune dysregulation in infertility treatment, where many endocrine systems get affected. This may put the patient at risk of development of auto-inflammatory symptoms and conditions. Post COVID era, there is a need to be observant of development of vasculitis in different settings, with atypical manifestations. Management consists of correct diagnosis and optimizing immunosuppressive agent of choice.

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**Abstract N°: 6948**

**calciphylaxis: a complication of renal failure not to be missed**

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**Introduction & Objectives:** Calciphylaxis is a cutaneous ischemic infarct caused by occlusion of blood vessels in the subcutaneous fat and dermis. Severe pain and propensity for infections make calciphylaxis highly debilitating with an annual mortality of 40% to 80%. The objective of this case is to recognize the clinical features and risk factors for calciphylaxis

**Materials & Methods:** we report the case of a patient with end-stage renal failure who developed calciphylaxis on the distal phalanx of the right hand.

**Results:** a 74-year-old type 2 diabetic patient on insulin, with end-stage chronic renal failure, attended our dermatological consultation with a spontaneously painful necrotic lesion on the distal phalanx of the fifth finger of the right hand and on the median side of the wrist, which had been established on 15 days previously. The patient showed high uremia (69mg/dL) and hypercalcemia (11 mg/dL) 010. He was receiving vitamin D supplementation, which was interrupted. radiography showed calcifications of the hand's vascular network. Given the clinical and paraclinical features, the diagnosis of calciphylaxis was retained.

Calciphylaxis can be classified as uremic (in patients with ESRD) or nonuremic (in patients with normal renal function or earlier stages of chronic kidney disease).

Patients without ESRD have a better prognosis (1-year mortality, 25 to 45%) than those who have ESRD (1-year mortality, 45 to 80%), probably because of the differences in coexisting conditions and the location of lesions.

**Conclusion:** Calciphylaxis is a complex microvascular calcification disorder typically manifested as painful cutaneous lesions. The occlusion is initiated by vascular wall calcification and is completed by thrombosis. Until recently, treatments have been limited to controlling risk factors and optimizing wound care.

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**Abstract N°: 7030**

# Infliximab in the Treatment of Refractory Livedoid Vasculopathy: A Case Report

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**Introduction & Objectives:**

Livedoid vasculopathy (LV) is a chronic, relapsing skin disease with symptoms of livedo reticularis, leg ulcerations and atrophie blanche. Lesions are generally distributed symmetrically on the dorsum of the feet, ankles and lower extremities. Patients are accompanied by pain and tenderness in the affected areas. Anticoagulants, anti-platelets, fibrinolytics, vasodilators, anti-inflammatory agents, immunosuppressants and supportive agents can be used in the treatment

**Materials & Methods:**

In this case report, a male patient with LV who was resistant to conventional treatment agents and was treated with infliximab will be presented.

**Results:**

An 18-year-old male patient with no known internal disease applied to our clinic with a complaint of painful lesions on his legs. In the patient's history, it was learned that he had colour changes in his legs for the last few years, but this was the first time he complained of rash and pain and he did not receive any treatment. During dermatological examination, palpable purpura and occasionally atrophie blanche scars were observed on bilateral legs. There was no history of trauma, medication use or infection.

The patient was diagnosed with LV based on clinical examination and histopathological evaluation. No systemic involvement was detected in the examinations. MTHFR C677T mutation was detected as positive in the patient's thrombophilia panel.

In the first step, 1 mg/kg systemic steroid treatment was started. While the patient's pain completely resolved, a partial regression was detected in the rashes on his legs. Afterwards, methotrexate, hydroxychloroquine, pentoxifylline, cyclosporine, enoxaparin treatments administered although the patient's complaints continued. Upon there is data in the literature indicating that TNF-α inhibitor agents can be effective in the treatment of LV. It was decided to start the patient with 5 mg/kg infliximab treatment. With infliximab treatment, a significant regression was detected in the patient's lesions and no new lesions appeared. The patient continues to be followed up under infliximab treatment.

**Conclusion:**

Microvascular thrombus, which plays a role in the pathogenesis of LV, causes ischemia, infarction, pain and hyperesthesia. At the same time, endothelial proliferation and perivascular fibrin deposition cause low tissue perfusion and impaired wound healing. Inflammatory cytokines such TNF-α have been reported to regulate the expression of genes that impair the function of endothelial cells and induce PAI-1 production. Therefore, TNF-α may play a role in the emergence of LV.

Recently, several cases of LV have demonstrated that anti-TNF-α agents and Janus-activated kinase (JAK)

inhibitors may have a positive therapeutic response for typically refractory cases.

LV has been reported to be associated with prothrombotic states, various hypercoagulable conditions, including methylenetetrahydrofolate reductase (MTHFR) polymorphism. MTHFR an important enzyme in homocysteine metabolism, causes increased plasma homocysteine level, and high homocysteine level is a well-known risk factor for atherosclerosis.

In the light of the case reports in the literature that indicating the effectiveness of adalimumab and etanercept in LV, we decided to treat our patient with infliximab and contribute to the limited beneficial therapeutic medications list in LV patients.

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**Abstract N°: 7032**

# Cystic lymphangioma of the upper lip in a child: a rare case with dermoscopic examination

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**Introduction & Objectives:**

Cystic lymphangiomas (CLs) are a rare, benign malformation of lymphatic vessels that almost exclusively affect children under two years of age, mostly in the cervical and axial regions. Oral lesions are relatively rare and usually occur on the dorsum of the tongue.

**Materials & Methods:**

Here, we report an unusual occurrence of CL on the upper lip of a 6-year-old child.

**Results:**

A healthy 6-year-old girl with an unremarkable birth and developmental history presented with progressively enlarging asymptomatic swelling of the upper lip with multiple vesicles discharging clear fluid following minor trauma. These lesions had been present since childhood and had enlarged and regressed intermittently without completely resolving. Mucocutaneous examination revealed a diffuse, painless, ill-defined swelling of the central region of the upper lip with clusters of deep-seated clear vesicles located on the mucosal side of the swelling area. The tongue, labial mucosa and floor of the mouth showed no abnormalities. Dermoscopic examination showed multiple clustered reddish lacunae of different sizes. The diagnosis of CL was made. The patient was referred for carbon dioxide laser treatment.

**Conclusion:**

CLs arise from abnormal lymphatic proliferation and lymph sac sequestration. They are typically diagnosed in infancy and early childhood, with few cases reported in adults. CLs are extremely rare in the oral cavity, with the anterior dorsum and lateral border of the tongue being the most common sites. Less frequently involved sites include the palate, buccal mucosa, gingiva, and lips. The diagnosis of CL is usually straightforward based on its clinical appearance and behavior. Dermoscopy can provide additional clues to the diagnosis, especially in unusual sites, as in our case. The most frequent dermoscopic characteristics of CL are lacunae and vessels. Lacunae are well-defined structures that are clustered and have a round to oval shape. Their histopathological correlation is the presence of dilated, thin-walled vessels in the papillary dermis. Their colour depends on their content, with whitish to yellow colouration being due to lymphatic fluid. The pinkish or reddish colour is due to the presence of red blood cells in the dilated lymphatic channels. They may be dark and violaceous, or blue or black if they are partially or totally thrombosed. A two-tone lacuna is named “hypopyon sign”. The white or yellowish colour of lacunae and the ‘hypopyon sign’. The white or yellowish colour of the lacunae and the hypopyon sign are highly characteristic of CL.

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# A puzzling case of Angiolymphoid Hyperplasia with Eosinophilia: diagnosis and management challenges

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**Introduction & Objectives:**

Angio-Lymphoid Hyperplasia with Eosinophilia (ALHE) is a benign, rare, vascular proliferative disease of unknown etiology. It mostly affects middle-aged adults, manifesting as non-specific red-brown pruritic papules or subcutaneous nodules located on the head and neck. The diagnosis is based on the histopathologic features. The treatment is still challenging, numerous options being proposed, with limited efficacy and risk of recurrence.

**Materials & methods:**

We report the case of an adult woman that presented multiple pruritic purple, bleeding papular and nodular lesions on the right retroauricular and laterocervical area.

**Results:**

A 40-year-old female patient with no relevant personal or family history presented to our Department with multiple erythematous-purple, intensely pruritic papules and nodules located on the right retroauricular and laterocervical area. The lesions had appeared 3 months earlier, with progressive growth in number and size. The patient denied previous trauma, insect bite at these locations, and only applied a topical combination of fusidic acid and bethamethasone valerat cream, with no improvement. A thorough physical examination revealed no additional abnormalities. Laboratory workup did not show any abnormalities, including eosinophilia or elevation of immunoglobulin E levels. An excisional biopsy of one of the nodules showed a reactive vascular proliferation consisting of blood vessels lined by plump endothelial cells with a rich lympho-plasmacytic infiltrate with histiocytes and eosinophils. These findings supported the diagnosis of ALHE, excluding other conditions such as Kimura disease, angiosarcoma, cutaneous lymphoma, cavernous hemangioma, or cutaneous metastasis. We started the patient on oral propanolol 40 mg daily, associated with topical corticosteroids and antihistamines. At the time of submission, after 4 weeks of treatment, the lesions had decreased in size and were less pruritic, with good tolerability and no adverse effects. The patient was advised to continue with oral propranolol, knowing that the previous reported cases that were treated successfully required at least three months of continuous treatment.

**Conclusion:**

ALHE is a rare disease but it has characteristic histopathological findings that can help the diagnosis. There are a large variety of proposed treatments but not enough data on most of them. Surgical excision is the treatment of choice in forms with fewer lesions. This was not the case of our patient. Other treatment options are pulsed-dye laser, CO2 laser, cryosurgery, systemic or intralesional corticotherapy, imiquimod, tacrolimus, isotretinoin, interferon alfa2a, anti-IL-5 antibodies, or dupilumab. Oral propranolol might be of use in selected patients. Although it is thought to be a bening disease, it can significantly affect the patient's quality of life. This warrants further research and efforts to find an effective cure and a unified therapeutic approach.

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**Abstract N°: 7369**

**Leriche syndrome revealed by unilateral cutaneous necrotic ulcerations**

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**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.

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**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 all-cause 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.

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**Abstract N°: 7464**

# Urticarial vasculitis with hypereosinophilia revealing celiac disease in a 57-year-old patient

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**Introduction & Objectives:**

Celiac disease is a common chronic enteropathy, linked to an abnormal immune reaction against ingested gluten in genetically predisposed patients. Over the last few years, there have been multiple reports of the association between CD and several cutaneous manifestations that may improve with a gluten-free diet (GFD). We report the case of a patient diagnosed with celiac disease presenting with urticarial vasculitis and blood hypereosinophilia.

**Materials & Methods:**

This is a 57-year-old female patient with a history of tuberculous pleuropulmonary tuberculosis in 2019, treated and declared cured, who presents with fixed, purpuric, and pruritic urticarial lesions located on the trunk, abdomen, back, and thighs, evolving for 1 month and leaving behind sequellae of hyperpigmented macules. The clinical picture was accompanied by digestive symptoms evolving for 1 week, including abdominal pain, nausea, vomiting, intermittent diarrhea, and abdominal bloating.

**Results:**

Investigations including a blood count, enteroscopy, and jejunal biopsy revealed hypereosinophilia at 6000/uL, hypocomplementemia, pleuropulmonary tuberculosis, and chylous ascites without identified cause. A skin biopsy revealed eosinophilic vasculitis, confirming urticarial vasculitis. ANCA search was negative.

Bone marrow aspiration: medullary hypereosinophilia at 22% (normal 2-3%).

Bone marrow biopsy: mature eosinophilic hyperplasia raising the possibility of hypereosinophilic syndrome or eosinophilic leukemia.

Fibroscopy: Erosive antral gastritis, erosive bulbous duodenitis, with histology suggestive of celiac disease: subtotal villous atrophy with cryptic hyperplasia and intraepithelial lymphocytosis estimated at 30%, and positive anti-transglutaminase, anti-gliadin, and anti-endomysium antibodies.

The patient was placed on a gluten-free diet and oral corticosteroid therapy, with good progress.

**Conclusion:**

Certainly! Blood hypereosinophilia is a complex aspect of hematological pathology, revealing its immunological and clinical intricacies. Recognizing it as a potentially revealing sign, especially when symptoms are vague, emphasizes the need for a thorough diagnostic approach, highlighting the various possible etiologies ranging from parasitic causes to tumor-related and drug-related implications.

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# "Not Just a Children's Diagnosis: Uncovering the Adult-Onset Challenges of Henoch-Schönlein Purpura, a case report"

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**Introduction & Objectives:**

Henoch-Schönlein purpura is a small vessel vasculitis predominantly seen in children, where it is known as the most common form of systemic vasculitis. In contrast, adult-onset HSP is rarer and tends to present more severe complications, including a higher risk of significant renal involvement and potential associations with malignancies or other systemic diseases. This paper details a case of HSP with renal in a 64-year-old, illustrating the unique diagnostic challenges and management of adult vasculitis, thus contributing to the broader understanding and awareness of adult HSP

**Materials & Methods:**

A 64-year-old male patient presented with purpuric lesions that initially appeared on his feet and spread to his legs, thighs, and upper limbs over the course of 20 days. He experienced accompanying abdominal pain and inflammatory polyarthralgia in the knees. The dermatological examination revealed symmetrical petechial purpuric lesions, some necrotic, concentrated on the lower extremities, along with hemorrhagic blisters and petechiae on the buccal floor. Lab results indicated elevated CRP at 24.5 mg/L and proteinuria (1.84 g/24h), with normal liver and coagulation profiles. However, his kidney function deteriorated within 2 weeks, and a peak creatinine level of 5.7 mg/dL was observed. Biopsies of the skin and kidneys confirmed a diagnosis of HSP. Treatment with oral prednisolone and cyclophosphamide resulted in significant improvement.Case report

**Results:**

HSP is a small vessel vasculitis rarely reported in adults. It manifests as a clinical tetrad: non-thrombocytopenic palpable purpura, abdominal pain, arthritis, and renal involvement. HSP can pathologically be considered a form of leukocytoclastic vasculitis that may involve not only the skin but other tissues.A 64-year-old male patient was admitted to the dermatology department presenting with purpuric lesions that had appeared days before his admission. The initial manifestation of these lesions was on the feet, but they gradually spread to include the legs, thighs, and upper limbs. Accompanying symptoms included diffuse abdominal pain of a spasmodic nature and inflammatory polyarthralgia affecting the elbows and knees. His medical background was unremarkable, and there were no recent infections or medications reported. Despite affecting children predominantly, genetic or molecular factors may contribute to its prevalence.** Adult-onset HSP has a low incidence and presents with varying symptoms. A retrospective study of 250 adults identified the most common symptoms as purpuric rash (96%), arthritis (61%), gastrointestinal (GI) disease (48%), and renal disease (32%). The skin rash is symmetric and tends to occur in crops with gravity-dependent distribution. Arthritis is transient or migratory, while GI symptoms, present in 48% of cases, include colicky pain or ulcer bleeding. Renal involvement, ranging from microscopic hematuria to nephrotic syndrome, is detected in up to 85% of cases and may lead to renal insufficiency in 30% of adults.** Diagnosis relies on clinical presentation and tissue biopsy revealing leukocytoclastic vasculitis with IgA deposition. Elevated serum IgA occurs in 60% of cases, and urine analysis may indicate hematuria or proteinuria. Other diagnostic markers include inflammatory markers and IGF-binding proteins.** Treatment of mild cases is

often self-limiting, with NSAIDs or colchicine for severe skin lesions. Glucocorticoids help manage GI symptoms. Aggressive therapies like corticosteroids or cyclophosphamide are reserved for severe nephritis. Plasmapheresis and immunoglobulin are used in refractory cases, while dapsone offers antioxidant effects.

**Conclusion:**

HSP is uncommon in adults and may be under-diagnosed. Diagnosis requires high clinical suspicion and confirmation through immunofluorescence for IgA deposits in leukocytoclastic vasculitis.