Unusual Benign Tumor in a Pediatric Patient: A Case report

Mariana Ramirez Posada¹, Cristina Velez², Ana Cristina Ruíz Suárez², GEOVANNA AYALA³

- ¹Stanford University, Dermatology, Palo Alto, United States
- ²Universidad CES, Dermatopathology, Medellin, Colombia
- ³Universidad del Valle, Dermatology, Cali, Colombia

Introduction

Panfolliculoma is a rare benign adnexal tumor characterized by follicular differentiation. Its occurrence in the pediatric population is exceedingly uncommon, making it an important differential diagnosis when evaluating solitary tumors in children.

Case Report:

A 7-year-old girl was brought to the dermatologist by her mother due to a long-standing, asymptomatic papule on her nose. Physical examination revealed a 3 mm pink papule on the nose. Dermoscopy showed superficial scaling and arborizing vessels.

A skin biopsy was performed, revealing a benign adnexal tumor with differentiation towards all components of the hair follicle, particularly the infundibulum, cystic structures, inner root sheath, and follicular stroma. Based on these findings, a diagnosis of panfolliculoma was established.

Discussion:

Panfolliculoma is a benign adnexal tumor with follicular differentiation, first described by Ackerman et al. in 1993. It is extremely rare, with fewer than 40 cases reported in the literature, and its occurrence in childhood is even more uncommon. This case represents the youngest patient reported in the literature. The tumor typically presents as a solitary papule on the head or trunk in adults. A distinguishing feature of this tumor is its differentiation towards all elements of the hair follicle. The cellular components include bright trichohyalin granules, clear outer root sheath cells, matrical cells, cyst formation, and abortive hair shaft formation, with no evidence of cellular atypia. Simple excision is the recommended treatment. The differential diagnosis includes trichoblastoma, other adnexal tumors, and basal cell carcinoma.

Conclusion:

Panfolliculoma is a rare tumor with a high potential for misdiagnosis, both clinically and histologically. It is essential for dermatologists and pathologists to have a comprehensive understanding of its characteristics and potential differential diagnoses.

Epidemiology survey of patients with pediatric psoriasis in the the Japanese Society for Psoriasis Research from 2017 to 2023.

Emi Nishida¹, Akimichi Morita²

- ¹Nagoya City University West Medical Center, Dermatology, Nagoya, Japan
- ²Nagoya City University Graduate School of Medical Sciences, Dermatology, Nagoya, Japan

Introduction & Objectives:

Psoriasis is a chronic and refractory skin disease. And its prevalence steadily increases from the age of 1 to 18 years in a linear fashion. Pediatric psoriasis patients often require treatment from childhood through adolescence and into adulthood. However, long-term treatment may increase the risk of complications and adverse events. On the other hand, the indications for the treatment of pediatric psoriasis are gradually expanding, so we decided to investigate pediatric psoriasis patients newly registered in the Japanese Society for Psoriasis Research (JSPR) epidemiological survey between 2017 and 2023 in order to understand the epidemiology of pediatric psoriasis in Japan.

Materials & Methods:

The Japanese Society for Psoriasis Research (JSPR) has conducted annual epidemiological surveys of patients with psoriasis since 1982. The JSPR partnered with medical institutions throughout Japan and used its own questionnaire to conduct annual surveys and collect data regarding newly diagnosed Psoriasis cases (from April of the previous year to March of the survey year). A total of 131 medical institutions participated in the surveys for the present study, conducted from 2017 to 2023. The survey was designed to acquire information about patient characteristics, lifestyle habits, disease type and severity, family history, past history and comorbidities, exacerbating factors, focal infection, distribution of lesions, and current treatments. Only data from the completed surveys were included. Pediatric psoriasis is defined as psoriasis in those aged 15 years or younger.

Results:

A total of 204 patients (110 men [53.9%] and 94 women [46.1%]) were enrolled from 2017 to 2023. Its prevalence steadily increases from the ages of 0 to 15 years in a linear fashion. The male-to-female ratio for adults in Japan is 2:1, but for the age groups 0-5, 6-10, and 11-15, the ratios are 1.36, 1.00, and 1.261, respectively. In addition, in other considerations, some patients had an atopic disposition (23 cases [11.3%]). A total of 19 patients (9.3%) had a family history of psoriasis.

Conclusion:

From these results, there is no difference in the male-to-female ratio in pediatric psoriasis in Japan unlike in adult cases. The cause is expected to be more related to genetic background. As a consensus is being reached worldwide regarding pediatric psoriasis, we hope that the epidemiological data on pediatric psoriasis in Japan will provide a basis for considering future treatment strategies and management of pediatric psoriasis, and we believe that it is necessary to continue to accumulate data in the future.

Paronychia in Selumetinib-treated patients

Luca Rapparini¹, Ginevra Martelli¹, Alessandra Gelmetti¹, Stephano Cedirian¹, Francesca Pampaloni¹, Ilaria Cecconi², Bianca Maria Piraccini¹, Michela Starace¹, Iria Neri²

¹University of Bologna, Bologna ²IRCCS AOU Bologna, Bologna

Introduction & Objectives:

Neurofibromatosis type 1 (NF1) management, especially in pediatric patients with inoperable plexiform neurofibromas, has been revolutionized by targeted therapies. Selumetinib, a MEK1/2 kinase inhibitor, significantly reduces tumor volume, improving symptoms and quality of life. However, dermatological adverse events may affect adherence. Effective management of side effects is crucial to maintaining therapy continuity and maximizing benefits.

Paronychia, often leading to periungual pyogenic granulomas, can cause pain and functional limitations, impairing daily activities. Therapeutic approaches include clobetasol under occlusion, betamethasone/fusidic acid, and topical beta-blockers. In severe cases, surgery or chemical matricectomy may be needed. If these treatments fail, dose reduction or discontinuation of selumetinib may be required.

Materials & Methods:

This retrospective study included 23 patients, eight of whom developed paronychia after initiating selumetinib. We analyzed clinical and dermoscopic presentations, treatments, outcomes, comorbidities, and other selumetinib-induced toxicities, as well as the time to symptom onset. The severity of paronychia was graded using the MASCC classification.

Results:

The study involved eight male patients (median age: 12 years, range: 9-19), with plexiform neurofibromas in various body areas. Paronychia appeared at an average of 20 weeks after starting selumetinib, exclusively affecting the toes (19 toes across all patients), with the hallux being the most affected. The severity was classified as grade 2 or 3. Three patients also developed an acneiform rash.

Treatment followed a stepwise approach. At the first evaluation (T0), clobetasol under occlusion was administered. After six weeks (T1), 55% of patients had complete resolution and discontinued therapy, while non-responders received second-line treatments, including continued clobetasol, timolol gel, or intralesional triamcinolone acetonide. At week twelve (T3), extending first-line therapy improved response rates, but second-line treatments resulted only in partial improvement. Two patients permanently discontinued selumetinib due to adverse effects, and one required a dose reduction.

Conclusion:

Clobetasol under occlusion proved effective in treating paronychia, achieving complete resolution in 55% of patients at six weeks, with improved outcomes at twelve weeks. Second-line therapies yielded only partial responses. Selumetinib can cause significant dermatological complications, sometimes requiring dose adjustment or discontinuation. Unlike other targeted therapies (e.g., EGFR inhibitors), high-potency steroid therapy under

occlusion alone was insufficient for complete remission. A multidisciplinary approach is essential to balancing the benefits of plexiform neurofibroma treatment with effective management of skin complications, ensuring the best possible quality of life for NF1 patients.

The role of ultrasound in the therapeutic decision and monitoring in infantile hemangiomas.

Lula María Nieto Benito¹, Pablo Fonda¹, Leire Sánchez-Los Arcos¹, Loreto Carrasco-Santos¹

¹Hospital Central de la Defensa Gómez Ulla, Dermatology, Madrid, Spain

Introduction & Objectives:

Infantile hemangiomas (IH) are the most common vascular tumor in childhood. Characteristically, they appear within a few weeks after birth and progress through three phases, with a variable duration and not all cases necessarily go through all, leading to different IH variants. Therefore, depending on the characteristics of the IH (size and location), the patient, and potential associated complications, each case must be individually evaluated. Regardless of the management approach, early assessment is essential, as delayed intervention may limit and hinder treatment response if needed. Ultrasound imaging is a valuable diagnostic tool and it is particularly useful for IH as it helps in differential diagnosis, lesion characterization, and assessing whether treatment, especially with propranolol, may be beneficial.

Materials & Methods:

A 6-month-old girl with no significant medical or obstetric history, born at term with appropriate weight and height for gestational age. She presented with a lesion on the extensor surface of her left arm, absent at birth but progressively appearing and growing from the fifth to sixth week of life. Physical examination revealed a 5 cm red-violet tumour with macroscopically visible vessels and a palpable deep component, suggestive of a mixed IH. Ultrasound imaging showed a well-defined, dermohypodermal, hypoechoic lesion with hyperechoic structures inside and intense Doppler signal. Due to the predominant vascular component, oral propranolol treatment was initiated after discussing options with the parents and considering the therapeutic expectations based on the patient's age and the lesion's evolution. Monthly follow-up showed good treatment tolerance. Three months after achieving maximum clinical regression (without further size reduction) and maintaining a negative Doppler signal, propranolol was discontinued, with no recurrence observed six months post-treatment.

Results:

The use of systemic beta-blockers has revolutionized the treatment of IH. Although effective, the timing of treatment initiation is crucial for achieving an adequate response. The earlier the treatment begins, particularly during the early proliferative phase, the higher the efficacy and the lower the risk of sequelae. Ultrasound is an appropriate diagnostic method for pediatric patients; its use in IH can be highly beneficial, especially in advanced cases, to assess the IH phase and determine the presence or absence of vascularization via color Doppler imaging. The vascular density of IH has been associated with a greater therapeutic response to oral propranolol. Determining the optimal duration for propranolol treatment can also be challenging. In addition to being crucial for treatment monitoring, ultrasound and color Doppler imaging can help determine the optimal time for treatment discontinuation.

Conclusion:

Ultrasound and color Doppler imaging are non-invasive and highly useful tools for objectively supporting the decision to initiate systemic beta-blocker treatment in IH, especially in uncertain cases or late-growing lesions. Furthermore, they are indispensable for treatment monitoring and crucial in optimizing the appropriate timing for drug discontinuation.

The Management of Juvenile Psoriasis at the University Hospital Center

ouissal hormi¹, zerrouki nassiba^{1, 2}, zizi nada^{1, 2}

¹Department of Dermatology Venereology and Allergology, CHU Mohammed VI, Oujda Morocco, Oujda, Morocco

²Laboratory of Epidemiology, Clinical Research, and Public Health, Faculty of Medicine and Pharmacy, Mohammed First University, Oujda, Morocco, Oujda, Morocco

Introduction & Objectives:

Psoriasis is a chronic inflammatory dermatologic condition that affects individuals across all age groups. Pediatric psoriasis, representing one-third of cases, significantly impacts the quality of life of affected children and their families. Despite its clinical importance, pediatric psoriasis differs from adult psoriasis in terms of epidemiology, clinical presentation, and therapeutic options. The management of juvenile psoriasis remains a challenge due to the lack of clear guidelines.

The primary aim of this study was to evaluate the epidemiological, clinical, therapeutic, and evolutionary profile of juvenile psoriasis at the University Hospital Center. We sought to assess the clinical characteristics, triggering factors, treatments used, and long-term outcomes in pediatric patients diagnosed with psoriasis.

Materials & Methods:

This retrospective, descriptive, monocentric study was conducted over a 10-year period (June 2014 to July 2024) at the dermatology department of the University Hospital Center. All children hospitalized with a diagnosis of psoriasis during this period were included.

Results:

The study included 20 children, with a mean age of 8.5 years (±3.2), and a slight male predominance (55%). The most common age group was 6-11 years. Family history of psoriasis was positive in 45% of cases, and 20% of children were overweight. Psychological trauma was identified as the primary trigger for psoriasis in 55% of patients, followed by infections in 25%. The average age of onset was 3.2 years, with an average disease duration of 4.87 years. The majority (75%) of patients had seen a dermatologist before hospitalization, with a mean of 4.9 consultations. The mean CHILD-DLQI score was 13.9, indicating a significant quality of life impairment. Local treatments, including corticosteroids and emollients, were prescribed to all patients. Systemic therapies included acitretin, methotrexate, and secukinumab. Biopsy was performed in 40% of cases. The average hospitalization duration was 16.1 days.

Conclusion:

Juvenile psoriasis is a chronic condition that significantly affects children's quality of life, with psychological trauma as a major trigger. Treatment is complex and individualized, with both local and systemic options available. Further research is needed to improve the management and long-term outcomes of juvenile psoriasis, as current evidence remains limited, particularly in pediatric populations.

Dual-Target Approach to Pediatric Localized Scleroderma: Impact of Calcineurin Inhibition and Proteolytic Enzyme Therapy

Andrey Mun¹

¹Tashkent Pediatric Medical Institute, Dermatovenereology, Tashkent, Uzbekistan

Introduction & Objectives:

Localized scleroderma (LS) in pediatric patients is a chronic autoimmune dermatosis characterized by progressive fibrosis and immune dysregulation. This study aimed to evaluate the immunological and clinical efficacy of a new treatment strategy combining standard therapy with a calcineurin inhibitor (Tacrolimus) and a proteolytic Papain enzyme (Cucumazim).

Materials & Methods:

The study included 68 children (22 boys, 46 girls; mean age 8.74±0.8 years) with active LS treated at the Tashkent Pediatric Medical Institute from 2018 to 2021. Patients were divided into three groups: a comparison group receiving only traditional therapy, main group I receiving therapy according to clinical guidelines, and main group II receiving standard therapy supplemented with Tacrolimus and Cucumazim. Clinical response was assessed using LoScAT, mLoSSI, and LoSDI indices, along with immunological markers (T- and B-cell subsets, cytokines, CICs) before and after therapy.

Results:

Baseline analysis showed marked T-cell immunodeficiency (reduced CD3+, CD4+, CD8+) and elevated activation markers (CD20+, CD25+, CD95+), with significantly increased IL-1 β , IL-2, IL-6, and TNF- α . By day 21, main group II showed faster and stronger clinical improvement: erythema decreased 2.1-fold (vs. 1.3-fold in the comparison group and 1.6-fold in group I); lesion compaction softened 1.4-fold (vs. 1.1-fold in group I); new lesions reduced 3.6-fold (vs. 1.4-fold in group I). IL-2 fell 6.4-fold; TNF- α by 2.5-fold; IL-1 β by 2.2-fold. CICs dropped to 25.6±1.34 units in group II (vs. 44.5±1.87 in group I and 69.9±2.52 baseline). Prognostic modeling indicated recovery in 28–34 days in group II vs. 41–48 days in group I.

Conclusion:

The combination of Tacrolimus and Cucumazim significantly enhanced outcomes in children with localized scleroderma. Acting on both inflammation and fibrosis pathways, it normalized immune markers more effectively than standard therapy alone. These findings support the integration of calcineurin inhibitors and proteolytic enzymes into LS treatment protocols to improve remission rates and shorten recovery time.

More than just a papule- deciphering insidious case of multiorgan xantogranuloma juvenile.

Anna Baran*¹, Julia Kobylińska¹, Marharyta Vezhnavets¹, Magdalena Kamińska², Marcin Płonowski², Maryna Krawczuk-Rybak², Jadwiga Małdyk³, Iwona Flisiak¹

- ¹Medical University of Bialystok, Dermatology and Venereology, Bialystok, Poland
- ²Medical University of Bialystok, Pediatric Oncology and Hematology, Bialystok, Poland
- ³Medical University of Warsaw, Pathomorphology Department, Warsaw, Poland

Introduction & Objectives:

Juvenile xanthogranuloma (JXG) is a rare disease but most common non-Langerhans cell histiocytosis. It affects mostly male children under the first year of life. It usually presents as solitary yellowish nodules located on head and neck or fewer, as multiple disseminated nodules. Extracutaneous inclusions are rarity. Diagnosis is based on clinical and dermoscopic picture, biopsy and differential diagnosis. The aim was to present a case of misleading numerous papules in the course of multifocal JXG.

Materials & Methods:

A 5-year old boy presented to a dermatologist with a five weeks history of numerous, small, yellowish papules and nodules on the whole body. This appointment was preceded by three independent consultations with equal diagnosis of molluscum contagiosum. 10% KOH solution prescribed earlier didn't improve patient's condition. Finally, the last dermatologist challenged prior diagnosis. Dermoscopy showed a setting sun pattern and the child was urgently referred to the Pediatric Hospital with suspicion of JXG.

Results:

Infectious diseases specialist excluded a contagious or infectious basis. First skin biopsy result came with an imprecise picture of histiocytosis. While waiting for the second opinion, a rapid progression continued with mucous membranes and eyes involvement. Laryngologists' fiberscope examination revealed lesions on the soft palate and the middle throat. Ophthalmologist discovered nodules near the external angle on the skin of the lower right eyelid, on the skin of left eye's eyelids and on the conjunctiva of the left eye. All lesions had a morphology similar to ones described on the skin. PET scan showed metabolically active osteolytic lesion in the right pubic bone, in the skin and numerous small non-active lung nodules. Laboratory tests did not reveal any abnormalities. Ultimately histopathological reassessment by another pathomorphologist exposed histiocytic cell infiltration with foamy cytoplasm, single giant cells. Cells were presenting infiltration of CD163, factor XIII, weak S-100, and negative response for CD1a, Langerin, c-kit, ALK-1. Thereupon juvenile xanthogranuloma was uncovered. Considering the disseminated course of the disease, danger of potential further visceral occupancy, chemotherapy with prednisolone and vinblastine were introduced. Unfortunately due to an exacerbation of lesions and electrolyte disturbances during the therapy patient was hospitalized again. He is still under supervision of the Oncology Unit, continuing treatment with conitnuous clinical improvement.

Conclusion: This case shows how misleading and extending the right diagnosis pinkish or yellowish papules in a child might be. At the beginning the diagnosis was believed to be molluscum contagiosum. Probability of occurrence of a poxvirus infectious disease in a child is far higher than a rare form of non-Langerhans histiocytosis the more diffused. Although rare, JXG should be included in the differential diagnosis. Given that JXG has many faces and extracutaneous involvement is possible, patients should be subjected to in-depth investigations.

Systemic involvement is potentially fatal and the course of the disease depends on location of lesions. In the case of multifocal involvement a multidisciplinary approach within chemotherapy, surgical or immunosuppressive treatment is recommended. Individual approach to the patient, with humility and caution are priceless in physician's work.

Single-cell RNA sequencing reveals transcriptomic landscape and potential targets for giant congenital melanocytic nevi

Wei Tan¹, Yeqiang Liu¹, Dan Deng²

¹Shanghai skin disease hospital, Shanghai, China

Introduction & Objectives: The pathogenesis of Giant congenital melanocytic nevi (GCMN) remains poorly understood, presenting significant challenges for its treatment. To date, no treatment guidelines for GCMN have been established. Moreover, single-cell technologies have not been applied in GCMN research, impeding a comprehensive understanding of the disease at the cellular level. The aim is to elaborate on the transcriptomic landscape of cells in the skin microenvironment of GCMN, explore the molecular mechanism of melanocyte pathological changes, and investigate their interactions with other cells.

Materials & Methods: We used a combination of single - cell and bulk RNA sequencing of lesional and non - lesional skin samples from patients with GCMN and healthy skin samples. Moreover, we also conducted western blotting, qRT-PCR, immunohistochemical, and multiplex immunofluorescence to validate our results. Additionally, we isolated primary cells and explored the impact of TBX2 knockdown on the biological functions of melanocytes.

Results: Patients with GCMN have a higher density of melanocytes and enhanced functions in melanin-synthesis-related processes. TBX2 serves as a marker gene and core transcription factor for a melanocyte sub-population that is specifically expressed in GCMN. It is closely associated with melanocyte proliferation and melanin synthesis. Knockdown of TBX2 in primary melanocyte cells can downregulate melanin synthesis. Additionally, abnormal activation of the PTN-PTPRZ1 and IGF1-IGF1R signaling pathways has been detected in GCMN melanocytes.

Conclusion: The study offers new insights into GCMN's molecular mechanisms and potential therapeutic targets.

²Shanghai Children's Medical Center, Shanghai, China

Capillary malformation-arteriovenous malformation syndrome and termography of cutaneous lesions: A multicenter Spanish study.

Francisco Javier Del Boz-González¹, Juan Carlos Hernández Rodríguez², Marina Romero Bravo¹, José Bernabeu Wittel², JOSE ANTONIO LLAMAS¹, JOSE MANUEL DE LA TORRE GARCIA³, Juan Manuel Segura-Palacios⁴, Gloria Garnacho-Saucedo⁵, MARIA TERESA MONSERRAT GARCIA²

Introduction & Objectives:

Capillary malformation–arteriovenous malformation (CM-AVM) syndrome is a rare genetic disorder characterized by the presence of multiple vascular lesions commonly referred to as "capillary malformations" ("CMs"). However, up to one-third of these patients may develop non-quiescent fast-flow vascular malformations (FFVMs) affecting the skin or other organs.

Thermographic devices detect infrared radiation emitted from various surfaces and produce images indicating the temperature of a specific region. Recent technological advancements have made these devices more sophisticated, affordable and widely available through online platforms, making thermography a practical tool in various clinical settings.

Thermography is a well-recognized tool for assessing vascular lesions, including vascular malformations. Our objective was to explore its application specifically in CM-AVM syndrome.

Materials & Methods:

We conducted a retrospective observational study, including pediatric patients (14 years old or younger) diagnosed with CM-AVM syndrome across five Spanish hospitals from 2015 to 2024. Inclusion criteria required patients to have at least three characteristic "CMs" and/or a confirmed mutation in the RASA1 or EPHB4 genes, which are implicated in the pathogenesis of the syndrome. We analyzed their epidemiological, clinical, and imaging data, which encompassed Doppler ultrasound, cranial and spinal magnetic resonance imaging (MRI), and thermography. We used the FLIR ONE Gen3, an infrared camera that connects to a smartphone to capture thermal images.

Results:

A total of 28 pediatric patients were included in the study, comprising 19 boys and 9 girls, with an average age of 7.9 years. Genetic analysis revealed RASA1 mutations (12 patients) and EPHB4 mutations (in 10). Lesions were most frequently observed on the arms and trunk (89%, 25/28), suggesting a predilection for these areas. Imaging studies detected FFVMs in 5 patients (18%, 5/28), with only one case involving the CNS. Telangiectasias, Bier spots, and epistaxis appeared with similar frequency in patients harboring RASA1 and EPHB4 mutations, suggesting no significant phenotypic differences based on genetic variation in our series.

Thermography was performed on 53.5% of the patients (15/28), and at least one hyperthermic lesion was

¹Hospital Regional Universitario de Málaga, Dermatology, Málaga, Spain

²Hospital Universitario Virgen del Rocío, Sevilla, Spain

³Hospital Universitario Virgen Macarena, Sevilla, Spain

⁴Hospital Universitario Costa del Sol, Marbella, Spain

⁵Hospital Universitario Reina Sofía, Córdoba, Spain

observed in 93.3% of these cases (14/15). Generally, smaller lesions (less than 1.5 cm) tend to be isothermal on thermography, while larger lesions, such as "herald patch"-type lesions, are more likely to appear hyperthermic.

The literature describes actual CMs as typically appearing isothermal in thermographic images, while FFVMs are characterized by hyperthermia due to their elevated blood flow. In our study, "CMs" appeared hyperthermic on thermography, which aligns with their nature as quiescent high-flow malformations. Our thermographic findings correlated well with previous ultrasound results, as larger lesions with more vivid vascular coloring often exhibited more prominent Doppler activity, reinforcing the utility of thermography.

Conclusion:

Thermography offers a non-invasive, accessible, fast and easy-to-interpret complementary test that provides immediate and valuable information. Its ability to detect subtle changes in vascular activity can aid in the early diagnosis, monitoring, and management of this syndrome.

Clinical case: Kaposiform hemangioendothelioma and Kassabach Merritt phenomenon

Karen Lema*¹, Jorge Bonifaz-Araujo², Hernán Luna³

¹HOSPITAL ENRIQUE GARCÉS, DERMATOLOGY, QUITO, Ecuador

²Skin Pro, Quito, Ecuador

³Dermoluna, Ibarra, Ecuador

Introduction & Objectives:

Kaposiform haemangioendothelioma is a rare benign vascular neoplasm, with an incidence of around 007/100.000 cases per year

We present the case of a 2 year old female, who presented with erythema in her left foot since birth associated afterwards with swelling, pain and growth of the foot. She was hospitalized for further evaluation. Laboratory, imaging and histopathological tests were done and they confirmed Kaposiform haemangioendothelioma with Kassabach-Merrit phenomenon.

Systemic therapy with corticosteroids was initiated with good results. The patient was transferred to a tertiary level of care hospital where she received vincristine and sirolimus with favorable outcomes.

Materials & Methods:

A 2 years old, female, born in Esmeraldas, at birth, presented erythema on the left foot that later was associated with swelling, foot enlargement and pain.

On physical exam: Dermatosis localized on the left lower extremity that involves the foot and a third of the distal leg, characterized by an erythematous purpuric tumor of 15x8x5 centimeters. The left foot is hard, painful and twice the size of the right foot, with limited range of motion. (figure 1 and 2).

Laboratory tests demonstrated Platelets of 60,000. Imaging: Left foot x-ray: Osteolytic destruction of third, fourth and fifth toes. Ultrasound of the left foot: dilatation of the veins and arteries throughout the left lower extremity with pulsatile flow and fistular pattern. Dilated epifascial vein. Unable to identify an afferent vessel or specific fistula.

Histopathology: Monochromatic cells of fusiform appearance surrounding blood vessels with extravasation of red blood cells forming cellular clusters.

Immunohistochemistry shows endothelial cells with positivity for the vascular endothelial markers CD31 and CD34, which confirmed the diagnosis: kaposiform hemangioendothelioma and Kassabach Merritt phenomenon. (Figure 3).

Systemic therapy with corticosteroids was started due to severe consumption thrombocytopenia that reached values of 14,000, with no signs of active bleeding associated with the underlying pathology up to that time.

Prednisone 2mg/kg was indicated and control tests performed at 72 hours reported platelets of 23,000, however due to the complexity of the pathology the patient was transferred to a tertiary level of care to initiate chemotherapy.

Results:

HEK is a rare disease with a prevalence of 0.07/100 000 cases per year (1), which manifests at birth in approximately 50% of cases and the rest in childhood (2), it is characterized by a benign vascular tumor, with aggressive local behavior and high mortality rates (3) in the presence of the Kassabach Merrit phenomenon.

Currently there are multiple treatments available, however there are no clear established protocols (7), first of all surgical intervention should be considered when it is possible, otherwise systemic therapy should be initiated.

Conclusion:

The diagnosis of kaposiform hemangioendothelioma should be considered in the presence of congenital hemangiomas, consequently periodic controls of these lesions are required in order to assess their evolution and identify early changes that may suggest the presence of HEK and avoid the appearance of FKM.

In case of infantile hemangiomas suggesting the presence of this pathology, complementary examinations should be performed (imaging, laboratory and histopathological), which may support or rule out the clinical suspicion.

Febrile Exanthem: A Diagnostic Dilemma in a Rare Pediatric Case of Incomplete Kawasaki Disease

Hilmi Kilgin*¹, Narmin Zahedi², BURHAN ENGIN¹

¹Cerrahpasa Medical Faculty Hospital, dermatology and venereology, Istanbul, Türkiye

Introduction & Objectives:

Kawasaki Disease (KD), or acute mucocutaneous lymph node syndrome, is a systemic vasculitis primarily affecting children under five. Diagnosis is based on fever lasting ≥5 days and at least four of five criteria: conjunctivitis, cervical lymphadenopathy, oropharyngeal changes, polymorphous rash, and peripheral extremity changes. When fewer than four criteria are met, the condition is classified as incomplete Kawasaki Disease (IKD), posing significant diagnostic challenges. This case highlights the diagnostic dilemma of IKD in a pediatric patient and emphasizes the importance of differentiating it from infectious and non-infectious diseases.

Materials & Methods:

An 8-year-old male with no prior medical history presented with an 8-day history of fever (maximum temperature: 38.5°C), erythematous rash, and bilateral conjunctivitis. Examination revealed scattered erythematous papules on the anterior trunk and maculopapular lesions on the extremities. Oral examination showed cheilitis but no strawberry tongue. Acute scrotal erythema and tenderness were noted, with Doppler ultrasound confirming epididymitis. Laboratory findings included elevated C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), and leukocytosis. Viral panels for Mycoplasma, Parvovirus, Rubella, HIV, Cytomegalovirus (CMV), and Epstein-Barr virus (EBV) were negative. Abdominal ultrasound ruled out gallbladder calculi. Echocardiography was planned to assess coronary artery involvement, and pediatric cardiology was consulted for early initiation of intravenous immunoglobulin therapy.

Results:

The patient exhibited several IKD criteria, including prolonged fever, polymorphous rash, conjunctival injection, cracked lips, and acute epididymitis. Although the patient did not meet the full criteria for KD, the clinical presentation strongly suggested IKD, prompting early pediatric cardiology consultation for coronary artery evaluation.

Conclusion:

Multiple mucocutaneous signs indicative of KD were observed, including erythematous macules and papules that presented as morbilliform, urticarial, pustular, scarlatiniform, or erythema multiforme-like eruptions. Additional findings included ulceration at the Bacillus Calmette-Guérin (BCG) vaccination site, perineal erythema, hand-foot edema, and late-stage periungual desquamation. Oropharyngeal changes consisted of hyperemia, dry fissured lips, and a strawberry tongue. Diagnosing IKD remains challenging, especially in cases with limited clinical features. Viral exanthems should be prioritized in the differential diagnosis of pediatric patients presenting with fever and rash. The clinical overlap of IKD with conditions such as BCGosis, toxin-induced staphylococcal and streptococcal diseases, Mycoplasma-induced rash and mucositis, systemic-onset juvenile idiopathic arthritis, meningococcemia, and multisystem inflammatory syndrome in children (MIS-C) linked to SARS-CoV-2 complicates recognition. Early identification is essential to prevent cardiovascular complications, particularly coronary artery lesions, a major cause of morbidity in KD. This case underscores the importance of maintaining a high index of suspicion for IKD,

²Cerrahpasa Medical Faculty Hospital, dermatology and venereology, İstanbul, Türkiye

even when the full diagnostic criteria are not met. Prompt intravenous immunoglobulin therapy can reduce the risk of severe complications, and long-term cardiovascular monitoring is required.

Assessing Quality of Life in Infants and Toddlers with Dermatological Conditions - Validation of the InToDermQoL Questionnaire-German Version

Juliane Traxler*¹, Neuza da Silva¹, Matthias Augustin¹, Hagen Ott^{2, 3}, Sanna Hoffmann³, Regina Fölster-Holst⁴, Maria Baumeister⁵, Petra Staubach-Renz⁶, Rachel Sommer¹

¹Institute for Health Services Research in Dermatology and Nursing (IVDP), University Medical Center Hamburg-Eppendorf, Hamburg

²MUC-iSPZ Hauner, Munich University Center for Children with Medical and Developmental Complexity, Ludwig-Maximilian-Universität München, Munich, Germany

Introduction & Objectives: Infants and toddlers represent an important population for paediatric dermatoses. As skin diseases not only cause physical discomfort but also affect quality of life (QoL), which can hinder psychosocial and physical development in this age group, addressing QoL in clinical practice is crucial. However, instruments for children below the age of four are currently lacking. To fill this gap, the skin-generic proxy QoL questionnaire *Infants and Toddlers Dermatology Quality of Life*(InToDermQoL) was developed simultaneously in seven European countries. While the German version (InToDermQoL-GV) was pilot-tested, a comprehensive psychometric evaluation is still pending. Hence, the aim of this study was to evaluate its reliability and validity.

Materials & Methods: Parents of children aged 0-4 years with any skin condition attending dermatology or paediatric clinics filled in a pen-and-paper survey including sociodemographic and clinical information, the InToDermQoL-GV, as well as validated generic [PedsQLTM Infant Scales; PedsQLTM 4.0 Generic Core Scales – toddler version] and disease-specific [i.e., Infants' Dermatitis Quality of Life Index (IDQOL)] questionnaires of QoL. The attending physicians provided clinical information. We examined the factor structure, internal consistency, convergent validity with PedsQLTM and IDQOL, and known-groups validity across disease severity levels of the InToDermQoL-GV; two-week test-retest reliability and minimal detectable change (MDC) scores were investigated in 35% of families.

Results: The sample consisted of parents of 148 children [56.1% male; age groups (n): < 1 (44); 1 – 2 (68); 3 – 4 (36)] with various skin conditions (atopic dermatitis 79.7%, hemangioma 8.1%, bacterial infection 3.4%). Exploratory factor analysis indicated a unidimensional structure of the InToDermQoL-GV for children < 1 year, and a two- and three-factor structure for the two older age groups, respectively. The InToDermQoL-GV showed excellent internal consistency (Cronbach's alpha: .904 – .922) and good construct validity with PedsQLTM and IDQOL across age groups. Known-groups validity analyses showed significantly higher InToDermQoL scores among severely affected children compared to children with no or mild burden only among 1-2-year-old children but not the other two age groups, Test-retest reliability was found to be good for infants and children aged 3 - 4 years but not 1 – 2-year-old children (ICC = .430). MDC scores ranged between 8.88 and 15.56 points (Table 1).

Conclusion: The** InToDermQoL-GV showed predominantly good psychometric properties in children ≤ 4 years with any skin disease. Its use allows assessing disease burden holistically in order to tailor pharmacological and non-pharmacological treatment, and it will contribute to a better understanding of the short- and long-term impact of skin diseases on children and their families.

³Pediatric Dermatology and Allergology, Children's Hospital Auf der Bult, Hannover

⁴Dermatology University Hospital Kiel, University Medical Center Schleswig-Holstein, Kiel

⁵Specialist Clinic Gaißach (Rehabilitation clinic for chronic diseases in children, teenagers and adults), Gaißach

⁶Department of Dermatology, Johannes Gutenberg University, Mainz

Table 1. Internal consistency (Cronbach's α , McDonald's ω), 2-week test-retest validity (intraclass correlations, ICC), standard error of measurement (SEm) and minimal detectable change (MDC) of the InToDermQoL-GV per age group.

Age group	n (baseline)	Cronbach's α	McDonald's ယ	n (retest)	ICC	SEm	MDC
< 1 year	44	.912	.921	11	.791	3.77	10.46
1-2 years	68	.904	.902	27	.430	5.61	15.56
3-4 years	36	.922	.922	13	.814	3.21	8.88

Paediatric blaschkitis: a rare case of multilinear acquired blaschko-linear dermatosis in an 11-year-old girl

Marra Aghajani¹, Matthew J Verheyden², Adrian Cachia³

¹The Skin Hospital, Sydney, Australia

²Hunter New England Local Health District, Newcastle, Australia

³Kossard Dermatopathologists,, Sydney, Australia

Title: Paediatric blaschkitis: a rare case of multilinear acquired blaschko-linear dermatosis in an 11-year-old girl

Introduction & Objectives: Blaschkitis is a rare, acquired inflammatory dermatosis following the lines of Blaschko, classically seen in adults. Paediatric cases are infrequently described in the literature and may be confused with more common dermatoses such as lichen striatus or atopic dermatitis. We present a unique case of paediatric blaschkitis characterised by multilinear distribution and spongiotic histology, adding to the evolving spectrum of Blaschko-linear acquired inflammatory skin eruption (BLAISE).

Materials & Methods: An 11-year-old girl presented with a three-month history of a mildly pruritic papular eruption involving the left leg, buttocks, and abdomen. Past medical history included eczema and hayfever, and family history was notable for atopy. Prior treatment with topical corticosteroids and oral antibiotics had no effect. Clinical examination revealed well-demarcated erythematous plaques with overlying scale and haemorrhagic crusting, arranged in broad, Blaschko-linear bands. A skin biopsy was performed for histopathological confirmation.

Results: Histology demonstrated spongiosis, patchy parakeratosis, and mild to moderate perivascular lymphocytic inflammation in the superficial dermis, with occasional eosinophils and sparing of the deeper dermis. No fungal elements were identified. Absence of lichenoid or deep periadenxal inflammation supported the diagnosis of paediatric blaschkitis. Lesions resolved with emollient therapy and have not recurred.

Conclusion: This case represents a rare paediatric presentation of blaschkitis, distinguished from lichen striatus by its broader, multilinear distribution, truncal involvement, and lack of lichenoid histopathology. While historically considered distinct entities, lichen striatus and blaschkitis may exist along a shared spectrum (BLAISE). Our report reinforces the relevance of clinical-histological correlation and contributes to growing recognition of blaschkitis in the paediatric population. Improved awareness of this condition may prevent misdiagnosis and unnecessary treatment.

Zinc deficiency masquerading as infectious dermatitis

Bruny Carolina Llamas Castellanos¹, Juan Sebastian Puentes-Ochoa¹, jairo jimenez¹, yuliney guerrero¹, jeymi natera², MARGUI LIZET GUERRA PEREZ³

- ¹Universidad de Cartagena, Cartagena, Colombia
- ²Universidad Libre de Barranquilla, Barranquilla, Colombia
- ³Fundación Hospital Infantil Napoleón Franco Pareja Casa del Niño, Cartagena, Colombia

Introduction & Objectives:

Zinc is an essential mineral with numerous enzymatic functions, immune regulation, and wound healing. Acquired acrodermatitis enteropathica is often underdiagnosed due to its low prevalence, variable clinical presentation, and laboratory challenges.

Materials & Methods:

A 2-year-old girl with no medical history presented with a one-year history of periorificial and acral lesions, along with significant hair loss. She had been treated with clindamycin and was diagnosed with bullous impetigo, with only partial improvement.

One year later, she was readmitted with anorexia, worsening of the skin lesions and complete hair loss. Examination revealed symmetrical erythematous, scaly, vesiculobullous plaques on periorificial, acral areas, and perianal region, along with paronychia, pachyonychia, and alopecia universalis.

Laboratory tests and a skin biopsy were performed, considering acquired acrodermatitis enteropathica. All laboratory results were normal. Biopsy findings were inconclusive. The zinc levels were reassessed, confirming deficiency and zinc therapy was instituted with significant skin and hair improvement.

Results:

Zinc is an essential mineral, absorbed in the small intestine. Deficiency can be inherited or acquired, and clinically it presents with a triad of diarrhea, alopecia, and periorificial and acral skin lesions. Reduced dietary intake, malabsorption, or increased elimination can contribute to zinc deficiency.

Diagnosis relies on classic dermatologic findings: symmetrical papulosquamous, vesiculobullous lesions on periorificial, anogenital, and acral areas. It is confirmed by laboratory tests such as zinc, however, zinc values may be misleading due to variables like inflammation, stress, trauma, time of day, serum albumin, and the phlebotomy technique. Alkaline phosphatase may aid in diagnosis.

In this case, initial laboratory tests were normal, delaying diagnosis. Due to persistent clinical suspicion, zinc levels were repeated, and therapy was initiated. After supplementation, the patient showed marked clinical improvement within 48 hours.

Conclusion:

Early recognition of zinc deficiency's cutaneous signs is key, as periorificial vesiculobullous lesions may mimic other skin conditions, delaying therapy. We highlight the need for a high index of suspicion, particularly when laboratory results are inconclusive.

A case of post-inflammatory naevus comedonicus following BCG (Bacillus Calmette-Guérin) vaccine

Terri Chiong¹, Valerie Ho¹, Emily Gan¹

¹KK Women's and Children's Hospital, Singapore, Singapore

Introduction & Objectives:

We present a unique case of acquired naevus comedonicus (NC) secondary to Bacillus Calmette-Guerin (BCG) vaccine in a paediatric patient, a complication not previously reported for this widely administered vaccine.

Materials & Methods:

Not applicable

Results:

A 3-year-old Chinese Singaporean girl presented to the Paediatric Dermatology clinic with a one-year history of chronic skin pits on the left gluteus, at the site of her previous BCG vaccine, with features consistent with nevus comedonicus. The BCG vaccine was administered at birth, following which there was swelling and episodic purulent discharge from the vaccine site for 4 months, before it healed to form an atrophic scar. At age 2, her mother noted pits overlying the scar, from which dark material could be expressed. Examination revealed a 2 by 2 cm cluster of dilated follicular ostia of irregular sizes and shapes, filled with dark keratin plugs, on a background of scarring. Based on the clinical presentation, the diagnosis of non-inflammatory naevus comedonicus (NC) secondary to BCG vaccination was made, and tretinoin 0.025% cream was prescribed.

We postulate that our patient's BCG vaccine abscess led to inflammatory destruction of the hair follicles, causing scarring and alteration of follicular architecture, resulting in the development of NC.

NC is a rare benign hamartoma characterised by dilated follicular openings filled with dark keratin plugs. While typically congenital in children, NC can occasionally occur following local tissue injury, as seen in cases following herpes zoster reactivation in adults.

BCG vaccination, commonly administered to prevent miliary tuberculosis (TB) and TB meningoencephalitis, is known to cause local reactions and abscesses, particularly when administered on the gluteus. To our knowledge, this is the first reported case of NC development after BCG vaccination so far. Therefore, this condition may be under-recognized or under-reported.

Conclusion:

This case highlights a rare instance of acquired NC in childhood, and is the first documented case of NC development after BCG vaccination. There needs to be heightened awareness of this potential complication, as it may be overlooked. Treatment may improve cosmetic outcomes and reduce suppurative complications.

Ichthyosis: An Overview of Epidemiological, Clinical, and Therapeutic Characteristics based on 80 Cases at Mohammed VI University Hospital, Marrakech

Oumaima Markouk¹, maryem aboudourib¹, layla bendaoud¹, said amal¹, ouafa hocar¹

¹Chu Mohamed Vi Marrakesh , dermatology, Marrakech

Introduction & Objectives:

Ichthyoses represent a heterogeneous group of monogenic keratinization disorders clinically characterized by the presence of scales on the skin's surface. This study aims to explore the epidemiological, clinical, and therapeutic features of ichthyosis.

Materials & Methods:

This is a retrospective descriptive study **executed** in the pediatric dermatology **consultation** at the diagnostic center of Mohammed VI University Hospital in Marrakech over a 12-year period (from January 2012 to October 2024). Demographic and clinicobiological data were extracted from the consultation register, then entered and analyzed using SPSS software.

Results:

We included 80 patients, comprising 43 girls and 37 boys, with a sex ratio of M/F: 0.86. The average age was 3.9 years, with ages varying from 1 day to 20 years. A family history of consanguinity was found in the parents of 50 patients (62% of cases). The symptoms typically began in the neonatal period in 69% of cases, with 41% presenting as collodion baby. Other forms of initial presentation included erythroderma in 45% of cases, harlequin baby in 10%, and the presence of bullous lesions in 3,8%. The main cutaneous symptoms were the presence of skin flakes in all patients, xerosis was noted in 70% of cases, erythema in 31.3%, palmar-plantar keratoderma (PPK) in 23.8%, and bullous lesions in 6.25% of cases.In terms of localization, involvement of the skin folds was observed in 21.3%, scalp involvement in 50%, extremities in 20%, palmar-plantar areas in 23.8%, and generalized affection in 80% of cases. The main associated symptoms included pruritus in 48 patients (60% of cases), ectropion in 33 patients, and eclabium in 5 patients. The types of ichthyosis identified based on clinical appearance were nonsyndromic ichthyosis in 75 patients (94% of cases), of which 72% were lamellar ichthyosis, 14.6% were congenital ichthyosiform erythroderma, 5.3% were bathing suit ichthyosis, 4% were epidermolytic ichthyosis, and 4% were vulgar ichthyosis. As for syndromic ichthyosis, only 5 cases were recorded, all of which were associated with Netherton syndrome. Regarding complicated forms of ichthyosis, 28 patients presented ocular complications such as conjunctivitis and keratitis, 7 patients had infectious complications, 4 patients had auditory complications and 4 patients had anemic syndrome. From a therapeutic perspective, all patients received symptomatic treatment with emollients and cleansing gels. Keratolytics were used in 15 patients, and retinoids were prescribed for 32 patients (40% of cases) to treat severe cases and moderate forms unresponsive to topical therapies. Regarding analytical data, a significant correlation was noted between early manifestation of the symptoms and the type of ichthyosis: congenital ichthyosiform erythroderma, epidermolytic ichthyosis, and bathing suit ichthyosis all showed neonatal period. Finally, a statistically significant correlation was found between the type of ichthyosis and the presence of complications: lamellar ichthyosis was associated with ocular and auditory complications, whereas congenital ichthyosiform erythroderma and epidermolytic ichthyosis were linked to infectious complications.

Conclusion:

The originality of our study **resides** in the clinical variability of hereditary ichthyosis cases, with a predominance of the lamellar form and a significant presence of parental consanguinity.



Dermatology Consultations from the Paediatric Emergency Department: A Five-Year Retrospective Analysis of Diagnoses and Managements

Nurten Gozgen¹, Nermin Karaosmanoglu¹, Sinem Ayşe Örnek*¹

¹Health Science University Ankara Training and Research Hospital, Dermatology, Ankara, Türkiye

Introduction & Objectives: Paediatric cases constitute 11–33% of all dermatology consultations. However, there are few studies in the literature focusing on patients referred from paediatric emergency departments to dermatology. This study aimed to evaluate the demographic characteristics, presenting symptoms, dermatological diagnoses and managements of patients referred from the paediatric emergency department to the dermatology department.

Materials & Methods: This retrospective study included 486 patients at Ankara Training and Research Hospital between January 1, 2019, and December 31, 2023 (Ethical approval: E-24-112). Patients' demographic features, clinical features, consultation details, paediatrician's and dermatologist's diagnoses (preliminary and final, respectively), and managements were recorded and analysed. Diagnoses were classified into ten categories: dermatitis, drug eruptions, erythematous-squamous dermatoses, hypersensitivity reactions, nail disorders, physical agent-related conditions, skin appendage disorders, skin infections and infestations, vasculitis, and others.

Results: A total of 486 patients [infant (21.6%), preschool-aged (20.6%), school-aged (29.8%), and adolescent (28%); M/F ratio = 1.08; mean age = 95.4 ± 66.6 months] were included. Most were first-time visitors (97.3%). Triage codes were red (46.7%), yellow (48.4%), and green (4.9%). The mean consultation response time was 48.2 minutes; 80.9% of consultations were requested outside working hours. The most common symptoms were itching (61.1%) and redness (44.1%). Preliminary diagnoses were documented in 31.1% of patients, with 58.3% matching the final diagnosis. The most frequent preliminary diagnoses fell under the categories of skin infections and infestations (61.6%), hypersensitivity reactions (14.6%), and vasculitis (7.3%), while the most common final diagnoses fell under skin infections and infestations (53.9%), hypersensitivity reactions (16.5%), and dermatitis (12.2%). The highest accuracy rates of the preliminary diagnoses were for nail disorders (100%), drug eruptions (75%), skin infections and infestations (68.1%), and hypersensitivity reactions (63.6%), while final diagnoses most frequently differed from preliminary diagnoses in erythematous-squamous dermatoses (100%), physical agent-related conditions (100%), dermatitis (94.1%), and skin appendage disorders (85.7%). The most frequently prescribed treatments were topical antibiotics (40.3%), systemic antihistamines (27.2%), and topical steroids (24.7%). Hospitalization was required in 1.9% of patients, while 7.8% underwent further tests and 4.3% had biopsies.

Conclusion: Most paediatric dermatology consultations from the emergency department were non-urgent and managed with outpatient treatments. The low rate of preliminary diagnoses and notable discrepancies in certain conditions highlight the need for improved dermatological training among paediatric emergency physicians to enhance diagnostic accuracy and optimize patient care.

Rare Cases of Malignant Melanoma Arising from Giant Congenital Melanocytic Nevi: Clinical, Genetic, and Pathological Insights

Wei Tan¹, Yegiang Liu¹

¹Shanghai Skin Disease Hospital, Shanghai, China

Introduction & Objectives: Giant congenital melanocytic nevi (GCMN) carry a low but significant risk of malignant transformation into melanoma. We report three unique pediatric cases of GCMN-associated melanoma, characterized by distinct clinical presentations, histopathological features, and genetic profiles, underscoring the multifaceted nature of this condition.

Materials & Methods: A retrospective analysis was performed. Data included clinical assessments, histopathological reviews, and whole-exome sequencing (WES) for genomic profiling.

Results:

Case 1 (4 years): Presented with congenital multiple black macules/patches on the back, progressive dorsal skin nodule, and an enlarged nodule in the right inguinal region. PET-CT showed metastatic disease. Histopathology confirmed melanoma arising from GCMN with lymph node metastasis. WES revealed NRAS mutations.

Case 2 (5 years): Born with congenital black macules with a central nodule and an enlarged nodule in the left inguinal region. Surgical resection and lymph node biopsy diagnosed melanoma arising in GCMN with lymph node metastasis. WES did not find the pathogenic genetic variants associated with melanoma.

Case 3 (6 years): At age 3, the patient presented with an egg-sized lump in right lumbar-sacral part. Postoperative pathology showed atypical hyperplasia of congenital melanocytic nevus cells. Two years later, a lump recurred in the same area. Moreover, the patient had a history of epilepsy and intellectual disability. Histopathology confirmed melanoma arising from GCMN. WES revealed SMARCA2 mutation. Bone scans and enhanced CT both illustrated tumor invasion. Collectively, the final diagnosis was melanoma arising in GCMN with lymph node metastasis plus neurocutaneous melanosis.

Conclusion: These cases exemplify heterogeneous pathways in GCMN malignant transformation. They emphasize the importance of early surveillance for nodular evolution, lymphadenopathy, and neurological symptoms in GCMN management.

Infectious Dermatoses in Schools: What Parents Know and What Needs to Change

Maryam Ghaleb¹, Ouiame eljouari¹, Gallouj Salim¹

¹CHU - Mohammed VI University Hospital Center, Dermatology and Venerology department, tanger

Introduction & Objectives:

Infectious dermatoses represent a significant problem for the school environment, from the point of view of child health and tranquility for the parents. This paper aims at assessing the parents' understanding about the infectious dermatoses, its prevalence, preventive measures, and handling of cases at school level.

Materials & Methods:

This will be a cross-sectional study that will be conducted in the dermatology department at CHU Mohammed VI in Tangier, Morocco, between May 2024 and September 2024. Data collection and analysis were made through a Google Form questionnaire that has been distributed to parents via social media and email.

Results:

A survey of 410 parents revealed that 43.2% have children aged 3-5 years and 26% have children aged 6-10 years. Nearly 70% of these children attend preschool or primary school. In terms of family structure, 42% of parents have two children, indicating a medium size family.

The survey revealed that 92% of parents are aware of infectious dermatoses, with tinea and herpes being the most recognized (98%), followed by varicella (90%), pediculosis (85%) and warts (80%). Approximately 86.2% of parents reported that their children had experienced infectious dermatoses, with chickenpox being the most common (65.8%), followed by pediculosis (53.6%) and impetigo (41%). Other less common conditions include herpes (31%), hand, foot, and mouth disease (27%), warts (13%), tinea (9%), measles (8%), roseola (7.8%), molluscum contagiosum (6.3%), and scabies (3%). 79.5% of parents reported that infectious dermatoses were common in schools.

Regarding sources of information, 40.5% of parents use the Internet, 38.5% consult a doctor, 12.8% refer to books or brochures, and 6.8% rely on the school. 60% of parents have been taught how to treat infectious dermatoses.

Preventive measures include teaching children about hygiene (20.3%), avoiding close contact with sick children (17.7%), notifying parents and restricting personal belongings (16.2%), cleaning school facilities (15.9%), and monitoring children's skin (13.7%).

When their child shows signs of infectious dermatoses, 43.8% of parents consult a doctor, 31.3% inform the school and 12.5% use home remedies or self-medication. 70% of parents teach hygiene daily and 12.4% weekly.

Regarding school-led awareness programs, 45.5% of parents are dissatisfied, 31.9% are partially satisfied, and 22.7% are completely satisfied. Suggested improvements include stricter enforcement of hygiene (32.9%), regular health checks (26.5%), educational programs for children (24.4%), and distribution of brochures at the beginning of the school year (15.9%).

Conclusion:

Awareness and prevention of infectious dermatoses in schools remain a major concern for parents. Increased

collaboration between parents and schools, along with increased awareness through the distribution of brochures and hygiene education for children, seem promising measures to contain the spread of these diseases.

Phakomatosis Pigmentovascular caesioflammeo-marmorata diagnosed immediately after birth

Julia Perches Ferreira¹, Mariana Pinheiro Genari¹, Mariana Cunha Cesar¹, Guilherme Gonçalves Nascimento¹, Tainá Polonio Angeli¹, Cristina Laczynski¹, Lucia Mioko Ito¹, Carlos D'Apparecida Santos Machado Filho¹

¹FMABC - Centro Universitário Faculdade de Medicina do ABC, Santo André - SP, Brazil

Introduction: Phakomatosis pigmentovascularis is a rare, sporadic genetic syndrome characterized by the occurrence of vascular and pigmented nevi with or without extracutaneous manifestations.

Case Report: Female, weight 3070g, Apgar 8/9, gestational age 39 weeks, maternal typing A positive, newborn typing: A negative, rapid HIV and syphilis test non-reactive. Cesarean delivery due to non-reassuring cardiotocography. Newborn at birth had erythematous spots outlined all over the body. There were no complications during birth. After initial care, the newborn was referred to the dermatology outpatient clinic. The dermatological examination revealed erythematous lesions on one side of the body, with vascular spots on the face, upper trunk, arms, legs and feet and dermal melanocytosis on the abdomen and legs. An evaluation by an ophthalmologist denied ocular alterations. Abdominal ultrasound and cranial tomography showed no alterations. The patient evolved uneventfully during hospitalization and was discharged with a diagnosis of Phakomatosis Pigmentovascular of the probable subtype V-Cesiomarmorata. Follow-up was initiated at a specialized dermatology outpatient clinic for periodic monitoring and follow-up with genetic study to definitively confirm the diagnosis of phakomatosis pigmentovascular caesioflammeo-marmorata.

Conclusion: The case presented reveals the early diagnosis of a rare disease with the possibility of some important future changes. Support is offered to the patient and family regarding the investigation of recent complications and the exact diagnostic outcome with the genetic study that will be performed.

Health-Related Quality of Life in Infants and Toddlers with Dermatological Conditions and Their Parents – Validation of the German Version of the Family Dermatology Life Quality Index (F-DLQI)

Juliane Traxler*¹, Neuza da Silva¹, Matthias Augustin¹, Hagen Ott^{2, 3}, Sanna Hoffmann², Regina Fölster-Holst⁴, Maria Baumeister⁵, Petra Staubach-Renz⁶, Rachel Sommer¹

¹Institute for Health Services Research in Dermatology and Nursing (IVDP), University Medical Center Hamburg-Eppendorf, Hamburg

²Pediatric Dermatology and Allergology, Children's Hospital Auf der Bult, Hannover

³MUC-iSPZ Hauner, Munich University Center for Children with Medical and Developmental Complexity, Ludwig-Maximilian-Universität München, Munich, Germany

⁴Dermatology University Hospital Kiel, University Medical Center Schleswig-Holstein, Kiel ⁵Fachklinik Gaißach, Gaißach

Introduction & Objectives: Paediatric dermatoses often pose additional caregiver burden on parents and impair their health-related quality of life (HRQoL). To assess this impact, the Family Dermatology Life Quality Index (F-DLQI), a 10-item generic dermatology-specific questionnaire, was developed. As its German version had not been previously validated and the burden of paediatric dermatoses on the HRQoL of both the affected children and their parents is generally understudied, the aims of this study were twofold: (1) to investigate the psychometric properties of the German version of the F-DLQI and (2) to test the associations between the parents' and their children's HRQoL.

Materials & Methods: Parents of an infant or toddler aged 0-4 years with any skin disease were invited to fill in a pen-and-paper survey, including sociodemographic and clinical information, as well as the German versions of the F-DLQI and of the Infants and Toddlers Dermatology Quality of Life (InToDermQoL) questionnaire. We tested the F-DLQI's structural validity using exploratory factor analysis, internal consistency, known-groups validity across children's disease severity levels, age groups, and sex; two-week test-retest reliability and minimal detectable change (MDC) scores were investigated in 37% of families.

Regression analyses were conducted on parental burden (F-DLQI), sociodemographic and clinical variables to identify variables associated with children's HRQoL as measured with the InToDermQoL.

Results: The sample consisted of parents [90% mothers; mean age = 33.5 ± 5.1 years, range: 21-45] of 126 children with various skin diseases [59% male; mean age = 1.92 ± 1.11 years, range: 0-4]. The German version of the F-DLQI showed no floor or ceiling effects and good internal consistency (Cronbach's alpha: .896) and test-retest reliability (ICC = .867). The MDC score was 9.31 points (Table 1). Regarding construct validity, the German version of the F-DLQI discriminated between disease severity levels but not between children's age groups nor between sexes. Based on exploratory factor analysis, unidimensionality of the instrument was assumed.

The only significant correlates of InToDermQoL were children's current complaints (β = .083, ρ < .001) and F-DLQI (β = .041, ρ < .001), explaining 77% of the total variance.

Conclusion: The** German version of the F-DLQI validly and reliably measures HRQoL in parents of children ≤ 4 years with any skin disease. Convergent validity and responsiveness remain to be evaluated. The generalizability of the findings is somewhat limited as the present sample consisted mostly of mothers. Nevertheless, the tool can provide relevant clinical and scientific insights into parental HRQoL, which is an important but often overlooked

⁶Department of Dermatology, Johannes Gutenberg University, Mainz

target in clinical care and research.

Table 1. Internal consistency (Cronbach's α ; McDonald's ω), 2-week test-retest validity (intraclass correlation, ICC), standard error of measurement (SEm) and minimal detectable change (MDC) of the German version of the F-DLQI.

n (baseline)	Cronbach's α	McDonald's ω	n (retest)	ICC	SEm	MDC
126	.896	.897	46	.867	3.36	9.31

An algorithmic approach towards diagnosing hereditary photodermatoses

Nibedita Patro*¹, Dharshini Sathishkumar², Maitreyee Panda³, Rahul Mahajan⁴

¹Hi-Tech Medical College, Bhubaneswar, India

²Christian Medical College, Vellore, India

³IMS & SUM Hospital, Bhubaneswar, India

Introduction & Objectives: The congenital photosensitivity disorders present as cutaneous signs and symptoms secondary to photosensitivity, extracutaneous manifestations and a predisposition to malignancy. These are secondary to various defects in DNA nucleotide excision repair, double strand break repair and in metabolism of biochemical substances. Diagnosis of these conditions mainly depend on clinical findings as the molecular analysis is not always feasible. This review focuses on forming an algorithmic approach towards clinical diagnosis of the various congenital photosensitivity disorders as early diagnosis is the cornerstone to minimize long term complications.

Materials & Methods: A systematic review of all the related articles collected after a thorough literature search over last 10 years using key words, "congenital AND photosensitivity NOT acquired" and the individual diseases was done. The literature search focused mainly on systematic reviews, meta-analysis, reviews, randomized controlled trials, clinical trials and case series. All the articles describing the clinical features, diagnosis, and management relevant from a dermatologist's perspective were included.

Results: A total of 264 articles were included in the review. An algorithm (Figure 1) for diagnosis of the different congenital photosensitivity disorders based on the various clinical presentations has been proposed. The diagnosis depends mainly on relevant history and clinical presentation. The history of age of onset, course of the disease, family history, history of consanguinity, history suggestive of systemic involvement and exposure to any photosensitizing agent form an important part in diagnosing hereditary photodermatoses. The congenital photosensitive disorders present symptomatically within the first few years of life and roughly around, from birth to first few weeks of life (congenital erythropoietic porphyria(CEP), hepatoerythropoietic porphyria(HEP), during infancy (Xeroderma pigmentosum(XP), Cockayne syndrome(CS), Trichothiodystrophy(TTD), Rothmund–Thomson syndrome(RTS), Bloom's syndrome) and one to five years (Erythropoietic protoporphyria (EPP), Hartnup disease). Other pediatric photodermatoses usually present in the school going age (>5 years). The children presenting during infancy can again be approached according to their skin manifestations: freckle predominant (XP, CS, UVS(S)) or poikiloderma predominant (RTS, BS, Kindler syndrome).

Conclusion: An early suspicion and diagnosis of the different congenital photosensitivity disorders is the cornerstone behind prompt institution of prevention and treatment, and decreasing the associated morbidity. Identification on the basis of clinical findings forms the mainstay of diagnosis in congenital photodermatoses. Genetic analysis for specific mutations and next generation sequencing further confirm various syndromes accurately. Once diagnosed life-long follow up for early detection and management of various complications must be the rule.

⁴Postgraduate Institute of Medical Education and Research, Chandigarh, India

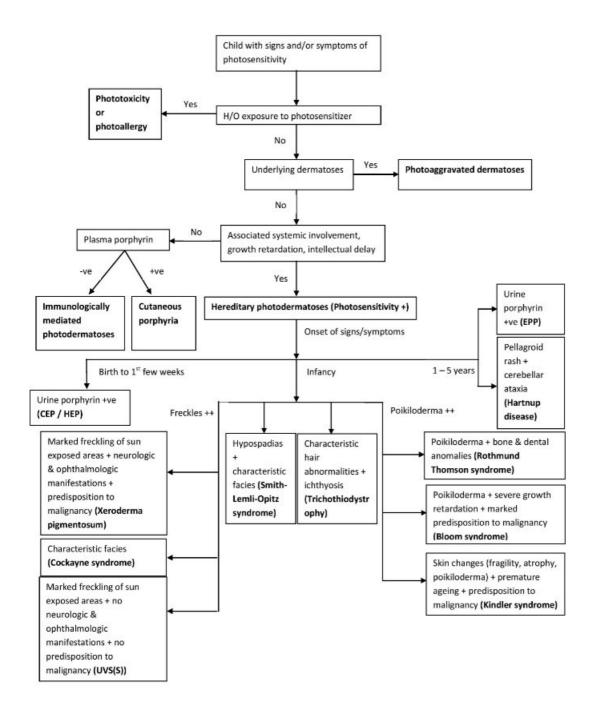


Figure 1. Algorithmic approach to hereditary photodermatoses

Genetically Confirmed Neurofibromatosis Type 1 Presenting with Multiple Juvenile Xanthogranulomas in a Filipino Child: A Case Report

Alyssa Felsophie Silor*¹, Amanda Chung¹, Carmela Augusta Dayrit-Castro¹

¹University of the Philippines-Philippine General Hospital, Department of Dermatology, Manila, Philippines

Introduction:

Neurofibromatosis type 1 (NF-1) is a multisystem genetic disorder with tumor growths, mostly on the skin and nervous system. The presence of juvenile xanthogranuloma (JXG), a benign non-Langerhans cell histiocytosis presenting as reddish-yellow papules or nodules, has been reported in NF-1 but is not part of the NF-1 diagnostic criteria. We present the first documented case of a Filipino child presenting mostly with multiple JXG together with scattered café-au-lait macules.

Case Presentation:

We present a case of a 2-year-old Filipino male born to a nonconsanguineous couple who presented with numerous, generalized, reddish-yellow nodules, histopathologically proven to be xanthogranolumas, together with café-au-lait macules, minimal axillary freckling, and developmental delays in language, communication, and social domains. No Lisch nodules or optic glioma were seen. Genetic testing revealed a pathogenic mutation in the neurofibromin gene clinching the diagnosis of neurofibromatosis type 1. Screening tests for leukemia were done and were within normal limits. Ongoing surveillance is in place to monitor for the development of leukemia.

Discussion:

In young children, the presence of multiple JXG with at least one NF-1 diagnostic criterion like café au lait macules should raise suspicion for NF-1. Genetic confirmation is still recommended to clinch the diagnosis. Since the association of juvenile myelomonocytic leukemia (JMML) in NF-1 patients with JXG has been reported, it may be prudent to screen and monitor patients for this. Conservative management of the cutaneous tumors is recommended as there may be possible spontaneous regression of JXG. NF-1 is commonly associated with developmental delays as in this case, and this requires early intervention. This case highlights the importance of early recognition of NF-1 in children with JXG, the role of genetic testing in confirming the diagnosis for prognostication, and the need for ongoing surveillance by a multidisciplinary team.

When Hypopigmentation Deceives: guttate leukoderma uncovers rare entity

Ahmed Nouh¹

¹Al-Azhar University, Cairo Branch New, Dermatology, Venereology and Andrology, Continuous medical education unit, Cairo, Egypt

Title: When Hypopigmentation Deceives: guttate leukoderma uncovers rare entity.

Introduction & Objectives:

Guttate leukoderma in pediatric patients presents a broad differential diagnosis. While pityriasis versicolor is a common etiology, rare entities may mimic its clinical presentation and delay appropriate management. This presentation highlights the importance of maintaining diagnostic vigilance in persistent or atypical cases of pediatric hypopigmentation.

Case Presentation

An 8-year-old girl was brought to our dermatology clinic with a 12-month history of hypopigmented macules and papules, mainly involving the upper back and lower abdomen. The lesions were initially diagnosed as pityriasis versicolor and treated intermittently with topical antifungals, with no sustained improvement. Upon returning from a prolonged absence, the father noted marked progression, prompting re-evaluation.

Clinical examination revealed mottled hypopigmented small macules coalescing into larger patches, interspersed with ivory-white, flat-topped papules with a smooth surface. The absence of fine scaling and non-responsiveness to antifungal therapy raised suspicion of an alternative diagnosis—specifically lichen sclerosus et atrophicus (LSA) or an atypical dermatosis.

A 3 mm punch biopsy was performed. Histopathology demonstrated numerous clear cells in the lower third of the epidermis, with mild acanthosis, hyperkeratosis, papillomatosis, and a sparse superficial perivascular lymphohistiocytic infiltrate. Special stains (PAS, CEA, CK7, EMA) revealed strong positivity in the basal and suprabasal epidermis, confirming a diagnosis of Clear Cell Papulosis (CCP).

Conclusion:

Clear Cell Papulosis is an extremely rare, benign dermatosis predominantly affecting children under the age of 6. First described by Kuo et al. in 1987, it typically manifests as asymptomatic, hypopigmented to depigmented macules and papules localized to the abdomen (83%), pubic area (34%), and axilla (27%). This case underscores the importance of considering CCP in the differential diagnosis of persistent pediatric leukoderma and highlights the critical role of histopathology and immunohistochemistry in rare dermatologic entities.

Erythematous papules in a febrile newborn: a challenging diagnosis

Inês Abreu^{1, 2}, Gustavo Silva^{1, 2}, Filipe Monteiro^{1, 2}, Sonia Fernandes^{1, 2}, Pedro de Vasconcelos¹, Paulo Filipe^{1, 2, 3}

¹Unidade de Local Saúde Santa Maria, Dermatology, Lisbon, Portugal

Introduction & Objectives:

Hemophagocytic syndrome, or hemophagocytic lymphohistiocytosis (HHL), is characterised by excessive and aggressive immune activation, which can progress to multi-organ failure and death. It predominantly affects children from birth to 18 months of age but can affect adolescents and adults. LHH can be familial or sporadic and can be triggered by various factors: infections, neoplasms, autoimmune diseases (macrophage activation syndrome) and immunodeficiencies. Early diagnosis and timely treatment are essential for a favourable prognosis. However, the rarity of this entity and its varied clinical presentation often make diagnosis difficult.

Materials & Methods:

This is a case report of a one-month-old male infant admitted with persistent fever and skin lesions. Clinical data were collected from medical records, including physical examination, laboratory and imaging studies, dermatological assessment, and skin biopsy.

Results:

A one-month-old male infant was hospitalised for five days due to fever without a focus, which had progressed for a week. Dermatology evaluation was requested due to a dermatosis that had been present since admission. Dermatological examination revealed a monomorphic dermatosis, with erythematous, tumid papules, some annular, distributed over the scalp, frontal region, trunk and upper limbs. The complementary study revealed thrombocytopenia, hyperferritinemia, hypertriglyceridemia, hypofibrinogenemia and an increase in inflammatory parameters (leucocytosis and C-reactive protein). Abdominal ultrasound revealed splenomegaly. These five findings, combined with fever, led to the diagnosis of LHH. Diagnostic hypotheses included a cutaneous manifestation of LHH or neonatal lupus. The skin biopsy was compatible with LHH. Both mother and baby were positive for anti-SSA antibodies, establishing the diagnosis of neonatal lupus and thus the most likely cause of LHH. Cardiac involvement was ruled out. Due to an initial clinical deterioration, the infant was transferred to intensive care and a therapeutic protocol with emapalumab and systemic corticosteroids was instituted. There was a complete resolution of the dermatosis.

Conclusion:

This case highlights the importance of recognising rare cutaneous manifestations of LHH, especially in autoimmune contexts, and reinforces the Dermatologist's assistance in diagnosis. Timely recognition of LHH can be decisive, allowing rapid intervention and improving prognosis.

²Faculdade de Medicina da Universidade de Lisboa, Clínica Universitária de Dermatologia, Lisbon, Portugal

³Gulbenkian Institute for Molecular Medicine, Dermatology, Lisbon, Portugal

Cystic Fibrosis Beyond the Lungs: A Case of Skin Peeling and Respiratory Distress in a Resource-Limited Setting

Menali Gamage*¹, Sriyani Samaraweera¹, Nithya Gunawardena²

¹Lady Ridgeway Hospital for Children, Colombo, Sri Lanka

²Teaching Hospital Kandy, Kandy, Sri Lanka

Introduction & Objectives:

Cystic fibrosis (CF) is a multisystem genetic disorder primarily affecting the respiratory and gastrointestinal systems. While respiratory symptoms are well recognised, extrapulmonary manifestations such as skin involvement are uncommon and often overlooked. In resource-limited settings, the diagnosis and management of CF remain particularly challenging due to limited access to diagnostic tools and specialised care. We report a rare presentation of CF with prominent skin peeling and respiratory distress in a child, highlighting the diagnostic complexity and the need for clinical vigilance in low-resource environments.

Materials & Methods:

A four-month-old infant with a history of recurrent cough presented with respiratory distress and skin peeling for one month. There was no history of diarrhoea or meconium ileus, but she was a product of consanguinity, with a sibling who had died with similar skin and respiratory symptoms. Examination revealed generalised oedema, flaky paint dermatitis, and signs of acute protein-energy malnutrition, without jaundice. There was no history of bowel surgery, and stool microscopy was negative for ova, cysts, or parasites. Serum amylase and bilirubin levels were within normal limits. Abdominal ultrasonography was unremarkable, with no hepatobiliary obstruction or pancreatic abnormalities. Hypoalbuminaemia with fat globules seen in the stool full report led to a clinical diagnosis of cystic fibrosis. However, no obvious lung changes were identified on imaging. The child was started on empiric pancreatic enzyme replacement and nutritional optimisation.

Results:

Cystic fibrosis should be considered as a differential diagnosis in any infant presenting with skin manifestations and recurrent respiratory symptoms. It is possible for chest radiographs to appear normal in infants with CF, particularly in the early stages of the disease.

Conclusion:

Cystic fibrosis, an autosomal recessive multisystem disease caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene, is uncommon in non-Caucasians. Defective chloride channels on the apical membrane of epithelial cells lead to increased viscosity of exocrine secretions in the bronchial, intestinal, pancreatic, and reproductive systems. Classic manifestations in children include meconium ileus, diarrhoea, malnutrition, chronic cough, and recurrent respiratory infections. It remains a clinical diagnosis, supported by sweat chloride testing and genetic confirmation where available.

When the Skin Stays Silent: A Case of Neuro-Sweet Syndrome in a Child Treated with Dapsone

Menali Gamage*¹, Sriyani Samaraweera¹, Jayamini Seneviratne¹, Janani Thavarajah², Amanda Danthanarayana³

¹Lady Ridgeway Hospital for Children, Colombo, Sri Lanka

Introduction & Objectives:

Sweet's syndrome typically presents with characteristic cutaneous lesions alongside systemic symptoms. However, when skin manifestations are absent, as in neuro-Sweet syndrome, diagnosis becomes significantly more challenging, particularly in children. This rare entity can mimic infectious or autoimmune meningoencephalitis, often leading to misdiagnosis and delays in appropriate treatment. We present a paediatric case of neuro-Sweet syndrome with no active cutaneous involvement, successfully managed with Dapsone, highlighting the importance of clinical suspicion and the role of steroid-sparing therapy in long-term management.

Materials & Methods:

A five-year-old child previously diagnosed with Sweet's syndrome presented with severe headache and fever of two weeks' duration while on a tapering dose of steroids. There were no skin lesions suggestive of Sweet's syndrome at the time of presentation. Inflammatory markers were elevated, but cerebrospinal fluid (CSF) analysis revealed aseptic meningitis.

His first presentation occurred at one month of age with high fever and a pustular rash; histology confirmed Sweet's syndrome. His clinical course was complicated by five episodes of febrile aseptic meningoencephalitis, a rare manifestation of neuro-Sweet syndrome, which was unresponsive to antimicrobial treatment but responded to steroids. Over time, he became steroid-dependent, and his growth was significantly impaired. Although his skin remained disease-free during this period, we initiated Dapsone therapy. He subsequently achieved good control of neurological symptoms and was able to successfully reduce his prednisolone dose.

Results:

Extracutaneous manifestations of Sweet's syndrome can mimic infectious causes, particularly in immunocompromised patients, often leading to extensive investigations and delays in appropriate treatment. In children, treatment is especially challenging due to growth failure and immunosuppression associated with steroid therapy. The vulnerability of children to infections limits the use of other immunosuppressive drugs as steroid-sparing agents. Dapsone, with its ability to cross the blood-brain barrier and its anti-inflammatory and neuroprotective effects, including inhibition of myeloperoxidase activity and reduction of oxidative stress, emerges as an effective steroid-sparing option for neutrophilic dermatoses, including neuro-Sweet syndrome.

Conclusion:

Combination therapy with Dapsone and Prednisolone can achieve disease remission while minimizing the side effects associated with long-term steroid use.

²Teaching Hospital Jaffna, Jaffna, Sri Lanka

³National Hospital of Sri Lanka, Colombo, Sri Lanka

Succesful treatment of prurigo nodulare in adolescent age with Dupilumab

Aniko Dozsa^{1, 2}, Erzsebet Szakos^{1, 2}

¹University of Miskolc, Faculty of Health Sciences, Miskolc, Hungary

²Borsod-Abauj-Zemplen County Central Hospital and University Teaching Hospital, , Department of Pediatric Dermatology, , Miskolc, Hungary

Introduction & Objectives:

Prurigo nodulare (PN) is a chronic inflammatory skin disease, that can cause severe pain and itching. Clinically it presents as 1-2 cm-sized pruritic, firm subcutaneous nodules or papules on extremities. The course of the disease is prolonged, with the typical age of onset being over 50 years. Hence, pediatric cases are rarely reported on.

Dupilumab, a fully human VelocImmune-derived monoclonal antibody, blocks the shared receptor component (IL-4R α) for interleukin (IL)—4 and IL-13, thus inhibiting signaling of these central drivers of type 2 inflammation. Worst Itch Numeric Rating Scale (WI-NRS), sleep numering rating scale (SNRS), investigator global assession (IGA) scoring, PN-S scoring were carried out.

Materials & Methods:

Hereby the authors present a case of a 13-year-old girl with PN, successfully treated with dupilumab.

Results: A 13-year-old girl had been suffering from prurigo nodularis for 2 years. The patient presented with dozens of pruritic, firm, brownish nodules on her upper and lower extremities. She had loss of selfconfidence and suffered from persistent pruritus and legpain.

Laboratory examinations showed all paramaters within normal range. X-ray and abdominal ultrasound were negative. She had no inflammation in tonsils. She was also tested for syphilis and HIV, with negative results. Imaging (X-ray and abdominal ultrasound) showed no abnormalities, and tests for HIV, syphilis, and tonsillar inflammation were negative.

IGA score was 3, WI-NRS 6, SNRS 7, PN-S moderate (min. 20 nodules). Topical corticosteroid treatment was not sufficient. She felt as she is shameful, because of her skin. To reduce severe symptoms, dupilumab therapy was initiated. As the patient was only 13- years old and 40kg, first dose was 2x300mg subcutaneous injection, and 300mg every following week, - Given her age (13 years) and weight (40 kg), the initial dose was 2x300 mg subcutaneously, followed by 300 mg every second week.

By week 16, the firm nodules had reduced in size, became soft and pruritus had improved significantly. IGA score was 1, WI-NRS 2. SNRS 3. The lesion count and number decreased lesions, however PN-S was still in moderate range. During the therapy, no new lesions developed. Dose interval was extended to every 4 weeks - Patient got dupilumab on every 4th week, and she is on week 42, The patient has experienced no side effects. We plan to continue her therapy.

Conclusion: : Dupilumab is reported to be efficient in prurigo nodulare in adults. This case shows itmay also be a successful therapeutic option in adolescents. Her moderate PN improved significantly within 16 weeks, with continued regression of symptoms over 42 weeks and no side effects. Dupilumab effectively targets Th2-regulated IL-4/IL-12 pathway, which plays a role in inflammation and pruritus induction.

Maintaining Infant Skin Microbiome Integrity While Enhancing Stratum Corneum Hydration: Effects of an acidic-pH Emollient

Joachim W. Fluhr*^{1, 2}, Michaela Arens-Corell³, Marijane Bevanda-Franjić⁴, Nicole Braun⁴, Lina Praefke³

¹Charité - Universitätsmedizin Berlin, Institute of Allergology, Berlin

²Fraunhofer Institute ITMP, Immunology and Allergology IA, Berlin

³Sebapharma, Boppard, Germany

⁴Eurofins Consumer Product Testing GmbH, Hamburg, Germany

Introduction & Objectives:

The first year of life is critical for the development of a balanced skin microbiome and epidermal maturation. These processes are closely linked to immune regulation and may influence the risk of developing atopic diseases. While emollients are often used to improve skin hydration and maintain skin condition in infants, their effects on the composition and stability of the infant skin microbiome have not been thoroughly investigated. Evidence from adult studies suggests emollients may influence microbial diversity, but data in infants—whose skin physiology and microbial colonization differ significantly are limited. This study examined whether the regular application of a acidic pH baby body milk alters skin microbiome composition and evaluated its effect on stratum corneum (SC) hydration.

Materials & Methods:

Thirty-three infants (mean age 8.0 months; range 5–13 months) were enrolled in an open-label, intra-individual study (conducted by Eurofins, Dermscan Poland). A defined area on the front upper leg was treated with a pH 5.5 body milk twice daily for 28 days, while an adjacent untreated area served as control. Swab samples were collected at D-7, D0, and D28 for microbiome analysis via 16S rRNA gene sequencing. Alpha diversity (Shannon, Chao) and beta diversity (Bray-Curtis PCoA) were used to assess microbial diversity. SC hydration was measured with a capacitance-based Corneometer.

Results:

Microbiome composition remained stable over time and between treated and untreated areas. No significant differences were observed in alpha or beta diversity. The dominant genera included *Staphylococcus* and *Streptococcus*, with consistent relative abundances of *S. epidermidis*, *S. hominis*, and *S. salivarius*. SC hydration in the treated area increased by 27% from baseline and was 24% higher than the untreated area at D28. No adverse events were reported. Parent-reported acceptability was high (92–94%).

Conclusion:

The data show that twice per day application of an acidic-pH emollient does not alter skin microbiome composition or diversity in infants over a 28-day period. The observed microbial stability suggests that the product is microbiologically neutral to evolving skin microbiome of infants, even when used regularly. The significant increase in SC hydration confirms its moisturizing effect. Together, these findings indicate that such emollients can be used to support hydration of infant skin without disrupting the developing microbial ecosystem. This may be of clinical relevance when considering preventive skincare strategies in early life, particularly for infants with sensitive skin or a family history of atopic disease. Longitudinal studies are needed to assess whether such emollient use contributes to reduced incidence of barrier-related conditions, such as atopic dermatitis, during

early childhood.

Clinicodemographic Determinants and Therapeutic Outcomes in Infantile Hemangiomas: A 10-Year Retrospective Cohort Study

Apoorva Sharma¹, manavi gupta¹, rahul mahajan¹, dipankar de¹, sanjeev handa¹

¹Department of Dermatology, Venereology and Leprology, Postgraduate Institute of Medical Education and Research, Sector 12, Chandigarh- 160012, India., Chandigarh, India

Introduction & Objectives: Infantile hemangiomas (IH) are the most common benign vascular tumors in infancy, affecting nearly 5% of newborns. Characterized by a unique growth pattern involving rapid proliferation followed by gradual involution, most IHs are self-limiting. However, a subset can be complicated by ulceration, bleeding, disfigurement, or functional impairment, necessitating medical intervention. The introduction of oral and topical beta-blockers, particularly propranolol and timolol, has significantly transformed the therapeutic landscape, relegating corticosteroids to second-line therapy. Despite this progress, clinical outcomes remain variable and depend on several patient-specific and lesion-specific factors. This study aimed to investigate the clinicodemographic profile and treatment outcomes of patients with IH managed at a tertiary care pediatric clinic over a 10-year period (2016–2025). Specifically, it sought to identify key factors influencing therapeutic response, with particular focus on anatomical site involvement, duration of therapy, and presence of complications.

Materials & Methods: A retrospective cohort design was employed, analyzing medical records of IH patients. Parameters including age at presentation, gender, weight, birth history, anatomical site, syndromic associations, treatment modality, dosage, and duration were documented. Descriptive statistics summarized the clinicodemographic data. Inferential analyses using chi-square tests, multiple linear regression, and logistic regression models identified predictors of treatment response. Multicollinearity was assessed and adjusted using variance inflation factors. Tukey's post-hoc test further explored the role of anatomical site on therapeutic outcomes.

Results: A total of 308 patients were included, with a female-to-male ratio of 1.82:1 and median age at presentation of 7 months. The head and neck region was most frequently involved (51.9%), followed by multi-site (18.2%) and trunk (16.8%) involvement. Only three cases were syndromic. Oral beta-blockers (propranolol or atenolol) were used in 35.4% of cases, while 27.9% received topical timolol. Combination therapies and corticosteroid use were less common. The median treatment duration was 10 months. A complete response was observed in 55.8% of patients, partial response in 41.2%, and no response in 1.9%. Regression analysis revealed that longer treatment duration significantly improved response (p = 0.01), and complicated hemangiomas were associated with significantly poorer outcomes (p < 0.01). Site of involvement also impacted therapeutic success (p = 0.03); lower limb and multi-site IHs showed better response rates, while genital and rare-site hemangiomas fared worse.

Conclusion: This study underscores the importance of therapy duration and lesion location in determining IH treatment outcomes. While beta-blockers remain highly effective, responses vary significantly based on lesion complexity and anatomical distribution. The findings support the need for individualized treatment strategies and highlight the limitations of a one-size-fits-all approach. Future prospective studies with standardized protocols and extended follow-up are essential to validate these observations and refine therapeutic guidelines for IH management.



Early Intervention with Topical Calcineurin Inhibitors in Pediatric Segmental Vitiligo: a Case Series Challenging Prognostic Expectations

Alessia Paganelli¹, Mauro Picardo¹

¹IDI-IRCCS Istituto Dermopatico dell'Immacolata, Rome, Italy

Introduction & Objectives: Vitiligo is a chronic autoimmune skin condition characterized by melanocyte destruction and subsequent depigmentation. Segmental vitiligo (SV), a less common subtype, typically arises early in life and progresses rapidly within the first year before stabilizing. Unlike non-segmental vitiligo (NSV), SV frequently presents with early leukotrichia and poliosis. Treatment of SV is often challenging due to its rapid stabilization and poor response to conventional therapies. Although topical calcineurin inhibitors (tCNIs) are established in NSV, evidence for their use in pediatric SV is limited. The aim of our work was to evaluate the efficacy of tCNIs in pediatric patients with early-stage SV.

Materials & Methods: Four pediatric patients (ages 8–11) with early-onset SV were treated with tCNIs twice daily, either topical tacrolimus 0.1% ointment or pimecrolimus 1% cream. All patients were evaluated clinically and under Wood lamp prior to treatment and after three months. Clinical and familial history, previous treatments and cutaneous findings were recorded. Photographic documentation was obtained at baseline and follow-up.

Results: All the selected patients showed significant clinical improvement after 3 months of therapy, with visible perifollicular repigmentation and lesion reduction. In one patient, leukotrichia also showed partial reversal, despite such rare finding typically being associated with poor therapeutic response. Interestingly, most of the patients had familial history of autoimmune diseases (including vitiligo and other immune-mediated conditions). No adverse effects were reported. The therapeutic response appeared to correlate with early initiation of treatment (<1 year from onset).

Conclusion: Topical calcineurin inhibitors can be an effective and well-tolerated option for treating pediatric SV, especially when initiated early. Our findings suggest that early immune modulation may preserve melanocytes and improve outcomes, even in cases with traditionally poor prognostic indicators such as leukotrichia. However, further studies are warranted to confirm these findings and establish standardized treatment protocols.

Hyphema as an Initial Manifestation of Juvenile Xanthogranuloma: A Rare and Vision-Threatening Presentation in Infancy

Najat Chebbawi¹, Fatima-Zahra Le Fatoiki¹, fouzia hali¹, soumiya chiheb¹

¹1 Rue des Hopitaux, Department of dermatology-venereology, CHU IBN ROCHD, CASABLANCA, Casablanca, Morocco

Introduction & Objectives:

Juvenile xanthogranuloma (JXG) is the most common form of non-Langerhans cell histiocytosis in children. While typically confined to the skin, rare extracutaneous involvement may occur, particularly ocular manifestations, which can lead to serious complications. We aim to highlight the importance of early dermatologic and ophthalmologic evaluation in infants presenting with atypical hyphema.

Materials & Methods:

We report the case of a 6-month-old male infant referred to the ophthalmology department for right-eye viral keratitis with associated spontaneous hyphema and suspicion of an intraocular mass. Dermatologic consultation was requested following the discovery of yellowish papulonodular cutaneous lesions.

Results:

Clinical examination revealed multiple well-demarcated, yellowish papulonodular lesions on the left nasolabial fold, chin, and trunk, present since the age of 2 months. Histopathological analysis showed normoacanthotic epidermis with orthokeratosis and a dermal infiltrate composed of lipid-laden histiocytes, Touton-type multinucleated giant cells, and a lymphoplasmacytic inflammatory background, confirming the diagnosis of JXG.

No visceral involvement was found on abdominal ultrasound. A therapeutic abstention strategy with close dermatologic and ophthalmologic follow-up was adopted.

Ocular involvement in JXG is uncommon but potentially severe, especially in children under two years of age. It is typically characterized by spontaneous hyphema, which may lead to secondary glaucoma if not promptly recognized. Multiple forms of JXG are more often associated with systemic involvement and an increased risk of hematologic malignancies such as juvenile myelomonocytic leukemia.

Given these risks, infants with multiple cutaneous JXG lesions should undergo thorough evaluation, including ophthalmologic screening, and families should be informed about alarming ocular symptoms.

Conclusion:

This case underscores the importance of considering JXG in the differential diagnosis of spontaneous hyphema in infants. Early multidisciplinary management is crucial to prevent irreversible visual complications and to screen for potential systemic associations.

Investigating Papular Lesions on the Face of Pediatric Patients: Following Clues Leading to Differential Diagnoses

Maria Clara Cronemberger Guimaraes Serzedo*¹, Luciana Vilela Gomide¹, Adriana Schikiera Martinelli Salathiel¹, Rafael Fantelli Stelini¹, Andrea Fernandes Eloy da Costa França¹, Renata Ferreira Magalhães¹, Elisa Nunes Secamilli¹

¹UNICAMP Universidade Estadual de Campinas, CAMPINAS

Introduction & Objectives:

Papules on the facial region of pediatric patients can indicate a range of diagnoses across various etiologies. Due to the clinical similarity of these lesions, misdiagnoses are not uncommon, often resulting in delayed implementation of appropriate therapeutic plans. We describe three cases of children presenting with facial papular lesions that were clinically similar but differed in etiology and histopathological findings.

Materials & Methods:

We present three cases of patients with papular lesions located on the face - clinically similar but differing in their etiopathogenic origins.

Results:

The first case involves an 11-year-old boy with micropapules on the forehead and nasal dorsum that progressed to the trunk over three years. Histopathological examination revealed distortion of follicles surrounded by dermis with mucin deposition, consistent with follicular mucinosis. Alopecia could be a diagnostic clue but was not present in this case. Treatment with topical tretinoin 0.025% on alternate days led to more than a 50% reduction in lesions. The second case involves an 8-year-old girl erroneously diagnosed with molluscum contagiosum, presenting multiple hypochromic, solid papules without umbilication, located in the bilateral malar region, for three years. A biopsy of one lesion revealed findings consistent with trichoepithelioma. Watchful waiting approach was adopted, with potential future surgical removal. The third case involves a 5-year-old boy with multiple hyperchromic papules on the bilateral malar region and nasal dorsum, present since birth and showing progressive growth. He also had diffuse hypochromic macules, a shagreen plaque on the forehead, focal epilepsy, mild cognitive impairment, and bilateral subependymal nodules in the lateral ventricles, on magnetic resonance imaging. These findings suggest angiofibromas related to tuberous sclerosis complex. For this condition, some studies indicate that topical sirolimus 1% can be highly effective.

Conclusion:

Thus, the three reported cases underscore the importance of pursuing diagnostic clues to ensure accurate diagnoses and tailored treatments. Although each child presented with facial papular lesions, distinct diagnostic and therapeutic strategies were required. The first case emphasized the significance of histopathological examination in confirming follicular mucinosis. The second case highlighted the need for diagnostic vigilance in the presence of atypical lesions, thereby avoiding misdiagnosis; histopathology ultimately confirmed trichoepithelioma. The third case demonstrated a more complex syndromic presentation consistent with tuberous sclerosis complex, warranting a multidisciplinary management approach. Therefore, these reports reinforce the value of thorough clinical assessment, histopathological confirmation, and individualized care in pediatric dermatology.

Juvenile Tuberous Xanthoma with type IIa Hyperlipedemia

Bhuvnesh G. Shah*¹, keya sheth²

¹Government Medical College, Department of Dermatology, Surat, India

Introduction:

Tuberous Xanthomas are nodular lesion which develop over pressure prone areas such as elbows, knees, achilles tendon and buttocks. There is alteration in lipid metabolism leading to hyperlipidemias.

Here we report a case of Juvenile Tuberous Xanthoma with type IIa hyperlipidemia.

Case Report: A 4year girl came with multiple yellowish waxy lesions over elbows, hands, legs, buttocks and face since3 years. It started on the right hand and progressed gradually. Lesions were bilaterally distributed. Lesions were not tender and were fixed. Parents were asymptomatic.

A complete physical and systemic examination was done. Systemic examination was normal. There was no arcus juveniles, organomegaly, lymphadenopathy.

Complete investigation was done, includes CBC, Liver function test, Renal function test, X-ray, Glucose tolerance test, Ultrasonography, fundus examination.

Results:

Altered lipid profile was noted, with increase in LDL(low density lipoprotein) (476mg/dL) and increase in total cholesterol (694mg/dL). Other investigation were within normal limit. Father also had raised LDL. Electrocardiogram suggested sinus bradycardia. Biopsy from nodule was suggestive of infiltration of dermis with admixture of foam cells, histiocytes and lymphocytes. All these finding differentiated it from other paediatric xanthomas such as Phytosterolemia, Cerebrotendinous xanthomatosis and Alagille syndrome. The diagnosis of Juvenile tuberous xanthoma with Type Ila familial hypercholesterolemia was confirmed. Patient was referred to physician and started with statin. Parents were advised screening for heart disease and lifestyle modification was stressed upon. Manual removal of certain lesion was undertaken in phases.

Conclusion:

Xanthomas are localised lipid deposit occurring due to alteration in lipid metabolism. Tuberous Xanthomas are firm yellow nodules that mostly occur over pressure sites. When it occurs in young generation, a severe form of hyperlipidemia is suspected. There are various clinical types of xanthomas and are associated with various hyperlipediemias.

There is increased risk of atherosclerotic diseases and pancreatitis .The World Health Organisation recommends Fredrickson classification and it is based on the type of lipoprotein present in excess. Familial hypercholesterolemia is characterized by high total cholesterol and high LDL cholesterol along with normal triglycerides.

The clinical significance in present case was to foresee an early development of heart complications like myocardial infarction, coronary artery diseases and pancreatitis which can develop during first 2 decades of life.

²skin care institute, surat, India

Pharmacokinetics and safety of afamelanotide in adolescent and adult erythropoietic protoporphyria (EPP)

anna minder*1

¹Stadtspital Zürich Triemli, Zurich, Switzerland

Pharmacokinetics and safety of afamelanotide in adolescent and adult erythropoietic protoporphyria (EPP)

EADV CUV052 Abstract (oral presentation)

Introduction & Objectives:

Erythropoietic protoporphyria (EPP) is an ultra-rare inherited disorder of haem metabolism, where systemic accumulation of the highly photoreactive haem precursor protoporphyrin IX (PPIX) causes debilitating phototoxicity following brief exposure to visible light (within minutes). Symptoms are present from early childhood, resulting in conditioned behaviour that has a severe impact on patient quality of life.

Afamelanotide is a synthetic analogue of naturally occurring α -melanocyte stimulating hormone (α -MSH) and the only approved treatment for EPP. Afamelanotide binds and activates MC1R on epidermal melanocytes, resulting in epidermal eumelanogenesis which, due to eumelanin filtering and absorbing light, prevents PPIX photoactivation and phototoxicity in EPP. Moreover, eumelanogenesis exerts antioxidant properties within cells, thereby neutralizing the inflammatory effects of free radicals which are generated following PPIX photoactivation.

Currently, afamelanotide is licensed for adults only, leaving a significant unmet need for treatment of paediatric EPP, with only a small number of adolescent patients treated off-label. CUV052 was the first interventional study to assess afamelanotide in the adolescent EPP population (12-17 years).

Materials & Methods:

Eligible participants were patients with EPP aged 12-70 inclusive and a weight greater than 50 kg. Participants received one 16mg afamelanotide implant (SCENESSE®) and had plasma PK samples taken at regular timepoints. The primary objective was to determine and compare the pharmacokinetic (PK) profiles of afamelanotide in adolescents and adult EPP patients. Primary endpoints were area under the plasma-concentration time curve (AUC0-t) and maximum plasma concentration (Cmax); secondary endpoints included area under the curve extrapolated to infinity (AUC0-∞), time at Cmax (tmax) and apparent half-life (t1/2).

Results:

In total, 28 participants (14 adolescents 8F:6M, 13-17 years) and 14 adults (4F:10M, age 18-55 years) consented and were enrolled. PK analyses showed that afamelanotide exposure was higher in EPP adolescents compared to adults, although at levels consistent with historical data from healthy volunteer studies. Afamelanotide was eliminated at a similar rate in adults and adolescents, as evidenced by similar terminal rate constants and plasma concentration half-lives.

Safety analyses showed afamelanotide was well tolerated by all study participants. Of the 28 TEAEs reported in adolescents, only 8/28 (28.6%) were assessed as related to afamelanotide. By comparison, 25 TEAEs were reported in adults, of which 14/25 (56%) were assessed as related. All related treatment-emergent adverse events (TEAEs) were mild in severity and resolved during the study, with no serious related TEAEs reported.

Conclusion:

In conclusion, PK profiles for afamelanotide were higher in adolescents versus adults and consistent with previous data. Afamelanotide was well tolerated in both adolescents and adults. Data from this study adds to the safety and efficacy profile for afamelanotide in the adolescent population.

Character count (excluding title and headings): 2642 without spaces

Isolated Granuloma Annulare in a Pediatric Patient: A Case Report with Clinicopathologic Correlation

Serap Öztürkcan*¹, Hazan Aksoy¹, Göksu Dalgıç¹, Peyker Temiz²

 1 Manisa Celal Bayar University, Dermatology and Venereology , Manisa, Türkiye

Introduction & Objectives:

Granuloma annulare (GA) is a non-infectious, inflammatory, granulomatous skin disorder characterized by necrobiotic granulomas. It presents as smooth, circular, skin-colored or mildly erythematous papules and plaques. The condition is usually benign and self-limited but has been linked to conditions like HIV and malignancies. The most common form is localized GA (LGA), while other variants include generalized GA (GGA), subcutaneous GA, perforating GA, and patch-type GA.

GA affects children, adolescents, and young adults, with the generalized form more common in older adults. The female-to-male ratio is approximately 2:1, and lesions can appear anywhere on the body, often asymptomatically. In some cases, tenderness is noted. This case report presents a pediatric case of localized GA with histologic features of the interstitial variant, emphasizing the significance of clinicopathologic correlation.

Materials & Methods:

A 6-year-old male patient presented with asymptomatic, isolated lesions on the dorsal aspect of the left foot that had appeared three days prior. He reported a similar, self-resolving episode five months earlier. Clinical examination revealed multiple small, mildly erythematous papules, with no pain, pruritus, or infection. No history of drug use or trauma was noted.

A punch biopsy was performed with differential diagnoses of atypical granuloma annulare and piezogenic papules. Histopathologic evaluation aimed to identify focal dermal collagen degeneration, interstitial histiocytic infiltration, and mucin accumulation. The biopsy was examined for histological confirmation of interstitial-type GA.

Results:

Histopathologic findings showed scattered histiocytes between collagen bundles and mucin deposition, with no palisading granuloma structure. These features were consistent with non-infectious granulomatous dermatitis, specifically the interstitial variant of GA. The patient was treated with a short course of topical corticosteroids and emollients. On follow-up, most lesions had resolved, although some post-inflammatory hyperpigmentation persisted. The patient remained asymptomatic, with no further complications.

Conclusion:

Granuloma annulare presents with various clinical and histological forms, including the interstitial variant. This case underscores the importance of clinicopathologic correlation, particularly in pediatric localized GA. Although the etiology and pathogenesis of GA remain unclear, immune mechanisms like Th1-dominant delayed-type hypersensitivity are suggested. GA has been linked to diabetes, infections, and certain medications, especially TNF- α inhibitors.

Most GA cases resolve spontaneously, but treatment may be necessary for persistent or cosmetic concerns. Topical corticosteroids, calcineurin inhibitors, and intralesional steroids are first-line treatments for localized cases.

²Manisa Celal Bayar University, Pathology, Manisa, Türkiye

For generalized or refractory cases, systemic therapies like hydroxychloroquine, methotrexate, and PUVA may be used, though none provide a definitive cure. This case highlights the need for differential diagnosis, particularly in atypical forms, for proper management.

Juvenile Systemic Scleroderma in a 9-Year-Old Child: A Case Report

Ouissal Laadime¹, Bendaoud Layla¹, chatti manel¹, Mariem Aboudourib¹, ouafa hocar¹, Amal Said¹

¹Arrazi mohammed VI university hospital,, dermatology and venereology,, marrakesh, Morocco

Introduction:

Systemic sclerosis (SSc) is a rare, multisystemic autoimmune disorder of unknown etiology, characterized by skin fibrosis and variable visceral involvement. It predominantly affects adults, with a strong female predominance. Diagnosis relies on clinical criteria established by the American College of Rheumatology (ACR) and the presence of specific autoantibodies (AAbs), which play a crucial role in disease classification. Juvenile systemic sclerosis (JSS) is exceedingly rare, with limited cases reported in the literature. We present a case of JSS in a young girl, highlighting its diagnostic and therapeutic challenges.

Materials & Methods:

We present in this case, A 9-year-old girl, born to a first-degree consanguineous marriage, presented with a history of Raynaud's phenomenon during winter. At age 8, she developed bilateral, symmetrical inflammatory polyarthralgia affecting large, medium, and small joints, followed by progressive digital sclerosis over several months in the absence of treatment.

The patient was admitted to the pediatric emergency department with dyspnea at rest and palpitations, without other associated symptoms.

Results:

On examination, she was tachycardic (140 bpm) and tachypneic (54 breaths/min). Clinical evaluation revealed scleroderma affecting the hands and feet, multiple fingertip ulcers, telangiectasias, arthralgia and myalgia and signs of right heart failure (jugular vein distention, hepatojugular reflux, and a systolic murmur at the tricuspid area)

Urgent echocardiography and chest CT identified a large pericardial effusion, right heart dilation, pulmonary artery dilatation, and interstitial lung opacity with massive pleural effusion.

Immunological workup revealed positive antinuclear antibodies (ANA) at a titer of 1.1, while anti-Scl70 and anti-DNA antibodies were negative. Inflammatory markers were within normal ranges.

Conclusion:

Juvenile systemic sclerosis is an exceptionally rare entity with distinct clinical and immunological features compared to adult-onset SSc. Given its potential severity, early recognition and prompt intervention are crucial to prevent irreversible organ damage and improve prognosis. Increased awareness among clinicians can aid in timely diagnosis and tailored management, ultimately enhancing patient survival and quality of life.

Guselkumab Pharmacokinetics and Immunogenicity in Pediatric Psoriasis: Phase 3 PROTOSTAR Study

Vikash Sinha¹, Herta Crauwels¹, Miriam Zimmermann*², Obinna Obianom¹, Bart van Hartingsveldt¹, Meg Jett¹, Gao Jiang¹, An Vermeulen¹

¹Johnson & Johnson, Spring House, PA, United States ²EMEA Medical Affairs, Johnson & Johnson, Zug, Switzerland

Introduction & Objectives:

PROTOSTAR evaluated guselkumab (GUS), a selective IL-23p19 subunit inhibitor, in pediatric study participants (aged \geq 6-<18 years) with moderate-to-severe plaque psoriasis (PsO; ClinicalTrials.gov: NCT03451851). GUS pharmacokinetics (PK) and immunogenicity were assessed to determine whether PK exposure achieved with pediatric weight-based (WB) dosing was comparable with that established for the approved adult dose regimen. In prior studies, mean steady-state (SS) trough serum GUS concentration in adult PsO participants was approximately 1.2 μ g/mL.

Materials & Methods:

In Part 1 (Weeks [W]0–16), participants were randomized to GUS, placebo, or an open-label etanercept reference arm. At W16 (primary endpoint), Part 1 participants entered GUS withdrawal/retreatment, GUS continuation, or crossover to GUS study periods (W16–52). Part 2 evaluated continuous, open-label GUS treatment in a single arm of participants aged \geq 12 years (W0–52). Participants received a WB GUS dose of 1.3 mg/kg for participants <70 kg or 100 mg for participants \geq 70 kg. PK and immunogenicity endpoints (Parts 1 and 2) included serum GUS concentrations through W16 and W44, and proportions of participants with antibodies (Ab) to GUS, neutralizing Ab (NAb), and Ab titers.

Results:

In Parts 1 and 2, 41 and 28 participants received GUS, respectively. In Part 1, mean serum GUS concentrations at W16 were slightly lower in participants aged \geq 6-<12 years (2.83 µg/mL) vs \geq 12-<18 years (3.61 µg/mL); however, ranges largely overlapped. Similar W16 concentrations were observed for the <70 kg and \geq 70 kg groups (3.53 and 3.19 µg/mL, respectively). In Parts 1 and 2, SS trough serum GUS concentrations were achieved by W20 and maintained through W44. In Part 2, W20 mean SS trough serum GUS concentrations were 1.50 and 1.54 µg/mL for the <70 kg and \geq 70 kg groups, respectively. Among 114 GUS-treated participants with available samples through W44, 21 (18.4%) tested positive for Ab to GUS; none had NAb. Ab titers were generally low (80.0% had titers \leq 1:160). The development and titers of Ab to GUS did not impact GUS PK or clinical response.

Conclusion:

The observed PK for GUS in pediatric PsO participants receiving WB dosing was generally comparable with PK in adults with PsO.

Efficacy and Safety of Combined Pulsed Dye laser and Propranolol vs. Monotherapy with Pulsed Dye Laser or Propranolol for Infantile Hemangioma: A Systematic Review and Meta-Analysis

Mishari Alrubaiaan*¹, Ahmed Almajed², Abdulelah Alghamdi³, Yazeed Alowairdhi⁴, Rahaf Alothman⁵, Faisal Alghamdi⁶, Reem Bin Idris⁷, Rayan Alkhodair^{8, 9, 10}

¹King Saud Bin Abdulaziz University for Health Sciences, Riyadh, Saudi Arabia. , College of Medicine, Riyadh, Saudi Arabia

²King Saud Bin Abdulaziz University for Health Sciences, Riyadh, Saudi Arabia., Riyadh, Saudi Arabia

³King Saud Bin Abdulaziz University for Health Sciences, Riyadh, Saudi Arabia. , Division of Dermatology, Riyadh, Saudi Arabia

⁴King Abdulaziz Medical City, Department of Dermatology, Riyadh, Saudi Arabia

⁵King Abdulaziz University, jeddah, Saudi Arabia

⁶King Saud Bin Abdulaziz University for Health Sciences, Riyadh, Saudi Arabia

⁷King Saud University, Riyadh, Saudi Arabia

⁸King Saud bin Abdulaziz University for Health Sciences, College of Medicine, Department of Dermatology, Riyadh, Saudi Arabia

⁹King Abdullah International Medical Research Center, Riyadh, Saudi Arabia

¹⁰King Abdullah Specialized Children's Hospital, Division of Pediatric Dermatology, Department of Pediatrics, Riyadh, Saudi Arabia

Efficacy and Safety of Combined Pulsed Dye Laser and Propranolol vs. Monotherapy with Pulsed Dye Laser or Propranolol for Infantile Hemangioma: A Systematic Review and Meta-Analysis

Introduction & Objectives:

Background

Infantile hemangioma (IH) is one of the most common benign vascular tumors in pediatrics. propranolol is the first-line treatment for IH, pulsed dye laser (PDL) is used as an adjuvant therapy. The efficacy and safety of combined PDL and propranolol therapy compared with monotherapy remain unclear.

Objectives:

To determine the efficacy and safety of combined PDL and propranolol compared to monotherapy with either treatment.

Materials & Methods:

This systematic review and meta-analysis according to PRISMA guidelines. Databases included: PubMed, Google Scholar, Web of Science, Ovid, and Ebsco. Patients treated with PDL, propranolol, or both were included. Outcomes included complete clearance, healing time, residual lesions, side effects, and discoloration. Statistical analysis was performed using Review Manager 5.4 and Comprehensive Meta-Analysis v3 software

Results:

Six studies involving 1,788 patients were included: 139 received combined PDL and propranolol therapy, 1,101

received PDL therapy, and 548 received propranolol therapy. complete clearance was significantly higher with combined therapy than with PDL alone (OR = 13.03, 95% CI: 4.72-35.96, P < 0.001) and propranolol alone (OR = 2.90, 95% CI: 1.00-8.39, P = 0.05). There was no significant difference in healing time between combined therapy and monotherapy (SMD = -0.083, 95% CI: -1.011 to 0.845, P = 0.861). Adverse events were similar in all groups, and there was no significant difference between combination therapy and monotherapy (RR = 1.19, 95% CI: 0.26-5.32, P = 0.82 for PDL alone and RR = 1.39, 95% CI: 0.75-2.58, P = 0.30 for propranolol alone). Combination therapy was associated with a lower risk of residual lesions compared with PDL alone (RR = 0.71, 95% CI: 0.53-0.97, P = 0.03), but not significantly different from propranolol alone (RR = 1.19, 95% CI: 0.81-1.73, P = 0.37)".

Conclusion:

Combination therapy of PDL and propranolol increased the complete clearance rate of IH without increasing side effects. However, all treatments resulted in similar healing times. randomized controlled trials are needed to investigate the long-term efficacy and safety of combination therapy for IH.

Atypical Age of Onset in Eosinophilic Pustular Folliculitis: Expanding the Pediatric Spectrum

Maria Eduarda Rinaldi¹, Jenniffer Taveras¹, Gabriela Cortines Blanc¹, Julia Ricardo Passos Cazelli¹, Karime Menezes¹, Luiza Soares Berenbaum¹, Mariana de Gusmão Nunes¹, Fernando Manoel Belles de Moraes¹

¹Institute of Dermatology professor Rubem David Azulay, Rio de janeiro, Brazil

Introduction & Objectives:

Eosinophilic pustular folliculitis (EPF), also known as Ofuji's disease, was first described in Japan in 1965. It is a rare, chronic dermatosis characterized by a recurrent course. Although its etiology remains unclear, EPF is frequently associated with immunosuppressed states; nevertheless, it has been reported across a wide range of population groups. Clinically, it manifests as pruritic papulopustular lesions of non-infectious origin, with eosinophilic infiltration observed on histopathological examination. We report a case with an atypical clinical presentation, followed by a review of the pathology and a discussion of the diagnostic and therapeutic challenges involved.

Materials & Methods:

A comprehensive review of the literature was carried out for this case, noting that eosinophilic folliculitis in children is an exceptionally rare and poorly documented condition, with only a limited number of reports available to date.

Results:

A 7-year-old male patient presented to our clinic with pruritic erythematous-violaceous papular lesions that appeared approximately one month prior, initially localized to the plantar region. These lesions progressively disseminated across the entire body, with a marked predilection for the flexural areas, where they appeared more clustered. Isolated pustules were also noted, predominantly on the lower limbs. He had previously been treated with antihistamines, systemic corticosteroids, and empirical therapy for scabies, without achieving significant clinical improvement. Histopathological examination revealed perivascular and perifollicular dermatitis with follicular involvement and intense eosinophilia, consistent with eosinophilic folliculitis.

Conclusion:

Eosinophilic pustular folliculitis (EPF) is a chronic, pruritic, eruptive dermatosis with a wide geographic distribution, characterized by three main clinical variants: the classic form, predominantly affecting young adult males; the immunosuppression-associated form, typically observed in HIV-infected patients; and the infantile form, occurring between 12 and 36 months of age. The present case does not exhibit a typical epidemiological pattern.

Diagnosis is established based on clinical presentation and histopathological findings. Clinically, EPF is characterized by sterile, follicular, pruritic papules and pustules, which tend to coalesce into chronic and recurrent polycyclic or annular erythematous plaques.

Topical corticosteroids represent first-line therapy. In extensive or refractory cases, alternative treatments such as indomethacin, phototherapy, isotretinoin, calcineurin inhibitors, and oral antibiotics may be considered. The multifactorial nature of EPF etiology is reflected in the favorable response to various therapeutic approaches.

This case underscores the necessity of considering a broader epidemiological spectrum in EPF and highlights the challenges associated with its therapeutic management.

Clinical and Endocrinological Insights into Pre-Adolescent Acne: Beyond the Surface

Pelin Koçyiğit¹, Gizem Nur Öztürk*¹, Merve Alızada¹, Feride Ongun¹, Sıla Şahin¹, Elif Özsu²

¹Ankara University of Medicine, Dermatology, Ankara, Turks and Caicos Islands

Introduction & Objectives: Acne vulgaris, a pilosebaceous unit disorder, is increasingly observed in the preadolescent period (ages 7–12), raising concern due to potential hormonal and metabolic implications. Although it shares mechanisms with adolescent acne such as increased sebaceous activity and hormonal changes, distinct endocrine and clinical features suggest a need for targeted evaluation and management. This study aims to analyze the clinical presentations, hormonal and metabolic parameters and risk factors in preadolescent acne patients.

Materials & Methods: A retrospective analysis was conducted on 30 patients aged 7–12 years diagnosed with acne vulgaris between January 2024 and January 2025. Dermatological findings, family history, pubertal signs, BMI percentiles and hormonal/metabolic profiles including** FSH, LH, DHEAS, cortisol,** SHBG, 17-hydroxyprogesterone, testosterone levels, lipid profile** and any potential triggering factors were assessed. Spearman's and Kruskal-Wallis tests were performed using SPSS v23.

Results: The study included 30 patients (80% female) with a mean age of 11.2 years. Clinical acne grading was distributed as comedonal (n=11), papulopustular (n=11) and nodulocystic (n=8). Overall mean age of acne onset was 11.5 years whereas, it was 9 years in the nodulocystic group. Facial involvement was predominant. Family history was positive in 20 patients, including 75% of nodulocystic cases. Obesity (≥95th percentile) was identified in 6 patients (20%) and 3 patients were overweight. Dyslipidemia was detected in 4 patients, all of whom exhibited significantly earlier acne onset (p=0.094). Clinical signs of hyperandrogenism were observed in 56.6% of patients including hirsutism (37%), seborrhea (29.6%) and acanthosis nigricans (11.1%). Laboratory confirmed premature adrenarch was present in only one patient. Although LH levels were within the normal range in all patients, a significant positive correlation between acne severity and relatively higher LH levels (r=0.73, p=0.0013) was identified among pubertal female patients. Systemic isotretinoin was initiated in 10 patients, with treatment responses lower than expected.

Conclusion: Preadolescent acne, with its rising prevalence, may indicate early hormonal activity. Our findings support previous literature indicating a female predominance and facial distribution. Comedonal acne was not predominant and nodulocystic type was more prevalent than expected. The mean age of acne onset was significantly lower in the nodulocystic group. Family history of severe acne was present in 75% indicating a possible genetic predisposition. Obesity, found in 20% of patients, did not correlate significantly with acne severity. However, early-onset acne in patients with dyslipidemia suggested a potential role for metabolic factors. While over half of the patients showed clinical signs of hyperandrogenism, only one had laboratory confirmed endocrinologic pathology. The significant correlation between relatively high LH levels and acne severity implies that LH-driven androgen secretion may contribute to pathogenesis of acne during puberty. This study offers a comprehensive evaluation of preadolescent acne integrating a wide spectrum of clinical and endocrine factors. Further larger-scale studies are warranted to elucidate the unique characteristics of preadolescent acne.

²Ankara University of Medicine, Pediatrics, Pediatric Endocrinology, Ankara, Turks and Caicos Islands

Table: Clinical and Metabolic Characteristics by Acne Subtype

Feature	Comedonal acne	Papulopustular acne	Nodulocystic Acne
	(n=11)	(n=11)	(n=8)
Mean age of acne	10	11	9
onset (years)			
Mean BMI (kg/m²)	23.5	17.2	23.5
Obese/overweight (%)	45.5%	27.3%	25%
Dyslipidemia (n)	2	2	-
Menarche reached	0	80%	100%
(female %)			
Clinical	5	8	4
hyperandrogenism			
signs (n)			
Hormonal* laboratory	1	-	-
abnormality (n)			

^{*:} Total and free testosterone, dihydroxyepiandrosterone sulfate (DHEAS), follicle-stimulating hormone (FSH), luteinizing hormone (LH), thyroid-stimulating hormone (TSH), prolactin (PRL), sex hormone binding globulin (SHBG), 17-hydroxyprogesterone (17-OHP), fasting cortisol and adrenocorticotropic hormone (ACTH)

Sjögren-Larsson Syndrome: What Can We Learn from Six Cases?

Ana Clara Maia Palhano*¹, Fábio Augusto Peroni Garcia¹, Thais Kohatsu Yanase¹, Luciana de Paula Samorano Lima¹, Maria Cecília Rivitti-MacHado¹, Zilda Najjar Oliveira¹

¹University of Sao Paulo, São Paulo, Brazil

Introduction & Objectives:

Sjögren-Larsson Syndrome (SLS) is a rare autosomal recessive genetic disorder resulting from mutations in the ALDH3A2 gene, which encodes the fatty aldehyde dehydrogenase (FALDH) enzyme¹. This condition causes lipid accumulation that impairs skin, eye, and nervous system function, typically presenting as ichthyosis, spasticity, and delayed neuropsychomotor development (DNPM) ¹, ². We describe and analyze the clinical features and family history of six patients with SLS seen at a tertiary dermatology center.

Materials & Methods:

Data from 6 patients diagnosed with SLS were analyzed (Table 1). Patient ages ranged from 12 to 43 years. The diagnosis was based on clinical criteria defined by the classical triad of the disease. All patients exhibited ichthyosis, typically presenting as lamellar scaling with associated pruritus. In addition, 2 of the 5 patients showed palmoplantar hyperkeratosis. Spasticity was observed in all cases, with variations in pattern including spastic diparesis in four patients and spastic quadriplegia in 2 of them. Altered DNPM was also present in all individuals. The evaluation of skeletal abnormalities revealed scoliosis in one patient and foot deformities in two others, including plano-valgus feet. Two patients had a history of prematurity and history of consanguineous parents. Genetic testing was performed in 2 cases and confirmed ALDH3A2 gene mutations. Extracutaneous manifestations varied among the patients, with bilateral sensorineural hearing loss reported in 1 case and auditory processing disorder in another.

Results:

In SLS, the accumulation of long-chain fatty alcohols leads to alterations in cell membrane integrity, impairing the cutaneous barrier and resulting in ichthyosis². Ichthyosis is a major diagnostic marker, often present at birth or within the first months of life¹. Pruritus is a frequently described manifestation, likely related to barrier disruption or elevated levels of leukotriene B4³. Palmoplantar hyperkeratosis, seen in two of the six patients, is not typically considered a hallmark of the syndrome. Prematurity, presented in two of our patients, is common and is likely due to abnormal lipid metabolism. Neurological manifestations were also predominant, with spasticity and delayed DNPM observed in all patients³. The spasticity reflects the systemic impact of the enzymatic deficiency, which disrupts neuronal myelination³. Our data also highlight extracutaneous findings such as bilateral sensorineural hearing loss and auditory processing disorder. Dysphagia, observed in one patient, is a less frequently reported and may be secondary to spasticity or other neuromuscular abnormalities³. Although genetic testing was not performed in all cases, diagnosis was based on consistent clinical criteria ^{1,2,4}.

Conclusion:

The clinical variability observed in this case series emphasizes the importance of thorough clinical and laboratory investigation when SLS is suspected.

Photodynamic therapy for pediatric genital warts

Federica Li Pomi¹, Mario Vaccaro², Francesco Borgia²

¹University of Palermo, Palermo, Italy

²University of Messina, Messina, Italy

Introduction & Objectives:

Genital viral warts in pediatric patients present both therapeutic and psychosocial challenges. Conventional treatments may be painful, poorly tolerated, or associated with high recurrence rates. Photodynamic therapy (PDT) has emerged as a non-invasive and effective approach for viral warts.

Materials & Methods:

Four pediatric patients with clinically diagnosed genital viral warts underwent treatment with conventional PDT. Each patient received three sessions at monthly intervals. For each session, 10% 5-aminolevulinic acid (ALA) in polyethylene glycol ointment (Biosynth AG, Staad, Switzerland) was applied under occlusion for 3 hours. Illumination was performed using a diode red light source emitting at 630 nm (S630, Alpha Strumenti, Milan, Italy), positioned 50 mm from the skin surface, delivering a skin irradiance of approximately 160 mW/cm². The light exposure lasted 8 minutes, for a total light dose of 75 J/cm².

All patients received instructions for home care, including the application of a bland emollient cream and local non-alcoholic disinfection solution. Clinical follow-up was performed after each session and at 3 months post-treatment.

Results:

All four patients achieved complete clinical resolution of the lesions at the three-month follow-up. The treatment was well tolerated, with only mild erythema and a transient burning sensation during illumination. No recurrences were observed at follow-up. Cosmetic outcomes were excellent, with no residual scarring or post-inflammatory changes

Conclusion:

Conventional PDT with 10% ALA is a safe, effective, and non-invasive option for the treatment of genital viral warts in pediatric patients. Its high tolerability and excellent cosmetic results make it a valuable alternative to traditional destructive therapies, especially in sensitive anatomical areas and in young patients.

Pediatric Lichen Sclerosus: A Self-Limited Disease? A Long-Term Follow-Up Case Series

Carlos Nogueira*¹, Filomena Azevedo², Carmen Lisboa Silva^{2, 3}

- ¹Unidade Local de Saúde de Braga, Dermatology and Venereology, Porto, Portugal
- ²Unidade Local de Saúde de S. João, Dermatology and Venereology, Porto, Portugal
- ³Faculdade de Medicina da Universidade do Porto, RISE-Health, Pathology Department, Porto, Portugal

Introduction & Objectives:

Pediatric genital lichen sclerosus (LS) has historically been considered a self-limited condition, with expected remission at puberty. However, growing evidence challenges this assumption, suggesting possible persistence of symptoms and anatomical changes into adulthood. Our study aimed to evaluate the clinical course of childhood-onset genital LS into adulthood, through a retrospective data review and direct patient follow-up.

Materials & Methods:

We conducted a case series analysis of all pediatric patients (≤18 years) diagnosed with genital LS in the Dermatology Department of a tertiary care hospital, before 2010. Medical records were reviewed for demographic data, clinical presentation, treatment, and documented follow-up visits. Additionally, patients were contacted to determine current symptoms, anatomical changes, and need for continued management. If needed, clinical reassessment was offered.

Results:

Five patients were included. Three of them were female and the mean age at diagnosis was 11 years. All presented with hypopigmented atrophic patches and local pruritus, with at least one year of ongoing symptons before the diagnosis. In four cases, the diagnosis was confirmed with histopathological examination. Initial treatment involved high-potency topical corticosteroids with variable tapering and maintenance strategies, with clinical remission achieved in all cases during pediatric follow-up and eventual medical discharge. After a mean follow up time of 16,2 years, only two patients reported sustained remission without symptoms or signs of LS. One male patient experienced relapse during adolescence and early adulthood, evolving with significant scarring and urethral narrowing, necessitating prolonged dermatological and psychiatric care, as well as several urological surgeries. Two female patients also reported relapse of symptoms, requiring medical observation and reinstatement of topical treatment, which they mantain, without complications.

Conclusion:

Our findings support previous literature questioning the notion that pediatric LS always resolves at puberty. Only two of our patients remained asymptomatic, while three cases had chronic relapsing symptons during and after puberty, one of which with major complications. These results align with published data, which describe persistent symptoms and physical signs beyond puberty in 60–75% of patients. Delayed diagnosis and inconsistent treatment adherence may contribute to worse outcomes. Long-term follow-up into adolescence and early adulthood is crucial to monitor for relapses and prevent sequelae. Our case series, though limited by small sample size, highlights the need for continued surveillance and reinforces the safety and effectiveness of potent topical steroids as first-line treatment.

Pyoderma Gangrenosum as an Uncommon Presentation of Myelodysplastic Syndrome in a Pediatric Patient: A Case Report

Adam Yu¹, Airiss Chan², Loretta Fiorillo^{2, 3}

- ¹University of Alberta, Faculty of Medicine and Dentistry, Edmonton, Canada
- ²University of Alberta, Department of Dermatology, Edmonton, Canada
- ³University of Alberta, Department of Pediatrics, Edmonton, Canada

Introduction & Objectives:

Pyoderma gangrenosum (PG) is a rare, ulcerative neutrophilic dermatosis, often associated with systemic diseases such as inflammatory bowel disease and hematologic malignancies. Pediatric cases are uncommon, comprising less than 5% of all PG diagnoses, and present unique diagnostic challenges due to clinical variability and low index of suspicion. We report a case of PG in a 3-year-old male with underlying myelodysplastic syndrome (MDS), where initial misdiagnosis led to delayed treatment and surgical complications.

Materials & Methods:

We conducted a retrospective chart review of a pediatric patient with pyoderma gangrenosum (PG) in the setting of myelodysplastic syndrome (MDS). Clinical history, treatment course, imaging, histopathology, and microbiological findings were reviewed. Diagnostic evaluation included skin biopsy, bacterial cultures, and serial photographs of lesion evolution. A multidisciplinary approach involving dermatology, hematology, infectious diseases, and surgery was employed. Treatment response was assessed following immunosuppressive therapy.

Results:

A previously healthy 3-year-old boy with recently diagnosed MDS presented with a non-healing right forearm wound following minor trauma. The lesion evolved rapidly, accompanied by fever, and was initially treated as cellulitis with intravenous antibiotics. Despite temporary improvement, the wound deteriorated after scab disruption during venipuncture, suggesting pathergy. Surgical debridement and bilateral advancement flap closure were performed. Cultures grew methicillin-sensitive Staphylococcus aureus; however, the wound dehisced and progressed after surgical intervention. Dermatology evaluation was sought after failure of conventional treatment.

On examination, a 2–3 cm ulcer with violaceous undermined borders and significant surrounding erythema was noted. Histopathology revealed ulceration with dense neutrophilic infiltrates and abscess formation and were negative for infectious organisms. Based on the clinical features, histopathologic findings, and poor antibiotic response, PG was diagnosed. Immunosuppressive treatment with intravenous methylprednisolone (1 mg/kg/day), topical clobetasol, and fluticasone resulted in rapid improvement. The patient achieved full remission after two months of corticosteroid therapy.

Conclusion:

PG should be considered in pediatric patients with underlying hematologic conditions who present with non-healing, rapidly progressive ulcers, particularly in the context of pathergy and antimicrobial resistance. A multidisciplinary approach and heightened clinical awareness are critical for timely diagnosis and effective treatment

Napkin Psoriasis in Infancy: A Predictor of Adult Psoriasis?

Corrado Zengarini*^{1, 2}, Federico Bardazzi², Alessandro Pileri^{1, 2}, Martina Mussi^{1, 2}, Bianca Maria Piraccini^{1, 2}, Michelangelo La Placa^{1, 2}, Alessandra Gelmetti^{1, 2}, Miriam Leuzzi^{1, 2}

¹University of Bologna, Department of Medical and Surgical Sciences, Bologna, Italy ²IRCCS Azienda Ospedaliero-Universitaria di Bologna, Dermatology Unit, Bologna, Italy

Introduction & Objectives:

Napkin psoriasis (NP) is considered a skin manifestation in itself and is debated whether it needs to be considered a variant of infantile psoriasis limited to the diaper area. Its potential association with the development of adult psoriasis remains unclear. Our study aimed to evaluate the long-term risk of psoriasis in individuals diagnosed with NP during infancy and to identify potential predictive factors for disease progression.

Materials & Methods:

We conducted a retrospective review of medical records from a single pediatric dermatology unit, including patients diagnosed with NP between 1990 and 2022. Follow-up data were analysed to determine progression to psoriasis in later life. Collected variables included demographics, NP characteristics, family history, and outcomes. Statistical analyses included frequency analysis, Fisher's exact test, t-tests, and univariate regression.

Results:

Of 130 patients with diaper dermatoses over 32 years, 17 met the inclusion criteria. Among them, 7 developed psoriasis in adulthood, yielding a prevalence of 41% and an annual incidence of 1.15%. The odds ratio (OR) for developing psoriasis was 23.44 (95% CI: 8.33-65.92; p < 0.0001). However, NP duration, presence of distant lesions, and family history were not significantly associated with progression to psoriasis.

Conclusion:

Infants diagnosed with NP appear to have a significantly increased risk of developing psoriasis later in life compared to the normal Italian incidence. While limited by its retrospective nature and small sample size, this study highlights the relevance of long-term follow-up in NP patients and calls for further research into the underlying mechanisms linking NP to adult psoriasis.

Generalized Granuloma Annulare in an Infant Clinically Manifested as Papules After Vaccination: A Rare Case

Ferdi Öztürk¹, Elif Nida Altiparmak Altinel¹, Şaduman Balaban Adım², Hayriye Saricaoglu¹

¹Uludag University Faculty of Medicine, Dermatology and Venereology, BURSA, Türkiye ²Uludag University Faculty of Medicine, Pathology, BURSA, Türkiye

Introduction & Objectives: Granuloma annulare (GA) is a common, benign, self-limiting inflammatory skin disorder that can affect both children and adults. Generalized granuloma annulare (GGA) accounts for about 15% of GA cases and typically consists of multiple papules and plaques, with or without annular formation, on the trunk and extremities. Most commonly it affects adults aged 50 years and older, children aged about 10 years, and rarely infants.

Materials & Methods: We present a case of generalized granuloma annulare (GGA) in a 4-month-old girl following vaccination. At 2 months of age, she received the BCG vaccine, the first dose of diphtheria-tetanuspertussis (DTaP), Haemophilus influenzae type b (Hib) vaccinations, and conjugated pneumococcal vaccine (CPV). The patient also received the second dose of the DTaP-Hib and CPV vaccinations 5 days prior to the onset of the lesions. One month after the BCG vaccination and 5 days after the second dose of the DTaP-Hib and CPV vaccinations, asymptomatic, slightly erythematous papular lesions emerged, initially localized to the trunk, and subsequently spread to the extremities, with a marked increase in the number of lesions over the course of several days. Prior to her examination, she had not received any treatment other than the application of a moisturizing cream. Dermatological examination revealed diffuse 1-2 mm skin-colored papular lesions on the trunk and extremities and an erythematous, crusted BCG vaccination scar on the left upper arm. Skin biopsy revealed changes consistent with palisaded granulomas, characterized by peripheral histiocytes and occasional multinucleated giant cells, along with central mucinous degeneration. Routine laboratory tests were normal, and Darier's sign was negative. The lesions were considered to be GGA triggered by vaccination. The lesions showed a tendency to regress, and hydrocortisone acetate cream 0.5% was recommended for 1 week. On re-evaluation one month later, the lesions had healed with post-inflammatory hyperpigmentation. It was also noted that the patient had received the third dose of the DTaP-Hib vaccine immediately after the last visit. No increase in the lesions was observed.

Results: Although the patient's lesions were diagnosed as vaccination-triggered GGA, the simultaneous administration of the BCG, DTaP-Hib, and CPV vaccinations caused confusion regarding which vaccination had triggered the lesions. However, after the third dose of the DTaP-Hib vaccination, there was no recurrence of the lesions, which ruled out the possibility that this vaccination had triggered the lesions. The BCG and CPV vaccinations were considered to be responsible for triggering the lesions. If the lesions do not recur after receiving repeated CPV by 12 months of age, they can be definitively attributed to the BCG vaccination.

Conclusion: The exact cause of GA remains unknown, but it is believed to be triggered by multiple factors. Vaccinations are an infrequent cause of GGA. Cases have been reported following BCG, hepatitis B, diphtheria/tetanus, pneumococcal, and SARS-CoV-2 vaccinations. A few cases have been documented in the literature. This case highlights the rare occurrence of GGA in infants and its association with vaccination.

Kikuchi-fujimoto disease presented as aseptic meningitis: a case Report

ayat al zadjali¹, Jawahir Al Sharqi², Nawal Al Rasbi³

¹MOH, muscat, Oman

²omsb, muscat, Oman

³sultan Qaboos university hospital, muscat, Oman

Introduction & Objectives:

Kikuchi disease is a form of necrotizing lymphadenitis with characteristic histological appearances, first described in 1972 independently by Kikuchi and Fujimoto. Although the initial descriptions were in people of Asian origin, the disease has now been reported in individuals of all races. The pathogenesis is unclear, but a viral or post viral hyperimmune reaction has been suggested as a possible mechanism. Many subsequent studies have failed to detect the presence of specific infectious agents through serologic or polymerase chain reaction (PCR) testing. It is most likely that there is no single inciting infectious agent, and that the development of Kikuchi's disease represents a hyperimmune reaction to certain viral or bacterial antigenic stimuli. Here, we present the case of a 9-year-old child with fever and signs of meningitis, later diagnosed with KFD. This case highlights the overlap clinical presentation, diagnostic challenges, and management of KFD in a pediatric patient.

Materials & Methods:

A 9-year-old boy presented with a three-week history of persistent fever, headache, photophobia, cervical lymphadenopathy and discrete macular rashes lasted for 4-5 days and then resolved with hyperpigmentation.

Treated initially as meningitis until cerebrospinal fluid (CSF) analysis revealed lymphocytic pleocytosis without infectious etiology. Laboratory findings included bicytopenia, elevated ferritin, and transaminitis. Diagnostic imaging and infectious workup were unremarkable. Definitive diagnosis was established by lymph node biopsy with histopathological and immunohistochemical findings consistent with KFD.

Results:

Histopathology of the excised lymph node revealed necrotizing lymphadenitis with nuclear debris and vasculitis. Immunohistochemistry showed positive cluster of differentiation (CD3), positive CD68 in histocytes, and negative CD30. The patient's symptoms resolved with supportive care and a 14-day antibiotic course. Follow-up showed complete resolution of symptoms, including lymphadenopathy, without recurrence.

Conclusion:

Kikuchi-Fujimoto Disease is a rare, self-resolving condition primarily found in Asia, affecting adult women aged 20 to 35. In children, males are more affected, with a higher ratio in some studies. Its clinical presentation can mimic serious diseases, including painful cervical lymphadenopathy, fever, rash, joint pain and hepatosplenomegaly, often leading to misdiagnosis as lymphoma. Immunohistochemical testing can show positive results for lysozyme, CD68, Myeloperoxidase (MPO), CD4, CD163 for histocyte and CD8 positive T-cell and CD20 positive B cell for lymphocyte. In this case, CD3 was positive in necrotic foci, CD68 in histiocytes, and CD30 was negative. KFD is a self-limiting, with most cases resolving within six months with supportive management and often involving non-steroidal anti-inflammatory drugs (NSAIDs). Corticosteroids may be used for severe or relapsing cases. Despite being a benign disease, yet some fata cases were reported.

A Rare Culprit Behind Eyelid Changes in a Child

Catalina Valencia Marín*¹, luisa lopera Botero², lina vanessa gómez gómez³

- ¹Universidad Pontificia Bolivariana , Dermatology, Medellín, Colombia
- ²Universidad Pontificia Bolivariana , Medellin, Colombia
- ³Hospital Pablo Tobón Uribe , Medellín, Colombia

Introduction & Objectives:

Lichen sclerosus is a chronic inflammatory dermatosis most commonly affecting the anogenital region. Extragenital presentations, particularly on the face, are rare and may be overlooked. Eyelid involvement is especially uncommon and often misdiagnosed. We present the case of a 10-year-old girl with isolated eyelid lichen sclerosus, initially mismanaged as atopic dermatitis, in whom dermoscopy played a crucial diagnostic role.

Objectives

- 1. Raise awareness of eyelid lichen sclerosus as a rare but important differential diagnosis in pediatric patients with persistent hypopigmented plaques in the face.
- 2. Highlight the role of dermoscopy in recognizing characteristic features of lichen sclerosus and aiding in non-invasive diagnosis.

Materials & Methods:

A 10-year-old girl with a personal history of allergic rhinitis presented to the dermatology clinic with a 3-year history of a hypopigmented lesion at the lateral canthus of the right eyelid, accompanied by pruritus. She denied any other symptoms. A KOH test performed previously was negative.

She had been treated with low-potency topical steroid for 10 days and oral antihistamines, but reported no improvement.

On physical examination, an ivory-colored plaque measuring 7×5 mm was observed on the right upper and lower eyelids, with irregular but well-defined borders, involving the palpebral margin. Dermoscopy revealed white structureless areas with follicular plugging.

The diagnosis of extra-genital lichen sclerosus was made, so treatment with topical steroid and topical calcineurin inhibitors was initiated.

Results:

Lichen sclerosus is a chronic, inflammatory disease that primarily affects the anogenital region. However, extragenital involvement is observed in up to 20% of cases, often involving the trunk, shoulders, and upper extremities, while facial involvement remains exceedingly rare1. Among facial presentations, the eyelids are an uncommon site, with only some case reports in the literature 2.

Eyelid lichen sclerosus typically manifests as asymptomatic, white, atrophic plaques that may resemble other diseases posing a diagnostic challenge3,4. Lesions may display features such as madarosis or even ptosis, as seen in both adult and pediatric presentations. A history of local trauma is sometimes present 4.

Dermoscopy has emerged as a tool in its assessment. Characteristic features include white to yellow structureless areas, linear irregular vessels, follicular plugs, and perifollicular scales1. These findings correlate with histopathologic features such as epidermal thinning, follicular plugging, dermal hyalinization, and lichenoid lymphocytic infiltrate1,2.

Timely recognition and treatment are essential to prevent scarring complications. Topical calcineurin inhibitors and corticosteroids remain first-line therapies2,3.

Overall, eyelid lichen sclerosus represents a rare but important diagnostic consideration for persistent, hypopigmented eyelid lesions. Dermoscopy is a valuable tool that can aid in accurate diagnosis and may reduce the need for invasive procedures in selected cases.

Conclusion:

Eyelid lichen sclerosus is a rare but significant diagnostic consideration in children with refractory hypopigmented lesions. This case highlights the importance of clinical suspicion and the role of dermoscopy as a valuable, non-invasive tool that can support clinical suspicion and guide early intervention.

Topical Treatment Failure in Infantile Scabies: The Impact of Palmar-Plantar Skin and Fist Clenching Habits

Martina Mussi^{1, 2}, Iria Neri¹, Marco Adrano Chessa^{1, 2}, Alessandra Gelmetti^{1, 2}, Bianca Maria Piraccini^{1, 2}, Michelangelo La Placa^{1, 2}, Corrado Zengarini^{1, 2}

¹S. Orsola-Malpighi Polyclinic, Dermatology, Bologna, Italy

²Alma Mater Studiorum - Università di Bologna, Department of Medical and Surgical Sciences, Bologna, Italy

Introduction & Objectives:

Scabies is a widespread parasitic skin infestation increasingly affecting infants, in whom treatment failure is not uncommon. Unique behavioral and anatomical characteristics of neonates and infants may contribute to therapeutic resistance. This study aims to highlight the role of palmar-plantar occlusion in reducing permethrin efficacy and to propose a targeted modification of the standard treatment protocol.

Materials & Methods:

A 12-month prospective observational study was conducted at a single pediatric dermatology unit. Sixteen infants (<12 months) with clinically and microscopically confirmed scabies were treated with permethrin 5% cream using the standard 1-7-1 regimen. Clinical data, disease severity score, number of household members, ethnicity, and presumed source of contagion were recorded. Infants with persistent lesions localized to palmar-plantar areas at 2-week follow-up underwent a modified treatment protocol: an additional three-night localized application of permethrin to the palms and soles. All household contacts were treated simultaneously with standard therapy.

Results:

All patients (n=16) experienced localized persistence of scabies at the palmar-plantar level despite standard treatment. Following the modified protocol, complete resolution was achieved in 100% of cases at the 4-week evaluation. Behavioral observations showed that all patients exhibited habitual hand-clenching and frequent foot movements, and 93.7% wore socks during treatment. These behaviors may have contributed to occlusion-related treatment failure. No systemic side effects were reported; mild, self-limited irritation occurred in two cases (12.5%).

Conclusion:

Palmar-plantar occlusion represents a significant and underrecognized factor in scabies treatment resistance in infants. Modifying treatment to include localized applications of permethrin in occluded areas appears to overcome resistance and improve outcomes. This approach, combined with caregiver education to avoid occlusive behaviors, may optimize scabies management in this vulnerable population.

Utilizing reflectance confocal microscopy as a minimally-invasive, early diagnostic tool of multiple skin cancers on 10-year-old child with xeroderma pigmentosum

Claudia Lee*1, Tracy Funk1

¹Oregon Health and Science University, Dermatology, Portland, OR, United States

Introduction & Objectives:

Xeroderma Pigmentosum (XP) encompasses a group of rare, genetic conditions in which patients have a significantly higher risk of developing numerous skin cancers, beginning in childhood. Because pediatric patients may be unable to undergo skin biopsy and skin cancer treatment without sedation, children with XP often experience multiple general anesthesia events during their lifetime. Reflectance confocal microscopy (RCM) may be a useful tool for pediatric XP patients for identification of and management of concerning lesions with a goal of most effectively treating skin cancers while minimizing general anesthesia events when possible.

Materials & Methods:

We present a 10-year-old boy diagnosed with XP-A who presented with 3 skin lesions concerning for malignancy based on clinical and dermoscopic findings and characterized the confocal microscopic findings to non-invasively determine which lesions will require removal under general anesthesia.

Results:

On RCM, the first lesion (superior left cheek) revealed a meshwork pattern with bundles of heterogeneous melanocytic nests and folliculotropism. The second lesion (inferior left cheek) demonstrated an undefined pattern at the dermo-epidermal junction and multiple aggregates of atypical dendritic melanocytes Both these lesions were highly suspicious for melanoma based on combined dermoscopy-confocal features. The third lesion (right temple) revealed irregular epidermal architecture with enlarged keratinocytes, tumor islands with nuclear palisading and clefting which was highly suggestive of basal cell carcinoma. All three lesions were subsequently biopsied and dermatopathology confirmed a diagnosis of melanoma in-situ for the first and second lesions on the left cheek and basal cell carcinoma for the third lesion on the right temple.

Conclusion:

For many patients with XP, biopsy fatigue and phobia pose a unique challenge with legitimate consequences to the patient's mental well-being, especially in pediatric populations. RCM is a painless and non-invasive tool that may be useful for reducing unnecessary biopsies and selecting which lesions may reasonably combine diagnostic and treatment procedures in a single encounter to minimize procedure fatigue in these uniquely susceptible populations.

PELVIS syndrome associated to amniotic bands but not on the same limb

Aïcha Salhi*¹, madassi sarrah¹, fodil tassadit¹, kentouri mohamed², foughali mohamed³, aitbenamar abdelhamid²

¹University of Algiers, dermatology, Alger Centre, Algeria

²University of Algiers, surgery, Alger Centre, Algeria

³University of Algiers, cardiology, Alger Centre, Algeria

Introduction & Objectives: PELVIS syndrome describes the constellation of perineal infantile hemangioma IH, external genitalia malformations, lipomyelomeningocele, vesicorenal abnormalities, imperforate anus, and skin tag. Myelopathy, especially spinal dysraphism is the most common extradermal anomaly. Other acronyms like SACRAL and LUMBAR are used to describe this entity. The last one also includes bone defects of the lower body and arterial anomalies. Diagnostic is retained if a perineal segmental IH is associated to one of these abnormalities. Amniotic band syndrome (ABS) is a congenital condition consisting of *in utero* entrapment of fetal structures within fibrous amniotic constriction bands leading to limb defect or atrophy. ABS affects limbs or digits in 80 % of cases but rarely concerns thorax, abdomen or pelvis. We report a case of PELVIS syndrome associated to ABS.

Materials & Methods: A 4 months old male infant was referred to our institution for evaluation of a segmental IH covering the perineum and the entire left lower limb from buttocks to toes. During pregnancy the mother received aspirin and low molecular weight heparin for history of repeated abortion. Birth was at full term by vaginal delivery. He presented with laparoschisis, omphalocele, bladder extrophy, congenital dislocation of the left hip, anal imperforation and malformation of the external genitalia organs. The left upper limb had a constriction band encircling a shortened forearm, a small puffy hand and incomplete fingers. A surgery allowed for a parietal repair with colostomy. Six days later the bladder extrophy recurred due to a dehiscence. Whole spine MRI identified lumbo sacral spina bifida occulta and no myelomeningocele. Abdomino pelvic MRI revealed pelvic left ectopic kidney and developmental dysplasia of the hips. No internal genitalia organs were identified and no external genital differenciation was observed. On the 20TH day of life a segmental IH developed over the entire left lower limb and perineum through the coalescence of small punctuate angiomatous elements. Propranolol allowed IH healing and planing surgery while preventing bleeding..

Results: The pathophysiological mechanisms underlying ABS and IH are not fully understood, although they share a common risk factor: villous biopsy. In ABS, constriction induces a reduction in blood flow, leading to progressive ischemia which induces the development of a hypotrophic limb and ultimately its amputation. IH would be induced by released hypoxia inducible factor after acute ischemia following sudden circulatory arrest and therefoe hypoxy inducible factor release which stimulates vascular endothelial growth factor and then endothelial cells proliferation. A sudden circulatory arrest is not comparable to a progressive one. This might explain why IH are not observed in areas downstream of the constriction ring. We reviewved three other published cases of IH associated to ABS but they were never located on the same anatomic territory.

Conclusion: This case report of an infant with ABS involving the upper limb and PELVIS with IH affecting the lower limb allows comparison of the mechanisms at their origin

Impact of social media on the perception of skin diseases in adolescents and young children

Meryeme BOUTAAROURT*1, ouiame el jouari1, salim gallouj1

¹Mohamed VI Hospital Center in Tangier (CHU of Tangier), Dermatology and Venereology, TANGIER

Introduction & Objectives:

Over the past decade, social media has become a key platform especially among youth. For visible skin diseases like acne or eczema, it offers support but can also cause stigma and risky behaviors.

This study explores how adolescents and preadolescents are exposed to skin-related content, its psychological and behavioral effects and its impact on self-esteem, perception of dermatoses and healthcare-seeking aiming to guide future interventions.

Materials & Methods:

This descriptive cross-sectional study was conducted from January to March 2025 through an online questionnaire shared on social media, consisting of 21 questions. It targeted adolescents (aged 13-18), children (≤12) and parents responding on behalf of their children.

Data were analyzed with SPSS 21 using percentages for qualitative variables.

Results:

A total of 524 responses were collected; 58% from adolescents, 12% from children responding directly and 30% from parents (22% for children, 8% for adolescents). The sample was predominantly female (67%). Chronic skin conditions were reported by 41% mainly acne (60%), seborrheic dermatitis (22%), atopic dermatitis (18%) and vitiligo (7%). Social media use was almost universal (94%) with Instagram (78%), TikTok (65%), YouTube (52%) and Snapchat (38%) being the most used. Among adolescents, 72% reported spending more than two hours per day online.

The dermatology-related content viewed was diverse; 62% watched personal testimonials, 58% followed influencer advice, 49% saw skincare advertisements and 43% viewed "before/after" videos. Only 36% had access to educational content from healthcare professionals. The perceived impact was significant; 49% believed social media promoted unrealistic skin ideals and 55% reported negative emotions such as shame, stress or insecurity. About 31% changed their skincare behavior after exposure to online content.

Moreover, 21% consulted a dermatologist based on content seen online and 38% tried advice found on social media with 12% reporting worsened symptoms. Photo editing to hide skin imperfections was reported by 87% and 65% experienced social withdrawal due to their skin. While 36% felt encouraged to take better care of their skin, 51% believed social media increased awareness of dermatological conditions and 42% perceived it as a source of stigma.

Conclusion:

This study highlights the significant influence of social media on how adolescents and young children perceive skin diseases. It reveals widespread use of digital platforms with exposure to a wide range of dermatology-related content largely dominated by personal testimonials and influencer advice, while educational material from

healthcare professionals remains limited.

The findings reveal a dual effect; on one hand, increased awareness of skin conditions and motivation to improve skincare; on the other, the reinforcement of unrealistic beauty standards, emotional distress and risky behaviors such as self-treatment or excessive photo editing. The high rate of social withdrawal and efforts to conceal skin imperfections reflect the pressure of online image norms and the importance of appearance in digital spaces.

These observations underscore the importance of early media literacy, inclusive representation of skin conditions and reliable content. Collaboration between dermatologists and content creators could help combat stigma, support healthy self-image and transform social media into a more constructive space for young users navigating skin concerns.

Early Cutaneous Markers and Metabolic Derangements in Childhood Obesity - a cross-sectional study in 171 patients.

prathibha kuchana*1, niti khunger1

¹Vardhman Mahavir Medical College, Dermatology, New Delhi, India

Early Cutaneous Markers and Metabolic Derangements in Childhood Obesity - a cross-sectional study in 171 patients.

Introduction & Objectives:

Overweight and obesity has been associated with whole plethora of cutaneous and systemic manifestations. Cutaneous manifestations can act as markers for underlying metabolic derangements.

The objective of the study is to evaluate cutaneous manifestations and metabolic profile in overweight and obese, children and adolescents.

Materials & Methods:

In this cross-sectional descriptive study, 171 patients aged 5 to 18 years classified as overweight and obese according to IAP gender specific BMI charts were included. After informed consent from the parents/guardians, detail dermatological examination was done and investigated for metabolic profile.

Results:

Out of 171 patients (male:88, female:83) 63 were children and 108 were adolescents. The most common cutaneous manifestation was acanthosis nigricans (74%), acrochordons (32%), striae (25%), acne (16%) followed by seborrheic dermatitis, keratosis pilaris, pityriaisis alba and others. 15% had metabolic syndrome and most of them were obese. 12% had deranged LDL, 4% had deranged triglycerides, 36 % had deranged HDL, 4% had deranged blood sugars, 55% had insulin resistance, 65% had vitamin D deficiency, 15% had insufficient vitamin D levels. Acanthosis nigricans was significantly associated with insulin resistance.

Conclusion:

Cutaneous markers associated with obesity in children help in detection of underlying metabolic disturbances, there-by preventing long term complications.

Riga-Fede disease: a new diagnostic challenge

Safa Djebbas¹, Mansoul Tarek¹, Boussaid Riadh¹, Chehad Ahmed Samaouel¹

 $^{
m 1}$ University hospital Dr Abdesselam Benbadis Contantine Algeria, dermatology, Constantine , Algeria

Introduction & Objectives: Riga-Fede disease is a benign ulceration of the oral mucosa, secondary to repetitive dental trauma, most often caused by the eruption of the primary lower central incisors. Although this lesion was first described in 1881, few cases have been published. We report a case in a four-month-old infant.

Materials & Methods: A four-month-old infant from a consanguineous marriage, born at term vaginally, was brought to the clinic with a painful lesion of the floor of the mouth that had been developing for a month and preventing feeding. Oral examination revealed a rounded, well-limited ulceration measuring approximately 1.5 cm in diameter, covered by an adherent whitish coating with an indurated base, located opposite the lower central incisors. The infant was in good general condition. Biological findings included normocytic anaemia (11 g/dL) and CRP (3 mg/L). On the basis of all the anamnestic and clinical data, we accepted the diagnosis of Riga-Fede disease. A smoothing of the incisal edges was proposed, pain ceased afterwards, feeding was resumed without difficulty and the ulceration disappeared after three months.

Results: In 1881, Antonio Riga first described a congenital or neonatal incisor in a newborn. In 1890, Francesco Fede published new cases. The diagnosis of Riga-Fede disease is purely clinical. Early management is essential, as the lesion can lead to deformation and mutilation of the tongue, with major somatic repercussions and, in extreme cases, retarded growth. Dental trauma is the most frequent cause. Treatment is conservative (smoothing, resin, dental protection), except in the case of supernumerary teeth or excessive mobility. Extraction is then the rule. Anatomopathological examination is only necessary in the absence of healing after treatment of the cause, to search for another diagnosis.

Conclusion: Ulcerations of the oral mucosa can have many etiologies that are sometimes difficult to identify, particularly in children. The diagnosis of Riga-Fede disease is sometimes one of the diagnostic challenges.

Malignant ectomesenchymoma of the forearm: unexpected clinical outcome in a pediatric patient

Aleksandra Osińska*¹, Magdalena Woźniak², Andrzej Materniak², Michalina Boruch², Marta Druszcz¹

¹Medical University of Lublin, Student Scientific Society at the Department of Pediatric Radiology, Lublin, Poland ²Medical University of Lublin, Department of Pediatric Radiology, Lublin, Poland

Malignant ectomesenchymoma of the forearm: unexpected clinical outcome in a pediatric patient

Introduction & Objectives:

Malignant ectomesenchymoma (MEM) is an uncommon and rapidly progressing malignant tumor affecting the nervous system or soft tissues, composed of both mesenchymal and neuroectodermal components. It primarily occurs in pediatric patients, with the pelvic region, retroperitoneal area, and genitourinary system being the most frequently affected primary sites. Current literature cites approximately 70 documented cases of MEM globally.

Materials & Methods:

A 22-month-old child was hospitalized in the Pediatric Hematology and Oncology Department for a follow-up ultrasound of the soft tissues and related laboratory evaluations.

Results:

During the consultation, the parents reported observing a painless nodular mass on the right forearm. An ultrasound examination revealed an intramuscular, polycyclic, well-defined, and heterogeneous lesion measuring 57x20x27mm on the palmar side of the forearm, with a chaotic blood flow pattern observed in the Color Doppler assessment. The sonographic findings were inconclusive and raised suspicion for a neoplastic condition. A biopsy was conducted, and histopathological analysis confirmed the presence of ectomesenchymoma. The patient was classified for chemotherapy for non-RMS-like conditions following the CWS - VAIA III protocol and initial staging categorized as HRG group; IRS III, N1.

Conclusion:

Due to the rarity of MEM and its heterogeneous nature, making a diagnosis is difficult. This results in a prolonged diagnostic and therapeutic process. In the presented case, the ultrasound findings indicated malignant features, including a subfascial position and a lesion size exceeding 5 cm. Furthermore, the irregular vessel morphology, including trifurcations and a disorganized, tortuous distribution of vessels with avascular areas likely indicating necrosis, suggested a chaotic vascular architecture. Surgical intervention remains the primary treatment strategy. The use of chemotherapy and radiotherapy is generally discouraged in children due to their potential adverse effects on neurodevelopment.

A deeper insight into the tumorigenesis of MEM may aid in identifying more suitable systemic treatments within current therapeutic frameworks.

Multiple juvenile xanthogranulomas in a 4-month-old-girl.

Armine Adilkhanyan¹, Tatevik Harutyunyan¹, Inga Mkhitaryan¹, Sylvie Fraitag², Alain Hovnanian³

¹Arabkir joint medical center, ICAH, Dermatology, Yerevan, Armenia

²Necker Hospital for sick children, Pathology, Paris, France

³INSERM UMR 1163, Imagine Institute and University of Paris, Paris, France

Introduction & Objectives:

Juvenile xanthogranuloma (JXG) is a benign non-Langerhans cell histiocytosis of unknown etiology, most commonly occurring in infants and children. It is characterized by solitary or multiple asymptomatic, yellowish cutaneous papules and nodules on the head and neck, trunk, and extremities. Mostly it presents with solitary lesion. Skin lesions of JXG often develop in the first year of life, although multiple lesions are more commonly present at birth or occur during the first 6 months of life. Extracutaneous involvement in JXG is rare, including the eyes, central nervous system, liver, kidney, spleen, lungs, heart, and bone marrow, and is most often associated with multiple JXG. Cutaneous manifestations in JXG may precede systemic involvement.

A diagnosis of JXG is based on characteristic clinical features, histopathology, and immunohistochemistry. Typical histologic findings in JXG are dense infiltration of histiocytes and Touton giant cells with a wreath of nuclei surrounded by foamy cytoplasm in the papillary and reticular dermis. Immunohistochemistry of JXG is typically strongly positive for CD68, and negative for S100 protein, CD1a, and Langerin.

In case of multiple JXG, additional examinations, such as ophthalmological examination, complete blood count, liver and renal function tests, chest X-ray, ultrasound of abdomen, sometimes brain MRI or CT scan, must be performed. JXG is usually self-limiting with skin lesions regressing in 3 to 6 years. Even multiple JXG resolves spontaneously in most cases.

Materials & Methods:

A 4-month-old girl presented with a history of asymptomatic, multiple yellow-orange papules and nodules, which had appeared since the age of one month and had gradually increased in number and size. Physical examination revealed numerous, yellowish-orange papules and nodules measuring 5-10 mm in diameter on the head, face, trunk, and lower extremities. She had no signs of extracutaneous involvement.

Results:

Laboratory investigations, including a complete blood count, serum lipid levels, renal and liver tests, were within normal range. Abdominal ultrasound examination, and an ophthalmologic examination revealed no abnormalities. A biopsy of a nodule on her leg was performed. Histopathological examination showed the proliferation of histiocytes throughout the dermis. There were no giant multinucleated cells. Immunostaining revealed multiple CD68+ cells and CD163+ histiocytes which did not express ALK nor TRK. S100 protein and CD34 were negative.

All these findings were consistent with the diagnosis of JXG.

Conclusion:

The correct diagnosis of JXG is essential in order to implement a regular follow-up with systemic evaluation to detect possible systemic involvement.

We could establish the diagnosis of multiple JXG in our young patient on the basis of the clinical findings and the results of immunohistopathology. Her follow-up to date has shown no extracutaneous involvement.

Congenital ichthyosis in children

Narjess Er-Rachdy¹, Ouissal Essadeq¹, laila benzekri¹, nadia ismaili¹

¹Departement of dermatology, Ibn Sina university hospital, Mohammed V university, Rabat, Morocco, rabat, Morocco

Introduction & Objectives:

Ichthyoses are a heterogeneous group of congenital or acquired disorders characterized by excessive epidermal shedding (resembling fish scales) due to a keratinization defect. They typically begin at birth or in the early years of life.

Materials & Methods:

This is a prospective monocentric and descriptive study conducted over an 30-month period at the Dermatology Department of Ibn Sina University Hospital in Rabat, Morocco.

Results:

We report a series of 40 cases, with an average age of 3 years and a female predominance.

Twenty-eight patients presented with lamellar ichthyosis, characterized by thick, dark scales that were generalized over the entire body in 80% of cases and localized to the extremities in 20% of cases. Nail dystrophy and palmoplantar keratoderma (KPP) were found in 50% of cases, and ectropion was observed in 8 patients.

The Netherton syndrome was identified in 6 patients, all of whom had erythematosquamous pruritic lesions and fragile hair, with a bamboo hair appearance seen in 3 patients through trichoscopy.

One case involved a harlequin baby, a female neonate with thick hyperkeratotic plaques interspersed with deep fissures covering the body, associated with severe ectropion and eversion of the lips (eclabium). The newborn also had hypoplasia of the left hand due to an amniotic band.

Conclusion:

Ichthyoses are complex syndromes with multiple clinical forms. Quality of life is often impaired, and treatment is symptomatic and palliative. There is a significant benefit in prenatal diagnosis, with hope for future advancements in gene therapy.

Castelman's disease whith pityriasis lichenoid

Narjess Er-Rachdy¹, Ouissal Essadeq¹, laila benzekri¹, nadia ismaili¹

¹Departement of dermatology, Ibn Sina university hospital, Mohammed V university, Rabat, Morocco, rabat, Morocco

Introduction & Objectives:

Castleman's disease (CD) is a rare, poorly understood lymphoproliferative disorder that share common lymph node histological features. Two clinical entities have been described: Unicentric with a confined disease to a single anatomic lymph node and multicentric characterized by generalized lymphadenopathy and more aggressive clinical course. It has three histological subtypes: hyaline vascular, plasma cellular and mixed type. Most reported cases of CD in the neck were of the hyaline vascular type and the most common sign was an asymptomatic neck mass. We report here the existence of a hyaline-vascular type of CD in a girl with chronic pityriasis lichenoide.

Materials & Methods:

A 5-year-old girl, presented with pustular and bullous lesions and ulceration. Physical examination revealed multiple firm, non-fixed masses on both sides of the neck.

Results:

A skin biopsy was in favor of chnonic pityriasis lichenoid. Ultrasound and CT scan showed multiple bilateral laterocervical adenomegaly and adenopathy. A cervical lymph node biopsy was performed in favor of castelman's disease in its hyalinizing vascular histological form.

Conclusion:

CD is a rare lymphoproliferative disorder, it is mostly reported in adults. Multicentric form, however, presents with systemic symptoms along with multiple lymph node hyperplasia.

Giant congenital melanocytic nevus and neurofibromatosis type 1

Narjess Er-Rachdy¹, Ouissal Essadeq¹, laila benzekri¹, nadia ismaili¹

¹Departement of dermatology, Ibn Sina university hospital, Mohammed V university, Rabat, Morocco, rabat, Morocco

Introduction & Objectives:

Giant congenital melanocytic nevus (GCMN) is a rare variety of Congenital melanocytic nevus characterized by its size, and its potential for transformation into malignant melanoma, it is infrequently associated with other findings which make the clinical picture complex. We report here a rare association of GCMN and neurofibromatosis type 1 (NF1).

Materials & Methods:

A 5-year-old child, presented with giant congenital nevus and multiple nodules soft, with a smooth surface, non pulsatile, non tender and freely mobile corresponding to neurofibromas and plexiform neurofibromas. Numerous medium-brown macules on the face, trunk, and extremities and he had no axillary freckling.

Results:

Ophthalmological assessment and neurological examination were both normal. Skeletal X-ray and brain and spinal MRI were normal. Biopsies performed at multiple sites were all in favor of a congenital nevus and neurofibroma.

In our patient, the diagnosis of type I neurofibromatosis was based on the presence of two clinical diagnostic criteria: café-au-lait macules and neurofibromas.

Conclusion:

In cases in which surgical excision is not feasible as in our patient, close clinical and dermoscopic follow up with biopsy of clinically suspicious areas is recommended.

Jacquet Erosive Dermatitis in a Child with DOCK8 Mutation- Clinical Improvement After Allogenic Hematopoietic Stem Cell Transplantation

Tubanur Cetinarslan¹, Göksu Dalgıç¹, Pınar Şahin², Gülcihan Özek³, Neslihan Edeer Karaca², Serap Aksoylar³, Peyker Temiz⁴, Coşkun Ekemen⁵, Aylin Türel Ermertcan¹, Regina Fölster-Holst⁶

Jacquet Erosive Dermatitis in a Child with DOCK8 Mutation- Clinical Improvement After Allogenic Hematopoietic Stem Cell Transplantation

Introduction & Objectives:

Dedicator of cytokinesis 8 protein (DOCK8) deficiency is a rare autosomal recessively inherited combined immunodeficiency, caused by mutations in the DOCK8 gene. Jacquet erosive diaper dermatitis (JED) is a rare and severe form of chronic irritant contact dermatitis.

Materials & Methods:

Herein, we report a girl with JED underlying DOCK8 mutation whose lesions regressed completely after allogenic hematopoietic stem cell transplantation.

Results:

A 9-year-old girl presented to our clinic with chronic, painful, non-healing ulcerative lesions in the genital and gluteal regions. She had been diagnosed with a dedicator of cytokinesis 8 (DOCK8) gene mutation one year prior to presentation. Over the following years, she experienced recurrent sinopulmonary infections, including otitis media and pneumonia, as well as frequent gastrointestinal and urinary tract infections, leading to multiple hospitalizations. Dermatological examination revealed multiple erythematous and eroded plaques in the inguinal and gluteal areas, crusted ulcerated lesions on the dorsal aspects of the hands and feet. Although treated with various topical agents under presumptive diagnoses of eczema and fungal infection, her condition failed to improve. A skin biopsy confirmed the diagnosis of JED. Concurrent wound culture revealed methicillin-resistant Staphylococcus aureus and treatment with systemic teicoplanin and topical mupirocin was initiated. At the age of ten, the patient underwent allogeneic hematopoietic stem cell transplantation (HSCT). All skin lesions completely regressed after HSCT. The patient is currently receiving regular IVIG and fludrocortisone therapy.

Conclusion:

Skin is the most commonly affected organ in DOCK8 deficiency. Cutaneous manifestations of DOCK8 deficiency include eczema, viral infections, and skin abscesses. Our patient is the first reported case of JED in a pediatric patient with DOCK 8 mutation.

¹Manisa Celal Bayar University, Dermatology and Venereology, Manisa, Türkiye

²Ege University, Pediatric Allergology and Immunology, İzmir, Türkiye

³Ege University, Pediatric Hematology, İzmir, Türkiye

⁴Manisa Celal Bayar University, Pathology, Manisa, Türkiye

⁵Ege University, Pediatrics, Division of Infectious Disease, İzmir, Türkiye

⁶Universitätsklinikum Schleswig-Holstein, Campus Kiel, Department of Dermatology, Venereology and Allergology, Kiel, Germany

Reports on the treatment of JED in the literature are scarce and inconsistent. There have been reports of patients with JED successfully treated with topical antibiotics, antifungals, and zinc oxide. In our case, minimal clinical healing was observed despite the use of topical and systemic antibiotics, and topical and systemic antifungals. On the other hand, currently, the only curative treatment for DOCK8 deficiency is HSCT. In our case, all lesions regressed after HSCT, and this raises the question of whether there may be a connection with immune deficiency in patients diagnosed with JED. Although the underlying factors have been reported as urinary and fecal incontinence, presence of detergent remnants in the diaper, infrequent diaper change, rough toilet paper usage, and chronic diarrhea, it is not known why this dermatitis occurs only in some individuals and not in others despite the presence of these factors. There is no history of urinary incontinence in our case, and the fact that she is 9 years old creates confusion about the underlying etiological factors, especially since this dermatitis is seen in diaper period. There is no report in the literature on the relationship between immunodeficiency and JED, and more studies are needed on this subject to understand whether there is a relationship between these two different conditions or whether they occur coincidentally.

Kaposiform haemangioendothelioma: a case report of sirolimus as monotherapy

Shihab Siddiquee*¹, Dev Tilakaratne¹, Catherine Boyd¹, Emma Ryan², Lachlan Warren²

¹Royal Darwin Hospital, Darwin, Australia

Introduction & Objectives:

A newborn female (born at term) was noted at birth to have a large indurated violaceous plaque over her right hip and inguinal region. We discuss the investigations, diagnosis and subsequent management of this rare presentation of a Kaposiform haemangioendothelioma.

Materials & Methods:

Magnetic resonance imaging showed a large infiltrative mass involving the skin, superficial and deep subcutaneous layer, and gluteal muscle, with multiple small supplying arteries and veins, with overall appearance consistent with a Kaposiform haemangioendothelioma (KHE). Histopathology of a skin punch biopsy showed a vasoformative lesion composed of scattered ectatic thin-walled vessels lined by endothelium, and nodules of tightly packed capillary-sized vessels lined by spindled to epithelioid endothelial cells. Immunohistochemistry showed the capillary nodules were positive for endothelial markers (CD31, CD34, and ERG) and negative for GLUT-1, and the overall features were compatible with tufted angioma/KHE spectrum. Multidisciplinary team input was obtained, and clinical correlation favoured a diagnosis of KHE. Blood tests in the first two months of life showed a progressively increasing and elevated D-dimer, without clinical evidence of Kasabach-Merritt phenomenon (KMP). Following relevant screening for infections and consultation with paediatric oncology, the patient was commenced on oral sirolimus at a total daily dose of 0.7 mg/m^2 (divided into two doses). Prophylactic oral trimethoprim/sulfamethoxazole was also commenced following consultation with the paediatric infectious diseases team, as the patient resided in a melioidosis-endemic region.

Results:

After commencing sirolimus the Kaposiform haemangioendothelioma plaque stabilised in size without ulcerating, and induration was palpably reduced. Sirolimus was well tolerated, with no significant side effects observed during follow-up by the dermatology and paediatric teams. Sirolimus blood levels were maintained within a therapeutic range (5-10 ng/ml).

Conclusion:

Kaposiform haemangioendothelioma is a rare, locally aggressive vascular tumour with a high mortality rate (up to 30%) reported in the setting of Kasabach-Merritt phenomenon, a consumptive coagulopathy which leads to severe thrombocytopaenia and a haemorrhagic diathesis. Sirolimus (an inhibitor of the mammalian target of rapamycin pathway) has previously been reported as a treatment for KHE and KMP, typically with concurrent systemic corticosteroids. This case highlights the role of dermatologists in recognising this uncommon and potentially life-threatening condition, the importance of multidisciplinary management, and positive outcome of sirolimus as monotherapy for Kaposiform haemangioendothelioma.

²Women's and Children's Hospital, Adelaide, Australia

Spontaneous remission of congenital aleukaemic leukemia revealed by «Blueberry Muffin Baby»

Garance De Pontbriand¹, Heloise Machut¹, Alaterre Camille¹, Duployez Nicolas², Roynard Pauline³, Guermouche Helene³, Brigitte Nelken⁴, Julian Thalhammer⁴, Olivier Philip¹, Catteau Benoit¹

Introduction & Objectives:

The aim of this case report is to highlight the difficulty of managing congenital aleukaemic leukemia cutis (CALC).

Materials & Methods:

We present a pediatric case exhibiting a congenital aleukaemic leukemia cutis with spontaneous remission.

Results:

A 1-month-old newborn presented few-days-after-birth with purplish nodules infiltrated over the back, abdomen, and right shoulder, without organomegaly. A "Blueberry-Muffin" syndrome was suggested. Skin biopsy revealed dendritic plasma cell infiltration with blastosis, in favor of neonatal acute myeloid leukaemia (AML). The blood count was normal and there were no circulating blasts. The bona marrow aspiration was normal; there was no sign of other extramedullary disease. No molecular abnormalities were identified in bone marrow or blood. Optical genome mapping (Bionano Genomics) revealed a t(5;6)(q35.1q23.3) on skin biopsy leading to a NPM1::CCDC28A fusion, previously described in AML. Therefore, the diagnosis was congenital aleukemic AML with strictly cutaneous expression. The therapeutic attitude consisted in active surveillance. The clinical course was favorable at 10 months, with almost complete regression. Monthly clinical and biological follow-up was uneventful.

Discussion:

Congenital leukaemia (<28 days of life) is extremely rare (~3cases/million-live-births) and the prognosis is poor (overall survival at 24 months: 23%). The biology of neonatal leukaemias is often different from later onset leukaemias and driven by fusion transcripts or trisomy 21. Moreover, AML is more common than lymphoid leukaemia. Its origin is intrauterine and therefore the disease is often advanced at presentation with hepatomegaly, splenomegaly, cutaneous infiltration, central nervous system infiltration and hyperleukocytosis. Spontaneous remission does occur in some infants (5% for AML), and is particularly associated with t(8;16); sometimes the remission is sustained, but careful monitoring is needed as relapse is frequent.

However, CALC should probably be considered differently. The prognosis seems better: among the small number of patients reported with CALC (15 patients), only three were treated immediately, with prolonged remission, five had prolonged spontaneous remission, six were found with acutisation of the leukemia (3 prolonged remission, 2 deaths, 1 unknown); 1 died of an other reason.

Conclusion

Here is another example of spontaneous remission (only 10-months-follow-up).

¹Dermatology department, Lille University Hospital, Lille, France

²Laboratory of Hematology, Lille University Hospital, Lille, France

³Institute of Medical Genetics, Jeanne de Flandre Hospital, Lille, France

⁴Department of Paediatric Haematology, Lille University Hospital, Lille, France

Acitretin in the Management of Pediatric Generalized Pustular Psoriasis: A Clinical Case

Wijdane tebbaai¹, layla bendaoud¹, ghita erramli¹, meriem aboudourib¹, said amal¹, ouafa hocar¹

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

Introduction & Objectives:

Generalized pustular psoriasis (GPP) is a rare, potentially life-threatening autoimmune inflammatory dermatosis. The pediatric form accounts for about 2%, with the generalized pustular type being even rarer at 7% of childhood psoriasis cases. We report a case of GPP in a child treated with acitretin.

Materials & Methods:

A 10-year-old child with a family history of psoriasis (father) presented with dry erythroderma and generalized non-follicular pustules persisting for one month, accompanied by scarlatiniform desquamation, inflammatory joint pain, and scaly scalp, without nail involvement.BSA >90%, GPPASI score of 10.8 (severe), and DLQI of 11.No drug intake was reported. Symptoms developed in a context of low-grade fever.A clinical diagnosis of GPP was made.Blood tests showed neutrophilic leukocytosis and an inflammatory syndrome. Pre-acitretin workup was normal.Treatment included topical corticosteroids, body cream, scalp lotion, and oral acitretin 10 mg every other day to improve adherence and reduce side effects.

Results:

Managing pediatric GPP remains challenging due to a lack of randomized controlled trials and standardized guidelines. Treatment should be tailored individually, weighing risks and benefits. Common first-line systemic treatments include oral acitretin, cyclosporine A, and methotrexate. Acitretin is the only systemic psoriasis treatment that is not immunosuppressive.** Its mechanism involves regulating cell proliferation, differentiation, apoptosis, and has immunomodulatory, anti-inflammatory, and anticancer properties. Numerous case reports show its rapid efficacy in severe pediatric psoriasis.** One case series reported 88% efficacy within three months of treatment. A Chinese report showed 96.5% efficacy with low-dose acitretin (0.3–0.4 mg/kg/day), with or without IL36RN mutation. A French multicenter retrospective study (June 2013–June 2014) of 154 children with psoriasis found acitretin as the most effective first-line agent in plaque and pustular psoriasis. It may also be suitable for infants. In a retrospective study of 174 pediatric patients, adverse effects occurred in 24%, most commonly skin irritation (10.3%) and lip dryness (9.2%). Biological abnormalities affected 22.4% (e.g., triglycerides 10%, alkaline phosphatase 5.9%), with only 4.1% requiring treatment discontinuation. Thus, acitretin appears safe in pediatric populations. In the absence of pediatric-specific monitoring guidelines, some authors recommend following adult protocols.

Conclusion:

Acitretin has a favorable safety and efficacy profile in children, especially given its non-immunosuppressive nature. Despite some differing scientific opinions, available data support its use. Further studies are needed to validate its role in pediatric care.

Severe and Refractory Bullous Pemphigoid in a Child: A Case Report

tinhinane benbrahim¹, sahel houria¹

¹faculté de medecine d'alger, bab el oued , Algeria

Introduction & Objectives:

Bullous pemphigoid (BP) is classically a disease of the elderly. Its occurrence in children is rare. With appropriate treatment, the clinical course is generally favorable. We report a case of severe and recalcitrant BP in a four-year-old girl.

Materials & Methods:

Case Presentation:

A four-year-old girl had been suffering from severe BP since the age of one, confirmed by histological and immunohistochemical studies. The condition was associated with marked and persistent eosinophilia (up to ten times the normal range). Two bone marrow aspirations yielded negative results.

The patient was treated with multiple therapies without clinical improvement: systemic corticosteroids at 1 mg/kg/day, dapsone at 2.5–5 mg/kg/day, cyclosporine at 2.5–5 mg/kg/day, and five courses of intravenous immunoglobulins (IVIg). She subsequently developed corticosteroid dependence.

Currently, she is being treated with a combination of mycophenolate mofetil, dapsone at 2.5 mg/kg/day, and prednisolone at 1.5 mg/kg/day. After two months of this regimen, the patient showed a marked clinical improvement: complete resolution of existing lesions, no new blister formation, and a gradual tapering of corticosteroids was initiated without signs of relapse. However, follow-up remains short to assess the long-term outcome.

Results:

Discussion:

BP is rare in the pediatric population and typically responds well to corticosteroids. Refractory cases are exceptional. Three recently published case series reported a total of nine pediatric patients successfully treated with topical or primarily systemic corticosteroids. Only two children required additional therapy with either dapsone or mycophenolate mofetil. Other authors have highlighted the potential benefit of treatments such as cyclosporine, azathioprine, and plasmapheresis.

In our case, the combination of systemic corticosteroids, dapsone, cyclosporine A, and IVIg proved ineffective. Clinical improvement was only achieved following the introduction of mycophenolate mofetil.

Conclusion:

Pediatric BP is a rare condition with poorly established therapeutic guidelines. We report a case of severe and refractory BP in a child, highlighting the need for individualized treatment approaches and further research in this population.

Effectiveness of Non-Pharmacological Interventions in Reducing Anxiety and Pain in Pediatric Dermatology Patients

Samiha Mohsen*1, 2, Megan Park1, 2, Cathryn Sibbald1, 2

¹University of Toronto, Toronto, Canada

²Hospital of Sickkids, Division of Pediatric Dermatology, Toronto, Canada

Introduction & Objectives: Dermatological procedures (including venipuncture, skin-prick testing, and burn care) and chronic dermatoses (atopic dermatitis, psoriasis) can be associated with significant pain and anxiety in children. While pharmacological interventions such as topical anesthetics are commonly used, they are not always sufficient or feasible. Non-pharmacological interventions (NPIs) offer promising adjunctive strategies to alleviate pain and anxiety in pediatric dermatology settings. We aimed to evaluate the effectiveness of non-pharmacological interventions in alleviating pain and anxiety among pediatric patients undergoing dermatological procedures or suffering from dermatological conditions. Secondary objectives were to assess safety.

Materials & Methods:

A systematic review was performed following PRISMA guidelines. We searched MEDLINE, Embase, and the Cochrane Central Register of Controlled Trials from database inception to March 26, 2025. Studies were included if they examined NPIs in children (0–18 years) undergoing dermatological procedures or living with chronic dermatologic conditions and measured outcomes related to pain and/or anxiety. Two reviewers independently screened studies and extracted data. Study quality was assessed using the Joanna Briggs Institute (JBI) critical appraisal tools. Due to heterogeneity in interventions and outcome measures, a narrative synthesis was produced. The certainty of evidence was assessed using the Grading of Recommendations, Assessments, Developments, and Evaluations framework.

Results:

Twenty-eight studies were included reporting on 1865 patients. Interventions included medical clowning, distraction techniques, hypnosis, virtual reality, music therapy, and mind-body therapies. Most studies reported some reduction in pain or anxiety compared to standard care. For needle-based procedures, music therapy was associated with a likely reduction in pain (moderate certainty of evidence). Medical clowning was also associated with decreased pain and anxiety (low certainty of evidence) (Table 1). For dermatology-specific procedures, such as cryotherapy and skin-prick testing, some interventions, including DistrACTION cards and video-based distraction, were associated with reductions in pain and anxiety (low certainty of evidence).

Conclusion:

Some non-pharmacological interventions may be effective in reducing procedural pain and anxiety in pediatric dermatologic settings. These strategies represent valuable adjuncts to standard care and should be integrated more broadly in pediatric dermatology practice. Further research could clarify optimal patient and intervention characteristics.

Table 1: GRADE assessment of the certainty of evidence for association between non-pharmacological interventions and procedures involving needles in pediatric patients

Total Participants			Certainty asses	ssment		Effect	5-4-1-4-	Immedia		
(N2 of studies)	Study design	Risk of bias	Inconsistency	Indirectness	Imprecision	Direction and Assessment	Certainty	Importance		
There is likely an association between music and reduced pain in pediatric patients undergoing procedures with needles										
212 (4)	observational	not serious	serious	not serious	not serious	Three out of four studies showed that music therapy reduce pain	⊕⊕⊕○ Moderate	CRITICAL		
There may be an association between medical clowning and reduced pain in pediatric patients undergoing procedures with needles										
133 (2)	observational	not serious	not serious	not serious	Very Serious	All studies reported that medical clowning reduced pain	⊕⊕○○ LOW	CRITICAL		
There is an uncertain association between distraction using a TV and reduced pain in pediatric patients undergoing procedures with needles										
29 (2)	observational	serious	not serious	not serious	Very Serious	All studies reported that TV distraction reduced pain	⊕○○○ Very LOW	CRITICAL		

Localized Pustular Psoriasis in Children: Clinical Characteristics, Diagnostic Challenges, and Treatment Strategies

Fadoua Chemsy¹, Fatimazahra Elfatoiki¹, Hanane Rachadi¹, Soumya Chiheb¹

¹ibn roched university hospital, dermatology and venerology, casablanca, Morocco

Introduction & Objectives:

Localized pustular psoriasis (LPP) in children is a rare and often unrecognized variant of psoriasis characterized by the presence of pustules overlying well-defined erythematous plaques. The condition not only poses diagnostic challenges due to overlapping features with other pustular dermatoses, but also raises concerns about optimal management and long-term prognosis in the pediatric population.

The aim of this presentation is to review and discuss the clinical features, differential diagnosis, and current therapeutic approaches for localized pustular psoriasis in children. Emphasis is placed on early recognition, accurate diagnosis, and individualized treatment planning to improve outcomes.

Case report:

A 13-year-old boy with no significant past medical history presented to the clinic with a localized dermatosis of the feet that had developed since the age of three years. The patient had been seen several times and had been treated with oral and topical antibiotics without improvement.

Clinical examination revealed a hyperkeratotic, erosive, and fissured erythematous plaque in one area, superinfected and crusted in another, located on the medial border of the right foot and opposite the left medial malleolus, with no cutaneous involvement of the rest of the integument, mucous membranes, or phanera.

The patient was started on antibiotics and underwent skin biopsy, which revealed psoriasiform hyperplasia with neutrophil microabscesses consistent with pustular psoriasis.

The patient was placed on dermocorticoid and reparative cream with marked improvement.

Conclusion:

Localized pustular psoriasis is a rare form of psoriasis and even more unusual in children. It is characterized by the presence of sterile pustules on limited areas of skin, most commonly the palms, soles, or around the nails. LPP is often misdiagnosed as eczema or infection, delaying treatment. Biopsy may be required to confirm the diagnosis of LPP by revealing the subcorneal pustules and neutrophilic infiltrate.

Treatment strategies range from topical agents, including corticosteroids and vitamin D analogues, to systemic therapies for refractory cases. Biologic therapies are emerging as promising options, with some case series documenting favorable outcomes even in younger patients.

Chronicity and psychosocial impact of psoriasis in children emphasize the need for a multidisciplinary approach.

Localized pustular psoriasis in children remains a diagnostic challenge due to its rarity and clinical overlap with other pediatric pustular dermatoses. Early diagnosis is critical to prevent chronicity and ensure appropriate treatment.

Three paediatric cases of primary cutaneous CD4+ small/ medium T-cell lymphoproliferative disorder.

Nerea Infante-Gonzalo¹, Lucero Noguera-Morel², Angela Hernandez-Martin², Isabel Colmenero-Blanco³, Mireia Segui-Olmedilla⁴, Jose Luis Rodriguez-Peralto⁵, Mar Llamas-Velasco⁴, Daniel Azorin-Cuadrillero³, Luis Blanco-Santana³, Ander Ezkurra-Altuna⁶, Francisco Arias⁷, Antonio Torrelo-Fernandez²

Introduction & Objectives:

Primary cutaneous CD4+ small/ medium T-cell lymphoproliferative disorder (PCSM-TCLPD) is an uncommon and heterogeneous clinical entity, mostly described in adults. A few cases are known in children, which appear as a solitary erythematous and asymptomatic lesion on the head, neck, trunk or upper limb. This entity is responsive to local treatment, the most common one being surgical excision, although spontaneous regression after biopsy has been described.

Materials & Methods:

We present three paediatric cases from a children's hospital, two boys aged 13 and 2, and a girl aged 7 years old.

Results:

The eldest boy bore a 6mm pinkish/ fleshy-coloured papule with teleangiectasias on the back, which had been incidentally discovered a month earlier and was excised, with two and a half years of follow-up.

The middle-aged girl exhibited a 15mm erythematoviolaceous nodule on the forehead, which had been evolving for three months and spontaneously regressed after a punch-biopsy, followed by a month of follow-up.

The youngest boy showed an erythematous nodule on the right forearm for two months, which also regressed after a punch-biopsy and has been followed-up for two and a half years.

None of them exhibited adenophaties nor accompanying symptoms.

Histological analyses showed images of a diffuse lymphocytic proliferation (with a nodular architecture being observable in the excision specimen) in the dermis and/or hypodermis made up of small or medium-sized cells with little or no atypia, accompanied by histocytes which in one cases included also touton-type giant cells.

IHC techniques highlighted an overwhelming CD3:CD20 (T/B proxy) ratio in all cases, with a variable majority between 65-90% in favour of CD4 over CD8, as well as inconstant loss of T markers, with no loss in one case, partial loss of CD7 in another and an almost complete one, also of CD7, in the third. CD30 was negative, as was EBER-ISH.

¹University Hospital Donostia, Dermatology, Donostia, Spain

²University Children's Hospital Niño Jesús, Dermatology, Madrid, Spain

³University Children's Hospital Niño Jesus, Pathology, Madrid, Spain

⁴University Hospital La Princesa, Dermatology, Madrid, Spain

⁵University Hospital 12 de Octubre, Pathology, Madrid, Spain

⁶University Hospital Donostia, Pathology, Donostia, Spain

⁷University Hospital Gregorio Marañon, Pathology, Madrid, Spain

Monoclonal rearrangements of TCR were confirmed in all cases.

All these findings were compatible with a PCSM-TCLPD. The patients underwent clinical follow-up without relapse nor other issues after 1 to 30 months.

Conclusion:

We present a series of three PCSM-TCLPD cases in paediatric patients. The average age was 7 years. All cases consisted of single lesions that appeared on forehead, back and forearm. Surgical excision was performed in the eldest of them. In the other two cases, spontaneous regression after biopsy was observed. The histological images were consistent with those described in the literature. No noteworthy events were recorded upon clinical follow-up.

PCSM-TCLPD ceased to be considered a lymphoma by the WHO in 2016 due to its excellent prognosis and its low rate of progression. Although most of the reported cases belong to adults, a few (very rare) cases in children are known. As in adults, the presentation is that of a solitary lesion, with an indolent evolution and an excellent prognosis. Staging is not required and clinical follow-up is enough.

Due to its uncommonness, increasing awareness of PCSM-TCLPD diagnoses is necessary in order to better classify this condition and create clinical guides for its handling. This entity should be known to avoid additional work-up and/or unnecessary invasive treatments.



Long term outcomes of pre-pubertal onset vulvar lichen sclerosus: a cohort study examining disease activity, treatment adherence and quality of life

Alexandra Savage¹

¹Price of Wales Hospital, Dermatology, Sydney, Australia

Introduction & Objectives: Vulvar lichen sclerosus (VLS) is a chronic inflammatory dermatosis affecting the vulvar and perianal skin. VLS has a bimodal age of onset, in pre-pubertal and post-menopausal age groups. Whilst in adults, VLS tends to run a chronic course, the long-term outcomes of pre-pubertal onset VLS is largely unknown. If left untreated or if inadequately treated, VLS can result in scarring and permanent deformity of the vulva, impaired sexual function and a 2 to 6% lifetime risk of malignant vulvar squamous neoplasia. This study aimed to establish the long-term disease activity, factors that influence disease progression, and the physical and psychological complications of pediatric VLS persisting into adulthood.

Materials & Methods: A cohort study conducted in a dermato-gynecology practice involving women who were diagnosed with VLS pre-menarche. A retrospective chart review of 135 case records was completed. Data on demographics, treatment protocols, follow-up duration, and clinical outcomes were extracted. The remaining data collection occurred prospectively through (i) study questionnaires, including quality of life as assessed by the Vulvar Quality of Life Index (VQLI), and (ii) in-person clinic appointments with a dermatologist. Treatment response was categorised as partial, complete, remission, or recurrence.

Results: One hundred and thirty-five cases were screened. After exclusions, retrospective data was collected for 68 cases of VLS. The mean age of diagnosis was 7.5 years and the mean duration of follow-up was 12.8 years. 62% of cases achieved remission. Of people who were adherent, 92.3% achieved complete resolution or remission, compared to 56.6% of people who were non- or partially adherent. Although treatment adherence was generally high following initial diagnosis, this reduced from menarche into early adolescence. People who were adherent with treatment were 60% less likely to have vulvar structural abnormalities than people who were not fully adherent. In addition, the VLQI was significantly lower (higher quality of life) among people who were adherent (mean = 1.6/45) compared to people who were partially or non-adherent (mean = 5.2/45).

Conclusion: This study informs clinicians of the long-term prognosis of VLS diagnosed pre-pubertally: 62% of cases achieved remission, and the rates of remission were higher in those who adhered to treatment. Structural changes/scarring were reduced in those who adhered to treatment. Treatment adherence is a critical determinant of remission, reduction in structural complications, and improved quality of life. Early recognition, comprehensive education, and treatment combined with sustained follow-up are essential to prevent long-term sequelae such as scarring, and to increase the chance of disease remission, in affected individuals.

 Table 3: Treatment response by treatment adherence and symptom duration prior to diagnosis

	Treatment Response	Negative (Partial or Recurrence)	Positive (Complete or <u>Remission)</u>	p-value
Adherence				
n (%)	Adherent	1 (7.7)	12 (92.3)	-
	Partial- or non-adherent	23 (43.4)	30 (56.6)	-
	Odds Ratio (95% CI) - crude Odds Ratio (95% CI) - adjusted	-	9.2 (1.1-76.0)	0.04
	for Initial PGA	-	8.7 (1.0-77.5)	0.05
Pre-Diagnosis Symptom Duration				
n (%)	<3 months	3 (18.7)	13 (81.3)	0.12
	>3-12 months	10 (47.6)	11 (52.4)	
	>12 months Unknown	9 (50.0) 3 (23.1)	9 (50.0) 10 (76.9)	

Familial Vitiligo Post-SARS-CoV-2: Insights into Viral-Induced Autoimmunity and Excimer Laser Efficacy

Ana-Maria Toma¹, Alice Brinzea², Roxana Nedelcu²

¹Elias Emergency University Hospital, Bucharest, Romania

²Carol Davila University of Medicine and Pharmacy, Pathophysiology, Bucharest, Romania

Introduction & Objectives:

Viral infections have been implicated in the pathogenesis of vitiligo, with evidence suggesting that viruses may trigger autoimmune responses that lead to melanocyte destruction. Chronic viral infections, such as those caused by Cytomegalovirus (CMV), Epstein-Barr virus (EBV), Herpes Zoster, HIV and more recently, COVID-19 have been associated with the onset or exacerbation of vitiligo. Vitiligo is known to impose a significant psychological burden on patients, and new therapies such as the 308-nm excimer laser have shown promise in promoting repigmentation and improving quality of life.

Materials & Methods:

We present the case of a 10-year-old male patient who developed well-circumscribed depigmented macules and patches of irregular shape, which progressively expanded over several months. The lesions were characterized by clearly defined borders, with normal surrounding skin and no associated erythema, scaling, or inflammation. The lesions initially appeared on the trunk, face, and limbs, following a recent mild SARS-CoV-2 infection. The patient had no history of toxic exposures or other infections correlating with the onset of vitiligo. Based on the typical clinical appearance of the lesions and confirmation with Wood's lamp examination, the diagnosis of vitiligo was established.

A notable aspect of this case is the development of vitiligo in the patient's father following the same SARS-CoV-2 infection, with similar but less extended lesions. Since the diagnosis, he has chosen not to pursue treatment.

Results:

Initial treatment included a course of topical corticosteroids, followed by long-term therapy with topical tacrolimus. However, lesions on the periocular area, knees, elbows, and hands, which remained refractory to these treatments, were subsequently treated with excimer laser therapy (Exciplex), following the typical regimen, which began with lower doses and was gradually increased to higher ones, adapted according to the patient's response and tolerability. This approach resulted in significant repigmentation and a favorable clinical outcome.

Conclusion:

This case highlights a potential association between SARS-CoV-2 infection and the onset of vitiligo, observed not only in the patient but also in his father, adding a new possible viral trigger for the condition in predisposed individuals. The patient responded positively to excimer laser therapy (Exciplex), a newer phototherapy that has proven effective for treating active vitiligo lesions. This is particularly important due to the necessity to provide a reliable treatment option, alleviating the psychological burden associated with vitiligo, especially in children. Early diagnosis and personalized treatment approaches are crucial for optimizing both clinical outcomes and the mental well-being of patients.

Acute hemorrhagic edema of infancy after bacillus Calmette-Guerin vaccination

Rajaa Bousmara*¹, Fatima-Zahra Elfatoiki¹, Fouzia Hali¹, Soumia Chiheb¹

¹UHC Ibn Rochd , Dermatology and Venereology , Casablanca, Morocco

Introduction & Objectives:

Acute hemorrhagic edema of infancy (AHEI) is a rare and benign form of leukocytoclastic vasculitis. Numerous precipitating factors, including drugs, infections, and vaccination have been reported. We report the case of a 3-week-old boy with AHEI following bacille Calmette–Guérin (BCG) vaccination. To the best of our knowledge, this is the second reported case after BCG vaccination.

Case report:

A 3-week-old male infant presented to our dermatological department with a 15-day history of progressive erythematous and indurated plaques with edema on the face and extremities. There was no previous history of infection or use of medications. However, the patient had been vaccinated with the BCG vaccine 3 days before presentation. Physical examination revealed purpuric and annular plaques on the face and extremities. The left eyelid was edematous with a large cockade and purpuric plaque. There was no mucus membrane involvement. The rest of the physical examination showed no systemic symptoms. Skin biopsy revealed leukocytoclastic vasculitis. Direct immunofluorescence was negative. Chest X-ray and abdominal ultrasonography were normal. Laboratory blood tests revealed neutrophils measuring 18000/mm3. Renal and liver function tests were normal. A diagnosis of AHEI was made. The patient's skin lesions spontaneously improved without receiving any treatment over 3 weeks, and no relapse was observed.

Conclusion:

Although vaccinations are rarely implicated in the occurrence of this disease, this diagnostic hypothesis should be raised following BCG vaccination. It is important to reassure parents about this post vaccination effect because it is benign and resolves spontaneously.

Idiopathic aseptic facial granuloma treated with ivermectin cream1% and tacrolimus 0,03%

Ana Estela Ribeiro¹

¹Santa casa de misericórdia de São Paulo, Dermatology, São Paulo, Brazil

Introduction & Objectives: A healthy 4-year-old girl had an inflammatory nodule in the left malar region for 5 months and recurrent hordeolum on the eyelids since the age of 2. On examination, some micropustules and follicular spicules were also noted around the nose and mouth. She came to the consultation accompanied by her father who had papulopustular rosacea and some areas of the face with pityriasis folliculorum. A standard superficial skin biopsy(SSSB) was performed which showed 97 *Demodex folliculorum* per cm2. (normal up to 5D/cm2).

After this result, a SSSB was collected from the malar region of the girl which showed 16D/cm2.

The hypothesis of infantile rosacea with aseptic granuloma was made. Initially, the patient was treated with oral ivermectin (3 mg - the child's weight was 16 kg) and sulfamethoxazole plus trimethoprim for 15 days. After thirty days, there was no improvement.

Then topical ivermectin cream 1% was prescribed to be applied on the entire face, including the eyelids once a day, at night (the father was an ophthalmologist consented) for 90 days, and tacrolimus 0.03% on the granuloma once a day, at morning. After this period, there was a very significant improvement in the lesions. The patient continued using ivermectin 1% cream every another day for 3 months.

The granuloma and hordeolum healed without scars and the patient was followed for more 2 years, and there were no recurrences of either the hordeolum or the granuloma.

Idiopathic facial aseptic granuloma is one of the lesions that can appear in infantile rosacea, as well as hordeolum. In this patient, the presence of pityriasis folliculorum led to the search for Demodex. In adults, rosacea has been associated with the presence of Demodex, but not in children.

Infantile rosacea with aseptic granuloma and hordeolum may be associated with *Demodex folliculorum* and responded to treatment with topical ivermectin and topical tacrolimus.

Materials & Methods:

Results:

Conclusion: Infantile rosacea with aseptic granuloma and hordeolum may be associated with *Demodex folliculorum* can be treated with topical ivermectin and topical tacrolimus.



Macular Lymphocytic Arteritis: The Diagnostic Challenge of an Isolated Pediatric Vasculitis

Nouha Touhami¹, Kenza Benothmane¹, Nadia Ismaili¹, Laila Benzekri¹

¹Ibn Sina Hospital, Dermatology, Rabat, Morocco

Introduction & Objectives Macular lymphocytic arteritis (MLA) is a rare cutaneous vasculitis, newly recognized as a distinct entity. It primarily affects small dermal vessels with a lymphocytic infiltrate, without systemic manifestations. Predominantly observed in North African women, it mimics cutaneous polyarteritis nodosa (PAN), differing significantly in prognosis and treatment. We report a case of MLA in a North African adolescent to highlight its distinctive features and the importance of accurate diagnosis.

Materials & Methods A 13-year-old North African girl, with no notable medical history, presented with a painless rash on all four limbs, slowly progressing over two years. Examination revealed non-infiltrated, erythematous macules with a branching pattern on the forearms and lower limbs, without systemic symptoms. Histological analysis of a skin lesion showed a dense interstitial and perivascular lymphocytic infiltrate, focally damaging vessel walls. Fibrin deposits were present without fibrinoid necrosis or thrombosis. Laboratory tests showed no systemic inflammation; immunologic tests, cryoglobulinemia, and hepatitis B/C serologies were negative. Imaging (chest X-ray, abdominal ultrasound) showed no systemic involvement. MLA was diagnosed.

Results This case is unusual, as MLA is typically reported in middle-aged women. The clinical presentation was strictly cutaneous, with non-infiltrated, erythematous macules in a branching pattern, painless and lacking systemic signs—unlike cutaneous PAN, which is often painful, inflammatory, and potentially necrotic. Histology revealed a dense lymphocytic infiltrate damaging vessel walls, without fibrinoid necrosis or neutrophils, consistent with MLA and distinct from PAN, which features segmental necrosis and mixed-cell infiltrates. Paraclinical findings supported the diagnosis: absence of systemic inflammation, negative immunology and viral serologies, and no systemic involvement on imaging. These findings confirm the cutaneous nature of MLA. This case stresses the importance of recognizing MLA to avoid misdiagnosis and inappropriate immunosuppressive therapy. Though chronic, its course appears benign. Given the unclear natural history, regular follow-up is recommended, especially in young patients.

Conclusion MLA is a rare, likely underdiagnosed vasculitis, particularly in North African females. Its chronic, indolent, and cutaneous course, coupled with distinct histological features, allows differentiation from PAN. Accurate diagnosis through clinical, histopathological, and multidisciplinary evaluation is essential to avoid unnecessary treatment and ensure proper follow-up.

Cutis Marmorata Telangiectatica Congenita Associated with Infantile Myofibromatosis: A Case Report

roumili raouia¹, kholoud afeissa¹, mira chikhaoui¹, Houria Sahel¹

¹Bab El Oued University Hospital, bab el ouad, Algeria

Introduction & Objectives:

Infantile myofibromatosis is a rare, benign soft tissue tumor characterized by the development of nodules involving the skin, striated muscle, bone, and, in exceptional cases, visceral organs. It presents with a wide spectrum of clinical manifestations depending on the extent and location of involvement.

Cutis marmorata telangiectatica congenita (CMTC) is an uncommon congenital vascular malformation, typically presenting with livedo and telangiectasias in a diffuse or segmental distribution.

We report a rare case of co-occurrence of these two distinct conditions.

Materials & Methods:

A 14-month-old girl, with no significant medical history, was initially diagnosed with congenital cutis marmorata telangiectatica congenita (CMTC).

At the age of one month, she developed a nodule on the left thigh. Physical examination revealed a reticular livedo with fine, regular meshes, partially blanching with pressure, non-infiltrated, involving both lower limbs. A firm, immobile 1.5 cm nodule with an erythematous surface was observed on the left thigh.

Hemihypertrophy of the left thigh was noted, along with muscle atrophy of both legs.

Doppler ultrasound of the lower limbs showed an indurated, echogenic appearance of the soft tissues, with solid oval structures embedded within the muscles of the left leg.

Muscle MRI revealed multiple subcutaneous and intramuscular nodular lesions with similar imaging characteristics.

Biopsy of one of the nodules, followed by histopathological examination, confirmed the diagnosis of infantile myofibroma.

Based on the clinical presentation, imaging findings, and biopsy results, a diagnosis of infantile myofibromatosis was established. A staging workup was performed and showed no evidence of visceral involvement.

Results:

We report a rare case of infantile myofibromatosis associated with cutis marmorata telangiectatica congenita (CMTC). This association has been described only once in the literature, in a child presenting with multiple nodules, CMTC, and hemiatrophy. In that report, the authors suggested that the coexistence of these conditions may be coincidental.

The pathophysiological link between these two entities remains unclear. Is this simply a fortuitous association, or could there be an underlying common mechanism? Further cases and studies are needed to explore this potential relationship.

Conclusion:

We report the second case in the literature of an association between cutis marmorata telangiectatica congenita and infantile myofibromatosis. This rare coexistence raises the question of a potential pathophysiological link, which remains to be elucidated.

Annular lipoatrophy of the ankles: A case report

Widad Sellami¹, Saliha Takheroubt¹, Issam TABLIT¹, Samira Zobiri¹

¹Dermatology Clinic, Mustapha University Hospital, Algiers, Algiers

Introduction & Objectives:

Annular lipoatrophy of the ankles is an acquired lipoatrophic panniculitis, primarily affecting children, a rare condition where only about twenty cases have been described in the literature. We report a new case, whose interest is to make this entity known and to be able to suggest early diagnosis in order to improve its management.

Materials & Methods:

A 6-year-old girl, with no particular medical history, consulted for distal circumferential atrophy of the legs and ankles evolving for six months. The evolution began with a unilateral inflammatory plaque on the right ankle, rapidly becoming bilateral, with ascending extension. After three months, the inflammatory lesions evolved into progressive cicatricial atrophy. Dermatological examination showed non-sclerosing subcutaneous atrophy forming a circumferential band with an erythematous upper limit of the lower third of the legs and ankles in a bilateral and symmetrical manner, and was asymptomatic. Neuromuscular examination was without abnormality. An autoimmunity assessment

showed elevated TSH and anti-thyroid antibodies, confirming Hashimoto's thyroiditis. Histology showed nonspecific lobular panniculitis with lipophagy, without signs of vasculitis. Treatment with corticosteroid therapy 0.5 mg/kg/day for six months allowed stabilization of the lesions, without the appearance of new involvement with a one-year follow-up.

Results:

Annular lipoatrophy of the ankles (ALA) is a rare entity of poorly known etiology, first described in 1970, with only 24 cases reported in the literature, most of them in children and mainly in girls. It initially manifests as bilateral erythematous or nodular inflammatory lesions at the ankles, evolving into ascending annular atrophy, sometimes up to the knees. Arthralgias and myalgias may accompany this evolution, without muscular deficit. Imaging shows inflammatory edema in the initial phase, followed by subcutaneous atrophy. The frequent association with autoimmune diseases, particularly dysthyroidism, as in our patient, suggests a dysimmune origin. Diagnosis is based on clinical presentation, histology not being essential. Treatment remains disappointing, with no evidence of efficacy on the prevention of atrophy, although early management may limit sequelae.

Conclusion:

We report an additional case of ALA, highlighting its similarities to the characteristics reported in the literature. ALA has a stereotyped presentation that should be known to clinicians for early and appropriate management.

Treatment of Pediatric Alopecia Areata with Dupilumab

Cecilia Iborra García-Trevijano¹, marta pérez¹, Jose Fuentes Ros¹, Pablo Díaz-Calvillo¹, MERCEDES PICO VALIMAÑA¹

¹Hospital Universitario Puerto Real, Cádiz, Spain

Introduction & Objectives:

Alopecia areata (AA) is the third most common dermatologic condition in pediatric consultations. It is estimated that 20% of patients develop symptoms during childhood. However, treatment options are limited after failure of conventional therapies such as corticosteroids or contact immunotherapy. This poses a therapeutic challenge, highlighting the need to explore alternative approaches and share clinical outcomes. The objective of this study is to describe the clinical experience and therapeutic response to dupilumab in three pediatric patients with refractory alopecia areata.

Materials & Methods:

We present the experience of a Spanish hospital in treating AA in three pediatric patients. Data on demographic, clinical, laboratory, therapeutic and outcome variables were collected from medical records and analyzed.

Results:

- Patient 1: Male with atopic dermatitis, developmental delay, and microcephaly. Experienced progressive hair loss since age one, evolving to total alopecia, including eyebrows, by age three. After a confirmatory biopsy and failure of multiple treatments (topical corticosteroids, oral corticosteroids, topical minoxidil, methotrexate), dupilumab was initiated at age six (300 mg/month), resulting in a reduction of the SALT score to 20% within six months.
- Patient 2: Female with alopecia universalis since age four. Started dupilumab at age six (300 mg/month) after no response to corticosteroids, minoxidil and methotrexate. Achieved complete hair regrowth after two years of treatment.
- Patient 3: Male with alopecia universalis since age five, ventriculomegaly, and mutism. Biopsy confirmed AA and thyroid and autoimmunity panels were unremarkable. He received unsuccessful treatment with topical corticosteroids, oral corticosteroids, topical minoxidil and methotrexate before starting dupilumab at age ten (300 mg/month), which was discontinued after 14 months due to lack of response.

Conclusion:

While new treatments like baricitinib (JAK1/JAK2 inhibitor) for adults and ritlecitinib (JAK3/TEC inhibitor) for patients over 12 have recently been approved, no targeted therapies are currently available for younger pediatric patients. Although AA is mainly driven by a Th1-mediated immune response, its association with IL-4 and IL-13 gene polymorphisms, as well as its frequent overlap with atopic dermatitis, has prompted the off-label consideration of dupilumab as a potential treatment option. However, literature reports both improvement and worsening of AA with this drug, preventing generalized recommendations in the absence of concomitant atopic dermatitis. In our experience, we observed an improvement of 80%, according to the SALT score, was observed in a child with atopic dermatitis, a complete response in a non-atopic female patient, and no response in a non-atopic male patient; suggesting that dupilumab may be beneficial in selected pediatric cases, regardless of atopic status. Further study is needed, and more detailed case reports may help identify potential responder profiles.

Aggressive and Recurrent Cellular Dermatofibroma

Olivier Philip¹, Stéphanie MALLET¹, Julien Antonietti¹, Nausicaa Malissen¹, Nathalie Degardin², Olympe Gaudray¹, Philippe Petit³, Audrey Maryse Aschero³, Angélique Rome⁴, Nicolas André⁴, Franck Tirode⁵, Richard Marie-Aleth¹, Nicolas Macagno⁶

Introduction & Objectives:

Dermatofibroma is a common cutaneous soft tissue tumor. The aim of this case report is to highlight the potential for local aggressiveness in atypical dermatofibroma.

Materials & Methods:

We present a pediatric case exhibiting local aggressiveness, for which whole-exome RNA-sequencing was performed.

Results:

An 8-year-old child with no prior medical history presented with a 6-month history of a pigmented, indurated, and tender nodule on the shoulder, measuring approximately 1 cm. Histopathological analysis confirmed the diagnosis of cellular dermatofibroma. One year later, the tumor recurred locally, accompanied by two satellite nodules. A wide excision was performed but revealed positive margins, necessitating an additional procedure with skin graft reconstruction. Clinical and radiological follow-up, including MRI at one year, showed no evidence of recurrence.

Discussion:

Dermatofibroma is typically diagnosed clinically. Most cases are benign, with recurrence rates below 1% following complete excision. However, lesions larger than 1 cm and incomplete excision are recognized risk factors for recurrence.

When the clinical diagnosis is uncertain, histopathological examination can easily confirm it and identify subtypes associated with higher recurrence rates. Histology also confirmed the absence of features indicative of high-grade malignancy, such as significant mitotic activity, necrosis, or pleomorphism. Three rare histological variants—cellular, aneurysmal, and pleomorphic—are associated with higher recurrence rates (10–20%). Our case was classified as an aneurysmal and cellular dermatofibroma. Although rare, malignant transformation has been reported, with cases of both local and metastatic recurrence.

Further molecular analysis using whole-exome RNA sequencing confirmed the presence of a PRKCG fusion and transcriptomic signatures associated with favorable prognosis in mesenchymal tumors, including low CINSARC and G2/M scores. The presence of a PRKCG fusion gene, as found in our patient, supports the clonal nature of dermatofibromas, challenging the previous assumption that they result from a reactive process.

¹Timone Enfants Hospital and Aix Marseille University, Dermatology, Marseille, France

²Timone Enfants Hospital and Aix Marseille University, Pediatric Plastic surgery, Marseille, France

³Timone Enfants Hospital and Aix Marseille University, Pediatric radiology, Marseille, France

⁴Timone Enfants Hospital and Aix Marseille University, Pediatric Hematology and Oncology, Marseille, France

⁵Center Léon Bérard, Lyon, France

⁶Timone Enfants Hospital and Aix Marseille University, Pathology, Marseille, France

Conclusion:

More studies are needed to identify clinical, histopathological or molecular predictive markers associated with this increased risk of recurrence in dermatofibroma, notably in the pediatric population.

The Diagnostic Triad: Clinical, Dermoscopic, and Histopathological Insights into rare paediatric Telangiectasia Macularis Eruptiva Perstans

Keshav Yadav*1, prakhar srivastava1, niti khunger1

¹Safdarjung Hospital, New Delhi, India

The Diagnostic Triad: Clinical, Dermoscopic, and Histopathological Insights into rare paediatric Telangiectasia Macularis Eruptiva Perstans

Introduction & Objectives:

Telangiectasia Macularis Eruptiva Perstans (TMEP) is a dermatological entity with unique clinical and histopathological features. It is a rare variant of cutaneous mastocytosis, is predominantly seen in adults and is characterized by erythematous-brown macules with telangiectasias. 1 Diagnosing them can sometimes be challenging due to unusual presentations. Dermoscopy can be helpful in these cases, and the diagnosis can later be confirmed by histopathology.

Materials & Methods: Not applicable

Results: Case Synopsis:

A 9-year-old boy presented with a one-year history of persistent, asymptomatic, reddish-brown lesions predominantly on the lower limbs. The reddish-brown lesions were initially insidiously observed on the left upper thigh, gradually increasing in size, number, and distribution to involve both legs. There was no pruritus, urticaria, or other systemic symptoms.

On examination, the lesions on the lower extremities consisted of multiple well to ill-defined erythematous-brown macules, 0.5–3 cm in diameter, with subtle telangiectasias. These macules were non-blanchable, and the Darier sign was negative. Mucocutaneous and systemic examinations were unremarkable. Dermoscopy of the lower limb lesions revealed a homogeneous erythematous-brown background with prominent dotted, linear, and branching vascular patterns. Irregular, tortuous vessels with perivascular brownish pigmentation were also noted.**
Histopathological examination of the leg lesions revealed perivascular mononuclear inflammatory infiltrate in the papillary dermis with telangiectatic capillaries. The presence of numerous mast cells was confirmed by Giemsa staining and CD117 (c-KIT) immunohistochemistry (IHC). CD68 staining identified macrophages in the papillary dermis. Clinically other differentials like Necrobiosis lipoidica, cutaneous histiocytosis, lichen aureus were also kept, but were ruled out on histopathology and these findings confirmed the diagnosis of TMEP. He was started on topical mid-potent corticosteroid and oral second generation antihistamine for management.

Conclusion:

In conclusion, this case of TMEP in a 9-year-old boy with predominant involvement of the lower limbs underscores the importance of considering rare dermatological entities even in atypical age groups and locations. Key learning points from this case include the unusual presentation of TMEP in a child, a condition typically seen in adults, and its predominant localization on the lower limbs, contrasting with the more common trunk and upper extremity involvement. Dermoscopy proved to be a valuable tool, revealing characteristic vascular patterns that aided in narrowing the differential diagnosis. However, the definitive diagnosis relied on histopathological examination, which confirmed the presence of increased mast cells through Giemsa and CD117 staining. Therefore, meticulous

clinical evaluation, coupled with the judicious use of dermoscopy and histopathology, can aid in accurate diagnosis, especially in cases with unusual presentations of rare conditions like TMEP.

Not Just Another Eczema: A Rare Pediatric Case of Sulzberger-Garbe Dermatosis

Przemysław Hałubiec*¹, Magdalena Jaworek², Anna Wojas-Pelc³, Andrzej Jaworek³

¹Jagiellonian Univeristy Medical College, Chair of Dermatology, Doctoral School of Medical and Health Sciences, Cracow, Poland

²Jagiellonian Univeristy Medical College, Institute of Paediatrics, Chair in Paediatric Neurology, Cracow, Poland ³Jagiellonian Univeristy Medical College, Chair of Dermatology, Cracow, Poland

Introduction & Objectives:

Sulzberger-Garbe dermatosis is a rare, chronic skin condition originally described by Marion Sulzberger and William Garbe in 1937, typically affecting middle-aged men. Characteristic features include discoid, exudative lesions with lichenification, primarily involving the trunk, extremities, and genital area, that are accompanied by severe pruritus. Although it may occur in patients regardless of age or sex, pediatric cases are exceedingly rare and often misdiagnosed as atopic dermatitis or nummular eczema.

We aim to present the diagnostically challenging case of early-onset Sulzberger-Garbe dermatosis.

Materials & Methods:

A 14-year-old boy with a long-standing history of seasonal, recurrent eczematous eruptions since infancy was referred to our dermatology clinic due to progression of skin lesions and intense pruritus. There was no personal or family history of atopy. Previous treatments with low-to-medium potency topical glucocorticosteroids (with or without topical antibiotics) failed to adequately control the condition.

Physical examination revealed erythematous, infiltrated plaques with erosions and post-inflammatory dyspigmentation, predominantly on the extensor surfaces of the limbs. The trunk and face were unaffected.

Laboratory testing revealed mild relative eosinophilia (5.6%) in the complete blood count. Other routine tests (liver enzymes, creatinine, fasting glucose, C-reactive protein) were within normal limits. Patch testing, prick testing, total and specific IgE testing were negative.

Histopathological examination of a biopsy specimen from the upper limb revealed features of chronic eczema corresponding to Sulzberger–Garbe dermatosis.

Results:

Based on the chronic course and clinical morphology, unresponsiveness to previous treatment, negative allergic and autoimmune workup and supporting histological presentation, a diagnosis of Sulzberger-Garbe dermatosis was established.

The patient was treated with potent topical corticosteroids (daily 0.05% betamethasone dipropionate, later switched to 0.05% clobetasol propionate in weekend therapy) and oral antihistamines (10 mg rupatadine 1 x daily). Additionally patient was educated on the proper use of moisturizers and prescribed with cholesterol ointment.

Only a partial remission was achieved using topical therapy and recurrent flares with vesicles, crusts, and oozing were observed, particularly in winter. In view of insufficient control, patient was scheduled for phototherapy.

Conclusion:

This case highlights a rare pediatric manifestation of Sulzberger–Garbe dermatosis. Although classically described in adult males, the condition should also be considered in adolescents with chronic, pruritic dermatoses unresponsive to conventional treatment. Comprehensive differential diagnosis – including allergy screening and histopathologic evaluation – is essential to exclude other dermatoses that share similar clinical features.

Menkes Syndrome: Status epilepticus diagnosed by a hair. Report of two cases.

jose carrasco muñoz¹, Irene Albert Cobo¹, Noelia Jara Rico¹, Marina Senent Valero¹, Laura Berbegal de Gracia¹

¹Hospital General Universitario Dr. Balmis, Department of Dermatology, Alicante, Spain

Introduction:

Pili torti is a congenital hair shaft anomaly characterized by fragile hair that twists at irregular intervals. It may occur in isolation or in association with other syndromes such as Menkes or Bazex. We report two cases from our hospital.

Clinical case:

Our cases are two male infants aged 3 and 4 months old. Both were admitted to the Intensive Care Unit due to epileptic seizures with poor response to antiepileptic treatment. A Dermatology consultation was requested for diagnostic and therapeutic support in light of the complementary test results: status epilepticus on electroencephalogram, persistent hyperlactacidemia on blood gas analysis, and decreased serum copper and ceruloplasmin levels. In one of the patients, cerebral MRI revealed tortuosity of the circle of Willis. Dermatological examination showed light-colored, sparse, and brittle hair, along with skin pallor in both patients, and microcephaly in one of them. Hair samples were taken for microscopic analysis, revealing *pili torti*, hair shaft fractures, and hair shaft nodes. Based on these findings, a presumptive diagnosis of Menkes syndrome or trichopoliodystrophy was supported. Genetic testing was requested, and subcutaneous copper-histidine treatment was initiated. Subsequently, in a few weeks both patients showed improvement in their status epilepticus and were discharged for outpatient follow-up.

In both cases, genetic testing confirmed pathogenic hemizygous variants in the ATP7A gene. Both patients continue treatment with copper-histidine.

Discussion:

Menkes syndrome is a neurodegenerative, X-linked recessive disorder caused by mutations in the ATP7A gene, which regulates copper metabolism. There is a decrease in copper and ceruloplasmin levels in plasma and in organs such as the brain, liver, bone, hair, and skin. It should be suspected in patients around 2–3 months of age who present with psychomotor delay, seizures, and brittle hair, among other findings. Trichoscopic examination typically reveals *pili torti, monilethrix*, and *trichorrhexis nodosa*. Blood tests show decreased copper and ceruloplasmin levels. Definitive diagnosis is established through genetic testing. Treatment is administered via subcutaneous copper-histidine, as oral supplementation is ineffective. However, neurological deterioration leads to death in most cases within the first three years of life.

Conclusion:

Dermatologists should be familiar with this condition, as early diagnosis of Menkes syndrome is essential, since treatment with copper-histidine in the early stages may improve neurological outcomes.

congenital lymphedema of milroy: an uncommon clinical observation

Imane Hakim¹, Bendaoud Layla¹, Mariem Aboudourib¹, Amal Said¹, Hocar Ouafa¹

¹Mohammed VI University Hospital, Dermatology Department, Biosciences Research Laboratory, FMPM, Cadi Ayyad University, Marrakech, Morocco

Introduction & Objectives:

Lymphedema is defined as an accumulation of lymphatic fluid in the interstitial spaces. It affects the lower limb electively. Two forms are distinguished: primary and secondary. Milroy's disease is a primary congenital lymphedema, hereditary, rare, autosomal dominant with partial penetrance.

The aim of our work is to draw attention to an extremely rare clinical presentation that is little known to practitioners.

Observation:

The patient was 10 years old, with a history of bilateral swelling of the feet since birth. Dermatological examination revealed two large, edematous feet that were painful to palpation, with bilateral papillomatosis over the 4th toe, associated with diffuse intertrigo between the toes. As part of the paraclinical workup, a karyotype and lymphosintigraphy were requested but not performed, due to insufficient resources. The rest of the polymalformative work-up, notably the chest X-ray, cardiac ultrasound and abdominal ultrasound, were without anomalies. Treatment consisted of physiotherapy, bandaging, compression stockings and local antimycotic therapy, with long-term follow-up. The evolution was favorable, with healing of the intertrigo inter-orteil, improved function of both limbs and reduced pain, as well as a slight reduction in the size of both feet.

Discussion:

Milroy disease is an autosomal dominant genetic disorder with partial penetrance and an incidence of less than 1 per 100,000 births. In our patient, the lymphedema present at birth testifies to its hereditary nature. In their study, Baulieu et al. noted that 4 children had a family history. This was not the case with our patient. However, the literature reports sporadic cases linked to de novo genetic mutations. Limb walking function was preserved, as in the studies by Carver et al. Kitsiou-Treli. Whereas, Baulieu noted a relative functional impotence in the adolescent group. This may be explained by the young age of our child, and it is to be feared that this may occur over the years. We found a diffuse inter-torteil intertrigo. Infectious complications described in the literature included erysipelas, pleuropneumopathy, digestive infections and myocarditis, hence the need for long-term follow-up. The diagnosis was made clinically. Lymphography was not performed. Molecular biology, however, was useful for diagnosis, as it looked for a mutation in the gene coding for vascular endothelial growth factor (VEGF), inducing hypoplasia or aplasia of the lymphatic vessels. Treatment was symptomatic. The literature reports the value of local topicals and macrolides in the treatment of cutaneous complications. The treatment of lymphedema with manual lymphatic drainage and multilayer bandaging (followed by elastic compression with compression stockings) is described in the literature. Our patient's evolution was favorable, marked by a reduction and stabilization of the edema. Baulieu, Carver, Smeltzer et al. also reported a stabilization or even complete regression of edema with age. However, it may reappear after trauma or pregnancy in girls.

Conclusion:

Milroy disease is an autosomal dominant inherited disorder with partial penetrance in most cases. This case illustrates not only the rarity of the disease, but also the diagnostic difficulties associated with the inadequacy of expensive paraclinical investigations in a developing country. Treatment is symptomatic, requiring regular follow-up throughout life to prevent complications.

Seborrheic Pemphigus in an Adolescent: A Rare Case Successfully Managed with Dapsone

Yasmine Farai¹, Mohamed Amine Ennaciri¹, Ilyass Anouar¹, Mohamed El Amraoui¹, Youssef Zemmez¹, Rachid FRIKH¹, Hjira Naoufal¹

¹Mohammed V Military Teaching Hospital, dermatology, rabat, Morocco

Introduction & Objectives: Seborrheic pemphigus (SP) is a rare autoimmune blistering disease, usually seen in adults, and often mistaken for seborrheic dermatitis. It is exceptionally rare in children, with only a few reported cases. Diagnosis is challenging, and treatment, particularly in adolescents, requires balancing efficacy with the safety of immunosuppressive therapies. We report the case of a 15-year-old girl successfully managed with dapsone.

Materials & Methods: The patient had a 6-month history of pruritic bullous lesions, initially localized to the auricular region, which evolved into erosions and extended to the trunk and back, sparing the face. Scalp involvement appeared later. Despite extensive cutaneous involvement, her general condition remained stable, with no systemic manifestations. Histological and immunofluorescence findings revealed intraepidermal IgG and C3 deposits, consistent with SP. The patient was started on oral dapsone at 50 mg/day, later increased to 75 mg/day. After one month, significant clinical improvement was observed, with reduced lesions and inflammation, and no adverse effects.

Results: Pemphigus in children is rare and often difficult to diagnose due to atypical presentations. Systemic corticosteroids are typically used but may cause long-term side effects like growth retardation. To reduce these risks, steroid-sparing agents such as dapsone are considered. Dapsone has shown efficacy in autoimmune blistering diseases, including pediatric pemphigus vulgaris and foliaceus. Reports suggest it can lead to rapid improvement and allow corticosteroid reduction. Although responses may vary, dapsone is generally well tolerated, with a favorable safety profile. Monitoring for hematologic toxicity, particularly in G6PD-deficient patients, is essential. In our case, dapsone led to clear improvement without adverse effects, consistent with existing reports.

Conclusion: This case underscores the diagnostic challenges of SP in adolescents and supports dapsone as an effective, steroid-sparing treatment for mild to moderate disease. Early diagnosis and management are essential to improving outcomes.

Case series: Effectiveness and safety of dupilumab for liver transplant recipients in paediatric patients with eczema

Angela Tewari¹, Emilia Duarte Williamson¹, Shafiah Muna Abdul Gafoor¹

¹King's College Hospital, denmark hill, United Kingdom

Introduction & Objectives:

An eruptive eczematous eruption after oral tacrolimus use has been described in the literature in adults (1). Histology shows eczema (with lymphocyte and eosinophils) and thought to be secondary to an immune switch due to selective Th1 blockage (2). This type of eczema is highly symptomatic and patch testing to topical tacrolimus has been reported to be negative.

Materials & Methods:

We report a case series of 4 patients (4-13years, mean 8years) with a background of liver transplantation, treated successfully with Dupilumab.

Results:

Case 1: A 4-year-old boy had a partial matched liver transplant at age 3 months for unexplained acute liver failure and began oral tacrolimus. 9 months later he was troubled by an extremely itchy scaly eruption on the torso, abdomen and hands which did not respond to super-potent topical steroids. Skin biopsy showed spongiosis only. The EASI score was 31, his IgE was 26000kU/L (ref 0-51.9). Dupilumab was initiated at age 3 and trialled slowly with regular LFTs. 6 months later the EASI had markedly improved to 10, the quality of life markers had improved dramatically and a reduced IgE at 264kU/L.

Case 2: A 7-year-old girl with severe hand and foot eczema, refractory to topical steroids. She had a maternal partial liver transplant secondary to biliary atresia and portal vein thrombosis at 1 year. Oral tacrolimus was started at the time of transplant. 5 years later with extremely symptomatic eczema, Dupilumab treatment was initiated with an improvement in the itch within 4 months.

Case 3: A 13-year-old boy with a liver transplantation secondary to giant cell hepatitis. He was immunosuppressed with mycophenolate mofetil and oral tacrolimus from age 2. Dupilumab was initiated and he has been on treatment for the past two years with no reported side effects. His EASI score has improved from 24 to 5.

Case 4: 7-year-old boy of skin type 6 with a liver transplant age 1 year for biliary atresia and secondary portal hypertension and a history of ITP needing IVIG and rituximab. The patient has been on oral tacrolimus. 2-3 years later he began developing perianal eczema which then progressed to a more typical presentation affecting the popliteal and antecubital fossae and multiple episodes of infections and 5 years later he began dupilumab (EASI 34). 9 months on his EASI is 5 and he is much happier.

Conclusion:

Our case series highlights that in line with other case series, Dupilumab is a safe and effective treatment. Our cohort of paediatric patients with severe eczema have had liver transplantations and thus have had no live vaccines throughout childhood. We report no side effect sequelae at present.

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Carbamazepine-Induced Erythema Multiforme Major in a Child: A rare case report

Fadoua Chemsy¹, Fatimazahra Elfatoiki¹, Hanane Rachadi¹, Soumiya Chiheb¹

¹ibn roched university hospital, dermatology and venerology, casablanca, Morocco

Introduction & Objectives:

Erythema multiforme major (EMM) is a rare but potentially serious condition affecting both the skin and mucous membranes. It typically presents with characteristic 'target' lesions and can cause significant discomfort, particularly in children. While infections particularly herpes simplex are the usual culprits, medications such as antiepileptic drugs, including carbamazepine, can also be responsible.

In this report, we present the case of a child who developed EMM after starting carbamazepine. Our aim is to raise awareness of this rare but important drug reaction, to emphasise the importance of early recognition and the urgency of stopping the offending drug to prevent life-threatening complications.

Case report:

The patient was 14 years old and was being treated for epilepsy and behavioural problems with mental retardation following febrile meningitis at the age of 5 years. She was admitted because of a febrile generalised rash that appeared 12 days after taking carbamazepine, without digestive, respiratory, muscular or joint signs, and without any suggestion of photosensitivity, Raynaud's phenomenon or hair loss.

On clinical examination, the patient was conscious, restless, normotensive, normocardiac, eupneic, febrile at 38°C, with infiltrated erythematous lesions, 0.5 to 2 cm in diameter, cocardiform in shape, isolated in places and confluent in others, scattered over the body, with facial and palmoplantar involvement, and negative Nikolsky's sign. Mucosal involvement was manifested by lip oedema with cheilitis, macroglossia and hypersialorrhoea, as well as OGE oedema and erosions of the genital and anal mucosa. There was no conjunctival or endonasal involvement.

Blood count was normal, liver and renal function tests were unremarkable, CRP was slightly elevated at 23.7 mg/L, and viral serologies and immunological tests were negative. Skin biopsy was consistent with erythema multiforme toxidermia.

Pharmacological investigation suggested carbamazepine.

The patient was placed on dermocorticoid and healing cream, with immediate discontinuation of the offending drug, and showed marked improvement.

Conclusion:

Although drug-induced erythema multiforme major (EMM) is uncommon in children, it remains a true dermatological emergency requiring rapid intervention. Among the most commonly implicated drugs, carbamazepine - despite its proven efficacy in the treatment of epilepsy - carries a known risk of triggering severe skin reactions, including EMM, Stevens-Johnson syndrome and toxic epidermal necrolysis.

Although skin rashes can be alarming, the key challenge is to identify the cause promptly and avoid unnecessary investigations or delays.

In our patient, the close timing between the introduction of carbamazepine and the onset of symptoms, together with the absence of infectious causes, left little doubt as to the drug's responsibility. What made the difference was the speed of intervention, early recognition of the reaction and immediate withdrawal of the drug led to a full recovery without complications.

This case reminds us that there is a responsibility behind every prescription, especially when treating vulnerable populations such as children. Clinicians should remain vigilant for new cutaneous or mucosal symptoms following initiation of antiepileptic treatment.

Episodic dimpling on skin of lower extremities on a 15-day-old girl: "Infantile transient smooth muscle contraction of the skin"

Marina López de Dicastillo Cía¹, Isabel Ibarrola¹, Clara Miguel-Miguel¹, Paula Rodríguez Jiménez¹, Irene Palacios¹, Juan Ignacio Yanguas Bayona¹

¹Hospital Universitario de Navarra, Pamplona, Spain

Introduction & Objectives:

Infantile transient smooth muscle contraction of the skin is a benign and self-limiting phenomenon that appears during the first weeks of life. It consists of transient ripples or dimpling on the skin, often affecting the lower extremities, caused by exposure to cold or friction (even spontaneous cases have been described). It is believed that this pilar smooth muscle contraction is due to the immaturity of the autonomic nervous system or smooth muscle hypersensitivity. Histologically, it has been shown that the skin is normal, without smooth muscle hyperplasia or other structural abnormalities. These episodes typically resolve by around two years of age without any sequel.

Materials & Methods:

We present a 15-day-old girl with episodic dimpling on lower extremities when exposing to low temperatures.

Results:

The healthy newborn was brought to consultation due to multiple small, skin-colored dimples on her thighs and legs. From a few days after birth, the baby suffered multiple daily episodes of minutes of duration with spontaneous resolution.

The skin sometimes had an urticarial appearance, so it was decided to perform an analysis with tryptase values. They were within range, so mastocytosis was ruled out. Congenital smooth muscle hamartoma was also considered but excluded due to the bilateral distribution of the lesions and the absence of hypertrichosis or pigmentary changes typically associated with this condition. No skin biopsy was performed, as the history and physical exam suggested diagnosis of infantile transient smooth muscle contraction. One year later, the patient is under follow-up and the frequency of episodes has significantly decreased, supporting the diagnosis.

Conclusion:

Infantile transient smooth muscle contraction of the skin is a rare entity in infants. It seems to be related to immaturity of autonomic nervous system, so it could be included within the physiological phenomena of the skin in newborns.

Recognition of this condition is essential to prevent misdiagnosis and unnecessary interventions. Increased awareness among dermatologists and pediatricians can help reassure families and avoid invasive procedures in newborns with this harmless presentation.

Zinc isn't always low: Atypical Acrodermatitis Enteropathica in an infant with Cancer

Sara Serghini¹, Hind Mansouri¹, Bendaoud Layla¹, Mariem Aboudourib¹, Amal Said¹, Ouafa Hocar¹

¹Mohammed VI University Hospital of Marrakech, FMPM, Cadi Ayyad University, Biosciences and health laboratory, Dermatology-Venerology, Marrakech, Morocco

Introduction & Objectives:

Acrodermatitis enteropathica (AE) is a rare dermatological condition linked to zinc deficiency, typically presenting with a triad of acral and periorificial dermatitis, alopecia, and diarrhea. The congenital form results from genetic defects in zinc transport, while acquired forms may occur in premature infants or those with malnutrition, systemic disease, or suboptimal feeding. Diagnosis is often supported by low serum zinc levels; however, normal levels do not exclude the diagnosis. This case report describes a 2-month-old infant with perianal erosive lesions suggestive of AE, despite normal zincemia, who showed rapid improvement following zinc supplementation. The aim is to emphasize the limitations of serum zinc levels and the value of clinical judgment and therapeutic trials.

Materials & Methods:

A 2-month-old male, under treatment for abdominopelvic rhabdomyosarcoma, was referred to dermatology for persistent painful perianal erosions evolving over a month. He was not exclusively breastfed. Clinical examination revealed two erythematous, well-demarcated erosive plaques with white debris in the perianal region. No alopecia or diarrhea was present.

Laboratory investigations revealed a normal serum zinc level (1.16 mg/L; normal range: 0.6–1.50 mg/L), microcytic hypochromic anemia, and thrombocytopenia. Despite the normal zincemia, the clinical features were consistent with AF.

The patient was started on oral zinc supplementation (1 mg/kg/day), local wound care (sterile saline and healing cream), and nutritional support with a high-protein, high-calorie diet.

Results:

Within a few days of initiating oral zinc therapy, there was rapid and marked improvement of the perianal lesions. This favorable response, in the absence of other interventions, strongly supported the diagnosis of acquired acrodermatitis enteropathica, likely secondary to functional zinc deficiency associated with chronic illness.

Conclusion:

This case underlines that AE can occur despite normal serum zinc levels, particularly in infants with systemic diseases such as cancer. Inflammatory states, hypoalbuminemia, and metabolic disturbances can mask true zinc deficiency by altering zinc distribution and transport. Given that serum zinc reflects only a small fraction of total body zinc, clinical suspicion and therapeutic response should guide diagnosis. Early recognition and zinc supplementation can result in rapid improvement, avoiding delays in care. This report supports the concept of "pseudo-acrodermatitis enteropathica"—a functional zinc deficiency diagnosable by clinical presentation and treatment response rather than lab values alone.

Alpha-Fetoprotein: Navigating Between Hepatoblastoma and Infantile Hepatic Hemangioma

Himeur Zoulikha¹, abuhammad ahmed¹, cherifi sara¹, salhi aicha², dahmani boumedine¹

¹Tlemcen Hospital, tlemcen

²algiers Hospital, algiers

Introduction & Objectives:

Infantile hepatic hemangioma (IHH) is the most common benign liver tumor during the first year of life and ranks as the third most frequent hepatic tumor in children overall. While many IHH lesions remain asymptomatic, severe complications such as hypothyroidism, consumptive coagulopathy, and congestive heart failure can occur in some cases. We present the case of an infant with miliary cutaneous hemangiomatosis associated with a hepatic hemangioma and hypothyroidism, who also exhibited markedly elevated alpha-fetoprotein (AFP) levels—initially suggestive of hepatoblastoma.

Materials & Methods:

A 4-month-old female infant, born at term, presented with miliary cutaneous hemangiomas (11 lesions) and abdominal distension. Clinical examination revealed a 44 cm abdominal circumference, visible collateral venous circulation, and a flattened umbilicus. A palpable mass was detected spanning both upper abdominal quadrants. Abdominopelvic computed tomography (CT) revealed multinodular hepatomegaly, a splenic angioma, and minimal ascites. Laboratory tests showed severe anemia (hemoglobin 7 g/dL) without thrombocytopenia. Renal and hepatic function tests, as well as electrolyte levels, were within normal limits. Thyroid function testing showed elevated thyroid-stimulating hormone (TSH) at 28.7 μ IU/mL (reference range: 0.62–8.05) and a free T4 of 22.95 pmol/L (reference range: 6.2–30.1). Serum AFP was markedly elevated at 1000 IU/mL, initially raising suspicion of hepatoblastoma. However, the patient was started on empiric treatment for infantile hepatic hemangioma. AFP levels normalized during follow-up. The infant was treated with oral corticosteroids (2 mg/kg/day) and levothyroxine (4 μ g/kg/day), with clinical and biochemical improvement.

Results: Infantile hepatic hemangioma is the most frequent benign hepatic tumor in infants. The differential diagnosis includes hepatoblastoma, metastatic neuroblastoma, and focal nodular hyperplasia. AFP is a widely used biomarker for pediatric liver tumors, particularly hepatoblastoma. Physiologically, AFP levels are high at birth and decrease exponentially, reaching near-adult levels by 8 to 9 months of age. Although AFP elevation is most commonly associated with malignant hepatic tumors, it can also be observed in cases of IHH. However, in IHH, the elevation is generally less pronounced and may not be sustained. The source of AFP in IHH cases is believed to be surrounding hepatocytes or reactive hepatic tissue, rather than the hemangioma itself. Hence, while AFP is a valuable diagnostic marker, its interpretation must be context-specific. Persistent and markedly elevated levels, particularly beyond the expected age-related decline, are more indicative of hepatoblastoma.

Conclusion:

Alpha-fetoprotein remains a pivotal biomarker in differentiating between benign and malignant hepatic tumors in infants. In cases such as IHH, AFP may be elevated, but typically to a lesser extent and in a transient manner. Accurate diagnosis requires correlation with imaging findings, clinical context, and, when necessary, histopathological confirmation. Recognizing this distinction is vital to avoid misdiagnosis and to guide appropriate therapy

beyond the vulva: extragenital manifestations of lichen sclerosus

Safa Gueroum¹, el fetoiki fatima zahra¹, hali fouzia¹, chiheb soumiya¹

¹University Hospital Center Ibn Rochd - Casablanca, Dermatology and Venerology Department, CASABLANCA

Introduction & Objectives:

Lichen sclerosus (LS), also known as lichen scleroatrophicus, is a chronic inflammatory sclerosing dermatosis that predominantly affects females and primarily involves the anogenital region. Extragenital involvement is uncommon but can occur on the wrists, palms and soles, nipples, and face. We report a case of vulvar lichen sclerosus in a child, associated with diffuse extragenital manifestations.

Materials & Methods:

Clinical Case An 8-year-old girl, with no significant medical history, presented with a one-year history of pruritic hypopigmented lesions initially affecting both feet, followed by the appearance of achromic lesions on the eyelids and genital area.

Clinical examination revealed scattered hypopigmented macules on the face and upper eyelids, as well as pearly, atrophic, sclerotic papules with central depression and a shiny, striated surface under oblique lighting, located around the ankles and on the dorsal aspects of both feet. Genital examination showed an achromic lesion on the mons pubis, with indistinct borders and a similarly striated and shiny surface, but no distortion of vulvar anatomy.

A diagnosis of lichen sclerosus was suspected and confirmed by skin biopsy. The patient was treated with daily topical tacrolimus, which resulted in clinical improvement.

Results:

Conclusion:

The uniqueness of this case lies in the rarity of childhood-onset lichen sclerosus, particularly in its extragenital form. Early and appropriate management is essential to prevent long-term functional, psychological, and aesthetic complications, especially when diffuse involvement includes the face, as in our patient.



Uleritema ophryogenes in an adolescent patient: a case report

Catalina Valencia Marín¹, María Camipaniagua 96¹, Camilo Arias Rodríguez¹, Maria Catalina cuellar martinez¹, Paula Andrea Arroyave Ramírez¹

¹Universidad Pontificia Bolivariana, Medellin, Colombia

ntroduction & Objectives: Ulerythema ophryogenes is a rare, benign dermatologic condition that typically presents in childhood. It is characterized by inflammatory follicular keratotic papules that may lead to scarring, atrophy, and alopecia, particularly affecting the eyebrows. While it may occur as an isolated finding, it is also associated with various congenital syndromes. This case report describes the clinical, dermoscopic, and histopathological features of ulerythema ophryogenes in an adolescent patient, highlighting diagnostic challenges, the importance of early recognition to prevent aesthetic sequelae, and contributing to the limited literature on this uncommon condition.

Materials & Methods: A 16-year-old male presented with a four-year history of asymptomatic erythematous macules on the cheeks. Clinical examination revealed reticulated erythematous macules in the medial facial areas, extending laterally across both cheeks with a mottled appearance. Dermoscopy showed perifollicular accentuation of light brown to reddish pigmentation on a background of generalized erythema. Histopathology revealed a thin, compact stratum corneum and keratin plugging surrounded by a mononuclear inflammatory infiltrate within a follicular infundibulum, consistent with follicular keratosis. Sebaceous glands were preserved, and no alopecia was noted. These findings confirmed the diagnosis of ulerythema ophryogenes. Treatment with 0.05% topical retinoic acid was initiated, yielding a satisfactory clinical response.

Results: Ulerythema ophryogenes, also known as atrophic facial keratosis pilaris atrophicans, is a rare, benign skin disorder with no malignant potential. It typically manifests in childhood, most often between the ages of 5 and 12, and is characterized by inflammatory follicular keratotic papules that may cause scarring, atrophy, and alopecia, particularly in the eyebrows. The condition can occur in isolation or with congenital syndromes such as Noonan, Cornelia de Lange, and Rubinstein-Taybi. Its true incidence remains unclear due to frequent misdiagnosis. Clinically, erythema and small follicular keratotic papules are often more palpable than visible. Lesions begin on the lateral eyebrows and may extend to the forehead and cheeks. Over time, atrophy and eyebrow loss may develop, with affected areas often feeling rough. Diagnosis is confirmed by clinical and histopathological evaluation. Although the condition stabilizes with age, scarring and eyebrow alopecia are typically permanent. Photoprotection is essential to prevent UV-induced flare-ups. Management is conservative, with mild cleansers and keratolytics like 5% lactic acid, 5% urea, or 2% salicylic acid.

Conclusion: Ulerythema ophryogenes is a rare, benign, progressive skin disorder that primarily affects the face, causing significant aesthetic consequences. Its etiology is not fully understood, but genetic factors play a role. It may occur in isolation or with congenital syndromes. Diagnosis is based on clinical and histopathological findings. Treatment focuses on symptomatic relief and aesthetic improvement using keratolytics, topical retinoids, and, in selected cases, laser therapy. Early recognition and preventive measures improve quality of life and minimize aesthetic burdens in young patients.

Clinical features, systemic evaluation and a novel KMT2A pathogenic variant in Wiedemann- Steiner syndrome

Amadeu José Rodrigues Queiróz¹, Charles Lourenço¹

¹Faculty of Medicine of São José do Rio Preto, São José do Rio Preto, Brazil

Introduction & Objectives:

Hypertrichosis is defined as the growth of hair in any region of the body. It can be classified as generalized or localized; congenital or late-onset; lanugo or terminal vellus hair. We describe a rare case of Wiedermann-Steiner syndrome related to a pathogenic variant never described before.

Materials & Methods:

Case report.

Results:

A 13-year-old female patient with a history of intellectual disability has had increased hair volume on all skin since birth. On physical examination, she has short stature, with long terminal hair on the trunk, limbs, especially on the forearms, and bilateral mandibular region; thick arched eyebrows, and downslanted palpebral fissures. She has hypertelorism and convergent strabismus (figure 1). There is no gingival hyperplasia or history of medication use.

Due to the congenital pattern, generalized with antecubital predominance, a panel was performed to evaluate genes related to regional and generalized hypertrichosis syndromes (table 1). Hormonal evaluation was normal.

Genomic DNA obtained from the submitted sample is enriched for targeted regions using a hybridization-based protocol, and sequenced using Illumina technology. Variants are reported according to the Human Genome Variation Society guidelines.

Confirmation of the presence and location of reportable variants is performed as needed based on stringent criteria using one of several validated orthogonal approaches. A pathogenic variant, c.11131C>T (p.Gln3711*), was identified in KMT2A. This sequence change creates a premature translational stop signal (p.Gln3711*) in the KMT2A gene. It is expected to result in an absent or disrupted protein product. Loss-of-function variants in KMT2A are known to be pathogenic. This variant is not present in population databases (gnomAD no frequency). This variant has not been reported in the literature in individuals affected with KMT2A-related conditions. The KMT2A gene is associated with autosomal dominant Wiedemann-Steiner syndrome (WDSTS), lymphoma and leukemia. WDSTS is a rare disorder characterized by hypertrichosis cubiti (hairy elbows), back hypertrichosis, intellectual disability, postnatal growth delays and short stature. Affected individuals typically have dysmorphic facial features including thick arched eyebrows, long eyelashes, downslanted palpebral fissures, wide nasal bridge and depressed nasal tip. Additional clinical features reported in affected individuals include feeding difficulties, microcephaly, variable skeletal and cardiac anomalies, strabismus, hyperopia, immunodeficiency, and aggressive behavior. Some individuals with KMT2A variants have a presentation consistent with Cornelia de Lange syndrome, which is similar to WDSTS but may also involve digital anomalies and more generalized hirsutism, without evident hypertrichosis cubiti. Due to the possible systhemic manifestation of KMT2A-related disorders, it is important to acess neurological, ophthalmologic, dental, immunologic and cardiovascular evaluation, growth hormone and thyroid assessment and evaluation for metabolic bone disease (table 2). Evaluation for seizures may be indicated for some individuals depending on the specific KMT2A variant identified.

Conclusion: With this case report, we aim to contribute to the literature by expanding the pathogenic variant database and presenting the clinical findings. And more data are necessary to better support systemically our patients.

Pagetoid reticulosis in early childhood: A rare case of mycosis fungoides in a 3-year-old male

Laura Rodríguez Suárez¹, mónica maya gómez¹, elkin peñaranda contreras¹

¹Unisanitas, Bogotá, Colombia

Introduction & Objectives: Pagetoid reticulosis (PR), also known as Woringer-Kolopp disease, is an unusual cutaneous lymphoproliferative disorder considered a solitary variant of mycosis fungoides (MF). It is usually characterized by a single, erythematous, persistent, slow-growing plaque on the extremities. This case aims to report the youngest male patient with PR reported in the literature to our knowledge, a finding that broadens the known age spectrum of this disease and underscores the need for heightened clinical suspicion in pediatric patients.

Materials & Methods: We report the case of a 3 year old male patient with a 7-month history of a well-demarcated erythematous plaque with infiltrated borders and slight surface scaling on the left forearm. On the posterior aspect of the left leg, there is a smaller lesion with the same characteristics.

Results: Skin biopsy of the lesion on the left forearm reported prominent intraepidermal epidermotropic lymphocytes that are hyperchromatic, some exhibiting convoluted nuclei. Immunohistochemistry demonstrated strong positivity for CD2, CD3, CD5, and CD8, with over 90% loss of CD7 and a marked reduction of CD4. Histopathological and immunophenotypic analysis demonstrated an atypical CD8+ T-cell lymphoid infiltrate, confirming the diagnosis of mycosis fungoides, pagetoid reticulosis variant.

Conclusion: This report describes the youngest male case of pagetoid reticulosis to date, characterized by a unique CD8+/CD4-/CD7- immunophenotype. Early consideration of MF pagetoid reticulosis variant in pediatric solitary plaques can lead to prompt biopsy, accurate diagnosis and appropriate management.

Congenital Smooth Muscle Hamartoma in Infants: A Clinicopathologic Study of Five Cases

arisa hirayama^{1, 2}, Satomi Kobayashi^{1, 3}, Michiko Kawakami¹

¹International Catholic Hospital, dermatology, shinjuku, Japan

²The Jikei University Hospital, dermatology, minato, Japan

³Tokyo Women's Medical University Hospital, dermatology, shinjuku, Japan

Background:

Congenital smooth muscle hamartoma (CSMH) is a rare, benign dermal tumor characterized by proliferation of smooth muscle bundles in dermis, presenting at or shortly after birth as slightly erythematous plaques, with or without hypertrichosis.

Objective:

To describe the clinical and histopathologic features of CSMH in five Japanese infants.

Methods:

We retrospectively reviewed five infant cases pathologically diagnosed with CSMH. Clinical data included color, size, location of the lesions, and presence of hypertrichosis. Skin biopsies were takened from all cases, and

histopathological findings, as well as immunohistochemical findings for calponin, a-smooth muscle actin (a-SMA), and desmin, were analyzed.

Results:

All lesions were noted at birth, presenting as slightly erythematous plaques. The size of the lesions ranged from 15 \times 15 mm to 50 \times 60 mm. The lesions were located on the upper arm, abdomen, buttok, knee, and back. hypertrichosis was present in all five cases. Histopathological findings revealed nodular proliferation of smooth muscle bundles in the dermis, extending partially into the subcutaneous tissue. Tumor cells were positive for calponin, a-SMA, and desmin. Masson's trichrome staining demonstrated dark red, irregularly oriented bundles.

Conclusion:

CSMH should be considered in the differential diagnosis of congenital erythematous plaques. While diagnosis is often made based on clinical findings, histopathologic examination is usuful for confirming the diagnosis and excluding other conditions. Hypertrichosis can be frequently observed in Japanese cases with CSMH and should be distinguished from Becker's nevus.

Dupilumab-induced guttate psoriasis in monozygotic twin sisters

hua qian¹, yang Gu*¹

¹Children's Hospital of Soochow University, Suzhou, China

Introduction & Objectives:

Atopic dermatitis(AD) is a chronic recurrent inflammatory disease, and dupilumab, a human monoclonal antibody targeting IL-4/IL-13 pathway, is the firstly approved biological drug for AD in pediatric patients. However, paradoxical psoriasiform eruptions after dupilumab therapy have been reported as a rare adverse effect. Guttate psoriasis during dupilumab treatment in adults has been reported, while in pediatric patients is scarce.

Materials & Methods:

We present two cases of dupilumab-induced guttate psoriasis in monozygotic twin sisters.

Results:

In our case, two 4-year-old monozygotic twin sisters with a 3-year history of uncontrolled AD, characterized by recurrent lesions on the extensor extremities, hands and lips, were initiated on dupilumab therapy. After 6-month treatment, the twin sisters simultaneously developed erythematous, scaly papules on the trunk. Skin biopsy specimen from trunk of one sister revealed psoriasiform hyperplasia, epidermal hyperplasia, perivascular lymphocytes infiltration and parakeratosis, confirming the diagnosis of guttate psoriasis. The dupilumab was discontinued and initiated with topical glucocorticoid ointment. At follow up, two sisters both had nearly complete improvement of her psoriasis lesions, while one sister had recurrent hand atopic dermatitis.

Conclusion:

Guttate psoriasis can appear during dupilumab treatment in twin atopic dermatitis children, highlighting the potential role of genetic predisposition in this adverse reaction.

Generalized pustular psoriasis in children : expérience of dermatology départment ,university hospital of Constantine

Houda Bounneche¹, Mansoul Tarek², Boussaid riyad², Chehad Ahmed Samaouel²

Introduction & Objectives: Pustular psoriasis is a rare but severe form of psoriasis, representing 1–5% of cases globally. Its pediatric form is much rarer and presents significant diagnostic and therapeutic challenges. This study aims to better characterize it

Materials & Methods: We conducted a retrospective, single-center epidemiological study based on medical records of patients under 16 years hospitalized for generalized pustular psoriasis (GPP) in the Dermatology Department of Constantine University Hospital from January 2019 to December 2024. Data were collected using a predefined form and analyzed with IBM SPSS v20.

Results: Nineteen pediatric cases were recorded over five years (male-to-female ratio: 2.8). Most patients (94.47%) were from Eastern Algeria. A personal history of plaque psoriasis was found in 52.6%, and a family history in only one case. The mean age at consultation was 9.15 years; the first flare occurred at a mean age of 7.26 years (range: 1–15 years). Upper respiratory tract infections preceded flares in 52% of cases. Two patients had Down syndrome. The annular form was seen in 47.4% and the Von Zumbusch type in 52.6%. Flexural involvement was present in all patients with Von Zumbusch type, but only in one annular case. Scalp involvement occurred in 78.95%, and nail involvement in 84.21%. No joint involvement was noted. A biological inflammatory syndrome was found in 36.84%, neutrophilic leukocytosis in 52.63%, and neutrophilic cholangitis in one case. Infectious complications were reported in four patients. Skin biopsies (performed in 52.63%) showed Kogoj-Lapière multilocular spongiform pustules. Treatments included ciclosporin (63.2%), methotrexate (21.1%), adalimumab (15.8%), systemic corticosteroids (15.8%), and acitretin (10.5%). Mean hospitalization was 2.68 weeks (1 to 5 weeks). Complete remission occurred in 63.2% and partial remission in 36.8%.

Discussion: GPP is rare in children, typically appearing in the first decade. In our series, onset ranged from 1 to 15 years (mean: 7 years), with a male predominance. Upper respiratory tract infection was the main triggering factor. Flexural involvement was seen in all Von Zumbusch cases and rarely in annular forms, confirming literature findings. Four patients experienced fever and sepsis, requiring urgent care to prevent life-threatening complications. Only one patient had hepatic involvement (neutrophilic cholangitis). While therapeutic options in adults include phototherapy, retinoids, methotrexate, and biologics, pediatric data are limited. In our series, ciclosporin was the most used drug.

Conclusion: We report a new series of 19 pediatric GPP cases, detailing clinical, paraclinical, and therapeutic characteristics.

¹Department of Dermatology, University Hospital of Constantine, Algeria, Constantine

²Department of Dermatology, University Hospital of Constantine, Algeria, Constantine, Algeria



Emerging treatments for dermatologic diseases in infants, children, and adolescents: a systematic review of clinical trials on biologics and small molecule inhibitors

Alireza Jafarzadeh*¹

¹Department of Dermatology, Rasool Akram Medical Complex Clinical Research Development Center (RCRDC), School of Medicine, Iran University of Medical Sciences, Tehran, Iran, Tehran, Iran

Introduction & Objectives:

Recent advancements in the treatment of pediatric dermatological conditions have emerged with the introduction of biologics and small molecule inhibitors (SMIs). These therapies target specific inflammatory pathways, which may enhance treatment outcomes for diseases like atopic dermatitis, psoriasis, and alopecia areata.** This systematic review seeks to assess the effectiveness and safety of biologics and SMIs for dermatologic conditions in children and adolescents, with an emphasis on randomized clinical trials.

Materials & Methods:

We performed an extensive literature search across PubMed, Scopus, and Web of Science, following PRISMA guidelines. Studies included in the review were those that analyzed systemic treatments using biologics and SMIs in subjects under 18 years of age. We extracted data on participant demographics, treatment regimens, effectiveness outcomes, adverse effects, and follow-up details. The risk of bias in the studies was determined using the Cochrane Risk of Bias Tool (RoB2).

Results:

From an initial pool of 1,454 studies, 49 articles fitting the inclusion criteria were identified, encompassing 6,372 cases. The review found that biologics such as Dupilumab, along with investigational JAK inhibitors like Abrocitinib and Upadacitinib, exhibited considerable efficacy in treating various conditions, particularly atopic dermatitis and psoriasis. Dupilumab specifically demonstrated significant improvements in both disease severity and quality of life. While most reported adverse events were mild to moderate, some serious adverse events were noted with certain treatments.

Conclusion:

Biologics and SMIs show great promise as therapeutic options in pediatric dermatology, offering better efficacy compared to traditional treatments. Despite these encouraging findings, additional research is needed to verify their long-term safety, especially in relation to growth and development in younger patients. Future investigations should aim to include a broader range of patient demographics and dermatological conditions beyond those currently studied.



Erythema Multiforme in children: Clinical Aspects and Management Considerations

kaouthar Ben Abdessalem¹, Bendaoud Layla¹, Mariem Aboudourib¹, Hocar Ouafa¹, Amal Said¹

¹Mohamed VI University Hospital Center, Bioscience and Health Laboratory, Faculty of Medicine and Pharmacy of Marrakech, Cadi Ayyad University, Marrakech, Morocco

Introduction & Objectives:

Erythema multiforme (EM) is an acute, self-limited, immune-mediated mucocutaneous condition characterized by the abrupt appearance of targetoid lesions, often triggered by infections. In children, *Mycoplasma pneumoniae* and *Herpes Simplex Virus* are among the most common etiologic agents. Although usually benign, EM with mucosal involvement (EM major) can mimic more severe diseases such as Stevens-Johnson Syndrome, requiring careful diagnostic distinction. This study aims to describe the clinical presentation, etiological factors, laboratory findings, and therapeutic approaches in three pediatric cases of erythema multiforme managed in our department.

Materials & Methods:

We retrospectively reviewed the medical records of three pediatric patients (two boys and one girl), aged 9, 10, and 12 years, who were admitted to the emergency department with erythema multiforme. Data collection included epidemiological profile, clinical findings, laboratory and imaging results, identified infectious agents, and administered treatments.

Results:

All three children presented with similar clinical features: erosive cheilitis with hemorrhagic crusting, restricted mouth opening, bilateral conjunctivitis, diffuse targetoid skin lesions, productive cough, dysphagia, and fever peaking at 39°C, associated with systemic malaise. None of the patients reported recent medication intake or a preceding viral-like illness. Respiratory multiplex PCR testing identified *Mycoplasma pneumoniae* in two cases and *Enterovirus* in one. Chest X-rays were normal in the patient with *Enterovirus*, while the two *Mycoplasma*-positive patients exhibited bilateral interstitial infiltrates. Laboratory workup revealed neutrophilic leukocytosis and elevated C-reactive protein (CRP) levels exceeding 50 mg/L in all cases. Treatment included intravenous acyclovir (10 mg/kg every 8 hours), azithromycin (250 mg/day for 5 days in *Mycoplasma* cases), paracetamol (15 mg/kg every 6 hours), and supportive local care for mucosal lesions.

Conclusion:

Erythema multiforme in children is often triggered by infectious agents, with *Mycoplasma pneumoniae* being a frequent cause in EM major cases. Despite its dramatic clinical presentation, the prognosis is generally favorable with appropriate supportive care and targeted treatment when the infectious etiology is identified. Prompt recognition and differentiation from more severe mucocutaneous syndromes such as Stevens-Johnson Syndrome are essential to ensure proper management and avoid unnecessary interventions

Lupus eruption in Tuberous Sclerosis Complex: A revealing dermatologic challenge

omaima el hafa¹, salma ait seddik¹, Bendaoud Layla¹, Mariem Aboudourib¹, ouafa hocar¹, Amal Said¹

¹Mohammed VI University Hospital, Dermatology, Marrakech, Morocco

Introduction & Objectives:

Tuberous sclerosis of Bourneville (TSB) is a rare, autosomal dominant genetic disorder characterized by the formation of benign tumors, or hamartomas, in various organs. The disease results from mutations in the TSC1 and TSC2 genes, which respectively code for the hamartin and tuberin proteins, negative regulators of the mTOR signaling pathway. These mutations lead to constitutive activation of this pathway, promoting cell proliferation and tumor formation. In systemic lupus erythematosus (SLE), a similar activation of the mTOR pathway has been observed, suggesting a common pathophysiological mechanism with TSB.

Materials & Methods:

We report the case an 11-year-old female patient, followed since the age of 5 for TSB, hospitalized for a pruritic vesicular eruption evolving for two months on the trunk and limbs, giving way to hyperpigmented scars, associated with palmoplantar keratoderma, without other clinical manifestations. Clinical examination revealed symmetrical pinkish facial papules, achromic mountain ash macules, a sorrel skin patch, and hyperpigmented post-vesicular lesions on the trunk and limbs. Psoriasiform palmoplantar keratoderma and a gingival fibroma on the upper gum were also observed. Skin biopsy revealed vacuolar degeneration of basal keratinocytes, lymphohistiocytic infiltrate and leukocytoclastic vasculitis. Direct immunofluorescence revealed a granular lupus band. Biological workup showed normocytic mornochromic anemia with positive coombs test, increased ESR, normal renal function and negative 24h proteinuria, C4 hypocomplementemia, and positive NAAs, concluding SLE. Rapamycin treatment was initiated in the setting of TSB. However, the clinical course will have to be followed over the long term to monitor the therapeutic response and observe the possible appearance of signs of lupus.

Results:

This patient, a carrier of a mutation in the TSC complex responsible for TSB, subsequently developed SLE. The coexistence of these two conditions raises the hypothesis of a pathophysiological link involving the mTOR pathway, hyperactivated in TSB and already recognized as a key player in SLE. This excessive activation could induce an immune imbalance, favoring the production of type I interferon by plasmacytoid dendritic cells, a central mechanism in lupus. In addition, the PI3K/Akt pathway, which regulates mTOR, contributes to the expansion of autoreactive B cells and chronic inflammation, reinforcing the link between these two pathologies. This association opens up new therapeutic prospects, notably with mTOR inhibitors such as sirolimus, already studied in certain refractory forms of lupus.

Conclusion:

Our case joins studies suggesting a common pathophysiological mechanism involving the mTOR signaling pathway, both in the pathogenesis of tuberous sclerosis of Bourneville and in the immunological disorders of systemic lupus erythematosus. The use mTOR inhibitors could represent an innovative strategy in this type of patient, offering a potentially beneficial new therapeutic approach in the management of these pathologies.

Lichen planus pemphigoides in childhood: a rare presentation

Oumaima Zouine¹, Hanane Baybay¹, Zakia Douhi¹, Meryem Soughi¹, Sara Elloudi¹, Fatima Zohra Mernissi¹

¹CHU HASSAN II , Fes, Morocco

Introduction & Objectives:

Lichen plan pemphigoides (LPP) is a rare autoimmune blistering disorder characterized by the coexistence of lichenoid papules and tense bullae. It typically occurs in adults aged 30 to 50, with pediatric cases being extremely rare. The exact etiology remains unclear, but drug exposure is a known trigger in some cases, while others are idiopathic. This case report describes a diagnostically challenging idiopathic LPP in a child, emphasizing the importance of a comprehensive clinical, dermoscopic, and histopathological approach for accurate diagnosis.

Materials & Methods:

An 8-year-old boy with no significant medical history and no prior drug exposure presented with a two-month history of chronic pruritic lesions on the knees, followed by the sudden appearance of tense bullae on the extremities. Clinical examination, dermoscopy, and histopathological analysis were performed to confirm the diagnosis. Direct immunofluorescence (DIF) was also conducted to evaluate the presence of autoimmune markers.

Results:

The patient presented with multiple violaceous to skin-colored papules with slight scaling distributed over the elbows, knees, anterior legs, and the dorsal surfaces of the hands and feet. Tense bullae, some hemorrhagic, were observed on erythematous skin of the palmar and plantar surfaces, including areas that appeared clinically uninvolved. Mucosal examination revealed isolated cheilitis with fern-like Wickham striae on the lips, without involvement of other mucosal sites, and the scalp was clinically normal. Dermoscopy of the papules revealed the presence of classic Wickham striae, supporting the lichenoid nature of the disease. Histopathological analysis from a lichenoid lesion on the knee confirmed interface dermatitis consistent with lichen planus, while a second biopsy from a bullous lesion demonstrated subepidermal blistering with eosinophilic infiltration and dermoepidermal separation. Direct immunofluorescence further supported the diagnosis by revealing linear IgG and C3 deposition along the basement membrane zone, a hallmark finding in LPP. The patient was treated with potent topical corticosteroids and dapsone, resulting in significant improvement with rapid bullous resolution and pruritus control.

Conclusion:

This pediatric case of idiopathic LPP illustrates the importance of recognizing its dual presentation and distinguishing it from other lichenoid or bullous dermatoses. The combination of clinical, dermoscopic, histopathological, and immunopathological findings is essential for accurate diagnosis. Early recognition and appropriate management, as demonstrated in this case, can significantly improve patient outcomes. The favorable response to dapsone and topical corticosteroids further supports their role in managing this rare pediatric condition.

Efficacy of Topical 2% Carteolol in the Treatment of Infantile Hemangiomas: A Dermoscopic and Clinical Evaluation

Esranur Ünal*¹, Muhammed Burak Yücel¹, saadet nurşah güneş¹, yılmaz ulaş¹, fatma türkan mutlu¹, Bengü Nisa Akay²

¹Kayseri City Hospital, Kayseri, Turkey, Kayseri, Türkiye

Introduction & Objectives: Infantile hemangiomas (IH) are the most common benign vascular tumors in infants, often requiring treatment in 10-15% of cases due to complications. While systemic propranolol is the first-line treatment, its potential for systemic side effects raises interest in topical therapies. This study investigates the efficacy of topical 2% carteolol in treating IH and its dermoscopic changes over time.

Materials & Methods: A total of 15 patients with IH were treated with topical 2% carteolol drops applied twice daily for 3 months. Clinical and dermoscopic images were captured at baseline, 1 month, and 3 months. Dermoscopic features such as erythema, clods, coiled vessels, dotted vessels, and others were evaluated. Visual Analog Scale (VAS) scores were used to assess treatment response.

Results: The most frequent dermoscopic findings at baseline were coiled vessels, followed by serpentine and dotted vessels. At 1 and 3 months, significant improvements were observed, with reductions in erythema and coiled vessel scores (p<0.05) and at 3 months with reductions in clod score (p<0.05). The skin-colored structureless area significantly increased, indicating lesion regression (p<0.05). VAS scores showed a significant decrease over the 3-month period, with a positive correlation between baseline dotted vessel scores and treatment response (r:0.656, p:0.008).

Conclusion: Topical 2% carteolol is an effective and safe treatment for IH. Dermoscopic monitoring provides valuable insights into treatment progression, showing significant changes in erythema, clod, and vascular patterns. The presence of dotted vessels at baseline may serve as a predictor of a favorable treatment response. Further studies with longer follow-up are needed to validate these findings.

²Ankara University, Faculty of Medicine, Ankara, Türkiye

Vaccine-Induced Lichen Planus in a 12-Year-Old Patient: A Rare Pediatric Case

Sara Nejjari¹, Fatima-zahra Agharbi¹, Inas Chikhaoui¹, Ghita Basri¹, Soumia Chiheb¹

¹Cheikh Khalifa Bin Zayed Al Nahyan Hospital, casablanca

Introduction & Objectives:

Lichen planus (LP) is a chronic inflammatory skin disorder characterized by pruritic, violaceous, flat-topped papules and plaques. While LP is commonly associated with various triggers, including medications and infections, its induction following vaccination is a rare phenomenon. This case report presents a 12-year-old patient who developed generalized LP after receiving a tetanus vaccine following a puncture wound from rusty metal. The dermoscopic features of LP in this pediatric patient are discussed, highlighting the importance of considering vaccine-induced LP in the differential diagnosis.

Materials & Methods:

A 12-year-old male patient with no significant medical history presented with pruritic, violaceous, flat-topped papules and plaques distributed over his trunk and extremities. The lesions developed approximately two weeks after receiving a tetanus vaccine administered following a puncture wound from rusty metal. Clinical examination confirmed the diagnosis of generalized LP. Dermoscopy was performed using a polarized light dermatoscope, revealing characteristic features of LP, including Wickham striae, white lines over a violaceous background, and follicular hyperkeratosis. A skin biopsy was performed, confirming the diagnosis of LP with typical histopathological findings.

Results:

Vaccine-induced LP is an uncommon adverse reaction, with few cases reported in the literature. The pathogenesis is thought to involve an immune-mediated response triggered by the vaccine, leading to the development of LP. In this case, the temporal association between the tetanus vaccination and the onset of LP supports the hypothesis of vaccine-induced LP. Dermoscopy plays a crucial role in the non-invasive diagnosis of LP, allowing for the visualization of characteristic features such as Wickham striae and follicular hyperkeratosis. These dermoscopic findings were evident in our patient, aiding in the diagnosis and differentiation from other dermatological conditions. The rarity of vaccine-induced LP underscores the importance of considering this diagnosis in patients presenting with new-onset LP following vaccination.

Conclusion:

This case highlights the importance of considering vaccine-induced LP in the differential diagnosis of new-onset LP, especially in pediatric patients with a temporal association between vaccination and the development of skin lesions. Dermoscopy serves as a valuable tool in the non-invasive diagnosis of LP, providing characteristic features that aid in differentiation from other conditions. Clinicians should be aware of this rare adverse reaction to ensure timely diagnosis and appropriate management.



Coexistence of Juvenile Plaque Psoriasis and Cold-Induced Cutaneous Vasculitis Mimicking Chilblain Lupus Erythematosus in a 13-Year-Old Patient

Mihaela Cojocaru¹, Dirzu Diana-Stefania¹, Sinigur Diana¹, Ioniuc Ileana-Katerina², Cozorici Monica-Iuliana²

¹Railway Clinical Hospital, Dermatology, Iasi

²Sain Mary Children's Emergency Hospital Iasi, Iasi, Romania

Introduction & Objectives: Psoriasis vulgaris (PV) is a chronic, immune-mediated inflammatory skin disorder with a complex genetic background. Chilblain lupus erythematosus (CHLE) and cold-induced vasculitic lesions can share overlapping clinical features, particularly in pediatric patients, complicating diagnosis. This overlap is further obscured when PV presents in acral or atypical forms, mimicking other dermatoses.

Materials & Methods: We present a 13-year-old female with a 2-year history of erythematous-squamous plaques, initially on the scalp and later involving the trunk and limbs. She had a positive family history of PV and showed limited response to various topical treatments. On examination, numerous well-demarcated erythematous plaques with silvery-white scales and crusts were noted, some with an atrophic center. Additionally, intermittent painful violaceous plaques appeared on her toes, blanching with pressure and worsened by cold exposure.

Despite the suggestive clinical picture and family history supporting PV, the acral lesions raised suspicion of CHLE. Serological tests and skin biopsies from psoriasiform and acral lesions were performed. Histopathology confirmed PV in truncal plaques and unspecified vasculitis with a prominent perivascular and parietal lymphocytic infiltrate in acral lesions. Direct immunofluorescence of the acral lesion showed coarse fibrinogen deposits in the basement membrane zone and dermal papillae tips, with no Ig or complement. Autoimmune markers, including ANA and SARS-CoV-2 serology, were negative. An abdominal ultrasound revealed incidental cortical cysts in the left kidney, without systemic lupus or other systemic involvement.

Final diagnoses were PV and chilblain-like cold-induced vasculitis (CHLV).

Results: This case highlights the rare coexistence of juvenile PV with cold-induced acral vasculitis mimicking CHLE. The absence of autoantibodies and immune complex deposition excluded lupus. Isolated fibrinogen deposits suggested non-specific vascular injury. This presentation emphasizes the diagnostic value of histopathology and immunofluorescence in pediatric dermatoses with overlapping features.

Conclusion: We report a rare pediatric case of coexisting PV and CHLV, with negative autoimmune workup and isolated fibrinogen deposition. Since CHLE may precede systemic lupus erythematosus (SLE) and cold-induced vasculitis often follows a benign yet recurrent course, accurate differentiation is vital for prognosis and treatment. Defining clear criteria for distinction remains essential in clinical practice.

Beyond the Wound: A Practical Approach to Keloid Management in a Low-Income Pediatric Population

Manuela Betanzo*¹, Emilia Neves¹, Montserrat Cendoya¹, Carolina Droguett¹, Nicolás Opazo¹, Denisse Tabak¹

¹Universidad de Santiago de Chile, Santiago, Chile

Introduction & Objectives: Keloid scars are an exaggerated tissue response to injury, characterized by excessive fibroblast proliferation and collagen overproduction. Their recurrence after primary surgical excision can approach 100%. This study aims to evaluate keloid recurrence following a combined treatment protocol of surgery and intralesional corticosteroids, both pre- and post-surgically.

Materials & Methods: A retrospective cross-sectional study was conducted at a public hospital in Chile, including patients under 18 years of age diagnosed with keloid scars and treated between January 2021 and December 2023. Inclusion criteria were patients with clinically diagnosed keloids who received treatment following a standardized institutional protocol consisting of intralesional Depot Betamethasone administered at three time points: 21 days prior to surgery, intraoperatively, and 21 days post-surgery. Patients treated with corticosteroid monotherapy or other modalities were also included for descriptive analysis. Exclusion criteria included patients with other forms of pathological scarring (e.g., hypertrophic scars) or those who underwent alternative or incomplete treatment regimens. Demographic data (age, sex), treatment modality (monotherapy vs. combined), and recurrence rates were collected from patient records. Recurrence was defined as clinical evidence of keloid regrowth at the surgical site during follow-up. Data were analyzed descriptively.

Results: A total of 29 patients treated for keloids were included, with an average age of 24 years and a female predominance (72% vs. 28%). Seventy-seven percent were treated with corticosteroid infiltrations: 47% as monotherapy and 30% combined with surgery. Three percent were treated solely with compression therapy. All surgical patients received corticosteroid infiltrations 3 weeks before, during, and 3 weeks after the procedure. Of these, 14% experienced recurrence.

Conclusions: Multimodal treatment proved to be more effective than monotherapy. While surgical excision alone has a recurrence rate ranging from 45% to 100%, combining it with corticosteroid infiltrations reduced recurrence to 29% in reported literature. In this study, the triple corticosteroid protocol was associated with a notably lower recurrence rate of 14%, suggesting that serial administration may be superior to single-dose therapy in preventing keloid regrowth.



Baseline characteristics, and treatment patterns of pediatric and adolescent patients in a Canadian phase IV study (CANDID) of dupilumab for the management of atopic dermatitis.

Vimal H. Prajapati*¹, Melinda Gooderham², Chih-ho Hong³, Kim A. Papp⁴, Jason K. Lee⁵, Andrew Ferrier⁶, Kirsten Walker⁷, Irina Turchin⁸, Maksym Breslavets⁹, Sanjay Siddha⁴, Dave N. Adam⁴, Maryam Shayesteh Alam¹⁰, Kerri Purdy¹¹, Rachel Asiniwasis¹², Irene Lara-Corrales¹³, Michelle Ramien¹⁴, Rashpal Bhogal¹⁵, Maryam Piram¹⁶

Introduction & Objectives:

Insights into the real-world treatment of atopic dermatitis (AD) are key to clinical decision making. The aim of this analysis was to characterize patients who receive dupilumab for moderate-to-severe AD in a real-world setting within Canada in patients from pediatric and adolescent cohorts.

Materials & Methods:

The CANDID phase 4 study is an ongoing, prospective, observational study of patients with moderate-to-severe AD who receive dupilumab according to Canadian-specific prescribing information. We report baseline characteristics, and treatment patterns for patients enrolled from October 4, 2023 to March 2, 2025. Analyses are descriptive; no formal statistical comparisons were performed

Results:

Of 305 patients enrolled in the study, 56 (18.4%) were under the age of 18. Of these, 22/56 were between the ages of 6 and 11 (pediatric) and 34/56 between 12 and 17 years (adolescent).

Pediatric and adolescent patients had a high disease at enrollment prior to dupilumab start with mean (\pm standard deviation) scores being 30.7% (21.0%) for body surface area, 20.4 (11.4) for Eczema Area and Severity Index, 7.2 (2.3) for Peak Pruritus Numerical Rating Score, 6.0 (2.8) for Skin Soreness Numerical Rating Score, 4.0 (3.1) for Sleep Disturbance Numerical Rating Score, 15.2 (5.6) for Children's Dermatology Life Quality Index, and

¹University of Calgary , Calgary, Canada

²Queens University, Kingston, Canada

³University of British Columbia, Vancouver, Canada

⁴University of Toronto, Toronto, Canada

⁵Toronto Asthma and Allergy Clinic , Toronto, Canada

⁶Stratica Dermatology , Edmonton, Canada

⁷University of Saskatchewan , Saskatoon, Canada

⁸Dalhousie University, Fredericton, Canada

⁹Centre for Medical and Surgical Dermatology, Toronto, Canada

¹⁰Probity Medical Research , Barrie, Canada

¹¹Dalhousie University , Halifax, Canada

¹²University of Saskatchewan , Regina, Canada

¹³Sick Kids Hospital, University of Toronto, Toronto, Canada

¹⁴Alberta Children's Hospital, University of Calgary, Calgary, Canada

¹⁵Sanofi Canada, TORONTO, Ontario, Canada

¹⁶CHU Sainte-Justine, Université de Montréal, Montreal, Canada

14.6 (7.5) for Dermatitis Family Impact.** In the previous 12 months, 96.4%, 21.1%, and 8.2% patients were treated with topical corticosteroids, systemic corticosteroids, and conventional systemic non-corticosteroid immunosuppressants, respectively.

Conclusion:

Canadian pediatric and adolescent patients enrolled in CANDID demonstrated considerable multidimensional burden of disease across signs/symptoms despite extensive previous use of topical and systemic AD treatments. This is the first real world evidence assessment of pediatric and adolescent Canadian patients aged 6-17, demonstrating the burden of disease including family impact and that of living with atopic dermatitis.

Clinical spectrum and outcomes in pediatric mastocytosis: a retrospective study

Iuliana-Ștefania Dohotaru¹, Ana Preda¹, Alexandra Andreea Galatanu¹, Alina Suru^{1, 2}, Adelina-Maria Sendrea^{1, 2}, Carmen Maria Salavastru^{1, 2}

¹Colentina Clinical Hospital, Pediatric Dermatology Department, Bucharest, Romania

Introduction & Objectives:

Mastocytosis represents a group of diseases characterized by the infiltration of mast cells into various organs and tissues. Cutaneous mastocytosis, the most common type in children, includes three subtypes: mastocytoma, urticaria pigmentosa and diffuse cutaneous mastocytosis. Systemic mastocytosis is rarely reported in the pediatric population. This study aims to analyze the clinical manifestations of cutaneous mastocytosis in children, along with associated symptoms, disease progression and management strategies.

Materials & Methods:

We performed a retrospective study on fifteen patients diagnosed with cutaneous mastocytosis between January 2020 and March 2025.

Results:

Among the fifteen cases analyzed, eleven patients were diagnosed with urticaria pigmentosa, which manifested as an eruption of erythematous-brown macules and papules of variable shapes and sizes, predominantly localized on the trunk, face and extremities. Four patients presented with mastocytomas, two with solitary lesion and two with multiple lesions, clinically described as irregular brown-beige plaques, localized on the trunk. Out of the eleven patients diagnosed with urticaria pigmentosa, two demonstrated atypical clinical manifestations. One patient presented with well-demarcated erythematous macules on the scalp, while the second patient exhibited soft, pearly-white papules confined to the vulvar region. Mean age at diagnosis was 2 years old, with a male predominance (9 cases). Two patients reported a personal history of food allergies. Local symptoms consisted of mild pruritus in two cases. The Darier sign was positive in 14 cases. One patient had a history of flushing episodes. Skin biopsy was performed in seven patients, confirming the diagnosis in five; in two cases, the histopathological findings were non-specific, and the diagnosis was established based on clinical features. All patients were advised to undergo serum tryptase level assessment and molecular analysis for c-KIT mutation. No c-KIT mutations were identified. Mild elevation of serum tryptase levels was detected in one patient, who was subsequently referred to a hematology specialist. Elevated serum IqE levels were found in two patients, and mild eosinophilia was observed in one. Abdominal ultrasound examinations, performed in four patients, revealed no specific abnormalities. Management included topical corticosteroid therapy for all patients. Systemic ketotifen was prescribed in five cases. Additionally, all patients received written guidance on trigger avoidance and general recommendations for the management of mastocytosis. The clinical course was favorable in the majority of cases, with no systemic progression of the disease observed during follow-up.

Conclusion:

Pediatric cutaneous mastocytosis is typically a benign disorder with skin lesions that frequently regress spontaneously around puberty. However, this favorable prognosis is not guaranteed in all patients. Systemic involvement remains uncommon in children, but constant follow-up is essential to promptly identify any potential

²"Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

progression. While the clinical presentation is typical in the majority of cases, clinicians should remain vigilant for any atypical manifestations. Given that therapeutic management is predominantly symptomatic with limited efficacy, patient education, trigger avoidance, and careful monitoring constitute the mainstay of effective long-term care.

Onychomadesis and onychomadesis-mimicking disorders in pediatric dermatology

Khatuna Kudava¹

¹European University, Dermatology, Tbilisi, Georgia

Introduction & Objectives: Onychomadesis, a specific form of nail dystrophy, is a constant subject of interest for pediatric dermatologists. Onychomadesis caused by blockage of the nail matrix, is clinically manifested by the separation of the nail plate from the matrix, its permanent attachment to the nail bed, and eventual replacement of the damaged nail with a new one. This process is often considered a complication of various viral infections, autoimmune and critical illness, and drug treatment. Similar dystrophic changes in the nails may also develop as a result of local infection of the nail matrix. A complete definition of the problem is important for selecting the correct management.

Materials & Methods: 23 patients aged 4-7 years with onychomadesis and onychomadesis-mimicking disorders of the nails were presented. A complete dermatological history was collected; microscopic and cultural examinations were used to identify the etiological cause. Structural changes of the nails were fully evaluated using a digital dermoscope.

Results: None of the patients had traumatic nail injuries. Dermatoscopic examination revealed features of nail dystrophy in all of them. Patients were divided into two groups. The first group included 9 patients with classical multiple onychomadesis of the fingernails and toenails, detachment of the proximal areas of the nail plates from the nail matrix and nail bed, and attachment of the medial and distal parts to the nail bed. None of these patients had paronychia. Microscopic examination did not reveal any fungal infection. In the anamnesis, 4 patients in this group had a history of hand, foot, and mouth disease associated with an unknown type of enterovirus, and 5 patients had a history of hand, foot, and mouth disease associated with a Coxsackie virus. The time from the end of the infections to the appearance of onychomadesis ranged from 28 to 45 days. It took 1.5 to 3 months for healthy nails to grow. During this time, the damaged nail plate remained attached to the nail bed along with the growth of the new nail. Complete nail growth occurred without specific treatment. The second group included 14 patients with onychomadesis-mimicking disorders of fingernails associated with paronychia. Nail dystrophies developed acutely, within a few days of the onset of infection. Eight patients were diagnosed with a single nail disorder, and six patients- with more than one nail. The clinical presentation was similar: the nail plate was detached from the nail bed with the medial and distal parts of the nail attached to the nail bed. Within two to three weeks, all damaged nails were completely separated from the nail bed. As a result of microscopic and cultural examinations, 9 patients were diagnosed with candidal infection, 5 with streptococcal infection. All of them were treated with antifungal and antibacterial treatment, respectively, with complete recovery.

Conclusion: Onychomadesis associated with hand, foot, and mouth disease present as a subacute secondary complication that develops later; with multiple nail involvement, without paronychia. Onychomadesis-mimicking disorders associated with candidal and streptococcal paronychia develops as an acute process. Proper management is important for effective treatment of onychodistrophis. Onychomadesis associated with hand, foot, and mouth disease does not require specific treatment, while in the case of onychomadesis-mimicking disorders, it is necessary to treat the infection that causes it.

Two cases of pediatric melanoma with uncommon germline mutations: genetic and follow-up insights

Gaia Bellesi¹, Sandra Peternel^{1, 2}, Marijana Vicic^{1, 2}

¹Clinical Hospital Centre Rijeka, Department of Dermatology and Venereology, Rijeka, Croatia

Introduction & Objectives:

Juvenile melanoma (JM) is rare and often differs from adult melanoma in presentation and pathology, making diagnosis challenging. While most JM cases are sporadic, a subset may arise within hereditary cancer syndromes. Germline mutations in genes such as *CDKN2A*, *BAP1*, *TERT*, *POT1*, and other DNA repair genes have been linked to melanoma predisposition. Though familial clustering is uncommon in JM, a personal or family history of malignancy should prompt genetic evaluation. We report two pediatric melanoma cases with pathogenic germline variants identified through blood-based molecular testing, emphasizing the relevance of genetic assessment and sustained follow-up.

Materials & Methods:

Two children diagnosed with melanoma underwent full clinical evaluation, including surgical excision, sentinel lymph node biopsy, imaging, and genetic counseling. Detailed family histories were recorded. Germline testing was conducted on peripheral blood using Next-Generation Sequencing and Multiplex Ligation-dependent Probe Amplification. Both cases were managed through a multidisciplinary team of dermatologists, pediatric oncologists, surgeons, and geneticists.

Results: Patient 1 (female, 6 years at diagnosis): A pigmented lesion on the right thigh showed recent color and shape changes. Excision revealed spitzoid melanoma (Breslow 1.8 mm). Family history on the paternal side included melanoma, lung, and breast cancer. Re-excision and sentinel lymph node biopsy (SLNB) were negative; thoracic computed tomography was normal. Germline genetic testing of the patient identified a heterozygous pathogenic *CHEK2* splice donor variant (c.444+1G>A). The patient remains free of disease recurrence throughout 4 years of follow-up. The patient's parents declined their own genetic testing.

Patient 2 (male, 4 years at diagnosis): A long-standing pigmented lesion on the right calf showed morphological changes over two years. Excision revealed Spitz melanoma (Breslow 3.8 mm). Re-excision and SLNB were negative. Family history included a maternal grandfather with colon cancer and grandmother with breast cancer. Germline testing revealed a pathogenic *RAD50* variant (c.2260_2266del) and variants of uncertain significance in *ATM* and *PRKAR1A*. The patient remains free of disease recurrence throughout 3 years of follow-up. The same *RAD50* mutation was confirmed in the patient's mother, who is now under surveillance for increased risk of cancer.

Conclusion:

These cases illustrate the need to consider hereditary cancer syndromes in pediatric melanoma patients, especially when family history suggests increased cancer risk. Although *CHEK2* and *RAD50* mutations are not typically associated with melanoma, their presence may reflect emerging genetic contributors and justify individualized long-term monitoring due to broader malignancy associations. *CHEK2* mutations are linked to elevated risks for breast, colorectal, prostate, thyroid, kidney, and lung cancer, while *RAD50* mutations have been associated with breast, ovarian, intestinal, hematologic, and other malignancies. Early identification of pathogenic germline

²University of Rijeka, Medical Faculty, Rijeka, Croatia

variants supports risk-adapted care and family-wide cancer prevention strategies.

Pediatric Psoriasis: Epidemiological, Clinical and Therapeutic Study

Hiba Kherbach¹, mhaimer soukaina¹, safae el msayryb¹, ezzaki chaimaa¹, chaouche mohammed¹, sokaina chhiti¹, radia chakiri¹

¹UHC SOUSS-MASSA, DERMATOLOGY, AGADIR, Morocco

Introduction & Objectives:

Psoriasis is a chronic inflammatory skin disease** whose incidence, in children, increases with age.** It differs from adult forms in clinical presentation and treatment. This study analyzes its epidemiological, clinical, therapeutic, and evolutionary aspects in patients at our university hospital.

Materials & Methods:

This retrospective descriptive study, spanning fourthy months (from January 2021 to May 2024) (January 2021–May 2024), includes all children under 15 diagnosed with psoriasis in pediatric dermatology consultations.

Results:

A total of 73 children were included: 68.50% girls and 31.50% boys, with a sex ratio (M/F) of 0.46. All patients were under 15 years old, with a median age of 9.52 years. Parental consanguinity was noted in 2.73%. Atopy (8%) was mainly atopic dermatitis. Family history of psoriasis was reported in 8.21%. Psychological trauma was found in 2.33% of cases. Plaque psoriasis was most common (87.67%), followed by guttate (8.22%), mixed forms (12.33%), nummular, and follicular types (4.11% each). Pustular psoriasis occurred in 2.74%, and no erythrodermic cases were recorded. One patient had psoriatic arthritis. PASI scores did not exceed 25. Two patients had DLQI scores above 21, requiring psychiatric consultation. Diagnosis was clinical, except for three biopsied cases. 95.90% received topical treatment, mainly dermocorticosteroids (95.90%) and emollients (71.23%). Two patients required systemic treatment (acitretin). Response was favorable in 61.64% of cases, with 20.55% in complete remission, 28.76% partially responding, and 12.33% relapsing.

Conclusion:

Pediatric psoriasis is a common chronic inflammatory dermatosis, typically diagnosed through clinical examination. Topical therapies remain the mainstay of treatment, given the generally favorable prognosis. Effective management requires parental involvement and attention to the psychological impact of the disease on the child.

CLOVES Syndrome: A Condition to Recognize

Mounira bouselsal 1 , tarek mansoul 1 , amel gherfi 1 , khadidja bouarroudj 1 , asma bouhila 1 , lilia keghouche 1 , ahmed samaouel chehad 1

¹University Hospital Center (CHU) of Constantine Dr. Abdessalem Benbadis, dermatology, Constantine, Algeria

Introduction & Objectives:

CLOVES syndrome (Congenital Lipomatous Overgrowth of the trunk with Vascular malformations, Epidermal nevi, Skeletal anomalies) is a sporadic, polymalformative syndrome caused by mutations in the *PIK3CA* gene. It is characterized by asymmetric body overgrowth, truncal lipomatous hamartomas, and various vascular and skeletal malformations. We report a new case of this rare condition

Materials & Methods:

We present a 14-month-old infant, born at term following an uncomplicated pregnancy to non-consanguineous parents, referred for evaluation of a polymalformative syndrome. Clinical examination revealed left-sided hemihypertrophy and lower limb overgrowth present since birth; an extensive verrucous epidermal nevus involving the trunk and left thigh; a mid-dorsal capillary malformation (port-wine stain); a partially thrombosed venous malformation of the right forearm; a "sandal gap" deformity of the toes; hypertrophy of the second toe; digital anomalies with restricted finger mobility; and a pectus excavatum. Abdominopelvic ultrasound and echocardiography showed no abnormalities. A diagnosis of CLOVES syndrome was established. The patient was started on curative-dose enoxaparin and sirolimus at a dose of 0.6 mg/m².

Results:

CLOVES syndrome is a rare, sporadic, and complex disorder belonging to the spectrum of mosaic overgrowth syndromes. It is caused by activating somatic mutations in the *PIK3CA* gene, located on chromosome 3q26.32. Clinically, it is characterized by congenital adipose tissue hyperplasia, lipomatous masses—particularly involving the thoracic and abdominal walls—capillary and venous malformations, lymphatic anomalies, and arteriovenous malformations. Epidermal hamartomas may be present at birth or develop during childhood. Skeletal anomalies frequently include scoliosis, asymmetry of limb bones, genu recurvatum, polydactyly, syndactyly, foot widening, and a widened first interdigital space (sandal gap). Detection of *PIK3CA* mosaic variants in affected tissue, when feasible, currently enables a definitive diagnosis. Differential diagnoses mainly include Proteus syndrome and Klippel-Trenaunay syndrome. Management is multidisciplinary, involving dermatologists, pediatricians, radiologists, psychologists, orthopedic surgeons, and vascular surgeons. As in other *PIK3CA*-related disorders, sirolimus has shown promising therapeutic potential.

Conclusion:

The diagnosis of CLOVES syndrome should be considered in the presence of varying degrees of congenital asymmetric overgrowth, vascular malformations, epidermal hamartomas, and extremity anomalies.

Photoprotection in Children: Knowledge, Practices, and Risk Perception

Hiba Kherbach*¹, mhaimer soukaina¹, safae el msayryb¹, ezzaki chaimaa¹, chaouche mohammed¹, sokaina chhiti¹, radia chakiri¹

¹UHC SOUSS-MASSA, DERMATOLOGY, AGADIR, Morocco

Introduction & Objectives:

Ultra-violet (UV) exposure is a major risk factor for skin cancer, with cumulative effects starting in childhood. Despite photoprotection recommendations, high-risk behaviors persist due to limited awareness. This study evaluates children's knowledge, practices, and awareness of sun exposure risks and photoprotection measures.

Materials & Methods:

We conducted a cross-sectional descriptive and analytical study using a questionnaire completed by children on our city's beaches and surrounding areas, as well as through social media to reach various regions of our country.

Results:

A total of 972 children participated, with the 14–17 age group most represented (50%) and a sex ratio (F/M) of 2.85. Private school attendance was 57.4%, and 66.7% had moderately pigmented skin. Among the 83.3% engaging in outdoor activities during vacations, 50% preferred staying in the sun, and 37% had over 30 minutes of direct exposure. Sun protection was used by 63%, including sunscreen (51.9%), though only 10.7% reapplied it every two hours, while 67.9% applied it once. At the beach, 53% wore a t-shirt, and among hat users (46.3%), 88.9% wore caps, while 11.1% chose wide-brimmed hats. Sunglasses were worn by 40.7%, and 75.9% actively sought shade. Awareness of sun risks was high (88.9%), with 53.7% recognizing the link to skin cancer, 40.7% to pigmentation disorders, and 53.7% to skin aging. However, 46.3% believed sunlight benefits mental health. Photoprotection awareness came mainly from parents (48.1%), social media (27.8%), and physicians (7.4%). While 92.6% knew phototype I is most sensitive, 14.8% believed wearing dark-colored clothing on sunny days was necessary.

Conclusion:

Our findings highlight the need for increased awareness campaigns, particularly targeting younger children, boys, and public school students. The involvment of parents, schools, and healthcare professionals is essential to promote protective behaviors and reduce sun-related risks from an early age.

Neonatal purpura due to severe congenital protein C deficiency

Fatim Ezzahra Afryad¹, Bendaoud Layla¹, Mariem Aboudourib¹, Amal Said¹, Ouafa Hocar¹

¹Mohammed VI University Hospital, Marrakech, Morocco, dermatology and venerology, Marrakech, Morocco

Introduction & Objectives:

Severe congenital protein C deficiency (homozygous deficiency or compound heterozygosity) is a rare but potentially life-threatening hereditary coagulopathy. It manifests within the first hours of life as extensive purpura or even widespread skin necrosis, contrasting with a preserved general condition and a negative infectious work-up. It often reveals a homozygous deficiency in a newborn whose parents are heterozygous carriers. We report the case of a newborn, born to consanguineous parents, who presented with extensive ecchymotic purpura revealing a protein C deficiency.

Materials & Methods:

This is a male newborn, on day 3 of life, hospitalized in the neonatal unit due to the onset, approximately 12 hours after birth, of ecchymotic purpura in a sock-like distribution on the left foot. The lesions worsened over the following 48 hours, with rapid extension to other areas and the formation of hemorrhagic bullae.

Results:

On clinical examination, the patient was pale, reactive, slightly hypotonic, afebrile, and hemodynamically and respiratory stable. Skin examination revealed extensive macular ecchymotic plaques on both feet in a sock-like distribution, the left thigh, the left hand and forearm in a glove-like pattern, and the abdomen, all topped with hemorrhagic bullae on both hands and feet. Initial work-up showed a negative infectious screening, anemia, thrombocytopenia, and a low prothrombin time (PT). A hematological origin was thus suspected, and a full coagulation panel revealed undetectable protein C activity. On day five of life, the newborn died following a ventricular hemorrhage with hydrocephalus.

Conclusion:

Prenatal diagnosis, early recognition of skin lesions, and immediate replacement therapy combined with anticoagulant treatment are essential to improve prognosis and prevent the devastating consequences of severe protein C deficiency.

Acute Hemorrhagic Edema of Infancy: A Report of Five Cases

kaouthar Ben Abdessalem¹, Bendaoud Layla¹, Mariem Aboudourib¹, Hocar Ouafa¹, Amal Said¹

¹Mohamed VI University Hospital Center, Bioscience and Health Laboratory, Faculty of Medicine and Pharmacy of Marrakech, Cadi Ayyad University, dermatology, Marrakech, Morocco

Introduction & Objectives:

Acute hemorrhagic edema of infancy (AHEI) is a rare, benign leukocytoclastic vasculitis that typically affects infants between 4 and 24 months of age. Clinically, it presents with striking purpuric and targetoid skin lesions accompanied by localized edema, but the general condition of the patient often remains well preserved. Despite its generally favorable prognosis, rare but severe complications, such as intussusception, may occur. This case series aims to describe the clinical spectrum of AHEI and to underscore the importance of clinical vigilance in detecting early signs of potential complications.

Materials & Methods:

We retrospectively analyzed five cases of infants diagnosed with AHEI and managed in our dermatology department. Data collected included patient demographics, clinical presentation, preceding events, laboratory results, imaging findings, treatment, and outcomes. The diagnosis was based on clinical features, supported by laboratory and imaging investigations when necessary. Follow-up was conducted to monitor for potential complications.

Results:

The study included five infants aged 2 to 13 months (3 males, 2 females). Four patients had a history of recent infection or flu-like symptoms prior to onset, with no recent episodes of vaccination. All patients presented with characteristic targetoid or pseudo-targetoid purpuric lesions predominantly affecting the face and lower limbs, along with peripheral edema. The trunk was generally spared.

- **Case 1:** A 2-month-old infant developed purpura with facial and limb involvement following watery diarrhea. The condition was complicated by acute intestinal intussusception.
- Case 2: A 10-month-old presented with limb edema and purpura after a flu-like illness, with normal imaging and favorable evolution.
- Case 3: A 13-month-old had fever and annular purpura on the lower limbs, with laboratory evidence of inflammation; no complications occurred.
- Case 4: A 9-month-old infant presented with targetoid purpura after a recent respiratory infection, associated with palpebral edema and limb involvement. Clinical and ophthalmologic assessments were unremarkable.
- Case 5: A 7-month-old, afebrile and stable, had widespread purpura and limb edema, with normal laboratory findings and spontaneous resolution.

Laboratory findings were variable: inflammatory markers were elevated in three cases, while others remained within normal limits. Imaging was performed in selected cases and contributed to the diagnosis of intussusception in one patient. No systemic treatments were administered; all patients were managed with close clinical monitoring. Four patients had complete resolution within 7–10 days without sequelae.

Conclusion:

Acute hemorrhagic edema of infancy is a rare vasculitic condition with a dramatic cutaneous presentation but typically benign and self-limiting course. Clinical diagnosis is paramount, as laboratory tests are often non-specific. While the overall prognosis is excellent, complications such as intussusception, though uncommon, justify close monitoring during the acute phase. This case series emphasizes the importance of recognizing AHEI as a distinct clinical entity to avoid unnecessary interventions, while remaining alert to signs suggestive of complications. Early identification and vigilant follow-up are essential to ensure favorable outcomes.

Pattern of restrictive dietary practices in children with chronic dermatosis: a cross-sectional study

Mihira Kommineni¹, Irene Mathews*¹, Ssv Prasad¹, Mahendra m¹, Swetalina Pradhan¹

¹All India Institute of Medical Sciences, Patna, India

Introduction & Objectives:

Chronic dermatoses in children are a source of significant anxiety to their families. Caregivers often implement dietary restrictions seeking cure or in an attempt to manage flares, driven by cultural beliefs or local practices. Such practices are concerning, especially in children from low-income families, who already face challenges in accessing balanced nutrition.

These practices are prevalent, yet there is a lack of comprehensive scientific research examining their frequency and the potential effects on children's overall health and quality of life.

This study aims to assess the prevalence and patterns of dietary restrictions practiced among children with chronic dermatoses and to explore the factors influencing these practices.

Materials & Methods:

This cross-sectional study included all children aged 4–16 years with chronic dermatoses who presented over a two-month period to the Dermatology Outpatient Department of a tertiary care centre in Eastern India. Data on dietary restriction practices and the probabable motivations were collected using a structured questionnaire.

Results:

A total of 95 children with various chronic dermatoses were enrolled; 62% were female and 38% male. Atopic dermatitis was the most common diagnosis (21 cases, 22.1%), followed by vitiligo (21.1%) and psoriasis (11.6%).

Thirty-five children (36.85%) were found to be on various dietary restrictions. The most commonly avoided foods were fish (in 28.4% cases), chicken (22.1%), eggs (21.1%), mutton (20%), sour foods (17.9%), dairy (10.5%), and brinjal (3.2%). There is a high prevalence of protein and iron deficiencies among the children in Eastern India. This pattern of dietary restrictions, predominantly focused on avoiding animal-source foods, may exacerbate these issues.

The most common diagnoses among the thirty-five children practicing dietary restrictions were vitiligo (13 cases, 37.1%), followed by psoriasis (25.7%) and atopic dermatitis (5.7%).

Among children with vitiligo, a significantly higher proportion were found to be following diet restrictions (65%), as compared to children with other chronic dermatoses (20%), p < 0.001. This may be because vitiligo is linked to significant social stigma in communities with skin of colour and provokes significant anxiety in caregivers.

Children from low-income families were less likely to follow diet restrictions (33.9% vs 41% in higher-income families), but this was not statistically significant (p=0.48). The caregivers attributed these practices to advice from family members (in 45.7% of cases), allopathic practitioners (31.4%), practitioners of other systems of medicine (20%), and other sources (2.9%).

Conclusion:

Restrictive diets are commonly practiced among children with chronic dermatoses, with variations across different communities. Dietary restrictions were most frequently observed in children with vitiligo and primarily involved the avoidance of animal-source foods. Dermatologists should be aware of such practices in their communities, as they can impact the nutrition of children with chronic dermatoses. These issues should be addressed through counselling and awareness initiatives.

A curious case of acute fever with vesicular rash in an infant

Shubhangi Gupta*1, manjaree morgaonkar1

¹dr. dy patil medical college, hospital and research centre, pimpri, dermatology, pune, India

Introduction & Objectives:

Fever with vesicular rash in infants can present a significant diagnostic challenge, often leading to misdiagnosis and unnecessary interventions. While maculopapular eruptions are common in pediatric viral illnesses, vesiculobullous presentations warrant careful consideration of both infectious and non-infectious etiologies. We describe a case of chikungunya virus—associated vesicular rash in a nine-month-old infant initially misdiagnosed as Staphylococcal scalded skin syndrome (SSSS). Our objective was to highlight the clinical features, discuss plausible mechanisms of bullae formation in chikungunya, and propose a diagnostic algorithm to guide clinicians faced with fever and vesicular eruptions in this age group.

Materials & Methods:

A nine-month-old male presented with three days of high-grade fever (max 38.5 °C) followed by one day of erythematous rash that progressed to dusky vesicles and flaccid bullae over the trunk and extremities. Initial management with oral amoxicillin-clavulanate and paracetamol was instituted for presumed SSSS. On dermatology evaluation, detailed history, morphological assessment of lesions, and systemic examination were performed. Laboratory investigations included complete blood count, liver and renal function tests, HIV serology, blood and blister fluid cultures, Tzanck smear, and chikungunya-specific IgM ELISA. Differential diagnoses considered were SSSS, hand-foot-mouth disease (HFMD), varicella, herpes simplex and zoster, and chikungunya.

Results:

Laboratory parameters were within normal limits; HIV serology, bacterial cultures, and Tzanck smear were negative. Chikungunya IgM ELISA returned positive, confirming acute infection. No multinucleated giant cells were observed, effectively excluding herpesvirus etiology. The infant received supportive care—adequate hydration, antipyretics, and skin care—and recovered fully over ten days without complications. The evolution of petechiae, purpuric macules, and flaccid bullae in the absence of mucosal involvement or focal staphylococcal infection, combined with positive serology, established the diagnosis of chikungunya-induced vesiculobullous rash.

Conclusion:

Chikungunya virus infection should be included in the differential diagnosis of acute fever with vesicular or bullous rash in infants, particularly in endemic regions during post-monsoon months. Proposed mechanisms for bullae formation include virus-induced keratinocyte necrosis and immune-mediated endothelial damage leading to capillary leak and purpura. Early recognition prevents unnecessary antibiotic use and invasive investigations. We present a simplified diagnostic algorithm to assist clinicians in distinguishing chikungunya from other vesiculobullous disorders in infancy, thereby promoting timely, appropriate management and reducing healthcare burden.

Cutaneous Manifestations as Initial Indicators of Neurological Dysraphism

Zineb Mernissi¹, aboudourib meriam¹, Bendaoud Layla¹, Ouafa Hocar¹, Amal Said¹

¹Mohammed VI University HospitalBioscience Laboratory, Faculty of Medicine and Pharmacy of Marrakech, Cadi Ayyad University, Department of dermatology and venerology, Marrakech, Morocco

Introduction & Objectives:

Cranial and spinal dysraphisms are a group of malformations related to abnormal neural tube closure. Often asymptomatic at birth, these malformations are typically discovered in the context of midline or paramedian cutaneous anomalies, neurological syndromes, orthopedic complications, or polymalformative syndromes. We conducted this study to investigate the dermatological manifestations that serve as initial indicators of neurological dysraphism.

Materials & Methods:

This is a retrospective descriptive study including 27 patients, identified through a review of the pediatric dermatology consultation records at the Mohamed VI University Hospital Center in Marrakech, over the period extending from August 2009 to September 2024.

Results:

The mean age of the patients was 4 months, and 60% of the cases were male. The dermatological signs of cranial dysraphisms included occipital hemangioma in 8 cases, midline scalp nodule in 5 cases, midline scalp hemangioma in 5 cases, and cutis aplasia in 2 cases. The cutaneous manifestations of spinal dysraphisms included lumbosacral hemangioma in 5 cases and lumbosacral achromic nevus in 2 cases. Medical imaging was recommended for 19 cases but was not performed for 8 patients due to limited resources or the need for sedation. Ultrasound was performed for 2 patients, CT scan for 3 patients, and magnetic resonance imaging (MRI) for 6 patients. Radiological investigations revealed 2 cases of meningocele, 2 cases of cutaneous mass associated with cranial bone defect, and normal findings for 7 patients.

Conclusion:

The term dysraphism refers to abnormalities in the closure of the neural tube. In clinical practice, it also encompasses closure defects in the midline skin, vertebrae, paravertebral muscles, and meninges. Most cases of occult spinal dysraphism are diagnosed based on cutaneous signs, which are present in approximately three-quarters of cases. These signs are typically located along or near the midline of the lumbosacral region, and less frequently in the thoracic or cervical areas. Suspicious cutaneous lesions suggestive of occult spinal dysraphism warrant a thorough family history check and complete physical examination, with a particular focus on the neurological assessment. Dermatological findings are categorized according to their associated level of risk, which guides the clinical and paraclinical approach. Currently, no standardized guidelines exist regarding the optimal imaging modality for evaluating suspected dysraphism based on cutaneous anomalies. Magnetic resonance imaging (MRI) remains the most appropriate modality for the detailed evaluation of the spinal cord; however, its use is limited by factors such as cost, availability, and the frequent need for sedation in young children. These limitations were especially evident in our context, where MRI was indicated but could not be performed for 8 patients. High-resolution ultrasonography (US) is a non-invasive screening tool that may be employed in infants under 6 months of age, prior to ossification of the vertebral bodies. In our study,** ultrasound** was used for the

diagnosis of meningocele in two patients with cutis aplasia and in three cases of midline scalp hemangioma. Nonetheless, it is a technique that is operator-dependent and less sensitive than MRI.

Physiological and Transient Dermatoses in Neonates: A Moroccan Study on Epidemiology and Associated Factors

lalla safia echarif¹, nada nacir¹, meryem aboudourib¹, layla bendaoud¹, said amal¹, ouafa hocar¹

¹Faculty of Medicine and Pharmacy in Marrakech, Biosciences and Health Laboratory, Marrakech, Morocco

Introduction & Objectives:

Neonatal skin undergoes significant adaptive changes after birth, frequently presenting as physiological or transient dermatoses. Although most of these conditions are benign, accurate identification is essential to prevent unnecessary medical interventions. This study aimed to evaluate the prevalence of physiological and transient dermatoses in healthy Moroccan new-borns and to investigate potential associations with maternal, neonatal, and paternal factors.

Materials & Methods:

This 3-month cross-sectional study (July-October 2024) examined 300 full-term neonates at the maternity ward of Mohammed VI University Hospital. After obtaining Ethics Committee approval and maternal written consent, we included only healthy neonates with uncomplicated histories. Two physicians collected maternal/neonatal data via questionnaire and performed complete dermatological examinations. The 300-patient sample size was biostatistically determined to be representative. Data were analyzed using SPSS (p<0.05 significant).

Results:

Our study found physiological dermatoses in 99% of neonates, most commonly sebaceous hyperplasia (55.6%), Mongolian spots (42%), and Epstein pearls (32%), while transient dermatoses affected 28.3%, primarily erythema toxicum (25.3%). Maternal factors showed 46% aged 18-25 years, 63.7% multiparity, and surprisingly low folic acid supplementation (46.7%) despite adequate iron intake (84.7%). Neonatal characteristics revealed male predominance (55%), day-1 examination (93%), and phototype III prevalence (49%). Key associations included: maternal dark phototype correlating with sebaceous hyperplasia, Mongolian spots and Epstein pearls (all p<0.05); parental skin tone influencing genital hyperpigmentation (darker) and vernix caseosa (lighter); folic acid supplementation linked to increased transient dermatoses (p=0.04), especially erythema toxicum (p=0.016); and male sex associated with transient neonatal pustulosis. Notably, maternal age and delivery mode showed no significant associations.

Conclusion:

Recognizing common neonatal skin patterns in healthy Moroccan newborns is critical to preventing unnecessary medical interventions. Our findings highlight the influence of ethnic background and maternal factors on these benign conditions, underscoring the need for culturally adapted clinical guidelines in neonatal care.

Lichen in Children: Unveiling the Hidden Burden through a Case Series

Imane Hakim¹, Bendaoud Layla¹, Mariem Aboudourib¹, Amal Said¹, Hocar Ouafa¹

¹Mohammed VI University Hospital, Dermatology Department, Biosciences Research Laboratory, FMPM, Cadi Ayyad University, Marrakech, Morocco

Introduction & Objectives:

Lichen is a chronic inflammatory dermatosis that occurs preferentially in middle-aged adults. It is uncommon in children. Various clinical forms have been described in the literature. We describe the epidemiological and clinical features of lichen in children in a series of 34 cases.

Materials & Methods:

This is a retrospective descriptive study including all cases of children with lichen who consulted our dermatology department, over a period of 14 years and 8 months from January 2010 to August 2024.

Results:

Over a period of 14 years and 8 months, 34 cases of lichen were recorded. There were 25 girls and 9 boys. The sex ratio was M/F = 0.36. Mean age was 10 years (range 4-16 years). Phototype ranged from III to IV. The mean duration of lesion development was 19.4 months. One patient had a similar case in the family. Clinical forms were dominated by lichen planus in 67.64% of cases: pigmentogenous lichen in 8 cases, classical lichen in 5 cases, actinic lichen in 2 cases, lichen annulare and follicular lichen in one patient each. Nail lichen was found in 7 patients. Scleratrophic lichen in 11 cases. No involvement of the oral mucosa was noted. Histological studies were carried out on 20 children, confirming our clinical diagnosis and eliminating other differential diagnoses (16 skin biopsies and 4 nail biopsies). Peladic dermatitis and vitiligo were associated in one case each. Treatment was based on strong to very strong class dermocorticoids combined with topical tacrolimus, in localized cutaneous and scleratrophic vulvar forms. In generalized forms (cutaneous and nail), general corticosteroid therapy was indicated, either orally or in the form of monthly injections. A depigmenting treatment combined with emollients was also used. The evolution was marked by regression of clinical lesions and partial improvement without recurrence in the majority of patients.

Conclusion:

We report a large series of cases of lichen in children. It confirms the data in the literature regarding the rarity of this entity and the partially satisfactory response to treatment. Most therapies lack solid evidence of efficacy, and a more lucid consensus on the treatment of pediatric lichen is urgently needed.

The Hidden Ocular Face of McCune-Albright Syndrome

Imane Hakim¹, Bendaoud Layla¹, Mariem Aboudourib¹, Amal Said¹, Hocar Ouafa¹

¹Mohammed VI University Hospital, Dermatology Department, Biosciences Research Laboratory, FMPM, Cadi Ayyad University, Marrakech, Morocco

Introduction & Objectives:

McCune-Albright syndrome is a rare hereditary disorder characterized by the association of fibrous dystrophy of the bone, café-au-lait skin patches and endocrine abnormalities. Ocular manifestations such as retinal vasculitis may also occur, although this is less frequent.

The aim of our work is to draw attention to an extremely rare clinical presentation through a case associating McCune-Albright syndrome and retinal vasculitis.

Observation:

This 20-year-old patient, with a history of cerebral aneurysm in 2020, and depression under treatment for two years, presented with an isolated progressive decrease in visual acuity that had been evolving for 10 months. The ophthalmological examination revealed impaired visual acuity on the right (counting fingers at 2 metres), and preserved visual acuity (10/10) on the left. The fundus showed papillary hyperhaemia (papilla 1/10), poor macular reflex and exudative occlusive vasculitis with centrolateral haemorrhage and hyalitis on the right. Examination of the left eye was unremarkable. Fluorescein angiography showed papillitis, occlusive venous vasculitis and laserized ischemia with persistent active neo-vessels close to the macula. In addition, dermatological examination revealed congenital café-au-lait spots, segmental with irregular Maine coast margins, blashko-linear stopping on the midline of the face, "S"-shaped on the thorax, "V"-shaped on the back and linear on the left upper limb. The diagnosis of retinal vasculitis in McCune-Albright syndrome was made in view of the cutaneous involvement, endocrine disorders (precocious puberty) and ocular involvement, with no bone involvement found. The patient was put on a corticosteroid bolus with slight improvement. A panretinal photocoagulation laser was indicated to inactivate the territories of the corticosteroids.

Discussion:

McCune-Albright syndrome is a fascinating example of the complexity of genetic diseases. The main features of McCune-Albright syndrome include: bone dysplasia, leading to bone fragility or deformity; endocrine disorders, including precocious puberty, thyroid disorders and other hormonal imbalances; and pigmented café-au-lait skin patches often appearing asymmetrically and well-defined. Retinal vasculitis, although not systematically associated with the syndrome, may be the result of systemic inflammation or an altered immune response. This can lead to symptoms such as visual blur, reduced visual acuity and halos or floating spots. Ocular complications can have a devastating impact on patients' quality of life. Treatment of McCune-Albright syndrome is usually multidisciplinary, involving endocrinologists, ophthalmologists and other specialists. Management of the symptoms of retinal vasculitis may include corticosteroids or other immunosuppressive drugs, depending on the severity of the inflammation. Increased awareness of McCune-Albright syndrome and its manifestations may lead to earlier diagnosis and better symptom management.

Conclusion:

McCune-Albright syndrome, and the retinal vasculitis that can be associated with it, illustrate the unique challenges encountered in the management of complex genetic diseases. Thanks to a multidisciplinary approach, ongoing research and increased awareness, it is possible to significantly improve the quality of life of patients affected by this rare condition.

Safety and Use of Oral Minoxidil in Pediatric Patients: A Systematic Review and Meta-analysis

Omr Alassaf¹, Layan Almutairi*², Albatool Tumbukani³, Razan Alhumidi³, Abdulkarim Alawdah⁴, Samia Almutairi⁵, Salman Albadr⁶, Khamael Aljabri⁷, Joud Alyousef⁸, Hanin Mobarki⁹, Renad Almutairi¹⁰, Rose Aljuaid¹¹, Osama Alsharif¹²

Introduction & Objectives:

Oral minoxidil is increasingly being used off-label in children to treat various conditions, including hair disorders and refractory hypertension. Despite its growing application, concerns regarding its safety and efficacy remain.

Objectives: This systematic review and meta-analysis assess oral minoxidil's benefits and risks in pediatric patients across various dosages.

Materials & Methods:

A comprehensive search across different databases, depending on using different predefined terms, was conducted, and only prospective or retrospective cohort studies in English were included in this review.

Results:

Following the screening of 625 articles, 13 met the inclusion criteria, comprising a total of 364 pediatric patients. In the meta-analysis, adverse events occurred in 25.2% of patients with hair disorders (P=0.009) and 26.1% of patients with refractory hypertension (P<0.001), with high heterogeneity (I2=94-95%). Improvement in hair density was seen in 59.5% of patients (P<0.001, I2=68%), while hair loss stabilization occurred in 24.3% (P<0.001, I2=52%). These findings suggest that OM may represent a safer and more effective option for pediatric patients.

Conclusion:

Further large-scale, randomized controlled trials are recommended to confirm these results and establish optimal dosing guidelines.

¹University of Hail, College of Medicine, Hail, Saudi Arabia

²Princess Nourah Bint Abdulrahman University -, College of Medicine, Riyadh, Saudi Arabia

³Taif University, College of Medicine, Taif, Saudi Arabia

⁴KING FAISAL UNIVERSITY, College of Medicine, Al Hofuf, Saudi Arabia

⁵Prince Sultan Military Medical City, Dermatology Department, Riyadh, Saudi Arabia

⁶King Saud University, College of Medicine, Riyadh, Saudi Arabia

⁷Ummul Al Qura University - Makkah, College of Medicine, Makkah, Saudi Arabia

⁸Princess Nourah Bint Abdulrahman University -, College of Pharmacy , Riyadh, Saudi Arabia

⁹Jazan University, College of Medicine, Jizan, Saudi Arabia

 $^{^{10}}$ Imam Mohammad Ibn Saud Islamic University, College of Medicine , Riyadh, Saudi Arabia

¹¹King Saud bin Abdulaziz University for Health Sciences (KSAU-HS), College of Medicine, Riyadh, Saudi Arabia

 $^{^{12}}$ King Fahd Hospital, Dermatology Department , Pediatric Dermatology , Madinah, Saudi Arabia

Successful treatment of pyoderma gangrenosum in an adolescent with adalimumab

Thu Le Thi Hoai*1, Thuy Nguyen1, Le Huu Doanh1

¹National Hospital of Dermatology and Venereology, Hanoi

Introduction & Objectives:

Pyoderma gangrenosum (PG) is a rare, chronic inflammatory skin disease characterized by painful and rapidly progressive ulcers. This neutrophilic dermatosis predominantly affects individuals in their mid-40s and is rarely observed in pediatric patients. Recently, off-label medications for PG have been utilized, with systemic corticosteroids and cyclosporine remaining the first-line treatments. Emerging evidence suggests that adalimumab, an anti-TNF- α agent, demonstrates significant efficacy and safety in treating PG. However, literature on its use in pediatric patients is extremely limited. Here, we present a case of an adolescent with PG who achieved sustained, complete response to adalimumab.

Materials & Methods:

A case report was described based on meticulous clinical and laboratory examinations, with sufficient data of follow-up during treatment.

Results:

A 14-year-old female patient, otherwise healthy, presented with a 1-year history of pustules progressing to painful ulcers, initially on her left shin and later spreading to bilateral extremities. Prior to referral to our institution, she was treated with systemic corticosteroids combined with mycophenolate mofetil, with no significant improvement. She experienced multiple exacerbations upon corticosteroid tapering. At our center, a definitive diagnosis of PG was made based on histopathology showing necrosis, mixed cellular inflammation with predominant neutrophil infiltrate, and five minor criteria from the 2018 Delphi consensus: (1) exclusion of infection, (2) pathergy, (3) history of pustule ulcerating within 4 days of appearing, (4) peripheral erythema and undermining border, and (5) multiple ulcerations with at least one on the anterior lower leg. The patient was also screened for and ruled out inflammatory bowel disease, inflammatory arthritis, and hematologic disorders. After an insufficient response to high-dose systemic corticosteroids combined with cyclosporine, the patient was switched to adalimumab 40 mg subcutaneously every month. Initial improvement was observed within a week, and complete resolution was achieved with a PGAR 100 and a PGA score of 0. No adverse events were documented during her treatment. Systemic corticosteroids were tapered and discontinued after 6 months, and the adalimumab regimen was maintained 6 months followed by a reduction in frequency and discontinuation after a total of 10 months with controlled disease. To date, the patient has been free of treatment with no recurrence for 6 months.

Conclusion:

This case report highlights an adolescent with recalcitrant PG who experienced prolonged, complete resolution with adalimumab treatment. Adalimumab appears to be a promising option for refractory cases of PG in pediatric patients, though further studies are warranted.