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Abstract N°: 227

Long-term safety of ustekinumab in paediatric patients with moderate-to-severe plaque psoriasis: Results from an ongoing observational study

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Introduction & Objectives:

Ustekinumab (UST) was approved by the European Medicines Agency for the treatment of moderate-to-severe plaque psoriasis in adolescents (12–17 years of age) in 2015 and in children (6–11 years of age) in 2020, based on findings from the CADMUS trials. Interim results are reported from an ongoing post-authorisation safety study (EUPAS19506) in which the long-term safety profile of UST was evaluated in patients 6–17 years of age.

Materials & Methods:

Patients diagnosed with moderate-to-severe plaque psoriasis were enrolled in this ongoing, prospective, observational, multicentre study conducted in routine clinical practice. Patients initiated UST either within the 12-week period prior to their first study assessment or the following 2-month period. Clinical data and adverse events (AEs) are reported from the start of data collection (25 October 2017) to the time of data cut-off (16 January 2024). Results are reported by age cohort (younger: 6–11 years; older: 12–17 years).

Results:

At the time of data cut-off, 39 younger and 86 older patients (125 total) were enrolled in the study; median follow-up time was 1.13 years and 2.02 years, respectively. Baseline demographics and patient characteristics are reported in Table 1.

The most common location of psoriasis across both cohorts was the scalp (81.6%). Most patients (84.0%) received other psoriasis therapies during the study, the most common of which was a combination of topical betamethasone and calcipotriol (50.4%). Overall, 80.8%, 15.2%, 2.4% and 1.6% of patients were prescribed one, two, three or more than three lines of biologics, respectively, throughout their treatment history, including prior to study entry.

Seventeen patients (13.6%) permanently discontinued UST: two in the younger cohort (AEs: n=1; lack of effectiveness: n=1) and 15 in the older cohort (AEs: n=2; lack of effectiveness: n=10; other reasons: n=3). Overall, 92% and 86% of patients were still receiving UST at 1 and 2 years after treatment initiation, respectively (Figure 1). In total, 474 AEs were reported by 79 patients (63.2%); of these, 146 AEs reported by 43 patients (34.4%) were considered related to UST. Nasopharyngitis (n=15 [12.0%]), headache (n=8 [6.4%]) and pyrexia (n=8 [6.4%]) were the most frequently reported UST-related AEs (Table 2). A total of 15 serious AEs were reported by 11 patients (8.8%; younger cohort: n=1; older cohort: n=10); two of these AEs were considered related to UST (younger

cohort: acute urticaria [n=1]; older cohort: appendicitis [n=1]). No cases of malignancy or deaths were reported in either cohort.

Conclusion:

Overall, the proportion of patients still receiving UST at 1 and 2 years after treatment initiation was high. The results of this analysis are consistent with previously reported analyses and confirm the established safety profile of UST in children and adolescents with psoriasis.

Table 1. Demographics and baseline characteristics of enrolled patients, stratified by age group

Characteristic	Younger cohort (6–11 years of age) n=39	Older cohort (12–17 years of age) n=86	Overall N=125
Median age, years (IQR)	9.0 (7.0–11.0)	14.0 (12.0–15.0)	12.0 (11.0–14.0)
Sex, n (%)			
Male	15 (38.5)	33 (38.4)	48 (38.4)
Female	24 (61.5)	53 (61.6)	77 (61.6)
Median BMI, kg/m^2 (IQR)	17.4 (15.6–19.8)	20.3 (18.7–24.5)	19.7 (17.0–23.3)
Median time from psoriasis diagnosis to start of current UST treatment, years (IQR)	3.0 (0.1–4.2)	4.1 (1.7–8.7)	3.9 (1.4–6.0)
Median age at psoriasis onset, years (IQR)	5.9 (4.5–7.4)	9.1 (5.3–12.0)	7.0 (5.0–11.0)
Patients with at least one of the following psoriasis variants, n (%)			
Scalp psoriasis	32 (82.1)	70 (81.4)	102 (81.6)
Nail psoriasis	13 (33.3)	36 (41.9)	49 (39.2)
Palmoplantar psoriasis	9 (23.1)	30 (34.9)	39 (31.2)
Inverse psoriasis	5 (12.8)	7 (8.1)	12 (9.6)
Psoriatic arthritis	-	6 (7.0)	6 (4.8)
Other	1 (2.6)	4 (4.7)	5 (4)
Prior exposure to biologic therapy, n (%)	6 (15.4)	9 (10.5)	15 (12.0)

BMI, body mass index; IQR, interquartile range; UST, ustekinumab.

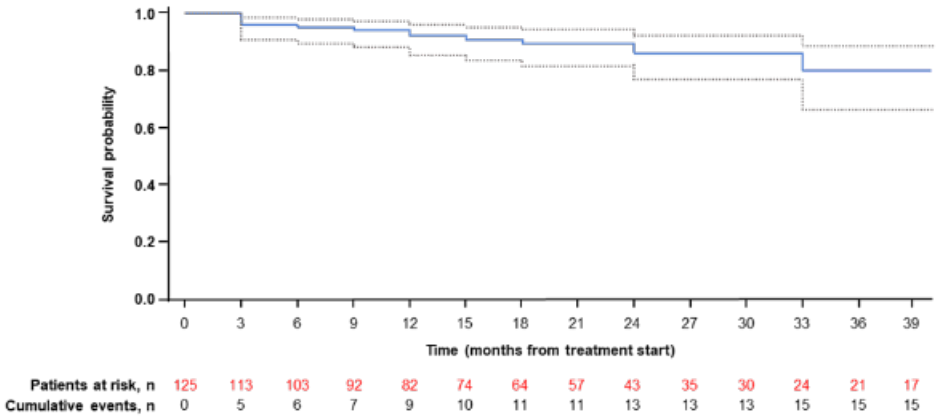
Table 2. Most common UST-related AEs, stratified by age group*†

AE	Younger cohort (6–11 years of age) n=39		Older cohort (12–17 years of age) n=86		Overall N=125	
	Patients, n (%)	Events, n	Patients, n (%)	Events, n	Patients, n (%)	Events, n
Any UST-related AE	10 (25.6)	29	33 (38.4)	117	43 (34.4)	146
Infections and infestations						
Nasopharyngitis	5 (12.8)	9	10 (11.6)	14	15 (12.0)	23
Influenza	3 (7.7)	3	4 (4.7)	4	7 (5.6)	7
Rhinitis	-	-	3 (3.5)	3	3 (2.4)	3
Sinusitis	1 (2.6)	1	1 (1.2)	1	2 (1.6)	2
Ear infection	-	-	1 (1.2)	2	1 (0.8)	2
General disorders and administration site conditions						
Pyrexia	2 (5.1)	3	6 (7.0)	7	8 (6.4)	10
Fatigue	-	-	7 (8.1)	9	7 (5.6)	9
Influenza-like illness	-	-	5 (5.8)	5	5 (4.0)	5
Respiratory, thoracic and mediastinal disorders						
Oropharyngeal pain	-	-	7 (8.1)	7	7 (5.6)	7
Cough	-	-	4 (4.7)	4	4 (3.2)	4
Nervous system disorders						
Headache	-	-	8 (9.3)	8	8 (6.4)	8
Hypoaesthesia	-	-	2 (2.3)	3	2 (1.6)	3
Musculoskeletal and connective tissue disorders						
Arthralgia	-	-	3 (3.5)	3	3 (2.4)	3
Pain in extremity	1 (2.6)	2	2 (2.3)	2	3 (2.4)	4
Myalgia	-	-	2 (2.3)	2	2 (1.6)	2
Gastrointestinal disorders						
Abdominal pain	-	-	2 (2.3)	3	2 (1.6)	3
Nausea	-	-	2 (2.3)	2	2 (1.6)	2
Skin and subcutaneous disorders						
Pruritus	1 (2.6)	1	1 (1.2)	1	2 (1.6)	2
Psychiatric disorders						
Insomnia	-	-	2 (2.3)	2	2 (1.6)	2

*AEs were determined to be related to UST if a relationship was considered possible, probable or very likely by investigators; †Any AE occurring in more than one patient in the overall population is reported here.

AE, adverse event; UST, ustekinumab.

Figure 1. Kaplan–Meier plot of time to UST discontinuation



Dashed grey lines represent 95% confidence intervals.
Data are only presented if at least 10% of the overall patients are at risk at the time point under evaluation.
UST discontinuation was defined as the time period (expressed in months) from first administration dose until the last administration dose + 1 day. Patients who terminated UST prematurely for any reason were considered as having an event, and the date of last administration dose was used in the calculation of time to event. Patients still ongoing were censored at the earliest of the following time points: death, study completion, withdrawal from study and data cut-off.
The cumulative number of events at each time point is displayed.
UST, ustekinumab.

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Abstract N°: 254

A case report of generalised lichen nitidus successfully treated with pimecrolimus 1% cream

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A case report of generalised lichen nitidus successfully treated with pimecrolimus 1% cream

Introduction & Objectives:

Generalised lichen nitidus is a rare variant of lichen nitidus, a condition of unknown aetiology characterised by multiple, tiny, skin-coloured papules on the chest, abdomen, extremities, and genitalia. Various treatments have been tried with mixed results. We report here a 10-year-old Chinese boy with generalised lichen nitidus successfully treated with pimecrolimus 1% cream. The latest literature of pimecrolimus cream used in the treatment of lichen nitidus is also discussed.

Materials & Methods:

A 10-year-old Chinese boy presented with a 12-month history of an asymptomatic rash. The rash started on the face for more than 3 months before it progressed to the trunk and limbs. He had no previous treatment and was otherwise well. Physical examination revealed multiple 1-3mm skin-coloured papules on the face, trunk, and limbs. Koebner's phenomenon was noted over the arms. Differential diagnoses including molluscum contagiosum, epidermodysplasia verruciformis, and lichen nitidus were considered. Histopathological examination of the papules was performed, and he was subsequently treated with pimecrolimus 1% cream. We also discussed the pimecrolimus response of lichen nitidus in our patient and the two previous case reports (Table 1).

Results:

Histopathology of his papules showed lichenoid tissue reaction involving slightly depressed areas of the epidermis, spanning up to five rete ridges with expansion of capillary dermis, with acanthosis of the rete at the edge. Typical claw-like appearance was not present. The overlying epidermis was partly atrophic and almost perforating in some lesions. The inflammatory infiltrate was composed of lymphocytes, occasional histiocytes, and a few epithelioid cells. The overall features were compatible with lichen nitidus. Treatment with pimecrolimus 1% cream twice daily produced gradual clearance of his papules over 8 weeks. All his lesions were cleared after 8 months of treatment. Follow-up for one year after treatment cessation showed no recurrence. Table 1 shows children to young adult age were affected. Our patient and the other child had generalised distribution whereas the adult had the localised variant. Improvement was observed at 2 weeks in the adult, and 8 weeks in our patient and the other child. Remission was achieved in the adult and our patient after 8 weeks and 8 months respectively. No recurrence was seen in the adult and our patient after treatment cessation for 3 months and 1 year respectively.

Conclusion:

Pimecrolimus cream can be an effective treatment for lichen nitidus, but longer duration of treatment may be required for the generalised variant. Spontaneous remission cannot be ruled out and further studies are needed to determine the pimecrolimus efficacy and treatment duration. However, this may prove challenging as generalised lichen nitidus is a rare condition.

Table 1: Summary of case reports of lichen nitidus treated with pimecrolimus 1% cream
Authors
Lee et al.
Farshi et al.
Present case

Table 1: Summary of case reports of lichen nitidus treated with pimecrolimus 1% cream

Authors	Age/years	Sex	Distribution	Koebner's phenomenon	Improvement (weeks)	Remission
Lee et al.	20	Male	Localised: Penis	Not specified	2	8 weeks
Farshi et al.	8	Male	Generalised: Torso, body, extremities, face	Not specified	8	Not specified
Present case	10	Male	Generalised: face, trunk, limbs	Yes	8	8 months

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Abstract N°: 563

Dermatologic Disorders of blood vassels and hair follicles: Atypical clinical findings in Silver-Russell syndrome. Mexican Chid Case.

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Introduction & Objectives:

Silver-Russell syndrome (SRS OMIM #180860), first reported by Silver and Russell, independently described pre and postnatal growth retardation, characteristic facial features, and body asymmetry. It's a rare genetic disorder with an estimated incidence of 1:30.000-1:100.000. Clinical presentation is an easily recognizable spectrum in typical cases, characterized by growth failure at birth, short stature, frontal bossing, triangular face, clinodactyly of 5th fingers, asymmetry and hemihypertrophy. Dermatologic disorders that include blood vessels (Capillary hemangioma of infancy) and hair follicles (hypo and hypertrichosis) have not been previously reported and could be atypical clinical findings in SRS.

Molecular testing in 30-60% of SRS of patients clinically diagnosed is caused by hypomethylation of the paternal imprinting center 1 located on chromosome 11p15.5

This case reports dermatologic disorders, including blood vessels and hair follicles, which are clinical findings not previously reported in SRS

Materials & Methods:

18-month-old male child with history of prenatal growth retardation. At 36 weeks of pregnancy, showed low birth weight and height. Physical examination somatometry under 3rd centile. Dolichocephaly, diffuse hypothichosis on scalp, multiple capillary hemangiomas of infancy (CHI) soft, bright-red vascular plaques, localized on superior parietal region of 4x4 cm, occipital region more than 5 irregular diffuse plaques 1x1 cm extending to right parietal region and satelital vascular plaques; triangular face, facial asymmetry, bilateral gonial angle hypertrichosis, prominent forehead, diffuse CHI on superior frontal region to superior nasal bridge area; long philtrum, small mouth, thin lips, narrow-ached palate; retrognathia; hands ungeal hypoplasia, 2nd right finger clinodactyly limitated to movement; low extremities: ankle ichthyosiform dermatosis, ungeal hypoplasia, 2nd-3rd syndactyly toes bilaterally, brachydactyly and camptodactyly of right feet; umbilical and right inguinal hernia and unilateral cryptorchidism. Cardiologic defect detected.

Results:

At birth, transfontanelar US revealed a right coroideo plexus cystic.

At month of age: Molecular sequence studies of the H19/IGF 2 genes, imprinting Control Region, chromosomal location 11p15.5. Positive mutation that causes SRS.

14 months of age, Cardiac ultrasound identified a wide membranous interventricular septal defect.

19 months old, radiological not visualize the carpal centers growth, considering a bone age of 3 months bilaterally.

Conclusion:

Silver-Russell syndrome is a clinically and genetically heterogeneous condition involving poor growth at birth; this is a primary abnormality with failure to thrive and postnatal growth retardation. Mainly diagnosed on clinical signs and symptoms that vary widely in severity among affected individuals, features are nonspecific. There not consensus on the clinical definition of the disorder.

Molecular tests are used to confirm the diagnosis of loss of methylation in the paternal imprinting. The present report shows the complete clinical and radiological phenotype in SRS, confirmed with the molecular analysis

The genetic heterogeneity with wide variability and a broad spectrum of symptomatology could be responsible, in our case, exhibited dermatological clinical findings not previously reported in SRS and considering the possibility of associated medical conditions

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Abstract N°: 575

Unusual eyelid bump: granuloma annulare in a child

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Introduction & Objectives:

Granuloma annulare (GA) is a low-frequency granulomatous inflammatory skin disease with an estimated incidence of less than 1%. It is located primarily on the back of the hands or feet but can infrequently occur in unusual places such as eyelids. We present the case of granuloma annulare on the eyelid of a toddler, an unusual location for a patient of this age.

Materials & Methods:

A 2-year-old female patient with no significant past medical history presented to the dermatology clinic with asymptomatic lesions in the right upper eyelid that had developed approximately one year prior. On physical examination, she had multiple 2-3 mm, flesh-colored, firm papules on the right upper eyelid that adopted an annular configuration. Dermoscopy revealed a pinkish-erythematous background, punctate vessels, and a whitish and yellow-orange surface.

Results:

Given the location of the lesion, a skin biopsy was defined. The histopathological findings showed skin with intact epidermis and in the dermis, an area of necrobiosis surrounded by a palisade of histiocytes and lymphocytes. The findings were consistent with a granuloma annulare-type granulomatous dermatitis. The lesion resolved after excision, and she had no recurrence during her follow-up three months later.

Conclusion:

GA is a non-infectious cutaneous granulomatous inflammatory disease with a low prevalence (0.1% to 0.4%). It can affect people of any age, predominantly in those under 30 years of age; it has a slight predominance in men in pediatric age and in women in adulthood. The etiology of GA is unknown; most cases are idiopathic and there is a possible relationship to a delayed hypersensitivity reaction and a cellular immune response to different antigens. The most affected areas of GA are: extension surface of the limbs, the back of the fingers and the palms of the hands (90%), the trunk (5%) and other areas, such as the eyelids (5%), as presented in our case. Localized GA, the most frequently found variant, is characterized by normochromic, erythematous or violaceous papules of firm consistency and a shiny surface with a central involution. The treatment of GA is controversial. Its self-resolving nature is well known in 80% of cases after two years. Compared to surgical excision, its benefit is diagnostic rather than therapeutic, given the risk of recurrence after excision. Therefore, management should be tailored to individual patients, considering the location and extent of the lesions, as well as the potential risks and benefits of intervention.

Granuloma annulare is a low-frequency entity that should be considered in the differential of annular lesions on the eyelids of pediatric patients.

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Abstract N°: 615

The untold ugly truth in pediatric genital diseases(=child sexual abuse (CSA)):

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Introduction & Objectives:

Childhood sexual abuse (CSA) is a global public health concern. It is a prevailing problem in all generations, socioeconomic strata and societies, most of them are hidden & especially in pediatric age group where the victim is helpless, dermatologist are most of the time the first line of contact with these problems, and they should be aware of the alarming signs when they see them

Objective : to shed light on this unspoken about problem in our society by showing many cases seen in different presentations suggesting CSA, and guiding the dermatologists through steps needed for early diagnosis; with a review of the literature concerning this important subject

Materials & Methods:

Every day we are confronted with an increasing number of children being brought to dermatology clinics complaining of different types of STD's putting the dermatologist under pressure of how to deal with this perplexing situation.

Results:

Twenty-six children both males & females with different age groups from infancy to adolescence presenting with STD's affecting the genital area were studied thoroughly & documented, we tried to explore the truth behind the source of their infection but the social restrictions were immense making us stand helpless against such a cruel and tragic occurrence and a serious infringement of a child's rights to health and protection. We also reviewed the literature concerning this important hidden problem, the contributing factors, its long-term effects on the patient and his family

Conclusion: CSA does really exist in our society, the dermatologist; being the first to see the child with STD's has an additional responsibly toward the child & his family by being cognizant of the medical & behavioral indicators that signal early abuse

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Abstract N°: 733

What lurks beneath? A tuft of hair to the vertex scalp

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Introduction & Objectives:

An 8-month-old boy was referred to Paediatric dermatology with a scalp lesion present since birth. The lesion was stable in size, but caused discomfort when touched. There were no other health or developmental concerns, and the patient has two healthy older siblings. Examination revealed a 2x2cm pink, soft, non-mobile crateriform nodule to the vertex scalp. Protruding from the central crypt was a tuft of hypertrophic hair. A rim of hypertrophic hair also surrounded the nodule. The nodule was surrounded by a 5x5cm macular pink patch. Appearances were in keeping with a combination hair collar sign (HCS) and hair tuft sign (HTS). The patient was referred for urgent MRI imaging to evaluate any underlying CNS communication or defect. We review the literature regarding these clinical signs, as well as the possible underlying neuroectodermal defects.

Materials & Methods:

A search of PubMED and SCOPUS was undertaken using the search terms “hair tuft”, “hair tuft sign”, “hair collar”, and “hair collar sign”. Abstracts were screened for relevance.

Results:

An MRI scan demonstrated the heterogenous scalp soft tissue. This appeared to be associated with a small defect extending through the skull vault and into a diminutive falcine sulcus. Appearances favoured an atretic cephalocoele.

The Hair Collar Sign (HCS) is a collar of hair surrounding a lesion on the scalp, typically present from birth. It can be a marker of underlying neuroectodermal defects including cephalocoele or heterotopic brain tissue1. The Hair Tuft Sign (HTS) describes hair extruding from an orifice or a congenital midline defect of the scalp. These tufts may overlie atretic meningocoeles or meningeal heterotopia2. Though these signs are indicative of potential underlying neuroectodermal defects, the cutaneous findings cannot predict what the underlying abnormality may be2.

A review of 56 cases that underwent CNS imaging, found 28.6% to have a skull bone defect with direct communication with the CNS. 25% had venous anomalies, and 12.5% had CNS anomalies. The most common final diagnoses were meningeal heterotopia (37.5%), atretic meningocoele (25%). Rarer causes included sinus pericranii, sebaceous hamartoma, dermoid cysts, and membranous aplasia cutis2. There is just one case report of an atretic cephalocoele associated with the hair collar and hair tuft signs3.

Conclusion:

Atretic cephalocoele is a rarely reported association with the hair tuft and collar signs. MRI is recommended to evaluate skull soft tissue masses to screen for underlying CNS malformations. This case demonstrates how cutaneous findings can be indicative of underlying anomalies, and the important role that dermatology plays in recognising these signs and arranging appropriate investigation.

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Abstract N°: 742

Orofacial granulomatosis as the only presenting sign of Crohn's disease in a paediatric patient

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Orofacial granulomatosis (OFG) is a rare disease that could affect all age groups and races. It may exist as an independent disease or may be part of the clinical manifestations of Crohn's disease, sarcoidosis, or Melkersson-Rosenthal syndrome.1 More common in younger patients with most children diagnosed with OFG subsequently found to have with Crohn's disease.2 Clinically, it is characterized by persistent enlargement of the soft tissues of the lip, oral mucosa, and the area around the mouth. The treatment of orofacial granulomatosis is challenging.1,2

We present a case of orofacial granulomatosis as a first sign of Crohn's disease in a child to highlight this association.

A 10 year-old girl presented with a 3 months history of persistent lower lip swelling associated with crusting, intermittent oral ulcerations and right cheek swelling. She had no gastrointestinal symptoms and no family history of inflammatory bowel disease (IBD). Full blood count, iron profile and faecal calprotectin were normal.

On examination there was lower lip swelling associated with crusting, bilateral cheilitis and right cheek swelling.

She was diagnosed with orofacial granulomatosis. Gastroenterology (GI) assessment was negative for inflamatorry bowel disease, a watch and wait approach was recommended. She had a good symptomatic response to oral tetracyclines and a topical calcineurin inhibitor. Three years later she experienced progression of her lip and cheek swelling. Further GI review including endoscopy, confirmed the diagnosis of Crohn's disease.

We present this case to illustrate the orocutaneous manifestation of IBD and to highlight the time lag between oral and gastrointestinal manifestations. A high index of clinical suspicion and regular clinical follow up is required to confirm the diagnosis.

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Abstract N°: 838

Baby Eczema Regimen Provides Clinical Eczema Improvement and Skin Benefits in Subjects With Mild to Moderate Eczema

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Introduction & Objectives: Eczema is a multi-factorial skin disease, affecting up to 30% of children, and is associated with erythema, scaly and oozing lesions, and severe pruritus. Eczema sufferers experience impact to both their physical and emotional well-being. Emollients are known over-the-counter remedies to help manage eczema symptoms by treating dry skin, increasing hydration, improving skin barrier, and reducing itch. Using an ideal emollient for eczema sufferers can help lessen eczema flare-ups and improve overall Quality of Life (QoL). A consistent, effective regimen can keep baby skin well moisturized and can help lessen eczema flare ups, itching, and dryness.**

Materials & Methods: A routine of a non-drying, gentle oat-containing moisturizing wash (used at least 3 times per week) and a 1% colloidal oatmeal moisturizing cream (applied twice daily) was evaluated for its tolerability and its unique ability to balance improvements in mild-to-moderate eczema baby sufferers, with significant improvement in eczema symptoms, skin benefits and QoL.

Results: A 4-week clinical study was conducted in male and female subjects with investigator-assessed mild-to-moderate eczema across diverse skin tones (N=29, ages 3 months-6 years). A statistically significant reduction ($p<0.05$) in Eczema Area Severity Index (EASI) and Atopic Dermatitis Severity Index (ADSI) was demonstrated as early as day 1 of regimen use. Investigator evaluations found significant improvements ($p < 0.05$) in skin hydration, barrier function, itch, and skin pH in both lesional and non-lesional sites. Eczema-related QoL assessment demonstrated statistically significant improvement in subject emotional well-being (as early as day 1) as eczema symptoms improved.

Conclusion: The colloidal-oat containing product was deemed well-tolerated on pediatric subjects with mild-to-moderate eczema, across all skin tones. Skin condition, including eczema severity, eczema lesions, skin hydration, and skin barrier function improved with use of the product regimen. As eczema symptoms improved, positive responses for infant's quality of life indicated improvements in baby's overall mood, comfort and the overall impact of dermatitis. The results from this study suggest a consistent regimen of a gentle oat-containing moisturizing wash and a 1% colloidal oatmeal moisturizing cream can be beneficial in managing symptoms of eczema and improving QoL in a pediatric population as early as day 1 of regimen usage.**

EA DV CONGRESS
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EUROPEAN ACADEMY OF DERMATOLOGY & VENEREOLOGY

Abstract N°: 901

Crusted (Norwegian) Scabies in a four-Month-old boy due to the local application of corticosteroids for the wrong diagnosis

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Introduction :

Scabies is a global health problem affecting more than 300 million individuals annually, with the highest prevalence in children younger than 2 years.

Crusted scabies (CS) is a highly infectious hyperinfestation variant of scabies with up to millions of Sarcoptes scabiei mites present on the skin surface.

We present a case of a 4-month-old boy misdiagnosed for atopic dermatitis who recieved local corticosteroids causing scabies to evolve into crusted scabies.

Case report:

A previously healthy four-month-old boy was admitted to our service with a history of atopic dermatitis that had been present for two months,

At that time, he was treated by a topical application of corticosteroids daily , and instead of the improvement the lesions have spreaded to the entire body and became crusty, leading to his transfer to our dermatologgy department.

At the physical examination, the patient presented multiple papules, crusted and erythematous lesions disseminated in the body, mainly affecting the trunk and the scalp. Fissures in the back were also evident.

In the face of this situation, our main diagnostic hypothesis was crusted or Norwegian scabies.

The parents and the two brothers complained of important pruritus, and disseminated lesions compatible with scabie too.

A skin scraping was performed on the baby, and with microscopy, the presence of mites, eggs, and feces was confirmed.

The baby was treated with Benzyl Benzoate at Day 1 and Day 8, and the family members were treated with ivermectin; the entire house also disinfected. At the 3-week follow-up, we noticed the improvment of all family members.

Discussion:

Crusted scabies, is a rare and highly contagious form of scabies, characterized by the presence of numerous parasites in the horny layer of the skin.

In most cases, it is associated with an underlying disease and mainly affects immunocompromised individuals.

The clinical presentation of scabies varies with age and immunologic status (normal host vs hereditary or acquired immunodeficiency), which often makes early diagnosis and treatment difficult.

Crusted scabies is an extremely contagious disease which is rarely reported in infancy, especially in healthy children, In a frensh study Fifteen (75.0%) children were treated with steroids before being diagnosed with scabies.

In our case, the wrong diagnosis led to the application of corticosteroids, which led to local immunosuppression and the spreading of the scabies, causing a crusted scabies condition in a few months old boy. Fortunately, the diagnosis was made at the right time, and the child evolved well under local treatment.

Conclusion:

Crusted scabies is a rare condition and even rarer in young children. The wrong diagnosis of scabies in babies can lead to a misplaced application of corticosteroids that can make things worse by creating immunosuppression and the spreading of the disease.

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Abstract N°: 914

Diffuse cutaneous mastocytosis: a new case

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Introduction & Objectives: Mastocytosis is a rare disease characterized by the accumulation of mast cells in various organs. Unlike in adults, this condition is usually indolent in children, with purely cutaneous involvement. Here's a new case report**.**

Materials & Methods: The case involved a 16-month-old infant from a non-consanguineous marriage, with no notable pathological history. Since the age of 6 months, he had presented with pruritic macules, some of which were brownish and others erythematous, initially on the back, then spreading to the whole body. Dermatological examination reveals multiple coarsely rounded brownish macules affecting almost the entire integument, without sparing the face. Darier's sign was positive. The rest of the somatic examination was unremarkable. The diagnosis of diffuse cutaneous mastocytosis was evoked, then confirmed by histology with the demonstration of a dermal mastocytocytic infiltrate. Avoidance of mast cell degranulation factors, particularly dietary and medicinal, was instituted, and the patient was put on a combination of antihistamines and dermoscorticoids, with good progression.

Results: Mastocytosis is characterized by the accumulation of mast cells in various organs. They are linked to an acquired activating point mutation in the c-KIT receptor tyrosine kinase, which is also the receptor for the main mast cell growth factor. Clinically, Darier's sign (swelling of a skin lesion after friction) is pathognomonic.

Among purely cutaneous forms, the most frequent clinical form is urticaria pigmentosa and solitary mastocytoma, while diffuse cutaneous mastocytosis is rarer. The latter presents as a maculopapular rash, sometimes bullous, generalized, red in color and pigmentogenic in evolution. Diagnosis is based on skin biopsy. Treatment is based on controlling the symptoms triggered by mast cell degranulation, phototherapy, antihistamines and dermocorticoids, and new treatments aimed at directly inhibiting mast cell proliferation pathways may be offered to patients.

Conclusion: Mastocytoses are a heterogeneous group of disorders, of which diffuse cutaneous mastocytosis is rare and usually has a benign course.

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Abstract N°: 1084

Oral sildenafil for macrocystic lymphangioma in a Filipino child: a case report

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Introduction & Objectives:

Lymphangioma is a rare developmental anomaly of the lymphatic system, commonly arising at birth, in infancy, or early childhood. Treatment is rarely curative. Sildenafil is an emerging non-invasive treatment option which can decrease lymphangioma volume and provide symptomatic relief. Five studies on sildenafil use for treatment of lymphangioma are in publication, with none available locally.

Materials & Methods:

This report describes a 16-year-old Filipino male presenting with multiple discrete to coalescing, erythematous to violaceous papules and plaques, with grouped clear to hemorrhagic fluid-filled vesicles on the left lower extremity since early infancy.

Histopathology showed features consistent with lymphangioma circumscriptum. Over the years, lesions increased in number and size, with development of severe leg pain and difficulty in ambulation. MRI revealed dilated vessels, hence, diagnosis of macrocystic lymphangioma was made. Furthermore, lower limb length discrepancy of 1.5 cm and a larger circumference on the left leg were noted.

Patient was then started on sildenafil at 1 mg/kg/day equivalent to 25 mg per tablet twice a day for a duration of 9 weeks.

Results:

On follow-up after 2 weeks, symptomatic improvement was noted with decrease in pain from previous 5/10 to 2/10, with greater ease in ambulation and performance of physical activities. There were no new lesions noted. Measurement of limb length and leg circumference remained the same. After 4 weeks, measurements were again taken. Circumference of left leg decreased by 1.5 cm from 29 to 27.5 cm. Due to good response and tolerance of medication, interval of follow-up was increased to monthly. After 9 weeks of treatment, leg circumference further decreased to 26 cm, a reduction of 10% from baseline. Lesions were flatter, less erythematous, more hyperpigmented, with complete resolution of pain.

Table 1. Leg length, circumference and pain scale during treatment with sildenafil

	Initial consult	2 weeks	4 weeks	9 weeks
	R	L	R	L
Length (cm)	87.5	89	87.5	89
Circumference (cm)	28	29	28	29
Pain		5/10		2/10

Conclusion:

A case of macrocystic lymphangioma treated with sildenafil in a Filipino child diagnosed clinically, radiologically, and histologically was presented. Given the recurrent course of this condition, even with surgical management, the use of a non-invasive approach such as sildenafil as potential treatment option for lymphangioma is an important emerging area of study.

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Abstract N°: 1091

idiopathic facial aseptic granuloma: a pediatric entity to keep in mind

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Introduction & Objectives:

Idiopathic aseptic granuloma of the face or cold pyoderma corresponds to a single, chronic, painless inflammatory nodule localized to the cheek, more rarely to the eyelid,

We report the case of a child with a nodule of the face who was wrongly treated with antibiotics.

The need for early diagnosis to avoid topical and/or systemic antibiotic treatment, which could lead to antibiotic resistance, especially as the duration of the disease does not appear to be shortened by the use of antibiotics.

Materials & Methods:

Patient R.Z, aged 4, presented with a nodule of the face that had been evolving for 6 months.

The patient was initially treated with local antibiotics, followed by well-conducted oral antibiotics, without any improvement.

Clinical examination revealed a single nodule on the face with an inflammatory appearance, purplish red in color, no telangiectasia, no papulopustular lesions and no ophthalmological signs consistent with rosacea.

All this evolved in a context of apyrexia and preservation of general condition.

Dermoscopy showed erythema with perifollicular hypopigmentation and follicular obstruction.

A bacteriological sample was taken with no abnormalities.

Results:

The diagnosis of idiopathic aseptic granuloma of the face was made based on the cold nature of the nodule, its chronic course, the negativity of bacteriological samples and the spontaneous regression of the nodule.

Cold pyoderma is a rare pediatric entity, first described in 1999. It typically presents as a single nodule, usually on the face, although other localizations have been reported in the literature, notably of the lower eyelid.

Bacteriological samples are always negative, as is skin biopsy.

In general, treatment consists of therapeutic abstention.

A case of a 7-year-old boy with trisomy 21 with an idiopathic facial aseptic granuloma of the cheek has been reported, successfully treated with low dose of isotretinoin.

The disease evolves chronically over several weeks to months, with spontaneous healing and sometimes scarring.

Some children may go on to develop rosacea.

Conclusion:

The importance of early diagnosis of this pathology lies in preventing antibiotic resistance and following up these children to detect any associated infantile rosacea.

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Abstract N°: 1116

Ocular Clues: Diagnosing Childhood Rosacea through Manifestations in the Eye

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Ocular Clues: Diagnosing Childhood Rosacea through Manifestations in the Eye

Introduction & Objectives:

Rosacea is a chronic inflammatory skin disease, of unknown etiology, more commonly seen in middle-aged women and it has rarely been reported in children. It is characterized by redness, erythema, telangiectasia, papules, pustules, glandular hypertrophy and/or phymas, in the central convex areas of the face. Ocular symptoms include photophobia, redness of the eyelids and conjunctiva, foreign body sensation, ocular burning, itching, lid hyperemia or recurrent chalazia. The delay in diagnosis of childhood ocular rosacea has been reported up to 7 years in the literature. Some authors have suggested criteria for the diagnosis of childhood rosacea, which are important to take into account since it allows us an early and accurate diagnosis, to avoid complications or sequelae.

Results:

A 2-year-old female patient, is brought by her mother to the clinic due to a three-month history of persistent and painful “little red balls” on her eyelids.

She had initially attended pediatric ophthalmology where she was diagnosed with chalazion and started treatment with antibiotic drops, without improvement and development of new lesions on cheeks described as “red dots”. Therefore, they went to an oculoplastic surgeon, who referred the patient to pediatric dermatology with suspicion of childhood rosacea.

On physical examination, the patient presented with labile and irritable crying, four erythematous papules on the right implantation edge of the upper and lower eyelashes and one in the left. Accompained with blepharitis, conjunctival erythema, countless erythematous papules and pustules on the cheeks, persistent malar erythema that spared the nasolabial fold accompanied by few fine telangiectasias evident on dermoscopy.

Based on the dermatological and ocular findings on physical examination, it was considered that the patient met criteria for infantile rosacea. Treatment was started with metronidazole gel 0.75% applied every night on the face, tacrolimus ointment 0.03%, 3 times a week on the face, azithromycin 4.5 cc orally for 8 weeks and daily sunscreen. We request a follow-up with pediatric ophthalmology and follow-up in one month in pediatric dermatology.

At the follow-up, the patient´s mother comments that ophthalmology started treatment with dexamethasone and ciprofloxacin in 0.3% eye drops for 10 days due to the presence of staphylococcus in culture of ocular secretion. She also rates an-80% improvement in her daughter’s clinical evolution. On physical examination, the patient is calm and smiling, she has two little red papules on the right lower eyelid, no lesions on the left, no conjunctival erythema, persistence of malar erythema without the presence of papules or pustules on the cheeks.

Conclusion:

The prevalence of ocular involvement in children is unknown. Eye involvement in children is associated with a

significantly major complications like corneal opacities and decreased visual acuity. That is why in addition to topical treatment, they must receive systemic treatment with antibiotics to avoid ocular sequelae and relapses. The prevalence of ocular involvement in children is unknown. Eye involvement in children is associated with a significantly major complications like corneal opacities and decreased visual acuity. That is why in addition to topical treatment, they must receive systemic treatment with antibiotics to avoid ocular sequelae and relapses.

EADV CONGRESS
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Abstract N°: 1199

Juvenile dermatomyositis: a severe case report

Ines Boudjabi*[1], Zaamouch Imene[1], Benchiheb Azzedine[1]

[1]University Hospital Abdesselam Ben Badis , pediatric , Constantine, Algeria

Introduction & Objectives:

Juvenile dermatomyositis (JDM) is a rare, often chronic, autoimmune disease characterised by muscle and skin involvement. It is the most common idiopathic inflammatory myopathy of childhood. Unlike adult-onset dermatomyositis, there is no association between malignant tumors and anti-TIF1γ antibody positivity. We report a case of a girl who presented this pathology in its severe form.

Materials & Methods:

A 10-year-old girl with no specific pathological history was referred to us for an onset motor deficit that had been evolving for one month.

Clinical examination: showed a symmetrical proximal muscle weakness predominantly in the limb-girdle region, with asthenia, myalgias, dysphonia, dysphagia, discharge of fluids through the nostrils and ineffective cough.

Dermatological examination revealed: macules arranged in bands on the backs of the hands opposite the metacarpophalangeal and interphalangeal joints (Gottron's sign).

Laboratory investigations revealed elevated serum levels of: creatine phosphokinase (CPK), lactate dehydrogenas (LDH), aspartate aminotransferase (AST), alanine aminotransferase (ALT) and troponin.

Electromyography was suggestive of a proximal-distal myogenic involvement. Muscle biopsy showed necrosis, inflammation and muscle regeneration. In spirometry: a restrictive ventilatory defect.

Based on Bohan and Peter's criteria the diagnosis of Juvenile dermatomyositis was made.

Results:

The goals of treatment are to control inflammatory myositis and prevent disease complications. There are no adequate controlled trials in the management of Juvenile dermatomyositis. Thus treatments are based on clinical experience. Treatment with high-dose oral Corticosteroids alone or in combination with in combination with another immunosuppressive medication (most commonly methotrexate) is the cornerstone of management.

In our patient's case, partial improvement (disappearance of dysphagia) was first obtained under corticosteroids with intravenous immunoglobulin infusion, then progressive and total (clinical and biological) after simultaneous use of methotrexate, corticosteroids and Physiotherapy.

Conclusion:

Juvenile dermatomyositis is a rare entity. The use of immunosuppressants such as Methotrexate provides a beneficial therapeutic complement.

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Abstract N°: 1202

Giant tuberous and tendinous xanthomas in a consanguineous marriage

Marwa Meribout*[1], Tarek Mansoul[1], Riadh Boussaid[1], Ahmed Samaouel Chehad[1]

[1]University Hospital Abdelhamid Ben Badis, Dermatology departement, Constantine, Algeria

Introduction & Objectives:

Materials & Methods:

Results:

INTRODUCTION:

Xanthomas are benign orange to yellowish tumors, due mostly to primary or secondary lipid metabolism dysfunction. They can reveal familial hypercholesterolemia where early diagnosis is crucial in order to prevent complications such as cardiovascular problems.

We report a case of tuberous and tendinous xanthomas which revealed familial dyslipidemia.

OBSERVATION:

An 8-year-old girl, of consanguineous marriage, living in a rural place, without familial antecedents, was oriented to our department for a 4-year history of big nodules over elbows and knees. The patient had multiple nodules of different sizes (the biggest was 4 X 5 cm), yellowish, soft, compressible and painless, extended on elbows, knees, buttocks, legs and Achilles' tendon. Moreover, she had limited mobility in large articulations due to the nodules.

Blood lipid test revealed a very high level of cholesterol (Cholesterol = 7.91 g/L, elevated LDL, but normal triglycerides and HDL levels). Complete blood count and thyroid examination were normal. Biopsy demonstrated a dermic infiltration by foamy histiocytes.

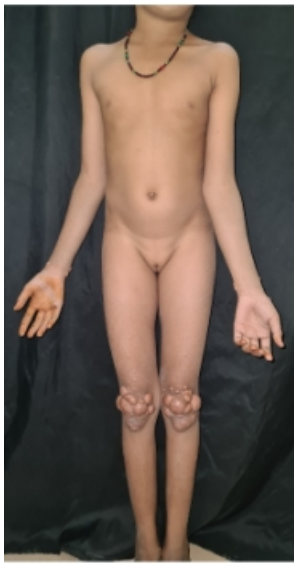
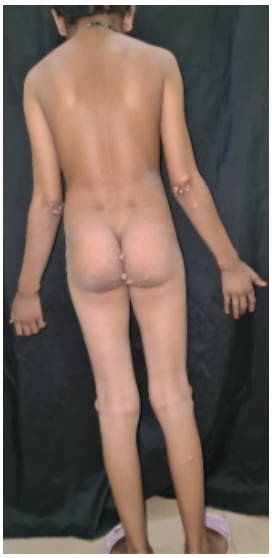
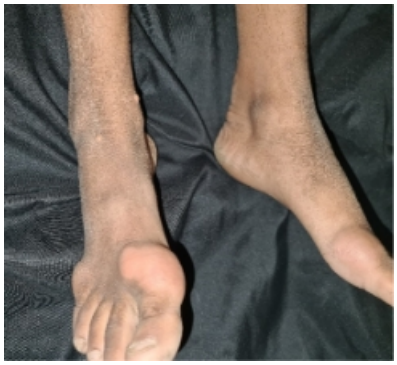
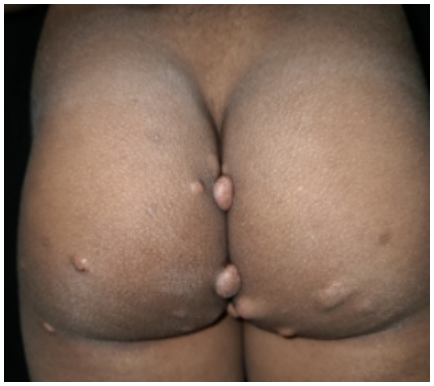
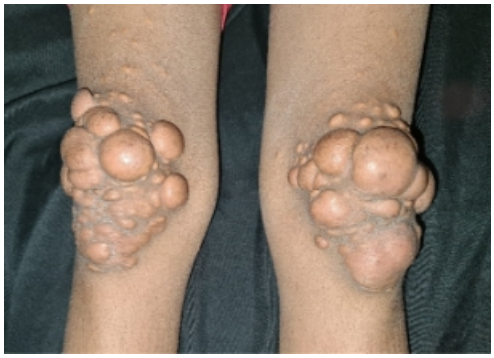
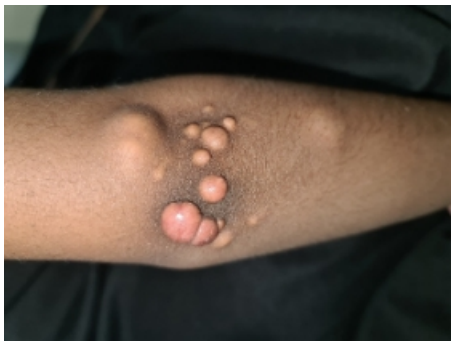
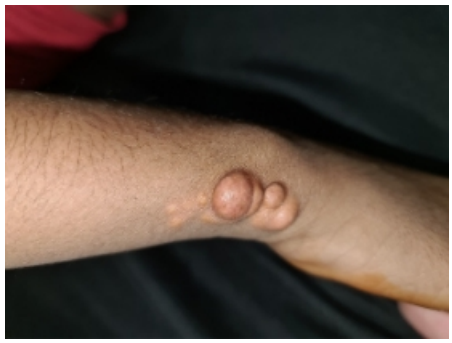
Diagnosis of Tuberous and tendinous xanthomas in the context of familial hypercholesterolemia (FH) was made and the patient was oriented to the endocrinology department for further evaluations.

DISCUSSION:

FH is a genetic disease due to LDLRAP1 gene mutation which encodes for « low-density lipoprotein receptor adaptor protein 1 » leading to high cholesterol blood level. It's has usually autosomic dominant transmission, rarely autosomic recessive. Our case is probably autosomic recessive or sporadic due to a negative family history. However, molecular genetic study is required for confirmation. FH can manifest through various clinical aspects of xanthomas including xanthelasma, and corneal arch, tuberous and tendinous xanthomas as in our case,

In addition to aesthetic and functional impact of xanthomas, FH can cause high morbimortality rate. In order to prevent cardio-vascular complications in young patients, early management by low fat diet along with hypocholesterolemiant therapy (statins or cholestyramine) is mandatory. Tuberous xanthomas are usually managed by laser, electrosurgery or conventional excision surgery for large lesions.

Conclusion:

EADV CONGRESS
AMSTERDAM 25-28 SEPTEMBER 2024
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Abstract N°: 1214

A Rare Case Report: Waardenburg Syndrome

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[1]Military Hospital Mohammed V, Dermatology Venerology, RABAT

Introduction & Objectives:

Waardenburg syndrome (WS) is a rare autosomal dominant disorder characterized by a spectrum of manifestations stemming from neural crest abnormalities. Initially described by Dutch ophthalmologist P.J. Waardenburg in 1951. It encompasses a diverse range of clinical features, including sensorineural hearing loss, pigmentary disturbances of the skin, hair, and iris, and dystopia canthorum. Despite its rarity, Waardenburg syndrome is genetically heterogeneous, with four recognized subtypes based on clinical presentations. We report a very interesting case of type 1 WS.

Materials & Methods:

A 7-year-old boy presented with piebaldism-like hypopigmented macular lesions over the forehead, middle parts of the forearms, and legs and decreased hearing in both ears since birth. Clinical examination showed confluent achromic macules and poliosis with no contrast enhancement under the Wood lamp. The audiometry revealed profound bilateral sensory-neural deafness. The ocular examination revealed dystopia canthorum, a broad nasal root, and heterochromia of the iris. The right iris was black, while the left was hazel. The central nervous system, musculoskeletal, and other systemic examinations were normal. The patient has two other normal siblings and normal parents in a consanguineous marriage. There is a family history of similar symptoms in the patient's extended family, including heterochromia iridis, premature graying of hearing, white forelock, and sensory neural deafness in various combinations.

Results:

Waardenburg syndrome is an uncommon autosomally inherited and genetically heterogeneous disorder of neural crest cell development with four recognized subtypes based on clinical presentations. The classic form is type I; dystopia canthorum is absent in type II; and limb abnormalities and dystopia canthorum are linked to type III. A variation known as type IV has been linked to Hirschprung's disease. The clinical presentation of the patient aligns with the classical features of Waardenburg syndrome. The presence of piebaldism-like hypopigmented macular lesions, profound bilateral sensorineural deafness, dystopia canthorum, broad nasal root, and heterochromia iridis, along with a positive family history, strongly supports the diagnosis of Waardenburg syndrome type 1. Additionally, the absence of contrast enhancement under the Wood lamp and normal findings on ocular examination, apart from heterochromia iridis, further support the diagnosis.

Conclusion:

This case report highlights the clinical presentation, genetic basis, and familial implications of Waardenburg syndrome. Early recognition and diagnosis of Waardenburg syndrome are crucial for appropriate management and genetic counseling, emphasizing the importance of awareness among clinicians to facilitate timely intervention and support for affected individuals and families.

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Abstract N°: 1226

Juvenile dermatomyositis: a severe report case

Ines Boudjabi[1], Zaamouch Imene[1], Benchiheb Azzedine[1]

[1]University Hospital Abdesselam Ben Badis , pediatric , Constantine, Algeria

Introduction & Objectives:

Juvenile dermatomyositis (JDM) is a rare, often chronic, autoimmune disease characterised by muscle and skin involvement. It is the most common idiopathic inflammatory myopathy of childhood. Unlike adult-onset dermatomyositis, there is no association between malignant tumors and anti-TIF1γ antibody positivity. We report a case of a girl who presented this pathology in its severe form.

Materials & Methods:

A 10-year-old girl with no specific pathological history was referred to us for an onset motor deficit that had been evolving for one month.

Clinical examination: showed a symmetrical proximal muscle weakness predominantly in the limb-girdle region, with asthenia, myalgias, dysphonia, dysphagia, discharge of fluids through the nostrils and ineffective cough.

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Laboratory investigations revealed elevated serum levels of: creatine phosphokinase (CPK), lactate dehydrogenas (LDH), aspartate aminotransferase (AST), alanine aminotransferase (ALT) and troponin.

Electromyography was suggestive of a proximal-distal myogenic involvement. Muscle biopsy showed necrosis, inflammation and muscle regeneration. In spirometry: a restrictive ventilatory defect.

Based on Bohan and Peter's criteria the diagnosis of Juvenile dermatomyositis was made.

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In our patient's case, partial improvement (disappearance of dysphagia) was first obtained under corticosteroids with intravenous immunoglobulin infusion, then progressive and total (clinical and biological) after simultaneous use of methotrexate, corticosteroids and Physiotherapy.

Conclusion:

Juvenile dermatomyositis is a rare entity. The use of immunosuppressants such as Methotrexate provides a beneficial therapeutic complement.

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Abstract N°: 1634

A Case of Hypohydrotic Ectodermal Dysplasia in a 1-year old Filipino male

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Introduction & Objectives:

Hypohidrotic Ectodermal Dysplasia (HED) is the most common Ectodermal Dysplasia, a group of rare, inherited multisystemic disorders exhibiting developmental aberrations in ectodermal structures including the hair, teeth, nails, and sebaceous and sweat glands. HED is commonly X-linked, presenting with abnormalities of the hair and teeth accompanied by inability to sweat. This case report aims to present a case of X-linked HED in a 1-year-old Filipino male.

Materials & Methods:

This is a case report of a 1-year-old Filipino male with a chief complaint of reduced sweating since birth and sought consultation at the Dermatology out-patient clinic of a tertiary government hospital in the Philippines.

Results:

Upon history taking and physical examination, he had scalp and facial papules, slowly growing, sparse eyebrow and scalp hair, periorbital wrinkling with hyperpigmentation, everted lips, frontal bossing, absent tooth, and negative palmoplantar starch-iodine test. Maternal male relatives had similar symptoms while females had little to no symptoms. He was diagnosed with HED. Skin punch biopsy with immunohistochemistry showed absence of eccrine glands on the plantar skin, which strengthened the diagnosis. Management of symptoms was mainly supportive and involved multidisciplinary collaboration with Pediatrics, Otorhinolaryngology, Dentistry, and Nutrition and Dietetics.

Conclusion:

This report underscores the critical role of Dermatologists in identifying key clinical signs of HED, which primarily relies on clinical assessment. A high level of suspicion is warranted when abnormalities in hair, teeth, and sweating are evident. It is crucial to consider other potential features in patients with symptoms of hypotrichosis, hypodontia and hypohidrosis, due to the varied manifestations of the disease. Early detection, comprehensive supportive treatment, and patient education are vital for improving the quality of life for those affected by HED.

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Abstract N°: 1796

Neonatal lupus : a case report

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Introduction & Objectives:

Neonatal lupus is a rare autoimmune disease resulting from the transplacental passage of maternal autoantibodies to the fetus. This condition develops in the fetus following the transplacental transmission of specific maternal antibodies such as anti-SSA/Ro, anti-SSB/La, or anti-ribonucleoprotein U1 (anti-U1-RNP) autoantibodies. We report the case of an infant born to a mother with systemic lupus erythematosus.

Materials & Methods:

A 4-month-old female infant began presenting with erythematous and scaly patches on her face for the past month. Additionally, she exhibited some annular scaly patches with erythematous borders and a clear center, extending to the folds, back, abdomen, and scalp. The rash was asymptomatic, with no reported itching or discomfort by the mother. Her mother had been diagnosed with systemic lupus erythematosus and had been treated with corticosteroids and hydroxychloroquine for 3 years. Laboratory tests showed elevated levels of anti-SSA and anti-SSB antibodies, with no abnormalities in the blood panel. Electrocardiogram and echocardiogram results were normal. The diagnosis of neonatal lupus was established. The rash improved with the use of topical corticosteroids and sun avoidance.

Results:

Neonatal lupus is an acquired condition that occurs in 20,000 newborns born to mothers with autoimmune diseases who transmit IgG autoantibodies transplacentally. The most common clinical manifestations of neonatal lupus are cutaneous manifestations, transient anemia, and rarely, complete heart block. The rash typically consists of erythematous annular lesions with slight central atrophy and raised margins, mostly localized on sun-exposed areas, particularly on the head and neck, less commonly on the trunk, and extremities.

Neonatal lupus can develop in newborns whose mothers are carriers of anti-SSA/Ro or anti-SSB/La antibodies and suffer from various connective tissue systemic diseases. However, in half of the cases, the mothers are asymptomatic, which may delay diagnosis and have a negative impact on the child’s prognosis.

Conclusion:

Neonatal lupus is a rare condition that can be life-threatening for the newborn. It is recommended to consider screening for antinuclear antibodies in every pregnant woman, as early treatment with hydroxychloroquine or intravenous immunoglobulins has been shown to be effective in preventing congenital heart block.

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Abstract N°: 1817

Acral skin peeling syndrome - a rare entity

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Introduction & Objectives: Levy and Goldsmith described peeling skin syndrome (PSS) in patients with continuous peeling of skin. Acral peeling skin syndrome (APSS), first described by Fox, is rare, autosomal recessive disorder caused by mutations in transglutaminase-5 (*TGM5*) gene. Prevalence is unknown, with no cases from India. It occurs due to separation of stratum corneum from stratum granulosum and is limited to dorsa of hands and feet. In majority, it develops shortly after birth. Symptoms are aggravated by heat, humidity and friction. Dermoscopy reveals structureless white areas and white/red globules. Histopathology findings are mild hyperkeratosis, splitting of epidermis between stratum granulosum and corneum. Management is largely symptomatic and preventive with emollients being mainstay of treatment.

Materials & Methods: A 2-year-old girl, born of non-consanguineous marriage, with normal mile stones and immunized till date, presented with asymptomatic, well demarcated, mildly hyperpigmented patches of peeling of skin, 5 x 6 cm on extensor aspect of bilateral forearms (between elbow to wrist joint), arms (left more than right) and 5x4 cm on lower limbs (between knee joint to ankle joint) with overlying small areas of peeling with complete sparing of dorsal aspect of hands and feet since 1 month. There was no similar involvement in past/family. Dermoscopy showed white globules, white structureless areas, some surrounded by hyperpigmented scales suggestive of PSS. She was treated with bland emollients with improvement.

Results and conclusion: APSS is a rare disorder which should be kept as a differential diagnosis for exfoliative diseases. Dermoscopy aids in diagnosis giving away need for biopsy.

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Abstract N°: 1898

Herpes Zoster in an infant in the post-covid era: Dermatoscopy with review

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Introduction & Objectives: Herpes zoster, a painful vesicular dermatomal eruption, is the result of reactivation of the varicella-zoster virus (VZV) from infected sensory ganglia. Traditionally, it is considered to be a disease of adults, in contrast to primary infection with VZV, which tends to occur mainly in children.

Materials & Methods: We report a case of infantile herpes zoster in healthy immunocompetent child at 7 months of age. Mother had primary varicella infection at 6th month of gestation. The infant presented with multiple vesicles in the trigeminal dermatome with erythema since 1 day. Symptomatically, the child did not have any fever and was not irritable. There were no similar vesicles anywhere else on body. Tzanck smear from the base of the vesicle showed multinucleated giant cells which supported the diagnosis of HZ in a child with vesicular rash.

The dermoscopic examination was done with DermLite DL4 in the polarized mode. Evolving lesions showed pale pink polylobular cloudy structures. Each lobule consisted of central brown globules with a grayish zone, which in turn was surrounded by a pale pink area with a grayish rim. Many coalesced lobules demonstrated brown and gray dots in the center.

As the child presented to us before 72 hours of skin eruptions, he was treated with oral acyclovir at the dose of 20 mg/kg/dose, five times a day for seven days along with topical antibiotic (fusidic acid cream).

Results: In infant, HZ generally occurs due to the reactivation of VZV acquired in utero from maternal varicella. The lesions of the child resolved completely in a weeks time. There were no secondary complications

Conclusion: Herpes zoster in infancy is rare but well described following intrauterine exposure to varicella-zoster virus (VZV). We hypothesize that this condition is rarely recognized because of the mild clinical manifestations in this age group and the expectation that maternal antibodies will be protective. It is likely that the vesicular lesions of herpes zoster in this age group are misdiagnosed as impetigo or other cutaneous disorders. With a high degree of suspicion, the dermatomal distribution of a vesicular eruption in infancy should point the clinician toward a correct diagnosis of herpes zoster. Maternal VZV antibody in infant offers protection for few months but the level decreases rapidly, and after 4 months, they remain no longer protected.

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Abstract N°: 1955

"Blood in the Sweat: An Uncommon Phenomenon in Hematohidrosis"

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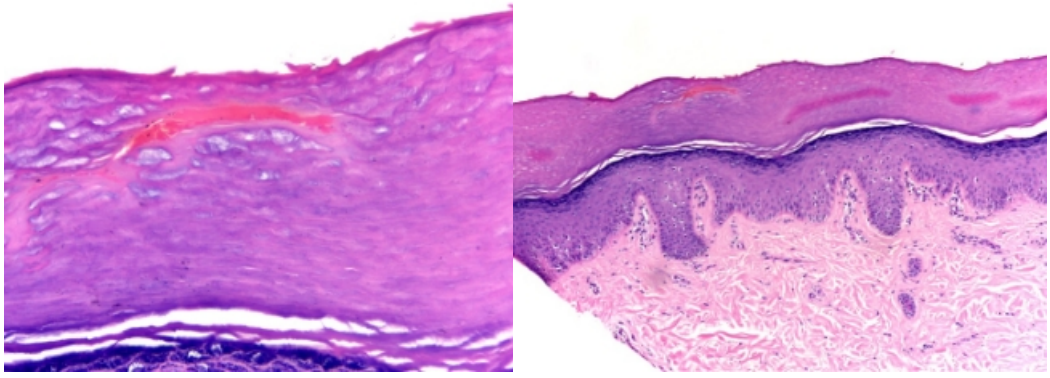
Introduction:

Hematohidrosis is a very rare clinical entity that consists of sweating blood. Its etiology has been unknown until now. The following case report presents a pediatric patient with hematohidrosis, demonstrated by a skin biopsy with findings of blood in the stratum corneum. There are currently no case reports with these findings.

Case Report:

An eight-year-old female patient, taken by her mother to the dermatology clinic, has been experiencing bloody sweating on her hands for 10 days. The mother reports three episodes of sweating in half an hour and more than ten at night. She also had an occasion of bleeding on her tongue. She denies any history of trauma in the affected areas. Additionally, the mother reports that she has been in a divorce process with the patient's father for a few months, and that this has caused family upheaval. At the time of the physical examination, her skin was intact. Blood tests showed platelet count and coagulation tests without abnormalities. A biopsy of the skin of the palm was taken with a punch, which resulted in the presence of bloody material in the stratum corneum, without obvious alterations in the glandular or capillary structures (Figure 1). Hematohidrosis was diagnosed and treatment with metoprolol and psychological support was prescribed, with which the patient improved.

Figure 1:



Discussion:

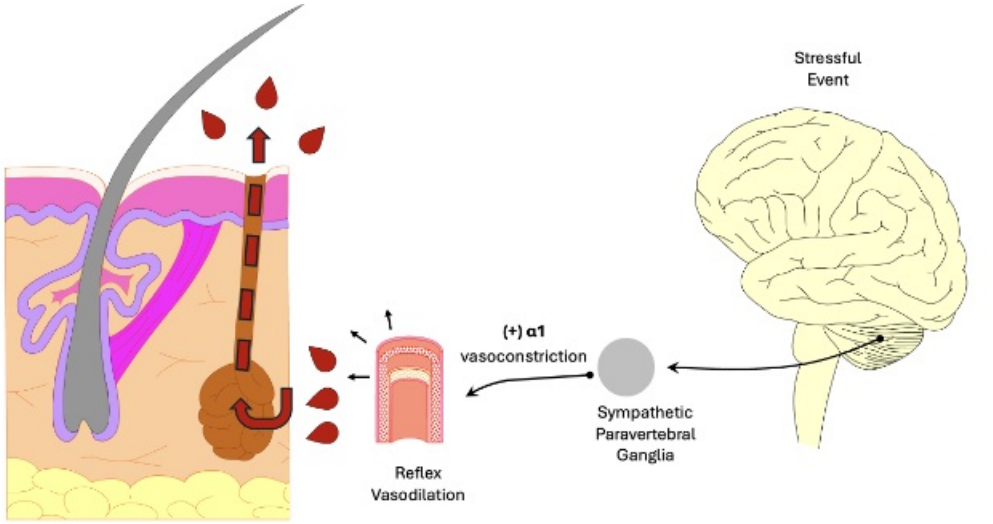
Hematohidrosis is a little-reported clinical enigma. In the literature, possible etiologies have been described such as systemic diseases that are associated with hemorrhage at other sites (rectal, vaginal bleeding, and sweat in tears), thrombotic thrombocytopenic purpura, and psychiatric disorders.

Multiple psychiatric disorders have been described as possible etiologies. Extreme mental stress has been reported in men sentenced to execution and in other stressful situations such as rape. Psychogenic purpura occurs where the blood itself generates a hypersensitivity reaction and causes a picture of ecchymotic lesions. Altered

psychological states, such as disorders in relationships with family members and bonds in pediatric patients, need for recognition in adolescents and anxiety in caregivers, can also be considered possible etiologies.

The proposed physiopathology consists of the fact that the blood vessels located around the sweat glands are organized to form a network, and that – under conditions of stress – pressure is generated and the vessels contract and then dilate and rupture in response to sympathetic activation. The extravasated blood reaches the sweat glands which is propelled, along with sweat, towards the epidermis (Figure 2).

Figure 2:



To diagnose the disease, disorders such as bleeding diathesis or dermatological diseases that could explain it must be ruled out. Currently, there are no effective treatments, but in the majority of the cases reported, support with psychological and psychiatric therapy has been shown to improve the episodes. Beta blockers have been used to inhibit sympathetic action or local administration of adrenaline to reduce vascular tone. Anxiolytics and antidepressants are also other options.

Conclusion:

Hematohidrosis is a rare entity little reported in the literature. The first case is reported with blood in the stratum corneum, possibly explained by anatomical alterations of the sweat gland vessels.

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Abstract N°: 2070

Adolescent with alopecia areata and concomitant atopic dermatitis recalcitrant to baricitinib treated successfully with upadacitinib

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Introduction & Objectives:

Upadacitinib is a selective Janus Kinase 1 (JAK1) inhibitor that was recently approved by the FDA for moderate-to-severe atopic dermatitis (AD) for adults and adolescents (above 12 years of age). JAK inhibitors (tofacitinib, ruxolitinib and baricitinib) are also proven to be effective in the treatment of alopecia aerate (AA). Data regarding upadacitinib as a treatment option for AA is starting to emerge, as well as for patients with concomitant AA and AD. However, data regarding this treatment in the pediatric population is lacking.

Materials & Methods:

Case report: 12-year-old boy with life-threatening peanut and milk allergy presented to the Pediatric Dermatology Clinic with severe AD and widespread multifocal AA. He was diagnosed with AD in infancy and with AA when he was 11-year-old. Topical therapy did not improve both conditions. Systemic therapy for AD included cyclosporine for several exacerbations which caused headaches. Methotrexate was attempted and stopped due to significant increase in liver enzymes, nausea and vomiting. Later on dupilumab was introduced but had only mild effect on his AD and resulted in severe conjunctivitis and worsening of AA. Baricitinib treatment was subsequently given for a period of 7 months (as part of a clinical trial), unfortunately without substantial improvement of both diseases. Due to the recalcitrant nature of the disease and the significant deterioration in the alopecia, upadacitinib 15 mg/d was initiated.

Results:

Upadacitinib led to an immediate and remarkable improvement. Complete hair regrowth was noticed only 8 weeks after commencing upadacitinib 15 mg/day. The remission in the alopecia was persistent with complete hair growth. However, after 9 months, due to insufficient control in the AD, upadacitinib dosage was increased to 30mg/day, with this dosage both conditions were controlled. The alopecia did not remit, and the excellent results were constant during 18 months of follow up. In addition, no side effects were observed with either dosage.

Data regarding upadacitinib as a treatment option for AA is lacking both in adults and especially in children. To date, only one additional case of an adolescent male demonstrated the efficacy of upadacitinib in patients with concomitant AD and AA. Both patients had a remarkable improvement with complete regrowth of all hair in short period (6 and 8 weeks). In addition, both patients had a substantial improvement in the AD status and responded well without any adverse effect to the treatment with upadacitinib. To our knowledge, our patient is the first reported pediatric patient with concomitant AD and AA recalcitrant to baricitinib that was treated successfully with upadacitinib.

Conclusion:

The newly introduced upadacitinib may hold the potential to treat both AA and AD in adolescent patients.

In light of the rapid, complete and persistence response we recommend treating adolescent patients with

concomitant AA and AD initially with upadacitinib 15mg/day. Later on, if not sufficient 30mg/day dosage should be tried. We believe this unique patient population with concomitant AA and AD can benefit greatly from this treatment. Additional prospective controlled studied should check the safety and efficacy for patients with both AD and AA and most importantly the safety profile and long-term side effect for the pediatric population.

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Abstract N°: 2142

Facial Lipoblastoma in a child

Maryem Aboudourib[1], Abid Meriem[1], el Atiqy Oum Keltoum[2], Amrani Moulay Driss[2], Benchemkha Yassine[2], Hocar Ouafa[1], Said Amal[1]

[1]Department of Dermatology and Venerology, Mohammed VI University Hospital, Bioscience and Health Laboratory, Cadi Ayyad University, FMPM, Department of Dermatology and Venerology, , MARRAKESH, [2]plastic surgery department,Mohammed VI University Hospital, , Cadi Ayyad University, FMPM, Morocco

Introduction & Objectives:

Lipoblastoma is a rare benign tumor arising from embryonic white fat which occurs in the early childhood. It usually arises on the extremities and considered as a rare cause of a pediatric head and neck masses. Thus, we present a case of a pediatric lipoblastoma of the cheek.

Materials & Methods:

A 4-year-old female presented with a slowly enlarging left cheek mass. growing progressively since birth. There was no history of trauma or infection. The mass was firm, mobile, non-pulsatile and painless causing noticeable facial deformity and skin heterogeneous pigmentation with cutaneous vascular dilatation. The size of the mass was approximately 4,5× 4 cm. Examination of the oral cavity was normal. No adenopathy or cranial nerves deficit was observed

Results:

Magnetic resonance imaging (MRI) showed an encapsuled mass, that measured 4,4 cm3.8 cm, with hyperintensity on axial T2 and T1, and reduced signal at fat suppression sequences demonstrating the fatty component of the lesion. The mass was crossed by internal septa with serpiginous circulation from the arteries. The tumor was completely removed by surgery. The pathologist confirmed that the mass was a lipoblastoma.

Results:

Lipoblastoma is a rare, benign adipocytic tumor that presents in children under 10 years old and has a preponderance for boys. The pathogenesis is unknown, though they are believed to arise from altered embryogenesis of human white fat and genetic predisposition, as chromosome 8 abnormalities may be implicated in the development of lipoblastoma. Usually, it presents as a painless subcutaneous soft tissue mass growing progressively. Symptoms are related to the location and size or mass effect of the lesion. Its rapid growth may cause compressive symptoms caused by airway obstruction or nerve compression. In our case, despite the tumor's size, there were no compression symptoms.

This tumor often arises in subcutaneous tissues of the trunk and extremities, but rarely involves the face and neck, as in the present case. Treatment of lipoblastoma is complete surgical excision without adjuvant therapies as seen with this child. Since this is a benign tumor, sub-total resection can be considered to avoid damaging important structures such as Stensen duct or branches of the facial nerve.

Conclusion:

Lipoblastomas are rare adipocytic tumors of the head and neck that should be considered in rapidly growing masses in children less than 10 years old. Imaging is important for surgical planning, but the definitive diagnosis is

made with histopathology

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Abstract N°: 2177

Blastic plasmacytoid dendritic cell neoplasm with skin compromise in children: A case series.

Yamila Babbini[1], María Buján[1], Paola Stefano[1], Marcela Bocian[1], Aldana Villagra[1], María Centeno[2], Cristian Sánchez La Rosa[3], Andrea Cervini[1]

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[2]"Prof. Dr. Juan P. Garrahan" Pediatrics Hospital, Department of Pathology, Buenos Aires, Argentina,
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Introduction & Objectives: Blastic plasmacytoid dendritic cell neoplasm (BPDCN) has been described as an aggressive hematological malignancy derived from plasmacytoid dendritic cells. It accounts for 0.7% of cutaneous lymphomas in children, affecting more frequently the skin and bone marrow, lymph nodes and the central nervous system. BPDCN presents as deep purple or brown patches, plaques, nodules, or tumors on the skin. The diagnosis is based on histopathology and mainly on immunohistochemistry. Immunophenotypic profiles are typically positive for CD4, CD56 and CD123, as well as TdT, CD43, BCDA2, TCL1 and HLA-DR. In pediatric cases, BPDCN is characterized by less aggressive clinical courses with a more favorable prognosis than adults. Although there is no standard treatment for childhood BPDCN, treating initially with acute lymphoblastic leukemia (ALL)-type chemotherapy schedule is seemingly effective.

To describe the epidemiological, clinical, and histological characteristics, as well as complementary studies, treatment and outcomes of patients with BPDCN seen at our department.

Materials & Methods: Descriptive, observational, cross-sectional, and retrospective study. The medical records of patients with a diagnosis of BPDCN confirmed by histopathology, immunohistochemical and/or flow-cytometry studies evaluated at the Department of Dermatology of a National Pediatric Referral Center from March 2008 to February 2024 were analyzed.

Results: Four patients were diagnosed with BPDCN. Two were female and the median age at diagnosis was 14 (6-15) years old. Two patients had previous antecedent: one patient had been treated as bilateral retinoblastoma resulting in the enucleation of the left eye and chemotherapy, while the other had right microtia with agenesis of the external auditory canal. The cutaneous presentations encompassed violaceous blue tumors observed in two patients, violaceous-red, indurated plaques in one patient, and violaceous-red nodules in another. Two patients showed localized lesions affecting the extremities and the other two patients had disseminated lesions. Two patients associated bone marrow dissemination, and one of them also had bone affection. None had central nervous system involvement. All cases tested positive for CD56 and CD123, with half displaying positivity for CD4 and the other 2 showed positivity for TDT. All patients received chemotherapy treatment according to ALL/NHL protocols achieving complete remission and remaining disease-free at a median of 20 (9-191) months. None of the patients died.

Conclusion: BPDCN is a rare malignancy that commonly presents on the skin. Its clinical manifestations can present multiple differential diagnoses. Skin biopsy, with immunohistochemistry and flow-cytometry studies are the tools for the correct diagnosis and treatment.The expertise of a dermatologist with this malignancy will aid in appropriate treatment. The outcome of this unusual disease is less aggressive in children than the prognosis described previously in adults.

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Abstract N°: 2302

Annular Lichenoid Dermatitis of Youth: Two Cases

Senem Yüksel Ergüder*[1], Tuğçe Aycan Akbal[2], Emine Beyza Koç Özdamar[2]

[1]University of Health Sciences Ankara Training and Research Hospital, Department of Dermatology, Ankara, Türkiye, [2]Ankara Training and Research Hospital, pathology

Introduction & Objectives:

Annular Lichenoid Dermatitis of Youth (ALDY) is a rare condition. Approximately 64 cases have been recorded, predominantly from Mediterranean countries. Although it primarily affects children and teenagers, there have occasionally been recorded cases among adult patients. Despite its clinical recognition, the etiology of ALDY remains elusive. ALDY presents a diagnostic challenge owing to its resemblance to other annular dermatoses such as hypopigmented mycosis fungoides, erythema annulare centrifigum, and granuloma annulare. However, its distinction from these entities is facilitated by histopathological examination. Herein, we aimed to present two cases of ALDY managed with topical corticosteroids and topical tacrolimus.

Materials & Methods:

Two cases of Annular Lichenoid Dermatitis of Youth were presented with their history, physical examination, pre and post-treatment photos, differential diagnosis tests, and histopathological examination.

Results:

Case 1: An 8-year-old female patient with persistent erythematous plaques in her axilla and groin was diagnosed with ALDY. Initially misdiagnosed as tinea corporis, treatment was unsuccessful. Laboratory tests showed results within the normal range, and serological tests were negative. A punch biopsy from the edge of the annular lesion revealed irregular acanthosis, orthokeratosis, focal parakeratosis, focal spongiosis, increased lymphocyte exocytosis, and focal basal degeneration in the epidermis. Basal vacuolar degeneration and apoptotic keratinocytes were evident at the rete tips. Immunohistochemical analysis revealed a predominance of CD8+ T lymphocytes. Diagnosis of ALDY was established, and treatment with 0.3% tacrolimus cream twice daily resulted in complete resolution of lesions after eight weeks.

Case 2: A healthy male patient with purple-brown hypopigmented patches, initially asymptomatic at age 5, was diagnosed with ALDY after a punch biopsy. Despite negative serological tests, the anti-HCV antibody was positive, prompting further investigation. However, subsequent HCV RNA testing returned negative. The patient remains under follow-up by the pediatric infectious diseases department. The patient underwent a punch biopsy, which revealed features consistent with lichenoid dermatitis, leading to the diagnosis of ALDY. Treatment with topical 0.1% mometasone furoate twice daily resulted in gradual resolution of lesions over eight weeks.

Treatment with mometasone furoate resulted in gradual lesion resolution over eight weeks.

Conclusion:

An essential connection between clinical and pathological findings is necessary for diagnosing ALDY, a unique form of lichenoid dermatitis. Awareness of annular lichenoid dermatitis of youth among dermatologists facilitates prompt diagnosis and appropriate management, ensuring optimal patient outcomes.

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Abstract N°: 2305

Annular Lichenoid Dermatitis of Youth: Two Cases

Senem Yüksel Ergüder*[1], Tuğçe Aycan Akbal[2], Emine Beyza Koç Özdamar[1]

[1]University of Health Sciences Ankara Training and Research Hospital, Department of Pathology, ankara, Türkiye, [2]Ankara Training and Research Hospital, Department of Pathology, ankara, Türkiye

Introduction & Objectives:

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Abstract N°: 2546

A case of linear IgA bullous dermatosis in atopic skin

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Introduction & Objectives:

Materials & Methods:

Results: A 6-year-old girl presented to our attention a vesiculobullous eruption, including ring-like configuration on dry skin, localized mostly on the upper extremities and on the trunk, and slight involvement of oral mucosa. Cutaneous manifestations were associated with intense itching, low-grade fever, arthralgia, and intense pain with difficulty walking. Her medical history was unremarkable, except for atopic dermatitis since early months of life and concomitant rhinovirus infection. Due to her general state of health, she was admitted to pediatrics recovery.

Laboratory investigations were normal (including complete blood count, glycemia, hepato-renal function, urinalysis, and autoimmunity tests), except for inflammation indices (erythrocyte sedimentation rate and c-reactive protein). A skin biopsy was performed on a new blister lesion localized on her left foot. Histopathologic examination with direct immunofluorescence showed subepidermal blisters associated with linear IgA deposits along the basement membrane zone. Based on the clinical presentation and histological examination, the patient was diagnosed with linear IgA bullous dermatosis (LABD). Treatment was started with oral prednisone 0.5 mg/kg/day in tapering dose and later continued with dapsone in oral solution (0.8 mg/Kg/day). A skin improvement was noticeable in just 5 days after starting dapsone, which the patient continued without side effects or worsening of atopic dermatitis.

LABD is a rare autoimmune disease and, to the best of our knowledge, only few cases of this bullous disease with atopic dermatitis coexisting together have been reported in literature.

Conclusion:

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Abstract N°: 2878

Effect of secukinumab on inflammatory linear verrucous epidermal nevus with a somatic mutation in CARD14

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Introduction & Objectives: Inflammatory linear verrucous epidermal naevus (ILVEN) is a rare skin disease that classically presents in infancy and extends over the skin for later months to years. It is characterized by pruritic, erythematous hyperkeratotic papules linearly distributed along Blaschko's lines. In this study, we reported a case presented with erythematous plaques with scales distributing all over the body following the Blaschko's lines cilinically diagnosed with inflammatory linear verrucous epidermal nevus (ILVEN). We treated her with secukinumab and finally and it turned out to be a significant improvement.

Materials & Methods: Histology of a skin biopsy taken from an erythematous verrucous plaque of the left lower limb was made; DNA from the affected tissue and blood was extracted, and whole exome sequencing (WES) was performed. We treated her with secukinumab and finally and it turned out to be a significant improvement.

Results: Histology of a skin biopsy taken from an erythematous verrucous plaque of the left lower limb showed alternated hyperkeratosis and parakeratosis. Within the dermis, there was a mild perivascular lymphocytic infiltrate. A heterozygous missense variant (c.412G□A, p.E138K) in *CARD14* was detected. The variant allele load was 24% in affected skin and 4% in the blood, which was proved to be as a somatic mutation. We treatment with secukinumab (75mg on week 0, 1, 2, 3 and 4 followed by 75 mg every four weeks). She had a significant relief of pruritis within four weeks, and at a 12-week follow-up, there was a significant reduction of the psoriasiform papules and plaques. Sustained improvement was observed within 52 weeks.

Conclusion: LVEN is a rare disorder, and the term is a clinical description.Genetic causes should be checked for this kind of children.

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Abstract N°: 2879

Granular parakeratosis: dermoscopic findings of an uncommon dermatosis

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Introduction & Objectives:

This case study presents a 3-year-old girl with recurrent pruritic skin lesions on her hips, ultimately diagnosed as granular parakeratosis (GP). Notably, the diagnosis was suggested by dermoscopy and confirmed by histology.

Materials & Methods:

Dermoscopic examination revealed distinctive features, including structureless brownish pigment surrounding central white areas, indicative of hyperkeratosis with retained keratinocyte nuclei.

Results:

These findings correlated with histological evidence of hyperkeratosis and parakeratosis with the presence of numerous small, eosinophilic granules within affected keratinocytes.

Conclusion:

Granular parakeratosis is an uncommon dermatological condition with distinct histological features. This report underscores the utility of dermoscopy in diagnosing GP, particularly in pediatric cases, suggesting the possibility of replacing histological exam with the less invasive dermoscopy. Further research is needed to establish definitive dermoscopic criteria for GP diagnosis.

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Abstract N°: 2942

Group A Streptococcal Blistering Distal Dactylitis: case series after COVID-19 pandemics in Hungary

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Group A Streptococcal Blistering Distal Dactylitis: case series after COVID-19 pandemics in Hungary

Introduction & Objectives:

Blistering distal dactylitis (BDD) first described in 1972 by Hays & Mullard is a previously rare, localized bacterial infection of the volar fat pad of the distal phalanx of the fingers. BBD is characterized by the acute appearance of medium to large, non-tender blisters filled with a thin, whitish fluid, which can become purulent and finally erosive. In the past, the causative organism was group A or B Streptococcus, but after 1992 Staphylococcus aureus bacteria grew most commonly from the culture of the fluid.

Materials & Methods:

The authors present cases of children with blistering distal dactylitis caused by Group A Streptococcus infection who attended the Department of Pediatric Dermatology at Heim Pál National Pediatric Institute, Budapest, Hungary, between January 2022 and August 2024.

Results:

The number of cases of blistering distal dactylitis associated with Group A Streptococcus infection significantly increased after COVID-19 pandemic. In 2021, Group A Streptococcus grew from 2,4% of the wound cultures, in 2022 this number was 3,5% and in 2023 the percentage of GAS infection of wound cultures increased to 9,8%. All cases treated with local therapy and systemic antibiotics healed completely. No severe complication was observed.

Conclusion:

Since the COVID-19 pandemic, the number of infections caused by Group A Streptococcus increased worldwide significantly, which can be explained by introduction of the toxigenic and rapidly spreading M1UK strain. Reduced GAS transmission and exposure during COVID-19-related restrictions leading to lack of herd immunity and coinfection with influenza, RSV and varicella could also supported the spread of GAS. The authors recommend close monitoring of the increasing number of the infections caused by Group A Streptococcus and spreading knowledge of the disease of blistering distal dactylitis.

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Abstract N°: 2948

Erosive pustular dermatosis of the scalp after premature prolonged rupture of the membranes.

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Introduction: Erosive pustular dermatosis of the scalp (EPDS) is an uncommon inflammatory skin disease characterized by the triad of erosions, crusts, and sterile pustules. Although it predominantly affects older adults, cases have been reported in childhood and at birth. We report a case of EPDS in a preterm newborn complicated by premature prolonged rupture of the membranes (PPROM).

Case report: A girl was born at 36 weeks of gestation with no problems by cesarean section indicated by PPROM, prolonged cephalic engagement and failure to progress. At birth, she had an erosive plaque encompassing 40% of the scalp, with central hemorrhagic areas and pink scarring. The lesion had a well-defined border and oval shape on the vertex and temporal scalp, with hair preservation all around the periphery. A culture was negative for bacterial and fungal organisms. Transfontanellar ultrasound showed no underlying bone or brain defects. A diagnosis of EPDS was consider in the context of PPROM and prolonged trauma. She was started on betamethasone valerate and gentamicin sulfate for 2 weeks, leading to a decrease in the ulcers and complete re-epithelialization was achieved.

Discussion: The incidence of EPDS is very low in children. Prior to this case, there have been 8 pediatric of EPDS published in the literature, 5 of whom were infants. The infantile cases seem to have developed in association with perinatal scalp injury. The most effective therapies include high-potency topical corticosteroids, but it usually resolves with cicatricial alopecia.

Conclusion: Pediatric EPDS should be considered in the setting of trauma-associated scalp wounds resistant to antibiotics. Early use of topical corticosteroids may increase the likelihood of hair repopulation.

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Abstract N°: 2975

Paediatric Spitzoid Neoplasms

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Introduction & Objectives:

Spitzoid lesions are a wide tumour class comprising Spitz nevus (SN), atypical Spitz tumour (AST) and Spitz melanoma (SM). These are all histologically characterized by spindle cell and epithelioid cell melanocytic proliferations, but while SN display benign histological features and SM display malignant histological features, AST present some atypical histological features in between SN and SM, yet insufficient to make a diagnosis of melanoma.

Herein we present a large series of paediatric-specific spitzoid neoplasms from a single centre with the aim of analysing the epidemiological, morphological and genetic aspects of paediatric spitzoid lesions.

Materials & Methods:

We conducted a single-centre-based retrospective survey on all histologically diagnosed spitzoid lesions of paediatric patients (1-18 years) of the last 10 years (2012-2022). Histopathological reports and electronic records of patients were used to retrieve relevant data regarding patients' features, clinical and dermatoscopical aspects of lesions when recorded, and FISH tests when present.

Results:

Of 255 studied lesions, 82% were histologically benign, 17% atypical, 1% malignant. Clinically, all the malignant lesions and most of the atypical ones were large (≥6 mm) and raised, while benign lesions were more likely to be small (≤5 mm), flat, and pigmented ($p < 0.0001$).

Dermatoscopical patterns, analyzed in 100 patients, further aided diagnosis: a starburst pattern suggested benignity (26% SN ($p = 0.004$)), while a multicomponent pattern pointed towards atypicality or cancer (56% AST, 50% SM ($p = 0.0052$)).

Eighty-five lesions were subjected to fluorescence in situ hybridization (FISH): positive results were more likely in atypical lesions (FISH-positive:71% AST; 29% SN vs FISH-negative:63% SN; 37% AST) ($p = 0.0038$).

Conclusion:

This study on pediatric spitzoid lesions disclosed that most (82%) were indeed harmless, but a concerning number (17%) showed unclear features (atypical), requiring careful evaluation, and 1% showed even malignancy. This highlights the importance of caution when dealing with spitzoid lesions in children, as some may appear harmless but have the potential to be cancerous.

EA DV CONGRESS
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Abstract N°: 2991

Cutaneous Xanthomatosis Revealing Familial Hypercholesterolemia: A Very Special Case

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Introduction & Objectives:

Xanthomas are yellowish cutaneous neoformations consisting mainly of macrophage-rich lipid granules made of cholesterol and triglycerides. This definition helps to distinguish them from lipomas composed of adipocytes, exogenous lipid overloads, and extracutaneous lipidosis within the context of lysosomal diseases. Cutaneous xanthomas can be eruptive, tendinous, tuberous, verruciform, or xanthelasma. We report the case of a child with multiple tuberous xanthomas revealing familial hypercholesterolemia.

Materials & Methods:

A 7-year-old girl, born of a consanguineous marriage (3rd degree), with no particular medical history, presented with papulo-nodular eruptions on the elbows/knees/hands/thighs/buttocks/in the Achilles tendon area, evolving for 5 years without itching, visual disturbances, diarrhea, neurological signs, or impairment of limb mobility in afebrile context. Dermatological examination revealed painless yellow-brownish nodules, smooth and firm, coalescing at times to form a ring-like appearance on the elbows, knees, buttocks, knee flexion folds (popliteal fossae), calcaneal tendons, and grouped papules on the dorsal aspect of both hands in the metacarpophalangeal joints area, as well as mobile and painless deep nodules on the 1st interphalangeal joints of all fingers. Dermoscopy showed centrally shiny yellowish-white areas with no specific vascular pattern and an erythematous border. Rheumatological examination was unremarkable. The rest of the examination of other systems was normal. Joint X-rays, chest X-ray, ECG, cardiac ultrasound, and Doppler ultrasound of the neck and limb vessels were normal. Laboratory tests revealed hypercholesterolemia at 5.79g/L and LDL hypercholesterolemia at 6.4g/L. A skin biopsy revealed xanthomatosis without signs of malignancy. Biological tests of the parents and younger brother also revealed hypercholesterolemia. The very high levels of LDL cholesterol, the presence of cutaneous xanthomas since the age of 4, and the absence of other abnormalities suggested a homozygous form of familial hypercholesterolemia in this child. The patient was managed with hygienic-dietary measures combined with statin therapy, and excision of unsightly lesions was planned using laser.

Results:

Familial hypercholesterolemia is a genetic disease with autosomal dominant transmission characterized by very high LDL cholesterol levels from birth and complicated by early cardiovascular diseases. It is characterized by the appearance of cutaneous xanthomas as early as the first decade and sometimes even present at birth. They are mainly located in the buttock region and on the extensor surfaces of the elbows and knees. LDL cholesterol levels are very high from birth. The homozygous form, which severely affects patients, is however much rarer and affects one person in 250,000 to 1,000,000. The average age of the first angina attack is around 11 years, and life expectancy does not exceed 20 years without treatment. Medical treatment combined with an appropriate diet is generally insufficient in homozygous patients. The treatment of choice is currently represented by LDL apheresis.

Conclusion:

In the presence of cutaneous xanthomas in a child with high cholesterol levels, familial homozygous hypercholesterolemia should be considered. Early management of the entire family prevents the occurrence of

complications

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Abstract N°: 3021

Incontinencia pigmenti in males. Presentation of three cases.

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Introduction & Objectives: Incontinentia pigmenti (IP) is a genodermatosis with dominant X-linked inheritance that produces multisystem involvement and is lethal in most male fetuses.

Materials & Methods: Case 1. A 55-day-old male with no relevant family history who, 48 hours after birth, began to have crusted vesicle lesions on the trunk and cheeks. Physical examination showed erythematous plaques and isolated pustules. General condition was normal and fever was absent. Analytical examination revealed hypereosinophilia, while a perivascular and interstitial inflammatory infiltrate with eosinophils and eosinophilic spongiosis was observed under histological examination, compatible with IP. Karyotype was requested, which was normal. An evaluation by pediatric neurology, ophthalmology and genetic study was requested.

Case 2. A 4-day-old male with no relevant family history, showing congenital lesions in the left leg, with posterior extension to the chest and right arm. Physical examination revealed multiple erythematous papules grouped in a linear pattern, with yellowish crusts and vesicles. PCR for HSV detection was performed and empirical treatment with acyclovir and cloxacillin was initiated. Skin biopsy was performed, which showed a perivascular inflammatory infiltrate in the superficial dermis composed of lymphocytes and eosinophils, as well as intraepidermal eosinophils, compatible with IP. Karyotype, genetic study and evaluation by neurology and ophthalmology were requested, but were not performed due to loss of follow-up.

Case 3. Male, 30 days old, under neurology follow-up due to muscle spasms, with normal EEG. His parents had had two previous abortions, of unknown sex. He had congenital injuries on his back. Physical examination showed erythematous-scaly plaques with linear configuration, drawing a “V”, with regular, well-defined edges, raised, with a flat surface and white adherent peeling. Biopsy revealed acanthosis, hyperkeratosis follicular corneous plugs, isolated intraepidermal eosinophils and dyskeratinocytes, compatible with IP. Karyotype was requested, which was normal. No mutations in IKBKG were observed in affected skin samples or blood samples. Up to date, ophthalmological and neurological evaluation, as well as dentition, have been normal.

Results: The cutaneous manifestations of IP begin in the neonate and are usually distributed along the Blaschko lines. The presence of typical IP lesions and compatible histology is sufficient to make the diagnosis. The most common mutation is the deletion of exons 4-10 in the IKBKG gene (previously known as NEMO), which encodes a protein that regulates cell apoptosis. It is a lethal syndrome for the male sex unless there is post-zygotic mosaicism, hypomorphic mutations of the IKBKG gene or Klinefelter syndrome (47XXY).

Conclusion: We present three cases of men with a diagnosis of IP, an entity that should be considered within the differential diagnoses of vesiculobullous lesions in male neonates.

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Abstract N°: 3027

Genetic syndromes with paediatric alopecia areata: A systematic review

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Introduction & Objectives: Numerous genetic studies strongly associate alopecia areata (AA) susceptibility to certain gene loci. However, there is a wide variation of phenotypes displayed by individuals with AA, especially in the paediatric population. Further linkage and association studies should be conducted to support the hypothesis that AA is a complex genetic trait that depends on the cooperation of several genes. This review aims to systematically search published studies to identify paediatric AA syndromes and their clinical features, to summarize the current state of their genetic elucidation.

Materials & Methods: A systematic search of MEDLINE, Embase, CENTRAL, PubMed, OMIM, and Orphanet databases was performed. All original case reports, case series, and observational studies describing AA in children (under the age of 18 years) with monogenic or chromosomal syndromes were included. Further data from OMIM, Orphanet, reviews, clinical guidelines, and basic science studies was used to retrieve additional comprehensive information on each syndrome, such as prevalence or pattern of inheritance.

Results: After title and abstract screening of 1426 studies, and full text review of 224 studies, 68 studies met the inclusion criteria and are summarized in this review. Overall, the search identified 33 genetic syndromes with paediatric AA. None of the syndromes included AA as a cardinal clinical feature; however, “alopecia,” or “hair loss,” is a cardinal feature in three of the included syndromes: Gomez-Lopez-Hernandez syndrome; hereditary hypotrichosis (Marie-Unna type); and incomplete antibody deficiency syndrome. Prevalence estimates were available for 78.8% (26/33) of syndromes, with 45.4% (15/33) of syndromes presenting in less than 1/1 000 000 individuals. Sixty-seven percent (22/33) of syndromes were fully genetically elucidated; 12.1% (4/33) were partially elucidated; 9.1% (3/33) were not genetically elucidated; and 12.1% (4/33) were syndromes with chromosomal abnormalities. Seventy-nine percent (26/33) of syndromes were described by only one report, while 21.2 % (7/33) were described in multiple independent reports. The following syndromes with paediatric AA were described more than once in independent articles: autoimmune polyglandular syndrome type 1; autoimmune polyglandular syndrome type 3; chronic atypical neutrophilic dermatosis with lipodystrophy and elevated temperature (CANDLE) syndrome; encephalocraniocutaneous lipomatosis, immune dysregulation, polyendocrinopathy, enteropathy, X-linked (IPEX) syndrome; Trisomy 21; and Turner syndrome.

Conclusion: This review identifies a total of 33 distinct syndromes associated with paediatric AA, summarizing their clinical and genetic features into a comprehensive catalogue. Despite the limited knowledge of these syndromes due to their low prevalence, this review provides insights into the range of genetic syndromes with paediatric AA, facilitating early prediction and diagnosis, and possibly personalized treatments.

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Abstract N°: 3046

A cutaneous nodule revealing Munchmeyer's disease

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Introduction & Objectives:

Cutaneous leishmaniasis is a parasitic disease caused by flagellate protozoa of the genus Leishmania. Meglumine antimonate or Glucantime® remains the first-line treatment for cutaneous leishmaniasis in Morocco, administered either systemically or intralesionally. The intralesional route is indicated for small, single lesions; periorificial locations are considered "at risk" in this technique, and practitioners tend to switch automatically to the systemic route in these locations.

Materials & Methods:

Our study is a retrospective compilation of cases of periorificial LC in children treated with intralesional infiltration of meglumine antimony, observed in our department.

Results:

We recorded 4 patients. The diagnosis of cutaneous leishmaniasis was confirmed by dermal smear (3 cases) or biopsy (1 case). The mean age was 6 years. The sex ratio (M/F) was 1. The mean duration of the disease was 4.7 months. The lesions were located on the tip of the nose in 3 cases and periorbitally in 1 case. All lesions were single and ranged in size from 1 to 3 cm. All patients were treated with intralesional infiltration of meglumine antimony. The number of infiltrations varied between 6 and 10 at a rate of 1ml per cm2 once or twice a week. Progress was favourable in all cases. No complications were observed. The average healing time after treatment was 3.2 months.

Discussion:

The periorificial regions are considered to be delicate areas with no indication for intralesional infiltration of meglumine antimony in the treatment guidelines for cutaneous leishmaniasis. This has led to a tendency to avoid these areas in daily practice, in favour of systemic treatment. This automatic switch to systemic treatment in children, with all its side-effects, sometimes seems to be abused. This is the case for small lesions where the only indication for systemic treatment is the site of the lesion. This corresponds to the lesion profile of the cases in our series. This is all the more true as the excessive precaution found in the recommendations is only really based on the scant literature available concerning intralesional treatment of these areas. Few articles have raised this question. There are a few case reports and 7 cases in a South American series comparing intralesional and intramuscular treatment with AM. Although rare, all these trials of IL infiltration in these "at-risk" regions have been carried out without notable incidents. This is the case in our series which is, to our knowledge, the only study reporting cases of cutaneous Leishmaniasis of periorificial site in children treated with Intra-lesional meglumine antimony.

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Abstract N°: 3130

COVID-19-induced Stevens-Johnson Syndrome-Toxic Epidermal Necrolysis Overlap: A case study in a 2-year-old male

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Introduction & Objectives:

Steven Johnson syndrome (SJS) and Toxic epidermolysis necrosis (TEN) are dermatologic emergencies that carry a risk of long-term morbidity and even fatality. SJS and TEN are immunologically-induced mucocutaneous reactions that cause blistering and epidermal detachment. The amount of skin detachment in relation to the body surface area (BSA) is the only difference between these two conditions. The majority of SJS/TEN cases are drug-related, but infections from *Mycoplasma pneumoniae* and *Herpes Simplex Virus* (HSV) have also been implicated in causing this condition.

Coronavirus disease (COVID-19) is a highly contagious and pathogenic coronavirus that emerged in late 2019 and caused a global pandemic. It typically manifests as an acute respiratory illness that endangers human health and public safety. Initial symptoms are typically non-specific including fever, cough, and lethargy. Multiple systems, including the respiratory, gastrointestinal, musculoskeletal, and neurologic, may also be implicated.

Currently, there are four cases of SJS/TEN in adults caused by COVID-19 have been documented in the literature. The majority of the published material also only demonstrates adult COVID-19-induced SJS or TEN, although a case of a 6-year-old male SJS or TEN has recently been reported. Extensive research on children is occasionally insufficient due to the lack of literature on such cases.

Results:

** The authors observed the disease course of a 2-year-old healthy male with no prior medical history. Patient initially presented with multiple atypical targets which coalesced to subsequently involve 25% of TBSA leading to a diagnosis of SJS-TEN. The patient tested positive for Covid-19 and required treatment with hydrocortisone in which he made a full recovery. Having ruled out drugs and other viral triggers, it is highly possible that COVID-19 was the culprit in this case.

Conclusion:

Certainly, there is much to learn about COVID-19 infection. The novel virus that caused the pandemic, which was initially isolated from the lower respiratory tract and believed to primarily affect the respiratory system, has been revealed to also induce gastrointestinal, cardiovascular, cutaneous, neurological, and hematological symptoms. Moreover, several case reports were already published about the cutaneous manifestations secondary to COVID-19 infection. However, there were only a handful of studies that have been reported on SJS/TEN caused by COVID-19 infection. This case report aimed to emphasize the potential for substantial COVID-19-induced SJS/TEN in the pediatric population.

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Abstract N°: 3163

Aplasia cutis congenita (ACC) due to maternal use of Carbimazole.

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Aplasia cutis congenita (ACC) due to maternal use of Carbimazole.

Introduction & Objectives:

Aplasia cutis congenita (ACC) is a rare congenital skin condition, characterized by localized absence of the skin and subcutaneous tissue. Many factors have been associated with developing ACC among which antithyroid medications were the strongest risk factor. A total of 61 cases of ACC were reported due to Methimazole/Carbimazole (MMI/CMZ). The underlying mechanism behind the potentially severe embryopathy caused by these two medications MMI and CMZ is still unknown. Here, we report a case of male infant with multifocal scalp ACC whose mother had Graves disease and was managed with carbimazole during pregnancy. By that, our case supports the idea of an MMI/CMZ embryopathy.

Materials & Methods:

Case report. And conducting extensive literature review.

Results:

We report a case of a neonate with ACC whose mother received Carbimazole during pregnancy. He was managed successfully by a conservative therapy, and there were no serious reported complications.To the best of our knowledge, this is the first case of ACC associated with intaking Carbimazole during pregnancy in Oma

Conclusion:

The correlation between MMI/CMZ and congenital malformations such as ACC is substantial, even though a conclusive interaction has not yet been demonstrated. Now that the notion of MMI/CMZ embryopathy has been established, larger epidemiological studies are needed to further quantify the risk of congenital deformity among mothers taking antithyroid medication, and a direct causal mechanism needs to be discovered. More studies are needed to corroborate results regarding early pregnancy shift from MMI/CMZ to PTU. Maintaining euthyroid throughout pregnancy is the key to reduce the risk of maternal, fetal, and newborn complications. Furthermore, new antithyroid with fewer side effects need to be established.~~

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Abstract N°: 3253

Analysis of COVID-19 Associated Skin Lesions in Children

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Introduction & Objectives:

The SARS-CoV-2 virus appeared at the end of 2019. More than four years later it is still ongoing issue. Most often, the virus causes respiratory symptoms, but other symptoms such as skin lesions have also been reported.

Our objective was to conduct a study in the population of children with COVID-19 and evaluate the nature of skin lesions, frequency of occurrence, associations with other factors.

Materials & Methods:

An anonymous survey was conducted from November 2021 till April 2022. A total of 434 questionnaires were included in the statistical analysis. Data was analyzed with Microsoft Excel and IBM-SPSS. Differences

were considered statistically significant when $p \leq 0.05$.

Results:

The questionnaire revealed that 52,5% (n=228) of participants were girls. Only children under 18 years old were included in the data analysis with the average age of the participants being 8,29 ± 4,8 years. 90,6% (n=393) of questionnaires were completed by parents. COVID-19 infection was confirmed by laboratory tests in 96,1%. (n=417) children. Further analysis was carried out with these cases.

The most common symptoms of the COVID-19 infection among children were fever 55,9% (n=223); nasal congestion 44,1% (n=184) and cough 39,6% (n=165). Mucous membrane damage occurred in 9,4% (n=39) of all children. Skin lesions are characteristic of 15,6% (n=65) of all children with COVID-19.

Skin symptoms were present in 60% (n=39) of girls, 40% (n=26) - boys. Average age – 7,84 ± 5,1 years. A statistically significant relationship was established between the age of patients and the appearance of skin lesions - among newborns and infants, skin rashes appeared more often than > 1 year old children (p=0,020). 2,2% (n=9) of all children with COVID-19 were hospitalized. Five of them had mucosal or skin lesions. It was found that children with mucosal or skin lesions were hospitalized more often than children with only COVID-19 (p=0,010).

The most common symptom accompanying the lesions was erythema in 56,9% (n=37) of cases. Cutaneous lesions usually last up to a week – in 35,4% (n=23) cases and in 52,3% (n=34) of cases occur during illness. Participants of the survey indicated that their skin lesions were similar to maculopapular rash in 23,1% (n=15) of cases, urticaria and vesicular exanthema each in 12,3% of cases (n=8), chilblain-like lesions and erythema multiforme each in 7,7% (n=5) of cases. A positive correlation was found between pain and duration of lesions (> 7 days) and erythema multiforme-like lesions (p=0,002, p=0,015, respectively), between erythema and chilblain-like lesions (p=0,043), between pain and urticaria (p=0,039).

A statistically significant association was found between skin lesions and other symptoms. They occur more frequently with headache (p=0,003), gastrointestinal symptoms (p=0,003), and mucosal lesions in at least one area (mouth/tongue/eye area) (p=0,000). Although in our study, only in 3,1% of cases skin lesions were the only

sign of the disease, the absence of general symptoms was statistically significantly higher among children with a skin lesion (p=0,020).

Conclusion:

These findings are significant and demonstrate the need for a comprehensive skin examination in children in order to suspect COVID-19. Early detection of the disease can help prevent the spread of the infection.

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Abstract N°: 3256

- **Differential dignosis of Psoriasis in Infancy- based on 20-year data from North East Hungary**

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Introduction & Objectives:

Psoriasis is a relatively common dermatosis, its incidence is 2% in developed countries in general population. However psoriasis rarely presents at birth and uncommon in infancy (age 0-3years). Diagnosis of infant psoriasis is based on its clinical appearance, and grouped as plaque, guttate psoriasis in most cases. Seldom represents as erytrodermia, pustular psoriasis, nail psoriasis.

Materials & Methods:

The authors reweived cases of database of the Hospital of Borsod-Abaúj Zemplén County, using its Medworks software system between 01. July 2004 and 31. December 2023.

Results:

In North-East Hungary, in our county, number of birth is approximately 7000 /year. In the past 20 years, 87 infants were diagnosed with psoriasis, 44 girls and 43 boys. The diagnosis of infant psoriasis was based mostly on its clinical appearance. Majority of patients had plaque psoriasis. 20% of infants had skin symptoms of psoriasis in further ages as well (persistant psoriasis), among them, regions affected were in most cases head and neck, genital area and trunk. Extremities, flexures were rarely affected.In non-persistant psoriasis, also head anfd neck, but also flexures were mostly affected areas. Differential diagnosis included atopic dermatitis, diaper rash, tinea corporis, Langerhans cell histiocytosis. In few cases, based on naked eye examination, it was not possible to differentiate the diagnosis, thus, dermoscopy, and histology was carried out. 50% of psoriatic infants had positive family history for psoriasis. Treatment of infants was topical corticosteroid combined with topical emollients.

Conclusion:

Infant psoriasis is a clinical diagnosis in majority of the cases, however its diagnosis can be challenging. Primary symptoms, age of onset, localization of skin symptoms, dermosycopy might give a clue. Rarely, histology examination is needed. Infant psoriasis complications are based on bacterial or fungal superinfection of psoriatic primary symptoms. In infants, prognosis of psoriasis is usually good, most cases regress for topical corticosteroid treatment combined with emollients rapidly. However 19,7% of these infants had symptoms of psoriasis in further ages.

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Abstract N°: 3271

Adalimumab originator versus biosimilars: a comparison in clinical response in a multi-center cohort of paediatric patients with psoriasis

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Introduction & Objectives: Pediatric psoriasis is a frequent disease with a prevalence ranging between 0.5 and 1.5% across Europe. It has a high impact on the quality of life of those affected especially in children or adolecents with moderate-to-severe disease. There are currently no international standardized guidelines for medical treatment of pediatric psoriasis. Moderate-to-severe disease may benefit from use of systemic therapy with biologics, which are considered a first-line systemic therapy. Tumor Necrosis Factor (TNF)- alpha has a key role in the disease's pathogenesis and two inhibitors of this molecule have previously been approved for paediatric psoriasis (etanercept and adalimumab). Additional biologics approved in childhood psoriasis include anti-IL12/23 (ustekinumab), and anti-IL17A (secukinumab, ixekizumab) antibodies. Biosimilars are a cost-effective alternative to off-patent biologic therapies. They offer a valuable pharmacoeconomic strategy to lower healthcare costs. Indeed, a substantial proportion of patients have limited access to therapy due to economics, health policies and clinical considerations, which creates clinical unmet needs. Adalimumab and Etanercept biosimilars have been proved to be comparable in terms of efficacy and safety to their reference products in adults. Data comparing biosimilars and originators are lacking in children. The aim of the study is to compare effectiveness and safety of adalimumab originator and biosimilars in two groups of patients.

Materials & Methods: A total of 100 patients with moderate-to-severe pediatric psoriasis were enrolled in a real-world, multi-center, international, retrospective, observational cohort study. Group 1 of patients included subjects already in treatment with Adalimumab originator and then switched to a Adalimumab biosimilar (n=30), gruop 2 included patients who started directly Adalimumab biosimilar (n=70). Effectiveness was evaluated by measuring the maintenance of clinical response in group 1 assessing the Psoriasis Area and Severity Index (PASI) changes at 6 and 12- months after switching. Effectiveness was evaluated in terms of PASI changes from baseline in group 2. Safety was assessed by reporting any adverse events occurred during the observational period.

Results: Preliminary data showed similar effectiveness and safety in both groups.

Conclusion: Although the number of patients investigated was low, this study represents the first real-world analysis comparing effectiveness and safety of biosimilars in pediatric psoriasis. The introduction of biosimilars also in pediatric patients can increase the number of patients able to receive biologics, allowing these patients to be treated earlier in the disease course, potentially modifying the course of their disease and reducing the risk of comorbidities. Long-term data on effectiveness and safety of Adalimumab biosimilars in paediatric psoriatic patients could confirm these preliminary observations.

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Abstract N°: 3287

Clinical evaluation of a topical formulation with Piroctone Olamine, Stearyl Glycyrrhetinate, Zinc Pidolate and Rhamnosoft in children with mild to moderate cradle cap.

Sonia Aladren[1], Javier Bustos[1], Albert Navasa*[1], Monica Foyaca[1]

[1]ISDIN

Clinical evaluation of a topical formulation with Piroctone Olamine, Stearyl Glycyrrhetinate, Zinc Pidolate and Rhamnosoft in children with mild to moderate cradle cap.

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Introduction & Objectives:

Cradle cap, or pityriasis capitis, is a subset of infantile seborrheic dermatitis (ISD). It is a very common, mostly self-limiting, chronic non-inflammatory scaling skin condition that presents between the third week and first couple months of life. The skin manifestations are marked by erythematous plaques with greasy-appearing yellowish scales. Cradle cap can be found in areas dense in sebaceous gland activity, such as the scalp, T line of the face and external ears. The objective of the study was to investigate efficacy and tolerability of a topical formulation containing Piroctone Olamine, Stearyl Glycyrrhetinate, Zinc Pidolate and Rhamnosoft on decreasing the signs and symptoms of cradle caps in children with mild to moderate cradle cap.

Materials & Methods:

The topical formulation was used at least twice daily in 11 children with mild or moderate cradle cap, 45% males, aged 2 to 40 months, during 29 days. The severity of symptoms of cradle cap such as scaling, crusting, erythema and oiliness were evaluated by the investigator using a scale of 5 scores (from 0=none and 4-severe) and Investigator Global Assessment (IGA) using a 5-point scale (from 0=no response to 4=complete response/clear). In addition, a self-assessment questionnaire was evaluated by the parents. These parameters were evaluated at baseline (D0), and after 29 days of treatment. The tolerability/acceptability was evaluated by the investigator through the clinical evaluation and asking the subjects or subjects' parents about unpleasant sensations.

Results:

After 29 days of use of the product, the study result showed a statistically significant ($p<0.01$, all) improvement of scaling score (-57%), crusting (-59%), erythema (-90%) and oiliness (-67%) compared to D0. In addition, the investigator reported that 100% of children showed an improvement of cradle caps, and 81% of them showed a moderate and excellent response at D30. 100% of parents reported that the discomfort feeling, shine, flaky or desquamation and redness of the cradle cap area decreased and improved the overall aspect at D30, and regarding the product, 100% reported the texture was pleasant and penetrated quickly. The product was very well tolerated.

Conclusion:

This topical formulation has shown an improvement of the cradle cap signs, with a visible improvement of appearance of children's skin and with well tolerance.

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Abstract N°: 3323

Case Series: Efficacy and safety of Spesolimab in pediatric GPP

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Introduction & Objectives:

Generalized pustular psoriasis (GPP) is a severe and potentially life-threatening subtype of pustular psoriasis, characterized by extensive pustules, pain, and systemic symptoms akin to sepsis. It is primarily observed in children and/or individuals with psoriasis. While biologics represent an advanced treatment option for GPP, effective treatments for pediatric patients remain scarce. Recently, the interleukin (IL)-36 receptor blocker, spesolimab, was approved for GPP in adults.

Materials & Methods:

Herein, we document the robust response of spesolimab treatment (15mg/kg, ivtt) in 4 GPP pediatric patients and assessed changes in the severity of their conditions.

Results:

The primary outcome parameter was the changes in GPPASI at day 15 . At day 15, all GPP patients achieved GPPASI 90. The mean score of GPPASI was 16.50±3.02 at baseline vs. 0.48±0.15 at day 15. The pustules subsided rapidly within 1 day, erythema and scale reduced gradually in two weeks with the mean score of GPPGA and CDLQI at baseline and day 15 was 2.50±0.58 vs. 0.00±0.00and 24.00±1.41vs.1.50±0.58, respectively. During follow-up, no unexpected safety signals were observed.

Conclusion:

Spesolimab may be a potentially effective and safe treatment for pediatric GPP.

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Abstract N°: 3469

Nail-patella syndrome in a mother-daughter dyad: a case presentation and review of the literature

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Introduction & Objectives:

Nail-patella syndrome (NPS), also referred to as hereditary onycho-osteodysplasia or Fong disease, is an autosomal dominant disorder involving the *LMXB1* gene that primarily encompasses the clinical tetrad of dysplastic fingernails, absence or hypoplasia of the patella, elbow deformities, and iliac horns. Renal involvement may initially manifest with proteinuria and can progress to end-stage kidney disease. Ocular hypertension and open-angle glaucoma are ophthalmologic complications associated with NPS. A unique presentation of NPS in a mother-daughter dyad presenting to the Dermatology clinic will be discussed, followed by a review of the pathogenesis of the disorder and current treatment guidelines.

Materials & Methods:

A chart review was performed to gather information on the patient's prior medical history and clinical course. Clinical photography was obtained to further characterize the patient's diagnosis. A literature search was performed on PubMed, including the search terms "nail patella syndrome", "Fong disease", and "hereditary onycho-osteodysplasia" to elucidate current guidelines for NPS.

Results:

A 21-month-old female infant was referred to Dermatology by her pediatrician for evaluation and further management of new papules present diffusely on her body. She presents with her mother to the dermatology clinic for the initial evaluation. On physical exam, in addition to several skin-colored papules with central umbilication characteristic of molluscum contagiosum, she is incidentally noted to have dysplastic fingernails bilaterally. When the patient's mother assists in positioning the infant for the physical exam, similar clinical features were apparent on her fingernails as well. The patient's patella was difficult to palpate bilaterally; the mother's patella was noted to be hypoplastic on exam. A diagnosis of familial NPS was made. The patient had a follow-up appointment with orthopedic surgery, and routine monitoring and referral to genetics were recommended.

Conclusion:

To date, no structured management recommendations put forth by a medical organization exist for NPS. Following clinical diagnosis, we typically recommend referral to and further evaluation with orthopedic surgery, ophthalmology, and medical genetics.

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Abstract N°: 3637

Connecting the Dots: Pediatric Psoriasis and the Metabolic Puzzle

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Introduction & Objectives:

Psoriasis is an immune-mediated inflammatory systemic disease with skin tropism & a chronic relapsing course. It affects about 1–2 % of the world's population & 22–33% (about 1/3) of cases starts in childhood. This imposes a negative impact on the children's quality of life

Currently, a changing viewpoint contends that psoriasis is not just a skin-limited condition, but rather**"cutaneous expression of systemic inflammation"***

In adults, this link between psoriasis & cardiometabolic comorbidities is well known, but the evidence is less clear in children

This work studies the latest evidence in this aspect

Materials & Methods: pediatric patients with psoriasis were examined with a review of old & recent literature regarding the association with the metabolic syndrome & the cardiometabolic consequences

Results: a clear relationship was found between psoriasis & cardiometabolic comorbidities

Conclusion: the guidelines for screening of cardiometabolic comorbidities & pediatric psoriasis that were set 2017 should be revised in order to reach early detection of life threatening cardiovascular comorbidities

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Abstract N°: 3662

Staphylococcus scalded skin syndrome-case report in 29-day-old male patient

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Introduction & Objectives: SSSS is a rare, severe, skin disease which is characterized by the detachment of the epidermis. This is caused by exotoxins A and B which are release from definite strains of Staphylococcus aureus bacteria.

Materials & Methods: We report a case of a 29-day-old male patient with SSSS who was admitted in our clinic. The first changes started two days before hospitalization, as erythema first on the face and flexural regions and within two days the changes spread to the hands, feet and body. He was treated by the family doctor as contact dermatitis, but there was no improvement, the rash is following by desquamation of skin that look like a burn.

Results: The clinical diagnosis on admission is confirmed based on the findings of tender erythroderma and desquamation with a scalded appearance, positive Nikolsky sign, and absence of mucosal involvement. After immediately treatment with intravenous antibiotics (Ceftriaxone 50mg/kg/body weight and Vancomycin 15mg/kg/ body weight) and local therapy (fusidic acid) there is a prompt improving of skin changes.

Conclusion: Staphylococcal-scalded skin syndrome (SSSS), is a potentially life-threatening disease and a pediatric emergency. Early diagnosis and treatment are essential to decrease the morbidity and mortality of this condition.

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Abstract N°: 3839

Cutaneous Rosai-Dorfman Disease presenting as soft-tissue tumors in a teenager

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Introduction & Objectives:

Rosai-Dorfman disease is an uncommon non-Langerhans cell histiocytosis with benign course. According to the 2016 Histiocyte Society classification, Rosai-Dorfman disease may be cutaneous and extracutaneous. Cutaneous Rosai-Dorfman disease (CRDD) is rare, representing <3% of cases typically occurring in adulthood. It is extremely rare in children, with < 10 reported cases and has unique epidemiological and clinical characteristics. The pathogenesis is unknown, but an association with viral infections that potentially lead to cutaneous immune dysregulation with macrophage is hypothesized.

We present this case of a male patient with CRDD secondary to Epstein-Barr virus infection.

Materials & Methods:

A previously healthy 9-year-old male developed a soft-tissue tumor in the abdominal wall and was treated with surgical resection, no histological study was done. One year later, the lesion recurred and new lesions arose in the groin, lumbar region and buttocks. He was referred to our institution for work-up and diagnosis.

Results:

Examination revealed multiple firm, subcutaneous nodules adhered to deep planes with different sizes (the largest 7 cm in diameter). Overlying skin was erythematous-violaceous in color. There was no lymph node involvement.

An excisional biopsy was obtained and reported a histiocytic proliferation with emperipolesis phenomenon in soft tissues, immunohistochemistry was positive for S100 and CD68, and negative for CD1a. With these findings, the diagnosis of extranodal Rosai-Dorfman disease was made.

He was referred to the Oncology service to rule out a systemic lymphoproliferative process. Diagnostic workup including blood count, acute phase reactants and immunological profile was normal, except for antibodies anti EBV (EBV VCA IgG +, EBV EA +, EBV EBNA +) suggesting recent active infection. A PET CT showed multiple hypermetabolic zones in subcutaneous tissue, without involvement of lymph nodes or bone, confirming the diagnosis of solely CRDD. Treatment with prednisone 1mg/kg/day and methotrexate 15/m2SC/week was started, with adequate clinical response manifesting as a decrease in the size of lesions.

Conclusion:

Cutaneous Rosai-Dorfman disease is a highly uncommon condition in the pediatric population. Clinical manifestations are varied and can easily lead to misdiagnosis, and it is always necessary to rule out extracutaneous involvement. It has a benign course with spontaneous resolution in up to 40% of patients; however, in cases with persistent lesions, like our patient, the use of methotrexate in combination with systemic steroids is indicated, with an overall cure rate of 28%. Further therapeutic options Include dapsone, thalidomide, cyclosporine, acitretin, imatinib, local radiation, cryotherapy and surgical resection.

We believe presenting this patient with CRDD is of great interest, because of the rarity of his diagnosis, florid

clinical manifestations, and the association with EBV infection.

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Abstract N°: 3897

Surgical management and long time follow up of nail matrix nevi in Asian children

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Introduction & Objectives:

Malignant melanoma has very distinct epidemiological characteristics, with clear differences in incidence and disease type between Caucasians and Asians.

Superficial spreading melanoma is more common in Caucasians, whereas acral melanoma is the most common disease type in Asians, accounting for over 40% of all cases, and this is also the case in Japanese.

Among Acral melanomas, those that occur on the soles of the feet are the second most common that occur on the nails. For this reason, pediatricians and obstetricians suspect nail apparatus melanoma, and many children visit dermatologists for surgery with nail pigmentation as their chief complaint. Radical surgery for nail apparatus melanoma often requires amputation of the fingers and toes, which is highly invasive, so the diagnosis must be accurate.

Among pigment nails, we attempted to clarify the differentiation of nail matrix nevi from acral melanoma, treatment methods, and effective methods for follow-up observation.

Materials & Methods:

A retrospective study of children aged 0 to 15 years who visited the National Cancer Center Hospital (Tokyo, Japan) with nail pigmentation between January 1992 and December 2022 and whose progress could be regularly followed.

Results:

The ages of the patients ranged from 1 to 15 years, 21 were boys and 16 were girls, and the sites of nail matrix nevi were fingers in 28 and toes in 9. The most common type was finger III. 36 cases were single cases, 1 case was multiple cases, and regular medical examinations were performed for many cases, but the most long-term follow-up was 26 years. Many cases of nail matrix nevi disappearing with age have been observed.

Two of the 37 surgical cases had pigmented lesions on the nails and skin around the nails, and surgery was performed because the skin lesions had enlarged. The histopathological diagnosis of both cases was nevus cell nevus.

There were no cases of acral melanoma during the observation period, including surgical cases and follow-up cases.

Conclusion:

Our study is a very large case series of Asian children with nail matrix nevi. Biopsy of nail matrix nevi is often technically difficult, and even if the nail is benign, the nail remains deformed after the biopsy. In addition, if the pigmented lesion is wide, surgery including the nail matrix will be required, resulting in permanent nail loss. On the other hand, since there were no cases of melanoma in our study, we believe that it is better to carefully observe the patient’s progress rather than immediately resorting to surgical treatment.

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Abstract N°: 4030

Unilateral precalcaneal congenital fibrolipomatous hamartoma, an atypical presentation of an underdiagnosed entity.

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Introduction & Objectives:

Precalcaneal congenital fibrolipomatous hamartoma is a benign entity, originally described as podalic papules of the newborn, presenting as plantar adipose nodules on the internal side of the heel, usually bilateral. Histologically it presents with lobules of mature adipose tissue surrounded by fibrous septa in the deep dermis and hypodermis. The aim of this study is to present a case of an atypical presentation of this underdiagnosed entity.

Materials & Methods:

Retrospective case presentation including anamnesis, physical examination and ultrasound examination.

Results:

We report an 8-month-old male infant referred by his paediatrician for presenting, since the age of 2 months, a 2 cm raised soft area on the sole of the left foot. On examination he presented a subcutaneous lesion, soft in consistency, poorly demarcated and with no epidermal changes. The initial suspicion was of precalcaneal congenital fibrolipomatous hamartoma, but due to its unusual unilateral presentation, we performed an ultrasound examination which showed small hypoechoic areas in the deep dermis and hypodermis intermixed with angulated hyperechoic bands, as well as dermal and hypodermal thickening, supporting the diagnosis. At the next visit the child had already started to ambulate without difficulty or discomfort with walking.

Conclusion:

Precalcaneal congenital fibrolipomatous hamartoma is characterised by the presence of bilateral soft subcutaneous nodules, with unilateral involvement being more uncommon. Retrocalcaneal cases have also been described in the literature. It was first described in 1990 as podalic papules of the newborn. The lesions are usually congenital or appear shortly after birth. In their evolution they usually grow with the child and do not usually cause discomfort or walking problems. There is no risk of malignancy.

Diagnosis is clinical, although when the presentation is unilateral it may cause confusion with other entities such as fibromatosis, lipomas, lipomatous nevus, dermal hypoplasia, haemangioma, lymphatic malformation, venous malformation and neurofibroma. In these cases, before considering a biopsy for histological study, an ultrasound examination can be performed, which will show an increase in soft tissue with hypoechoic areas mixed with hyperechoic areas in the deep dermis and hypodermis. It is important to know about this entity and to provide reassurance to the parents. In case of diagnostic uncertainty or to aid diagnosis, a soft tissue ultrasound will be very useful.

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Abstract N°: 4070

multiple juvenile xanthogranuloma, key considerations

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Introduction & Objectives:

Juvenile xanthogranuloma (JXG) is the most common form of non-Langerhans cell histiocytosis (NLCH). It occurs mainly in the pediatric age group (64% before the age of 7 months), with an estimated prevalence of 1 per million children. It manifests clinically as single or multiple (2-3 to more than 100) erythematous-yellowish papules or nodules located most frequently on the head and neck, with a typical “setting sun” dermoscopic pattern.

Materials & Methods:

Presentation of a case and review of the literature.

Results:

Systemic JXG is a very infrequent entity and usually presents with multiple cutaneous and/or subcutaneous nodules with involvement of 2 or more viscera. Systemic ocular involvement (0.24% of cases) is more frequent in patients with multiple JXG. Thus, the presence of 2 or more cutaneous JXG, an early onset (< 2 years) and the cutaneous micronodular form are suggested risk factors for ocular involvement.

Conclusion:

We present the case of an 8-month-old infant with multiple JXG and no extracutaneous involvement. In the presence of multiple JXG meticulous physical examination and clinically-based complementary tests are recommended. Finally, the latency period of systemic involvement can range from months to years and so a negative baseline study does not exclude possible subsequent onset.

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Abstract N°: 4136

A rare case of erythrokeratodermia variabilis in a 7-year-old child.

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Introduction & Objectives:

Erythrokeratodermia variabilis (EKV) is a rare subtype of heterogeneous group of skin diseases called erythrokeratodermi. It presents with migratory erythema and fixed hyperkeratotic plaques usually distributed on the extensor surface of extremities, buttocks, and face. EKV lesions commonly occur in the early stage of life.

Herein we report a case of erythrokeratodermia variabilis in a 7-year-old child.

Materials & Methods:

Results:

A 7-year-old boy came to our department for brownish and erythematous lesions that had started 2 years ago and had appeared from time to time. The red spots initially spread centrifugally and disappeared without any mark, however, the brownish lesions on the anterior part of trunk and arms did not disappear. He was seen by different physicians and was treated with various fungal treatments that were not beneficial. No similar lesions were detected in other family members. No systemic disease was detected.

The dermatological examination showed erythematous plaques located on the anterior part of chest, thighs and limbs, associated with brownish hyperkeratotic patches on both arms. No lesions were detected on the palm, sole, nail, hair, teeth, or mucosa.

The laboratory tests were within normal limits. No fungal elements were detected in potassium hydroxide preparation.

The histopathological examination of the hyperkeratotic lesion showed basket-like hyperkeratosis, papillomatosis, acanthosis, and perivascular lymphocytic infiltration of the dermis. The clinical and histopathological findings were consistent with EKV.

The patient was treated with topical corticosteroid ointment with good resolution at 2 months follow-up.

Conclusion:

EKV is a very rare type of EK, determined by an autosomal dominant gene of variable expressivity, characterized by reddish-brown plaques, which are hyperkeratotic and fixed in location, particularly over the knees, elbows and ankles; associated with figurate patches of erythema which vary in size, shape and location. There are two major subtypes of EKV: EK variabilis (Mendes da Costa), and EK progressiva symmetrica (Gottron). The classical EKV initially described by Mendes da Costa is characterized by two types of skin lesions: Figurate hyperkeratotic plaques and transient erythematous areas. The lesions have propensity to locate on the distal extremities, buttocks, and trunk. Hyperkeratotic plaques are particularly distributed on the face, hip, and extensor aspect of the limb.

Histopathological findings of EKV are non-specific. The findings include loose hyperkeratotic stratum corneum, wide plugged hyperkeratotic follicular openings, acanthosis, papillomatosis, perinuclear vacuolization and

parakeratosis.

No specific therapy is available for EKV: The erythematous patches of EKV can be treated with topical corticosteroids. Hyperkeratotic lesions usually show a good response to topical keratolitics, topical retinoids, tazarotene, alpha-hydroxy acid, and topical corticosteroid. However, patient’s response to retinoid therapy is only limited during the therapy course. The lesions reappear during the untreated period.

Medication choice should be made based on its side-effect because of its prolonged use.

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Abstract N°: 4146

Job's Syndrome: Insights from a pediatric case report.

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Introduction & Objectives:

Job's syndrome or hyper-immunoglobulin E syndromes (HIES) is a rare, heterogeneous complex of primary immunodeficiency disorders. It is characterized by extremely high serum immunoglobulin E (IgE) levels, recurrent cutaneous manifestations like chronic eczematous dermatitis and recurrent sinopulmonary infections.

We are reporting a case of Job's syndrome in a 3-year-old child who presented with classical symptoms of Job's syndrome.

Materials & Methods:

Results:

A 3-year-old male child born out of non-consanguineous parentage and uneventful pregnancy, was referred to our dermatology department with eczematous lesions on the neck, face, upper chest and pelvic region evolving for 15 days. Physical examination showed multiple eczematous lesions associated with pustular lesions on the neck, upper chest and pelvic region. There was no history of fever and lymphadenopathy. He had similar such episodes in the past along with episodes of upper and lower respiratory tract infections. His medical history was significant for eczema since newborn period. Family history was not significantly related to condition.

Biochemical investigations revealed high serum IgE levels 3621 IU/ml associated with hypereosinophilia 2381/mm3.

The rash had been treated with topical steroids (betamethasone dipropionate) and oral vitamin C with slight resolution.

Conclusion:

HIES, also known as Job syndrome, belongs to a group of primary immunodeficiency diseases characterized by elevated serum IgE levels, recurrent infections, and various clinical features.

Although most cases are sporadic, two distinct forms are being identified: Autosomal-dominant HIES, which develops due to STAT3 gene mutation and autosomal-recessive HIES caused by DOCK8 gene mutation.

Clinical features include recurrent eczema, frequent bacterial pulmonary infections, mucocutaneous candidiasis and cold abscess. Characteristic facial features that typically appear in late childhood include asymmetry, a prominent forehead, deep-set eyes, a broad nasal bridge, a wide fleshy nasal tip, rough facial skin, an increased inter-alar distance and prognathism.

The eczema in HIES is difficult to be distinguished from atopic dermatitis. However, a long course from early life with atypical distribution such as in the axilla and groin, associated to recurrent bacterial infections and resistant to conventional treatment are signs of HIES dermatitis rather than atopic dermatitis.

Diagnosis can be problematic as there is no clear criteria and symptoms are quite diverse: It is mainly diagnosed

using clinical presentation and laboratory findings: Serum IgE levels are elevated in all patients with HIES. Also, eosinophilia of greater than or equal to 700 cells µl is seen in most cases.

There is also a scoring system to evaluate for STAT3 probability.

Treatment options are few, mainly supportive treatment with antibiotics for cutaneous and pulmonary infections, and topical steroids for eczematoid rashes. The goal of therapy should be to prevent these infections, and if they occur, to aggressively treat them to prevent the development of complications that may lead to mortality.

Despite its rarity, understanding and accurately diagnosing HIES are crucial for effective management and treatment.

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Abstract N°: 4158

Anti-platelet therapy in venous malformations. Efficacy and safety profile of the acetylsalicylic acid:an observational retrospective study

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Introduction & Objectives:

Simple, combined or complex venous vascular malformations (MVVs), especially if extensive or multifocal, can be associated with a localized intravascular coagulopathy (LIC), characterized by the formation of endoluminal thrombi in the dysplastic vessels and consumptive coagulopathy. Acute or chronic pain and consequent functional limitation are the main clinical signs. Blood tests often show increased D-dimer level and hypofibrinogenemia.

Pharmacological treatment is required both to control the pain caused by thrombotic phenomena and to

prevent progression towards disseminated intravascular coagulation (DIC).

Low molecular weight heparins (LMWH) are the first-line treatment, in particular enoxaparin. However, if the treatment is started in childhood, the use of daily subcutaneous injections can negatively impact patient's quality of life. Oral drugs, in these cases, are a possible therapeutic alternative.

Antiplatelet agents, in particular acetylsalicylic acid (ASA), have been shown to have a beneficial effect,

reducing pain with acceptable adverse events.

The objective of the study is the evaluation of the efficacy (reduction of painful symptoms due to

LIC-related thrombotic events) and safety of ASA in patients suffering from different forms of MVVs.

Materials & Methods:

A retrospective observational study was conducted in consecutive patients followed at our Vascular

Anomalies Center. All patients were affected by simple, combined or complex MVVs and in treatment with ASA for 3 months (average time) at a dosage of 0.3 mg/kg/day.

For each patient, personal data, characteristics of the MVV and clotting tests were

collected. The pain was evaluated before (T0) and after ASA (T1).

The effectiveness of ASA was measured by comparing the intensity of pain before and after administration

of the drug.

For the assessment of pain intensity, two scales were used: for patients ≥8 years old, the pain Numerical Rating Scale (p-NRS); for patients <8 years, the Wong Baker Faces Pain Rating Scale (WBFPRS).

Furthermore, the onset of any adverse events that occurred during ASA therapy was recorded.

p-NRS/WBFPRS values were reported as means and standard deviations. To calculate the mean difference

and the corresponding 95% confidence interval, the Student' T test was used. The value of P<0,005 was considered statistically significant. Statistical analyzes were performed using R-foundation for statistical computing, Vienna, Austria.

Results:

34 patients (18 M, 16 F) with an average age of 12.53± 5.43 years were enrolled. 27 patients had simple MVVs; 3 patients had a venous-lymphatic malformation; 1 patient had a capillary-venous-lymphatic malformation; 3 patients with Klippel-Trenaunay syndrome.

The average p-NRS/WBFPRS values before the administration of ASA was 7 ± 2.46. The average p-NRS/WBFPRS values, considering the same patients, after the administration of ASA was found to be 3.37 ± 3.13. The mean difference was found to be -3.640 (95% C.I. -5.0031 to -2.2769; P<0.0001) and therefore statistically significant.

No patient developed side effects.

Conclusion: ** The results of our study are in line with the few data present in the literature. The administration of ASA resulted, in the analyzed sample, in a statistically significant average reduction in painful symptoms compared to same untreated patients. Furthermore, treatment with ASA is safe.

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Abstract N°: 4256

ullous pemphigoid in children: what's so special about it?

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Introduction & Objectives: Bullous pemphigoid (BP) is an acquired autoimmune subepidermal bullous disease that mainly affects the elderly. It is rare for the disease to begin in childhood, and even more so in infancy. We report a new case of PB in a child.

Materials & Methods: A child aged two years and 09 months was referred for a bullous eruption that had started 04 weeks previously. Initially on the two inguinal folds, it progressively spread to the trunk. This was a third child from a non-consanguineous marriage, born at term vaginally without complications, exclusively breast-fed and vaccinated as recommended, with no personal or family history. On dermatological examination, there were tense vesicular and bullous lesions of varying size and serous content on healthy or erythematous skin, associated with erosive-crusty lesions. Lesions were mainly located on the face, hands, feet, palms and soles, and perineal area. There were no mucosal lesions. Nikolsky's sign was negative. The child was apyretic, in good general condition, with normal staturoposterior development and a non-abnormal extracutaneous clinical examination. The blood count revealed a mild normocytic anemia with an aegerative value of 10.2 g/dL, and thrombocytosis of 570,000/mm3. The rest of the blood work-up was unremarkable. Anti-basal membrane and anti-intercellular space antibodies revealed high positivity to AgBP230 and AgBP180. histological examination of a skin biopsy showed the presence of a bulla by dermal-epidermal cleavage. The content of the bulla was serous and contained numerous inflammatory elements. Clinical, laboratory and histopathological data confirmed the diagnosis of infantile bullous pemphigoid. Treatment began with prednisolone 2mg/kg, with good lesion control. To allow weaning from corticosteroids, dapsone was introduced at 0.5mg/kg. Previous follow-up examinations had shown normal G6PD levels, and no further worsening of the anemia occurred with the medication

Results: The first case of bullous pemphigoide in a child was described in 1970 on the basis of an immunofluorescence diagnosis. Since then, the number of reported pediatric cases has steadily increased, prompting experts to propose diagnostic criteria for pediatric PB that included children and adolescents up to the age of 18. In 2008, Waisbourd-Zinman et al. noted that clinical presentations varied according to the age of affected children. In a review of the literature, they showed that the majority of cases of infantile PB occurred in small children, and that these small children presented a particular clinical picture with acral involvement. The distribution in older children was much less uniform, and included a subgroup of children with localized genital BP. IgG autoantibodies to the NC16A domain of BP180 are usually found. The frequency of these antibodies in childhood BP would be similar to that observed in adults.

Conclusion: This condition, although rare, should always be included in the differential diagnosis of bullous dermatoses in children. In contrast to what is observed in adults, the prognosis of PB in children is good.

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Abstract N°: 4282

Prevalence of Vitamin D Deficiency in Children with Inherited Ichthyosis: A Transversal Controlled Study

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Introduction & Objectives:

Inherited ichthyosis constitutes a heterogeneous group of cornification disorders. Its healthcare management is a challenge for most specialists doctors in dermatology. The development of vitamin D deficiency in patients with inherited ichthyosis has recently been observed, yet exact cause of such association is not properly understood. To date, few studies have investigated serum vitamin D status in patients with inherited ichthyosis.

The objective of this study was to determine the prevalence of vitamin D deficiency of patients suffering from inherited ichthyosis, and to identify the different factors associated with this deficiency.

Materials & Methods:

This was a cross sectional study performed in the University Hospital center at the dermatology department among two groups: patients with inherited ichthyosis and patients without this disease. Patients' clinical characteristics were collected. Serum concentration of 25-hydroxyvitamin D was determined among the two groups. Comparisons between the two groups conducted by bivariate analysis.

Results:

46 patients were included in the present study. There was a majority of males (60.9%) with a median age of 04.5 years (0.16 - 20). The majority (84.8%) had serum 25-hydroxyvitamin D below the optimal level of 30 ng/mL with 19.6% in sufficiency, 43.5% in insufficiency, 47.9% in deficiency. The median age among patients with inherited ichthyosis was 3 years old (0.16 - 20) and the sex ratio male/female was at 2.28. In addition, 87% was below the optimal level of 25-hydroxyvitamin D. The mean of serum level of 25-hydroxyvitamin D was lower at in patients with inherited ichthyosis 18.24 ± 10 ng/mL in comparison with control group 22.0 ± 15.0, (p = 0.19). Among patients with inherited ichthyosis, 56.5% had a lamellar form, 39.1% had a vulgaris form and 01 case had netherton disease. The dander scales were mainly thin in (60.9%). More than a half of patients had no erythema (60.9%) and no itching (65.2%). 65.2% had an ectropion. Moreover, oral retinoids were used as the main treatment in 52.2% of cases. The patient with netherton disease had a thin dander scale, an erythema, suffered from itching and had not an ectropion. Also, he is in insufficiency according to the classification of 25(OH). A bivariate analysis had identified that ichthyosis vulgaris form was mainly associated to ectropion (p = 0,02) and thick dander scales (p = 0.001). The mean of serum 25-hydroxyvitamin D was less among patients with lamellar ichthyosis form (19.71 ± 17.83) in comparison with patients with vulgaris ichthyosis form (17.28 ± 10.10).

Conclusion:

In summary, measurement of vitamin D levels should be recommended in patients with ichthyoses, especially for individuals with severe ichthyosis, the lamellar ichthyosis, and oral retinoids. This recommendation does not refer to distinct age groups. Treatment options proposed apart moisturizers, topical keratolytics and topical and systemic retinoids is recently systemic vitamin D analogues. Further studies are needed to confirm that vitamin D

levels are a consequence of the cornification disorders or are related to its physiopathology.

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Abstract N°: 4296

Acrodermatitis enteropathica-like lesions in a child revealing a celiac disease

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Introduction & Objectives:

Coeliac disease is an immune-mediated, inflammatory disease of the proximal small intestine caused by gluten sensitivity in genetically susceptible individuals. Zinc deficiency in patients with celiac disease is common, it is associated with a wide range of skin lesions. Herein, we report the case of an 18- month-old child who developed secondary acrodermatitis enteropathica revealing a celiac disease.

Materials & Methods:

Results:

An 18-month-old girl presented with recent onset of skin lesion in the perioral region and feet, with a history of hematemesis. Past and family histories did not suggest gastro-intestinal disease. She had been exclusively breastfed for 6 months, and after 6 months was continued on partial breast-feeding along with a complementary diet that included wheat and barley. On examination, there was a hemorrhagic gingivostomatitis with perioral erythema topped with multiple ulcers extending down to the neck, as well as erythema and edema of both feet. The patient received oral acyclovir and antibiotics for ten days with no improvement. Laboratory tests showed a microcytic hypochromic anemia (Haemoglobin was 8,9 g/dl) with a decreased alkaline phosphatase level at 49 U/l. He started on zinc supplementation at a dose of 20mg/day with a complete resolution of lesions after two weeks. Duodenal biopsy showed a loss of normal villous structure and an increase intraepithelial lymphocyte suggesting a celiac disease.

Conclusion:

Acrodermatitis enteropathica is a constellation of skin lesions resulting from severe zinc deficiency. Coeliac disease is a malabsorption syndrome in which the proximal small intestine is commonly affected. As the small intestine also plays a major role in maintaining zinc homeostasis, zinc deficiency is common in patients with celiac disease. It may result from excessive loss and from impaired absorption secondary to damaged intestinal epithelium. Dermatological manifestations include vesiculo-bullous, eczematous, dry and scaly or psoriaform lesions over the peri-oral, peri-anal and acral areas, the elbows, knees and cheeks bilaterally. The lesions can spread rapidly to a weeping desquamation and infection. The confirmation of the diagnosis is based on the dosage of the plasma zinc or the diagnostic test by zinc supplementation. In children, vesicles, erosions and crusting in the periorificial area can be mistaken for a herpetic gingivostomatitis or a bullous impetigo. In our case, the lack of improvement under antivirals and antibiotics as well as the involvement of the feet allowed us to reconsider the diagnosis.

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Abstract N°: 4304

Transitory dermatoses in Neonatal Intensive Care Unit (NICUs).

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Introduction & Objectives:

Dermatological affections are common among the pediatric population. Transitory neonatal dermatoses are skin conditions that appear in newborns immediately or shortly after birth and disappear spontaneously after a few days or weeks. The aim of this study is to determine the epidemiological profile of transitory neonatal dermatoses in Neonatal Intensive Care Unit, and their various clinical aspects.

Materials & Methods:

This is a descriptive cross-sectional study including newborns admitted for various medical conditions and presenting dermatological abnormalities in the neonatology department of the Souss-Massa University Hospital in Agadir, over a period extending from August 2023 to February 2024.

Results:

During the study period, 649 newborns were hospitalized. A total of 45% of newborns presented dermatological manifestations (N=292). The average age was 5 days. The sex ratio (M/F) was 0.9.

Transitory sebaceous hyperplasia of the newborn was the most frequent dermatosis (55.82%), appearing as pinhead micro-papules on the nose and cheeks. Milia was another frequently found dermatosis in our patients (20.89%), in the form of whitish papules on the face or trunk.

Neonatal pustular lesions were mainly represented by neonatal benign cephalic pustulosis (28.42%), which clinically consists of papules and pustules without retention lesions, followed by toxic erythema (11.98%) which appears in the form of papules and pustules on the body.

Capillary malformations, notably Unna angiomas, were observed in 22.94% of cases, sitting on the forehead with a “V” shape.

Meanwhile, pigmentation abnormalities were dominated by mongoloid spots (36.3%), presenting as bluish macules, located essentially in the lumbar region. Only one case of transient genital hyperpigmentation was reported.

Cytosteatonecrosis was observed in 5 newborns who presented neonatal sufferance, with a median age of 11 days. It is characterized by the presence of subcutaneous indurations. Only one patient had hypercalcemia complicated by a grade 2 nephrocalcinosis.

The rest of the neonatal dermatoses, in particular neonatal desquamation, were observed in 13.01% of cases.

Conclusion:

Neonatal dermatoses are multiple and diverse, hence the need for their knowledge in order to guide investigations in case of atypical presentation or persistence of lesions.

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Abstract N°: 4305

The profile of palmoplantar keratodermas in pediatric dermatology

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Introduction

Palmoplantar keratodermas (PPK) in children represent a group of rare but significant dermatological pathologies, which can have a profound impact on the quality of life of children, affecting not only their physical health but also their emotional and social well-being. The objective of our work is to study the epidemiological, clinical, and etiological profile of children followed for PPK in pediatric consultation.

Materials and Methods

This is a retrospective descriptive study conducted over a period of 5 years between January 2019 and January 2024, including children aged (0-15 years) followed for PPK in pediatric dermatology consultation.

Results

A total of 37 patients were included in the study, comprising 23 boys and 14 girls with a sex ratio of 1.64. The mean age was 8.62 years (2-15 years), and the mean duration of symptoms before consultation was 2.88 years (2 months to 12 years). The main medical history found in our series included atopy (4 cases), consanguinity (2 cases), and dental anomalies (2 cases). The mean age at onset was 5.6 years (4 months to 12 years). The keratosis was predominantly palmoplantar in 31 cases, plantar in 5 cases, and palmar in only 1 case. Skin lesions were erythematous-squamous in 11 cases, hyperkeratotic in 10 cases, and fissured in 26 cases. Pruritus was present in 17 patients, and pain was reported in 5 patients, mainly associated with fissured lesions. Biopsy was performed in 11 patients, revealing psoriasiform dermatitis in 7 patients, lymphocytic infiltrate with epidermotropism in one patient, spongiotic dermatitis in one patient, and spongiotic pustules in 2 patients. Mycological sampling was performed in 4 patients, showing Candida albicans in one patient, and being sterile in other cases. The causes of PPK were inflammatory in 32 cases (psoriasis, eczema, pityriasis rubra pilaris), hereditary in 3 patients (papillon-lefevre syndrome, clouston syndrome, variable erythrokeratodermia), one case of mycosis fungoides, and one case of candidal PPK. Most patients were treated with a combination of dermocorticosteroids, balneotherapy, and healing cream with good outcomes. Six patients were treated with acitretin, including 3 cases of hereditary PPK, 2 cases of pustular psoriasis, and 1 case of pityriasis rubra pilaris.

Discussion

Palmoplantar keratodermas in children represent a complex area of study within pediatric dermatology. It is a rare reason for consultation with multiple etiologies, which can be hereditary, acquired, or associated with genodermatoses. Its course is often chronic, requiring long-term therapeutic management. Our study demonstrates that a comprehensive approach, taking into account the diversity of clinical presentations, medical history, results of complementary examinations, and underlying etiologies, is essential for optimal management of PPK in children. This integrated approach aims to provide optimal treatment to minimize the functional impact of this disease on the child’s daily activities.

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Abstract N°: 4389

"Blueberry muffin baby" as presentation of congenital leukaemia

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Introduction & Objectives:

Materials & Methods:

Results:

Congenital leukaemia is defined as occurring within the first 28 days of life and represents less than 1% of all childhood leukaemias. The cutaneous infiltration by leukemic cells, referred as leucemia cutis, occurs in 25 to 30% of cases.

We describe a case of a 1-month-old male infant with a congenital dermatosis initially affecting the scalp and with subsequent dissemination. Uneventful full-term pregnancy, deliver and neonatal period. Physical examination revealed pallor but a good general status and multiple violaceous nodules on the scalp, as well as violaceous papules on the trunk, left axilla and genital area. There was no mucous membrane involvement, lymphadenopathies or hepatosplenomegaly. Blood analysis revealed hypoproliferative anaemia, neutropenia and serologies for the main congenital infections were negative. Abdominal ultrasonography was normal. Skin biopsy shown a dermal infiltration by leukemic cells and immunophenotyping confirmed the presence of 2 cellular populations of monocytic lineage. Similar cells were identified in the bone narrow, peripheral blood and cerebrospinal fluid. Clinical and laboratorial data led to the diagnosis of congenital acute myeloid leukaemia. Cytogenetic analysis of the bone marrow identified a previously undescribed translocation t(10;11)(q11;p15). The patient underwent chemotherapy with mitoxantrone, etoposide and cytarabine with complete resolution of the skin lesions within 2 weeks. After different chemotherapy regimens, the infant is waiting for a hematopoietic stem cell transplantation.

Congenital leukaemia cutis usually presents with a disseminated violaceous papulonodular eruption, described as "blueberry muffin baby" rash. In this clinical case, despite the absence of hepatosplenomegaly and hyperleukocytosis, pallor and anaemia are common manifestations of congenital leukaemia. Diagnosis is established by confirming skin infiltration by leukemic cells, detection of an immature hematopoietic stem cells proliferation in the bone marrow and exclusion of leukemoid reactions. The monocytic variant of acute myeloid leukaemia is the most common congenital leukaemia. Treatment is based on chemotherapy and hematopoietic stem cell transplantation, thought there is no established protocol. The prognosis is poor, with a 2-year survival rate of only 20%.

Conclusion:

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Abstract N°: 4414

Neonatal nephrocutaneous syndrome associated with EGFR mutation

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Introduction & Objectives:

Materials & Methods:

Results:

EGFR is a transmembrane protein with tyrosine kinase activity, involved in the differentiation of keratinocytes and regulation of skin inflammation. Homozygous germline mutation of the EGFR gene (Gly428Asp) is associated with a rare nephrocutaneous neonatal syndrome, described for the first time in 2014.

Newborn female with pre-term delivery at 30 weeks of gestation, due to premature rupture of membranes and polyhydramnios. Parental consanguinity and family history of a paternal uncle with an unknown congenital pathology, who died in the first months of life. On physical examination the patient had a very low weight, bilateral ankyloblepharon and arachnodactyly, associated with erythroderma and progeroid appearance with atrichia and scarce supracilia. Within 2 weeks, there was a progressive improvement in erythroderma with widespread fine icthyosiform scaling. There were also signs of skin fragility with erosions and purpuric macules. During the first two months of life there were several episodes of cervical, axillary and diaper area candidiasis, perianal dermatitis, impetigo, medium/external otitis and multiples episodes of sepsis. The patient also developed a persistent papulopustular eruption of the scalp. Blood analysis revealed a secondary hyperaldosteronism, associated with severe and frequent hydroelectrolytic disorders. Transthoracic echocardiography was normal. Complete exome sequencing revealed a homozygous EGFR mutation (Gly428Asp), leading to nephrocutaneous neonatal syndrome associated with EGFR mutation diagnosis. The patient remained hospitalized with daily emollient application, continuous correction of hydroelectrolytic imbalances and frequent cycles of antibiotics and antifungals (topical and intravenous). However, the persistence of this complex clinical course, disease's poor prognosis and loss of venous accesses, led to the decision of best supportive care in the second month of life. The patient died 2 weeks later.

Nephrocutaneous neonatal syndrome associated with EGFR mutation typically presents as premature newborns with intrauterine growth restriction, polyhydramnios and familiar history of consanguinity. The icthyosiform desquamation, cutaneous fragility, atrichia and scarce supracilia are common skin manifestations. Papulopustular eruptions are described in some cases. Although absent in our case, centrofacial dysmorphisms, nephromegaly, congenital cardiopathies, recurrent vomiting and diarrhoea, are frequently reported. The complex clinical course with frequent infections, sepsis and severe hydroelectrolytic disorders, is a distinctive feature of this syndrome. Despite the diagnosis confirmation by genetic testing, the disease remains without effective therapy, with most patients dying within the first 6 months of life.

Conclusion:

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Abstract N°: 4486

Erythema annulare centrifugum in a child with leukemia

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Introduction:

The erythema annulare centrifugum (EAC) is a rare inflammatory dermatosis of uncertain etiology and variable associations such as malignancies, autoimmune diseases, food, and medications. There is no predilection for race or gender and it is most prevalent between the 5th and 6th decades of life. It manifests as erythematous, annular, or polycyclic plaques and may present with scaling or pruritus. It reaches diameters ranging from 6 to 10 cm and progresses with central healing and centrifugal growth, with a predilection for the trunk and extremities. Cutaneous lesions are believed to result from a type IV hypersensitivity reaction to the etiological agent. EAC can be classified as superficial, generally more common, with desquamation and pruritus; and deep, with hardened edges, without desquamation or pruritus. The diagnosis is clinical, and a biopsy assists in confirming the condition. Investigation and exclusion of other underlying diseases and possible triggering agents should be performed, even though in the vast majority the conclusion is idiopathic. Differential diagnoses include dermatoses that present with annular lesions such as dermatophytosis, psoriasis, among others. EAC is usually self-limiting and tends to regress spontaneously or after treatment of the underlying cause.

Case report

A 10-year-old girl presented with erythematous lesions that had been progressively growing on her face and anterior chest over the past 15 days, without any associated symptoms. She denied recent infections or vaccinations, as well as the use of topical medications, having allergies, or a family history of skin diseases. She had been diagnosed with acute lymphoid leukemia a year ago and had been undergoing maintenance treatment with methotrexate and mercaptopurine for the last 5 months, along with prophylaxis using acyclovir, azithromycin, and sulfamethoxazole with trimethoprim. During the physical examination, erythematous annular plaques with centrifugal growth were observed, appearing poorly defined with a smooth surface. These lesions were found on her anterior chest as well as in the bilateral frontotemporal and malar regions, with the largest measuring 10 cm in diameter. Laboratory tests revealed no abnormalities. Histopathological examination indicated superficial and deep perivascular dermatitis, displaying a ‘cuff-like’ infiltrate consistent with the clinical diagnosis of centrifugum annular erythema. The lesions spontaneously regressed over the course of one month, and there have been no recurrences to date.

Discussion

Erythema annulare centrifugum has been described as a facultative paraneoplastic dermatosis associated with hematological malignancies such as leukemia and lymphoma. In this case, after excluding other etiologies, the hematological disease is believed to be the possible triggering factor for the cutaneous condition.

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Abstract N°: 4518

Cutaneous manifestations of microscopic polyangiitis in children and adolescents

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Introduction & Objectives: Although better recognized than previously, microscopic polyangiitis (MPA) associated with perinuclear antineutrophil cytoplasmic antibodies (pANCA) specific to myeloperoxidase (MPO) is a very rare disease in children.

Materials & Methods: We retrospectively analyzed the clinical, histological and serological parameters of childhood MPA, diagnosed from 2002 to 2023. Disease activity was assessed according to the Birmingham Vasculitis Activity Score (BVAS).

Results: Ten girls and one boy aged 12.0+/-2.4 years (range: 5-17 years) met the following criteria: (1) clinical manifestations of systemic small vessel involvement; (2) histological demonstration of pauci-immune necrotizing glomerulonephritis; and (3) serological findings of increased concentration of MPO-ANCA by ELISA test. The main clinical manifestations were: influenza-like symptoms (100%), hematuria/proteinuria (100%), purpura or palpable purpura (100%), pulmonary-renal syndrome (57%), acute renal failure (ARF) (29%), ischemic cerebral insults (29%), and necrotizing vasculitis of the skin (29%). Gangrene and cutaneous necroses were seen in patients who had end-stage renal disease (ESRD) and/or severe renal histopathological features. BVAS ranged from 22 to 36 (25.7±6.4) of possible maximal score 63. Patients were followed from 4 to 15 years (9±2 years). None of our patients died. Two of 11 children who had ARF progressed to ESRD; two developed chronic renal failure, and 7 normalized renal function. Gangrene and cutaneous necroses, ARF and central nervous system involvement at presentation were parameters of poor renal outcome.

Conclusion: MPA should be considered early in the differential diagnosis of children presenting with purpura or palpable purpura together with constitutional symptoms and hematuria. The diagnosis must be confirmed by the detection of MPO-ANCA. Cutaneous presentations have diagnostic (purpura) and prognostic significance (gangrene and cutaneous necroses). The early treatment enables a favorable prognosis of MPO-ANCA-associated MPA in children.

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Abstract N°: 4523

Lichenoid drug eruption in children

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Introduction & Objectives:

Lichenoid drug eruption is an uncommon adverse drug reaction characterized by papulosquamous eruption, with possible prolonged latency period to the time of lesion manifestation. It is difficult to distinguish it from Lichen planus due to their clinical and histological similarities. In children, lichenoid drug eruption is exceptionally rare with few cases published.

Materials & Methods:

This is a case of a 10 years old male who presented at our facility with a history of generalized skin

lesions for 8 weeks. The lesions started on the neck as a few scattered papules and within one

month they have progressively spread to the trunk, limbs, face and are associated with moderate

pruritus that is exacerbated by hot weather. Before presenting to our facility, he sought medical

treatment at a local facility where he initially received topical clotrimazole cream, with no relief. A

week later lesions were progressing and subsequently, he received ceftriaxone, metronidazole,

paracetamol and cetirizine, he reported improvement in pruritus but no regression in the lesions. He also has a history of being treated for upper respiratory tract infection with oral antibiotics and

analgesics in August and December 2023 unfortunately the mother cannot remember the drugs

given.

On examination, he had generalized coalesced violaceous and hyper-pigmented flat-topped papules and scaly plaques that are symmetrically distributed, with marked desquamation on the back. However, sparing the scalp, palms, soles, nails, and mucous membranes.

Results:

On dermoscopic examination: there was absence of Wickham striae, with elements of erythema and hyperpigmentation, and on histology, it showed infiltrates of inflammatory cells, predominantly of eosinophils and prominent vessels in mid dermis.

The patient was started on topical clobetasol ointment 0.05% for the body and betamethasone

ointment 0.1% for the face. Follow-up was in two weeks and there was a great improvement on the lesion, he presented with shiny post-inflammatory hyper-pigmented macules, patches and papules.

Conclusion:

In our setting, diagnosis of Lichenoid drug eruption can be challenging due to the limited number of

dermatologists in the country, especially in rural areas, where patients are seen by clinical officers with little knowledge in dermatological conditions.

Hence medical personnel at primary health care level need to be able to take a comprehensive

clinical, medical, and drug history as well as physical examination to aid in distinguishing

lichenoid drug eruption from other papulosquamous disorders, even when the culprit cannot be identified.

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Abstract N°: 4546

BCGites : A 40 case series of an overlooked complication

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Introduction & Objectives:

Antituberculous vaccination using the BCG, an attenuated live vaccine, is still routine at birth in many countries, including ours.

Materials & Methods:

A retrospective descriptive study of BCGitis cases in infants under 2 years old.

Results:

40 patients were included (15F / 25M). Vaccination occurred within the first 48 hours of life, except for two vaccinated at 1 month. Symptom onset varied from 1 to 24 months. Thirty infants had a locoregional form. Twenty-five patients had left-sided adenitis at the injection site, fistulized in 14 patients : axillary (n=18), lateral-cervical (n=3), submandibular (n=3), and supraclavicular (n=1). Three patients had lingering ulceration at the injection site, and 2 had fluctuating swelling in the left thigh, diagnosed as BCGitis histologically. All patients responded well to local care except for two: one with PCR-positive BK adenitis and one with BCG-confirmed tuberculous lupus requiring systemic antituberculous treatment (ATT).

Ten patients had disseminated BCGitis, nine of them immunocompromised (5 severe combined immunodeficiency, 2 chronic granulomatous disease, 1 IL-12 INFγ axis deficiency, and 1 HLA class 2 deficiency). Four patients had multiple cutaneous lesions, and 8 had systemic manifestations with positive BK in the bone marrow. Under systemic ATT, 6 rapidly passed away, and 4 improved well after 12 months of treatment.

Conclusion:

BCGitis frequency post-vaccination is low compared to the large number of vaccinated individuals, with locoregional forms being common due to technical errors, deep injection, and high doses. Strain variation and early vaccination (<1 month) correlate with increased risk. The unusual thigh location can be explained by vaccination errors at birth: either BCG injection into the thigh or shared use of the same needle for BCG and vitamin K injections. Treatment for BCGitis is not standardized, ranging from observation with monitoring to symptomatic treatment, systemic ATT or surgery. Most locoregional cases resolve spontaneously.

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Abstract N°: 4590

Paradoxical psoriasiform skin eruption in pediatric patients with inflammatory bowel disease treated with TNFα Inhibitors

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Introduction & Objectives:

Tumor necrosis α inhibitors (TNFαI) are a group of monoclonal antibodies used to treat various inflammatory diseases, including inflammatory bowel diseases (IBD), psoriasis, hidradenitis suppurativa, rheumatoid arthritis, ankylosing spondylitis, juvenile idiopathic arthritis and uveitis. Adverse events related to this treatment include infections, increased risk of malignancies, demyelinating disorders and skin reactions. One type of skin reaction induced by TNFαI is paradoxical psoriasiform eruptions. The mechanism of action of this adverse event is not fully understood, especially since TNFαI can be used as a therapeutic options for patients with psoriasis. Presumably, the inhibition of TNFα causes increased interferon α expression and homing of Th1 cells to the skin, resulting in psoriasiform dermatitis. Psoriasiform eruptions have been widely reported in adults, but data in children are relatively limited. We aimed to describe the clinical characteristics of the TNFαI-induced psoriasiform eruptions and therapeutic options in pediatric patients with IBD.

Materials & Methods:

We reviewed the medical charts of pediatric patients (aged <18 years old) with IBD who developed TNFαI-induced psoriasiform eruptions during 2006-2022.

Results:

Among 454 patients with IBD treated with TNFαI, 58 (12.8%) were diagnosed with TNFαI-induced psoriasiform eruptions, of whom 51 were included in the study. The female to male ratio was 1:1.3. The median age at skin eruption was 14.1 [IQR, 12.11-16.05] years. The median elapsed time to eruption appearance was 15 [IQR, 7-24] months after initiation of the treatment. All the patients were treated with topical steroids and 17 (33%) needed systemic treatment (phototherapy, methotrexate or acitretin). Sixteen patients (31%) needed to stop TNFαI treatment due to an intractable eruption. Female patients, patients with inflammatory alopecia and patients who were treated with methotrexate or phototherapy were more prone to stop TNFαI.

Conclusion:

TNFαI-induced psoriasiform eruptions are prevalent in pediatric patients with IBD and may appear even years after the treatment was initiated. In most patients these eruptions are manageable, but some need a change in treatment due to the uncontrolled eruption. Further studies are needed to determine whether a medication is needed for these patients that is targeted to both the gastrointestinal inflammation and the skin involvement, or if removal of the inducing agent is the key for cutaneous cure. Given the high prevalence and the complexity of managing these patients, multidisciplinary collaboration between pediatric gastroenterologists and dermatologists is recommended.

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Abstract N°: 4729

Changes in the concentration of micronutrient (MN) composition and hair structure in children with alopecia

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Introduction & Objectives: The science of micronutrients (MN) remains at the stage of amassing factual material. Hair is a reliable and non-invasive source of information about the MN content in the body, for its metabolism is slow and only long-term disturbances in the concentration of nutrients can be reflected in it. The attention of many researchers has been drawn to the hypothesis of the influence of exogenous and endogenous factors on the hair functional status, especially with regard to essential and toxic MN.

Today a tendency towards an increase in the number of paediatric patients with hair diseases ranging from excessive hair loss to various clinical forms of alopecia is observed.

Objectives. The study was aimed to identify the MN imbalance, manifested through the condition and appearance of hair, to follow structural changes in hair and to assess the influence of internal organ pathologies.

Materials & Methods: During one year 27 patients with various forms of alopecia and 15 basically healthy children were examined using various methods, which included laboratory-instrumental examination and counselling by appropriate experts. Hair MN content was determined via quantitative and qualitative emission spectral analysis using spectrographs STE-1 and ICP-51. The hair structure was studied employing scanning electron microscopy (SEM) performed with a TESLA BS-300 scanning electron microscope.

Results: There were 17 boys and 10 girls with focal, marginal and diffuse forms of alopecia, which at the disease onset was manifested by increased hair loss. Digestive disorders (chronic gastritis, hepatitis, intestinal dysbacteriosis) and endocrinopathies (thyroid antibody titre, clinical disorders of glucose tolerance) were detected for the first time and prevailed in 83% of examined subjects.

Resting on quantitative indices we established the difference between the ME content in hair of patients suffering from a variety of forms of alopecia and the norm. Statistically reliable in marginal alopecia is the increase in magnesium 30.7+-11.81 μg/g, chromium 1.54+-0.48 μg/g, manganese 2.9+-0.9 μg/g, iron 11.3+-3.64 μg/g, copper 3.71+-1.15 μg/g, barium 6.7+-2.6 μg/g, lead 3.11+-1.0.9 μg/g, and in focal alopecia a statistically true elevation of vanadium 0.57+-0.25 μg/g, in diffuse alopecia silicon 18.5+-6.6 μg/g, iron 8.75+-2.93 μg/g, copper 2.95+-1.02 μg/g was observed. SEM revealed several types of structural changes in the root structure: a) with remnants of root sheaths; b) atrophic, without root sheaths; c) with root sheaths. The stem structure showed protrusions and depressions, absence of a tile-like pattern.

In characterising the hypothetical mechanism of the possible toxic effect of the above-mentioned MNs, which hair content significantly statistically differs from that of the reference group, it can be assumed that it involves a complex interaction, which causes possible occurrence of increased hair loss.

Conclusion: Increased hair loss and various forms of alopecia are associated with significant MN imbalance. Changes in the structural and spectral makeup of hair in examined patients indicate the atrophic nature of alopecia and the expediency of providing patients with appropriate pathogenetic therapy taking into account the MN hair content.

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Abstract N°: 4831

Gianotti-Crosti syndrome - an underdiagnosed disease

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Introduction

Gianotti-Crosti syndrome is described as a rare and self-limiting disease characterized by a papular eruption, primarily affecting regions such as the face, buttocks, extensor surfaces of the upper and lower limbs, symmetrically, with possible itching. It predominantly occurs in children aged 2 to 6 years, and rarely in adults. There is no predilection for race or gender. Often, an infectious episode precedes the presentation. The pathophysiology of the disease is still uncertain but it is believed that the lesions result from a type IV hypersensitivity reaction in response to the causative agent. It has been associated with hepatitis B virus and Epstein-Barr virus, and occasionally linked to other agents such as herpes virus type 6, enterovirus, cytomegalovirus, parvovirus B19, adenovirus, influenza, hepatitis A, rotavirus, respiratory syncytial virus, among others, and more recently coronavirus. Although rare, the condition can also occur after vaccination. Diagnosis is clinical and laboratory tests are not specific. The main differential diagnoses include papular urticaria, scabies, hand-foot-mouth disease, and other viral rashes. In most cases, prognosis is excellent and treatment expectant, with spontaneous regression of the lesions within a year. When there is intense itching, antihistamines are indicated.

Case report

A 2-year-old boy presented with itchy skin lesions on his face, upper and lower limbs for 3 weeks. He was referred by the primary healthcare unit due to persistent lesions after treatment for scabies with permethrin lotion and ivermectin. Two weeks before the onset of the condition, he had experienced flu-like symptoms. The mother reported no other health issues or allergies, and confirmed that his vaccinations were up-to-date, although he did not receive any recent immunization. On physical examination, normochromic micropapules and erythematous-crusted papules were observed on the face, upper limbs, and lower limbs, symmetrically distributed and sparing palms and soles. There were no palpable lymph nodes or organ enlargement. Considering Gianotti-Crosti syndrome, the mother was informed about the benign course of the disease and the resolution of the lesions within weeks to months. An antihistamine was prescribed for itching and low-potency corticosteroid for excoriated lesions.

Discussion

We report this case to raise awareness about a frequently underdiagnosed condition. Although respiratory viruses are not the primary implicated, it is important to consider this diagnosis in children with skin lesions and a previous history of respiratory infections to avoid unnecessary procedures and treatments. The described lesions, typically papular with symmetrical distribution on the face, buttocks and limbs, serve as key indicators. Clinical diagnosis suffices and additional tests are not required.

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Abstract N°: 4908

A rare case of periocular infantile hemangioma

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Introduction & Objectives: Infantile hemangiomas (IHs) are proliferative, benign tumors of vascular endothelium that may be present at birth or more commonly become apparent in the first 2 weeks of life. The incidence is increased in preterm infants. Subcutaneous periocular hemangiomas are of particular concern since they may extend deep into the orbit, causing exophthalmos or globe displacement with only subtle cutaneous manifestations. Treatment with local or systemic therapy beta-blockers such as propranolol markedly improved the prognosis of IH.

Materials & Methods: We present a 11 month old baby boy with no family or personal history of interest. On the 4th day after birth the mother noticed a slightly red lesion located on the inner side of the lateral canthus of the left eye. His IHReS score was 8 points so he was referred by his GP to an expert center. The lesion grew rapidly and started to imply pressure on the left eyeball. After a consultation with a dermatologist, ophthalmologist and pediatrician the baby underwent an MRI. An orbital MRI showed tissue formation, in hypo signal T1, in hyper signal T2, containing serpiginous areas of vascular origin, this orbito-palpebral lesion implies pressure on the eyeball. PHACE (posterior fossa anomalies, hemangioma, arterial anomalies, cardiac anomalies, and eye anomalies) syndrome, Retinoblastoma and subconjunctival hemorrhage were excluded due to imaging studies. The decision after the results was to start a systemic therapy with Propranolol oral solution with slowly titrating the dose. The first week, the baby was given 1 mg/kg with good clinical responses and no adverse effects. In the second and third week the dose was 3 mg/kg leading to better clinical outcome – the edematous lesion rapidly shrunk up. Due to the good clinical response , the treatment was continued for 8 months.

Results: Predictive factors for ocular complications, such as astigmatism and amblyopia,

include size >1 cm in diameter, a deep component, and upper eyelid involvement. Our patient had two of the aforementioned: the diameter was more than 1 cm and there was a deep component of the hemangioma. Treatment was started as soon as feasible and before the completion of the proliferative phase, which resulted in rapid regression of the vascular tumor.

Conclusion: Hemangioma of any size or morphology in the periorbital location may pose a threat to vision. There is a risk of vision impairment, occlusion of the pupil,compression of the eyeball and expansion into the retrobulbar space, leading to amblyopia, strabismus, astigmatism. Treatment of infants with periocular IHs requires a multidisciplinary approach . Patients with periocular IH have a lower rate of amblyopia now compared with the pre propranolol era and emphasizes the importance of early treatment of periocular IH to prevent permanent visual sequelae.

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Abstract N°: 5056

Dentition development anomalies in facial segmental hemangiomas not related to PHACES

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Introduction & Objectives:

In recent years, the association between infantile hemangiomas (IH) affecting the maxillary or mandibular area and different alterations in dental development have been described, especially in the context of PHACES syndrome.

Our objective was to describe not previously found dental development alterations in two girls with facial IH who did not fulfilled PHACES syndrome criteria.

Materials & Methods:

We present the cases of two female patients aged 8 and 6 years respectively who were followed by our team because of segmental facial hemangioma with subsequent studies which did not reveal any abnormalities; therefore, they did not meet the criteria for PHACES syndrome.

Both patients were treated with oral propranolol, achieving an optimal and fast response.

Results:

In the first case, a delay in eruption of deciduous teeth was observed in the left inferior hemiarch, where the hemangioma was more intense. At the age of 5 years, enamel hypoplasia was observed, along with caries and tooth fusion. An orthopantomography was performed at the age of 7 years, showing agenesia of permanent incisives in the left inferior hemiarch, as well as other dentition anomalies. In the second case, on the right upper and inferior hemiarchs, where the hemangioma was located, hypoplasia and cavities on some primary and permanent molars, and one supernumerary tooth, were observed.

Conclusion:

We present two patients with facial segmental IH not related to PHACES, who developed multiple dentition abnormalities. To our knowledge, some of these abnormalities have not even been reported in the context of segmental IH.

Evidence of an association between IH and developmental anomalies seems to be strong. These anomalies may be severe, and not only in PHACES-related cases, as shown in these cases.

This association is probably underdiagnosed owing to the lack of protocolized dental studies on IH patients and follow-up. Therefore, we suggest screening for these anomalies with X-ray imaging during transitional dentition in cases of IH affecting the maxillary and mandibular areas.

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Abstract N°: 5143

palmoplantar psoriasic keratoderma in children : a retro-prospective study of six cases

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Introduction & Objectives: Psoriasis is a chronic inflammatory disease of multifactorial etiology, affecting 0.51 to 11.43% of adults and 0 to 1.37% of children. Palmoplantar psoriasic keratoderma in children (PPPK) is a common clinical subtype of childhood psoriasis , whose epidemiological characteristics remain unknown . The objective of our study is to determine the clinical and therapeutic epidemiological profile of this condition

Materials & Methods: This retrospective case study carried out on 06 children affected by PPPK, followed in the dermatology department of the EHU of Oran from January 2023 to March 2024. Epidemiological, clinical and therapeutic data were collected from their medical files .

Results: Six children were included (mean age: 10 years; M/F = 3/3, sex ratio: 1), the average age of onset of the disease was 4.17 years. Two children had a first-degree family history of psoriasis. The triggering factors identified were episodes of nasopharyngitis and school stress. None of the children was obese or overweight, only one child (16.67%) had PPPK only and the others (83.33%) had droplet lesions on admission, erythematous -squamous plaques < 30% of CS and scalp involvement. All had nail damage such as thimble punctures, trachyonichia and onychorrhexia . Only one child had associated juvenile idiopathic arthritis. The severity and quality of life scores used in our study at admission were the SPI and cDLQI. The average simplified psoriasic index (SPI) severity score was 9.25 and the childDLQI was 14.83. Five children were receiving treatment with acitretin , at dosages varying between 10 mg twice a week and 10 mg per day (0.1-0.25 mg/kg/day), while only one child was treated. with methotrexate at a rate of 20 mg per week, These treatments are well tolerated clinically and biologically, leading to a notable improvement in their condition, the only side effect observed was moderate cheilitis, often linked to the use of the acitretin.

Discussion : PPPK constitutes a common form of psoriasis in children, the association with other psoriasis conditions is common, notably nail involvement . Severity scores such as the PASI are mostly used by analogy with adults, without validation in the pediatric population .

We concluded that PPPK has a significant impact on the quality of life of the children included in our study, as indicated by the use of SPI and cDLQi scores . The severity seems mainly associated with pain caused by cracking and aesthetic discomfort, causing difficulty in writing, walking and practicing sports activities. Acitretin is indicated as first-line treatment for moderate to severe psoriasis in children.

Our study allowed us to evaluate the children’s quality of life, the progression of the disease and the effectiveness of the treatments. Overall, it is consistent with the data in the literature. However, a discrepancy is observed regarding the dosage of acitretin , which is much lower than recommended, but nevertheless gives satisfactory results. However, the small number represents a limitation in our study. Larger studies are needed to confirm these data.

Conclusion: our study highlights the significant impact of PPPK on children’s quality of life. However, the use of low doses of acitretin showed good improvement in their condition. Larger studies are needed to confirm these observations.

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Abstract N°: 5325

Revealing hidden type 1 diabetes : A case study on lipoid necrobiosis as a diagnostic indicator

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Introduction & Objectives:

Lipoid necrobiosis (LN) is a rare granulomatous dermatosis. It is reported in 0.3 to 1.2% of diabetic patients, preferentially located in the leg. The lesions appear as erythematous plaques, with central depressions.

We report the case of an 11-year-old child who presented with asymptomatic well-circumscribed erythematous plaques of the leg, 5 years before the discovery of her diabetes, the diagnostic of lipoid necrobiosis was confirmed.

Through this case, we will identify clinical, histological and therapeutic features of lipoidal necrobiosis.

Clinical case:

An 11-years-old child, not known to be diabetic, with a particular pathological history of erythemato-atrophic plaques on the right leg which had been treated long-term by self-medication for 6 years, with no improvement. The patient was admitted to the emergency room for a non-traumatic afebrile consciousness disorder subsequently revealing ketoacidosis. Secondary, a type 1 diabetes discovered following assessments carried out in the intensive care unit. Dermatological examination revealed shiny erythemato-telangiectatic plaques, 7cm and 3cm long axis respectively. A skin biopsy was performed confirming the diagnosis of lipoid necrobiosis. With good improvement under treatment.

Discussion:

Lipoid necrobiosis is a chronic non-infectious idiopathic granulomatous disease of the dermis. Due to its increased prevalence in diabetic patients, especially type 1, etiological theories mainly refer to microangiopathy. However, the association with poor glycemic control remains controversial.

The prevalence of diabetes in patients with LN remains controversial, ranging from 11 to 65% depending on the series. The LN in the diabetic patients reached electively the adult during the 3rd and 4th decades, with a female predominance according to a publication of 2017. LN lesions are generally solitary and localized to the lower limbs, often bilateral and on tibial margin and instep. Variable sizes can exist, from the size of a small coin to extensive lesions covering almost the entire the leg, from the knee to ankle. These lesions are exceptionally described on the face, scalp and trunk.

These lesions are often difficult to treat, leaving frequently significant scars. The course is usually chronic, even if spontaneous regression is observed in 20% of cases. And may be complicated by ulceration in 35% cases.

According to the bibliographic research, we found a single case reported in Lebanon of lipoid necrobiosis revealing type 1 diabetes in 2012, which makes our case quite rare.

Conclusion:

Despite years of research, the origin of Lipoid Necrobiosis remains unknown. Once installed, it remains difficult to take care of. The relationship between LN and diabetes continues to be studied. Fortunately, the incidence of LN

in patients with diabetes has become very low. This should probably be seen as a consequence of the constant improvement in the care of diabetic patients.

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Abstract N°: 5387

Subject Perception of an OTC Healing Ointment to Prevent/Heal Diaper Rash in Infants

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Introduction & Objectives:

Diaper rash is a frequent dermatitis in infants, commonly related to exposure to moisture, skin sensitivity, and chafing. This study elicited subjective feedback from parents who used a healing ointment (HO) to prevent and/or heal diaper rash in their infants.

Materials & Methods:

Blinded home-use test study, subjects aged 12 weeks to 12 months were treated for 2 weeks and an online recall survey was administered at Day 1, 7, and 14. Informed consent was obtained from parents/legal guardians. Parents reported they were primarily responsible for diaper changes and used ointment/cream/paste during diaper changes at least once per day. Subject infants had to be currently experiencing a slight to moderate diaper rash.

Results:

A total of 110 infants participated, who were predominantly Caucasian (63%). Most subjects (49%) had moderate rash, followed by 37% with mild severity and 14% with slight rash. No adverse events were reported. Results of the Day 1 questionnaire indicated an immediate relief occurred with HO, and this improvement continued throughout Day 14. Parents reported moisturized skin (93% at Day 1), smoother skin (90%, Day 1), less irritation (85% Day 1), and long-lasting soothing that was sustained between diaper changes (95% Day 14). In addition, they reported HO to be an elegant product, that applied easily, spread evenly, and was easy to wipe off.

Conclusion:

The healing ointment resulted in noticeable improvement in infants' experiencing diaper rash on their skin and overall comfort, yielding an overall positive experience.

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Abstract N°: 5395

Painless subcutaneous nodules in children; consider subcutaneous granuloma annulare

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Introduction & Objectives: The subcutaneous granuloma annulare (SGA) is a rare variant of the classic granuloma annulare (GA), which occurs exclusively in children. This rare clinical entity is characterized by the sudden appearance of painless, immobile subcutaneous nodules primarily located on the lower extremities and the scalp. Uncommon sites include the palms, soles, periorbital, and genital areas. Diagnosis is based on histopathological examination. Recurrence rates are higher compared to GA, but remission periods last longer; however, spontaneous resolution is expected within the next two years from the onset of the disease. The pathogenesis of the disease is unknown, but it has been associated with trauma, insect bites, diabetes mellitus, as well as infections such as tuberculosis, histoplasmosis, and streptococcal infection.

Materials & Methods: A twelve-year-old girl presented with clearly demarcated erythematous plaques on the right lower limb for six months. The eruption started from the ankle and then, after a four month interval, new lesions appeared on the anterior and posterior surfaces of the shin and thigh. The lesions on the thighs included subcutaneous, erythematous, painless nodules. The lesions on the right lateral ankle exhibited raised borders and clinically resembled lesions of lipoid necrobiosis. There were no accompanying symptoms. The patient's personal medical history was unremarkable.

Results: Laboratory tests were performed with no pathological findings. Screening for hemophilia was negative. Histological examination confirmed the diagnosis of granuloma annulare. The patient did not present symptoms suggestive of rheumatoid arthritis. Treatment with topical corticosteroids was administered without clinical improvement.

Conclusion: Granuloma annulare typically resolves spontaneously within months or years, making clinical observation an ideal approach rather than interventions such as surgical excision. Recurrence rates range from 40% to 80%, but they do not alter the diagnosis unless additional clinical factors suggesting rheumatic diseases are present. Differential diagnosis from other disease manifestations, such as rheumatoid nodules, is significant. Granuloma annulare is not associated with connective tissue diseases. It is important to include it in the differential diagnosis of subcutaneous nodules in the lower extremities and scalp in children with an unremarkable medical history to avoid unnecessary diagnostic tests and invasive procedures in a self-limiting disease.

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Abstract N°: 5396

What association exists between infantile acropustulosis and scabies? A report on two cases?

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Introduction & Objectives:

Acropustulosis infantile is an uncommon and little-known dermatosis that primarily affects newborns. It presents as a vesiculopustular exanthema of the palms and soles and is extremely itchy. Because of the location and nature of the lesions, it is sometimes mistaken for scabies. However, it frequently complicates scabies in children. We present two cases of scabies that are complicated by acropustulosis.

The objective of our study is to clarify the relationship between scabies and acropustulosis in children.

Case report:

Case 1

A one-and-a-half-year-old baby with no previous pathological history presented four months ago with erythematous skin that was diffusely located throughout the body and involved the palmoplantar area, accompanied by pruritic vesiculopustular skin lesions. Questioning revealed the presence of diffuse pruritus with nocturnal recrudescence in the mother. The diagnosis of scabies was accepted, and the patient was treated with benzyl benzoate. The progression was characterized by the preservation of palmoplantar vesiculopustular lesions and the absence of pruritus and skin blanching.

Case 2

A 4-year-old girl, with no previous medical history, presented for 6 months with generalized pruritus with nocturnal recrudescence of a familial nature. Clinical examination revealed vesiculopustular cutaneous lesions resting on erythematous skin localized on the extremities and interdigital areas, with a scabious nodule and scratching lesions.

The diagnosis of scabies was accepted, and the patient was treated with benzyl Benzoate.

The evolution was marked by the persistence of vesiculo-pustular lesions, impetiginized in places and excoriated in others, localized on the dorsal surface and edges of the feet, inter-toral spaces and hands: dorso-palmar surfaces and interdigital spaces.

Conclusion:

Infantile acropustulosis is a syndrome characterized by recurrent pruritic acral vesicopustules.

The etiopathogenesis is unknown, but some cases of infantile acropustulosis have developed during true scabies, suggesting a hypersensitivity reaction to Sarcoptes scabiei. Treatment has been disappointing, and the disease heals spontaneously after a few years.

The aim of our work is to highlight the importance of looking for a history of scabies in any child presenting with acral vesiculo-pustular lesions, before accepting the diagnosis of idiopathic acropustuosa.

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Abstract N°: 5400

Mycosis fungoides in children

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Introduction & Objectives:

Mycosis fungoides is the most common form of cutaneous T-cell lymphoma in adults. In children, the disease is much rarer and often under-diagnosed. In this presentation, we report four cases of mycosis fever in children.

The object of our work is to determine the clinical, histological, therapeutic and evolutionary profiles of MF in children.

Cases

We diagnosed four patients with a myosis fongoid (median age at first symptoms: 5.5 years; at diagnosis: 8.5 years; sex ratio: M/F = 4/1). At the moment of diagnosis, all patients were in stage I. Lesions were hypopigmented macules with fine scaling in all four cases. Pruritus was present in only one case. KPP was present in 2 cases. Localized depilation was present in only one case. Histologically, epidermotropism was present in all cases, with one case of pilotropism. On IHC, the CD3+ phenotype was present in all 4 cases, with 1 case of CD8+. Initially, patients received a local treatment of dermocorticoids combined with puvatherapy. A clinical response was obtained in two patients (complete remission 1 and partial remission 1), while two patients were treated with systemic treatment (methotrexate) with good evolution.

Conclusion:

MF is the most common form of cutaneous lymphoma in children. The hypopigmented form was the main clinical manifestation; pruritus was exceptional. The diagnosis is confirmed by a skin biopsy with an immunohistochemical study. Local therapy usually has a positive effect on pediatric MF. The evolution is mostly indolent, leading to delayed diagnosis, and the prognosis is generally favorable.

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Abstract N°: 5402

Epidermolyses bulleuses hereditaires: épidemiological and clinical aspects in the pediatric population

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Introduction & Objectives:

Epidermolyses bulleuses hereditaires are a rare, heterogeneous genetic disease with autosomal dominant or recessive inheritance characterized by epithelial fragility due to dermal-epidermal cleavage, manifesting as localized or generalized bullae and mucocutaneous erosions.

We present a descriptive study of EBH cases from the pediatric dermatology consultation service.

The aim of our study is to identify the epidemioclinical profile of EBH in children in our context.

Materials & Methods:

A retrospective descriptive study spread over a period of 11 years, from January 2013 to December 2023, collected cases of EBH managed in the pediatric dermatology consultation service

Results:

A total of 52 cases have been collected, of which 25 cases (48%) were male, with a sex ratio of 0.92. The mean age was 5.3 years, ranging from 0 to 23 years. Consanguinity was present in 37 cases (71.15%), with a similar sibling in 22 cases (42.3%). EBH cases were divided into three categories: simple EBH in 17 cases (32.7%), dystrophic EBH in 11 cases (21.1%), and junctional EBH in 5 cases (9.6%). Lesions included erosions and bullae in all cases, milium grains in 20 cases (38.5%), atrophy and dystrophic scars in 14 cases (26.9%), alopecia in 4 cases (7.7%), plantar keratoderma in 3 cases (5.7%), mucosal involvement in 22 cases (42.3%), nail involvement in 27 cases (52%), and dental anomalies in 11 cases (21.15%). Complications included anemia in 15 cases (28.8%), staturo-ponderal retardation in 12 cases (23%), infection in 12 cases (23.7%), syndactyly in 10 cases (19.2%), and chronic inflammatory syndrome in 2 cases (3.8%) complicated by amyloidosis. Six patients died (11.5%) and six were lost to follow-up (11.5%).

Conclusion: ### EBH are caused by mutations in genes coding for structural and functional proteins of the dermal-epidermal junction. Depending on the degree of cleavage in the skin, a distinction is made between simple, junctional, and dystrophic forms, as well as Kindler syndrome. The diagnosis is suspected clinically and confirmed by immunohistological examination of the skin. The severity of EBH varies widely, ranging from simple discomfort that allows a near-normal life to lethal forms. About therapy, there is no curative treatment available. The basic tenet of therapeutic care is multidisciplinary collaboration.

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Abstract N°: 5573

Extensive Nevil in early childhood with a difficult therapeutic approach

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Introduction: Epidermal nevi are benign, hamartomatous growths of the skin that are present at birth or develop in early childhood. They may be composed of a variety of epidermal cells and structures, including keratinocytes, sebaceous glands, hair follicles, apocrine and eccrine glands, and smooth muscle cells, and are thought to represent a form of cutaneous mosaicism. Lesions with prominent adnexal components (eg, sebaceous, follicular, and/or apocrine) are sometimes referred to as “organoid,” while lesions with primarily epidermal differentiation are known as “non-organoid” or “keratinocytic” nevi. Inflammatory linear verrucous epidermal nevus (ILVEN) is a rare variant of epidermal nevus. It typically presents in early childhood and is characterized by pruritic, erythematous, and hyperkeratotic papules that often coalesce into plaques. It is usually unilateral and most often located on the lower half of the body, with a linear distribution that follows the lines of Blaschko. In most cases, the diagnosis of linear epidermal nevus is made clinically, based upon the finding of verrucous papules and plaques in a linear distribution along the lines of Blaschko. If the diagnosis is in question, a skin biopsy may be necessary for histopathologic confirmation. **Case Report:** A 3-year-old girl, presenting skin allergies since she was born, which has worsened in the last 5 months. At certain periods, there is an increase in he number of lesions, associated with intense itching. The patient presents obesity and hyperactivity. Regarding family history, her grandmother is diagnosed with psoriasis. On physical examination, she presented erythematous-scaly plaques on the dorsum of the feet, back and buttocks on the left side of the body. A diagnostic hypothesis of NEVIL (left hemibody) and Psoriasis was made. Her mother chose not to undergo a biopsy for confirmation. Soap, moisturizer, vitamin D (if deficient) and antihistamine were prescribed. **Conclusion:** The management of epidermal nevi is difficult. Full-thickness excision provides definitive treatment for small lesions, but may not be an option for large or extensive lesions, due to the risk of disfiguring scarring. Multiple alternative surgical or destructive approaches have been reported, including shave excision, cryotherapy, deep chemical peels, and laser ablation. However, their efficacy is uncertain due to the lack of randomized trials or large observational studies with long-term follow-up. Topical therapies have a limited place in the treatment of epidermal nevi. There are isolated reports of successful treatment with topical retinoids, topical fluorouracil, topical corticosteroids, topical calcipotriol, and topical sirolimus.

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Abstract N°: 5632

Adolescent Follicular Keratosis on the Cheek in Skin of Color: Case series and literature review

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Introduction:

Follicular keratosis (FK) is a rare dermatologic disorder characterized by monomorphic skin-colored hyperkeratotic follicular papules on a hyperpigmented background. The underlying etiology is unclear; however, it is often linked to chronic skin friction. Herein, we report three pediatric patients of color with FK on the cheeks.

Case Presentations:

Patient 1, a 13-year-old African American female, developed FK on the left cheek after long-term mask use. Though initial treatment failure with tacrolimus, subsequent therapy with tretinoin yielded improvement. Patient 2, an 18-year-old Mexican American female, displayed a dark spot evolving from bumps on the right cheek, treated with tretinoin and lactic acid cream. Patient 3, a 16-year-old African American female, presented with hormonal acne and keratotic papules confined within hyperpigmented patches on the bilateral cheeks, showing significant improvement with tretinoin up-titration.

Conclusion:

Our report highlights treatment response to topical retinoids in a difficult-to-treat condition, as well as the predilection of this condition for the cheeks and children of color. Furthermore, this case series and literature review brings clarity to two synonymous conditions of traumatic anserine folliculosis (TAF) and FK, constituting the same medical diagnosis.

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Abstract N°: 5646

How to discuss skin, common skin problems and skin conditions with primary school students?

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Introduction & Objectives:

Skin issues such as warts, eczema, or vitiligo are a common experience for children in schools everywhere. These concerns are frequently overlooked, yet studies indicate a decrease in overall well-being. The repercussions of bullying on youngsters with skin conditions are noteworthy. Both children dealing with persistent skin conditions and their parents seek guidance on managing these issues within the school setting. Nonetheless, maintaining good skincare practices is crucial for all, encompassing sun protection to prevent skin cancer. Furthermore, misinformation regarding skincare spreads on social media platforms due to a deficit in fundamental understanding.

Our objective was developing educational materials for children aged 4-12 fostering knowledge, understanding, skills, and attitudes about their own and others' skin. The goal is to empower children to prevent health risks and enhance their well-being and resilience.

Materials & Methods:

Healthcare professionals, educational experts, and patient representatives collaborated to develop educational materials on skin, common skin problems and chronic skin conditions tailored for children aged 4-12. The curriculum was carefully developed to be substantively correct, fitting into the educational context and being appropriate for patient representatives. A children's council provided advice.

Results:

The result is a curriculum comprising five lessons tailored to specific age groups, covering skin basics, skincare, skin colour, temporary and long-lasting skin problems, understanding and acceptance of skin problems and sun protection. There are three additional lessons about rare skin diseases and one about acne for children aged 10-12 years. The strength of the curriculum is that it focuses on the skin of all children, while increasing understanding of skin problems and skin conditions. In 2022-2023, 600 teaching kits were distributed to 170 different cities in the Netherlands. Teachers evaluated the materials positively, finding them didactically sound and flexible to use.

Conclusion:

The curriculum Skin at School has been successfully developed, implemented and positively assessed by teachers. Further research on programme effects and the possibilities for expansion to other countries is needed.

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Abstract N°: 5649

Poikilodermatous mycosis fungoides in a child

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Introduction & Objectives:

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma. MF is rare in pediatric population and most of pediatric cases presented with hypopigmented MF. Poikilodermatous mycosis fungoides is a variant of MF, formerly referred to as poikiloderma vasculare atrophicans. The lesions are classically characterized by large patches/plaques of hypopigmentation and hyperpigmentation with atrophy and telangiectases.

We report a case of a child presented with** poikilodermatous mycosis fungoides.

Materials & Methods:

Clinical features, laboratoty results, histopathological and immunohistochemical findings were made to reach the diagnosis.

Results:

Ten - year- old male child presented with 2 year history of patches of poikiloderma (skin atrophy, mottled pigmentation and telangiectasia) on trunk and extremities with mild itching**. Hisopathological findings** showed atrophic epidermis with epidermotropism of atypical lymphocytes, basal hydropic degeneration, pigment incontinence and telangiectatic vessels. **Immunhistochemical findings** showed CD3+ and CD8+**.** Complete blood count, liver and renal tests were normal. Peripheral blood smear for Sezary cells was negative. Lymph node examination, computed tomography scans of the chest and ultrasonography of the abdomen and pelvis were normal.TNM staging was stage IB. **Treatment** was initiated using NBUVB phototherapy.

Conclusion:

poikilodermatous mycosis fungoides is uncommon vatiant of mycosis fungoides that is characterized, clinically by localized or diffuse patches of telangiectasias, mottled pigmentation and atrophy. This variant is rarely reported in pediatric population. It has a good prognosis with good response to phototherapy.

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Abstract N°: 5666

Challenging the limits: an unusual case of pemphigus foliaceus in the pediatric population

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Introduction & Objectives: Pemphigus foliaceus (PF) is a chronic autoimmune blistering skin disease. This condition is part of the pemphigus group, vesiculobullous diseases that can occur at any age, being more common in patients between 50 and 60 years old, although the average age at diagnosis varies significantly by country of origin and ethnicity, considering it a rare disease in the pediatric age group, with incidence rates ranging from 0.1 to 0.5 per-100,000 people per year. Due to its spectrum of clinical manifestations, it can be considered a true challenge in dermatology.

Materials & Methods: We present the clinical case of a pediatric patient from southwestern Colombia with a diagnosis of pemphigus foliaceus with the aim of illustrating the clinical and histopathological characteristics of this unusual disease in an age group where its presentation is rare.

Results: A 15-year-old male from a rural area of Cauca, Colombia, with no relevant medical history. Evaluated at a high-complexity pediatric clinic for a two-month history consisting of scaly plaques on the scalp associated with pruritic vesicular lesions, managed in primary care as tinea capitis with no clinical improvement. On physical examination, a patient with phototype VI was found, with a yellowish crust on an erythematous base on the scalp, as well as erythematous papulovesicular lesions, some desquamated and crusted with blood in the face, neck, pre-sternal, and interscapular regions, as well as some isolated lesions on all four limbs. No mucosal involvement was evident. Blood tests were normal, KOH and fungal cultures were negative, tests for HIV, HTLV I and II, and hepatitis B and C were non-reactive. Skin biopsy showed subcorneal acantholysis with partially detached blisters, granular keratinocytes, neutrophilic exocytosis, and few intraepidermal eosinophils, and dermal level showed a perivascular lymphoplasmacytic inflammatory infiltrate. Direct immunofluorescence of perilesional skin showed a net pattern around keratinocytes with positivity for IgG and C3, all findings compatible with pemphigus foliaceus. Treatment with prednisone 1 mg/kg/day was initiated and high-potency topical steroids were prescribed for skin and scalp lesions, with significant improvement within fifteen days.

Conclusion: We report a case of an uncommon and potentially serious blistering disease that generally affects adults and is very rare in the pediatric population, often being confused with other types of inflammatory or vesiculobullous diseases more common in this population. Therefore, clinical suspicion and appropriate diagnostic aids lead to early management and thus reduce the morbidity and mortality associated with this condition.

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Abstract N°: 5717

Blueberry Muffin Baby Syndrome: Recognizing the Alarm in Dotted Newborns

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Introduction & Objectives:

Blueberry Muffin Baby syndrome, a rare cutaneous condition observed in neonates, is characterized by disseminated reddish-purple or blue-gray inflammatory papulonodules resembling blueberry muffins. These lesions are indicative of dermal extramedullary hematopoiesis reactions and can arise from various etiologies, including congenital infections, neonatal hemolytic diseases, and tumor pathologies. Our objective is to elucidate the clinical features, diagnostic challenges, and potential underlying causes of this syndrome.

Materials and Methods/Observations:

We present the case of a newborn male born to non-consanguineous parents following a poorly monitored full-term pregnancy. The delivery was vaginal, accompanied by meconium-stained amniotic fluid and premature rupture of membranes lasting over 12 hours. Initial Apgar scores were low, and the infant exhibited respiratory distress, clinical examination revealed multiple variable-sized blue-gray maculopapular and nodular lesions diffusely distributed over the face, trunk, back, and limbs, consistent with Blueberry Muffin Baby syndrome. Additional findings included cyanosis of the extremities and respiratory distress with a Silverman score of 5/10, absent lung crepitations on auscultation, and a negative hyperoxia test. Laboratory investigations showed thrombocytopenia, leukopenia with neutropenia, anemia, and abnormal prothrombin time. A provisional diagnosis of respiratory distress due to early neonatal pulmonary infection was made, leading to the initiation of empirical bi-antibiotic therapy (amoxicillin + gentamicin). Unfortunately, the newborn succumbed to the condition 8 hours post-birth before further etiological investigations, including bone marrow examination, could be completed.

Results/Discussion:

Blueberry Muffin Baby syndrome manifests as disseminated papulonodules in newborns, exhibiting various shades from vivid red and purple to blue-gray. These lesions typically measure approximately 2 to 8 mm in diameter and are often generalized, predominantly affecting the head, neck, and trunk. Although they usually resolve within 3 to 6 weeks postpartum, these lesions may persist or evolve, acquiring a brownish hue. The syndrome represents a postnatal manifestation of dermal hematopoiesis, which may persist postnatally or indicate neoplastic infiltration. During the neonatal period, dermal hematopoiesis can be associated with various congenital infections, hemolytic diseases, or malignant conditions such as neuroblastoma. Despite the typical skin lesions observed in our patient, the exact etiology could not be determined due to the rapid deterioration and demise of the newborn.

Conclusion:

Blueberry Muffin syndrome serves as a cutaneous marker of prolonged fetal hematopoiesis and may signify serious underlying conditions in neonates. While congenital infections, hemolytic diseases, and tumors are common associations, prompt recognition and thorough diagnostic evaluation are imperative for appropriate management and prognosis. Differential diagnoses encompass various congenital vascular lesions, necessitating a comprehensive approach to neonatal dermatologic conditions.

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Abstract N°: 5790

Hospitalization Disease Burden of Varicella-Zoster Virus Infection in Post-Transplantation Pediatrics in China: The Imperative for Active Immunization in Immunosuppressed Pediatric Populations

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Introduction & Objectives:

Infections caused by the varicella-zoster virus (VZV) are common in the clinical practice of dermatology, the reactivation of the VZV is primarily influenced by the host's immune status, despite generally favorable outcomes in most patient groups, it often leads to severe and poor outcomes in immunocompromised individuals, especially among transplant recipients. Despite extensive assessments conducted in adult transplant patients, the scarce data highlighting the profound impacts of VZV in pediatric transplant recipients resulted in limited studies on the safety and immunogenicity of VZV vaccines in this population. This study aims to evaluate the necessity for VZV prevention in immunocompromised children.

Materials & Methods:

The face sheets of discharge medical records (FSMRs) related to VZV infections were analyzed within the timeframe from December 2016 to December 2022. This analysis was conducted utilizing data from the FUTang Update medical REcords (FUTURE) database, comprising information from 27 tertiary children's hospitals in China. Comparative assessments of clinical characteristics, length of stay (LOS), and economic burden were conducted among various groups in the present study.

Results:

For the retrospective analysis, 9,171,717 inpatient case records were extracted from the database, including 353 herpes zoster (HZ) and 6196 chickenpox diagnoses. Within this dataset, 60 children (aged 0-17) with a history of transplantation were hospitalized for either HZ or chickenpox, constituting 11.33% (40 out of 353) for HZ and 0.32% (20 out of 6196) for chickenpox cases. School-age children (7-12 years) and teenagers (13-17 years) represented over 81.67% (49 out of 60) of all VZV cases. No neonatal patients (0-28 days) were reported in the dataset. Of the 60 VZV cases, 70% were male, and the ratio of coinfection was higher in chickenpox patients (9 out of 20; 45%) compared to HZ patients (8 out of 40; 20%) (Table 1). The average duration of hospital stay was notably longer for both HZ (11 days) and chickenpox patients with a history of transplantation in comparison to non-transplant patients with the same infections (HZ: 11 days vs 7 days; chickenpox: 9 days vs 5 days; transplant vs non-transplant) (Fig. 1A). Additionally, the average cost of hospitalization was higher for pediatric patients with prior transplantation diagnosed with HZ and chickenpox, as opposed to age- and infection-matched patients without transplantation history (HZ: 16,637.3 RMB vs 5,386.4 RMB; chickenpox: 13,335.0 RMB vs 3,898.9 RMB; transplant vs non-transplant) (Fig. 1B). Also, the duration and cost of hospitalization did not exhibit significant differences between male and female transplant recipients, nor among different age groups (Fig. 1C-F).

Conclusion:

Our study reveals higher costs and longer hospital stays for transplant recipients with VZV infection were

significantly higher than the non-transplant patients.

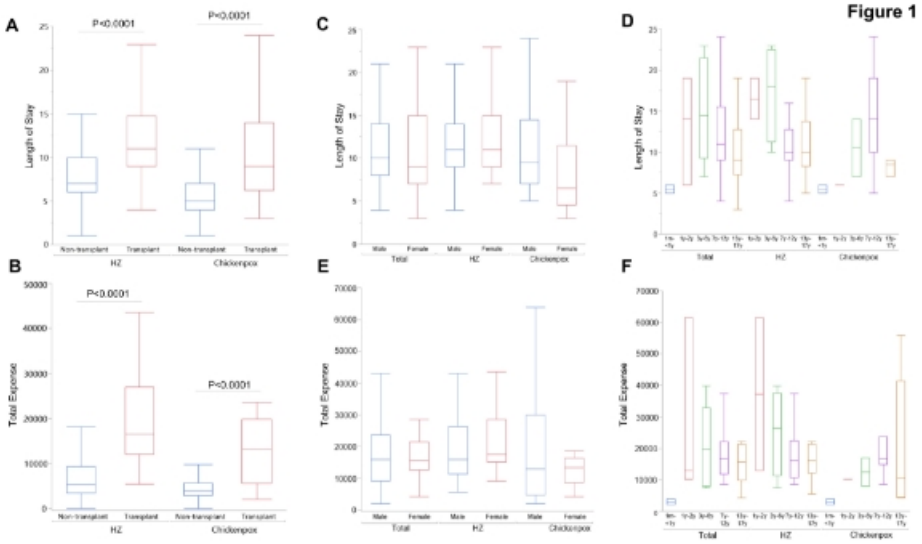

Figure1. Assessments of clinical characteristics, length of stay, and economic burden were conducted among various groups.

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Abstract N°: 5840

Childhood Prurigo: Epidemiological, Clinical, and Etiological Profile of 168 Patients at the University Hospital Center

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Introduction & Objectives:

Prurigo is a common dermatosis in children, impacting the quality of life of both children and their parents. The objective of our study is to define the epidemiological, clinical, and etiological profile of childhood prurigo.

Materials & Methods:

This is a retrospective study conducted over a six year period within the pediatric dermatology consultation at the University Hospital, including children aged 0 to 14 years with prurigo. Data were collected in an exploitation form, including epidemiological, clinical, and etiological data.

Results:

Results Among the 168 children included (89 girls and 79 boys), the mean age of symptom onset was 2.43 years. Prurigo accounted for 21% of pediatric dermatology consultations. Personal or family atopy was present in 37.50% of cases. Clinical manifestations were mainly papulovesicular lesions (74.40%), bullae (14.88%), and urticarial lesions (10.71%), with pruritus present in all patients. Affected areas were predominantly exposed areas and the belt area in 62.5% of cases. Three subgroups were identified: Perennial prurigo accounted for 25.60% of cases. Seasonal prurigo was found in 43.45% of cases, and non-seasonal seasonal prurigo was found in 30.95% of patients. Aggravation by certain histamine-liberating foods was noted in 35% of cases.

Conclusion:

Few studies have been conducted on childhood prurigo, highlighting the uniqueness of our work. The peculiarity of childhood prurigo is its primary nature, unlike in adults, often of allergic or non-allergic origin. The early age of lesion onset is linked to initial environmental sensitizations and the immaturity of the immune system. There are three forms of childhood prurigo, with seasonal prurigo being the most common form. Lesions predominantly affect exposed areas and are induced by mosquito bites. The study highlights a possible association with histamine-liberating foods, emphasizing the importance of allergological exploration. Further extensive studies are needed to better characterize this childhood dermatosis.

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Abstract N°: 5915

Long-term efficacy and safety of secukinumab over 4 years in children and adolescents with moderate to severe chronic plaque psoriasis: Results of a randomised phase III, open-label trial

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Introduction & Objectives:

Psoriasis is a chronic relapsing inflammatory skin disease that affects approximately 1% of children, with onset most common during adolescence.1 Secukinumab (SEC) is a fully human monoclonal antibody that selectively neutralises interleukin-17A, a key cytokine involved in the pathogenesis of plaque psoriasis.2 Efficacy and safety of SEC up to week 52 have been reported previously for the present pivotal phase III study in children and adolescents with moderate to severe chronic plaque psoriasis (NCT03668613).3 Herein, we report the long-term efficacy, safety and tolerability of SEC over a period of up to 208 weeks.

Materials & Methods:

Patients aged 6 to <18 years were randomised (1:1) to receive SEC of low dose (LD, 75/75/150 mg; N=42) or high dose (HD, 75/150/300 mg; N=42) based on two stratification factors: weight (<25 kg, 25 to <50 kg or ≥50 kg) and disease severity (moderate or severe). The study consisted of three periods: screening (up to 4 weeks), treatment (of 208 weeks) and post-treatment follow-up (of 16 weeks). Long-term efficacy (Psoriasis Area and Severity Index [PASI] 75/90/100 response, Investigator's Global Assessment modified 2011 [IGA mod 2011] 0/1 response, PASI score), Children's Dermatology Life Quality (CDLQI) 0/1 response, impact of treatment on physical development and safety over 208 weeks were assessed.

Results:

Overall, 79.8% (67/84) patients completed treatment. PASI 75/90/100 as well as IGA 0/1 responses remained high in both treatment groups from Week 12 through the end of treatment (EOT) (Week 208). At Week 208, SEC-treated patients showed sustained PASI 75/90/100 (SEC LD: 96.3%/88.9%/51.9%, SEC HD: 87.9%/81.8%/72.7%) and IGA mod 2011 0/1 (SEC LD: 85.2%, SEC HD: 84.8%) responses. Mean PASI score remained low in both treatment groups, from Week 12 through Week 208; at Week 208, mean percentage change in PASI scores from baseline was −95.7% (mean absolute score 0.76) with SEC LD and −94.5% (mean absolute score 1.07) with SEC HD. CDLQI 0/1 response remained high in both treatment groups from Week 12 through Week 208 (SEC LD: 75.0%, SEC HD: 88.2%). Long-term secukinumab exposure (~313.9 patient-years) in this study showed no new safety signals. The incidence of treatment-emergent adverse events was similar in both SEC dose groups (LD [78.6%] and HD [83.3%]). The most frequently affected system organ class included infections and infestations,

gastrointestinal disorders and skin and subcutaneous tissue disorders. The most reported adverse events by preferred term were COVID-19, nasopharyngitis and acne (**Table 1**). SEC did not have any adverse effect on the growth and development of patients or on their sexual maturation, as evaluated by Tanner score. Anti-drug antibody (ADA) development with SEC treatment for 208 weeks was low; seven patients (7/84, 8.3%) presented with positive treatment-emergent ADAs, with six having positive ADAs only during the treatment-free follow-up period.

Conclusion:

Both doses of SEC demonstrated sustained efficacy and improved health-related quality of life through the entire treatment period (up to EOT on week 208) in paediatric patients with moderate to severe plaque psoriasis. No new safety signals were identified. SEC was well tolerated at both doses (low and high). Growth and development data did not show any notable adverse effects with SEC treatment in the paediatric population.

References

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Table 1. Safety profile over the entire treatment period (safety set)

	SEC LD (N=42)	SEC HD (N=42)	Any SEC (N=84)
Total, TEAEs, n (%)	33 (78.6)	35 (83.3)	68 (81.0)
Death	0 (0.0)	0 (0.0)	0 (0.0)
Non-fatal SAEs, n (%)	4 (9.5)	2 (4.8)	6 (7.1)
Discontinuation due to any AE, n (%)	1 (2.4)	2 (4.8)	3 (3.6)
Most frequent AEs (by PT), n (%)			
COVID-19	12 (28.6)	9 (21.4)	21 (25.0)
Nasopharyngitis	13 (31.0)	7 (16.7)	20 (23.8)
Acne	7 (16.7)	2 (4.8)	9 (10.7)
Safety topics of interest, n (%)			
Infections and infestations (SOC)	27 (64.3)	29 (69.0)	56 (66.7)
Hypersensitivity (SMQ) (narrow)	7 (16.7)	1 (2.4)	8 (9.5)
Neutropenia (NMQ) (narrow)	4 (9.5)	2 (4.8)	6 (7.1)
Vaccination-related complications (HLT)	1 (2.4)	1 (2.4)	2 (2.4)
Inflammatory bowel disease (NMQ) (narrow)[a]	1 (2.4)	1 (2.4)	2 (2.4)
Suicidal ideation (PT)[b]	1 (2.4)	0 (0.0)	1 (1.2)

MedDRA version 26.0 was used for reporting.
AE, adverse event; HLT, high-level term; HD, high dose; LD, low dose; N, total number of patients; n, number of patients; NMQ, Novartis customized MedDRA Query; PT, preferred term; SAE, serious AE; SEC, secukinumab; SOC, system organ class; SMQ, standardized MedDRA Query; TEAE, treatment-emergent adverse event.

[a]One patient (HD group) had a mild AE of haemorrhagic diarrhoea (not diagnosed as IBD) and one patient (LD group) had a moderate SAE of Crohn's disease. The events were considered to be related to the study drug. Both events were resolved with concomitant treatment (Crohn's disease SAE was resolved with sequalae) and both led to permanent discontinuation of the study drug.

[b]The patient had a history of somatic symptom disorder, anxiety and depressed mood. Patient attempted deliberate self-harm, which was reported as a severe SAE of intentional self-injury. The event resolved on the same day of its onset without any concomitant treatment. The event was considered as not related to the study drug.

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Abstract N°: 5976

Title: Generalized Cutaneous Form of Parvovirus B19 Infection: A Case Report

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[1]uhc ibn rochd, dermatology, [2]uhc ibn rochd

Introduction & Objectives:

Parvovirus B19 infection is a common viral disease, primarily affecting children aged 4 to 10 years. we report a clinical case illustrating a generalized form of purpura associated with parvovirus B19 infection.

Observation:

A 6-year-old girl with no significant medical history was hospitalized for extensive febrile purpura initially affecting the perioral region and lower limbs, rapidly progressing to the trunk and upper limbs. The skin eruption presented as confluent petechial ecchymotic lesions, associated with cheilitis without necrotic, bullous, or ulcerated lesions. On examination, the patient had a fever of 39°C, with sub-centimeter right cervical and bilateral inguinal lymphadenopathies, erythematous and edematous throat. Laboratory tests revealed a slight elevation of CRP at 8.08 mg/L, without leukocytosis on CBC. Real-time PCR for parvovirus B19 DNA was positive, confirming a primary infection by the virus. Despite the absence of specific serology, other etiological investigations were negative. Symptomatic treatment with paracetamol, vitamin therapy, and empirical antibiotic therapy was administered, with a favorable outcome after 15 days, not requiring any specific treatment.

Conclusion:

The uniqueness of our work lies in the extent and atypical nature of the cutaneous involvement in Parvovirus B19 infection observed in our patient. However, it can lead to formidable complications such as cardiac (cardiomyopathy), hematologic (severe aplastic anemia), hepatic, and others. It highlights the importance of systematically testing for anti-B19 virus antibodies in patients with uncertain etiology cardiomyopathy or aplastic anemia.

This clinical case significantly illustrates the diversity of cutaneous manifestations associated with Parvovirus B19 infection and the importance of early diagnosis and management.

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Abstract N°: 6026

Trichotillomania in children and adolescent : Clinical and trichoscopic study

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Introduction & Objectives:

Trichotillomania (TTM) is a psychodermatological disorder characterized by a recurrent urge to pull out hair. It belongs to the group of obsessive-compulsive and related disorders according to the criteria of the Diagnostic and Statistical Manual of Mental Disorders 5 (DSM-5).

Diagnosis of trichotillomania in the paediatric population is often difficult and under-diagnosed. Trichoscopy is a non-invasive tool that can be used to differentiate it from other etiologies of alopecia in children.

Materials & Methods:

The study included eight patients attending department of dermatology between January 2019 and December 2022 diagnosed with TTM on the basis of the clinical history, physical examination and trichoscopic findings.

Results:

A total of eight patients were included. Two (25%) were males, and 6 (75%) were females with sex ratio 0.3. Average age at diagnosis was 13 years [9-17]. The mean disease duration was 5 months [1-12]. The associated comorbidities were depression (2 cases), mental retardation (1 case) and epilepsy (1 case).

Clinically, trichotillomania manifested as patchy alopecia in 6 cases and diffuse alopecia in two cases. The pull test was negative in all patients. The scalp was involved in all cases. The eyebrows were affected in one case. The frontoparietal region was the most affected area (5 cases), occipital region was affected in one case. The trichoscopic features were broken hairs of different lengthes (6 cases), black dots (5 cases), trichoptilosis (4 cases), V sign (3 cases), tulip hairs (3 cases), flame hairs (3 cases), coiled hairs (2 cases), yellow dots (3 cases), exclamation mark hairs (2 cases), haemorrhages (1 case). All patients were reffered to a child psychologist for additional management.

Conclusion:

Epidemiologic data in paediatric population are relatively scarce. Prevalence is estimated to be around 1–3%. Age at onset varies from 9 to 13 years and is more common in females. In our study, the most patients were adolescent females with a mean age at diagnosis of 13 years.

Clinically, trichotillomania presents with patchy alopecia or diffuse scalp involvement. The frontoparietal region is more often affected. Eyebrows, upper eyelashes, and pubic area are also commonly involved. These clinical data are similar to the clinical results in our serie.

Diagnosis of trichotillomania can be challenging, especially when patients deny or are unaware of hair pulling. The main differential diagnoses for scalp trichotillomania in paediatric patients are alopecia areata and tinea capitis.

The most common trichoscopic findings in trichotillomania are broken hairs and black dots. However, these features are not specific for trichotillomania. These data are similar to those found in our study.

The others trichoscopic signs found in our serie were trichoptilosis, V sign, tulip hairs, flame hairs, coiled hairs, yellow dots, exclamation mark hairs and haemorrhages. These data are in agreement with the data found in the literature. The most specific trichoscopic features in trichotillomania are trichoptilosis, V-sign, hook hairs, flame hairs, coiled hairs, tulip hairs, and hair powder.

Trichoscopy is a non invasive tool that can be used to diagnose trichotillomania, differentiate it from other causes of alopecia and avoid unnecessary biopsies which are often traumatic in the pediatric population.

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Abstract N°: 6044

Infantile post-vaccination bullous pemphigoid

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Introduction & Objectives:

Bullous pemphigoid (BP) is a rare autoimmune blistering disease typically associated with older individuals but occasionally presenting in pediatric cases, posing unique diagnostic and therapeutic challenges. We present a case of infantile BP occurring shortly after pentavalent vaccination in a 3-month-old male infant.

Results:

The patient displayed bullous eruptions initially localized to the hands and feet, which rapidly extended to involve the trunk and face, accompanied by striking palmoplantar involvement. Dermatologic examination revealed tense bullae on erythematous skin, urticarial plaques, and small vesicles. Histological examination confirmed subepidermal bullae with eosinophilic dermal inflammation and linear C3 deposition, supporting the diagnosis of BP. Treatment with topical corticosteroids resulted in rapid resolution of lesions over a 2-month period, highlighting the efficacy of this therapeutic approach in infantile BP.

Despite its rarity in pediatric populations, infantile BP demonstrates a favorable prognosis compared to adult-onset BP, often exhibiting a robust response to corticosteroid treatment and complete resolution of symptoms within a relatively short timeframe. However, the precise etiological factors underlying infantile BP remain poorly understood, necessitating further research efforts to elucidate its pathogenesis. Additionally, the distinct clinical features of infantile BP, including palmoplantar involvement, warrant consideration in diagnostic and therapeutic decision-making processes.

Conclusion:

This case underscores the importance of recognizing and promptly managing infantile BP, particularly in the context of recent vaccination, to optimize patient outcomes and minimize potential complications. Further studies are warranted to refine diagnostic criteria, elucidate the epidemiology of infantile BP, and optimize treatment strategies, ultimately improving our understanding and management of this rare autoimmune disorder in pediatric populations.

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Abstract N°: 6138

Congenital cutaneous aplasia : a report of 16 cases

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Introduction & Objectives :

Congenital cutaneous aplasia (CCA) presents as a rare dermatological anomaly characterized by localized absence of epidermis, dermis, and occasionally subcutaneous tissue, affecting diverse anatomical regions including the scalp, face, trunk, and extremities. Its clinical manifestations range from scleroatrophy to superficial or deep ulceration, with lesion sizes varying from a few millimeters to exceeding 10 cm in diameter. This study aims to assess the clinical features of cutaneous aplasias and associated presentations

Materials & Methods:

A retrospective descriptive study spanning 14 years was conducted, encompassing all cases of congenital cutaneous aplasia observed within our medical department, totaling 16 cases. Epidemiological and clinical data were systematically collected and the progression of these cases was monitored.

Results:

The majority of diagnoses were made during the neonatal period, with patients presenting for consultation within the first 18 months of life. Among the 16 identified cases, comprising 6 females and 10 males, a sex ratio of 0.6 was established. 33% of patients had consanguineous parentage, with two cases also exhibiting familial history of cutaneous aplasia. Notably, maternal infectious history and antenatal anxiety treated with benzodiazepines were observed in some instances, with a significant proportion of pregnancies being inadequately monitored.

Dermatological examination revealed a spectrum of manifestations, including translucent, scarred, ulcerated, and fibrinous plaques of variable shapes, exposing subcutaneous vasculature. Bullous lesions were observed in 26% of cases, often co-occurring with areas of aplasia and prone to friction-induced rupture. Oral mucosal involvement was evident, characterized by mucosal blistering and erosions. Predominant lesion sites were the upper and lower limbs, accounting for half of the cases, followed by involvement of the flanks and abdomen in 12% of patients. Scalp involvement was less frequent in our series, observed in only two patients: in a newborn with a large plaque several cm in size, with irregular borders, exposing the dura mater, and in a second patient of 18 months, with aplasia of less than 1 cm, associated with a cranial deformity resembling a “boat prow,” suggesting probable trigonocephaly.

5 patients (31.5%) were diagnosed with Bart syndrome, a form of dominant dystrophic epidermolysis bullosa, while malformations were observed in 31% of patients, including bilateral talipes equinovarus, radial club hands, cleft lip, craniosynostosis, and associated bone defects.

The treatment approach for CCA focused on symptomatic management, patient and family education, and psychological support. Conservative methods were preferred for less severe lesions with directed wound healing. Anticipatory management of complications, such as pain relief, antibiotic therapy, and addressing concurrent malformations, was advised. However, difficulties in follow-up were acknowledged, especially in cases requiring neurosurgical intervention.

Conclusion:

Congenital cutaneous aplasia poses as a rare yet clinically significant anomaly, warranting thorough evaluation in neonates to ascertain potential syndromic associations. While diagnosis is typically straightforward, the multifaceted nature of its management demands comprehensive interdisciplinary care.

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Abstract N°: 6205

Clinical profile and outcomes in infantile erythroderma

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Introduction & Objectives:

Infantile erythroderma can be due to a variety of primary dermatological as well as systemic disorders; and often presents a diagnostic challenge. There are few studies on this relatively rare condition, especially those describing long-term outcomes.

Our objective was to characterise the clinical manifestations, laboratory features, and outcomes in infantile erythroderma.

Materials & Methods:

The study included all infants with erythroderma presenting to the Department of Dermatology from August 2021 to July 2022. Following detailed evaluation, all participants were followed up for at least 3 months for evolving features.

Results:

Fourteen infants with erythroderma were identified during the study period. Two infants with the ichthyosiform erythroderma phenotype had underlying TGM1 and NIPAL4 mutations. Twelve infants had eczematous phenotypes belonging to two distinct morphological types viz. atopiform dermatitis (6 cases) or seborrheic dermatitis-like presentation (5 cases).

Five of the patients with atopiform lesions went on to satisfy the clinical criteria for atopic dermatitis (AD). All except one of these infants had overlapping features of seborrheic dermatitis in the form of flexural lesions (4 cases). In two patients, these flexural lesions resolved within 6 months of age. In the other two patients, persistent flexural lesions evolved into sebopsoriasis. Netherton syndrome was the cause of severe atopiform dermatitis in one infant.

Of the five infants with a seborrheic dermatitis-like presentation, while three had no underlying systemic abnormalities, in two infants the phenotype was a manifestation of systemic disease (zinc deficiency and hyper IgE syndrome).

These findings are summarized in **Figure 1.**

Conclusion:

Infantile erythroderma has a variety of causes including primary dermatoses and systemic disorders. A high index of suspicion and close follow-up for evolving features is needed for an accurate diagnosis.

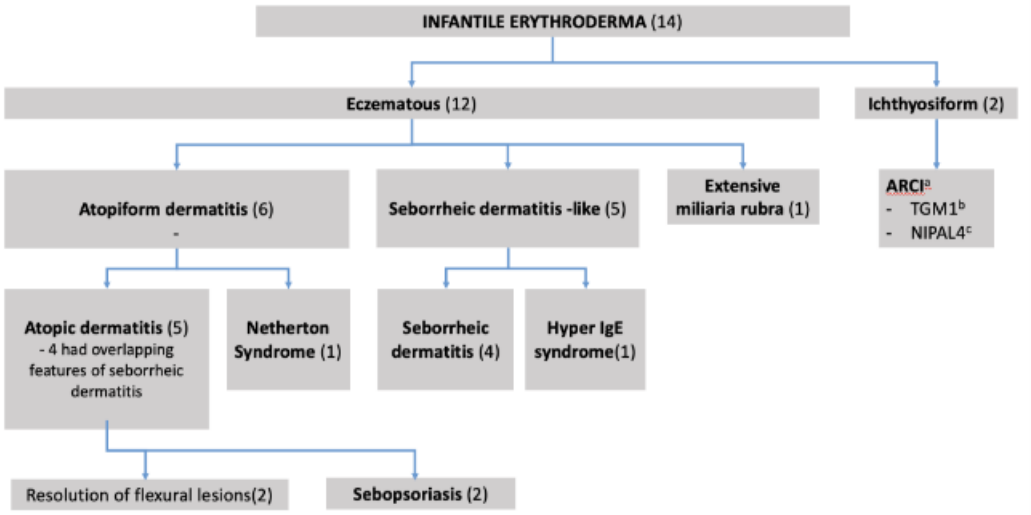
Atopic dermatitis was the most common cause of infantile erythroderma in our study, and most of these patients had overlapping features of seborrheic dermatitis with flexural lesions. A unique feature noted was that, in 50% of these patients, these flexural lesions persisted and evolved into sebopsoriasis coexisting with atopic dermatitis.

Figure 1. Morphological patterns and etiological diagnosis/ outcomes in infantile erythroderma

a Autosomal recessive congenital ichthyosis

b Transglutaminase 1

c Nipa-Like Domain- Containing 4



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Abstract N°: 6258

Epidermal Nevus Syndrome

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Introduction & Objectives:

Epidermal nevus syndrome (ENS) is a rare congenital syndrome characterized by the presence of epidermal nevi in association with developmental abnormalities of the skin, eye, nervous, skeletal, cardiovascular, and urogenital systems. It is often overlooked since some developmental abnormalities are noticed long after the neonatal period and often have lifelong implications.

Materials & Methods:

We report a case of a 20-year-old female with ENS who presented with hyperpigmented papules and plaques over the head and neck since birth and fleshy growths on the scalp and earlobe for three months. Born with no apparent abnormality, she was well till eight years old when she lost the ability to walk on her own. She presented as a paraplegic in a wheelchair and small for her age. Clavicles were bowed, short and placed closely together with remarkable upper and lower limb shortening, asymmetry and anterior bowing on the long bones bilaterally and kyphoscoliosis. She had associated patches of alopecia on the parietal region suggestive of aplasia cutis. She also had skewed, elongated lower incisor teeth and an angiomatous midline tongue lesion with a bifid uvula.

Results:

Skin biopsies taken from the hyperpigmented plaques were compatible with linear sebaceous epidermal nevus, while the biopsy of the fleshy lesions from the earlobe and scalp showed features of syringocytadenoma papilliferum. X-ray imaging showed bowing of the long bones, open physes and several incomplete or healing fractures.

Conclusion:

Some symptoms of epidermal nevus syndrome manifest long after early childhood; therefore, a high index of suspicion is required when faced with a child presenting with an epidermal nevus. The treatment of ENS is on a case-by-case basis. A multidisciplinary approach to the management of these patients can never be overemphasized. Having these patients on follow-up with at least yearly checkups is also essential.

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Abstract N°: 6291

Prevalence and pattern of genital, perineal, perianal and gluteal dermatoses in children.

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Introduction & Objectives:

Lesions predominantly involving genitals, perineal, perianal and gluteal area in children can be of various etiologies like infections (venereal and non-venereal), inflammatory (atopic dermatitis, seborrheic dermatitis, psoriasis, diaper dermatitis, gianotti- crosti, drug reactions), disorders of keratinisation, nutritional deficiency and congenital disorders. The prevalence across various age groups varies due to their gender, genetics, nutritional status, hygiene and habits, economic status and environment.

Materials & Methods: ** A prospective study was conducted from April 2022 to April 2024 to evaluate the prevalence and patterns of genital, perineal, perianal and gluteal dermatoses in children attending dermatology, venereology and leprosy OPD at our centre. Cases were confirmed by relevant bed side and laboratory investigations along with required histopathological evaluations as per the disease. Sample size of 430 children ranging by age from day zero to 18 years were included and they were grouped based on age as neonate (0-28 days), infants (28 days-1yr), toddler (1-3yrs), pre-school (3-5yrs), school age (5yrs- onset of puberty), early adolescence (12-14yrs), mid adolescence (14-16yrs), late adolescence (16-18yrs).

Results:

Out of 430 children, 256 (59.5%) were males and 174 (40.5%) females. Majorly the complaints were itching, pain, scaling, oozing, discharge of pus, blood or curdy white material/fluid. Duration of disease ranged from day zero to 11years. Family history of similar disease was present in cases of scabies and tinea infections. History of consanguinity was present in 3 (0.7%) cases keratinisation disorder. During an outbreak of hand foot mouth disease 69 (16%) cases presented for the same. Localisation of disease to genitals in 34 (7.9%) cases, genitals with inguinal folds and gluteal involvement in 148 (34.4%) cases and as a part of generalised disease in 282 (65.6%). Based on disease classification, 360 cases (83.7%) were of infective conditions out of which 6% were of sexually transmitted diseases, whereas 57 cases (123.3%) were of inflammatory conditions, of which 0.5% were of drug reaction. Other smaller groups included congenital disorders (1.9%), disorders of keratinisations (0.7%) and nutritional disorder (0.5%). On classifying according to age, in neonate and infants candidal infections (26.8%) and seborrheic dermatitis (14.3%) were the most common. In toddlers to school aged children, scabies (38%) and hand foot mouth disease (28.5%) were most common infections, whereas diaper dermatitis (3.6%) and Gianotti-Crosti syndrome (3.2%) were most common inflammatory conditions. In adolescents, scabies (45.4%) and intertrigo (14.5%) were the most common and 14 cases (9.2%) were of sexually transmitted infections.

Conclusion:

Complicated anatomy and special physiological factors like moisture, friction and occlusion of genital, perineal, perianal and gluteal areas result in multiple inflammatory and infective diseases, especially in paediatric population because of lack of proper hygiene and communicability of disease amongst peers and household members. Sexually transmitted diseases are also on rise in adolescent as well as school going children. Therefore, suggesting attempts for parental awareness about the rising child sex abuse.

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Abstract N°: 6296

Methotrexate as a systemic monotherapy: how worthy is it in pediatric morphea?

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Introduction & Objectives: Morphea is an autoimmune disorder that often manifests in childhood. According to the therapeutic guidelines, methotrexate is recommended as a systemic treatment for pediatric morphea. Systemic corticosteroids may be prescribed for severe or rapidly progressing cases, but long-term use is generally avoided due to side effects. This study aims to evaluate the efficacy and tolerability of methotrexate monotherapy in pediatric morphea.

Materials & Methods: A transverse, retrospective, single center study was performed on 11 patients diagnosed with morphea who were treated with methotrexate. The study was carried out from October 2019 to March 2024 in the Pediatric Dermatology Department of Colentina Hospital.

Results: Among 11 patients, females accounted for 63.6% and males for 36.4%, with a median age of 11 years. Regarding the type of morphea, there were 5 cases of mixed type (45.5%), 3 of linear form (27.7%) and 3 of plaque morphea (27.7%).** Six subjects (54,5%) were treated with methotrexate and short courses of systemic corticosteroids (5 cases with both therapies from the beginning of the treatment and 1 case when corticosteroids were prescribed later) while 4 patients (36.3%) received methotrexate only. Only 1 patient (9.2%) was treated with systemic corticosteroids after the cessation of methotrexate due to persistent leukopenia. All patients received topical therapy with vitamin D analogs, topical corticosteroids, tacrolimus, scar remodeling supportive therapy as a gel (Allium cepa extract, heparin, allantoin in combination as inhibitor of the fibroblasts activity) and oral therapy with avocado-soybean unsaponifiables (for the anti-fibrosis effect). All patients were treated with subcutaneous methotrexate, with a maximum dose of 15 mg/week for the majority of patients (63.6%); 12.5 mg/week for 18.2% and 20 mg/week for 18.2%, accompanied by 1mg/day of folic acid on days without methotrexate administration. Treatment was temporarily ceased in 45.45% of cases, due to upper respiratory tract infections or SARS-CoV2 infection (18.18%), adverse effects (27.27%) and in 1 case due to retroauricular herpes zoster. Adverse effects of methotrexate that required temporary suspension included leukopenia with lymphopenia (18.18%), neutropenia (9.09%), hepatic cytolysis (9.09%) with treatment resuming upon normalization of values. 63.63% of patients did not present any adverse reactions, 9.09% experienced dizziness and 18.18% experienced nausea after methotrexate injection, resolving within 12-24 hours. Considering the efficacy of methotrexate as monotherapy, 6 of the patients (54.54%) out of which 3 were of the mixed type presented a decrease in LoSCAT score: the average activity disease score (LoSAI) decreased by 77.77% and the average damage score (LoSDI) decreased by 28.26%, 2 patients (18.18%) showed disease progression, which required a change of the treatment in one case, while in the other one the methotrexate dose was increased from 15 mg to 20 mg. In 3 cases (27.27%), methotrexate was ceased after 4 doses due to persistent side effects.

Conclusion: Methotrexate remains the favorite treatment option in pediatric morphea, especially the mixt and linear type. In monotherapy it can reduce disease activity, slow progression, diminish dyspigmentation and fibrotic lesions and improve mobility limitations in affected joints. By regular follow-up, side effects can be avoided, with a favorable risk-benefit ratio.

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Abstract N°: 6310

Spectrum of Connective tissue disorders in pediatric age group

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Introduction & Objectives:

Connective tissue disorders encompass a diverse group of autoimmune disorders with varied manifestation involving skin, joints, and internal organs. These disorders can manifest in various ways, involving multiple organs and presenting unique challenges in pediatric patients. Understanding the complexities of connective tissue disorders in pediatric populations is crucial for early detection, intervention and improving outcomes for patients. Wide array of autoimmune connective tissue diseases affecting the skin can occur in childhood; amongst them more common are cutaneous lupus erythematosus, morphea and dermatomyositis.

Materials & Methods:

Case 1: A-11-year-old female with scaly erythematous pruritic plaque over face, neck, retro auricular area and whitish papules over bilateral knuckles and elbows since 7 months associated with difficulty in getting up from squatting position and photosensitivity. Skin biopsy was consistent with dermatomyositis. Muscle enzymes were raised. She was managed with oral methotrexate, hydroxychloroquine and sunscreen.

Case 2: A-11-year-old male with scaly erythematous pruritic plaque over upper eyelid, cheeks, neck, back associated with photosensitivity and sclerotic indurated skin over left flank since 6 months. Muscle enzymes were normal. Skin biopsy from face showed interface dermatitis with mucin deposition and biopsy from flank showed features of morphea, was diagnosed as amyopathic dermatomyositis with morphea, was managed with oral methotrexate, hydroxychloroquine and sunscreen.

Case 3: A-12-year-old female with history of photosensitivity, joint pain, painful ulcerative lesions healing with scarring involving upper extremities since 1 year. On evaluation found to have elevated ANA, dsDNA levels, and pancytopenia. Skin biopsy was suggestive of vasculitis confirming the diagnosis of SLE with vasculitis. She was managed with oral azathioprine, oral hydroxychloroquine and sunscreen.

Case 4: A-11-year-old male with itchy scaly erythematous plaques since 5 years associated with photosensitivity without any difficulty from getting up from squatting position. Muscle enzymes were normal. Skin biopsy was suggestive of dermatomyositis. Patient was managed with oral methotrexate and sunscreen.

Case 5: A-13-year-old male with itchy hyperpigmented lesion over the dorsum of left hand since 7 years. No other significant skin lesion. On evaluation ANA was normal. Skin biopsy was suggestive of morphea. Patient was managed with oral methotrexate and topical tacrolimus ointment.

Results:

We hereby report five cases of pediatric connective tissue disorders with varied cutaneous manifestations, diagnosed with clinic-pathological correlation along with serology. Managed with weight appropriate dosage of immunosuppressives.

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Abstract N°: 6311

Vaccine-Induced Subcutaneous Aluminum Granulomas in Children: A Case Series

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Introduction & Objectives:

Worldwide, vaccination programs that follow a regular schedule have been shown to considerably reduce the mortality rate of diseases that can be prevented by vaccination in children under five.

Aluminum salts serve as common adjuvants in vaccines, though they can induce contact allergy and the development of vaccination granulomas. These granulomas present as small, subcutaneous nodules at the injection site, often accompanied by pruritus and hypertrichosis. The differential diagnosis for palpable nodularity in the thigh of a child are foreign body reaction, local infection, vascular malformation and skin tumor. Localised hypertricosis should be distinguished from smooth muscle hamarthroma and lichen simplex cronicus.

Materials & Methods:

We report 4 cases of female patients, ages two to five, who had extremely itchy plaques on their lateral thighs that lasted for one to four years**.** These plaques appeared at the injection site of the HEXAVALENT vaccine (diphtheria-tetanus-pertussis-polio-HiB-HB), several months after the third dose. Local manifestations included subcutaneous nodules, hypertrichosis, hyperpigmentation, and excoriations. Additionally, a patient developed cellulitis as a result of prurigo lesions superinfection. According to the patients' medical records, they received the HEXAVALENT vaccine at the ages of 2, 4, and 11 months.

Case 1 underwent 2 ultrasound assessments of the antero-lateral thigh, followed by skin biopsy conducted by the pediatric surgeon. Case 2 had an ultrasound and a MRI scan to exclude malignancy, recomended by the orthopedic surgeon.

Results:

Case 1 involves a 3-year-old patient, who underwent 2 thigh soft tissue ultrasounds within a span of 3 months. Both scans revealed findings suggestive for a postvaccinal granuloma, characterized by hypoechoic cystic structures in the hypodermis. Subsequently, a skin biopsy was performed, unveiling a lymphohistiocytic infiltrate with histiocytes displaying finely granular basophilic cytoplasm.

Case 2 is a 2-year-old patient with a medical history of a dermoid cyst in the left eye and a thyroglossal duct cyst. She underwent ultrasound and MRI examinations of the proximal lateral thigh. Both imaging modalities displayed features suggestive of postvaccinal granuloma in the subcutaneous tissue, without involvement of the muscular fascia.

Cases 3 and 4 presented to the dermatology department, and there were no further investigations performed, due to the specific clinical appearance.

Every patient received treatment with short courses of topical corticosteroids under occlusion, resulting in significant improvement of pruritus.

All patients were referred to the allergology department for epicutaneous aluminium testing. A positive test would confirm the diagnosis, while a negative result does not definitively rule it out.

Conclusion:

Data from literature indicate that the lesions typically resolve gradually, although the symptoms persist over a prolonged period and the associated pruritus can be severe. Notably, considering the benign nature of aluminium granulomas and the importance of routine vaccination, it is recommeded that the patients proceed with their vaccination schedule.

Accurate diagnosis can diminish parental concerns and the need for unnecessary invasive investigations, such as biopsies or excisional surgery.

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Abstract N°: 6343

Achromic patches following hematopoietic stem cell transplantation

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Introduction & Objectives:

Graft-versus-host disease (GVHD) is a multiorgan disease that happen as a common complication of hematopoietic stem cell transplantation (HSCT). The skin is the most common affected organ, thus, 80% of patients who develop acute GVHD have skin involvement at the time of diagnosis. Acute GVHD typically presents as a maculopapular rash while chronic GVHD has a more widely presentation which includes poikiloderma, lichen planus-like lesions and sclerodermiform features. Vitiligo-like lesions have been described as an infrequent manifestation, reported in 1.78% to 8% of patients after HSCT.

Our objective is to describe the clinical case of an 8-year-old male patient diagnosed with combined immunodeficiency caused by ARPC1B deficiency who underwent HSCT and four months after developed vitiligo-like chronic GVHD. This clinical presentation is rare, and few cases are reported in the literature.

Materials & Methods:

An 8-year-old patient who was born with a pathogenic variant in the ARPC1B gene causing an autosomic recesive combined immunodeficiency characterized by impaired T-cell migration and proliferation, increased levels of IgE and IgA, and thrombocytopenia. He underwent HSCT from an haploidentical related donor (father) and developed acute GVHD of the skin (stage I) and gastrointestinal tract (stage III), responsive to photopheresis, topical tacrolimus, enteral budesonide and mesalazine.

Six months after HSCT, the pediatric dermatology department was consulted for widespread depigmented macules and patches that appeared two months prior.

Results:

Examination revealed achromic macules and patches in the trunk and extremities that accentuated upon Wood's lamp examination. Family history was negative for pigmentary disorders. A 4-mm punch biopsy was performed, and histology revealed vacuolization of the basal epithelial layer, lymphocyte exocytosis and decreased melanocytes in the basal layer. All of which confirmed the diagnosis of vitiligo-like graft-versus-host disease.

Treatment was initiated with topical tacrolimus and pimecrolimus with partial response in the two-month visit, consisting of some of the patches having repigmentation islands.

Conclusion:

GVHD is a common and well-known complication of HSCT, and it is associated with an imbalance between autoreactive and autoregulatory lymphocytes. It has been hypothesized that it acts as a triggering factor for autoantibodies and the subsequent appearance of autoimmune diseases. As vitiligo is an autoimmune T-cell mediated process, it is critical to consider that its appearance in these patients can be associated with GVHD so a timely evaluation can be made, and the appropriate treatment be given.

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Abstract N°: 6430

"The Boy with Four Ears": a Rare Isolated Finding of Bilateral Accessory Tragi or a Case of Goldenhar Syndrome

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Introduction:

Accessory tragi are relatively common congenital anomalies of the external ears, occurring in 3-6 per 1000 live births, but their bilateral presence is much rarer, affecting only about 10% of all cases. They are generally benign findings, however, in some cases they can be a sign of a more complex issue.

Results:

This is a case of an 8-year-old boy who was brought to the clinic due to the presence of congenital bilateral periauricular nodules, which the parents wished to have surgically removed. Upon further investigation, it became clear that the nodules were in fact bilateral accessory tragi - a single sessile lobed accessory tragus in the intratragal area on the right side; on the left a sessile nodular and a sessile lobed accessory tragi in the intratragal area, as well as a pedunculated spherical accessory tragus with a cartilage root in the pretragal area. It was decided to have all of the lesions surgically removed with elliptical excisions.

With the knowledge that accessory tragi can be a sign of an underlying syndrome, a full physical examination of the patient was performed. Mandibular hypoplasia and facial asymmetry, affecting the left side of the patient's face, were noted. Additionally, the patient suffered from a form of vertebral abnormality (kyphosis and possibly scoliosis) which required a consultation with an orthopedist. This cluster of symptoms pointed in the direction of a developmental disorder mainly characterized by craniofacial and vertebral anomalies - Goldenhar syndrome (a.k.a. Oculo-auriculo-vertebral spectrum (OAVS)). OAVS is a congenital disorder with a heterogenous phenotype that affects structures derived from the first and second branchial arch, as well as the vertebrae, and other systemic abnormalities. Due to the multisystem involvement of the disorder, patients require an interprofessional approach and a long-term follow-up. The patient's family was advised to arrange the necessary consultations, in order to establish the extent of involvement of the different organ systems, and to ensure the proper management of the child's disorder.

Conclusion:

With this case, we want to emphasize the need for a high index of suspicion when it comes to dealing with accessory tragi, which may seem as simply a cosmetic issue, but can in fact be a sign of serious congenital disorders, which require multidisciplinary management.

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Abstract N°: 6496

Epidemiology study of fungal infection

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Introduction & Objectives:

Fungal infection is one of the most widespread infection seen all over the world. In children it holds a special threat as it is often masked or neglected. It is equally seen in urban as well as rural set up. The presentation may vary to some extent.

The quality of life and the lifestyle has an indirect impact on occurence of the disease. This study was undertaken to understand those elements which has a link to occurence and reoccurrence of fungal infection.

Materials & Methods:

3500 cases of fungal infection in children under age 3-13 years.

Time frame of 6 months.

1700:- boys and 1800:- girls.

Children were on treatment for fungal infection.

Systemic diseases were eliminated.

Parent consent taken.

70 % cases were Tinea corporis. Remaining cases were Tinea cruris,Tinea faciei, Tinea scalp, secondarily infection, concomitant infection with other viral infection, and few along with atopic dermatitis.

Results:

Children residing in damp places were more affected. Children with temporary house shed faced more problems during summers due to indirect heat.

Incidence was high in children staying in overcrowded huts. The severely of disease was high in houses with poor ventilation. 10% Children with poor nutrition has more recurrence . 12 % rates were higher in families with more than 1 child.

Rates were low in children where parents had basic education (Educated above class 5-class 7) in comparison to child belonging to uneducated parents. Rates of fungal infection was high in females post menstruation.

Conclusion:

This study has not only helped in understanding the nature of the disease but also helped in preventing the reoccurrence to disease in children.

Highlighting the importance of basic hygiene especially for girls during mensuration has helped to reduce the rate

by 20%.

In boys simple suggestions like trimming hair short during summers, keeping toes open and dry, changing into extra pair of clothes after exercise has helped to eliminate fungal infection.

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Abstract N°: 6501

A retrospective epidemiological study of skin diseases among pediatric population attending a tertiary care hospital

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Introduction & Objectives:

The incidence of skin diseases in children is influenced by hereditary, social, and environmental factors. The disease pattern differs in a given population by different ecological factors. The aim of this study the prevalence of different dermatologic diseases in paediatric patients

Materials & Methods:

We retrospectively evaluated paediatric patients (0-14 years), visiting dermatology OPD between January 2021 and feburary 2024. Demographic and clinical profile of these patients were studied.

Results:

12564 children were included in the study (5699 females and 6865 males), with a male to female ratio of 1.2:1. A total of 1125 infants & 100 neonates were included.

Neonatal dermatoses constituted 0.9% cases. Erythema toxicorum neonatorum, seborrheic dermatitis ,milia, mongolian spots were the common neonatal dermatoses seen.

The most frequent diagnoses in 1yr to 14 years age group were : Infections (27%), eczema (2%), Atopic dermatitis (2%), Vitiligo (1.04%), Urticaria (1%), Alopecia areata (0.74%), Seborrheic dermatitis (0.76%), Acne (0.6%); Nevi, Vascular malformations and Hemangiomas (0.35%), Psoriasis (0.28%), icthyosis (0.08%).

Scabies (11.2%) was the most prevalent infection followed by Fungal infections (8%). dermatophytosis (7.4%) was the most common fungal infection.

Molluscum contagiosum (2%) was the most common viral infection. Leprosy (07cases) and tuberculosis (07 cases) were the mycobacterium diseases seen in our study.

Other dermatoses seen in our study were genodermatoses (0.02%) , nutritional dermatoses(0.63%).

Results:

Paediatric dermatoses includes various condition ranging from transient neonatal dermatoses to severe infections & inflammatory dermatoses. Additionaly these dermatoses cause loss of school days, low self esteem in children and anxiety in parents hence adequate knowledge of paediatric dermatoses is important for early diagnosis and subsequent management.

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Abstract N°: 6503

Deep vein thrombosis revealing a rare lymphomatous entity: non-specific peripheral T-cell lymphoma: case report

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Introduction & Objectives:

Non-specific peripheral T-cell lymphoma is a rare and heterogeneous entity among lymphomas, posing a real diagnostic challenge. Thromboembolic complications occur in 15–25% of cancer patients. Herein, we present a unique case entailing both diagnostic challenges and thromboembolic complications.

Case description:

A 45-year-old patient with no significant medical history was admitted to our department for nodular lesions present for 3 months, initially localized to the forearms, complicated by the onset of a painful swelling in the right upper limb and the extension of lesions to the rest of the body, with preserved general condition. The patient had undergone a biopsy elsewhere, indicating septal panniculitis with vasculitis. Clinical examination revealed painful dermo-hypodermic nodular lesions on the forearms, confluent into plaques measuring approximately 10 cm in the longest axis, firm, fixed with an ulcerative-necrotic center, evolving on a swollen and painful upper limb. Additionally, multiple non-painful, non-ulcerated nodular lesions were observed diffusely on the back, trunk, thighs, and soles of the feet, measuring between 20-50 mm in diameter, along with two left axillary lymphadenopathies. Investigations showed normochromic normocytic anemia, leukopenia, lymphopenia, decreased CD3, CD4, CD8, and NK lymphocyte subpopulations by flow cytometry, with elevated LDH levels at 416. Emergency doppler ultrasound of the right upper limb revealed extensive venous thrombosis necessitating anticoagulant therapy. Further examinations revealed thickening and tissue infiltration in the upper limbs, as well as bilateral inguinal and axillary lymphadenopathies, some with cortical thickening. Additionally, pulmonary nodular consolidation was detected in the left postero-basal lobe of the lung on a bodyscan. Skin biopsy showed T-cell lymphoproliferation, with immunohistochemical analysis revealing a CD3+ CD4+ CD8- CD20- CD56- CD30- CD68+ TdT- profile with a Ki-67 proliferative index of 30%, consistent with non-specific peripheral T-cell lymphoma. Over the course of a few weeks, there was rapid extension and evolution towards ulcerative necrosis, along with lesion superinfection. Chemotherapy was initiated for this patient following discussion in the oncology-hematology team.

Conclusion:

Non-specific peripheral T-cell lymphoma is a rare, heterogeneous entity that poses diagnostic challenges requiring collaboration between clinicians and pathologists. It is characterized by the presence of EBV in 30% of cases and a CD4+/CD8- immunohistochemical profile. Thromboembolic complications, the leading cause of morbidity and the second leading cause of mortality, are influenced by multiple risk factors such as immobility and tumor vessel compression. Chemotherapy appears to reduce clot formation and strength, but further studies are needed to better classify non-specific peripheral T-cell lymphomas in order to tailor therapies accordingly.

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Abstract N°: 6532

Photosensitivity in pediatric age group

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Introduction & Objectives: Diagnosis of photosensitivity in a pediatric age group is based on accurate history taking and careful physical examnination. It's rare entity seen in children and hence it's a challenge to diagnose on time and prevent further complications.

Materials & Methods:

Steps to accurately diagnose photosensitivity in children

\1. Accurate history taking.It should include age of onset, onset during which time of the year, when symptoms appear after sun exposure, duration of the eruption, any history of exposure to photosensitizers, and family history

\2. The physical examination should include careful observation of the distribution and morphology of lesions

3.To aid diagnosis Plasma porphyrins, autoimmune profile, a skin biopsy specimen, and phototesting may be useful

Results: Photosensitivity in children can be broadly classified as follows:-

1. Idiopathic photodermatoses:- Immunologically mediated),:- Polymorphic light eruption is the most common diagnosis. Followed by actinic prurigo and solar urticaria.

Polymorphic light eruption is most common entity seen in children.It appears as erythromatosus macule , papule or vesicle with pruritus arising after 1-2 days of sun exposure and resolving after 10 days on own

2 Photodeematoses secondary to exogenous agent or endogenous agents.

:- NSAIDs, anti-malarial, diuretic sunscreens and sandwood oil are few examples

3.Due to DNA defect:- Xeroderma pigmentoism, Bloom's syndrome are few examples.

4. Photoaggrevated dermatoses :- Lupus erythromatosus, pellagra, acne vulgaris, seborrhoeic dermatitis, atopic dermatitis.

Conclusion:

If the child develops a sunburn reaction, swelling, intense pruritus, skin fragility, or scarring even after limited sun exposure mainly over sun-exposed areas then Photosensitivity should be suspected. The immunologically mediated photodermatoses in children is uncommon than seen in adults. Systemic disorders, such as genetic or metabolic defects, should be considered in children who have strong family history, and who develop the symptoms early in life. PMLE is the most common pediatric condition seen.

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Abstract N°: 6541

Intra-lesional infiltration of meglumine antimony in pediatric periorificial leishmaniasis: still a contraindication?

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Introduction & Objectives:

Cutaneous leishmaniasis is a parasitic disease caused by flagellate protozoa of the genus Leishmania. Meglumine antimonate or Glucantime® remains the first-line treatment for cutaneous leishmaniasis in Morocco, administered either systemically or intralesionally. The intralesional route is indicated for small, single lesions; periorificial locations are considered “at risk” in this technique, and practitioners tend to switch automatically to the systemic route in these locations.

Materials & Methods:

Our study is a retrospective compilation of cases of periorificial LC in children treated with intralesional infiltration of meglumine antimony, observed in our department.

Results:

We recorded 4 patients. The diagnosis of cutaneous leishmaniasis was confirmed by dermal smear (3 cases) or biopsy (1 case). The mean age was 6 years. The sex ratio (M/F) was 1. The mean duration of the disease was 4.7 months. The lesions were located on the tip of the nose in 3 cases and periorbitally in 1 case. All lesions were single and ranged in size from 1 to 3 cm. All patients were treated with intralesional infiltration of meglumine antimony. The number of infiltrations varied between 6 and 10 at a rate of 1ml per cm2 once or twice a week. Progress was favourable in all cases. No complications were observed. The average healing time after treatment was 3.2 months.

Discussion:

The periorificial regions are considered to be delicate areas with no indication for intralesional infiltration of meglumine antimony in the treatment guidelines for cutaneous leishmaniasis. This has led to a tendency to avoid these areas in daily practice, in favour of systemic treatment. This automatic switch to systemic treatment in children, with all its side-effects, sometimes seems to be abused. This is the case for small lesions where the only indication for systemic treatment is the site of the lesion. This corresponds to the lesion profile of the cases in our series. This is all the more true as the excessive precaution found in the recommendations is only really based on the scant literature available concerning intralesional treatment of these areas. Few articles have raised this question. There are a few case reports and 7 cases in a South American series comparing intralesional and intramuscular treatment with AM. Although rare, all these trials of IL infiltration in these “at-risk” regions have been carried out without notable incidents. This is the case in our series which is, to our knowledge, the only study reporting cases of cutaneous Leishmaniasis of periorificial site in children treated with Intra-lesional meglumine antimony.

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Abstract N°: 6555

Lineal immunoglobulin A bullous dermatosis in children: about a case.

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Introduction & Objectives:

Linear immunoglobulin A (IgA) bullous dermatosis (LABD) is an autoimmune blistering disease notable for subepidermal blisters and linear IgA basement membrane antibody deposition. The incidence of LABD is 0.5 per million in western Europe, making it a rare disease. In children, the average age of onset ranges from four to five years. The most common presentation is blistered skin accompanied by mild itching or intense burning, typically affecting the face, perineal area, lower abdomen and thighs. The diagnosis of LABD requires a clinicopathologic correlation for a definitive diagnosis. There are currently no guidelines on the management of LABD, although usually dapsone is positioned as the first line of treatment.

Materials & Methods:

A 5-year-old girl with a 4-month-history of generalized blistering lesions accompanied by intense pruritus. The onset was not related to sun exposure, drug intake or systemic symptoms. The patient had erythematous plaques with annular advancing edges and vesicles located on the edges (string-of-pearls sign), as well as multiple serohematic crusts. The first pathology to be ruled out was LABD, a skin biopsy was performed, which later confirmed the suspicion, and a blood test including glucose-6-phosphate dehydrogenase (G6PD) level was requested. While waiting for the results, she was treated with topical and systemic corticosteroids, with clinical improvement but worsening when the dose was reduced, having in one of these moments great affectation of the perineal area.

Results:

For 5 months she has been treated with dapsone in oral solution, currently at a dose of 1mg/kg/day (at the beginning 2 mg/kg/day and concomitantly with systemic corticosteroids) with great improvement of the blisters, although with clinical worsening when reducing the dose.

Conclusion:

Dapsone is a first-line treatment in most of the cases, in children lower doses than adults (0.5-2mg/kg/day) are used. It is imperative to first check a G6PD level before initiating dapsone as G6PD deficiency can cause life-threatening hemolysis. Moreover, the clinical course can last for years and a balance must be achieved between the minimum effective dose of dapsone and no side effects. In cases of people with G6PD deficiency or allergic to dapsone, systemic corticosteroids are considered as the first option, although with important adverse effects derived from their prolonged use. There are other alternative treatments for LABD reported, most of them based on anecdotal evidence, but there is no definitive algorithm to follow in dapsone-refractory cases. We present a pediatric case affected with a difficult pathology to approach and manage.

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Abstract N°: 6582

a case of extensive pyoderma gangrenosum in a 5-year-old child successfully treated with intra veinous immunoglobulin

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Introduction & Objectives:

Pyoderma gangrenosum (PG) is a primarily sterile inflammatory neutrophilic dermatosis .Cases in infants and adolescents (< 15 years) account for only 4%. We report an extensive PG occuring in a 5-year- old girl with a good response to Intravenous immunoglobulin.

Materials & Methods:

A 5 year old girl was referred to our clinic due to a 2 years history of widespread painful ulcerations all over her body. Lesions started with an erythematous pustule in her leg that increased in size to form a nodule that broke into painful ulcer.Then gradually numerous similar non-healing ulcers developed on the rest of her body. Multiple courses of antibiotics were ineffective.

On examination, she was diagnosed with failure to thrive, with significant thinness and pallor. Cutanesous exam showed multiple ulcers of varying shape and size, with violaceous borders and granulation tissue at the base, partly covered by yellowish adhrent crusts. Ulcers were located on the back,trunc, buttocks and 4 limbs. Otherwise, there were two pustules on her left leg and multiple white atrophic rounded scars along her body. Biopsy specimens for infectious diseases screening (special stains, culture, PCR) were negative. Bacteriological sampling was sterile. Pathergy test was positive. Laboratory tests** showed severe anemia** (Hb: 62g/L), white blood cell count of 24 X 10^9/L with 86.7% neutrophils. Peripheral smear, chest-X ray, colonoscopy, HIV tests, immunoglobulins and complement levels were normal. Histopathological examination revealed a massive neutrophilic infiltrate in the dermis. The diagnosis of PG was made. The patient was treated with intravenous imunoglobulin (IVIG) at 2g/kg/d for 5 days. The ulcers started to heal within 1 week after the first IVIG course. The treatment was continued for 6 months with a monthly infusion allowing remission to be maintained up to 8 months.

Results:

Pyoderma gangrenosum is relatively rare in the pediatric population. In a Japanese survey, only 2 of 473 patients were less thant 10 years-old. Clinically, as in adults, the ulcerative type is most common. Diagnosis of pediatric PG is often delayed, likely because of its low prevalence among children and it can be misdiagnosed with other causes of ulcerative diseases. In our patient, the non-response to antibiotics, clinical features, pathergy phenomenon and characteristic histology were suggestive of diagnosis. PG has been reported among pediatric population in association with ulcerative colitis, Crohn’s disease, hematologic malignancies,various immunodeficiencies and auto-inflammatory syndromes.The screening for comorbidities were negative. Corticosteroids are often employed as a first line agent,** but their numerous side effects, especially on long-term use, are very limiting, particularly in children with the risk of stunted growth. Second line treatments are cyclosporine, TNFα inhibitors, immunosuppressants. IVIG were used in refractory PG with rapid effectiveness confirmed on pain and healing-time ulcerations. Our patient had a spectacular response to IVIG courses with a sustained remission after **8** months of follow-up.

Conclusion:

Pyoderma gangrenosum is very rare in infants. We highlight the challenging diagnosis in presence of persistent cutaneous ulcers in children that can delay the early management of the disease. IVIg could be an interesting first-line treatment instead of corticosteroids in pediatric population.

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Abstract N°: 6588

Juvenile dermatomyositis: increase in its incidence since the COVID pandemic.

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Introduction & Objectives:

Since the beginning of the COVID pandemic, we have registered an increase in the incidence of cases of juvenile dermatomyositis (JDM) in our hospital, compared to the previous two decades. Between the years 1999-2019 we registered 8 cases of JDM, and from 2020, 9 new cases have been diagnosed.

Materials & Methods: Prospective observational study. We gathered epidemiological, clinical and laboratory data from all children diagnosed with JDM since 2020.

Results: The age of the patients ranged between 8 and 13 years. Most were female (6/9 patients). Symptoms began during the autumn months in most cases. All but one patient had myalgias. In addition to the classic signs of JDM, involvement of the oral mucosa (6/9 patients) and presence of acral lesions (8/9 patients) were noted. Two patients were positive for anti-TIF1 gamma antibodies and four for anti-MDA5 antibodies.

Conclusion:

We present a series of patients with JDM diagnosed over the last four years in our hospital. We have noticed an increase in the incidence of JDM since the start of the COVID pandemic. Some of the skin manifestations described in this study, such as oral lesions and acral involvement, have been rarely reported previously. Like other patients described in the literature, children with anti-TIF1gamma antibodies presented greater severity and more extensive and recalcitrant skin involvement. Muscle biopsy has been left aside in recent years since MRI is a less invasive test. In the last two years, we have diagnosed patients with milder skin manifestations, which could be due to vaccination against COVID, an increase in the suspicion of the disease and early diagnosis, as well as greater access to healthcare centers compared to the beginning of the pandemic.

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Abstract N°: 6626

Annular lesions in a febrile infant

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Introduction & Objectives:

Kawasaki disease is an acute febrile systemic vasculitis that occurs predominantly in infants and children younger than 5 years old.

Classic Kawasaki disease is diagnosed in the presence of fever for at least 5 days, together with at least 4 of the 5 following main clinical features: Erythema and cracking of lips, strawberry tongue, and/or erythema of oral and pharyngeal mucosa, bilateral bulbar conjunctival injection without exudate, rash, erythema and edema of the hands and feet or periungual peeling, cervical lymphadenopathy (≥1.5 cm diameter), usually unilateral.

However, some patients with Kawasaki Disease do not meet all diagnostic criteria and may have an incomplete or atypical presentation of the disease.

Materials & Methods:

Male patient, 1 year 11 months old, previously healthy, with a history of 7 days of high-grade fever and difficult control who presented to the emergency department of our hospital due to cough, decreased consistency of stools, exanthema with annular lesions and cheilitis, vomiting of gastroalimentary content, and jaundice.

Results:

Laboratory work-up revealed microcytic hypochromic anemia (Hb 9.9 g/dL), hyponatremia (Na 130 mEq/L), elevated ALT and AST (56 IU/L and 65 IU/L respectively), cholestatic pattern hyperbilirubinemia (Total bilirubin 4.83 mg/dL and indirect bilirubin 1.88 mg/dL), hypertriglyceridemia (200 mg/dl), and hypoalbuminemia (2.8 g/dl).

Our Dermatology service was consulted due to the presence of a rash with annular lesions on the thighs, edema and macular lesions on the palms and soles, perioral mucositis, cheilitis, and perianal peeling.

With the data of long-standing fever and the rest of the clinical and laboratory findings, the diagnosis of incomplete Kawasaki disease was made, so a transthoracic echocardiogram was requested, reporting minimal pericardial effusion without hemodynamic repercussion and without coronary alterations.

Treatment was started with intravenous gammaglobulin at 2 g/kg/dose, ASA at 5 mg/kg/day and methylprednisolone at 2 mg/kg/day in two doses, on the 8th day of evolution; A respiratory viral panel was requested resulting positive for Influenza AH1N1, adding oseltamivir to treatment.

The patient improved and was able to be discharged after 8 days of hospital stay and in follow-up appointments, the mother mentioned that the patient had presented peeling of the palms and soles and the physical examination already showed some of Beau's lines.

Conclusion:

The skin manifestations in Kawasaki Disease are usually variable and non-specific, but the rash most frequently presents with erythematous maculopapular lesions, on the extensor surfaces and the trunk, erythema multiforme-

like lesions are an infrequent presentation.

Kawasaki Disease is now the most common cause of acquired heart disease in children in developed countries and is crucial to diagnose it early to prevent serious cardiac complications through timely treatment.

We present the case of a patient with incomplete Kawasaki disease, in which the rash presentation were annular lesions, demonstrating the important variability of cutaneous manifestations and the identification as part of this disease.

There is no definitive diagnostic test or pathognomonic clinical characteristic for Kawasaki Disease, so doctors must be attentive to the different forms of clinical presentation of the disease.

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Abstract N°: 6644

Two NRAS-mutated congenital melanomas in congenital melanocytic nevi

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Introduction & Objectives: Less than 0.3% of melanomas occur in pre-pubertal children. Less than 100 cases of congenital melanomas are described. We report a very rare case of two congenital melanomas in a patient with a giant congenital melanocytic nevi.

Materials & Methods: Case report.

Results: Patient born at term presenting a plaque with defined limits on the mid and lower back, buttocks, abdomen, genitalia and thighs with black, light brown and dark brown areas. She also had brown and black papules and nodules interspersed with the main plaque and papules and nodules on her legs bilaterally, dissociated from the main plaque. The patient received a clinical diagnosis of giant congenital melanocytic nevi of the trunk G1S1C2R2N2H0.

In the left lumbar region, the patient presented a hard tumor, measuring approximately 5cm, exophytic, infiltrated, attached to deep planes, painless. Clinically, melanoma, fibrosarcoma and rhabdomyosarcoma were hypothesized.

The investigation of central nervous system involvement using magnetic resonance imaging was negative.

Magnetic resonance imaging shows, in tumor investigation, the presence of a lesion with expansive behavior, with regular and well-defined contours, measuring approximately 3.9x3.5x2.5cm in its longitudinal, latero-lateral and antero-posterior axés, respectively, presenting hypersignal in liquid-sensitive sequences and signal intensity similar to muscle bellies on T1, superficially located in soft tissues of the lumbosacral region to the left of the midline, exophytic in character, and which undergoes heterogeneous enhancement after intravenous injection of the medium paramagnetic contrast. In deep planes, the lesion is in close contact with the subcutaneous cellular tissue, without evident signs of invasion of the underlying muscular structures.

Excisional biopsy of the tumor showed asymmetric, nodular and extensively ulcerated proliferation of epithelioid to spindle-shaped melanocytes with atypia, mitosis and solid and infiltrative growth. In the periphery, cells with a congenital pattern. There is positivity for Melan-A, HMB45 and S100, but there is loss of expression of melanocytic markers, suggesting dedifferentiation. Breslow was 15.6mm. Positive Gln61Arg NRAS was found. There was negative KRAS mutations(figure 1).

There was growth and change in consistency of the nodule, which had been present since birth, and increased to 1.2cm, in the left infrascapular region. A new excision was performed at 45 days of life. Histology and immunohistochemistry revealed the second melanoma. Breslow was 2.1mm.

The margins of both melanomas were enlarged. Staging showed that it was congenital melanoma in giant

congenital melanocytic nevus T4BN0M0-AJCC IIC and T3AN0M0-AJCC IIA.

Due to positivity for NRAS, treatment with trametinib was started.

Conclusion: Congenital melanomas are extremely rare tumors that present biological peculiarities that directly impact the investigation, staging and treatment process.

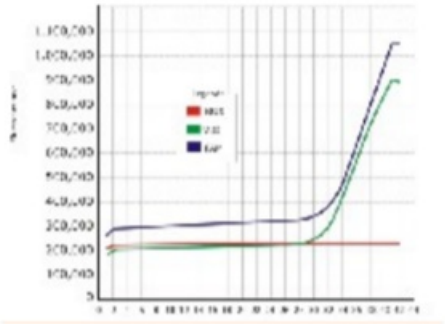

Figure 1: Allelic discrimination graph showing amplification of wild-type (green) and mutated (blue) alleles.

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Abstract N°: 6686

Dermatological involvement in pediatric patients diagnosed with leukemia

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Introduction & Objectives:

Leukemias have multiple clinical manifestations, including cutaneous manifestations, occasionally secondary to epithelial infiltration of neoplastic hematopoietic cells, which can occur in up to 77% of patients with leukemia diagnosis. These lesions can be classified as specific or nonspecific; specific lesions. Are produced by direct infiltration of hematopoietic cells into the dermal tissue, Nonspecific lesions include paraneoplastic lesions secondary to chemotherapy or infectious lesions. Cutaneous manifestations require patients to undergo long-term treatments complementary to chemotherapy, such as alkylating agents, antimetabolites, antibiotics, and steroids, which increase hospital stays and costs.

Objective: describe the dermatological manifestations and sociodemographic characteristics of pediatric patients diagnosed with leukemia.

Materials & Methods:

An observational, descriptive, retrospective study was conducted, including patients diagnosed with leukemia who presented dermatological manifestations and had been treated at a tertiary-level institution, between 2014 - 2019.

A non-probabilistic sampling of consecutive cases was conducted, the source of information was secondary through medical records. The data collection instrument used was a standardized form designed by the research group, including variables established in the study protocol. Data analysis was carried out using a SPSS program, employing univariate analysis through proportions, absolute, and relative frequencies.

Results:

49 patients were included. Median age of 11 years, 57% was males. The median age at diagnosis was 5 years, The most frequent leukemia was lymphoid leukemia (71.4%), with acute lymphoblastic leukemia (ALL) being the most common subtype at 94.2% (33).

Cutaneous manifestations: 59% of patients had both primary and secondary lesions, 31% had only primary lesions. Characteristics of the lesions: 86% were described as multiple, 14% were grouped, 47% were localized, 53% (26) were non-painful, Regarding color, 61.2% (30) were hyperpigmented. The dermatological diagnoses reported for the patients, 8% corresponded to pruritus, 6% Herpes Zoster, 4% atopic dermatitis, 4% scabies, 4% xerosis, and 2% Herpes Simplex. 6% of patients did not receive a dermatological diagnosis, and 57% were reported with other diagnoses, including graft-versus-host disease (6%), bullous dermatosis (4%), staphylococcal scalded skin syndrome (4%), and urticaria (4%). 84% of patients received specific treatments for the lesions, 10% did not receive treatment. The patients who received treatment, 43.9% showed improvement, 14.6% showed no evident changes, 2.4% worsened with treatment. Following this treatment, lesions emerged in 47% (23) of patients, there were no changes in lesions in 12%, lesions improved in 8%, worsened in 4%.

Patient outcomes at 6 months after diagnosis, 51% (25) entered remission, 28.5% did not achieve remission, 12.2% (6) experienced relapses, 2% (1) died.

Conclusion:

The challenge for all healthcare professionals involved in the multidisciplinary management of leukemia patients is early diagnosis. Recognizing paraneoplastic dermatoses associated with leukemia diagnosis is crucial. Although they are challenging to diagnose and sometimes overlooked, they can serve as the first warning sign of an underlying hidden malignancy

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Abstract N°: 6701

Maternal transfer of antibodies in bullous pemphigoid of infancy

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Introduction & Objectives:

Bullous pemphigoid is an autoimmune, blistering disease that is often rarely seen in children.

Materials & Methods:

We present a case of bullous pemphigoid occurring in a 2-month-old infant who developed a blistering eruption initially affecting the hands and the feet before spreading to other parts of the body. The diagnosis was confirmed on histology with direct immunofluorescence and autoantibody testing. Given the early age of onset of the condition, the mother was also tested for autoantibodies which were positive despite not displaying any clinical features of bullous pemphigoid.

Results:

The infant was treated with high-strength topical corticosteroid ointment and a short course of oral antibiotics with complete resolution within 2 months from diagnosis.

Conclusion:

Transplacental transfer of antibodies from the mother to foetus is a known mechanism that provides protection for the infant and forms the foundation for maternal immunization strategies, where antibodies may be present up to 6-12 months from birth. Vertical transfer of anitbodies has not been well described in infantile bullous pemphigoid. We postulate that the mother had low levels of BP180 and BP230 that were insufficient to have caused disease but through active transplacental transfer of antibodies, led to levels capable of inducing clinical disease in her baby. Serial testing of both mother and infant demonstrated absence of antibodies 6 months later.

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Abstract N°: 6710

Gianotti-Crosti syndrome and Nail matrix arrest overlapping

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Introduction & Objectives:

Gianotti-Crosti syndrome is a self-limited childhood exanthem that manifests in a characteristic acral distribution, associated with Epstein-Bar virus, hepatitis B, A and many others.

Nail matrix arrest is caused by a temporary arrest in nail plate formation and is reversible. Many conditions like systemic illness, drugs, fever, periungual dermatitis, trauma, and infection including viral are associated with nail matrix arrest.

Materials & Methods:

We present 2 years boy with symmetric, exanthematous, asymptomatic cutaneous eruption following low-grade fever, malaise, pharyngitis and symptoms of an upper respiratory tract infection. He was treated with antipyretic and antihistaminic. The laboratory tests were normal.

Results:

Because of typical diagnostic criteria for GCS like: monomorphous, flat-topped, pink-brown papules or papulovesicles 1-10 mm in diameter, the localization on four sites: cheeks, buttocks, extensor surfaces of forearms, extensor surfaces of legs, symmetric eruption, absence of extensive truncal lesions and scaling, we confirm the diagnosis. We treated him with low potent corticosteroid cream diluted with emollient, and antihistaminic. After 10 days the papules ended leaving hypo and hyperpigmentation and sporadic papules which last 1,5 month.

Two weeks after the regression, the nail matrix arrest disappeared with Beau's line (the transverse ridging of the nail plate) and onychomadesis We gave only the emollient cream and after a few month the nails became normal.

Conclusion:

To the best of our knowledge, there have been no clinical studies and not so many cases regarding the association between GCS and nail matrix arrest in Macedonia

The nail matrix arrest is usually associated with entero, coxsackie, citomegalo and echo viruses but the mother did not allow any blood examination to specify it.

This is one rare and interesting case of overlapping of two dermatosis, and in our opinion the best solution for cure the illness is education of patient about the disease.

In many cases there is no need of treatment.

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Abstract N°: 6741

A young girl with Stevens-Johnson Syndrome: an expanded spectrum of adenovirus infections

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Introduction & Objectives:

Dermatological manifestations of adenovirus infection are diverse. Severe mucocutaneous involvement is rare but has been described in isolated case series. SJS/TEN is potentially fatal. It is crucial to recognize the condition early and institute aggressive treatment in a timely manner.

Materials & Methods:

We present a case of a critically ill young child with adenovirus infection who developed SJS with severe mucocutaneous involvement.

Results:

A 6-year-old Malay girl with underlying Dravet syndrome presented with conjunctivitis, lip erosions and a maculopapular rash. She subsequently developed severe mucositis over multiple sites and a positive Nikolsky sign. A polymerase chain reaction (PCR) for respiratory viruses returned positive for adenovirus, and the skin biopsy showed subepidermal blister with full thickness epidermal necrosis and interface dermatitis consistent with SJS. She required intensive care and was also treated with systemic steroids, intravenous immunoglobulin and ciclosporin.

Conclusion:

We describe a rare case of life-threatening mucocutaneous eruption associated with adenovirus infection. Supportive care administered by a multi-disciplinary team is the mainstay of treatment in SJS. Early recognition of SJS/TEN is essential but remains a diagnostic challenge.

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Abstract N°: 6774

Guiding Light: Navigating the Complexities of Xeroderma Pigmentosum Variant (XP-C) in Pediatric Care

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Introduction & Objectives:

Xeroderma Pigmentosum (XP) is a rare autosomal recessive disorder characterized by extreme sensitivity to ultraviolet (UV) radiation, leading to progressive cutaneous and ocular manifestations. Among its variants, XP-C, resulting from mutations in the XPC gene, presents unique challenges in diagnosis and management, particularly in pediatric patients. We present a comprehensive approach to the management of XP-C in a 6-year-old female patient, emphasizing the importance of early intervention and multidisciplinary care.

Materials & Methods:

We conducted a thorough clinical evaluation of the patient, including dermatological and ophthalmological examinations, dermoscopy, and genetic testing to confirm the diagnosis of XP-C. A multidisciplinary team comprising dermatologists, ophthalmologists, and geneticists collaborated to develop a personalized management plan tailored to the patient's specific needs. The plan focused on strict photoprotection measures, topical treatments for cutaneous lesions, and regular follow-up assessments.

Results:

The patient presented with typical cutaneous manifestations of XP-C, including poikiloderma, xerosis, and pigmented lesions, along with ocular abnormalities such as conjunctivitis and decreased visual acuity. Dermoscopic examination revealed characteristic findings, aiding in the diagnosis of suspicious lesions. Genetic testing confirmed a pathogenic variant in the XPC gene, establishing the diagnosis of XP-C. Implementation of the management plan resulted in significant improvement in the patient's symptoms and quality of life.

Conclusion:

This case highlights the importance of early recognition and comprehensive management of XP-C in pediatric patients. By employing a multidisciplinary approach and implementing strict photoprotection measures, we can effectively mitigate the risk of cutaneous malignancies and ocular complications associated with this rare genetic disorder. Our experience underscores the need for increased awareness among healthcare providers and emphasizes the pivotal role of proactive interventions in optimizing outcomes for patients with XP-C.

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Abstract N°: 6792

Rare Paediatric Dermatoses: A Case Series

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Rare Paediatric Dermatoses: A Case Series

Introduction & Objectives: Paediatric dermatoses encompass cutaneous disorders of wide array of variation and aetiologies. Their early detection facilitates decrease in morbidity and appropriate management. Genodermatoses are conditions caused due to genetic mutation often presenting with characteristic changes in skin and majority associated with systemic manifestations. Understanding the inheritance is a necessity in such disorders so as to predict the risk of recurrence in families affected. Here we present 4 such cases

Case 1: A 12-year-old female born out of a 3rd degree consanguineous marriage presented with pruritic erythroderma since birth without history of collodion membrane, fluid filled lesions. She had growth retardation, absence of axillary and pubic hair. Histopathology showed parakeratosis, hypergranulosis, dense lymphocytic infiltrate in upper dermis. Hemogram showed anaemia and hypoproteinaemia. Ultrasonography revealed hypoplastic uterus. On genetic analysis CDSN gene deletion on the chromosome 6 was found. Hence, a final diagnosis of peeling skin syndrome was established.

Case 2: A 6-year-old male born out of a non-consanguineous marriage presented with mildly pruritic varioliform scars on the trauma prone areas of extremity and face since 4 years along with moniliform papules over eyelids. History of fluid filled lesions preceding the present lesions was present. Parents noticed hoarseness of his voice. A diagnosis of lipoid proteinosis was made and biopsy revealed hyperkeratotic epidermis with mild papillomatosis, mild spongiosis and hypergranulosis with dermal mild superficial perivascular lymphocytic infiltrate with PAS positivity thus confirming our diagnosis.

Case 3: A 10-year-old female born out of a non-consanguineous marriage came with recurrent angioedema involving periocular and labial region of 6 months duration. Antecedent history of trauma and infection was present in each episode which were controlled by injectable hydrocortisone and pheniramine maleate. Further investigations revealed C1 esterase inhibitor deficiency (106) with normal C3 C4 levels. She was well controlled on oral tranexamic acid.

Case 4: A 16-year-old male born out of 3rd degree consanguineous marriage presented with pruritic lichenified plaques all over the body of 11 years duration. He had coarse facies along with short stature. He also had skeletal deformities of cubitus valgus, scoliosis, and genu valgus. Absence of secondary sexual character and undescended testes was also observed. Histopathology revealed lichenoid reaction. On enquiry he had been receiving injectable growth hormone following which he developed the complaints. Karyotyping revealed pathogenic variant of SOS1 gene indicating Noonan's syndrome 4. Hence final diagnosis of Lichenoid drug reaction secondary to growth hormone in a case of Noonan's syndrome was made.

Conclusion: Each condition presents unique challenges and requires tailored management strategies including genetic analysis where required.

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Abstract N°: 6800

Epigenetic profiling of N6-Methyladenosine and 5-hydroxymethylcytosine for Kaposiform hemangionedothelioma in children

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Introduction & Objectives:

Kaposiform hemangionedothelioma (KHE) is a rare vascular neoplasm in children with high mortality. The predominent features of KHE is progressive angiogenesis with local invasion. Consumptive coagulopathy in KHE is also known as Kasabach-Merritt phenomenon (KMP), which is life-threatening. Although the incidence of KHE is low, it is crucial to understand the pathogenesis of to improve the long-term prognosis of patients. Current studies have found mutations and key genes inducing angiogenesis and lymphangiogenesis in KHE, however, the pathophysiology may be attributed to a combination of events. To this end, we investigated the characters of epigenetic modifications in KHE samples to improve our knowledge of this disease.

Materials & Methods:

To investigate the epigenetic pathogenesis of KHE in children, two epigenetic biomarkers were studied, an RNA modification biomarker known as N6-Methyladenosine (m6A) and a DNA modification biomarker known as 5-hydroxymethylcytosine (5-hmC). To understand if the epigenetic biomarkers were differentially expressed in KHE compared to normal skin, 4 tissue samples from lesions of patients diagnosed with KHE and 3 normal skin tissue samples from patients with nevi were collected. The normal skin tissues from nevi patients were served as the control group. To investigate the epigenetic changes, tissue samples from both groups were checked with three second-generation sequencings, including transcriptome sequencing, MeRIP sequencing and hMeDIP sequencing. Results were collected to analysis the underlying epigenetic mechanisms in KHE.

Results:

Firstly, results showed that compared to the control group, 701 differentially up-regulated genes and 1376 down-regulated genes were found in the KHE samples. Enrichment analysis of the differential gene expression results revealed that the differentially expressed genes in KHE tissues mainly participate in physiological processes highly related to the phenotypes of this disease, including cell signaling transduction, vasculogenesis and angiogenesis. Among them, the enrichment factor significantly increased in biological processes such as vascular related smooth muscle development, regulation of angiotensin levels, and endothelial cell morphology development. Secondly, in terms of epigenetic modifications, compared to the control group, low expressions of m6A in the gene coding region were found in the KHE group, while an enrichment of m6A in the 3 'UTR region was observed in the KHE group. Moreover, compared to the control group, a significant modification of 5-hmC occurred in the gene promoter region in KHE samples. Genes with high expression of 5-hmC mostly encode axons and extracellular structures, indicating the regulatory role of neuroendocrine regulation in the stimulation of KHE, while genes with low expression of 5-hmC mostly encode cytoplasmic proteins, participating in protein connectivity and kinase activity regulation.

Conclusion:

In summary, differential epigenetic modifications, including m6A and 5-hmC, participated in multiple key

pathological processes in the pathogenesis of KHE. This study could give us new insights into understanding the pathogenesis of KHE at an epigenetic level.

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Abstract N°: 6833

A Case Report on Leukocyte Adhesion Deficiency Type I: An Important Consideration in the Clinical Differential Diagnosis of Recurrent and Non-Healing Wounds in An Infant.

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Introduction & Objectives:

Leukocyte adhesion deficiency (LAD-I), is a rare (incidence < 1:1 million), autosomal-recessively inherited primary immunodeficiency disorder, affecting the leukocyte adhesion and migration. It results from mutated ITGB2 gene on chromosome 21q22.3, encoding for 2-integrin (CD18), which is essential for leukocytic chemotaxis. LAD-1 is characterized by umbilical cord complications, persistent leukocytosis and recurrent, non-healing, diverse infections lacking classical pus formation. Severe LAD-I is fatal before 2 years of age without allogeneic hematopoietic stem cell transplant which, however, is limited by transplant-associated toxicity and graft-versus-host disease. Herein, we report LAD-I in a 1.5- month-old infant, presenting with a non-healing perianal wound.

Materials & Methods:

A 1.5-month-old, 2nd born male infant, of 2° consanguinity, presented to us with a solitary, horse-shoe shaped, punched-out perianal ulcer, with a clean base deprived of pus. Ulcer was irresponsive to antibiotics and surgical debridement. Patient also presented with omphalitis of persisting umbilical cord (usually sheds by 2nd week) and marked leukocytosis (92.5x103/µl). Ulcer worsened despite adequate management leading to death from septicaemia. There was history of similar complaints and neonatal death (due to bronchopneumonia) in elder sibling, thus, raising suspicion of primary immunodeficiency in our patient. In conjunction with family history, clinical and laboratory presentations, we evaluated for LAD-I and CD18 deficiency was detected via flow cytometry, hereby confirming diagnosis of LAD-I.

Results:

There are three sub-types of LAD with varying mutations and severity. Literature review of LAD-I's clinical presentation reveals recurrent necrotic infections of the skin and mucous membranes, progressing to extensive ulceration which is secondary to lack of leukocyte recruitment at infective site. There is delayed umbilical cord detachment, omphalitis, etc. Leukocyte quantification with defective CD18 expression determines severity. Rapid identification of LAD-I is essential to refer to centres of expertise and early implementation of definitive therapy. We counselled parents for prenatal diagnosis in subsequent pregnancies.

Conclusion:

LAD-1 was first described in 1980, and since then, approximately 300 patients with LAD-1 have been reported worldwide. Parenteral immunoglobulins, topical granulocyte-macrophage colony-stimulating factor administration for wound healing are possibly effective treatments for LAD-1 patient, but we were unable to explore them due to cost restrains. In summary, in children, chronic wounds accompanied by leukocytosis, omphalitis and delayed umbilical cord detachment are "red flags" with respect to the possible diagnosis of LAD-1. Further studies are needed to examine the role of IL-23 and IL-17 for LAD-1 severity.

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Abstract N°: 6939

Atypical Hormonal Crisis

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Introduction & Objectives:

Hormonal crisis is a phenomenon characterized by an increase in the size of the genital organs of newborns due to fluid accumulation under the influence of maternal hormones. Our case report presents the instance of a newborn in whom this diagnosis, although benign yet concerning, was made, with the particularity lying in the importance of considering the corrected age of premature newborns in the diagnostic reasoning.

Results:

This concerns a newborn at 60 days of life, with a history of prematurity at 33 weeks of gestation, with a corrected age of 1 week. She presented with vulvar edema evolving over 10 days. Breast examination was normal, as was the rest of the skin and general examination. No genital bleeding or discharge was reported by the parents. Given the absence of other suggestive signs, the diagnosis of newborn hormonal crisis was established. Symptoms resolved spontaneously 1 week later.

Signs of estrogenic impregnation diminish around the second week of life and should disappear around the eighth . Although our newborn was 60 days chronologically, her actual age was 1 week, which perfectly aligns with the diagnosis. It is necessary to consider this because being born earlier does not mean that all organs are mature. The maturation process is the same for a prematurely born baby or a term baby. Therefore, there is immaturity to consider.

Newborn hormonal crisis is characterized by an increase in the volume of the labia majora in girls, making the vulva moist and congested. Fluid vaginal discharge, mucous, sometimes milky, or resembling menstruation may occur. In boys, there is a transient increase in the volume of the penis, scrotum, and testicles, followed by rapid regression. For both sexes, it results in breast enlargement, without infection: the breasts take on the appearance of a mandarin, are red and swollen, sometimes with a discharge of a whitish fluid resembling milk. It is imperative to emphasize the importance of not manipulating or attempting to empty them, as this could lead to infections, potentially resulting in a breast abscess.

Conclusion:

The various transformations of newborn hormonal crisis constitute a normal process requiring no treatment. Generally, everything returns to normal within two to three weeks. The baby's organs regain a very infantile appearance, which persists until the eve of puberty. Medical consultation is recommended in case of abnormal prolongation of the transformations.

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Abstract N°: 7018

congenital syringocystadenoma papilliferum in an unusual location

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Introduction & Objectives:

Report one new case of congenital syringocystadenoma papilliferum in an inusual location.

Results:

A 1 month-boy was referred for consultation because of a nodule on the back. The lesion, present already at birth and was growing slowly . Examination showed a soft, red papule , 6 mm in diameter, on the back. A deep excision was performed. The histology demonstrated a characteristic endophytic glandular and cystic proliferation also with verrucous epidermal hyperplasia an glands with epidermal connection. It also showed glands with double layer of cuboidal columnar epithelium with some stromal plasma cells. We made a diagnosis of congenital syringocystadenoma papilliferum (SCAP). The excision site was closed primarily. At the follow-up 6 months postexcision, the patient showed no evidence of recurrence.

Conclusion:

Syringocystadenoma papilliferum is predominantly a childhood tumor: in about half of those affected it is present at birth, and in a further 15–30%, the tumor develops before puberty. Young children may have a solitary nodule or multiple tiny papules with a smooth, flat or dome-shaped surface. Larger nodules may ulcerate, draining a serous fluid, which produces a central crust. The small papules may be arranged in a linear pattern. The differential diagnosis of SCAP in pediatric patients includes viral diseases (molluscum contagiosum and verruca vulgaris), or other tumors of skin appendage origin. Its diagnosis can be best confirmed by histopathology. Observations in adults indicate the rare development of basal cell epithelioma or adenocarcinoma within the lesions. The prophylactic excision of SCAP is of uncertain benefit, but in case of growing or ulcerating lesions, surgical removal is recommended. In anatomic areas unfavorable to excision or grafting, CO2 laser excision is a clinical treatment option.

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Abstract N°: 7033

Generalized granuloma annulare in an infant with spontaneous resolution: A case report

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Introduction & Objectives:

Granuloma annulare (GA) is a benign, self-limiting inflammatory dermatosis of unknown etiology. It affects patients between the ages of 3 and 50, with an incidence of approximately 0.04%. There are several subtypes of GA, including the localized, generalized, subcutaneous, perforating, and patch variants, each with distinct clinical features. In the pediatric population, the most common form of GA is localized, and generalized GA(GGA) accounts for only about 15% of all cases. Herein, we describe the case of an infant with a generalized granuloma that was histopathologically confirmed and had a spontaneous resolution.

Materials & Methods:

Case report

Results:

A 2-year-old female infant presented with a history of generalized asymptomatic papules and annular plaques over the trunk, bilateral upper and lower limbs,and buttocks evolving for 3 months and sparing palms and plants. Physical examination revealed multiple (> 10) papules and annular plaques over the trunk and extremities, approximately 0.1-0.6 cm in diameter, with central clearing. The papules had a reddish-light brown surface and a firm texture with distinct boundaries. There was no history of fever, pain, or drug intake. Histopathologic findings showed a normoacanthosic epidermis with orthokeratotic lamellar hyperkeratosis. The dermis was discretely fibrous, with a predominantly mononuclear, histiocytic, reticular inflammatory infiltrate around the locally altered collagen fibers. Hence, the patient was diagnosed to have an annular interstitial pattern of GGA based on clinical and histopathological findings. After a skin biopsy, the lesions healed completely spontaneously. No treatment was given, and the patient was advised to return regularly for follow-up.

Discussion:

GA is a benign, relatively common cutaneous disease that classically presents as papules, plaques, and nodules on the extremities with skin-colored, pink, or violaceous hues. It is often symmetrical, and lesions are typically asymptomatic.** The diagnosis depends on clinical and histologic features. There are four main variants of GA: localized GA, subcutaneous GA, generalized GA, and perforating GA. Children most commonly present with localized and subcutaneous forms. In the review by Patrizi et al., the authors concluded that multiple LGA is the most common variant of GA, and GGA has been rarely reported in children. GGA is defined by the presence of more than 10 lesions and truncal involvement. It was associated with pruritus in less than half of cases. . A typical sparing of palms and plants has also been reported.

GA is a palisading granulomatous dermatitis involving the mid and upper dermis, with normal epidermis. Histology is characterized by a central core of fragmented dermal collagen bundles surrounded by an infiltrate of fibroblasts, lymphocytes, and histiocytes arranged in a palisading fashion. Although GA is not uncommon, its etiology, presentation, and treatment are not well-defined. Steroids are often the most commonly prescribed

treatment. While localized GA tends to be self-limited, other subtypes, particularly generalized GA, may have high recurrence rates. Systemic treatment may be required for generalized GA, which is often resistant to treatment, but our patient had a spontaneous resolution after a skin biopsy.

Conclusion:

GA is a self-limiting, granulomatous inflammation. The etiology and pathogenesis of this condition are unknown, and it can present in various forms.

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Abstract N°: 7082

Neonatal pemphigus a bullous dermatosis of infants

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Introduction & Objectives:

Neonatal pemphigus is an autoimmune bullous dermatosis caused by the transfer of maternal IgG across the placenta when the mother develops pemphigus during pregnancy.

The clinical manifestations of this condition are transient and less severe than those of pemphigus.

We report a case of neonatal pemphigus in a newborn.

The aim is to shed light on this bullous pediatric pathology.

Materials & Methods:

A premature female newborn from a closely monitored delivery, born to a 35-year-old mother who had been treated for 2 years for histologically confirmed pemphigus vulgaris under oral corticosteroid therapy in combination with azathioprine, who had also received rituximab treatment which was discontinued following her pregnancy was admitted to the neonatal intensive care unit immediately post-partum following a caesarean delivery.

Her examination on admission revealed multiple bullous and post-bullous lesions located mainly in the perioral and genital areas, associated with a few satellite lesions. The examination revealed multiple pseudo-urticarial lesions with bullous evolution.

The evolution was marked by a spontaneous and progressive regression of the bullous lesions after three weeks.

Results:

Given the maternal history and the spontaneous evolution towards regression, the diagnosis of neonatal pemphigus was established.

Neonatal pemphigus occurs in 30-45% of children born to affected mothers, due to the transfer of maternal antibodies from the placenta to the fetus.

Although neonatal pemphigus is transient, has a benign course and is not associated with the development of future diseases, it can lead to misdiagnosis.

Indeed, several other bullous dermatoses may be present in the neonatal period, all of which differ considerably in terms of etiology, course and treatment, hence the importance of being aware of this entity, which does not require specific treatment or skin biopsy.

Conclusion:

In conclusion, neonatal pemphigus is a benign pathology, with a favorable evolution in the absence of treatment. It should be noted, however, that although a biopsy officially determines the disease, in this case it was not necessary, due to the maternal history and satisfactory clinical evolution during the patient’s period of clinical

observation in hospital.

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Abstract N°: 7094

Ichthyosis prematurity syndrome: a case report.

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Introduction & Objectives:

Ichthyosis prematurity syndrome, or congenital ichthyosis type 4, is a rare autosomal recessive genodermatosis characterized by the triad of prematurity, thick caseous desquamating epidermis, and perinatal asphyxia. It is associated with mutations in the *SLC27A4* gene. The objective is to raise awareness of this rare syndrome despite its distinctive features as we believe it is still underdiagnosed.

Materials & Methods:

Here, we describe a prematurely born baby patient (35 weeks of gestation) diagnosed with Ichthyosis prematurity syndrome.

Results:

A female neonate, delivered at 35 weeks of gestation from a consanguineous union. The birth weight was 2400 g. There was a history of polyhydramnios. The newborn exhibited a state of apparent death with an APGAR score of 3, necessitating respiratory support. Dermatological examination revealed multiple thick verruciform hyperkeratotic plaques with a cobblestone appearance, covering all his body, more prominent over the forehead and scalp. Hematological analysis indicated hypereosinophilia. However, due to resource constraints, mutation analysis of the SLC27A4 gene couldn't be performed. Sterile petroleum jelly therapy was initiated but unfortunately, the patient expired within 24 hours. Genetic counseling was offered to the mother for future pregnancies.

Conclusion:

We report a new case of Ichthyosis prematurity syndrome to emphasize its stereotypical clinical presentation at birth. Indeed, its immediate recognition allows for understanding the specific needs of this genodermatosis, which can impact the newborn's prognosis.

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Abstract N°: 7206

Chronic, non-healing pustular lesion in two pediatric cases: An unexpected diagnosis: Preauricular sinus

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Introduction & Objectives:

Preauricular sinuses are quite common congenital external ear anomaly. They are small pits located superior to the tragus and anterior to the helix. They usually occur due to the incomplete fusion of tubercles with each other and do not show gender predilection. They usually occur in antenatal period and are present at birth. They may be bilateral in 25% to 50% of cases. Bilateral preauricular sinuses are more likely to be familial and associated with renal anomalies. They are mostly asymptomatic unless infected.

Materials & Methods:

There is not much data in the literature about preauricular sinuses and most symptomatic patients don't seek medical attention. Therefore, most physicians may be unaware of them.

Results

We report 2 cases of preauricular sinus infection that misdiagnosed as chronic bacterial cutaneous infection. In the first case, a 7-year-old boy with no known medical history presented with right ear pain, discharge and swelling and redness of the preauricular region for seven months. He denied having fever, headaches. On dermatological examination, there was a tender and draining swelling anterior to preauricular sinus (Fig. 1). In the second case, a 5-year-old girl with a history of left ear pain presented with redness on the preauricular region. Her sister had Wilms tumor history and her father had bilateral preauricular sinuses. Dermatological examination revealed a tender redness anterior to preauricular sinus on left preauricular area (Fig. 2) and asymptomatic pit on right preauricular area. Both patients don't have any systemic complaints and or findings. Exudates were cultured and both patients were treated with systemic and topical antibiotics. Laboratory studies demonstrated no abnormalities. Due to recurrent infections, both patients were consulted to the Otorhinolaryngology department for surgical excision. Audiological evaluation was performed to prevent and/or manage hearing loss for both patients. Since 11p15 locus could be associated with both preauricular sinus and Wilms tumor, genetic tests were performed for second case. She was also consulted to Pediatric Surgery department to investigate any possible renal anomaly.

Conclusion:

Preauricular sinuses mostly have a favorable prognosis and have no associated mortality. Morbidity is associated with recurrent infections, ulceration, scarring, pyoderma, and facial cellulitis. Surgical treatment may be curative in recurrent infections. However, recurrence risk is still possible. On the other hand, in patients with preauricular sinuses, espacially in bilateral involvement and famial cases, the possibility of renal abnormalities should be considered.

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Abstract N°: 7353

A rare case of linear porokeratosis in a child

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Introduction & Objectives: We report a case of a 4-year-old female child with linear porokeratosis on the left lower limb.

Materials & Methods: A four-year-old female child was reffered to a dermatologist due to multiple pruritic brownish keratotic plaques on the left lower limb that were present at birth. She was otherwise healthy and there were no signs of developmental problems. No similar lesions were present in other members of her family. Examination showed multiple plaques with hyperkeratotic, peripheral rims on anterior aspect of the left lower limb extending up to the dorsum of the left foot. The lesions were arranged along the Blaschko lines. The clinical appearance was most likely suggesting the linear epidermal nevus, linear lichen planus or linear psoriasis.

Results: Dermoscopic examination showed that the rim of the lesions was surrounded by white bands with a flaky white homogeneous structureless areas in the center of the lesions. Histopathological examination showed the characteristic cornoid lamella and absent granular layer below the invagination. Upper dermis showed mild chronic inflammatory cell infiltration The patient was treated with tretinoin cream (0.05%) twice daily and oral antihistaminic for two months with subsequent skin irritation.

Conclusion: Porokeratosis is a rare disorder of keratinization characterized by one or more atrophic macules or patches, surrounded by a distinctive hyperkeratotic, ridge-like border called a "cornoid lamella". The pathogenesis of porokeratosis is unknown and multiple clinical variants of porokeratosis exist. Linear porokeratosis is a rare form of porokeratosis with skin lesions that typically present during infancy or early childhood. Malignant transformation occurs in a minority of cases. Treatment guidelines of the therapeutic options for pediatric porokeratosis are lacking and various topical, surgical and destructive therapies can be effective in some patients.

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Abstract N°: 7398

The importance of thorough medical history in diagnosis of vesicular lesions in children

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Introduction & Objectives: In childhood, skin changes of infectious origin are a common occurrence, with the most common causes being bacteria or viruses. Impetigo is one of the most common skin infections in childhood caused by bacteria (Staphylococcus aureus, beta-hemolytic streptococcus group A). The face, especially perioral and perinasal areas, is the predilection site. The disease starts as small vesicles with a thin covering, filled with clear fluid on a slightly erythematous base, which quickly ruptures, leading to erosion and a honey-colored crust. Differential diagnosis should also consider HSV and VZV infections. In this case report, we will present the importance of taking a thorough medical history in diagnosis of vesicular changes on a child's face.

Materials & Methods: A 13-year-old female patient with unremarkable personal and family history presented to a dermatologist due to vesicular skin changes on her face. Initially mistaken for an HSV infection, the patient had been treated with acyclovir by the pediatrician, resulting in the spread of the lesions. After a detailed medical history was taken by the dermatologist, it was concluded that the diagnosis was impetigo. Local treatment with mupirocin and oral therapy with cephalexin for 7 days were recommended, leading to complete regression of the lesions.

Results: Dermatological examination revealed limited erythematous eroded lesions with yellowish crusts and occasional vesicles filled with yellowish secretions on the skin of the left corner of the mouth spreading to the mandible, left cheek, and below the lower lip. Similar lesions up to 1 cm in size were present on both eyelids and in the frontal area near the right eyebrow.

Conclusion: Special attention should be given to obtaining an accurate medical history due to the clinical similarity between impetigo and HSV infection. HSV infection usually starts with prodromal symptoms and commonly appears unilaterally, for example, on the lip in the vermilion area, while the disseminated form is present in children with immunodeficiency or underlying dermatosis. In our case, there were no prodromal symptoms, and the lesions appeared on different parts of the face over several days. If standard antiviral therapy does not lead to improvement in skin lesions, other common causes of skin infections should be considered

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Abstract N°: 7417

Giant congenital nevus: an exceptional picture

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Introduction & Objectives:

A congenital nevus is a benign abnormal accumulation of melanocytes in the skin at birth. The size and number of nevi can vary, from a single lesion to innumerable lesions, with or without large lesions.

Clinical case:

This case report illustrates a newborn with a giant congenital nevus. This newborn, seen at H6 of age, is male, from a non-consanguineous marriage, and from a 2nd full-term pregnancy. Since birth, he has presented with a pigmented plaque measuring 15cm on the anteroposterior surface of the right arm, extending to the forearm, interspersed with healthy areas, sometimes smooth and sometimes thick, with clear contours, associated with similar smaller lesions all over the body, affecting the oral and genital mucosa, as well as the dander. The rest of the examination was unremarkable.

Discussion:

Giant congenital nevus is a pigmented skin lesion larger than the palm of the hand. Its incidence is 1/20,000. Diagnosis is clinical. Its giant size increases the risk of degeneration into melanoma. It may also be complicated by leptomeningeal melanocytosis, or be associated with other neuro-ectodermal tumors, making regular clinical surveillance essential. Treatment options must be decided on a case-by-case basis, and include surgical techniques (surgical excision, serial excision, etc.) and alternatives to surgery (dermabrasion, curettage and laser).

Conclusion:

Giant congenital nevi are exceptional, with a poor prognosis and difficult therapeutic management. Iterative excisions are proposed in conjunction with skin expansion techniques. CO2 lasertherapy is a promising method that offers improved curative and aesthetic results.

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Abstract N°: 7433

Erythema infectiosum / slap cheek disease / “fifth disease”

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Introduction & Objectives:

Erythema infectiosum (slap cheek disease, “fifth disease”) is a common viral exanthem illness found in children.

Materials & Methods:

We describe 5 clinical cases selected from a large series aiming to draw the attention to the disease.

Results:

The cause of fifth disease is the human parvovirus -B19 infection. Transmission is primarily via droplets from respiratory secretions and can spread via blood exposure. It occurs in spring and early summer months.

Diagnostic is based on clinical features and rarely is needed lab investigations.

Treatment is supportive and symptom controlling.

Conclusion:

The disease is very often described in children 5 to 15 years old, although younger children or, even, adults can be affected. It is of outmost clinical importance to understand the pathogenesis of each entity, to make the clear distinction and to recommend an appropriate treatment.

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Abstract N°: 7456

Boy with warm-like scars on his face

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Introduction & Objectives: : Keratosis pilaris atrophicans faciei is a disease that belongs to a group of hereditary follicular disorders known as a very rare entity. It is also known as a clinical feature called atrophoderma vermicularis. The usual prevalence of the patients is children between 1-10 years old, but it is also seen in adolescents and infants. The disease itself is characterized by facial inflammatory keratinization. Pathophysiology of the disease explains the possible mechanism of the syndrome caused by abnormal keratinization which leads to follicle inflammation caused by scales that fill the hair follicles. There are cases connected to gene mutations such as autosomal dominant and recessive mutations in the desmoglein 4 gene. The natural progression of the disease is unpredictable which makes the diagnosis highly uncertain.

Materials & Methods: The diagnosis was made based on clinical features and the appearance of skin changes.

Results:

A 5-year-old patient came with his mother due to the appearance of unusual skin lesions on the cheek area. The mother noticed skin lesions a few weeks before the check-up at a pediatric dermatology practice. She denies any other skin changes or lesions elsewhere on the son's body. There wasn't any connection to stings or insect bites and well as no appearance of acne on the face before. Boy was otherwise healthy, but his mother was complaining about dry skin on the cheeks and on the body, especially during the winter, but without prodromal eczema. There wasn't an acne history in the family. Atrophic pinpoint changes were visible on both sides of the cheeks without any inflammatory changes or hyperpigmentation. Hair and eyebrows were neat.

Conclusion:

It is a chronic disease that can appear as a part of certain syndromes so it is necessary to rule out other possible causes. The disease has a progressive course, but skin changes usually disappear after puberty, however, alopecia is permanent. Our patient, due to his age of 5, has limited options for treatment. Topical agents used in therapy are facial cleanses, creams with a higher percentage of urea, and topical retinoids, but it is not recommended at this age. Sun protection is necessary. The child will remain further in the follow-up.

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Abstract N°: 7482

Parvovirus-induced papular-purpuric gloves and socks syndrome in a thalassemia patient: a case report

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Introduction & Objectives:

Papular-purpuric gloves-and-socks syndrome (PPGSS) classically presents as focal acral papular-purpuric eruptions of symmetrical distribution, and is usually caused by parvovirus 19 infection. However, atypical presentations have been reported, especially in patients with underlying conditions.

Patient & Observation:

A 9-year old boy, with a background of thalassemia and asthma, presented with a 48 hour rash prevailing on the limbs, associated with fever and sore throat, and mild arthralgia. Examination found a stable patient, with symmetrical purpuric maculopapular lesions on the extremities, the flexor surfaces of the limbs, particularly the knees and elbows, and the anterior surface of the neck. No abdominal symptoms or lymphadenopathy were found. Blood count, hemostasis and creatinine tests did not show any abnormalities aside from mild anemia. The patient was given symptomatic treatment with good evolution. Parvovirus immunoglobulin M serology was positive.

Discussion:

Parvovirus B19 infection is a viral illness that can manifest in a variety of ways, ranging from asymptomatic to symptomatic depending on the patient's health.

Among its clinical signs, the most common are dermatologic and include erythema infectiosum ("slapped cheek" disease), papular-purpuric gloves-and-socks syndrome, Gianotti-Crosti syndrome, etc. However, other manifestations such as rheumatologic and hematologic are not uncommon, particularly in patients with underlying hematological abnormalities, in whom they can lead to aplastic cellular crises.

PPGSS most commonly manifests in adolescents and adults, and presents as an acute symmetrical eruption of petechiae and small purpuric papules affecting hands and feet in a gloves-and-socks distribution; often associated with pruritus, pain, erythema, and edema. However, this distinctive feature is not always essential for diagnosis, as intertriginous involvement, a more generalized purpuric-petechial eruption and/or truncal distribution are atypical presentations, often encountered in patients with hematological abnormalities such as thalassemia.

Fever, arthralgia, and generalized lymphadenopathy, which suggest an underlying viral etiology, are common, but differential diagnoses to consider include hand-foot-mouth disease, erythema multiforme, and vasculitis, commonly Henoch–Schonlein purpura.

Diagnosis confirmation through laboratory tests to detect immunoglobulin M (IgM) is not systematic, but may be required in patients with systemic manifestations or negative evolution, as PPGSS is usually self-limiting and clears spontaneously within 1 to 2 weeks with no long-term complications. Treatment should be symptomatic, and oral corticosteroids should be avoided.

Conclusion:

This case sheds light on atypical presentations of parvovirus B19 infection, especially in patients with underlying conditions, and on the importance of early recognition in order to avoid invasive biopsies and systemic treatments for a disease with usual favorable outcome.

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Abstract N°: 7517

Neonatal complicated cytosteatonecrosis: A case report.

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Introduction & Objectives:

Neonatal cytocsteatonecrosis is an acute hypodermatitis characterized by purplish-red nodules, more or less well-defined, present from birth in infants with complicated delivery. In the majority of cases, these lesions are limited and benign, but a few cases have been associated with hypercalcemia, which can lead to cardiac, neurological, renal complications, or even death.

It typically develops after a free interval within the first 15 days following delivery but may appear up to 30 days after birth. Lesions begin with erythema that rapidly progresses to purplish-red hypodermic plaques. These lesions are often painful. Preferred locations include the back, neck, and upper limbs.

Factors predisposing our patient to this condition were maternal gestational diabetes and neonatal distress. Diagnosis is usually clinical, and if there is doubt, a skin biopsy confirms the diagnosis.

Hypercalcemia is the most common complication, with secondary visceral calcium deposits frequently reported. The more frequent complications are nephrocalcinosis and renal calculi.

Treatment of neonatal cytocsteatonecrosis is not standardized and is primarily symptomatic. Conventional treatment for severe hypercalcemia involves hydration with isotonic saline, hypercalciuric diuretics, a low-calcium, low-vitamin D diet, glucocorticoids, and, in severe cases, calcitonin, bisphosphonates or ketoconazole.

Materials & Methods:

We report a case of complicated neonatal cytosteatonecrosis in a 2 month old infant.

Results:

Female infant, 2 months old, born from a well-monitored pregnancy complicated by gestational diabetes. Maternal age was 30 years, full-term vaginal delivery, birth weight 4.11 kg with a history of neonatal distress. The infant was admitted to the pediatric department for acute dehydration due to vomiting, with a weight loss of 1 kg, along with diffuse skin lesions appearing 20 days after birth.

General examination revealed a stable infant in terms of hemodynamics and respiration, afebrile, and dehydrated. Dermatological examination revealed hardened erythematous-purple nodules and plaques on the back, neck, buttock and thighs, with the presence of an ulceration and clear fluid discharge on the left thigh.

The diagnosis of cytosteatonecrosis was made. Laboratory tests revealed hypercalcemia at 170.14 mg/l, renal ultrasound found grade 3 nephrocalcinosis with the presence of a mass in the right adrenal compartment, and soft tissue ultrasound showed subcutaneous collections in the buttock regions. An abdominal scan revealed a calcified hematoma in the right adrenal compartment.

Management included correction of hypercalcemia through hydration, diuretics, stopping supplementation of calcium and vitamin D, and Cortancyl 5 mg/day.

Conclusion:

Neonatal cytosteatonecrosis is a well-described but rarely encountered condition in routine practice. It typically follows a benign course.

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Abstract N°: 7521

Sweet syndrome in an adolescent

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Introduction & Objectives:

Sweet syndrome (dermatosis acuta febrilis neutrophilica) is a neutrophilic dermatosis, more common in women. It is characterized by sudden erythematous and infiltrative skin lesions, accompanied by fever and peripheral blood neutrophilia. The lesions are most commonly located on the trunk, upper limbs and face. Sweet syndrome is considered a paraneoplastic syndrome as it can sometimes be associated with lymphoproliferative proliferations.

Materials & Methods:

We present the case of a 12-year-old girl who developed numerous pustular eruptions, papules and nodules on the lateral trunk bilaterally and in the armpit area.

Results:

Histopathological examination of the lesions confirmed the classic form of Sweet syndrome, and 6 months after the first lesion appeared on the skin, a diagnosis of acute myeloid leukaemia was made on the basis of a bone marrow biopsy.

Conclusion:

As Sweet's syndrome may be the first manifestation of the malignancy, the importance of appropriately prompt diagnosis and treatment is of immense importance to increase the patient's survival rate.

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Abstract N°: 7555

An unusual case of cervical cystic lymphangioma

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Introduction & Objectives:

Cervical cystic lymphangiomas are rare and benign. However, they can lead to significant complications, including aesthetic, phonatory, or swallowing issues, and can also be life-threatening due to airway compression or severe bleeding. Here, we report a case of an atypical cystic lymphangioma of the neck successfully treated with surgery and sirolimus.

Materials & Methods:

Results:

A 3-week-old female newborn, born full term from consanguineous parents, presented for consultation with a lateral cervical mass present since birth. General examination revealed a febrile newborn with a temperature of 38.7°C and a heart rate of 155 bpm. Dermatological examination revealed a left lateral cervical swelling measuring 10 cm in diameter with central ulceration and pus discharge, as well as a subcutaneous swelling on the right parietal scalp. Laboratory tests showed leukocytosis at 14,640 with neutrophilia at 10,180, anemia at 9.8, and thrombocytopenia at 32,000. Cervical CT scan revealed a cystic left lateral cervical mass measuring 11,5*7cm extending into the carotid and pre-vertebral fat spaces, containing homogeneous liquid content traversed by incomplete fine septa, consistent with a cystic lymphangioma. Cerebral CT scan revealed an extracranial collection beneath the right parietal periosteum consistent with a cephalohematoma. The patient was treated with antibiotics with good clinical and laboratory improvement. After controlling the infection, she underwent surgical excision with no postoperative complications, and received sirolimus at a dose of 0.8 mg/m2 per dose, twice daily with good progress. Histopathological examination showed lymphatic vascular ectasia with an inflammatory infiltrate rich in plasma cells and lymphocytes with micro-abscesses.

Conclusion:

Cystic lymphangioma is a congenital, histologically benign, lobulated abnormality originating from the lymphatic system. Cervical lymphangiomas represent a very severe condition, not only due to possible airway compression or massive bleeding but also because of the severe sequelae they may cause. Radiological imaging techniques such as CT and MRI assist in diagnosis and provide insight into the extent of the condition. However, the definitive diagnosis is confirmed through histopathological examination. Surgical excision remains the first-line treatment. However, sirolimus may offer an effective therapeutic approach for children with large complicated lymphatic malformations affecting the head and neck, especially in cases when complete surgical resection is not possible.

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Abstract N°: 7650

Experience in the use of reflectance confocal microscopy in an uncontrolled setting in pediatric dermatological patients: Prospective study of 47 patients at a University Hospital

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Introduction & Objectives:

Reflectance confocal microscopy (RCM) corresponds to a non-invasive imaging modality for diagnostic support, allowing in vivo quasi-histological resolution images of different cutaneous pathologies. It is a well-tolerated tool in the pediatric population and would lead to a decrease in the number of biopsies, thus reducing the costs and complications associated with sedation and general anesthesia in these patients. The objective of the following study is to describe the results and utility of RCM in the pediatric population with suspicious skin lesions.

Materials & Methods:

Uncontrolled prospective study of consecutive cases conducted between February 2021 and January 2024 at a university hospital. Data were extracted from patients' medical records, RCM reports, and pathological anatomy reports from the same clinical center. Patients under 18 years of age were considered pediatric patients. The location of the lesions studied was described based on Delphi dermatological anatomical consensus. The referral clinical diagnosis considered for statistical analysis was the one with the worst prognosis implicated. In the case of melanocytic lesions diagnosed by histopathology, an additional category, MPATH-Dx Version 2.0, was incorporated for subgroup analysis. Statistical analysis was performed using Microsoft Excel and RStudio software.

Results:

A total of 47 lesions in 41 patients were studied, with 65.9% (27) female and 34.1% (14) male. The mean age was 12.04 years (range 0-17 years). The most frequent locations of the studied lesions were the cheek (14.9%), upper back (14.9%), and thigh (12.8%). The referral clinical diagnoses were melanoma (97.9%; 46 lesions) and basal cell carcinoma (2.1%; 1 lesion). Out of the total lesions clinically considered melanomas, 12 were referred with an alternative diagnosis of spitzoid lesion. A clinical-RCM discordance of 89.36% was obtained with 87.23% potentially saved biopsies, significantly higher than observed in the adult population. Of the total biopsied lesions (8; 17.5%), the RCM-histopathological correlation was 50%, and when analyzing melanocytic lesions by MPATH-Dx version 2.0, it was 71.42%. Out of the total lesions without biopsy indication according to RCM, 2 were followed up with a new RCM without changes, and 4 biopsies were performed for a different indication, all with benign results.

Conclusion:

The percentage of potentially saved biopsies is significantly higher in the pediatric population, which establishes RCM as a valuable tool in this population in daily dermatological practice.

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Atypical Adams-Oliver syndrome associated to ciliary dyskinesia

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Introduction & Objectives: Aplasia cutis congenita is characterized by the absence or scarring of the skin present since birth. It can be isolated or associated with systemic comorbidities. Due to the possibility of systemic diagnoses, it is important to promptly assess the presence of other comorbidities and carry out a genotype-phenotype correlation, seeking greater accuracy in monitoring patients, reducing morbidity and mortality.

Materials & Methods: Case report.

Results: A 13-year-old male patient, children of consanguineous parents of Japanese origin, with ciliary dyskinesia due to two pathogenic variants of DNAH5 in homozygosity c.9365del(p.Leu3122*), presented with a congenital 4x3cm atrophic plaque in the right lumbar region adjacent to a light brown macule appearing on the straight from the plate and extends for 15cm. Due to the hypothesis of aplasia cutis, neuraxial MRI were performed, which were normal. Cardiovascular images showed heterotaxy syndrome compatible with left atrial isomerism; dilation of the azygos/hemiazygos system, with the infrahepatic inferior vena cava not being identified; inferior vena cava not visible, which may be related to a developmental anomaly. Research was carried out for specific mutations related to aplasia cutis. Patient presents homozygous DOCK6 mutation c.3800C>T(p.Ala1267Val). NF1, KCTD1, KRAS, HRAS, EOGT, DLL4, NOTCH1, RBP5, ARHGAP31 were negative.

Conclusion: Adams-Oliver syndrome presents with malformations of the central nervous system, limb defects, telangiectatic cutis marmorata and cardiovascular malformations. Aplasia cutis is usually located on the scalp. The case presented shows aplasia cutis in the lumbar region, not centered and associated with a mutation related to Adams-Oliver syndrome. In addition to ciliary dyskinesia due to the DNAH5 mutation located at 5p15.2, the patient has other vascular malformations not usually associated with ciliary dyskinesia. The case is suggestive of an association of Adams-Oliver syndrome or variation in presentation that may be due to a specific genotype-phenotic relationship due to a mutation in the DOCK6 gene at 19p13.2.