Clinical Cases in Dermatology
Series Editor: Robert A. Norman

Torello M. Lotti
Fabio Arcangeli *Editors*

Clinical Cases in Middle-Years Pediatric Dermatology

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Series Editor
Robert A. Norman
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Clinical Cases in Middle-Years Pediatric Dermatology

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Contents

Chapter 1
A 10-year-old Patient with Erosive Stomatitis, Conjunctival Hyperaemia and Fever

Alfonso Delgado Rubio and Fabio Arcangeli

A 10-year-old boy came to our observation because he had a high fever for two days and erosive-necrotic lesions on his lips. He also had edema of the eyelids and conjunctival hyperaemia (Fig. 1.1) for 4 days. There was erosive stomatitis (Fig. 1.2), round cutanous erythematous lesions mainly on the palms of the hands (Fig. 1.3) and on the soles of the feet too. On physical examination, we found a slight erythema on the genital mucouses.

He was treated with Aciclovir and antibiotic eye drops without any improvement.

His personal history was negative except an herpetic stomatitis that occurred a year earlier.

Laboratory tests and chest x-rays were normal. No viruses were isolated from oral secretions and serological investigations for EBV, CMV and other viruses were negative.

Based on the history and the photographs which diagnosis would you propose?

1. Kawasaki disease
2. Staphylococcal scalded skin syndrome (SSSS)
3. Erythema multiforme (EM)
4. Stevens-Johnson syndrome (SJS)

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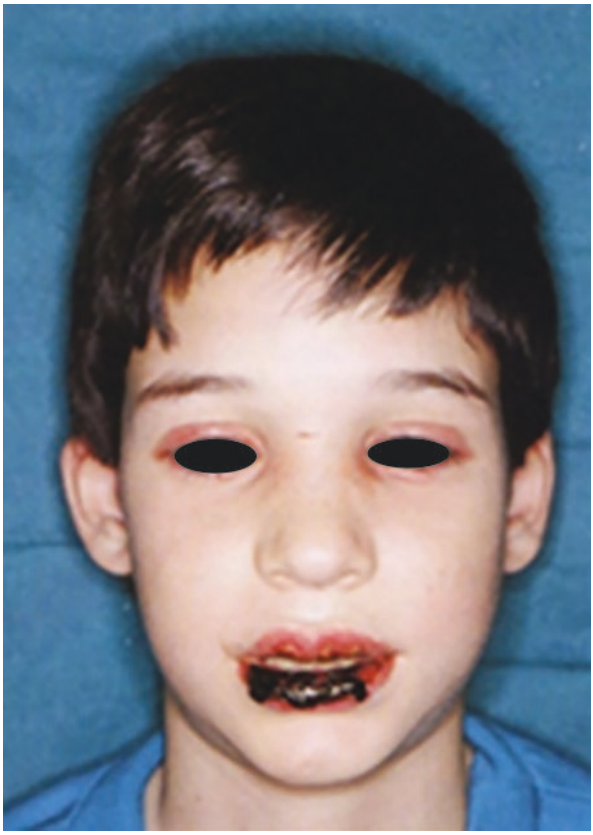

Fig. 1.1 Necrotic lesions on the lower lip and erythema on the eyelids

Diagnosis

Stevens-Johnson syndrome (SJS).

Discussion

Kawasaki Disease was ruled out because of the absence of diagnostic criteria such as a fever lasting five or more days, accompanied by four (typical Kawasaki) or two/three (atypical Kawasaki) out of the classical five findings. As it's knowhn these are bilateral conjunctival injection, oral changes (cracked and erythematous lips, strawberry tongue), cervical lymphadenopathy, extremity changes (erythema or palm and sole desquamation) and polymorphous rash.

Staphylococcal scalded skin syndrome (SSSS), due to an staphylococcal exfoliative toxin usually doesn't involve oral, conjuntival or genital mucosa and has superficial epidermal peeling.

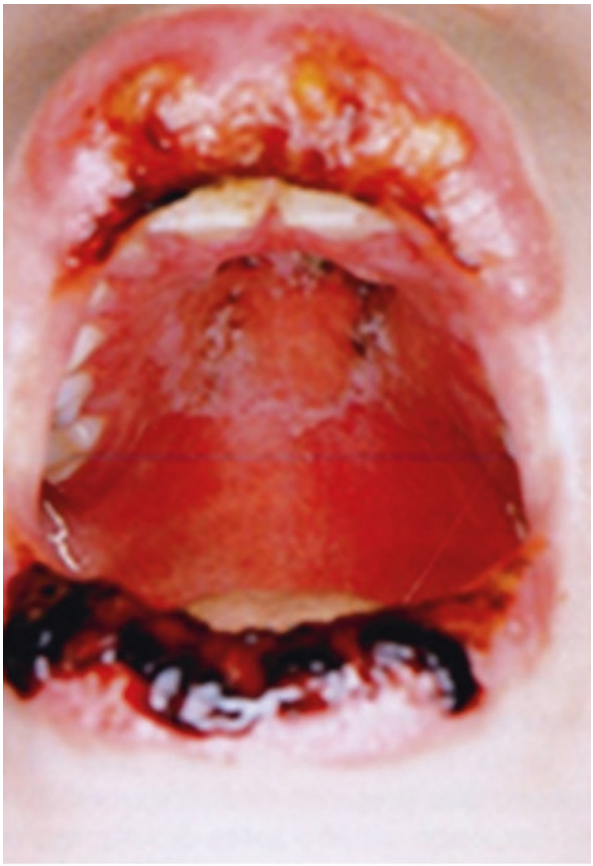

Fig. 1.2 Erosive stomatitis

Erythema multiforme (EM), caused more frequently by infectious agents (herpes and mycoplasma), can be excluded both in the minor and major varieties. In minor EM the mucous membranes are spared, in the major EM which, although it can involve the mucous membranes, similarly to SJS, it presents "typical target" lesions (cockade) on the skin (Fig. 1.4). In our case the cutaneous lesions had a so called "atypical target" representation as in every case of SJS (Fig. 1.5) [1]. Furthermore in EM systemic features are milder or absent and recurrences are more frequent [2].

Stevens-Johnson syndrome is an hypersensitive reaction that typically involves the skin and the mucous membranes. It is a minor form of toxic epidermal necrolysis (TEN), in which less than 10% of the body surface area (BSA) is detached. SJS is idiopathic in 25-50% of cases. Drugs (antibiotics, analgesics, NSAIDs, psychoepileptics etc) are most often implicated as the ethiology [3]. There is strong evidence for a genetic predisposition in cases of severe cutaneous adverse drug reactions [1, 4].

Typical prodromal symptoms—such as cough, headache, malaise, arthralgia—usually occur before the onset of the rash. The cutaneous lesions can begin as

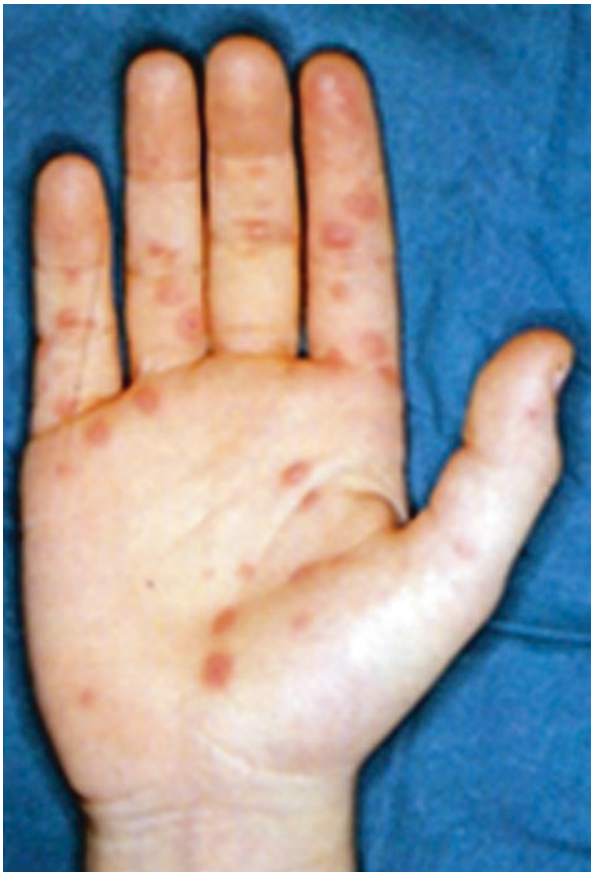

Fig. 1.3 Erythematous rounded lesions on the palm

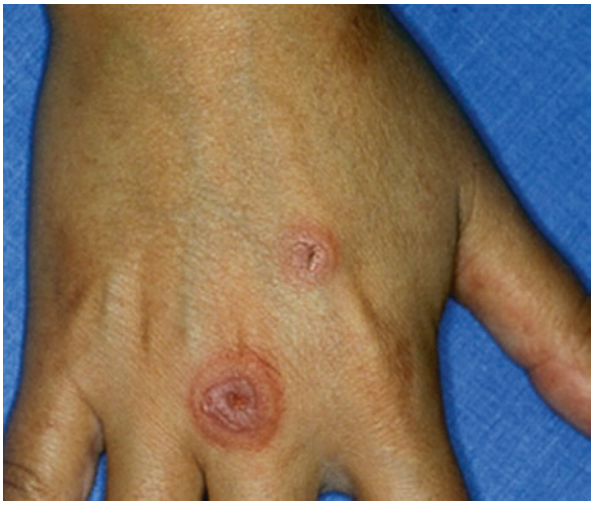

Fig. 1.4 Example of "typical target" lesion (cockade) with three concentric colour zones: (1) a darker centre with a blister or crust (2) a ring around this that is paler pink and raised due to edema (3) a bright red outermost ring

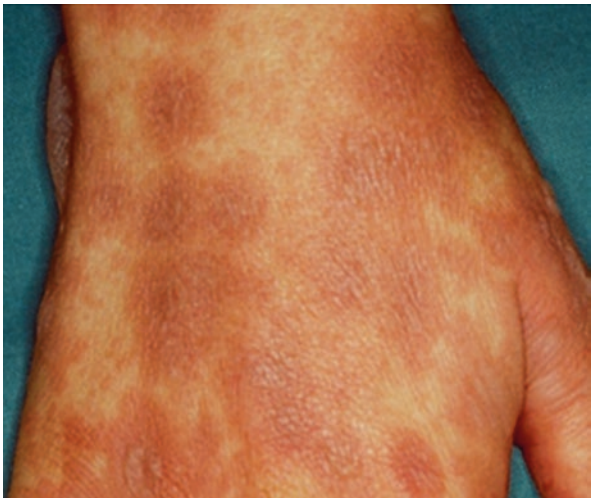

Fig. 1.5 Example of "atypical target" lesion with only two concentric colour zones: (1) a dark central zone of epidermolysis (2) a red outer area with faded borders

macules that develop into vesicles or bullae. The lesions have the appearance of a target but in contrast to the typical cockade shape of erythema multiforme, they only have two areas of colour. The lesions may become bullous and later rupture, leaving denuded skin. Shearing pressure on the involved erythematous skin may cause epidermal detachment (Nikolsky's sign) [5]. Peri-lesional erythema is a sign of disease activity and helps monitor treatment response [1]. They may occur anywhere, althought the palms, soles, dorsum of the hands, and extensor surfaces are most commonly affected. In addition to the skin, lesions in SJS may involve oral, conjuntival and genital mucosae. Fever is very common.

Delineation of a drug exposure timeline is essential, especially in the 1–3 weeks preceding the cutaneous eruption, because the withdrawal of the suspected offending agent is imperative to prevent possible evolution in TEN) (more than 30% of the body surface area detached) [1, 4, 6].

Stevens-Johnson syndrome and toxic epidermal necrolysis are potentially life-threatening diseases. The management of SJS/TEN should be undertaken in specialized centers such as dermatology departments or burn units, which has been shown to improve outcomes [7, 8]. Acute active management is controversial because of the lack of high-level evidence that any treatment is superior to supportive care alone [7].

However, the most common theraphy is oral or parenteral corticosteroid (prednisolone 1–2 mg/kg/day or equivalent), tapered rapidly within 7–10 days. Newer treatments such as cyclosporine (3–5 mg/kg/day) for 10–14 days, alone or in combination with corticosteroids [1, 4] and etanercept have shown promise [7].

Key Points

- SJS should be considered in all patients with "atypical target" lesions and mucous membrane involvement

- SJS is most frequently caused by drugs and if the offending drug is not discontinued it can evolve into more severe TEN
- The clinical distinction between SJS and EM is not based on the involvement or not of the mucous membranes but on the representation of skin lesions, “atypical target” in the case of SJS, “typical target” (cockade) in the ecase of EM

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Chapter 2
A Child with a Pruriginous Symmetrical Rash

Alina Suru, Alexandra Denisa Oprea, and Carmen Maria Salavastru

Short Clinical Story of the Patient

A 6-year-old female was referred to the Dermatology clinic for an erythematous, intensely pruritic rash consisting of excoriated papules symmetrically distributed on the cheeks (Fig. 2.1) and the extensor surface of the knees and elbows (Fig. 2.2). Other skin findings were represented by telangiectasia on her cheeks and the posterior upper trunk. The lesions evolved during the last six months and at onset had an urticarial and eczematous character affecting the extensor surfaces of the knees and elbows, but also the folds (axilla and popliteal fossa).

Considering the relatively long course of pruritic skin disease with little or no response to topical treatments, the suspicion of an autoimmune disease was raised. The necessity of a skin biopsy was explained to the parents; as they were reluctant to this maneuver, only laboratory tests were performed.

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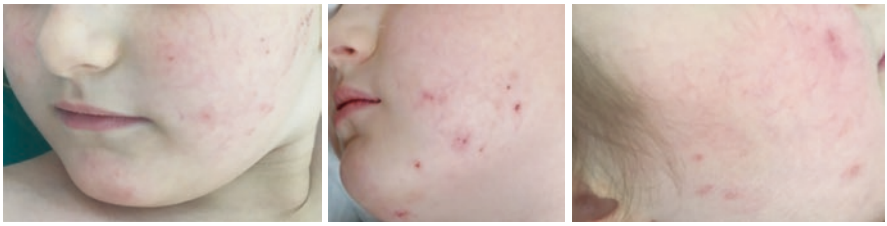

Fig. 2.1 Escoriated papules and telangiectasia of the cheeks

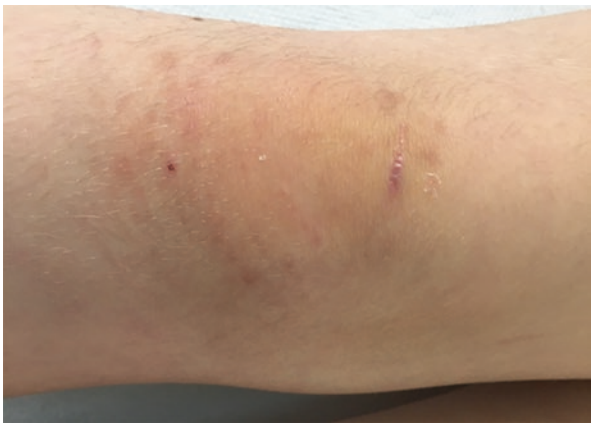

Fig. 2.2 Erythematous, crusted and hyperpigmented papules on the knees

In the serum, the presence of immunoglobulin G antigliadin (106 U/mL; reference, <12), IgA anti–tissue transglutaminase (>300 U/mL; reference, <12) antibodies was detected.

Other laboratory findings were eosinophilia, hyperfibrinogenemia and elevated antinuclear antibody levels. Therefore, an Extractable Nuclear Antigen Antibodies panel was performed and the high anti-DFS70 antibody titer was noted. The serum creatine kinase, aldolase, lactic acid dehydrogenase, and transaminase levels were within the normal range.

The patient had a BMI of 23 and denied any gastrointestinal symptoms.

The personal medical history noted recurrent herpes simplex virus infections and granuloma annulare at the age of five.

The family medical history mentioned a maternal great-grandmother with lupus erythematosus, but no medical documents were provided in this regard.

Based on the Case Description and the Photographs, What Is the Diagnosis?

1. Dermatitis herpetiformis
2. Atopic dermatitis
3. Amyopathic dermatomyositis

Diagnosis

Dermatitis herpetiformis (DH).
Celiac disease (CD).

Discussion

Childhood DH is rare and can present with atypical lesions including eczematous lesions and facial lesions. The incidence and prevalence are uncertain, but cases have been reported in children as young as 8 months; most children receive the diagnosis between the ages of 2 and 7 years [1].

Atopic dermatitis (AD) is valuable to mention in the differential diagnosis of this patient because of the polymorphic lesions with initial localization in the folds and of the initial laboratory tests (eosinophilia). Patients with an extensor distribution of eczema in later childhood are uncommon and may take longer to remit. AD is frequent in children; the most characteristically involved sites are the elbow and knee flexures, a mixture of erythema, crusting, excoriation, and hyper/hypopigmentation. The erythematous and edematous papules tend to be replaced by lichenification, but true eczematous lesions with vesiculation may occur [2].

Associations between DH and other autoimmune diseases must be kept in mind at diagnosis and during the follow-up of patients with DH [3]. In this case, clinically amyopathic dermatomyositis (CADM) was important in the differential diagnosis because of the positive family history, clinical features (telangiectasia on her cheeks and the upper third part of the posterior trunk), elevated titer of antibodies (anti-DFS70 antibody, antinuclear antibodies). Nailfold capillaroscopy was unremarkable. CADM is an autoimmune disease characterized by the presence of cutaneous manifestations of DM without muscle weakness and with normal serum muscle enzymes. The cutaneous lesions of dermatomyositis are often intensely pruritic, prominent on extensor surfaces, including the elbows, knees, metacarpophalangeal joints, and both the proximal and distal interphalangeal joints. A very characteristic feature of dermatomyositis is photo distributed poikiloderma, often involving the upper chest (V-neck sign) and the upper back (shawl sign) [4].

The pathophysiology in DH and GSE results from an IgA dominant autoimmune response to transglutaminase (TG) molecules. IgA/TG3 immune complexes are formed locally and are deposited within the papillary dermis resulting in the infiltration of activated neutrophils from the circulation. Degranulation of neutrophils releases proteases which cleave the lamina lucida and create a subepidermal blister [5]. Because the condition is so pruriginous, intact vesicles are rarely seen and the patient may simply present with excoriations. Several antibodies have been described in DH and CD: endomysial antibodies (EMA), deamidated gliadin peptide (DGP) antibodies and transglutaminase antibodies (TGA-IgA).

Recent guidelines for the diagnosis of CD accept a no-biopsy approach for children if TGA-IgA titer is more than 10 times the upper limit of normal and the EMA-IgA antibodies are positive [6].

Since both the skin disease and the intestinal disease resolve with strict adherence to a gluten-free diet and recur with return to a regular diet, it is proved that the dietary protein gluten is the cornerstone to the pathogenesis of the skin lesions [3]. Although over 90% of DH patients have evidence of GSE, only about 20% have gastrointestinal symptoms of CD [7].

Multiple studies have revealed an association between dermatitis herpetiformis and certain HLA types, particularly HLA DQ2 and HLA DQ8. A negative test for HLA-DQ2 and/or -DQ8 indicates that CD is unlikely, while a positive result does not confirm the diagnosis [6].

Based on the patient's medical history, clinical picture and laboratory test results, the diagnosis of dermatitis herpetiformis was established. Direct immunofluorescent skin biopsy should have been obtained and would have demonstrate IgA deposition along the dermoepidermal junction with concentration at the papillary tips. Since the parents were reluctant to this medical maneuver, the authors based their diagnosis on the clinical aspect and high titer of specific antibodies.

The patient was referred to the pediatric gastroenterology department, where according to the ESPGHAN 2020 guidelines, she was diagnosed with celiac disease with predominant cutaneous involvement.

Historically weight loss is a classic symptom of CD, but recent studies in adults and children indicate that obesity/overweight at disease onset is not unusual [8, 9].

The patient was instructed to follow a strict gluten-free diet. After three months of diet, the cutaneous signs and symptoms improved, with the alleviation of the pruritus and decrease of the papular rash. Considering this favorable response to the diet, the introduction of Dapsone was timed.

Key Points

- Childhood dermatitis herpetiformis is rare and may associate atypical presentation and facial involvement; the symmetrical pruritic rash is often distributed on the extensor surface of the elbows and knees.
- Dermatitis herpetiformis is a chronic autoimmune blistering disease closely associated with gluten-sensitive enteropathy
- Various laboratory tests may be very helpful in cases where skin biopsy may not be an option
- Dermatitis herpetiformis may increase the risk for autoimmune diseases and lymphoma, so accurate diagnosis and treatment are essential.

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Chapter 3
A Pediatric Patient with Photosensitivity

Neloska Lence, Tusheva Ivana, Filipovic Dejan, and Damevska Katerina

A 6-year-old boy presented to our clinic with pruritic, erythematous skin lesions on his face precipitated by the sun. The patient had similar prior episodes during spring and summer, involving sun-exposed areas that have been treated as contact dermatitis with topical corticosteroids. He was not taking any medications and had no known drug allergies. The parents denied a history of photosensitivity in other family members.

Physical examination revealed numerous symmetrically scattered tense vesicles and blisters on an inflamed base. Lesions involve both cheeks, helices of the ears, nose, lower labia, and the posterior neck (Figs. 3.1, 3.2, and 3.3). Mucous membranes and nails were spared, and Nikolsky's sign was negative. Routine laboratory findings were within normal limits. Testing for porphyrins in his urine and blood during skin eruptions were negative. Hepatitis B virus, Epstein-Barr virus (EBV), and cytomegalovirus (CMV) testing were all negative. Antinuclear antibodies and double-stranded DNA were also negative. The vesicular eruption resolved gradually within two weeks, leaving depressed scars (Fig. 3.4).

Based on the Case Description and the Photograph, What Is Your Diagnosis?

1. Infantile systemic lupus erythematosus
2. Polymorphous light eruption
3. Actinic prurigo

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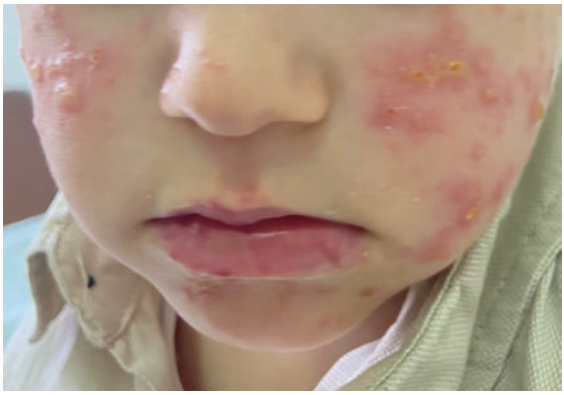

Fig. 3.1 Active lesions in various stages over the central part of the face

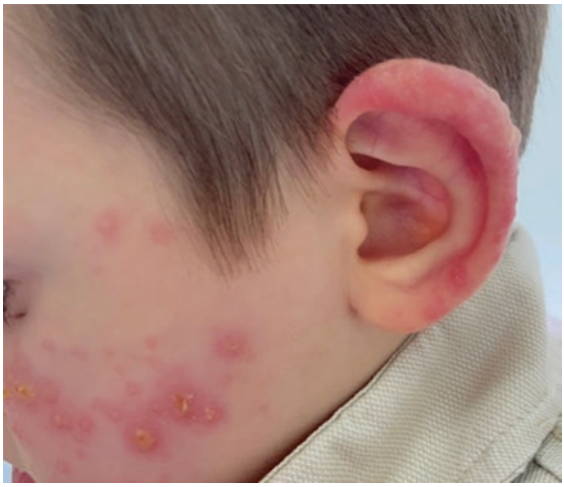

Fig. 3.2 Diffuse erythema and small vesicles on the auricular helix

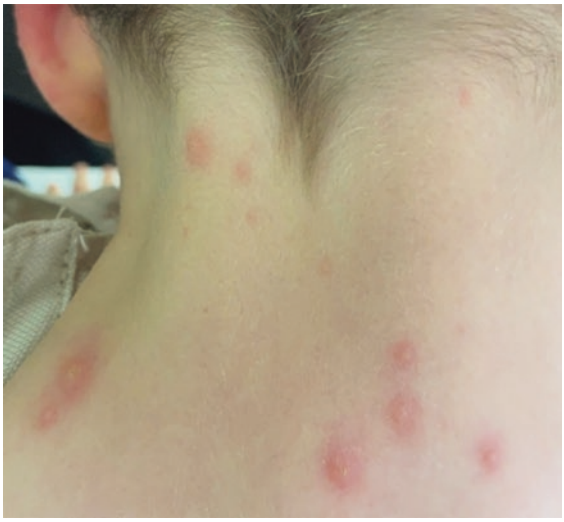

Fig. 3.3 Tense vesicles with surrounding erythema over the neck

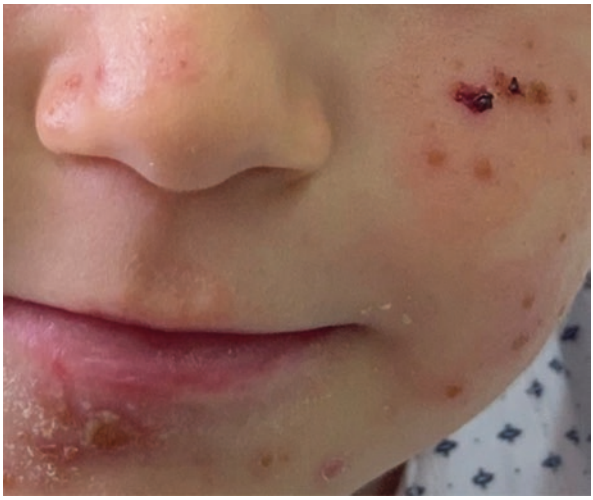

Fig. 3.4 Vesicles and papules were turning into necrotic crusts in the face a week after the first visit

4. Hydroa vacciniforme
5. Hydroa vacciniforme-like lymphoma
6. Protoporphyria

Diagnosis

Classical hydroa vacciniforme.

Discussion

The patient was diagnosed with classical hydroa vacciniforme (HV), an extremely rare photosensitivity disorder of childhood. The key to the correct diagnosis, in this case, is recognizing crops of papulovesicles or vesicles that appear on uncovered areas of skin and heal with vacciniform scars.

Classical HV, also known as Bazin's hydroa vacciniforme, was described by Pierre-Antoine-Ernest Bazin in 1862 [1]. HV usually develops in early childhood and resolves by puberty. Severe forms most often occur in male individuals. The primary occurrence and the recurrences have been reported in spring and summer. The most affected areas include the face, ears, hands, and lower limbs. Signs and symptoms develop about 30 min to 2 h after sun exposure. The flare-ups usually start as mild burning, followed by the development of umbilicated and sometimes necrotic papules on an inflamed base. Some patients may also develop keratoconjunctivitis, photophobia, lifting of fingernails and toenails, fever, and malaise [1–3].

The diagnosis is based on a cutaneous manifestation, laboratory tests, and biopsy. Histologic changes in an early stage include intraepidermal vesicle formation with focal epidermal keratinocyte necrosis and spongiosis. Perivascular neutrophil and lymphocyte infiltrate can also be present. Older lesions show necrosis, ulceration, and scarring. Phototesting can also be done and may show increased sensitivity to short-wavelength UVA. However, phototesting could not differentiate HV from other photodermatoses [3, 4].

HV-like lymphoproliferative diseases (HV-LPD) include EBV-positive HV, atypical HV, and HV-like lymphoma. HV-LPD is a cutaneous form of chronic active EBV infection with skin manifestations similar to HV. HV-like eruptions are distinguished from classical HV by developing lesions in both exposed and sun-protected skin and by the presence of systemic symptoms such as fever, lymphadenopathy, and hepatosplenomegaly. Patients with HV-LPD develop papulovesicular lesions, ulceration, and scars in the disease's early stage [5].

Photocutaneous porphyrias are a subset characterized by acute skin pain and chronic skin lesions as the disease's main features. Porphyrias with photocutaneous features include erythropoietic protoporphyria (EPP), X-linked protoporphyria (XLP), congenital erythropoietic porphyria (CEP), and porphyria cutanea tarda (PCT) with its rare variant hepatoerythropoietic porphyria (HEP). Almost all porphyrias are caused by inherited mutations in genes encoding enzymes regulating the heme biosynthetic pathway. Each porphyria is characterized by accumulated porphyrin precursors, intermediaries, or by-products. Skin fragility and blistering develop in PCT/HEP, CEP, VP, and HCP. Rapid-onset severe burning skin pain evoked by light exposure is the predominant complaint in EPP and XLP. Precise diagnosis of any symptomatic porphyria is possible by demonstrating disorder-specific biochemical profiles of porphyrins or porphyrin precursors in body tissues and fluids [6].

Polymorphous light eruption (PMLE), also known as prurigo aestivalis, is a common form of primary photosensitivity. The rash can take many forms, although, in one person, it usually looks the same each time it appears. It mainly occurs in young adults after the first exposure to intense sunlight during the spring or early summer. Lesions appear within hours of exposure to sunlight and stay for a few days. However, the vesicular form of PMLE is uncommon, and the lesions almost always heal without scarring. Juvenile spring eruption (JSE) is estimated to be a localized variant of a PMLE. The most common time for the presentation of JSE is in the early spring when the weather is still cold [2].

Photoavoidance remains a crucial factor in the care of patients with HV. Other treatments, such as antimalarials, azathioprine, cyclosporine, thalidomide, beta-carotene, and fish oils, are of uncertain efficacy [7]. Prophylactic photo-therapy with narrowband UVB or PUVA might be beneficial.

Key Points

- Hydroa vacciniforme is a rare pediatric disorder characterized by photosensitivity and recurrent vesicles that heal with vacciniform scarring.
- It is frequently associated with Epstein–Barr virus.

- Focal intraepidermal vesiculation, reticular keratinocyte degeneration, epidermal and upper dermal necrosis are pathognomonic histologic changes.
- The primary treatment for HV includes sun avoidance, sun-protective clothing, and sunscreen (UVA and UVB protection).

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Chapter 4
A Seven-year-old Boy with Stomatitis, Conjunctivitis and Dehydration

Orsola Ametrano, Filomena Barbato, Elena Sammarco, Stefania Gagliardi, and Giuseppe Ruggiero

Introduction

A seven-year-old boy is brought to the emergency room by his parents because of severe stomatitis with difficult feeding and dehydration. His parents reported that 10 days earlier the child had bronchitis treated with cefixime but after few days the symptoms had worsened with the onset of fever, purulent ocular discharge, stomatitis and erosions to the labial mucosa with difficulty in feeding. For this reason, acyclovir and nystatin were started without improvement. At the admission, the patient was in fair general conditions with axillary temperature of 38.2 °C, bruised lips, hyperemic gums (Fig. 4.1), bilateral conjunctivitis with yellowish secretions, submandibular lymphadenopathy, and papular lesion on the palm of the hand. Oral cavity was evaluated with difficulty because of intense pain. Blood tests showed increased white blood cells with neutrophilia and elevation of CRP (40 mg/l); renal and hepatic function test were in the normal range and the autoantibody screening tests were negative.

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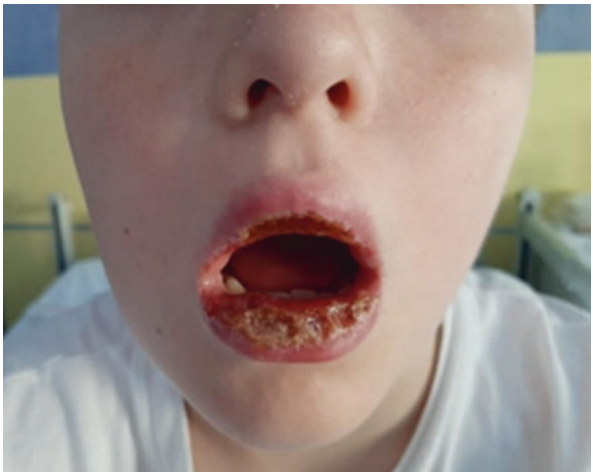

Fig. 4.1 Severe oral mucositis with hemorrhagic crusting in our patient

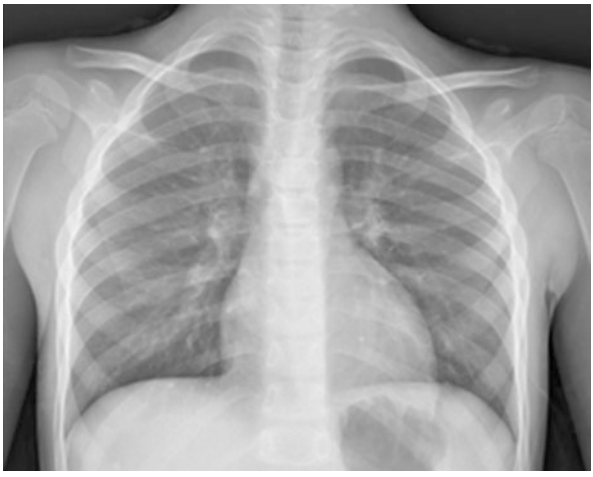

Fig. 4.2 Chest X-Ray

An infectious panel test for herpes viruses, streptococci, toxoplasma, cytomegalovirus and rubella virus was negative. IgM and IgG anti mycoplasma pneumoniae were present. A chest X-Ray was performed and an atypical pneumonia resulted.

Considering the Case Description and the Figs. 4.1 and 4.2 What Is your Diagnosis?

1. Atypical Kawasaki disease
2. Herpetic Gingivostomatitis
3. Stevens Johnson Syndrome (SJS)
4. Mycoplasma-Induced Rash and Mucositis (MIRM)

Diagnosis

Mycoplasma-Induced Rash and Mucositis (MIRM).

Discussion

Recently evidences have described a new entity named "Mycoplasma Induced Rash and Mucositis" (MIRM)) which is distinct from SJS. It's estimated that up to 25–33% of patients with M. pneumoniae infection has some cutaneous manifestations and MIRM occurs in up to 6.8% of cases [1, 2].

M. pneumoniae associated mucocutaneous disease mainly affects young males presenting with prodromal symptoms such as cough, malaise and fever approximately a week before the eruption. In contrast to SJS, 34% of patients with MIRM has no skin rash and an additional 47% has only a restricted skin involvement. The most commonly involved sites are oral (100%), ocular (92%), and urogenital (78%) mucosa [1].

Diagnostic criteria for MIRM are:

- Skin detachment of less than 10% of the body surface area
- At least two mucosal sites involved))
- Few skin lesions
- Evidence of atypical pneumonia
- Laboratory test confirming a M. pneumoniae infection.

Our patient satisfied all the criteria proposed as we found IgM and IgG for M. pneumoniae, two mucosal sites affected (oral and ocular mucosa) and an isolated macular-papular lesion on the palm of his right hand.

It's important to distinguish MIRM)) from SJS because of different prognosis: the 81% of patients with MIRM get a full recovery reoccurring only in about 8% of cases [3].

There are no evidence-based guidelines for treating MIRM and usually are used anti M. pneumoniae antibiotics, systemic corticosteroids and supportive care (pain control, mucosal care, and hydration). Even though antibiotic therapy prevents neurologic and pulmonary complications of M pneumoniae infection, it's not known if it also decreases the incidence/severity of the mucocutaneous involvement. The role of IVIG is still debated.

Key Points

- Keep in mind the diagnosis of MIRM in case of young patient with general discomfort associated with respiratory and mucosal involvement
- It's important to properly identify MIRM as infection-triggered disease to ensure appropriate treatment.

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Chapter 5
A Strange Dermatitis in a Young Boy

Giuseppe Ruggiero, Elena Sammarco, Filomena Barbato, Mario Diplomatico, and Orsola Ametrano

Introduction

A 10-years-old Caucasian boy came to our attention for five lesions which suddenly appeared the day before. They were localized to the abdominal and thoracic region appearing as red-brownish round macules with bullous or erosive area; only one presented with a figurate shape. The child had no mucosal involvement, fever or itching and Nikolsky's sign was negative. He reported that four lesions suddenly appeared the first day and that a fifth lesion appeared the day after (Figs. 5.1 and 5.2). Dermatoscopic examination of the figurate lesion showed an erythematous border, red-brownish reticule and a small ulcerated area (Fig. 5.3). He was treated with systemic Amoxicilline+Clavulanic acid and topical Ozenoxacin.

Based on the Case Description and Pictures, Which Is Your Diagnosis?

1. Bullous Impetigo
2. Dermatitis Artefacta
3. Pemphigus Vulgaris
4. Bullous Pemphigoid

After seven days of local and systemic antibiotic therapy, the child improved significantly (Fig. 5.4). This led to the suspicion that it could be a bacterial infection even if the morphological aspects and the contemporary appearance of the first four lesions still raised doubts about the diagnosis.

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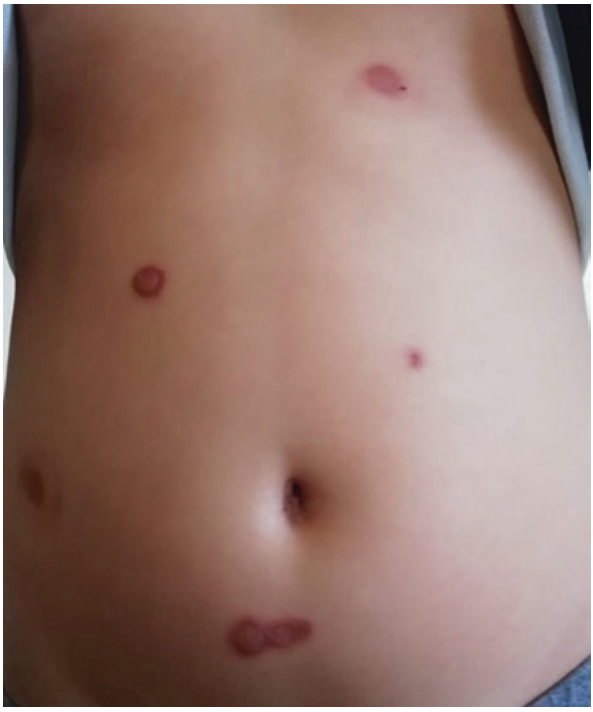

Fig. 5.1 Clinical aspect

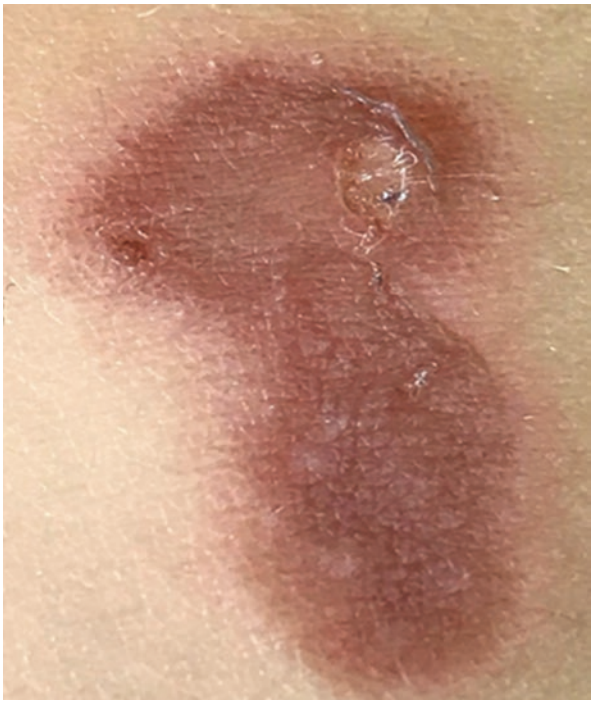

Fig. 5.2 Clinic aspect

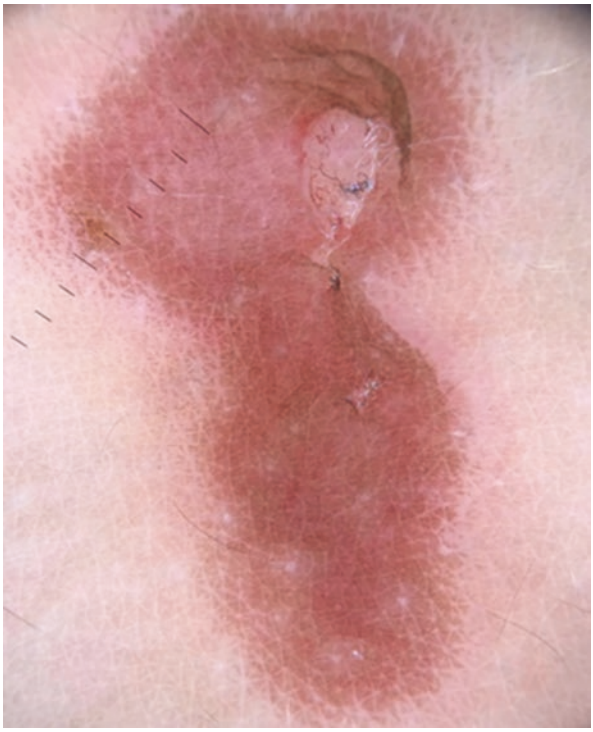

Fig. 5.3 Dermoscopic aspect

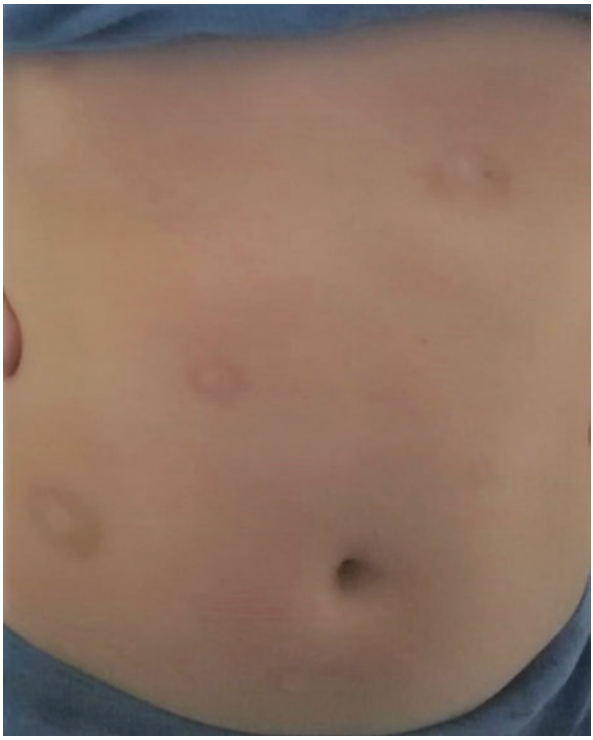

Fig. 5.4 Clinical aspect after seven days of therapy

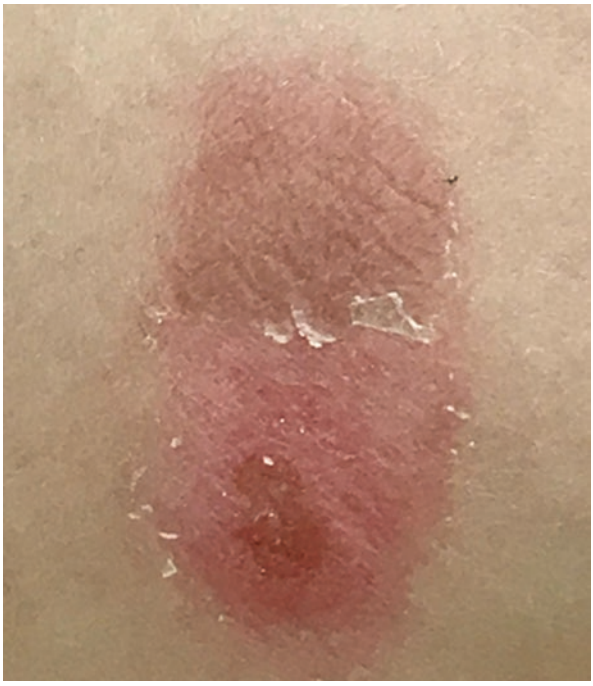

Fig. 5.5 Clinical aspect

In the suspect of Dermatitis Artefacta, we decided to follow up the child.

After one month, the child presented with a single erythematous rectangular lesion on his right clavicular region which had suddenly appeared. At examination, the lesion showed sharp margins, brown upper area and red-pink ulcerate lower area. Dermoscopic examination showed dotted and polymorphic vessels and a small ulcerated area, both present in the red-pink area (Figs. 5.5 and 5.6).

The diagnosis of Dermatitis Artefacta, induced by application of a warm object, was made. The patient was referred to the psychiatrist.

Diagnosis

Dermatitis Artefacta, Dematitis Factitia, Pathomimia.

Discussion

The anamnesis and morphological aspect of lesions excluded the Bullous Dermatosis. Bullous Impetigo is characterized by large, fragile, flaccid bullae that can break and emit yellowish fluid. Typical collarette of scales on its periphery and brown crust on the remaining erosions, after the bullae rupture, are pathognomonic.

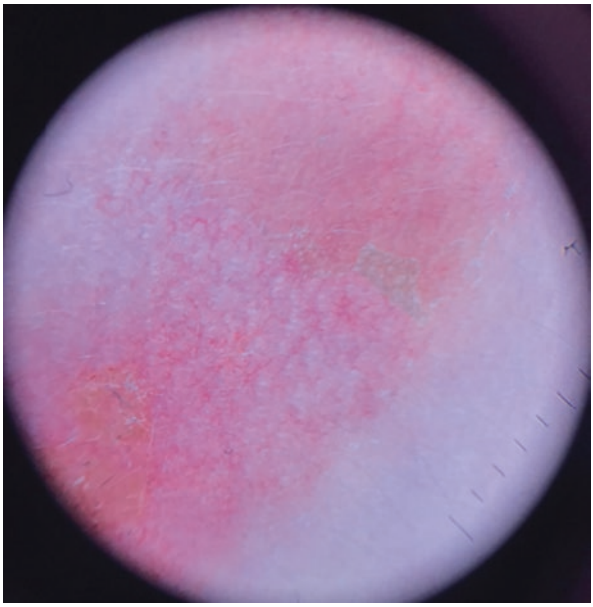

Fig. 5.6 Dermoscopic aspect

Despite the presence of some flaccid bullae, the absence of mucosal involvement, erosions and the negative Nikolsky's sign allowed us to exclude Pemphigus Vulgaris, which is a very rare disease in pediatric age [1–3]. Bullous Pemphigoid was excluded because of the absence of a prodromal phase (lasting days, weeks or months), the absence of erythematous-edematous lesions, and/or patches with indistinct limits of eczematous type, intensely itchy.

Bullous Pemphigoid was also excluded for the absence of gradual eruption of tense bullae overlying erythematous skin and their progression to erosions and crusts [4].

Dermatitis Artefacta is a psychocutaneous disorder, patients create lesions in skin, hair, nail or mucosae to attract attention or satisfy a psychological need. It is rare in children and it is mostly observed in late adolescence to early adult life. The cutaneous lesions may be blisters, erosions, burns, excoriations, urticarial lesions, hemorrhagic lesions or necrosis. The perilesional skin is normal. The mechanism of self-inflicted skin lesions can be mechanical (rubbing, scratching, suction), thermal (heat, cold), chemical (acids/bases contact or autoinoculation, drugs injection), infectious, electrical. The face is the most common site, followed by dorsum of hands and forearm of the non-dominant limb, but any part of the body can be affected. The patient is calm and quite during the medical examination, denies any knowledge of the cause of the lesions. Their relatives are frustrated, angry and asked a firm diagnosis, refusing, in most cases, the psychiatric referral offered [5, 6].

Key Points

- To suspect Dermatitis Artefacta when the cutaneous lesions do not show a clear evolution and they have a bizarre or geometric shape.
- As always, anamnesis should be carefully investigated.
- Always make sure to understand whether the injuries are self-induced or caused by others [7].
- When possible, to exclude other skin diseases on the basis of morphological characteristics, histological data and circulating autoantibodies [8].

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Chapter 6
A Ten-year-old Boy with a Nodular Lesion on the Upper Lip

Giuseppe Ruggiero and Domenico Di Maria

A ten-year-old male shows a lesion between the upper and vermilion border and the right labial commisure (Fig. 6.1). The lesion, appeared suddenly, was mobile, of hard and elastic consistency, about 5 × 5 mm in size.

The child had no injuries to the oral mucosa, or other injuries in other sites of the body and did not report having used dermo-cosmetic products and/or blunt objects.

The mother administered a single dose of betamethasone 1 mg orally.

Based on the Case Description and the Pictures, Which Is Your Diagnosis?

1. Pathomimia
2. Mucocele
3. Granulomatosis orofacialis

Diagnosis

Mucocele.

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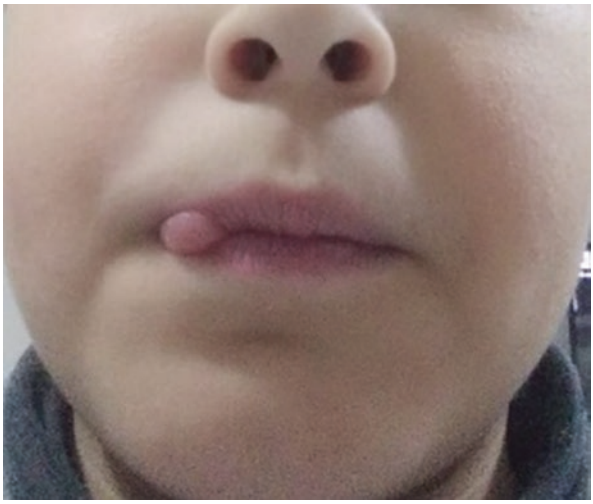

Fig. 6.1 A soft nodule near the left labial commisure

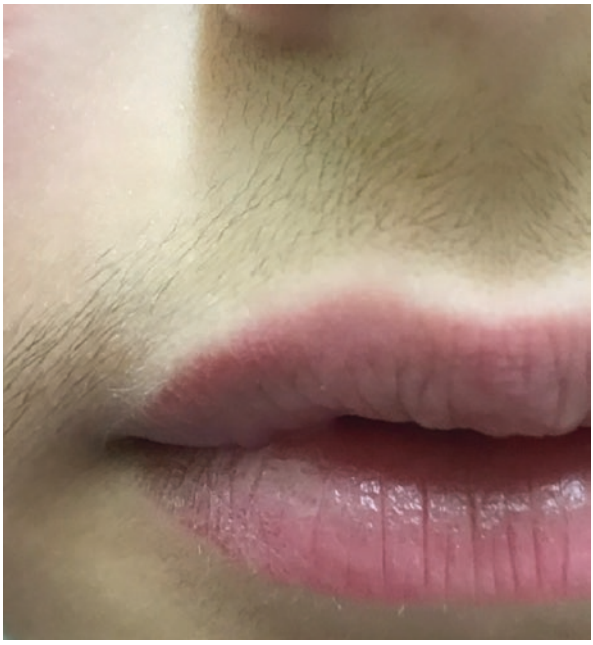

Fig. 6.2 The lesion was early broken and disappeared

Discussion

The next day the lesion was broken, revealing a mucinous substance and the absence of inflammation (Figs. 6.2 and 6.3). The anamnesis, the fast response to cortisone, the clinical appearance led to the diagnosis of upper lip's Mucocele.

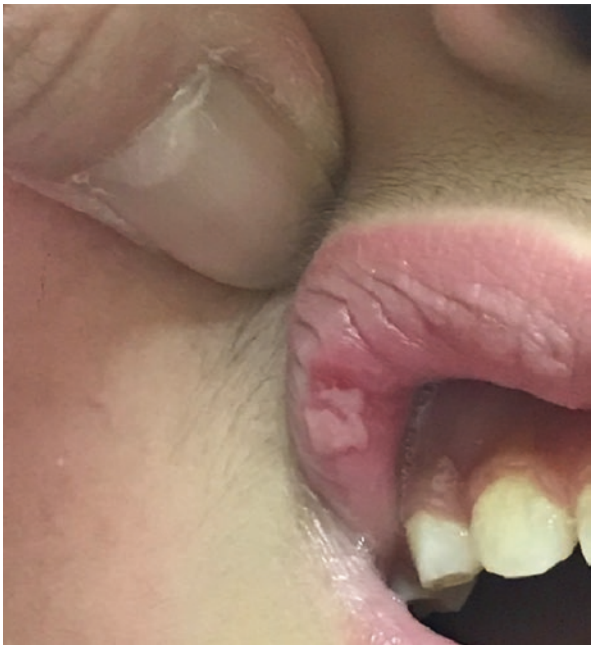

Fig. 6.3 No inflammation lesions were present after breaking, only a small flap of redundant mucosa

Oral mucoceles (OM) are benign soft tissue masses and are clinically characterized by single or multiple, painless, soft, smooth, spherical, translucent, fluctuant masses, which is usually asymptomatic. OM are cavities filled with mucus [1] and are the most common minor (accessory) salivary gland lesion affecting the general population. Minor salivary glands are found in most parts of the oral cavity except the gum. Mucocele can be due to extravasation and retention.

Extravasation mucocele results from a broken salivary glands duct and the consequent involvement of the soft tissues around this gland. Retention mucocele appears due to a blockage of the salivary gland ducts so the secretion is trapped in the gland [2]. When located on the floor of the mouth these lesions are called ranulas because the inflammation looks like the cheeks of a frog.

Diagnosis is clinical; the anamnesis should be carried out correctly and it is the key. The other important informations for a correct diagnosis are looking for previous trauma; lesion location, rapid onset, mobility, size changes, and the soft consistency [3].

Palpation can be helpful for a correct differential diagnosis with orofacial granulomatosis which has a hard consistency, does not appear suddenly and present no fluctuation.

The anamnesis and the typical site allow to exclude an artificial dermatitis mucosa having no inside epithelial components [4–8].

Key Points

- Oral mucoceles are often seen in a pediatric population, it mainly affects the lower lip in young children, but rarely as well the upper lip.

- A careful history and the palpation of the lesion allows to have a diagnostic orientation
- The type of extravasation is the most common.

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Chapter 7
A Young Boy with Fever and Wide Spread Pustules

Carmen Maria Salavastru, Ioana Ionescu, Sabina Andrada Zurac, and Adelina Maria Șendrea

Short Clinical History of the Patient

A 7-year-old male patient presented for a generalized eruption consisting of scaly, well-defined papules, plaques, and placards with non-follicular pustules on the surface, with an annular aspect of the lesions in certain areas (Figs. 7.1 and 7.2). The cutaneous findings were associated with a 5 days lasting high fever (>40 °C), malaise, rhinorrhoea and polyadenopathy.

The patient had a personal medical history of a similar localized eruption in the setting of chronic suppurative otitis and also of scaly erythema associated with pruritus on the scalp, 6 months before. Regarding the patient's family history, he mentioned that his maternal uncle was diagnosed with psoriasis, without being able to mention which clinical type.

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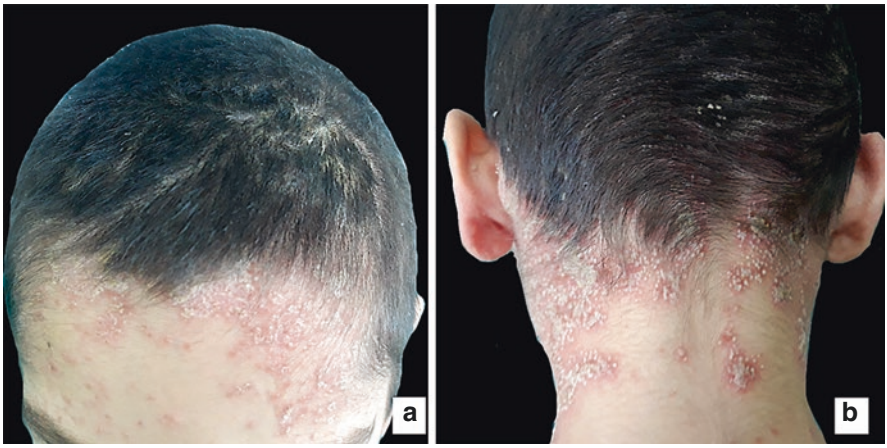

Fig. 7.1 Scaly erythematous papules and plaques associated with pustules on the scalp and forehead (**a**), and also on the posterior aspect of the neck (**b**)

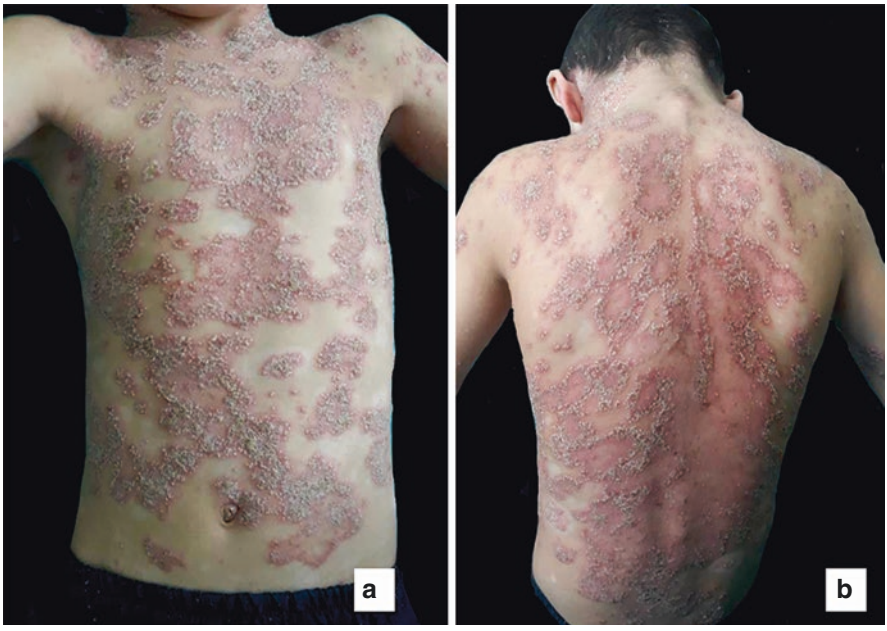

Fig. 7.2 Erythematous scaly papules, plaques, and placards with well-defined borders, associated with pustules on the surface, localized on the posterior (**a**) and anterior (**b**) aspect of the trunk

The patient was admitted to the Pediatrics Department, where the blood tests identified an inflammatory syndrome, leukocytosis and neutrophilia. Additionally, the patient tested positive for influenza type A.

A bacterial swab from one of the pustules was performed, with no identification of any bacterial growth. Under local anesthesia, a punch biopsy from a papular and

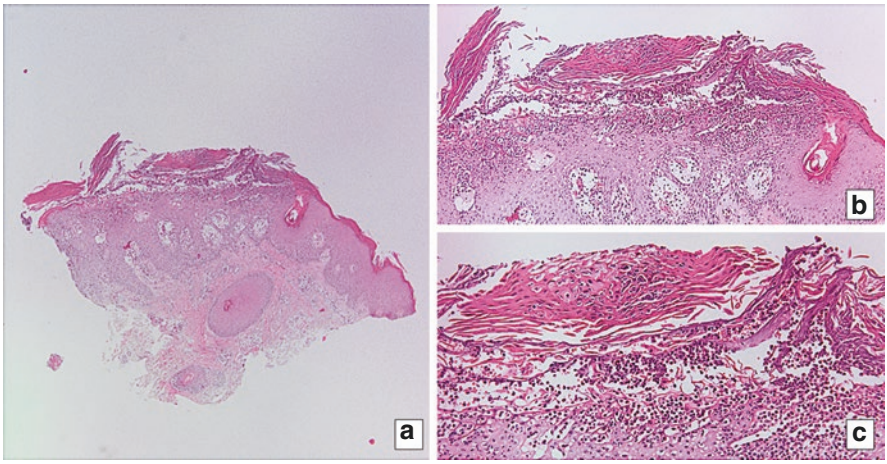

Fig. 7.3 Pathology image: psoriasiform acanthosis and neutrophils collection forming macropustule—HE 4X (**a**); pustule with neutrophils—HE 10X (**b**), HE 20X (**c**)

pustular lesion localized on the left buttock was performed and hematoxylin—eosin staining was conducted. The histopathological analysis (Fig. 7.3) revealed in the epidermis moderate hyperkeratosis, limited areas of hypo-/agranulosis, moderate acanthosis with focal fusion of rete ridges, subcorneal and intraepidermal vesicles and pustules containing numerous neutrophils, and moderate spongiosis near the neutrophilic collections. Moreover, the papillary dermis was significantly oedematous, with ectatic and tortuous capillary vessels, accompanied by quantitatively moderate inflammatory infiltrate.

Based on the Case Description and the Photographs, What Is Your Diagnosis?

1. Generalized pustular psoriasis, von Zumbusch
2. Acute generalized exanthematous pustulosis (AGEP)
3. Sneddon-Wilkinson disease (subcorneal pustular dermatosis)

Diagnosis

Von Zumbusch generalized pustular psoriasis.

Discussions

Pustular psoriasis represents an uncommon, clinically heterogeneous subtype of psoriasis, being an immune-mediated polygenic skin disorder, characterized by epidermic neutrophils accumulation, which leads to sterile pustules development [1, 2]. Pustular psoriasis can present with two clinical phenotypes: localized and

generalized, with the common pathogenic mechanism for both phenotypes being represented by an IL-36 pathway alteration, in which mutations of the IL-36 receptor antagonist determine an upregulation of the IL-1 production [3, 4]. The main clinical manifestation of generalized pustular psoriasis is represented by an eruptive generalized sterile pustulosis and it can present with an acute (also called von Zumbusch), subacute (annular pustular psoriasis) or chronic course. The localized subtype of pustular psoriasis comprises two entities: palmo-plantar pustulosis and acrodermatitis continua of Hallopeau [1].

Generalized pustular psoriasis is a potentially life-threatening, rare type of psoriasis in children (representing 0.6% to 7% of all cases of pediatric psoriasis), with a male predisposition and an onset age between 3 and 16 years old [5]. Various factors, such as infections, topical or systemic treatments, and psychological stress, may trigger or exacerbate the condition. There is a relationship between generalized pustular psoriasis and psoriasis vulgaris as the latter may develop before or after an episode of generalized pustulosis. Furthermore, von Zumbusch type of pustular psoriasis can present as the sole phenotype during a patient's evolution. Molecular genetic studies highlight the theory that generalized pustular psoriasis associated with psoriasis vulgaris and generalized pustular psoriasis have different etiologies, the former being associated with CARD14 mutation, while the latter is associated with IL36RN mutation [5, 6]. Both mutations determine a proinflammatory mechanism activation via the NF-kB signaling pathway.

Generalized pustular psoriasis usually presents with 2–3-mm sterile pustules overlying painful, erythematous skin, accompanied by fever, malaise, neutrophilic leukocytosis, and elevated C-reactive protein [7]. The pustules might be located in the periphery of expanding erythematous plaques or overlying erythematous skin [3]. Extracutaneous manifestations, like cholestasis, epigastric pain, arthralgia, pneumonia, and acute renal failure, may be seen [5, 6].

Although the diagnosis relies on the clinical picture and blood tests, the skin biopsy is performed to confirm the diagnosis. The hallmark histopathologic feature in pustular psoriasis is represented by the spongiform pustules of Kogoj. Other epidermal histopathological characteristics include an absent granular layer, parakeratosis, subcorneal neutrophilic infiltrates, and psoriasiform hyperplasia. In the dermis, dilated blood vessels are identified, along with a scarce inflammatory infiltrate [5, 7].

The differential diagnosis of generalized pustular psoriasis comprises cutaneous afflictions that present with widespread non-follicular pustules [1]. The acute onset, with fever, leukocytosis, and inflammatory syndrome may lead to a false diagnosis of systemic infection. Skin infections must also be ruled out, either primary skin infections or secondary infections of pre-existing dermatological conditions.

The most challenging differential diagnosis is represented by acute generalized exanthematous pustulosis (AGEP), which represents an acute, self-limited, drug-induced eruption consisting of non-follicular pustules developing on erythematous and edematous plaques, associated with fever and marked neutrophilia [2]. The histopathologic features may be similar to pustular psoriasis, but AGEP is characterized by the presence of eosinophilic spongiosis, vacuolar interface change, and

dermal eosinophilia [8]. Moreover, the patient's history is very important, as it usually identifies the prior culprit medication usage, with antibiotics being most commonly involved [1]. Disease resolution is seen at approximately two weeks after drug withdrawal [2].

Sneddon-Wilkinson disease, also known as subcorneal pustular dermatosis, is listed among the differential diagnoses of pustular psoriasis, as a rare, chronic, relapsing dermatosis. Clinically, it presents with crops of sterile, very superficial vesiculo-pustules that develop in an annular or polycyclic configuration on slightly erythematous flexural skin. The lesions might be pruritic and systemic manifestations are uncommon. Compared to pustular psoriasis, the histopathology report reveals subcorneal pustules, in the absence of spongiform pustules and other psoriasiform epidermal changes. Moreover, its successful response to dapsone treatment also makes it a distinct entity from pustular psoriasis [2, 9].

Treatment of childhood generalized pustular psoriasis is challenging, considering the small number of cases and the lack of randomized controlled trials in this special patient population. Besides trigger removal and general supportive measures, first-line therapy is represented by systemic treatment with methotrexate, acitretin or cyclosporine. As a second line treatment, infliximab can be used, either as monotherapy or in association with methotrexate, in severe, rapidly progressive or potentially life-threatening cases, that are unresponsive to other systemic medications. Corticosteroids should be used with caution. They may cause flares in patients affected concomitantly by psoriasis vulgaris [3]. Topical treatment with emollients and topical corticosteroids can be associated with the systemic treatment [1, 5, 10].

In this case, considering the patient's age, a systemic regimen with subcutaneous methotrexate 10 mg/week (associated with 5 mg folic acid) and topical treatments (with emollients and class III topical steroids) were prescribed. After sixteen weeks of treatment, remission was achieved, with complete clearing of erythema and pustules; post-inflammatory hypopigmentation occurred (Fig. 7.4).

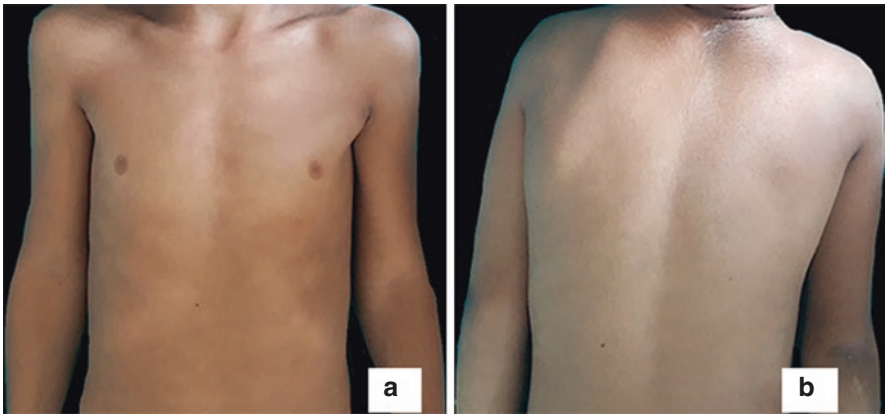

Fig. 7.4 The outcome of the skin eruption on the anterior (**a**) and posterior (**b**) aspect of the trunk after 16 weeks methotrexate treatment—postinflammatory hypopigmentation

Although uncommon in pediatric dermatology patients, acute generalized pustular psoriasis (von Zumbusch) should be considered when dealing with patients that present with generalized non-follicular, sterile pustules located on an erythematous background, in order to initiate as soon as possible the systemic therapy.

Key Points

- Pediatric acute generalized pustular psoriasis is a rare, potentially life-threatening entity characterized by eruptive generalized sterile non-follicular pustules, associated with systemic manifestations.
- A thorough differential diagnosis should be performed, with special attention towards acute generalized exanthematous pustulosis (AGEP).
- Treatment represents a challenge, but combined systemic and topical treatment, as well as supportive measures, are required.
- To have a better understanding and management plan for generalized pustular psoriasis in the pediatric population, further research and evidence-based studies are needed.

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Chapter 8
Acneiform Eruption in Prepubertal Children

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A 11-year-old, otherwise healthy boy, presented for a one-year history of facial flushing and burning sensation. Six months prior, he developed erythematous papules over the nose and forehead. Initial treatment with topical corticosteroids (TCS) and topical antibiotics showed no improvement. Also, he had an 8-month history of itching and redness in both eyes, treated with various topical agents. His family history was insignificant.

Dermatological examination revealed background erythema and telangiectasias on the central aspects of the face. Small, dome-shaped erythematous papules and tiny surmounting pustules with no comedones, symmetrically distributed over the nose, cheeks, and forehead, were observed (Fig. 8.1).

Based on the Case Description and the Photograph, What Is Your Diagnosis?

1. Perioral dermatitis
2. Rosacea
3. Acne
4. Angiofibromas
5. Demodicosis

Diagnosis

Childhood rosacea.

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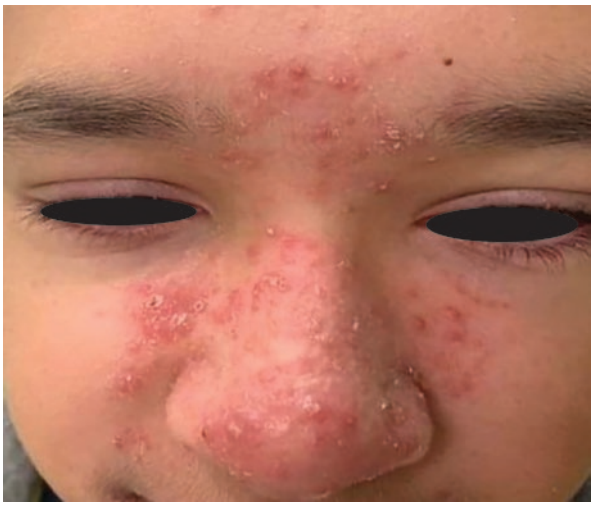

Fig. 8.1 Papulopustular lesions on a telangiectatic background in the central convex areas of the face. The papules coalescent into infiltrated erythematous plaques over the nose. Redness and scaling on the eyelids

Discussion

Topical metronidazole gel (0.75%) once daily was started. We observed significant improvement after two months.

Rosacea is a chronic relapsing inflammatory skin disease with a high prevalence among adults with fair skin. It is a phenotypically heterogeneous disease, classified into four major subtypes: erythematotelangiectatic, papulopustular, phymatous, and ocular. There is the frequent simultaneous occurrence of more than one subtype and one subtype's potential progression to another. Signs and symptoms of rosacea may vary over time, often fluctuating between exacerbation and remission [1]. Papulopustular rosacea (PPR), also known as inflammatory rosacea, is characterized by central facial erythema and transient papules and/or pustules, with or without telangiectases. Patients might exhibit severe intermittent flares of acute vasodilation (flushing of rosacea) [2].

Rosacea is very rare in the pediatric population, especially before puberty [3]. A family history of rosacea is sometimes reported. The clinical feature is similar to adults: the disease often begins with flushing and then progresses to papules and pustules' development. Children can present with vascular, papulopustular, and ocular findings. However, phymatous rosacea is only seen in the adult population. Sometimes pediatric rosacea tends to be discrete and similar to perioral dermatitis [4]. Granulomatous rosacea is extremely rare in the pediatric age group, with only a few cases reported in the literature. The youngest patient with rosacea fulminans, a rare exacerbated form of rosacea, was a 3-year-old girl who developed a sudden onset of pustules, cysts, and purulent discharging sinuses on the cheeks, chin, perioral, and paranasal areas [5]. Adult patients are often provoked by heat, sunlight, topical corticosteroids, spicy foods, hot liquids, certain soaps, cosmetics, and topical medications; however, it is unclear which of these factors are triggers of pediatric rosacea [3].

In adults, the prevalence of ocular involvement in rosacea varies from 30% to 50%; in approximately 20% of the patients, they precede cutaneous findings. Ocular rosacea is frequently misdiagnosed in the pediatric population. In children, ocular symptoms are often present at diagnosis, especially in girls. Some studies have suggested an increased risk of ophthalmic complications in children, as opposed to adults. However, there is no correlation between the ocular disease's severity and cutaneous rosacea [6].

Chamaillard et al. [7] studied 20 patients aged 1 to 15 years with cutaneous or ocular rosacea. The mean age at presentation was 4.6 years (median age, 42 months). Six patients had a family history of rosacea, and 13 had skin phototypes I or II. Among these 20 patients, 11 had both ocular and cutaneous rosacea.

Childhood rosacea can mimic many other inflammatory facial dermatoses. It is essential to rule out other papulopustular eruptions, including perioral dermatitis, steroid rosacea, acne, and angiofibromas. Despite clinical similarities, there are significant differences regarding the patient's age, clinical history, and presentation of the lesions.

Perioral (periorificial) dermatitis typically presents with red to flesh-colored papules and rarely pustules near the eyes, nose, and mouth. Although the etiology is unknown, many patients have had recent exposure to a topical or, less commonly, an inhaled or systemic corticosteroid [6].

The distribution of steroid rosacea to the eyelids and lateral face may distinguish it from the centrally located typical rosacea [8]. Steroid rosacea usually starts after discontinuing potent TCS. The majority present with a history of worsening of symptoms after sun exposure. Interestingly, eye problems are reportedly absent [9].

The onset of acne typically correlates with the onset of puberty. Prepubertal acne is the appearance of acne before true puberty due to the maturation of the ovary and testis. There is little adrenal production by gonads in this age, so the causative factor for prepubertal acne can be attributed to adrenal androgens. Lesions of prepubertal acne mainly consist of comedones, with or without inflammatory lesions. The central area of the face (mid-forehead, nose, and chin) is primarily affected. Lesions may appear before any other signs of pubertal development. Severe acne in prepubertal age raises the possibility of hormonal abnormality [10].

Facial angiofibromas are considered one of the most apparent clinical presentations of tuberous sclerosis (TS), an autosomal dominant hamartomatous disorder. Multiple facial angiofibromas are also found in multiple endocrine neoplasia type 1 (MEN-1) and Birt-Hogg-Dube syndrome. When they are extensive and bilateral, they are pathognomonic of TS. The angiofibromas start to appear between the 1st and 4th years of age and typically grow during puberty. Fibrous papules are solitary, dome-shaped, skin-colored to red, located around the nose and on the malar eminences. They can have tiny telangiectatic vessels located on the surface. In TS, angiofibromas can coalesce into plaques [11].

Demodicosis typically involves the periorificial regions in adults and the elderly population. Patients can present with fine, white-yellow, scaly changes of the sebaceous hair follicles. Papules and pustules also can be present. Erythema and inflammation are minimal. Ocular manifestations of demodicosis include chronic anterior

blepharitis, posterior blepharitis, meibomian gland dysfunction, recurrent chalazia, and keratoconjunctivitis. In children, demodicosis is linked to HIV, leukemia, Langerhans cell histiocytosis, acute lymphoblastic leukemia, and T-cell non-Hodgkin lymphoma. The diagnosis is easily confirmed by the presence of numerous Demodex mites [12].

Pediatric rosacea responds well to treatment, although it may take several months before a noticeable improvement. General measures include discontinuing applying all facial creams, including TCS and cosmetics. Standard topical therapies include metronidazole and erythromycin. Systemic treatment is primarily based on published case reports and small case series: tetracycline (500 mg twice daily tapered to 250 mg), minocycline (50–100 mg twice daily), doxycycline (50–100 mg twice daily or four times daily), erythromycin (30–50 mg/kg daily), clarithromycin (15 mg/kg twice daily for four weeks and then daily for four weeks), and azithromycin (5–10 mg/kg daily). Tetracyclines are not recommended for treatment in children younger than eight years of age [4, 13].

Key Points

- Rosacea in children, especially before puberty, is uncommon.
- An association of ocular symptoms and facial inflammatory dermatosis in a child should lead to suspicion of rosacea.
- Flushing, presence of telangiectasia, and the ocular findings allow the differentiation of rosacea from other facial eruptions
- Topical metronidazole and age-appropriate oral antibiotics are the main therapeutic agents of management.

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Chapter 9
Acquired Pigmented Lesion in a 6-year-old Child

Laura Ciulli and Antonio Iannone

A 6-year-old boy of Moroccan origin came to our observation for an asymptomatic pigmented lesion on the left knee. Brown-black in colour, it appeared about three years earlier and after a period of stability suddenly and rapidly increased in the last three months (Figs. 9.1 and 9.2).

On physical examination it appeared as a dome-shaped papular lesion of about 0.5 cm in diameter, with a homogeneous black colour, with slightly irregular surfaces and margins.

The dermoscopic examination shows pseudopods regularly distributed throughout the periphery (Fig. 9.3).

Based on the History and the Photographs Which Diagnosis Would You Propose?

1. Spitz-Reed nevus
2. Spitzoid melanoma
3. Atypical Spitz tumor

Diagnosis

Spitz-Reed nevus.

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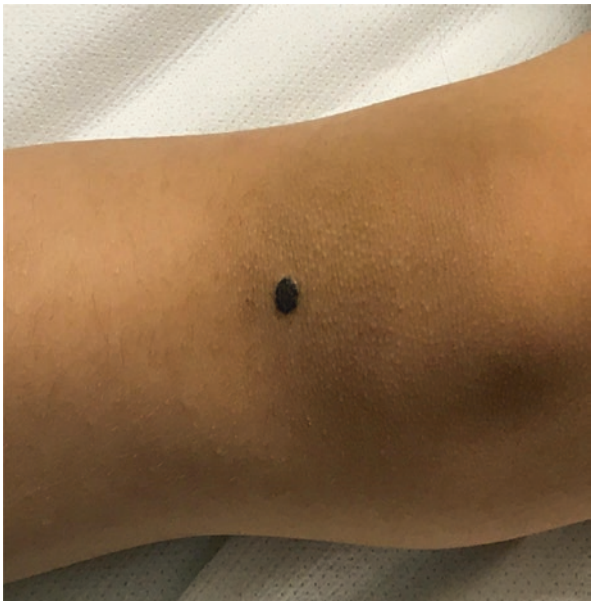

Fig. 9.1 Acquired papular pigmented lesion on the left knee

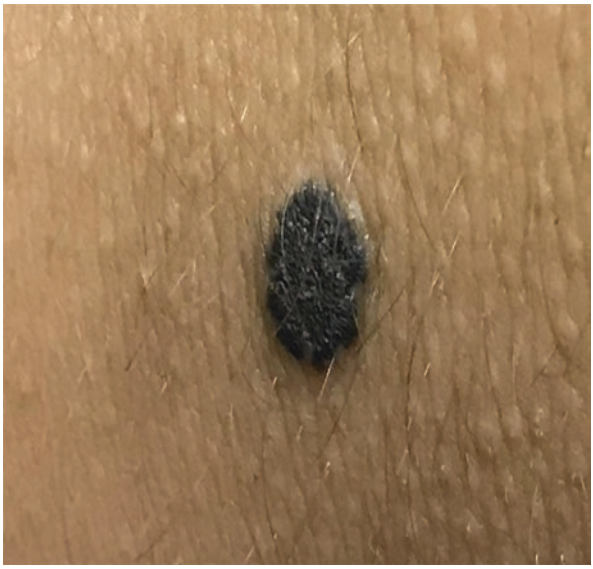

Fig. 9.2 Acquired papular pigmented lesion on the left knee

Discussion

A characteristic starburst pattern on dermoscopy—presence of pseudopods regularly distributed along the entire circumference of the lesion—is commonly observed in pigmented Spitz naevus (Spitz-Reed naevus) at the advanced stage. It

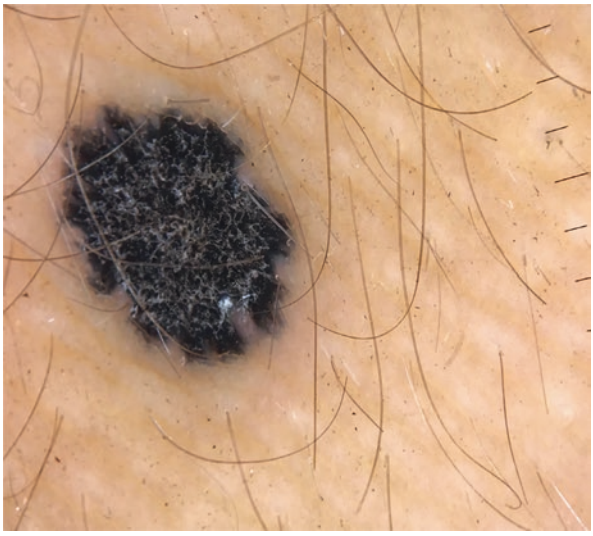

Fig. 9.3 Dermoscopy Examination. Homogeneous central black pigmentation with regular radial streaks at the periphery (starburst pattern)

occurs in about 60% of cases and is a clue for accurate diagnosis [1]. This pattern correlates with the histopathological feature of heavily pigmented nests of melanocytes along the dermoepidermal junction [2].

Nevertheless, the rapid growth of the lesion years after its onset—as occurs in our case—requires its diagnostic removal to exclude spitzoid melanoma or atypical Spitz tumor.

In fact the lesion was surgically excised under local anaesthesia and the hystopathological diagnosis was "Junctional Spitz nevus".

Spitz-Reed nevus belongs to the spectrum of melanocytic lesions histologically characterized by epithelioid or spindle cells. Classical Spitz nevus, first described by Sophie Spitz in 1948 as "melanoma of childhood" [3] is generally non-pigmented. Spitz-Reed nevus is the nevus originally described by Reed [4] as pigmented lesion. It is considered by most pathologists a lesion included in the spectrum of clinical and histopathological presentation of the Spitz nevus [5].

The classic Spitz nevus presents clinically as a usually amelanic (pink or red) papule, plaque or nodule, mainly localized on the cephalic area and on the lower limbs. The Spitz-Reed nevus is instead a variant characterized by an intense brown-black pigmentation more often localized on the lower or upper limbs [6].

The most frequent dermoscopic feature of Spitz-Reed nevus is the starburst pattern with the presence of symmetrical radial striae ending with club-like swellings. It is followed by globular pattern (large black-brown globules also arranged a bit like a starburst with a centrifugal spread) and homogeneous pattern.

Spitz-Reed nevus is characterized by rapid initial growth, then a stabilization phase followed in many cases by a slow involution until its disappearance. During these phases the Spitz-Reed nevus changes its dermoscopic pattern. Over time, the starburst and globular patterns are replaced by a multicomponent or non-specific pattern [1]. Dermoscopic monitoring allows a conservative management of all

lesions with a spitzoid appearance in pre-adolescents [7]. Nodular lesions with a diameter greater than 8 mm and those that continue to grow or resume growth months after their onset should be surgically removed [8]. Their diagnostic excicsion also quells parents anxiety.

Differential diagnosis include

1. Spitzoid Melanoma. Rare in pediatric age (0.8 cases per million inhabitants per year in the US). On dermoscopic examination it never shows a starburst pattern.
2. Atypical Spitz tumor. It belongs to a large group of borderline lesions of difficult clinical and histological classification. About 40% of these lesions are associated with regional metastases, but have a mortality rate of less than 5%. These Spitz tumors usually show a multicomponent dermoscopic pattern, and when melanotic, theyt can present with dotted vessels and white lines, unlike what was found in our patient [8].

Key Points

- The Spitz-Reed nevus, as well as the classic Spitz nevus, is a benign melanocytic neoplasm that most frequently affects the pediatric age
- The most frequent and most characteristic dermoscopic patterns of the Spiz-Reed nevus are the starburst and globular
- A conservative attitude, clinical and dermoscopic monitoring (approximately every three to six months), is currently considered the “standard of care”
- Lesions with a diameter greater than 1 cm, those ulcerated, those that continue to grow after the first 4-6 months and those that start to grow again after a stabilization phase must be surgically removed for an histopathologic evaluation.

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Chapter 10
An Erythematous Nummular Lesion with a Centrifugal Growth

Chiara Bordin and Annalisa Franch

A seven-year-old male presented for medical examination because an erythematous lesion had appeared in the left scapular region two days before.

The physical examination shows an ovalar erythematous macule 4 × 6 cm in diameter with a lighter central area and an hyperemic ring centrifugally spreading (Figs. 10.1 and 10.2).

There were no alterations on the surface and the lesion is smooth to the touch.

The patient has always been healthy. Any symptoms have been reported and the general physical examination did not show any other signs.

Based on the Case Description and the Photographs, Which Is Your Diagnosis?

1. Tinea corporis
2. Granuloma annulare
3. Erythema migrans

Tinea Corporis

It presents as a round inflammatory lesion with a red ring and shows a centrifugal growth. The edges, where the inflammation is progressing, is more evident while the central area has a partial resolution of the inflammation. Superficial alterations such as crusts and scales are usually present.

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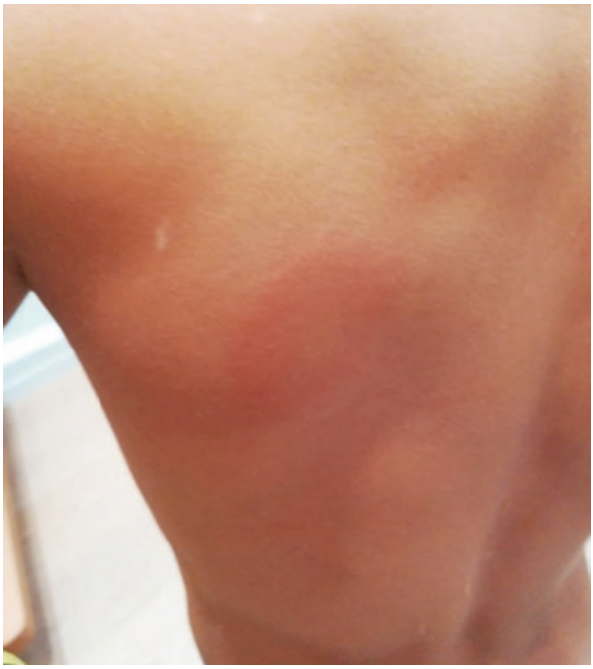

Fig. 10.1 The erythematous macule in the left scapular region

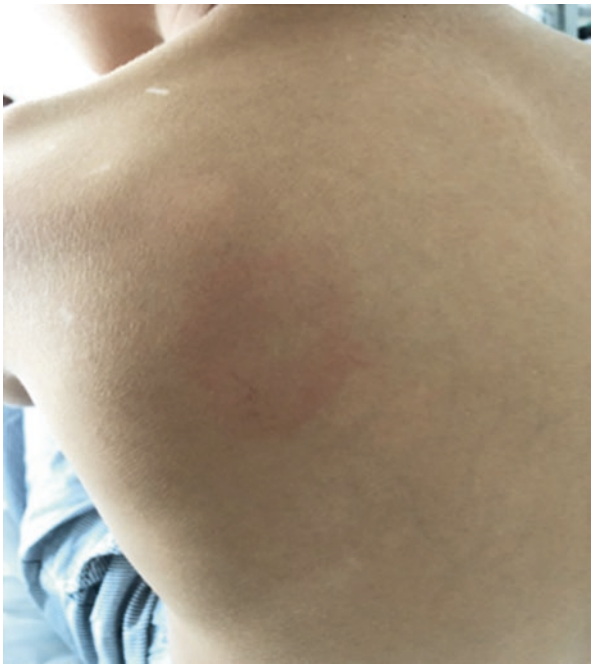

Fig. 10.2 The lesion after four days: the dimensions have increased, the central area is lighter while the surrounding area showed an hyperemic ring

Anular Granuloma

It is clinically characterized by round annular lesions without superficial alterations. The center of the lesions may be depressed and borders raised, composed of numerous dermal papules. Localized granuloma annulare is the most common form and is usually found on the dorsal surfaces of the feet, hands, and fingers. Lesions rarely appear on the trunk.

Diagnosis

Erythema migrans.

A useful contribution to the diagnosis was provided by a photo of the patient's back after an excursion in the woods around Bolzano, two weeks before (Fig. 10.3).

The image shows a tick, lodged in the skin of the left scapula, at the center of a small hyperemic papule. This image suggested a precise topographic correlation between the tick bite and the erythema migrans. As often happens, the tick was removed after the photo was taken and the episode has been forgotten. The papule disappeared during the following days.

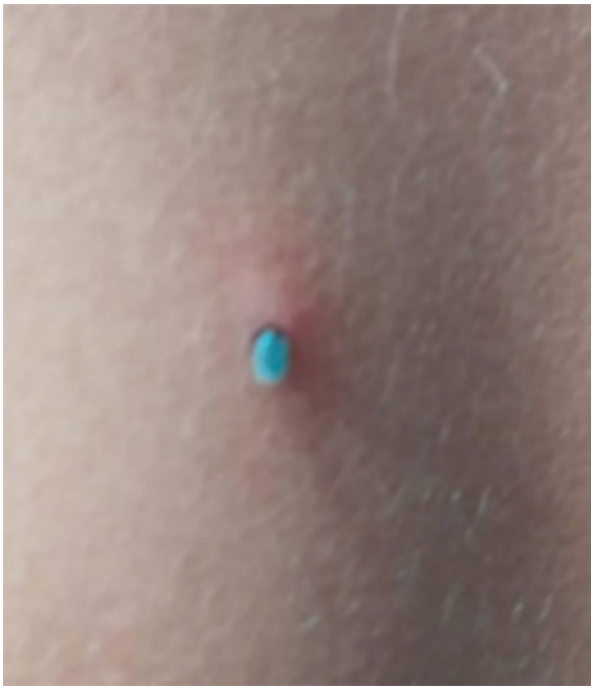

Fig. 10.3 Hyperemic papule presenting a tick in its center

Discussion

Erythema migrans is a pathognomonic sign of the initial stage of Lyme disease (borreliosis), a bacterial infection caused by the a tick bite. The bacteria responsible is the spirochete *Borrelia Burgdorferi*, which is carried by many animal species (deer, fawns, rodents, birds and also horses, dogs and cats).

Borrelia is spread to humans through the bite of a particular tick, the *Ixodes Ricinus.*

Lyme disease takes its name from the homonymous American town where the first case was recorded in 1975. It is common in many regions of the United States and in Europe (UK, France, Austria and adjacent states of East Europe),

In Italy, the regions where it's most common are Friuli Venezia Giulia, Trentino Alto Adige, Liguria, Veneto and Emilia Romagna - in particular the wooded areas between 600 and 1500 m of altitude.

The disease occurs mostly in the summer and autumn.

The incubation period is around three days to three weeks, on average two weeks, but it can reach one month. The disease, if not treated, can evolve in three stages:

Stage 1

Erythema migrans is the main feature of Lyme disease and is found in at least 75% of patients. It usually appears after a few weeks in the area of the tick bite as a small reddish papula. The lesion increases in size and, often becomes clearer in the middle, forming a ring shape. Later, in 50% of untreated patients, a lot of secondary lesions usually smaller in size appear. The skin lesions are commonly warm to the touch but don't hurt. In the majority of patients, the erythema migrans is accompanied by a flu-like syndrome with asthenia, fever, headache, muscle and joint aches [1].

Stage 2

After a few weeks or a month, if the disease has not been treated, neurological symptoms can appear. These are lymphocytic meningitis, meningoencephalitis, neuritis of the cranial nerves (especially paralysis of the facial nerve) and peripheral polyneuropathies. Cardiac involvement, with atrioventricular block, myocarditis, pericarditis can also appear [1].

Stage 3

Stage 3 symptoms are present in 60% of untreated patients, after a period which varies from a couple of weeks to a maximum of two years. The most typical symptom is arthritis of the major joints [1].

The diagnosis of Erythema migrans is suggested by the presence of an hyperemic macular or popular lesion which:

- increases in size showing a typical lighter center
- doesn't cause itching or pain
- occurs on the site of the tick bite
- becomes visible one to four weeks after a tick bite (but can appear from three days to even three months) and lasts for several weeks

Erythema migrans is pathognomonic and does not require any further laboratory investigations. Serological tests are not useful at this stage of the infection.

In contrast, the diagnosis of neuroborreliosis requires the assessment of serum and cerebrospinal fluid [2].

The administration of oral amoxicillin or doxycycline (after eight years of age) will prevent the progression to the more severe later stages of Lyme disease [3].

Routine antibiotic prophylaxis is not justified after a tick bite, even in an endemic area, as the risk of infection is low [3].

It is best to monitor the skin around the bite and to prescribe an antibiotic only if erythema migrans develops, thus avoiding unnecessary treatment and potential adverse effects [3, 4].

The overall risk of developing Lyme borreliosis after a tick bite is 2.6% [5].

The transmission of the borrelia burgdorferi usually occurs after an infected tick is left in the skin for more than 24 h. Therefore, after a potential exposure, looking for and removing ticks can help prevent infection [6].

Key Points

- Accurate diagnosis of Erythema migrans is essential to initiate an appropriate antibiotic therapy
- When Erythema migrans is promptly treated with appropriate antimicrobial agents, the prognosis is excellent
- Never dismiss the possibility of Lyme disease in asymptomatic individuals without positive anamnesis due to tick bites

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Chapter 11
Asymptomatic Papules of the Trunk

Mohamed Saeed Mohamed, Shady M. Ibrahim, and Mohamed L. Elsaie

Case Presentation

An 8 years old girl presented to the dermatology clinic with multiple asymptomatic papules on her trunk (Fig. 11.1). The lesion had history of two weeks; the parents of the patient gave no relevant familial or personal medical history. On physical examination of the skin, he had multiple asymptomatic pink, well-defined, soft, rounded papular lesions that release white material on compression. There is no evidence of apparent lymph node enlargement. The patient informed removal some of them with recurrence.

Dermoscopic examination revealed multilobular, white-pink amorphous structure in the center with a surrounding crown of vessels that did not cross the centers of the lobules (Fig. 11.2).

Based on the Case Description, Clinical and Dermoscopic Photographs, What Is Your Diagnosis?

1. Hemangioma.
2. Cutaneous horn.
3. Pyogenic granuloma.
4. Molluscum contagiosum

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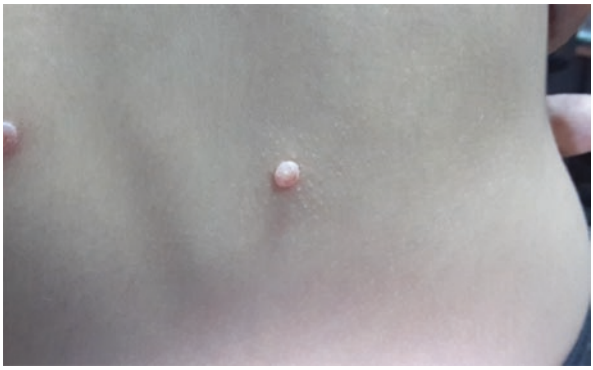

Fig. 11.1 Asymptomatic papule on the trunk of female patient 8 years old

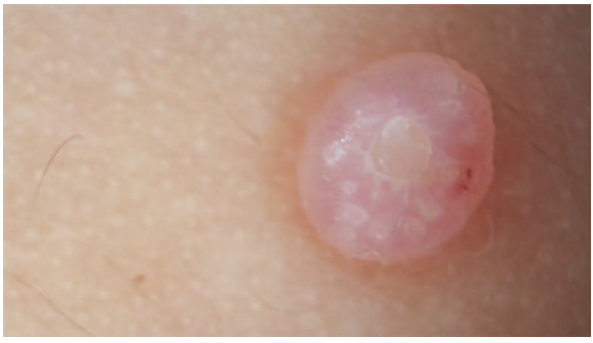

Fig. 11.2 Dermoscopic examination of the papulerevealed polylobular, white-pink, amorphous structure in the center with a surrounding crown of vessels that do not cross the centers of the lobules (Dermoscopy 3gen DermLite 3, magnification 10 X)

Diagnosis

Molluscum contagiosum.

Discussion

Molluscum contagiosum is a viral infection of the epidermal keratinocytes with characteristic intracytoplasmic inclusions. Lesions are generally multiple, translucent appearing with umbilicated skin-colored papules, located at various sites on the skin surface [1]. Lack of central umbilication may be observed [2]. Atypical lesions, small, single, initial lesions, inflammatory lesions and lesions with perilesional eczema also may be present [3]. This difficulty in diagnosing the condition may be present at the initial evaluation, during treatment or in the follow-up of the patient [4].

Although the clinical diagnosis of molluscum contagiosum is considered simple, Dermoscopy can help diagnosis especially in atypical presentation which revealed polylobular, white-pink, amorphous structure in the center with a surrounding

crown of vessels that do not cross the centers of the lobules. This pattern is caused (1) by inverted lobules of hyperplastic squamous epithelium that expands into the underlying dermis separated by fine septae of compressed dermis and (2) by vessels in this dermis. The recognition of this pattern is very helpful in the clinical diagnosis of molluscum contagiosum in adulthood [5].

Key Points

- Molluscum contagiosum is a viral infection of the epidermal keratinocytes with characteristic intracytoplasmic inclusions.
- Although the clinical diagnosis of molluscum contagiosum is considered simple but dermoscopy help in diagnosis with its characteristic pattern.

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Chapter 12
Atopic Dermatitis, Hidradenitis Suppurativa and Poikiloderma

Fabio Arcangeli and Elisa Sama

A seven-year-old girl presented with classil atopic dermatitis (eczema on the upper and lower limb folds, xerosis, history of wool intolerance and high temperatures, elevated IgE, allergic rhino-conjuntivitis) since six months of age. Hidradenitis suppurativa (acne inversa) had also appeared on the perigenital and groin areas for a year. This was the main reason for the dermatological consultation.

The patient was born at full term but with low weight. Slow weight gain, short stature, and teeth retardation were reported by her parents. Worsening of the facial erythema following sun exposure was also noticed.

The physical examination confirmed the reported dermatological diagnoses (Figs.12.1 and 12.2) but we also notited atrophic and dischromic lesions on the cheeks and extensor regions of the limbs (Figs. 12.3 and 12.4).

The examination of the oral cavity highlighted dental abnormalies referable to micro and hypodontia (Fig. 12.5). Laboratory tests documented only high level of IgE.

Based on the Clinical History and the Photographs Which Is Your Diagnosis?

1. Lupus erythematosus (LE)
2. Rhotmund Thomson syndrome
3. Kindler syndrome
4. Dyskeratosis congenital

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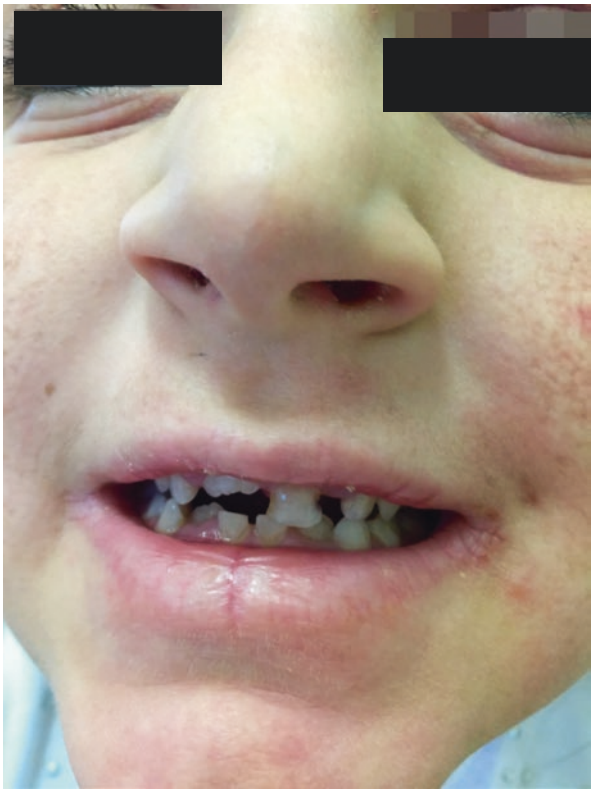

Fig. 12.1 Dennie-Morgan folds on lower eyelids (diagnostic marker of atopic dermatitis), cheilitis and perioral dermatitis

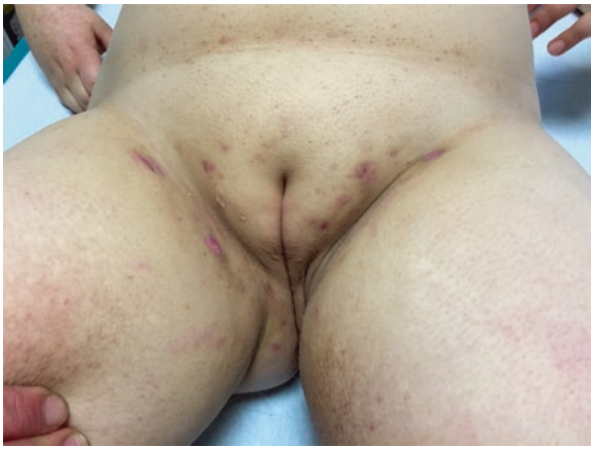

Fig. 12.2 Hidradenitis suppurativa (acne inversa). Blackheads, papules, nodules and scarring on the perigenital areas and groin

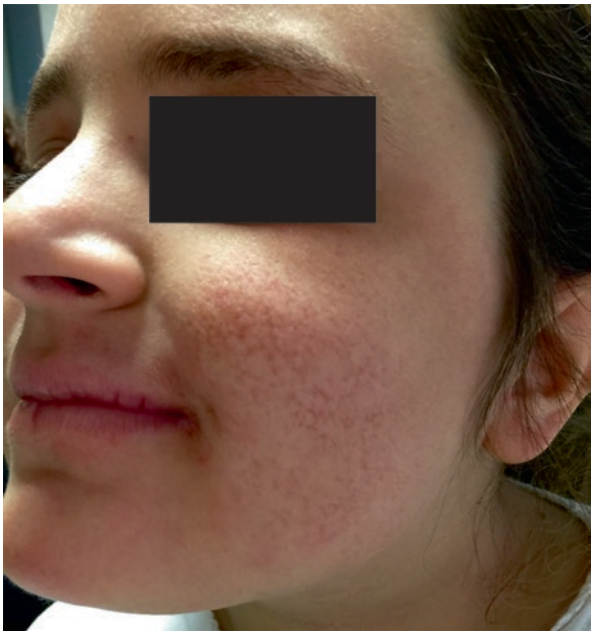

Fig. 12.3 Reticulated atrophy and punctate scars with dischromic appearance on the face

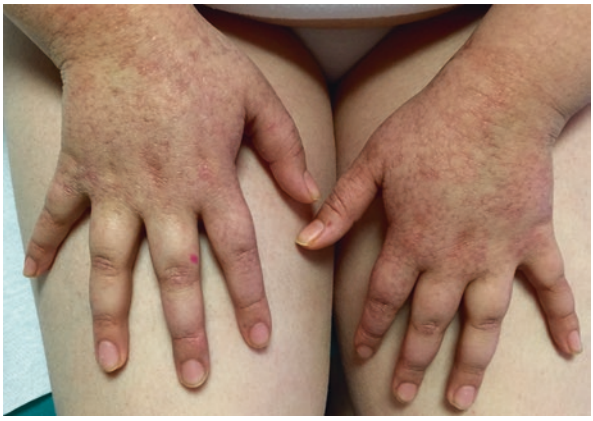

Fig. 12.4 Reticulated atrophy and punctate scars with dischromic appearance on the dorsal regions of the wrists and hands

Diagnosis

Rhotmund Thomson Syndrome (RTS).

Discussion

Rothmund-Thomson syndrome (RTS) is a rare autosomal recessive disorder characterized by: a facial rash, growth retardation, sparse or absent hair on the scalp,

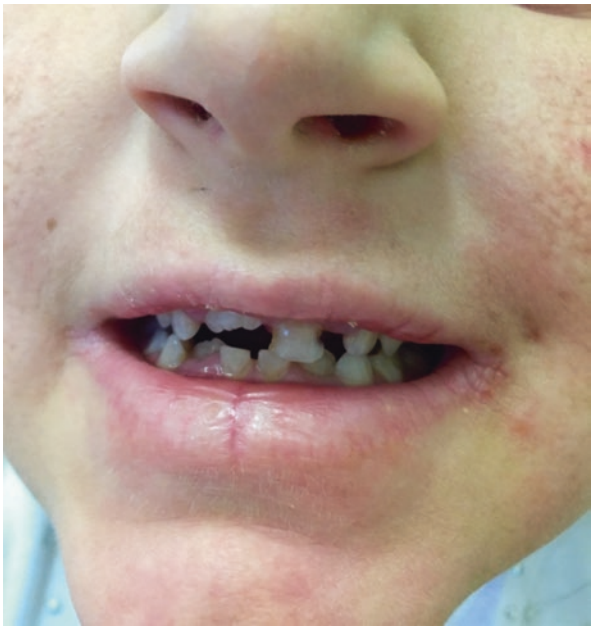

Fig. 12.5 Dental abnormalities

eyebrows and eyelashes, juvenile cataracts, skeletal abnormalities and a predisposition to early onset of cancer. The diagnostic hallmark is facial erythema, which appeares between three and six month, then extending to the extremities with the exception of the trunk. The evolution is towards punctate atrophy with reticular hypo- and hyper-pigmentation having the typical appearance of poikiloderma that persists throughout life [1].

There are two clinically defined forms of RTS: type I, characterized by poikiloderma, ectodermal dysplasia and juvenile cataracts with unknown etiology, and type II characterized by poikiloderma, congenital bone defects, an increased frequency of osteosarcoma and other skin cancers, caused by RECQL4 (8q24.3) mutation [1, 2]. The prevalence of RTS is unknown, about 300 cases have been reported in the literature. Type 2 RTS accounts for almost two-thirds of cases [1].

Patients may display few or many of the associated clinical features. The severity of each sign can also vary. Although some clinical signs suggest precocious aging, life expectancy is not impaired in patients who do not develop cancer [3].

The diagnosis is based on clinical findings (primarily at the age of onset, spreading and appearance of the poikiloderma) and molecular analysis for RECQL4 mutation.

Our patient had same of the clinical signs of RTS as poikiloderma, small stature, dental abnormality. Nevertheless her hair, eyelashes and eyebrows were normal, no skeletal abnormalities were clinically or radiologically detected. No cataracts and cancer have been found so far. She had atopic dermatitis and hidradenitis suppurativa, never priviously reported in association with RTS. The diagnosis of RTS was made according to typical lesions and mutation of RECQL4 gene. The patient was

advised to avoid sun exposure and undergo annual checkups for teeth, eyes, skin, and bones.

Poikiloderma occurs in a number of genodermatoses and other syndromes but with a subtly different pattern [3].

Lupus erythematosus. Poikiloderma has been reported as a feature of LE, but it does not appear early and is mainly expressed in the advanced phases of the disease.

Kindler syndrome is a hereditary and bullous syndrome, where poikiloderma arises generally at the age of two – three years. Blistering is induced by trauma and photosensitivity usually starts in early infancy. The patient commonly shows acral bullae, since birth or the first few days of life. The disease resembles epidermolysis bullosa. The blistering improves with age giving place to poikiloderma, mainly involving the face but also spreading to other sites. Additional features include syndactyly, dental abnormalities, stenosis of the oesophagus and anus, ectropion, chronic inflammation of oral mucosa and anhidrosis.

Dyskeratosis congenita is characterised by the tried: abnormal skin pigmentation, nail dystrophy and leukoplakia. The poikiloderma involves the face, neck, trunk and thighs. Its onset - like other clinical manifestations - occurs generally during childhood, though later than in RTS. The nail dystrophies are severe and are the first appearance, before the poikiloderma. Mental retardation is more common than in RTS. There is no photosensitivity.

Key Points

- The diagnosis of RTS is very probable when poikiloderma on the face, spreading to extremities, appeared in early infancy
- Poikiloderma is also a symptom of many systemic diseases, such as Lupus erythematosus, Kindler syndrome, dyskeratosis congenita and other
- Recognition of the age of onset and evolution of poikiloderma may be very useful to differentiate RTS from other syndromes
- The diagnosis of RTS should be made according to typical lesions and mutation of RECQL4 gene.
- The result of genetic testing is instructive and useful for a definitive diagnosis and future procreation guidance for the patient's family
- Genetic counselling should be provided for RTS type II patients and their families, together with a recommendation for cancer surveillance

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Chapter 13
Congenital Small Dimple on the Median Line of the Nose

Merli Yuri, Annalucia Virdi, Marco Adriano Chessa, Valeria Evangelista, and Iria Neri

A mother of two children brought her younger son to visit us for an irritating scalp problem.

After visiting and diagnosing him with psoriasis, our attention was drawn to the older daughter.

The 8 year old girl showed a small dimple with a tuft of hair in the center placed on the median line of the nose. The mother, when asked about it, claimed that she was born like this and that the pediatrician said not to pay attention to it (Figs. 13.1 and 13.2).

Based Upon History and Clinical Appearance, What Is Your Diagnosis?

- Nasal glioma.
- Adnexal tumor
- Dermoid fistula.
- Chickenpox scar
- Aplasia cutis.

Diagnosis: Dermoid Fistula

In the suspicion that it may be a skin sign of a cranial dysraphism we prescribed an MRI.

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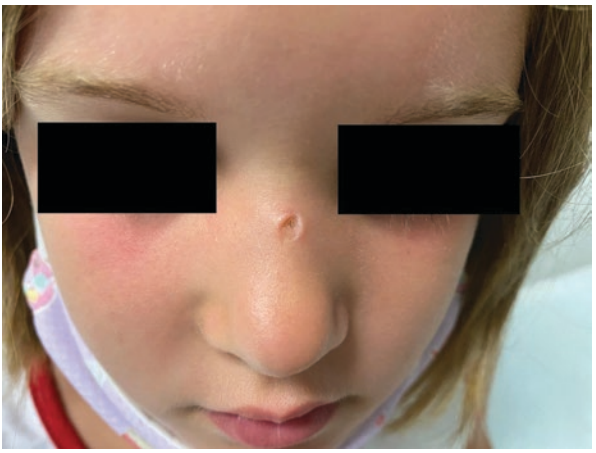

Fig. 13.1 A small dimple with a tuft of hair located on the median line of the nose

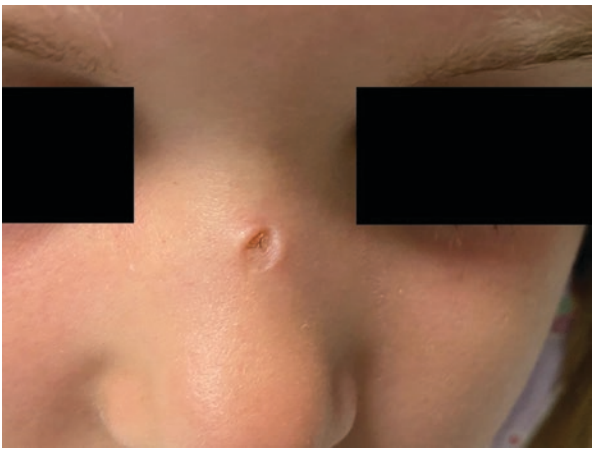

Fig. 13.2 Detail with the base of the dimple and a small tuft of hair in the center

A few weeks later the mother called us to inform us that her daughter has been diagnosed with "dermoid fistula with intracranial extension" and that neurosurgeons have taken over to correct the defect.

Discussion

The term "dysraphism" strictly refers to neural tube closure abnormalities but, conventionally, it also includes median closure abnormalities of the meninges, vertebrae, paravertebral muscles and skin. There is a macro division on the base of the site of the defect in craniofacial and spinal dysraphism. Severe or externalized dysraphism are detected mainly in the prenatal period through fetal ultrasound or, at most, at birth where newborns are immediately tended to by pediatric

neurosurgeons. Mild or occult dysraphism can be diagnosed later, during childhood or adulthood [1, 2]. Congenital malformations due to cranial dysraphism include: dermoid fistula, nasal glioma, cephalocele, aplasia cutis of the scalp and the sign of a "collar" or "tuft of hair". Skin lesions that reveal the presence of a possible cranial dysraphism are generally located on the midline or paramedian of the face and scalp, more specifically in the frontal-nasal area and in the vertex. These lesions may or may not have intracranial communications depending on the type of defect [1, 2]. Dermoid fistulas arise from an abnormal development of the frontal-nasal region with inclusion of the ectoderm during the fusion of the embryonic buds. They can arise anywhere, however the most common region is the head-neck region with most occurrences on the lateral third of the eyebrow, where there is never intracranial extension. On the other hand, when they are located along the median or paramedian axis, they are more frequently found on the nasal crest or at the occipito-temporal junction. In these cases the possibility of an intracranial extension is higher [1, 2].

Clinically they occur in 2 ways:

- Asymptomatic nodule is bluish or yellowish, non-pulsating and non-compressible.
- Isolated millimeter cutaneous dimple located on the line from the glabella to the columella.

The main complications are inflammation or infection. If there is communication with the intracranial structures, there is a great risk of bacterial meningitis which can be fatal in some cases. The first choice for instrumental examination is MRI which allows doctors to delineate the morphology of the lesions and their possible intracranial extension, along with early surgical treatment to avoid deformities and infectious complications [1, 2].

Key Points

- Cranial dysraphisms can manifest themselves with skin signs, even trivial, present from birth or later on, from infancy to adulthood.
- Any congenital skin lesion located in the midline should be suspected of an underlying dysraphic disorder.
- In case of suspicion of dysraphism, an MRI of the skull is the reference examination.

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Chapter 14
Cutaneous Lesions and Multisystem Inflammatory Syndrome in Children (MIS-C)

Kenan Barut, Defne Özkoca, and Zekayi Kutlubay

A 10-year-old male patient applied to the pediatric emergency department with fever, diarrhea and respiratory distress. He has had fever and diarrhea for one week. He was tachypneic, tachycardic and hypotensive upon arrival therefore, he was hospitalized in the pediatrics ward with the diagnosis of Multisystem Inflammatory Syndrome in Children (MIS-C). His vitals were: a body temperature of 38.7 °C, a heart rate of 130 beats per minute and a blood pressure of 70/50 mm mercury. Other systemic examinations revealed hepatosplenomegaly and a rash. His past medical history was unremarkable. His family history revealed that his mother has had SARS-CoV-2 infection one month ago.

The laboratory examination revealed leukocytosis with lymhopenia (white blood cells 16,300/mm^3, lymphocyte 500/mm^3), increased C-reactive protein (156 mg/L), increased b-natriuretic peptide (1963 pg/ml) yet the echocardiography was normal, increased D-dimer (21.94 mg/L) and increased ferritin levels (421 ng/ml). SARS-Cov-2 PCR test was negative but anti-SARS-CoV-2 IgG serology test was positive. The patient was diagnosed with MIS-C by the pediatrics department given the Anti SARS-CoV-2 IgG positivity and the cardiac and gastrointestinal manifestations. Treatment with intravenous immunoglobulin (IVIG), systemic steroids (prednisolone 2 mg/kg/day), low molecular weight heparine (2x0.4 mg/day), combined antibiotherapy with teicoplanin, cephotaxime and amikacine, milrinone, noradrenaline, n-acetyl-cysteine and vitamin-C was initiated immediately upon the diagnosis.

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Non-invasive ventilation with mask was provided for the respiratory distress. The fever, respiratory and cardiac symptoms subsided upon the second day of treatment. The patient fully recovered by the seventh day of treatment.

Dermatology consultation was requested due to the skin findings. The dermatologic examination revealed multiple annular erythematous plaques with central clearing and inner trailing collaret desquamation on the thorax and the abdomen shown in Figs. 14.1 and 14.2. The lesions were pruritic; have been present for several days, started singly and then distributed to the thorax and abdomen.

Based on the Case Description and the Photographs, What Is Your Diagnosis?

- Urticaria
- Pytriasis rosea like lesions due to SARS-CoV-2
- Insect bite
- Erythema multiforme

Diagnosis

The patient was diagnosed as pityriasis rosea like lesions due to SARS-CoV-2. The lesions subsided in a week with the daily use of topical steroid ointment.

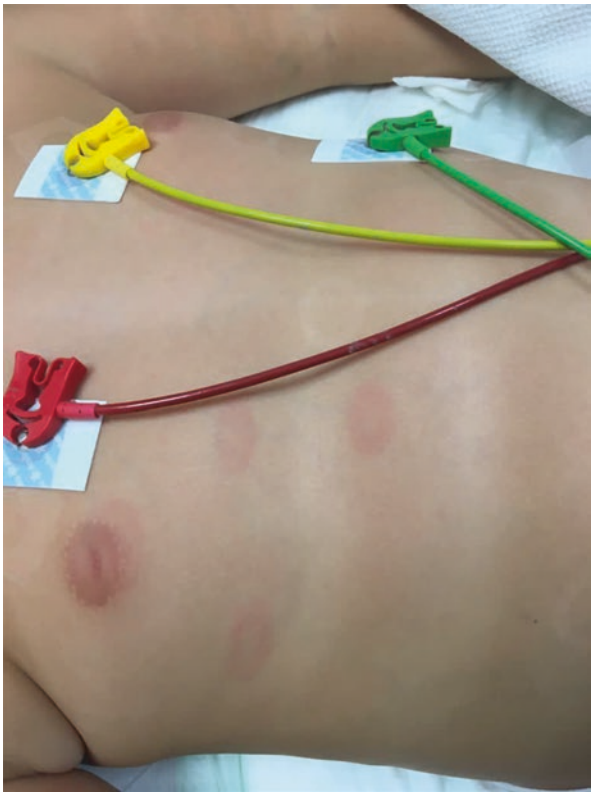

Fig. 14.1 Multiple annular erythematous plaques with central clearing and inner trailing desquamation located on the chest are noticed

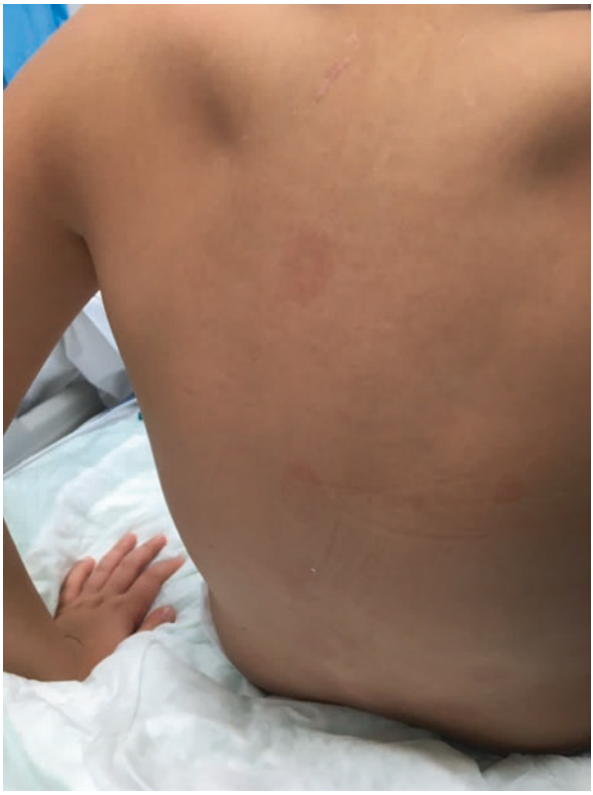

Fig. 14.2 Multiple annular erythematous plaques with central clearing and inner trailing desquamation located on the back are seen

Discussion

The SARS-CoV-2 infection is either asymptomatic or has a mild course in the pediatric population. However, it can have a severe, even fatal, course in minority of cases, which often are admitted to intensive care units. Multisystem inflammatory syndrome (MIS-C) in children is one of these severe presentations [1]. The constellation of fever, severe illness, involvement of two or more organ systems and the laboratory or epidemiologic evidence of SARS-CoV-2 infection is called MIS-C and it resembles Kawasaki disease, toxic shock syndrome and secondary hemophagocytic lymphohistiocytosis in its presentation [2]. The most commonly involved organ system is the gastrointestinal system, thus abdominal pain and diarrhea are very common. Cough and respiratory distress often occur as well [3]. The pathogenesis of MIS-C relies upon post-infectious immune dysregulation [2]. Cutaneous symptoms occur with MIS-C as well [1–3].

Cutaneous manifestations of the SARS-CoV-2 reported in the pediatric population are chilblain-like lesions, erythema multiforme, urticaria, vesicular lesions, morbiliform rash, petechia-purpura, erythematous exanthema and pityriasis rosea like eruptions [3]. The most commonly reported cutaneous manifestation of the

SARS-CoV-2 infection in patients with MIS-C is erythematous exanthema [4]. However, the cutaneous manifestations are nonspecific and any kind of eruption may be seen in patients with MIS-C [5, 6].

Pityriasis rosea has been associated with the infection or re-activation of the human herpes viruses 6 and 7 (HHV-6 and HHV-7). Pityriasis rosea-like eruptions have previously been reported with other viral infections and vaccination. The typical eruption is multiple erythematous annular plaques with collarets of trailing scale distributed in a Christmas tree like pattern along the cleavage lines. Pityriasis rosea-like eruption has been reported in association with the SARS-CoV-2 infection as well, presenting along with gastrointestinal symptoms, similar to the child presented in this case. Topical corticosteroids is sufficient in the treatment of this cutaneous symptom [7].

Key Points

- The constellation of fever, severe illness, involvement of two or more organ systems and the laboratory or epidemiologic evidence of SARS-CoV-2 infection is called MIS-C; a disease that has a cutaneous component as well.
- The most common cutaneous manifestation of MIS-C is erythematous exanthema; however any kind of eruption may be seen.
- Pityriasis rosea like eruption is one of the cutaneous manifestations of the SARS-CoV-2 infection and it has been reported in the pediatric population as well as in the adults.

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Chapter 15
Erythematous Patch on the Antecubital Fossa

Mohamed Saeed Mohamed, Shady M. Ibrahim, and Mohamed L. Elsaie

Case Presentation

A 6 years old boy was seen in the dermatologic clinic due to asymptomatic slightly erythematous patch on his left antecubital fossa (Fig. 15.1). The lesion had history of dating shortly after birth; the patient reported bleeding with minor trauma and no other associated symptoms. There was no relevant familial or medical history.

On physical examination of the skin, he had single erythematous well-defined round patch with central clearing about 4 × 4 cm plaque on his left antecubital fossa. Examination of other areas of the skin, hair, nail and oral cavity showed no abnormality. There is no evidence of apparent lymph node enlargement.

Dermoscopic examination of the patch revealed empty lacunae, decrease erythema, which slightly replaced by brown pigmentation with only residual of capillaries spots are detectable (Fig. 15.2).

Based on the Case Description, Clinical and Dermoscopic Photographs, What Is Your Diagnosis?

1. Hemangioma.
2. Melanocytic nevus.
3. Melanoma.
4. Post inflammatory hyperpigmentation.

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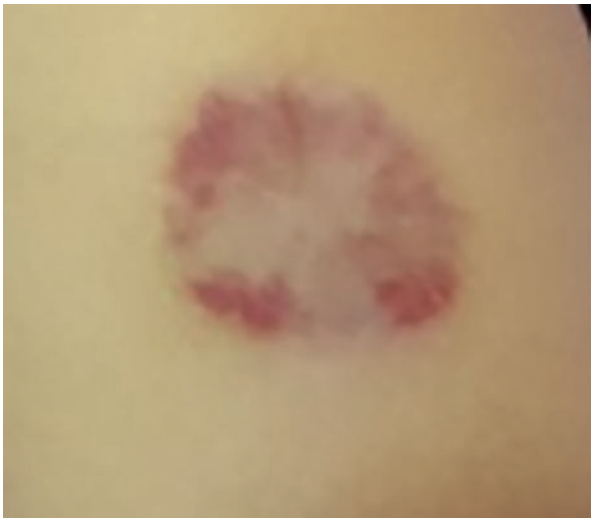

Fig. 15.1 Asymptomatic slightly erythematous patchon the left antecubital fossain male patient 6 years old

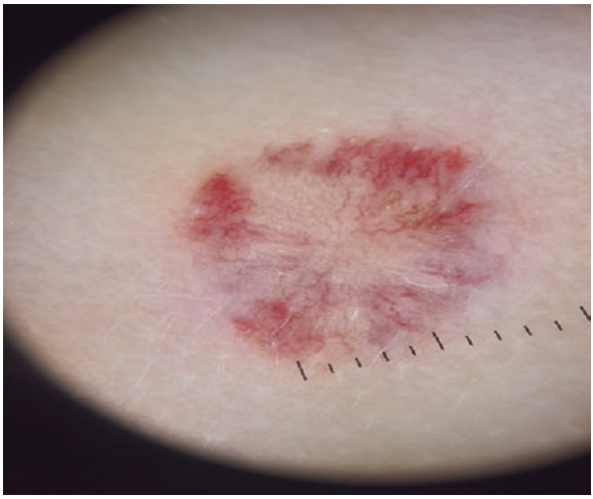

Fig. 15.2 Dermoscopic examination of the patch revealed empty lacunae, decreased erythema, which slightly replaced by brown pigmentation with only residual of capillaries spots (Dermoscopy 3gen DermLite 3, magnification 10×)

Diagnosis

Hemangioma (regression).

Discussion

Infantile hemangiomas (IHs) are the most common benign childhood tumors, characterized by a rapid growth phase (0–12 months) followed by a slow involution phase (5–9 years) [1, 2]. Up to 70% of IHs resolve with problems that can range

from telangiectasia, hyper/hypopigmentation, atrophic/ hypertrophic scars [3, 4]. About 30% to 34% of cases present complications requiring therapy (ulcerations/ hemorrhage/infection), or can be life-threatening (12%–15%) with eye/-mouth/ear functions impairment or organ compression [5, 6].

Dermoscopy of hemangiomas shows lacunae of variable sizes and dilated vessels against a red to reddish blue background [7].

The majority of infantile hemangiomas do not require any medical or surgical intervention. Historically, medical care of clinically significant hemangiomas had been limited to a few medications, including glucocorticosteroids (topical, intralesional, and oral), interferon alfa, and, rarely, vincristine and topical imiquimod [8]. Beta-blockers, most specifically propranolol, have been shown to induce involution of infantile hemangiomas and are now considered first-line treatment for problematic infantile hemangiomas [8]. Laser surgery is beneficial in treating both proliferating and residual vessels from hemangiomas. The flashlamp-pumped pulsed-dye laser has become the most widely used laser for selective ablation of vascular tissue in childhood [9].

Key Points

- Infantile hemangiomas (IHs) are the most common benign childhood tumors**.**
- Dermoscopic evaluation of hemangioma may help to assess the response of treatment**.**
- The majority of infantile hemangiomas do not require any medical or surgical intervention

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Chapter 16
Erythematous Ulcerated Perioral Nodule

Diego Abbenante, Valeria Evangelista, Giulia Veronesi, Alba Guglielmo, and Iria Neri

A 6-year old boy was admitted to the pediatric emergency department complaining of a 5-month history of an asymptomatic erythematous- plaque of the right perioral area. His past medical history was relevant for severe atopic dermatitis. Despite several topical (mupirocine, fusidic acid) and systemic (amoxicillin/clavulanic acid, trimethoprim/sulfamethoxazole) treatments had been used, the lesion did not show any improvement and it continued to increase in size. The physical examination revealed an inflamed plaque of about 3 centimeters with an overlyng crust involving the right perioral area (Fig. 16.1). The remainder of his examination was normal except for a bilateral cervical lymphadenopathy. Dermoscopy highlighted a central yellowish crust surrounded by a white starburst-like area (Fig. 16.2). An ultrasound examination of the lesion was performed and showed a thickening of the subcutaneous tissue containing a deep inhomogeneous hypoechoic area and a small, superficial fluid collection with diffused vascular signals within the lesion. A biopsy sample was taken and histopathology revealed the presence of epidermal hyperplasia and ulceration; a dense, mixed, granulomatous infiltrate and amastigotes within macrophages cytoplasm.

Based on the Case Description and the Photographs, What Is Your Diagnosis?

1. Erysipelas
2. Cellulitis
3. Cutaneous leishmaniosis
4. Granuloma faciale

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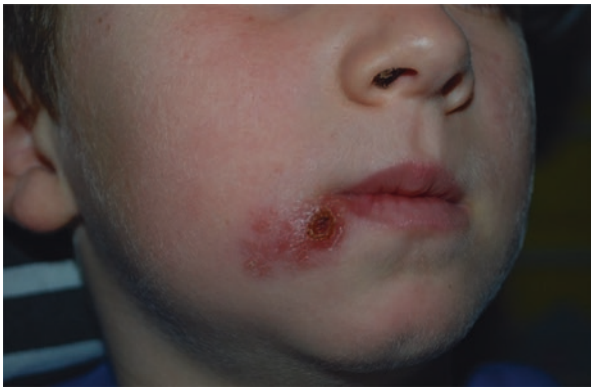

Fig. 16.1 Circumscribed inflammatory erythematous nodule of the perioral area

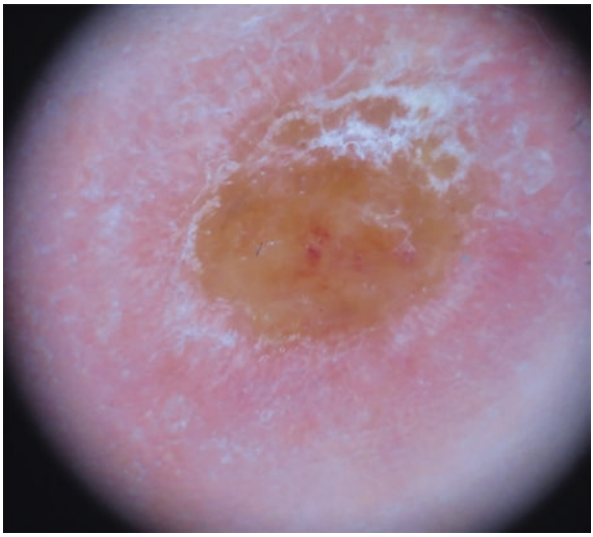

Fig. 16.2 Dermoscopy (×10): central yellow-orange crust surrounded by erythema and white radial strikes

Diagnosis

The lesion completely resolved after two treatments with intralesional Meglumine Antimoniate.

Discussion

Leishmaniasis is a vector-borne disease caused by protozoan parasites of *Leishmania* spp. It can be classified in three different forms: visceral, mucocutaneous and cutaneous (CL). Globally, the annual incidence of CL is estimated to be 0.7 to 1.2 million new cases per year [1].

The protozoa, transmitted by phlemotomes bites, are obligate intracellular parasite and infected dogs represent the reservoir. The incubation period for

symptomatic CL ranges from weeks to months but the infection may have a subclinical course.

CL usually involves the exposed areas and presents with an erythema at the site of bite that usually is asymptomatic. The lesion turns into an infiltrated plaque that progressively enlarges becoming an indolent nodule/plaque with a central erosion and crust and usually heal in a period of months to years leaving an atrophic scar. Satellite lesions can be observed.

Dermoscopy represents a useful noninvasive tool for CL diagnosis [2, 3]. The most frequently observed pattern is a diffuse erythema with some yellowish-orange areas. This pattern is not specific for CL as it can be observed in many other granulomatous disorders such as sarcoidosis, lupus vulgaris and foreign body reaction. At an early stage, yellowish tear-like structures and polymorphous vessels arranged in a radial pattern are detected. Advanced lesions show a central erosion/crust surrounded by white hyperkeratotic projections in a starburst-like configuration. Moreover, dermoscopy can be useful for treatment monitoring as, in case of response, it shows the presence of orange-brown structureless areas.

The diagnosis is based on clinical and dermoscopic findings and frequently requires the detection of typical Leishman-Donovan bodies in skin biopsy. In some cases, PCR identification may be needed [4].

Possible treatments for of uncomplicated CL include cryotherapy, paromomycine ointment, intralesional injections of pentavalent antimony and photodynamic therapy [4, 5].

Key Points

- The diagnosis of CL should be considered in patients with one or more chronic, painless, papulo-nodular skin lesions and a history of exposure in an area where leishmaniasis is endemic.
- Dermoscopy can be useful for a noninvasive diagnosis of CL and for treatment monitoring.
- Local therapy is preferred for treatment of uncomplicated CL.

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Chapter 17
Hair Loss in an 11-year-old

Anneliese Willems and Rodney Sinclair

An 11-year-old female presented with 11 months of progressive hair loss. Her initial symptoms were patches of hair loss in discrete areas throughout the scalp. Four months prior to assessment her symptoms has progressed to almost complete scalp hair loss (Fig. 17.1). Examination revealed associated patchy loss of eye lashes and eyebrows. Trichoscopy to the remaining hair bearing areas revealed micro-exclamation marks.

Based on the Case Description and the Photograph, What Is Your Diagnosis?

- Alopecia areata
- Trichillomania
- Tinea capitis

Diagnosis

Alopecia universalis variant of alopecia areata.

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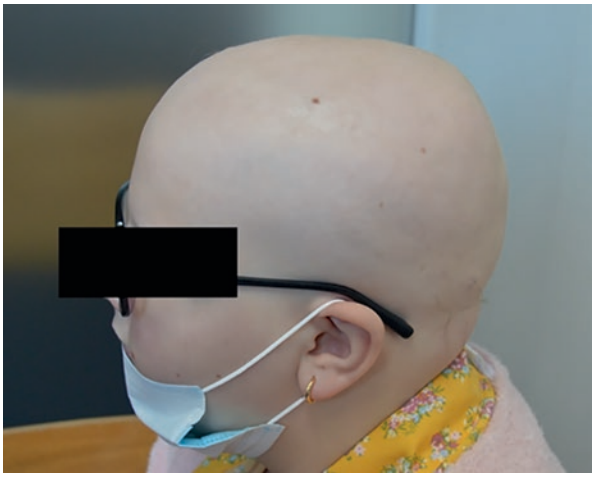

Fig. 17.1 11-year-old female presenting with almost complete scalp hair loss

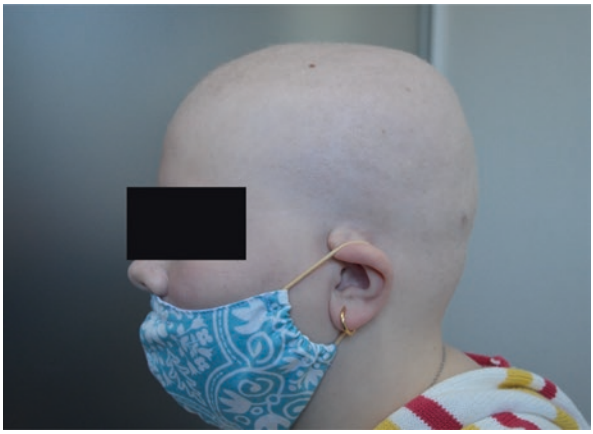

Fig. 17.2 Three weeks after initiation of combination of oral Baricitinib and sublingual minoxidil. Fine vellus hair growth can be seen across the entire scalp

Treatment

The patient was initially unsuccessfully treated with twice daily topical 5% minoxidil and daily use of a low-level laser cap. As both the patient and her parents were experiencing significant associated psychological distress, she was commenced on Baricitinib in a dose of 1.7 mg twice daily for the first month, reducing to 1.7 mg once daily thereafter in combination with sublingual minoxidil 0.3 mg daily. Both medications were well tolerated.

Within three weeks, fine vellus hair regrowth began to appear over her 90% of her scalp (Fig. 17.2). At 5 months (Fig. 17.3), her SALT score was 2 with good regrowth of both eyebrows and eyelashes. Regrowth corresponded with significant improvements to her psychological wellbeing.

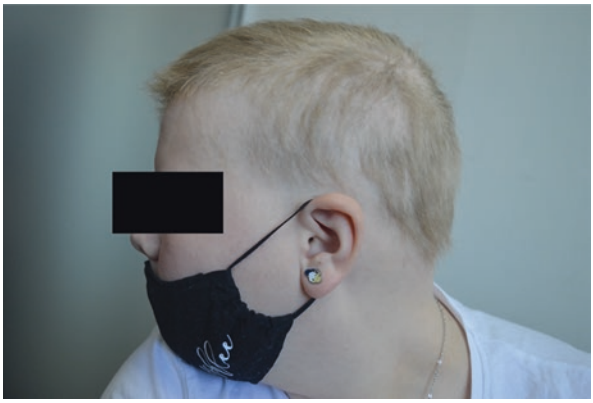

Fig. 17.3 Response to combination of oral Baricitinib and sublingual minoxidil at 5 months

Discussion

Alopecia areata (AA) is an inflammatory immune-mediated disorder resulting in hair loss [1]. Eyebrow, eyelash and nail growth may also be affected [1]. The severity of hair loss varies from an isolated small patch to alopecia totalis (AT), with complete loss of hair to the entire scalp, or alopecia universalis (AU) with entire body alopecia [2]. The natural history is also variable with up to 70% of patient achieving a spontaneous and durable remission within 12 months (acute alopecia areata), while for others the disease course may be protracted with multiple relapses and remissions over many years (chronic alopecia areata) [1, 3, 4].

Childhood onset is common and early age of onset is associated with a worse prognosis [5]. The psychological morbidity associated with AA is variable, however, children and adolescents suffering from AA tend to have higher rates of anxiety and depression [6–10].

There are no systemic treatments approved for the treatment of AA/AU [1]. Recent case reports, case series and genome-wide association studies support the use on Janus Kinase enzyme (JAK) inhibitors in the treatment of AA [11, 12] and a number are currently being investigated in phase 2 and phase 3 clinical trials. Baricitinib is a selective JAK 1/2 enzyme inhibitor, approved for use in the treatment of rheumatoid arthritis [13]. There have been a number of case reports of success with Baricitinib in the treatment of adults with AA/AU [14–16], but there are no reports of it use in children with AA/AU.

Minoxidil has been used in the treatment of alopecia areata for many years, both topically and orally. Fiedler-Weiss et al. reports regrowth in around 20% of patients treated with 5 mg twice daily [17]. High doses were used as minoxidil was assumed to act by stimulating vasodilation and thereby increasing hair follicle perfusion [18] and the dose threshold for any haemodynamic effect of minoxidil is 2.5 mg daily [19]. Recently the ASCT1 neutral amino acid transported in the keratogenous zone of the hair bulb a has been identified as the primary target for minoxidil induced hair growth and lower doses of minoxidil have been shown to produce hair growth

without inducing vasodilation [20]. Sublingual administration of minoxidil bypasses hepatic sulfation of minoxidil SULT2A1, further reducing systemic side-effects and increasing substrate avaiability for hair follicle SULT1A1 conversion to the more active metabolite minoxidil sulfate [21].

In conclusion we found the combination of oral Baricitinib and sublingual minoxidil to be highly effective in the treatment of this child with AA/AU. Further research will best further explore the potential application of these agents in paediatric patients with AA/AU.

Key Points

- There are currently no systemic treatments approved for the treatment of AA/AU
- AA/AU will often first present in childhood
- The psychological and cosmetic burden of AA/AU may be significant
- Oral Baricitinib and sublingual minoxidil are novel potential treatments for paediatric AA/AU sufferers

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Chapter 18
Hyperkeratotic Plaques on the Extensor Extremities and Punctate Palmoplantar Keratoderma in a Boy

Shashank Bhargava and George Kroumpouzos

An 8-year-old boy presented to dermatology with reddish lesions on his extremities that were predominantly observed on the elbows and knees, and thickening of the palms and soles since early childhood (Figs. 18.1, 18.2, and 18.3). The lesions were mainly distributed over the extensor surfaces. They started as erythematous, scaly papules and progressed into hyperkeratotic plaques covering large areas; however, they were not migratory in nature. The patient's developmental milestones were typical. The boy was born out of a non-consanguineous marriage, and none of his family members had similar lesions. Skin examination revealed well-defined, erythematous, hyperkeratotic plaques distributed symmetrically over the dorsal aspect of the fingers and feet, knuckles, elbows, and knees. Hyperkerototic, punctate lesions were present over pressure-prone areas on the soles and palms (Fig. 18.4). Toenails showed pits, and a median canalicular ridge was present on the big toes. The review of systems was unremarkable, and routine serologic parameters were within normal limits.

Based on the Case Description and Photographs, What Is Your Diagnosis?

- Erythrokeratoderma variabilis
- Epidermolytic ichthyosis
- Progressive symmetric erythrokeratoderma
- Olmsted syndrome
- Psoriasis

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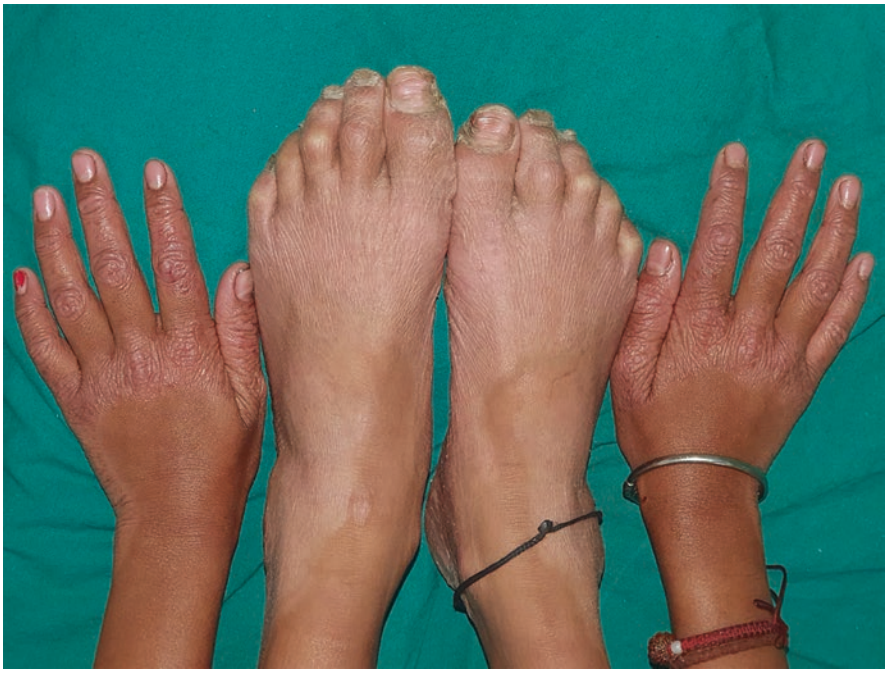

Fig. 18.1 Large hyperkeratotic plaques, almost perfectly symmetrically distributed over the dorsal feet and hands

Diagnosis

Progressive symmetric erythrokeratoderma (PSEK). The diagnosis is supported by the history and clinical findings, including the symmetric involvement of the extensor surfaces of the extremities and non-migratory nature of hyperkeratotic lesions.

Discussion

PSEK is a rare genodermatosis. It is an autosomal disorder of cornification with variable penetrance associated with frameshift mutations in loricrin gene [1]. Positive family history is observed in half of the cases - the rest are due to spontaneous insertional mutations in loricrin gene. PSEK usually manifests in the first decade of life, but cases with adult onset have have been reported [1]. Unlike erythrokeratoderma variabilis (EKV), the closest differential diagnosis, the neonate with PSEK has no skin lesions at birth.

The condition is characterized by well-defined, erythematous, hyperkeratotic plaques affecting the extensor surfaces of the extremities, buttocks, and

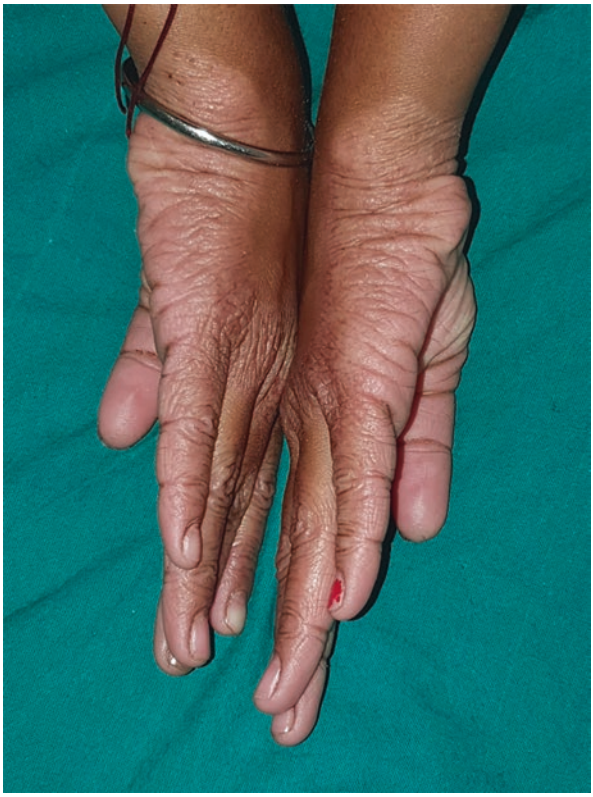

Fig. 18.2 Symmetric hyperkeratotic plaques on the lateral aspect of the hands

occasionally the face; the trunk is typically spared, although lesions on the chest and abdomen have been reported [1–3]. The lesions are almost perfectly symmetric. These hyperkeratotic plaques are slowly progressive, increasing in number and size throughout early childhood before either stabilizing, regressing or disappearing sometime later during life [4, 5]. Waxing and waning may exceptionally occur. Calloused skin on the palms and soles (PPK; palmoplantar keratoderma) is not uncommon, and can be disabling [5].

The histopathological features of PSEK include orthokeratosis, focal parakeratosis, mild or moderate acanthosis, normal or prominent granular layer, and a perivascular lymphocytic infiltrate in the papillary dermis [6]. PSEK is diagnosed based on the history and clinical findings, especially as loricrin gene analysis is difficult in hospital settings [1].

Here, we report a rare presentation of PSEK associated with punctuate palmoplantar keratoderma. We started the patient on topical keratolytics, emollients, and oral acitretin. Topical calcipqotriol has been helpful, but topical retinoids and steroids have shown variable efficacy [3].

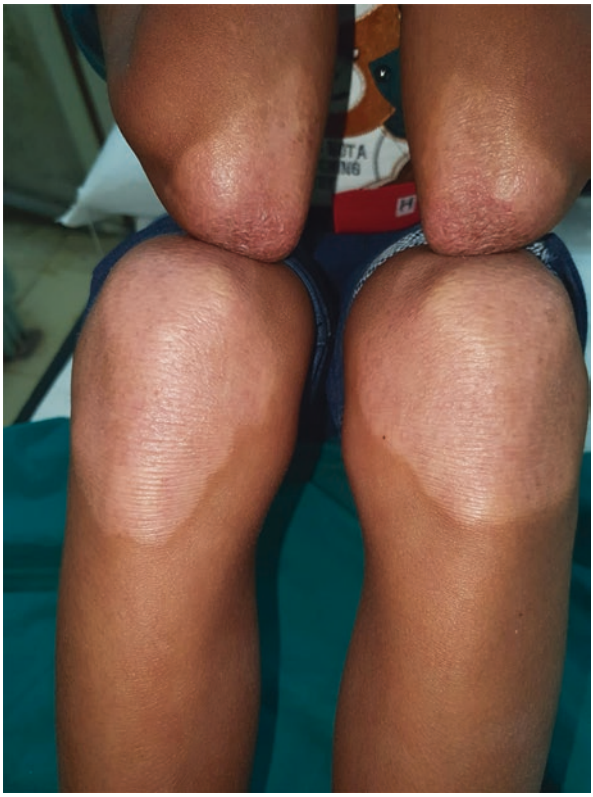

Fig. 18.3 Hypopigmented thickened skin with accentuated skin markings on the elbows and knees

Key Points

- Erythrokeratoderma is a rare group of genodermatoses - the majority of individuals with features of erythrokeratoderma belong to the clinical spectrum of EKV or PSEK.
- PSEK typically presents with symmetrically distributed, non-migratory, large, well-defined, erythematous, hyperkeratotic plaques. The lesions are strikingly symmetrical and affect predominantly the extensor surfaces of the extremities and sometimes the face.
- PSEK usually presents in early childhood and, unlike EKV, not at birth.
- Punctate type of PPK was noted in our patient - this is an exceptional association of PSEK.

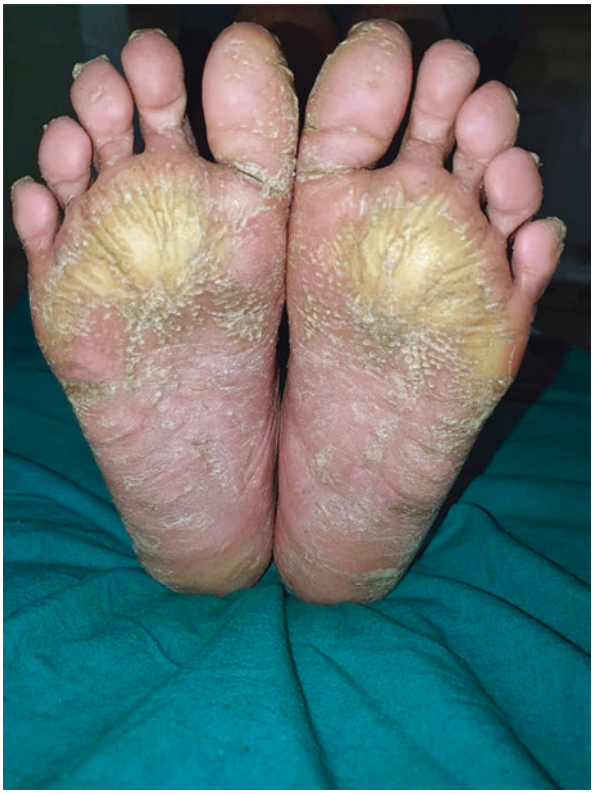

Fig. 18.4 Punctate keratoderma is noted on the soles

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Chapter 19
Hypopigmented Mycosis Fungoides in Pediatric: A Case Report

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A 6-year-old boy with non-itchy white spots on the face, trunk, and extremities of 4 months' duration. No history of drug and food allergies nor atopy in the patient and family. There were multiple macules and hypopigmented patches with fine scales (Fig. 19.1a). No significant improvements were seen after application of hydrocortisone and desonide cream.

Based on the case description and the photographs, what is your diagnosis?

1. Pityriasis alba
2. Postinflammatory hypopigmentation
3. Pityriasis lichenoides chronica
4. Mycosis fungoides

Diagnosis

Histopathological findings from skin biopsy revealed acanthosis, parakeratosis, epidermotropism, Pautrier microabscess, lymphohistiocytic infiltrates at the dermoepidermal junction. Immunohistochemical (IHC) staining was positive for CD3, CD8, granzyme, CD56 and negative for CD20 and perforin.

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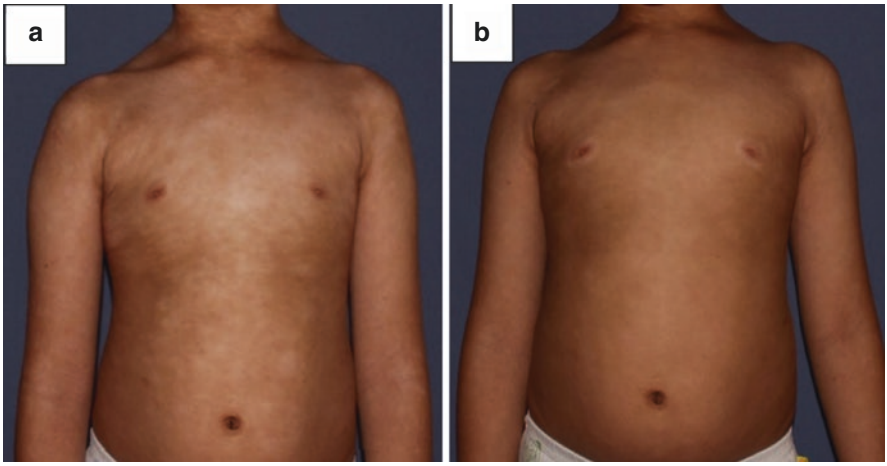

Fig. 19.1 Multiple hypopigmented macules and patches with fine scales. (**a**) Before phototherapy. (**b**) After 2 months of phototherapy

The diagnosis of hypopigmented MF was sought. Further evaluation for staging showed no involvement of internal organs, lymph nodes, and blood. He was given whole-body narrowband UVB (NB-UVB) phototherapy, moisturizer, and 0.025% fluocinolone acetonide cream twice daily on the patches. There was significant clinical improvement after 2 months of phototherapy (Fig. 19.1b).

Discussion

Hypopigmented MF was more common in Asian and dark-skinned patients [1, 2] and more frequently in pediatric patients than adults [3], as was seen in our patient. Zackheim and McCalmont refer to MF as 'the great imitator', a description previously given to syphilis [4]. The diagnosis of MF is often difficult to establish, because of the variation on clinical features and the differential diagnosis includes a variety of benign inflammatory skin disorders. The color of MF lesions can be pink to orange, purplish red, brownish, and less commonly, hypopigmented [1]. Hypopigmented lesions can mimic various skin disorders such as pityriasis versicolor, pityriasis alba, vitiligo, post-inflammatory hypopigmentation, progressive macular hypomelanosis, and pityriasis lichenoides chronica [1, 3, 5], similar to our patient. Hypopigmented MF lesions range from hypopigmentation to acromia, sometimes resembling vitiligo, with distribution mainly on the body and proximal parts of the limbs (especially buttocks, pelvis) and the lower leg [2].

Histopathological examination is important in diagnosing hypopigmented MF. Early patch stage of MF was characterized by epidermotropism with intraepidermal Pautrier microabscesses [3]. Fibroplasia is often absent and epidermal hyperplasia with interface dermatitis is common [1–3], which were found in our patient. Immunohistochemical staining is a key for MF diagnosis because it can

help in determining the proliferation of T cells. An increase of the CD4+/CD8+ ratio MF is usually marked [2, 3]. Phenotypes of T cells which are often identified are CD2+, CD3+, CD4+, CD5+, and CD8–, with reduced or absent of CD7 expression [3]. These features are consistent with findings in our patient. The distinctive feature of hypopigmented MF which can differentiate it from other MF types is the dominance of the CD8+ cell population, as we also found in our patient [1–3]. Damage to melanocytes due to infiltration of cytotoxic T cells that undergo neoplasia was the underlying cause of hypopigmentation. These tumor cells cause degeneration of melanocytes, abnormal melanogenesis, and disruption in melanosome transfer [2].

Immunophenotyping and TCR gene rearrangement can also help diagnosis, but they are rarely done for daily practice [2, 5]. Overall prognosis of MF depends on the extent of the disease and hypopigmented MF is associated with a better prognosis than other MF types [1]. Hypopigmented MF responds mostly to phototherapy, and lesion repair is usually characterized by repigmentation [3, 5].

Key Points

- Hypopigmented mycosis fungoides is a clinical variation of cutaneous T cell lymphoma which is more frequently found in children than adult patients.
- Lesions can mimic various skin disorders such as pityriasis versicolor, pityriasis alba, vitiligo, post-inflammatory hypopigmentation, progressive macular hypomelanosis, and pityriasis lichenoides chronica.
- Histopathology and immunohistochemical staining are necessary to confirm the diagnosis of hypopigmented mycosis fungoides.

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Chapter 20
Itching and Edematous Lesion of the Fifth Toe After Caribbean Holiday

Maria Manta and Fabio Arcangeli

A 6 year-old girl, in good health and vaccinated, came to the office for an itching on the left foot where her mother noted a small erythematous edematous lesion on the fifth toe. She thought it was an insect bite from her recent Caribbean holiday.

The itch continued for several days, disturbing her sleep. The original lesion became red and spread to the plantar surface of the foot. In the next few days the lesions extended to covering most of the plantar region and the itching increased (Figs. 20.1 and 20.2).

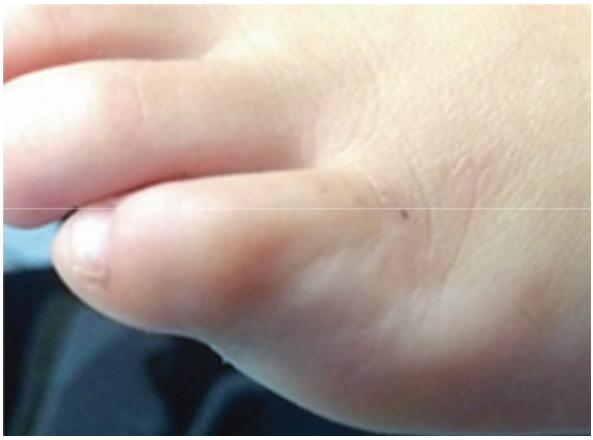

Fig. 20.1 A small erythematous edematous lesion on the fifth left toe

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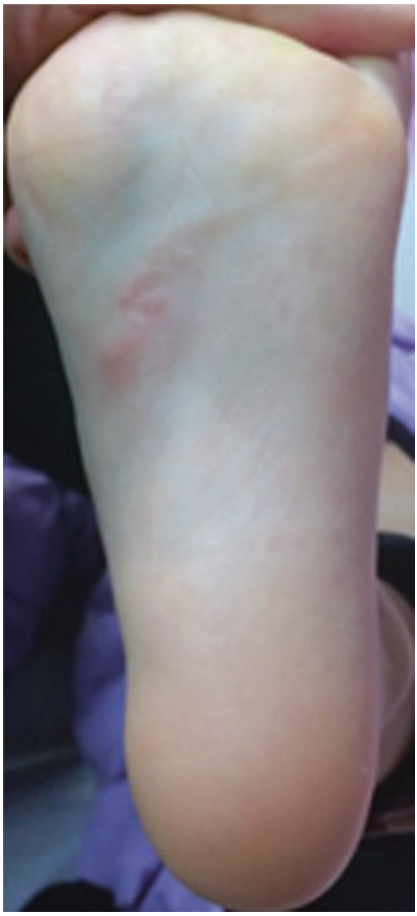

Fig. 20.2 Erythematous lesions spreading on the plantar surface

Based on the history and the photographs, which diagnosis would you propose?

1. Scabies
2. Allergic contact dermatitis
3. Tinea pedis
4. Larva migrans

Diagnosis

Larva migrans

Discussion

- Scabies is a parasite disease, characterized by severe nocturnal itching, which often involves different members of the family or community. Usually it presents lesions—such as papules and scabs—located on typical sites: wrists, interdigital hand folds, perigenital and axillary regions, without involvement—except for young children—of the plantar regions.
- Allergic contact dermatitis presents with erythematous lesions, sweating and scabs, on typical sites depending on the nature of the contact. Allergic contact dermatitis on the feet is almost always on both. The itch is usually irritating but rarely disturbs sleep.
- Tinea pedis shows erythematous and vesicular lesions, at times confluent in blisters, located in the folds, on the plantar feet. Sometimes it extends on the dorsal regions with well defined and curved margins. The itching is generally mild.

Larva migrans is a zoonotic infestation caused by penetration and migration in the epidermis of nematode's larvae (mainly *Ancylostoma braziliense*) which have biological cycles in the intestine of different kinds of animals (dogs, cats, farms and wild animals). This kind of infection is endemic in tropical and subtropical areas of Africa, South America, the Caribbean area and the South east of Asia. Cutaneous larva migrans is one of the most common skin diseases reported in travelers returning from tropical regions.

Nematodes larvae can be found on sandy beaches or on land. When humans are walking or lying down, larvae can penetrate the skin. Adult nematodes live in the animal intestine and their eggs are expelled in faeces. When eggs open larvae can infest sandy beaches or land. Humans who are walking or lying down can be penetrated by larvae in their skin. Parasites can not penetrate dermis and can not complete its biological cycle so that the infection resolves itself in a few weeks or months [1].

Larvae migrate in the skin by moving 2 cm/day, producing an inflammatory reaction that leaves an erythematous and itchy trace, with a serpiginous track [2].

The sites most affected are those that most easily come into contact with the infected sand (sole of the foot, palm of the hand and buttocks).

Clinically, cutaneous larva migrans is characterized by an itching erythematous track which is a migrating tortuous line. The diagnosis is mainly clinical, based on the history of travel to an endemic area and exposure to contaminated sand/soil and the characteristic serpiginous track.

Cutaneous larva migrans is self-limited, but treatment is often necessary to contain symptoms and any bacterial superinfection. Sometimes the psychological aspect must also be taken into account as often the patient or family does not accept the presence of the larva under the skin.Treatment options include a single oral dose of albendazole or ivermectin, topical thiabendazole and cryotherapy with liquid nitrogen [3].

Our patient was fine after a single application of liquid nitrogen a few millimeters ahead of the front of the larva.

Key Points

- Cutaneous larva migrans is one of the most common skin diseases reported in travelers returning from tropical regions.
- Classic cutaneous larva migrans presents with a unilateral, serpiginous, itching eruption on the hands, feet, or buttocks following direct contact with contaminated sand or soil
- Cutaneous larva migrans is self healing but therapy is often necessary due to intense itching
- In case of cryotherapy liquid nitrogen must be applied a few millimeters ahead of the front of the larva

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Chapter 21
Longitudinal Melanonychia in Pediatric Patient

Silvani Geani, Yuri Widia, Irmadita Citrashanty, Sawitri, Iskandar Zulkarnain, and Astindari

A healthy 9 years-old girl patient with an asymptomatic linear brown to black discoloration on her nail (Fig. 21.1). This discoloration has been present for about 9 months and has not changed in thickness or pigment intensity. There was no personal or family history of melanoma. History of trauma, medication or darkening spots on another part of the body was denied.

Based on the case description and the photographs, what is your diagnosis?

1. Longitudinal melanonychia caused by melanocytic nevi.
2. Nail melanoma.
3. Iatrogenic melanonychia.
4. Peutz-Jegher and Touraine syndrome characterized by LM.
5. Nutritional deficiencies.

On examination we observed a homogeneous black color band of the fourth fingernail of the right hand and bright brown band of the second toenail of the left foot. Dermoscopy investigation on fingernail revealed black, homogeneous, narrow and regular longitudinal lines in the nail plate, width 2 mm with pseudo-Hutchinson's sign. On toenail revealed bright brown, homogeneous, narrow and regular longitudinal lines with no Hutchinson sign (Fig. 21.2).

Diagnosis: longitudinal melanonychia caused by melanocytic nevi.

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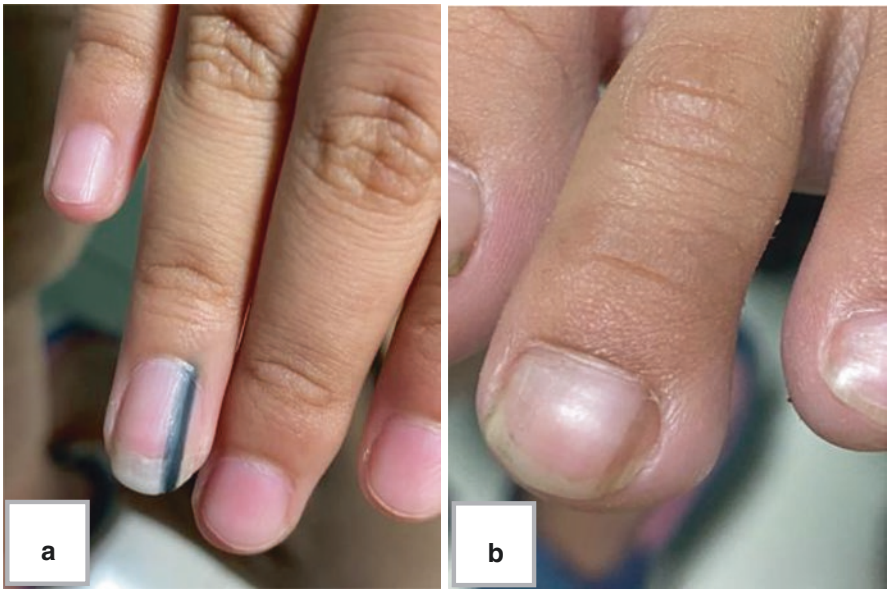

Fig. 21.1 (**a**) Longitudinal melanonychia of the fourth digit of right hand, and (**b**) second digit of left foot

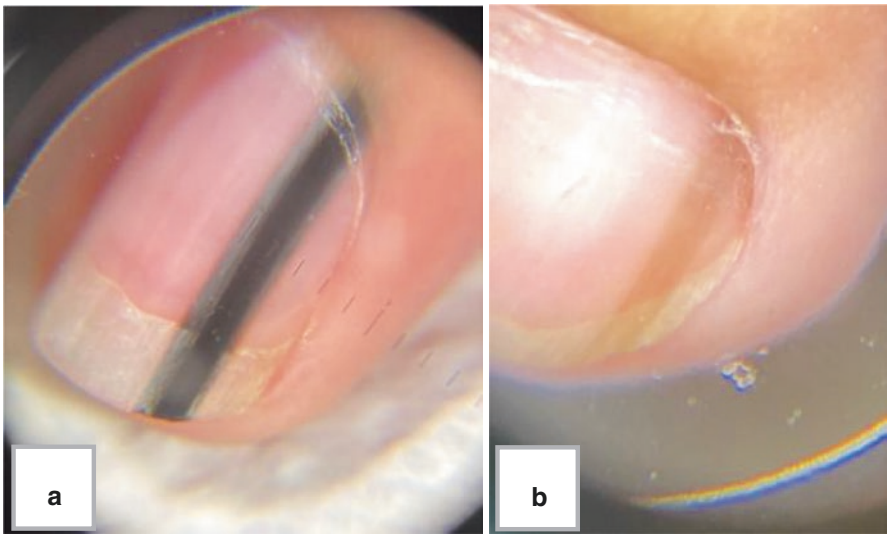

Fig. 21.2 (**a**) Dermoscopy on fingernail showing black homogeneous linear pigmentation and pigment visible through cuticle and proximal nail fold (Pseudo-Hutchinson's sign). (**b**) Bright brown homogeneous linear pigmentation on toenail

Treatment: No therapy was given to the patient. We advise the parent to monitor the changes of the lesions including increasing width or darkening of the lesion and around the nail, or other abnormality of the nail plate.

Discussion

Melanonychia represents a brown to black discoloration of the nail plate that may be induced by benign or malignant cause [1]. It most frequently presents as a longitudinal melanonychia, whereas transversal and total melanonychia are rarely [1, 2]. Almost all cases of LM in children are benign; most are due to melanocytic nevi or lentigos [2, 3]. Melanocytic nevi are the most common cause of melanonychia in children [4]. Clinical evaluation, which includes the use of the ABCDEF rule has limitations to distinguish benign and malignant lesions [3]. Therefore, dermatologists should use additional tools (dermoscopy) to increase the diagnostic accuracy, it can reduce unnecessary and invasive nail unit biopsy [3, 4]. The criteria of ABCDEF rule for our patient includes negative prognostic factors such as age between 50 and 60 years old, Asian ethnicity, presence of a band less than 3 mm, brown or black pigmentation, negative rapid change in pigmentation, multiple digit, thumb/hallux not involved, negative of Hutchinson's sign, and a negative family history. Pseudo-Hutchinson's sign is pigmentation observed through the distal portion of the proximal nail fold and cuticle, for transparency. It is commonly seen in benign lesions [3]. Regular or irregular lines in dermoscopy are classified according to parallelism, spacing, thickness, and color. Regular lines mean benign pole and irregular lines mean malignant pole [3]. LM in children should be followed every 3–6 months, with periodic clinical, photographic and dermoscopy evaluation [3, 5]. If there are no changes, "wait and see" is the best option [3]. It is recommended to excise all bands as a preventative measure after puberty [2].

Key Points

- Longitudinal melanonychia in children are probably benign and most are caused by melanocytic nevi.
- Dermoscopy is a useful tool for diagnosing LM and can reduce unnecessary and invasive nail unit biopsy.
- A "wait and see" is the best option for managing melanonychia in children.

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Chapter 22
Nail Dystrophy in a Young Boy

Torello M. Lotti and Fabio Arcangeli

A 11-year-old boy presented with a nail dystrophy of the right index finger that had started about 18 months earlier and had progressively worsened.

There was no previous trauma in his medical history. The boy always had good health. With the exception of discomfort during manual activities, no other symptoms were reported (Fig. 22.1).

Based on the clinical history and the photograph which is your diagnosis ?

1. Onychomycosis
2. Injury distrophy
3. Nail lichen planus (nail pterygium)
4. Nail psoriasis

Diagnosis

Nail lichen planus (nail pterygium)

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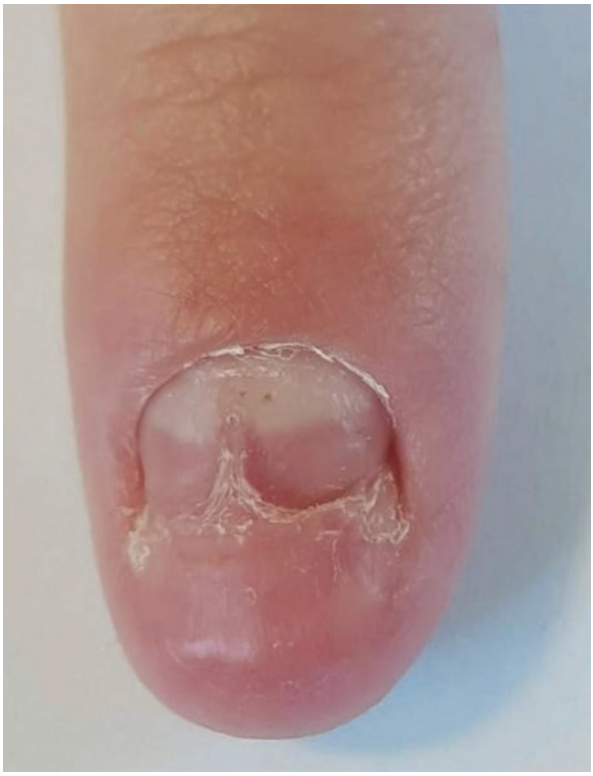

Fig. 22.1 Extension of the proximal nail fold over the nail plate creating split portions on the right index finger nail plate

Discussion

The case we presented consists of a nail deformity called "Pterygium unguis". In this condition the cuticle grows distally, distorting the proximal nail fold. It is typically caused by scarring of the nail matrix. It may occur in the central portion, resulting in the nail splitting or progress to complete nail loss. Pterygium unguis is mostly seen in patients with lichen planus, but it has also been reported in other conditions such as bullous diseases, toxic epidermal necrolysis, burns and leprosy.

In our patient associated oral lichen planus was detected upon physical examination and laboratory tests show auto-immune thyroiditis.

Onychomycosis Fungal infection is the most common disease in adult ungual disorders, but it is very rare in childhood. Toenails are much more likely than fingernails to be infected. The physical examination usually shows thickening and yellowish discoloration of the nail plate, distally or proximally. Formal diagnosis can be made by micro-biological tests.

Injury Distrophy Sometimes an important injury of the nail matrix can cause a pterygium or an acquired anonychia. In our patient no trauma was reported.

Nail Psoriasis Up to 90% of patients with psoriasis will develop nail alterations. They tend to be less common in children. An early detection of nail psoriasis is very important because it is strongly associated with psoriatic arthritis. Clinically we can find pitting, leuconychia, crumbling and red dots in the lunula when the nail matrix is involved. When the nail bed is affected it produces oil drop patch or salmon patch, onycholysis, nail-bed hyperkeratosis and splinter haemorrhages. Pterygium unguis has not been reported in psoriatic patients.

Lichen planus (LP) is an inflammatory disease which can affect skin, mucous membranes, hair follicles, and/or nails. The nail unit involvement in the course of LP was reported by Corsi [1] in 1937 and subsequently characterized by Samman in 1961 [2].

Nail abnormalities are estimated to occur in up to 10% of LP cases [2]. The muco-cutaneous lesions of LP may either precede or develop after the onset of the nail disease. They are usually mild and not related to the severity of the nail disease [3]. The typical clinical features of nail lichen planus (NLP) are nail plate thinning with longitudinal ridging and fissuring. Pterygium formation is usually observed in the advanced stage of nail involvement. Other nail abnormalities due to the matrix lesions may include trachyonychia, atrophy or scarring of the nail unit. The nail bed involvement usually produces onycholysis, hyperkeratosis, dyschromia and plate fragmentation [3, 4].

Although nail biopsy remains the gold standard for the diagnosis of NLP, the presence of pterygium, the most specific feature of NLP, enables diagnosis without the need of histological examination.

NLP affects mostly children, up to 40% [5]. Some authors have noted that psychological stress occurred in about 25% of cases [5].

Data from literature points at more common pterygium formation in patients with isolated NLP—18–75% [3, 5]. Isolated NLP affects primarily the fingernails rather than toenails and affects males more [3, 4].

NLP considerably reduces the quality of life due to impaired manual activity and cosmetic discomfort [6].

No curative therapy is known and the treatment of NLP is challenging due to the lack of treatment guidelines. Topical corticosteroids are commonly considered as first line treatment for NLP. When the disease involves up to three nails, an intralesional steroid injection is proposed. The literature reports other possible therapies such as topical tacrolimus, systemic corticosteroids, etanercept, methotrexate, retinoids and antimalarials.

Some authors recommended early diagnosis of NLP and an early start of systemic therapy with alitretinoin to prevent the development of pterygium and permanent nail damage [6].

Frequent relapses are common. Auto-immune hypo-thyroidism may contribute to the development of LP with a possible underlying associated mechanism [7].

Key Points

1. Pterygium is the most specific and almost pathognomonic finding of NLP
2. Biopsy is not necessary for the diagnosis of NLP when pterygium is present

3. NLP is more common in children and often occurs after psychological stress
4. The muco-cutaneous lesions of LP may either precede or develop after the onset of nail disease
5. No curative therapy is known but topical, intralesional and systemic corticosteroids are frequently proposed

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Chapter 23
Neurofibromatosis Type 1 with Congenital Pseudarthrosis Tibialis

Raden Mohamad Rendy Ariezal Effendi, Reiva Farah Dwiyana, Srie Prihianti Gondokaryono, and Inne Arline Diana

An 11-year-old girl presented to the hospital with her parents complaining of painless small tumors throughout the body and non pruritic multiple hyperpigmented macules on neck, trunk, and axilla. She also had painless skeletal abnormalities on left tibia that made her difficult to walk. The skin lesions started as hyperpigmented macules that appeared at birth, followed by skin-colored tumors that first appeared on her arms 5 years ago and gradually spread on almost the entire body (Fig. 23.1). There is no history of similar skin conditions in the family.

Based on the case description and the photograph, what is the possible diagnosis for the patient?

1. Legius syndrome
2. Neurofibromatosis type 1
3. Neurofibromatosis type 1-like syndrome
4. Neurofibromatosis type 2

Patient had a problem getting along with other children because she felt embarrassed with her disease. For this complaint, the patient was consulted to a psychologist and was suggested to improve personality and social interactions. There was no history of seizure and learning problems.

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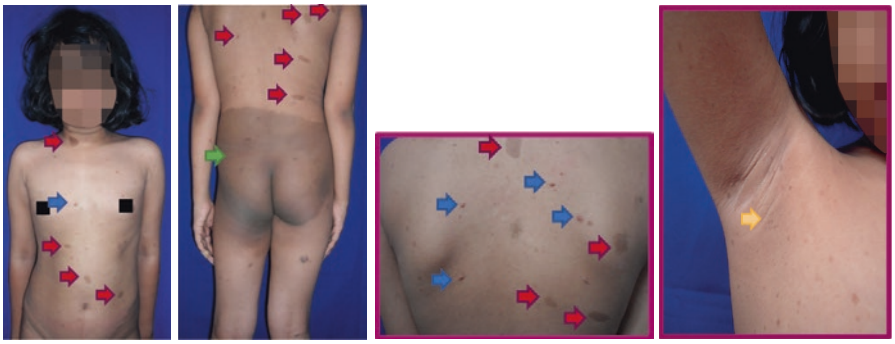

Fig. 23.1 Red arrow, café au lait macules; yellow arrow, Crowe's sign; green arrow, plexiform neurofibroma; blue arrow, neurofibroma

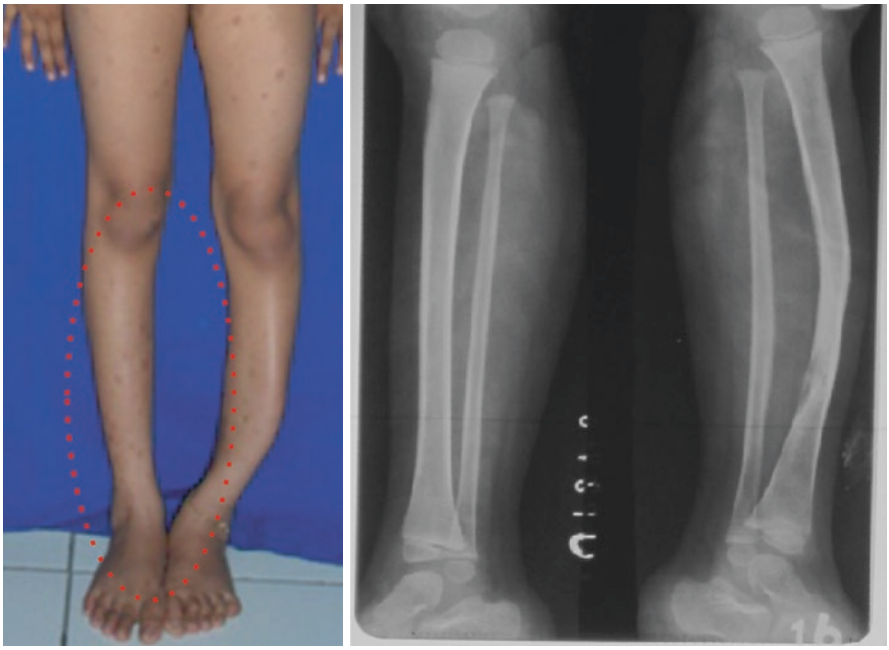

Fig. 23.2 Congenital pseudoarthrosis of left tibia

She had a history of visual problems since the past year. She was then referred to ophthalmology, pediatric, and orthopaedic surgery departments to look for other extracutaneous involvements. There was astigmatism myopia on both eyes and congenital pseudarthrosis of the left tibia (Fig. 23.2). Both parents did not give consent to do skin biopsy for histopathological examination.

Diagnosis

Neurofibromatosis type 1

Discussion

Neurofibromatosis (NF) is an autosomal dominant disorder characterized by benign tumor originating from the nerve sheath with varying clinical features [1, 2]. NF occurs due to mutation in the NF1 gene located on chromosome 17q11.2 and encodes for the protein neurofibromin [1]. This disease encompasses three distinct disorders: NF1, NF2, and schwannomatosis [3]. NF most often found in childhood, characterized by specific cutaneous features and alterations of the several organ and bone deformities [2, 3]. The typical picture of a cutaneous skin-colored tumor (neurofibroma) protrude just above the skin surface, softer than surrounding connective tissue, when pressed will invaginate into the skin or known as the "buttonholing" sensation, and can occur anywhere on the body [1, 2]. Other skin disorders in NF are café-au-lait macules which are flat, often the first manifestation of NF1 to appear and become more numerous both in size and amount as the infant grows [2, 4]. Another typical lesion on NF is axillary freckling or called Crowe's sign, which is a hyperpigmented macule measuring less than 5 mm and is the most specific sign of NF1 patients [1, 2]. About 80% freckling is found in the axilla, 1 but can also be found in other body folds. Plexiform neurofibroma (PN) is often associated with overlying hyperpigmentation. The affected skin may also appear thickened [1, 2].

Extracutaneous manifestations in NF1 have the ability to affect every organ system, including bone abnormalities, especially regarding long bones and are usually found in childhood [2]. Another complication in the bones of patients with NF1 and the most difficult to treat medically is congenital pseudoarthrosis (CPT). Pseudoarthrosis shows a bone abnormality in the form of 'false joint', unilateral deformities which can affect the tibia, radius, ulna, or long bones [1–3].

One type of this disease is NF1 which is established based on the classification and diagnosis criteria by the National Institutes of Health. The seven diagnostic features are: six or more café-au-lait macules larger than 5 mm in greatest diameter in prepubertal individuals, and larger than 15 mm in greatest diameter in post puberty individual; two or more neurofibromas of any type or 1 plexiform neurofibroma; freckling in the axillary or inguinal regions ; optic glioma; two or more iris Lisch nodules; a distinctive osseous lesions such as sphenoid dysplasia or thinning of long bone cortex with or without pseudoarthrosis; a first degree relative with NF1 by above criteria. Patients must meet 2 or more of the listed criteria [2].

Diagnosis NF1 in this patient established based on clinical appearance of more than six cafe-au-lait macules on the neck, trunk, and both upper and lower extremities, intertriginous freckling, neurofibromas in almost entire body, plexiform neurofibroma from flank to the bottocks. The complication as CPT on her left leg was

present with intact tibial bone and mild anterolateral angulation on radiographic examination. The lesions of cutaneous neurofibromas were treated by cryosurgery procedure within interval 2–4 weeks for a year, that resulted in smaller size of neurofibromas and remaining hypopigmented macules, as well as disappearance of some small lesions. There were no specific treatments for CPT in this patient and still observed by the orthopedic department.

Key Points

- Neurofibromatosis type-1 patients are predisposed to developing abnormalities in a number of body systems that can have a significant impact on quality of life.
- Further observation and comprehensive multidisciplinary approach are needed in management neurofibromatosis type-1.

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Chapter 24
Non-segmental Vitiligo in 10 Years Old Child

Rina Gustia and Miranda Ashar

A 10 years old boy patient was referred to outpatient clinic of Dermatology and Venereology Dr. M. Djamil Hospital Padang with chief complaint milky white macules that does not feel itchy and numbness under the right eyelid, bridge of the nose, right nostril, under the right nostril, above the upper lip, above the corner of the right upper lip since 2 weeks ago (Fig. 24.1).

Initially, ±5 months ago, milky white macules that did not feel itchy and numbness appeared as big as corn kernels on the upper lip. The previous appearance of white macules was not preceded by red patches. The milky white macules was increased and got wider to the upper lip, under the right eyelid, under the right nostril, bridge of the right nose, and above the corner of the right lip. The patient likes to scratch the milky white macules under the right nostril, so that the milky white macules under the right nostril is getting wider in 2 weeks. The patient likes to play ball with his friends under the sun. The patient does not use sunscreen or a hat to protect his face. None of the family members experienced milky white patches that did not itch and did not feel numb like the patient.

On examination, we found milky white macules that do not feel itchy and numbness under the right eyelid, bridge of the nose, right nostril, under the right nostril, above the upper lip, above the corner of the right upper lip. The Koebner phenomenon was positive. The patient's skin type according to Fitzpatrick was type IV, the VASI score was 0.5 and the DLQI score was 2 (small effect). On examination of the

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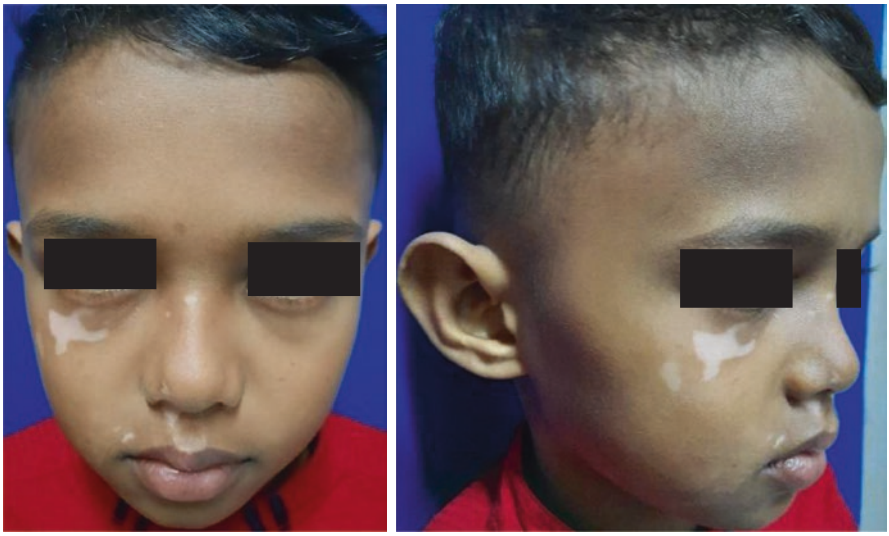

Fig. 24.1 Milky white patches macules under the right eyelid, bridge of the nose, right nostril, under the right nostril, above the upper lip, above the corner of the right upper lip

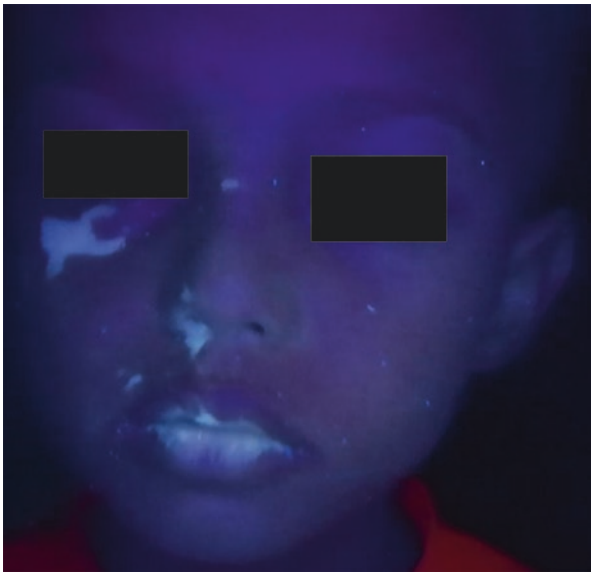

Fig. 24.2 There was a milky white fluorescence under the right eyelid, the bridge of the nose, the right nostril, under the right nostril, above the upper lip, and above the corner of the right upper lip

wood lamp, there was a milky white fluorescence under the right eyelid, the bridge of the nose, the right nostril, under the right nostril, above the upper lip, and above the corner of the right upper lip (Fig. 24.2). On dermoscopy, we found milky white macules with defined borders, perifollicular depigmentation, and leukotrichia (dark circles) (Fig. 24.3). On laboratory examination, the results obtained T3 2.29 nmol/L, T4 112 nmol/L, TSH 1.17 μIU/mL, and ANA profile was negative.

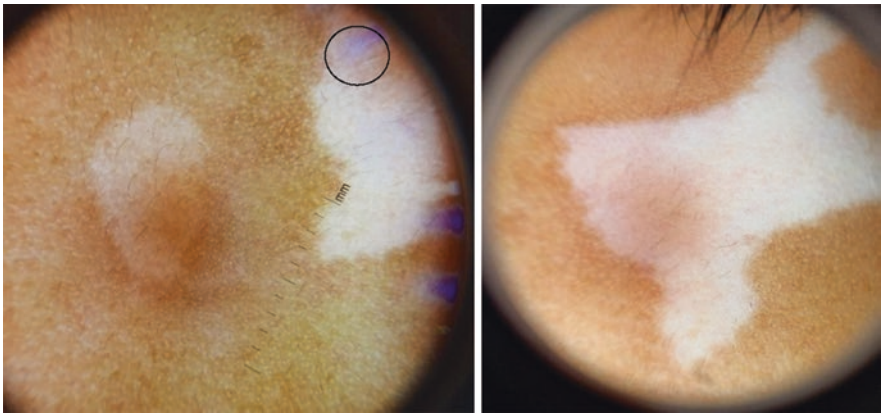

Fig. 24.3 There were milky white macules with defined borders, perifollicular depigmentation, and leukotrichia (dark circles)

Based on the case description and photograph, what is your diagnosis?

1. Non-segmental vitiligo
2. Post traumatic hypopigmentation

Based on medical history, physical examination, wood's lamp, and dermoscopy, diagnosis for this patient is Non-segmental vitiligo.

Treatment: we started mometasone furoate cream 0.1% twice daily on milky white macules for 3 months and wore sunscreen with SPF 30, applied 30 min before going out or exposed to the sun. Patients after using mometasone furoate cream 0.1% for 3 months still have not seen any improvement. In accordance with the algorithm of vitiligo management in children, the patient was given tacrolimus ointment 0.1% for 6 months.

Discussion

Vitiligo is characterized by asymptomatic milky white macules, possibly localized or generalized. Vitiligo in childhood is categorized as segmental and non-segmental. Segmental vitiligo (SV) is a depigmented macule along a dermatomal or quasi dermatomal pattern, without crossing the midline. Non-segmental vitiligo includes local and generalized variants [1].

Various therapeutic modalities are available for the treatment of vitiligo. However, all of these therapeutic modalities cannot be used in children. The modalities of vitiligo therapy in children consist of medical therapy (topical and systemic therapy), phototherapy, surgical therapy, cosmetic camouflage and depigmentation. Moderate potent topical corticosteroids are first-line therapy for children with localized vitiligo. Although high potency steroids are more effective in vitiligo, they are not recommended for use in children [2].

The use of sunscreen is also recommended for children who have vitiligo. Sunscreen aims to prevent sunburn, reduce sun damage, and prevent Köbner's phenomenon. Sunscreen can also reduce unaffected skin tone and discoloration of skin with depigmented lesions [3]. It is recommended that children use sunscreen after 1 year of age and with a sun protect factor (SPF) of at least 15 [4].

Corticosteroids can also cause local side effects such as striae, atrophy, telangiectasia and contact dermatitis. To reduce these side effects, in children, non-fluorinated corticosteroids with moderate potency can be used and the same efficacy for successful therapy. Corticosteroids can also be combined with other therapies such as topical calcipotriol. Combination of class potent or mid potency topical corticosteroid with calcipotriol showed 87% success rate and 95% repigmentation rate. The efficacy and safety of this combination are better than single therapy. To monitor the therapeutic response used an examination with Wood's lamp. Follow-up is done every 1–2 months. The use of topical corticosteroids in and around the eyelid must be monitored carefully, because it can cause increased intraocular pressure and cause glaucoma [5].

Key Points

- Vitiligo is a chronic pigmentation disorder characterized by the appearance of milky white macules on the skin due to progressive loss of epidermal melanocytes, it can occur in childhood.
- Various therapeutic modalities are available for the treatment of vitiligo. However, all of these therapeutic modalities cannot be used in children. In children, vitiligo therapy is usually based on the extent of the lesion.

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Chapter 25
Pediculid: An Autosensitization Dermatitis Developed as an-Id Reaction to Pediculosis Infestation

Ahmed Hassan Nouh and Mohamed L. Elsaie

Case Presentation

We report a 6 years old girl presenting with a 2 weeks generalized eczematous rash with a progressive nature. On examination polymorphic disseminated itchy rash was found consisting of groups of erythematous papules, vesicles and eczematous plaques. Lesions had a bluish grey shadows along with dusky-red erythema and fine white scales on the trunk, upper and lower limbs (Figs. 25.1 and 25.2). In addition, old slightly hyperpigmented patches and enlarged cervical lymph nodes were detected. According to her parents report, she only received ibuprofen for her fever but no other medications.

Further examination revealed massive infestation adult louse and nits. Feeding adult louse was filmed and residuals from newly hatched nymphs after moulting pictured under dermoscopic examination (Polarized DL4, ×10). Some impetiginized lesions on scalp were found as well (Figs. 25.3, 25.4 and 25.5).

Based on the case description, clinical and dermoscopic photographs, what is your diagnosis?

1. Pediculosis Capitis.
2. Scabies
3. Atopic Dermatitis
4. Pediculid.

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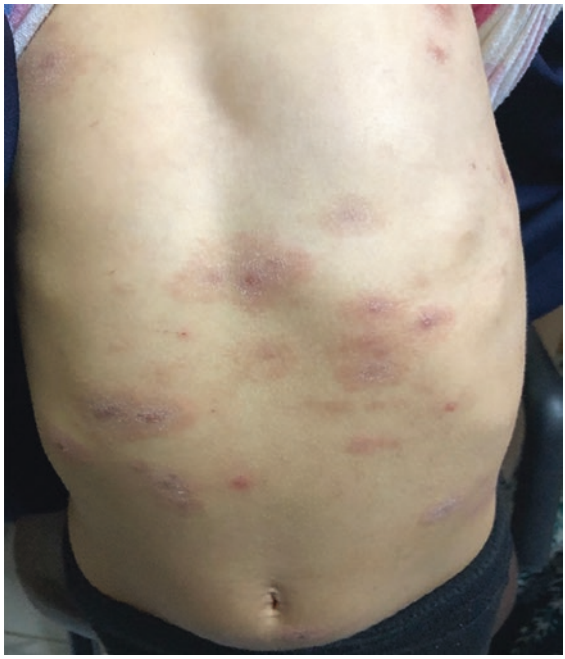

Fig. 25.1 Initial presentation of lesions

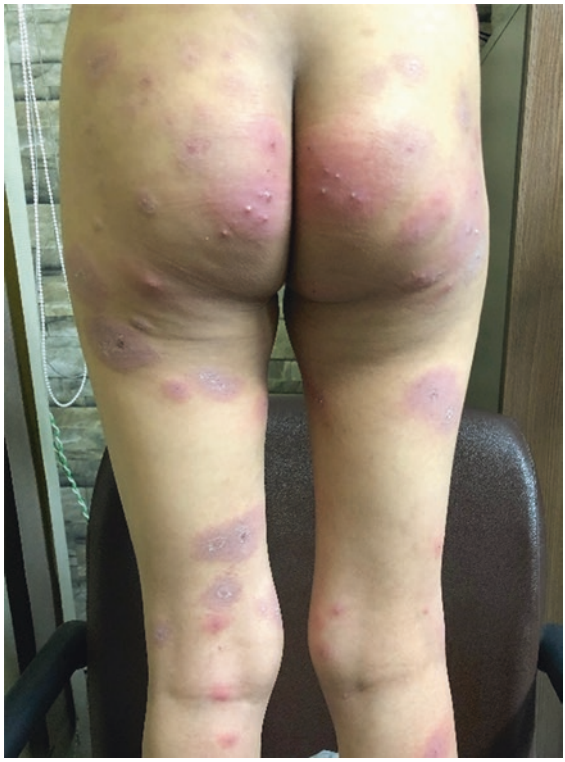

Fig. 25.2 Initial presentation of lesions

Fig. 25.3 Dermoscopic examination showing a feeding adult louse

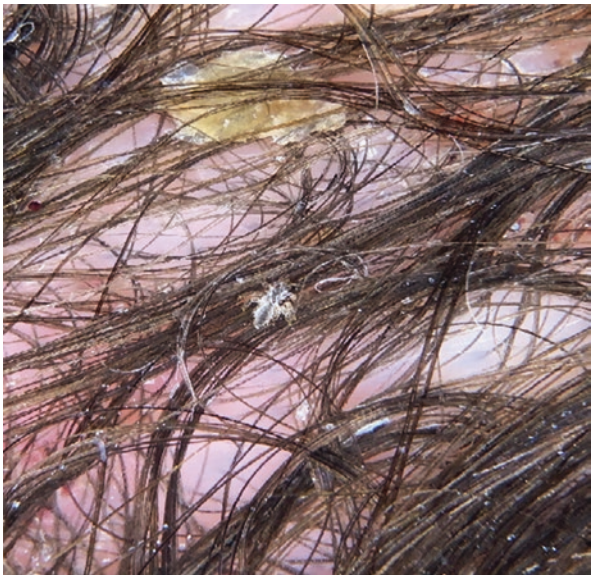

Fig. 25.4 Dermoscopic examination showing residual Chitin cover after nymph moulting

Fig. 25.5 Dermoscopic examination showing nits

Diagnosis

Pediculid

The diagnosis of Pediculid was made according to clinical picture and findings described above, as well as history and examination of clothes to exclude co-infestation with *Pediculus humanus corporis*. Treatment prescription included Potassium permanganate as wet dressing for impetiginized scalp lesions and for the same treatment purposes systemic antibiotic therapy with Amoxicillin/Clavulanic acid with general dosing according to age and weight. Cetirizine, an antihistamine was prescribed to relieve itching. Main treatment was oriented to eliminate head louse infestation—Ivermectine 1% lotion along with manual nits' elimination by special hair combing technique.

On follow up examination after 2 weeks full resolution of skin lesions was observed. Head louse infestation was eliminated but additional prophylactic treatment with ivermectine lotion was recommended once weekly and for 2 consecutive weeks (Figs. 25.6 and 25.7). The patient did not comply with the prophylactic treatment and came back with the same manifestation of ID-reaction after 2 weeks (Figs. 25.8 and 25.9). She was finally successfully treated by only Ivermectine 1% lotion to eliminate the Pediculosis.

Discussion

Pediculosis capitis is an infestation caused by *Pediculus humanus capitis* ectoparasite, a six legged parasite, nesting on the scalp [1]. The most common symptom seen in the patients is itching on the scalp, appearing approximately 7–10 days after

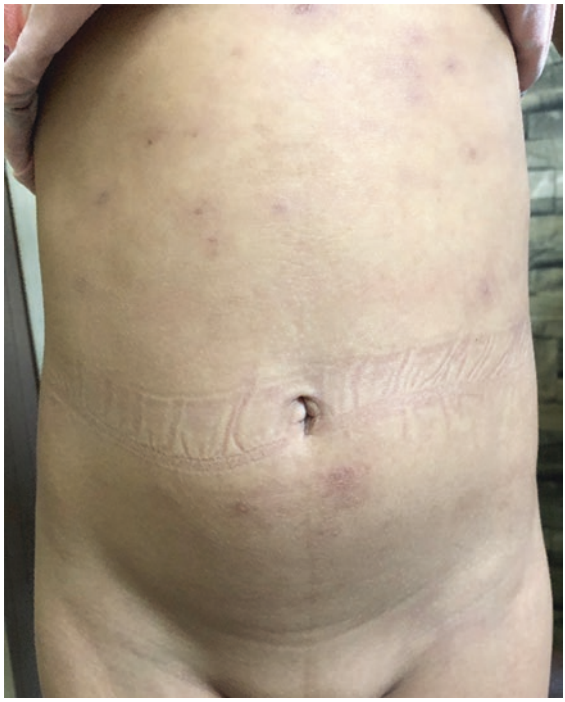

Fig. 25.6 Follow up examination status showing complete clearance of lesions

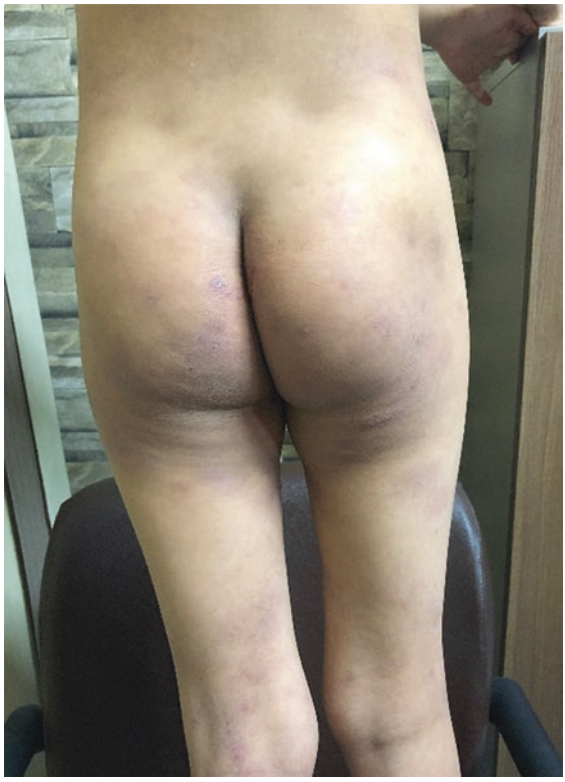

Fig. 25.7 Follow up examination status showing complete clearance of lesions

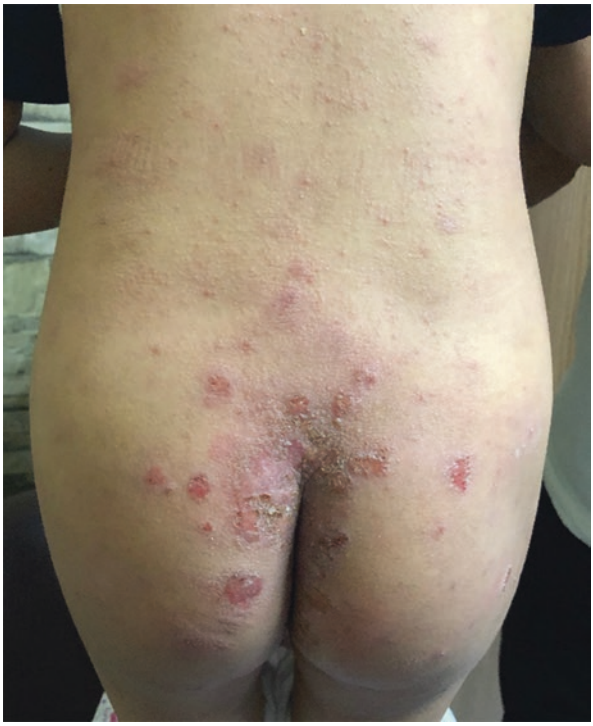

Fig. 25.8 Reappearance of skin lesions

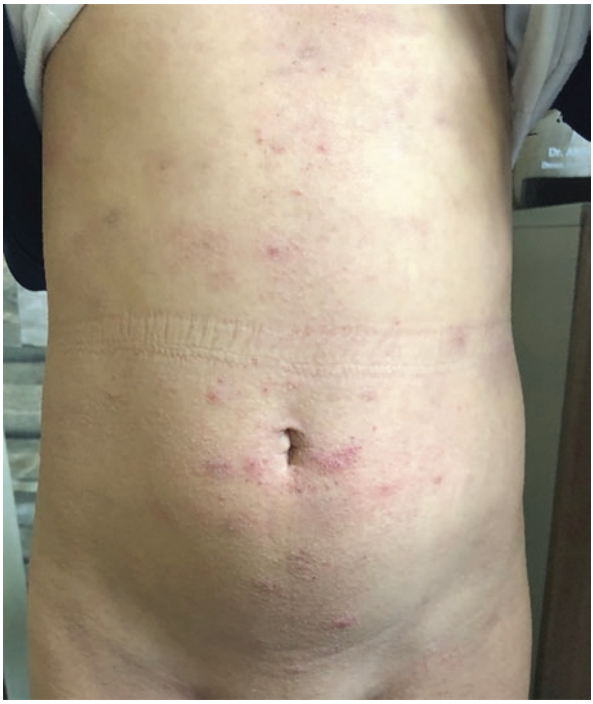

Fig. 25.9 Reappearance of skin lesions

sensitivity to the parasite's saliva or excrement antigens [1, 2]. Following the severe itching, excoriations, secondary bacterial infection, occipital or cervical lymphadenopathy might develop [3]. Autosensitization dermatitis is an acute dermatitis appearing as itchy erythematous, maculopapular or papulovesicular lesions away from the primary inflammation focus [4]. Although it is seen most frequently in leg ulcer patients, infections, trauma, irritant or allergic chemical substance contact and ionizing radiation may also cause irritation [5]. Even though the etiology of the eczematous reaction is not known for certain, autosensitization developing against the epidermal antigens is emphasized [6]. When being related with an infectious case, it is referred as dermatophytid, pediculid, bacterid or virusid in accordance with the etiologic factor [7]. The pruritic rash related with pediculosis capitis was first defined by Ronchese in 1946 [8]. Brenner and his colleagues were the ones who used the term of pediculid for the first time in 1984, by describing pruritic eruption related with pediculosis capitis [7]. To our knowledge there are only four pediculid cases in the literature. While generalized pruritic skin-colored papules existed on two patients with pediculosis capitis, the other case was a bullous pediculid identified in a patient with pediculosis pubis [7–9]. Hereby, with the presentation of a rarely reported pediculid case, the importance of exploring infection or infestation focus explore in patients who present with generalized pruritic maculopapular eruption was emphasized, before considering any complicated diagnosis requiring advanced examinations

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Chapter 26
Pruritic Erythematous Plaque on the Knee

Mohamed Saeed Mohamed, Shady M. Ibrahim, and Mohamed L. Elsaie

Case Presentation

A 9 years old male boy with no relevant history was seen in the dermatologic clinic due to a pruritic, erythematous plaque with months of evolution on his right knee (Fig. 26.1). The lesion had history of 3 years; the patient told he had no relevant

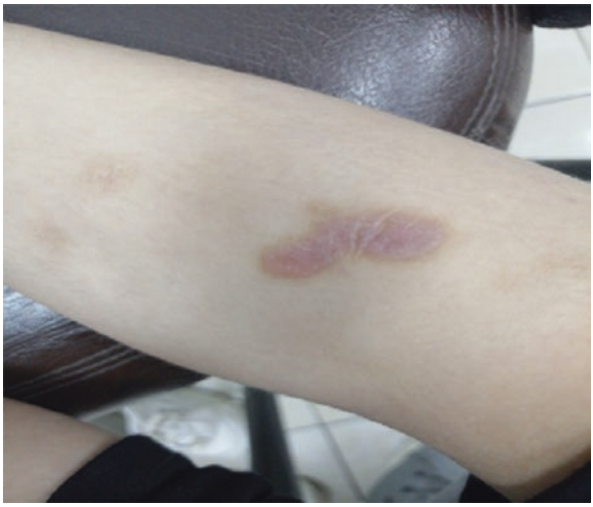

Fig. 26.1 Asymptomatic pruritic erythematous plaque on the right knee in male patient 9 years old

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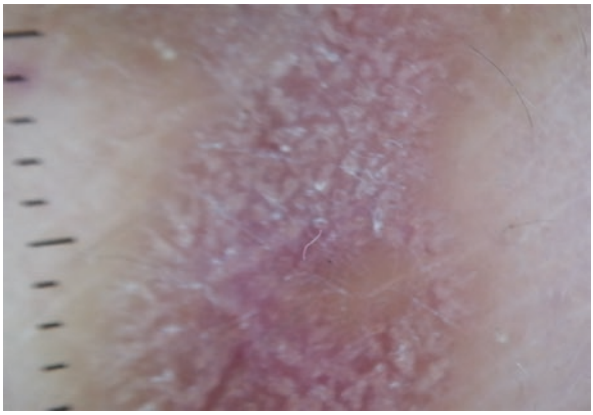

Fig. 26.2 Dermoscopic examination of the plaque revealed whitish striations (projections) with a "fern leaf" aspect on an erythematosus background, linear and rounded whitish streaks some of them are pigmented (Dermoscopy 3gen DermLite 3, magnification 10×)

familial or significant medical history apart of trauma on his right knee 5 years ago. On physical examination of the skin, he had single slightly scaly flesh colored well defined oval about 3 × 2 cm plaque on his right knee. Examination of other areas of the skin, hair, nail and oral cavity showed no abnormality. There is no evidence of apparent lymph node enlargement. Initially the patient tried topical steroid with slight response.

Dermoscopic examination of the plaque revealed whitish striations (projections) with a "fern leaf" aspect on an erythematous background (Fig. 26.2).

Based on the case description, clinical and dermoscopic photographs, what is your diagnosis?

1. Psoriasis.
2. Keloid.
3. Lichen planus.
4. Hypertrophic scar.

Diagnosis

Lichen planus

Discussion

Lichen planus is a chronic inflammatory disease that affects approximately 1–2% of the general population [1, 2]. About half of the patients with skin lesions have oral lesions, whereas about 25% present with oral lesions alone. They often occur bilaterally on the flexor surfaces of the extremities [3].

A detailed description of the peculiar striae and dots found on the surface of a lichen planus papule was given by Louis Frederic Wickham in 1895 and are referred as "Wickham's Striae" [4].

The classic skin lesion consists of a flat-topped polygonal papule that is slightly erythematous to violaceous. A thin and adherent scale can be observed on top of it. On the surface there are reticular or pinpoint whitish structures, Wickham striae (WS), pathognomonic of this disease [5].

Dermoscopy findings showed polymorphic pearly whitish structures that correspond to WS with arboriform "fern leaf" projections. At the border's linear vessels (radial capillaries) and erythematous globules can be seen, WS dermoscopic patterns [6]:

- Reticular (the most common).
- Circular.
- Radial streaming.
- Leaf venation.
- Starry sky/white dots [7, 8].

Key Points

- Lichen planus is a chronic inflammatory disease that affects approximately 1–2% of the general population.
- Dermoscopy is of high specificity and sensitivity in diagnosis of lichen planus.
- Wickham striae has different dermoscopic patterns which is clue for diagnosis (pathognomonic).
- Dermoscopy can improve the diagnosis and follow-up of patients with this dermatosis

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Chapter 27
Psoriasis Vulgaris in Children with Obesity

Armyta Denissafitri, Yuri Widia, Irmadita Citrashanty, Iskandar Zulkarnain, Muhammad Yulianto Listiawan, and Sawitri

An 11-years-old girl was brought to the Dermatology Emergency Room of Dr. Soetomo General Hospital Surabaya because of red patches with thick scales almost all over her body. The red patches first appeared on her scalp with dandruff 3 months ago. Gradually the red patches spread to all her body with an itchy and burning sensation. She already got treatment loratadine, dexamethasone tablets, and ointment from the general practitioner but there was no improvement. She had never complained like this before and there is no family with the same complaint. The physical examination of general status showed her body weight was 55 kg, based on BMI weight-for-age chart from CDC she was obesity (>95 percentile). We consulted the patient at the ENT and odontology department where there were no focal infections (Fig. 27.1).

Based on the case description and the photographs, what is your diagnosis?

1. Psoriasis vulgaris
2. Seborrheic dermatitis
3. Tinea corporis
4. Nummular dermatitis

On examination we observed multiple erythematous et hypopigmentation macules unsharply marginated with scales on the scalp, facialis, extremitas superior et inferior region, no geographic tongue. Multiple erythematous plaque with clear border, covered by scales on the trunk region. There were no subungual hyperkeratosis

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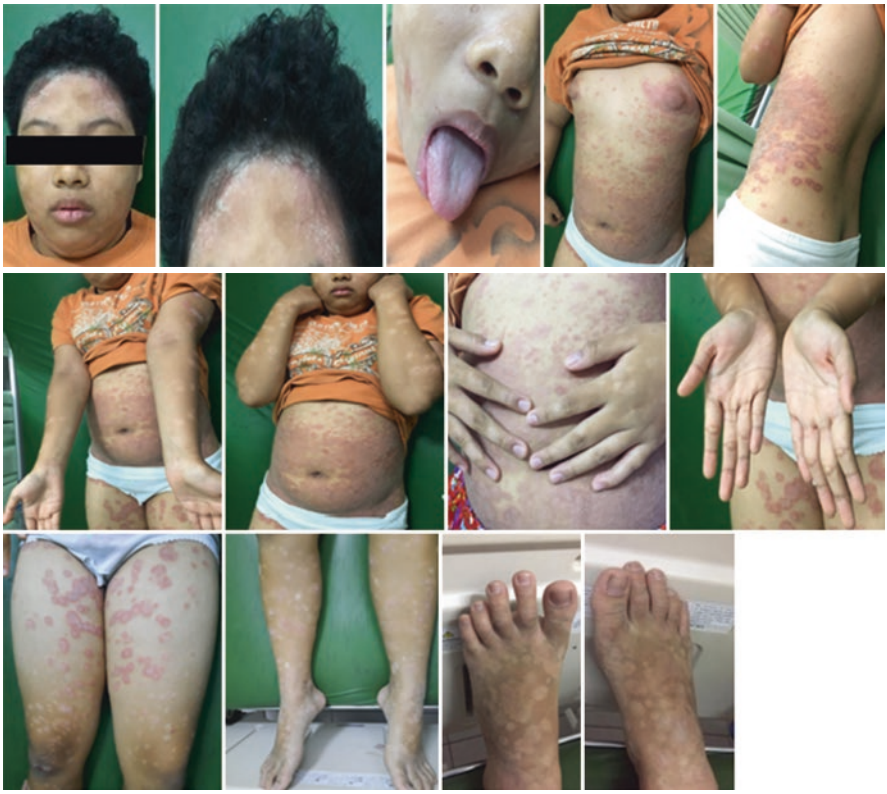

Fig. 27.1 Multiple erythematous macules with white thick scale almost all over her body

and no pitting nails. There were Auspitz signs and Karsvlek phenomena. The PASI (Psoriasis Area and Severity Index) score was 23.2.

Laboratory examination: Hemoglobin 12.7 g/dL (normal range: 13.3–16.6), leucocyte 9800/μL (3370–10,000), thrombocyte 340,000/μL (150,000–450,000), SGOT 20 U/L (0–50), SGPT 27 U/L (0–50), BUN 11 mg/dL (7–18), creatinine serum 0.63 mg/dL (0.6–1.3). Urine examination was normal. Potassium hydroxide examination there was no hyphae and no pseudohyphae. Histopathology examination was concordance with psoriasis vulgaris.

Diagnosis: Psoriasis vulgaris with obesity

Treatment: The patient was given topical desoximetasone 0.25% cream on the thick scale, mometasone furoate cream on the face, methotrexate 15 mg/week, folic acid 1mg/daily the day after methotrexate consumption, and emollient (vaseline album). After 1 week of methotrexate treatment, the lesion had significant improvement with the PASI score of 7.3 and no side effects (Fig. 27.2). We followed up the patient's condition after methotrexate treatment 1 year later, she was in good condition and never relapsed using only topical treatment.

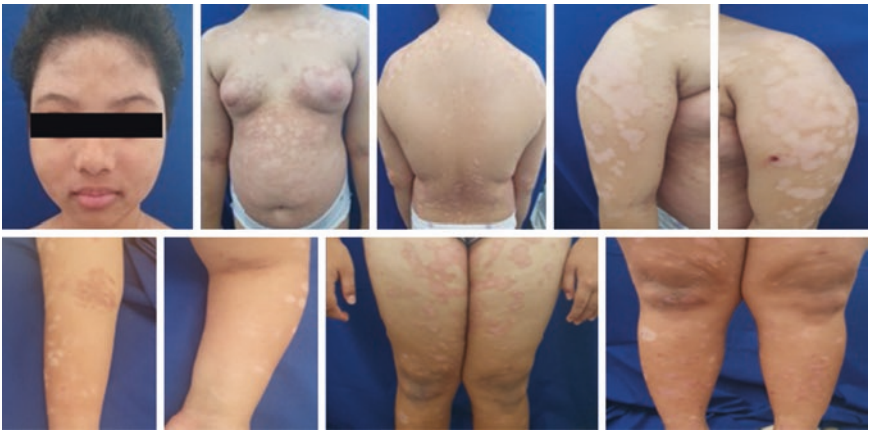

Fig. 27.2 Patient's progression after 1 week of methotrexate therapy

Discussion

The description of psoriasis in children is a well-demarcated erythematous plaque with silvery-white scale. Pruritus is a frequent symptom in children [1]. Childhood psoriasis is linked to several comorbidities with adult equivalence, one of the comorbidity is obesity [2]. Obesity is also related to greater disease activity and severity. In recent days, the correlation between psoriasis and obesity has become visible. Some studies hence showed the association between psoriasis and increased BMI. Obesity may be a trigger for psoriasis because it is associated with a low-grade systemic inflammatory state, owing to adipokines released by the interaction between adipocytes and macrophages [3, 4]. It is shown that intra-abdominal fat is actively secreting adipocytokines, promoting inflammation. The childhood-onset psoriasis is associated with HLA-Cw6. This specific gene is also associated with obesity. An individual with normal weight without HLA-Cw6 was 35 times less likely to occur psoriasis than obese individuals with the HLA-Cw6 gene. At this point, it can be said that there is an interplay between psoriasis and obesity, and hence adipokines may play an important role [3]. The standardized guidelines on the management and treatment of the psoriasis in children are lacking [2]. Systemic therapies should only be used in pediatric patients with moderate to severe psoriasis and methotrexate is one of the conventional therapy. To date, evidence on efficacy and safety of systemic treatments in pediatrics psoriasis is limited and evidence-based guidelines are scarce. The 'rule of tens', used in adults to indicate moderate to severe psoriasis, can also be applied in pediatric psoriasis as an indication for the use of systemic therapy. This modified 'pediatric rule of tens' would be PASI $\geq$10 and/or body surface area (BSA) $\geq$10 and/or the children's dermatology life quality index (CDLQI) $\geq$10 [5, 6]. Methotrexate is recommended as one of the first-line treatments and it has a degree B/C of recommendation approval by the European

Medicines Agency (EMA) [7]. The recommendations for methotrexate dosage is once weekly 0.2–0.4 mg/kg/week [5]. In this case, we gave the methotrexate 15 mg/week with a good result and no side effects.

The guidelines for the treatment of moderate to severe psoriasis vulgaris in Indonesia still use methotrexate as the first-line treatment, although it is not declared as a treatment for adults or children. Biologic agent therapy is available in Indonesia at very high price, but is not commonly used because it is not covered by government insurance. Methotrexate is available at low price, easy to obtain, and provides good therapeutic results with no severe side effects in Indonesia.

Key Points

Psoriasis vulgaris in children has a strong association with obesity as comorbidity. Methotrexate is one of the systemic treatment for moderate to severe psoriasis vulgaris in children.

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Chapter 28
Purpuric Eruption in a COVID 19 Positive Child

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An 8 years old male patient applied to the outpatient clinic complaining of rash on the upper and lower extremities for 5 days. The rash was treated from his medical physician with antihistaminic medication without improvement and then the child was sent to dermatology department. His medical history and family history were unremarkable. He has no history of trauma or allergies. His childhood vaccinations were performed on time. He was afebrile, eating and drinking well. He had been acting normally during the day.

The dermatological examination revealed multiple no pruritic rash that starts briefly as erythematous papules or urticarial wheals, petechial and purpura [1] (Fig. 28.1). The lesions change from red to purple to rust-colored before fading over a period of approximately 10 days. The rash is most commonly located in dependent areas that are subject to pressure, such as the lower extremities, belt line, and buttocks and upper extremities (Figs. 28.1 and 28.2). The purpura is seen on the extensor surfaces of the extremities. He had no other cutaneous pathology and her other system physical examinations were completely normal. According to the consultation note, his weight and height were in accordance with her age. We sent the patient to the laboratory and we didn't see the patient for 5 days. After 5 days the child came with the results of biochemistry and complaining of abdominal pain and vomiting. We send the patient to pediatric department for hospitalization. Keeping the pandemic in mind, his PCR-Cov-2 tests were requested and his nasal swab was

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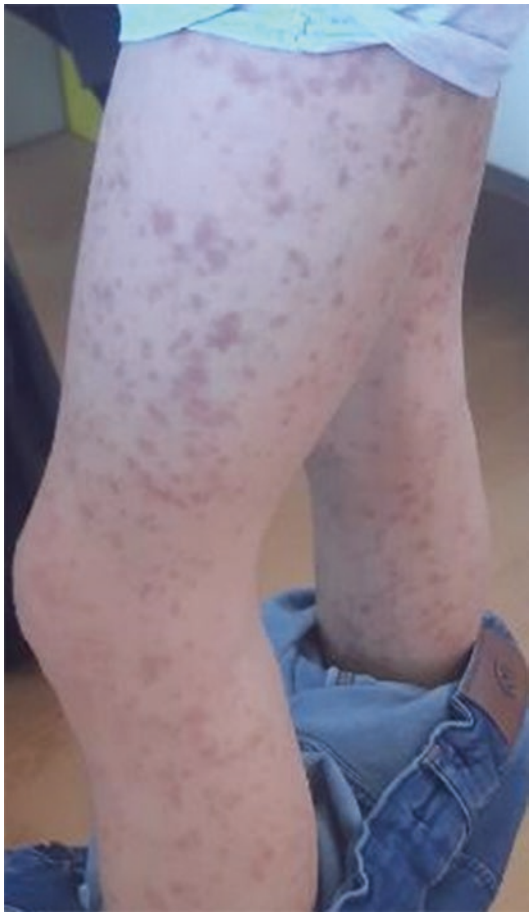

Fig. 28.1 Palpable purpura on the lower limbs

positive on Cov-2 infection: Furthermore, D-dimer was 2808 mg, CRP was 2.8. His heart sounds were normal. He also had lower extremity pitting edema. The rest of the examination did not reveal anything unusual. On examination, general condition of the patient was fair and vitals were stable. Abdomen was soft and non-tender.

On admission, his serum creatinine was 49 mmol/l, and he had proteinuria, and hematuria. In addition, serum complement testing for C3 and C4 were normal.

The blood pressure was TA 145/94 mmHg. During hospitalization complete blood account were performed, monitoring of the pressure, blood culture, coagulation profile, basic metabolic panel and Abdominal ultrasonography.

Based on the case description and the photograph, what is your diagnosis?

- Urticarial vasculitis
- Microvascular Angiopathy of SARS-Cov-2
- Henoch Schönlein Purpura
- Erythema multiforme

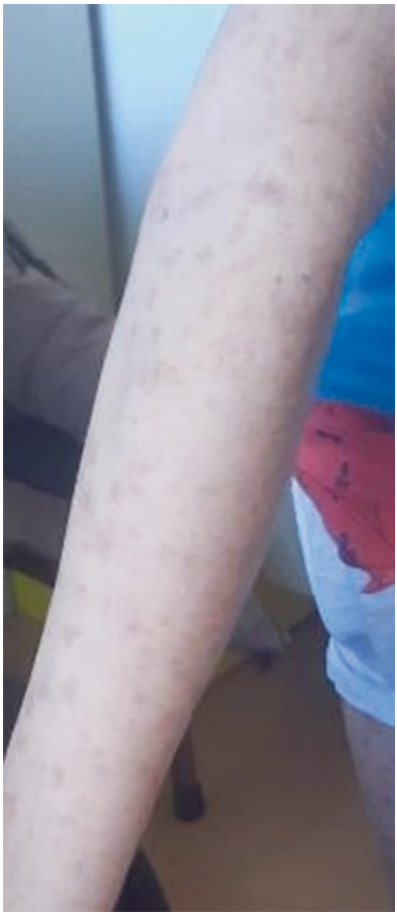

Fig. 28.2 Palpable purpura on the upper extremities

- Thrombocytopenic purpura
- Polyarteritis nodosa

Diagnosis Henoch Schönlein purpura

Henoch Schonlein Purpura (HSP) is the most common cause of non-thrombocytopenic purpura, Ig A mediated small vessel vasculitis of the childhood that affects several systems which is characterized with skin changes and abdominal pain, joint or renal manifestation [1].

The diagnosis is made due to the mandatory and supportive criteria of European league against Rheumatism, the Pediatric Rheumatology International Trials Organization and the Paediatric Rheumatology European Society (EULAR/PRINTO/PRES) in 2010. The mandatory criterion includes palpable purpura in the absence of thrombocytopaenia, while the supportive criteria involve at least one or more of the following: acute onset diffuse abdominal pain, acute-onset arthralgia or arthritis, renal involvement in the form of proteinuria or haematuria, and

histopathological evidence of leucocytoclastic vasculitis or proliferative glomerulonephritis with predominant IgA deposits [2, 3].

The dermatology condition is presented with palpable purpura distributed symmetrically over the lower limbs is typical, commonly extensor surfaces and buttocks. It may also involve arms, face and ears but usually spares the trunk. Purpuric lesions range from petechiae to larger bruises and may be proceeded by urticaria or erythematous maculopapular lesions. There may also be a painful subcutaneous oedema (hands, feet, sacrum, scrotum).

Joints (60–80%) specially affects the larger joints (knees, ankles) with pain, swelling and reduced range of movement. Swelling is usually periarticular oedema, synovial effusions are usually absent. Joint involvement can be debilitating but does not result in permanent damage.

Gastrointestinal (60%) symptoms are colicky abdominal pain, nausea and vomiting to GI hemorrhage, intussusception and rarely, pancreatitis, hydrops of the gallbladder and protein losing enteropathy.

Renal (20–60%) symptoms are 76% present within 4 weeks of disease onset and 97% present within 3 months of disease onset. Renal involvement has a wide range; microscopic hematuria, macroscopic hematuria, proteinuria, nephritic/nephrotic syndrome and renal impairment. Hypertension may be associated with nephritis or be an isolated finding. Most patients with renal involvement will have mild disease that recovers well. 5% will have long term morbidity [4–6].

The HSP can be trigger due to secondary upper infection of respiratory tract, medication, or malignancy [1].

The virus Covid-19 is caused by SARS Covid 2 which is a single stranded RNA virus from beta corona viridae family and its present as a respiratory symptom from a mild cold like to pneumonia and also with cardiac, thrombotic hepatocellular and dermatology symptoms.

During hospitalization child was treated with oral prednisolone for 7 days. Child also received other supportive care in the form of anti-emetics, antacids, analgesics and fluids. The cutaneous purpura markedly improved. Child did not develop any neurological manifestations. Patient was advised follow up in the OPD with serial urine analysis and BP monitoring after 2 weeks [7–9].

Key Points

- HSP is usually self-limiting and symptoms resolve within 6 weeks in most cases
- The HSP can be trigger due to secondary upper infection of respiratory tract as SARS CoV-2 infection.
- SARS-CoV-2 infection is usually asymptomatic or mild in children; and therefore may go unnoticed until the appearance of cutaneous findings.
- Mortality is very low severe abdominal pain and bloody stools are risk factors for significant renal involvement
- Renal manifestations generally present in the first 6 months but occasional cases take up to 12 months so prolonged monitoring is required.

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Chapter 29
Scaly Itchy Lesion of Glans Penis

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Case Presentation

Male patient 9 years old presented in the dermatologic clinic with itchy scaly lesion on his glans penis (Fig. 29.1). The lesion had history of 4 years, initially started as scaly lesions then became diffuse over the glans penis with no family history of the same lesion. On physical examination, the patient presented with whitish scales on the glans penis with slight erythema, the patient reported bleeding with minor trauma and no other associated symptoms apart of itching. There is no evidence of apparent physical lymph node enlargement. Initially the patient tried topical steroid application with remission but recurrence is often common.

Dermoscopic examination with non-contact mode revealed diffuse white scales, with application of interface media (alcohol), there is dotted blood vessels pattern arranged regularly and diffuse all over the lesion (Fig. 29.2).

Based on the case description, clinical and dermoscopic photographs, what is your diagnosis?

1. Psoriasis.
2. Eczema.
3. Bowen's disease.
4. Tinea circinate.

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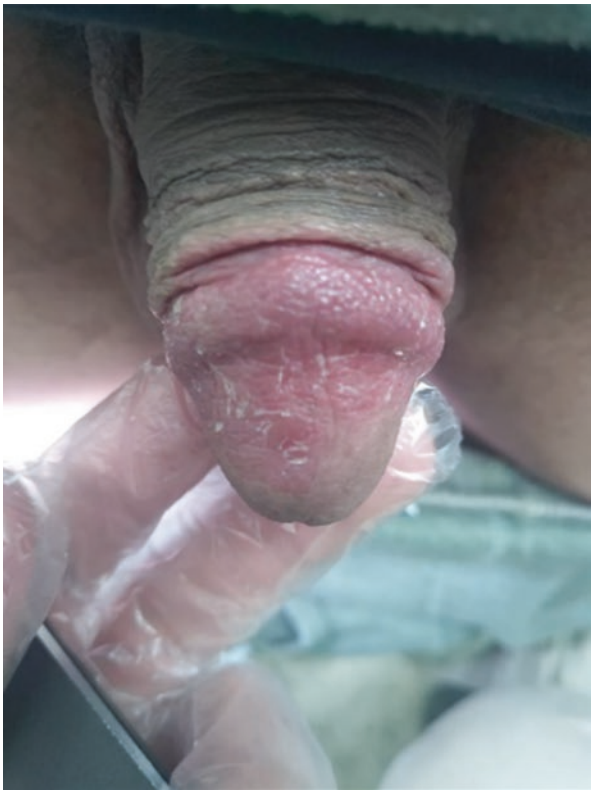

Fig. 29.1 Itchy scaly lesion on the glans penis of male patient 9 years old

Diagnosis

Psoriasis

Discussion

Genital psoriasis affects approximately 63% of psoriasis patients at least once in their lifetime. More than any other area on the body, genital lesions significantly impair patients' quality of life [1]. In 2–5% of psoriasis patients, lesions only occur in the genital region [2]. Genital psoriasis can occur in all age groups, from newborns to geriatric patients, with a slight predilection for younger male patients with relatively severe disease [3].

Low-to-mid-potency topical corticosteroids are the first-line treatment for genital psoriasis [4] but with special caution due to peculiar anatomy of the genitalia [5]. The most common differential diagnosis of psoriasis is eczema. Dermoscopy is a non-invasive office procedure, which facilitates the diagnosis of inflammatory skin

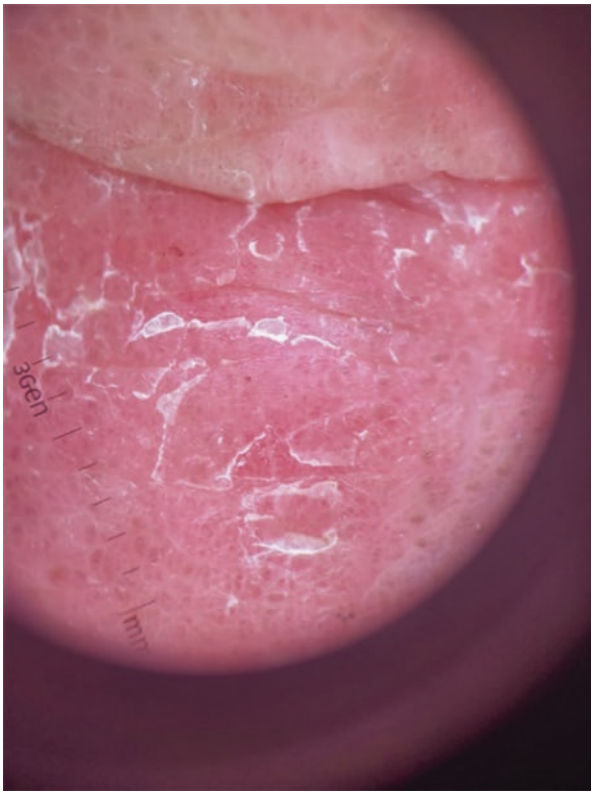

Fig. 29.2 Dermoscopic examination revealed diffuse white scales, with application of interface media (alcohol), there is dotted blood vessels pattern arranged regularly and diffuse all over the lesion (Dermoscopy 3gen DermLite 4, magnification 10×)

diseases.it is used for the evaluation of the type and the distribution of cutaneous blood vessels, as well as the color of the scale [6]. Dermoscopy of psoriasis is highly sensitive and specific shows white scales with diffuse red dots arrangement while in eczema the vessel have same pattern as psoriasis but in scattered arrangement [7].

Key Points

- Genital psoriasis affects approximately 63% of psoriasis patients.
- It affects and impairs psychological and sexual life of the patient.
- Dermoscopy is a non-invasive office procedure helping differentiation of psoriasis from other similar dermatoses as dermatitis.

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Chapter 30
The Impact of Malnutrition to *Pseudomonas aeruginosa* Infection in Children

Indah Purnamasari, Yuri Widia, Irmadita Citrashanty, Sawitri, Iskandar Zulkarnain, Muhammad Yulianto Listiawan, and Afif Nurul Hidayati

A 7-year-old girl came to the Dermatology Outpatient Clinic of Dr. Soetomo General Hospital Surabaya due to progressive wound and ulcer on her scalp since 2.5 months ago. According to her mother, it initially appeared as a small bump, filled with pus, spread around her scalp then turned into ulcers. The wounds and ulcers became more extensive and there was a history of slight fever, weakness, and pale skin (Fig. 30.1). Weakness conditions and pale appearances were also being complained by the patient's mother. Previous treatment with oral antibiotics and hospitalized for 1 week showed no improvement. The patient was in poor nutritional status but had a complete immunization history. There was no history of growth and development delay.

General examination showed malnutrition. Her body weight was 12 kg and 98 cm tall based on body mass index (BMI) weight-for-age chart from the Centers for Disease Control and Prevention (CDC) the patient was marked as moderate malnutrition (<5 percentile). The body temperature was 38.5 °C, conjunctiva looked anemic and the scalp appeared as wasted and fluffy hair.

Local examination showed wide erythematous necrotic ulcers on scalp region in size 15 × 10 cm and 15 × 7 cm, with a central black eschar and surrounding hyperemia along with purulent discharge (Fig. 30.1) and was foul-smelling.

Based on the case description and the photographs, what is your diagnosis?

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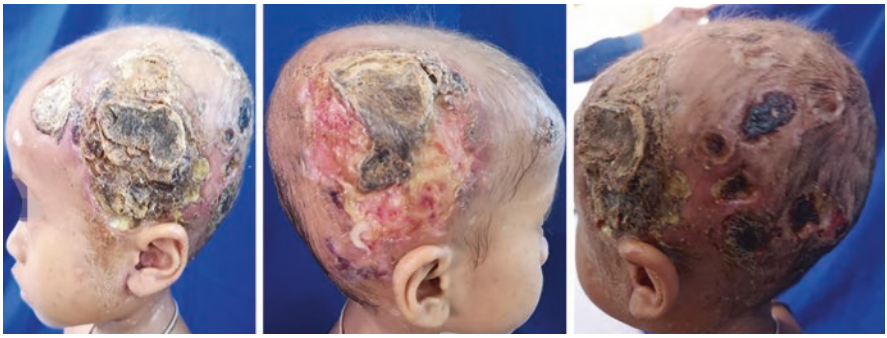

Fig. 30.1 Extensive ulcers with purulent discharge and necrotic eschar on first day of admission

1. *Pseudomonas* infection
2. Pyoderma gangrenosum
3. Tinea favosa
4. Kerion type tinea capitis
5. Malignancy

Laboratory examination: erythrocyte count 3.07×10^6, hemoglobin 7 g/dL, leukocytes 6300/μL, thrombocyte 474,000/μL (150,000–450,000), serum creatinine 0.3 mg/day, potassium 4.4 mmol/dL, sodium 140 mmol/dL, chloride 101 mmol/dL, SGOT 79 U/L, SGPT 91 U/L, albumin 3.1 g/dL and BUN 9 mg/dL. Gram negative bacteria was found on Gram staining. Potassium hydroxide test was negative. The bacterial culture from blood and pus swab were positive for *Pseudomonas aeruginosa* which sensitive to ceftazidime, cefepime, amikacin, piperacillin, piperacillin-tazobactam, ciprofloxacin, levofloxacin, fosfomycin, imipenem, and meropenem. The biopsy examination was not performed because her mother refused it.

Diagnosis: Chronic wound infections caused by *Pseudomonas aeruginosa* with moderate malnutrition.

Treatment: We immediately started comprehensive management to the patient in order to improve the general condition, giving the appropriate wound care and systemic antibiotic therapy. The improvement of general condition was achieved by giving packed red cells (PRC) blood transfusion until Hb>10 and correcting the nutritional deficiency in collaboration with the Pediatric Department. After two bags of transfusion, the hemoglobin increased to 11 g/dL and her body weight increased gradually during the subsequent 2 weeks to 14 kg. Wound care with wet dressing with normal saline, debridement necrotic tissue every 2 days and moist care with tulle. The antibiotic agent intravenous ceftazidime 600 mg was given every 8 h based on sensitivity results. Six-days after wound care, the central eschar sloughed and the ulcer improved to 8 × 4 cm and 9 × 4 cm in size, minimal exudate and with granulation tissue formation (Fig. 30.2). The ulcers started to heal after 3 months of treatment (Fig. 30.3). We also had collaboration with the Plastic Surgeon Department who helped manage the possibility of repairing the remaining scars.

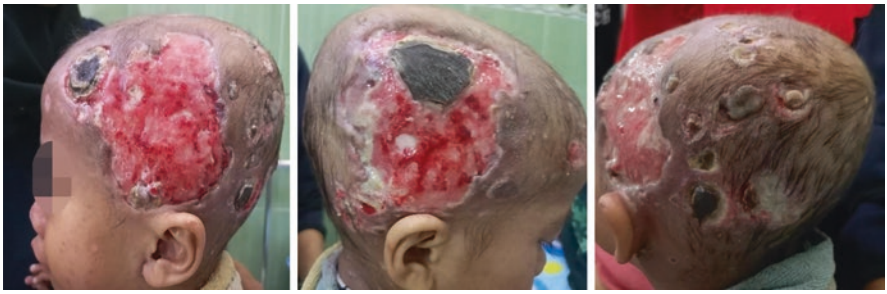

Fig. 30.2 The ulcers has been improved after sloughing of necrotic eschar on day 6 after wound care

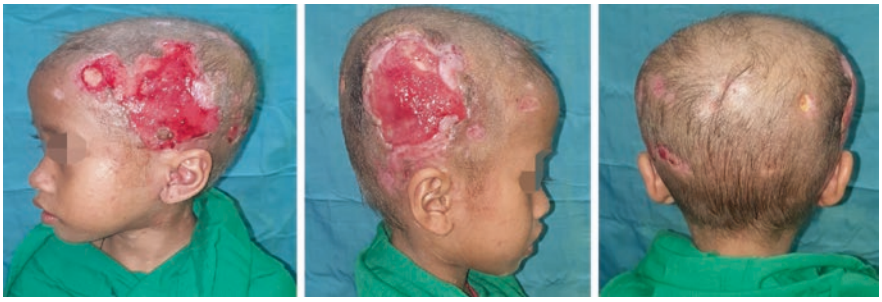

Fig. 30.3 The ulcers looked smaller, with a clean base, red in colour with granulation tissue but presented with bone exposed in some areas on the scalp

Discussion

The synergistic association between malnutrition and infection has been recognized for more than 50 years [1]. Few longitudinal studies clearly define malnutrition as a risk factor for the increased incidence and/or severity of infection [2]. The World Health Organization (WHO) defines malnutrition as deficiency, excess or imbalance in a person's intake of energy and/or specific nutrients in relation to their requirements [3].

Pseudomonas aeruginosa is an ubiquitous pathogen infecting virtually all tissues that can cause both acute and chronic infections, especially prevalent among in immunocompromised patients, pyoderma gangrenosum and burn wounds [4]. Infections in malnourished children can be due to *P. aeruginosa* and associated with higher mortality [5].

Classic pyoderma gangrenosum can occur on any skin surface, but is most commonly seen on the legs. The typical appearance is that of a necrotic skin lesion with a central eschar surrounded by a erythematous halo. The diagnosis of pyoderma gangrenosum is based mainly on clinical findings because biopsies show no specific diagnostic features. In many cases, however, a biopsy can help exclude other

conditions such as infections, malignancy, or cutaneous vasculitis. In histopathological examination of pyoderma gangrenosum, lymphocytes may be seen to infiltrate vessel walls with intramural and intravascular fibrin deposition indicative of vascular damage [6].

The chronic wounds in this case were originally suspected due to pyoderma gangrenosum because of the similarities of some clinical manifestations of a deep ulcer with a well defined border. The ulcer starts as a small papule or collection of papules, which break down to form necrotic wounds with eschar. Because in this case a biopsy examination was not performed, a pyoderma gangrenosum was not as a definitive diagnosis, but the bacterial culture in this case signified the presence of bacteria for *Pseudomonas aeruginosa* infection.

Comprehensive management is required, including counseling of parents [5]. It is crucial to recognize whether the wound is infected or not, identify the causative agent and give the appropriate systemic antibiotic therapy [7]. In this case, the patient had moderate malnutrition and anemia; due to that condition, correction of nutritional deficiency and administration of blood transfusion are the main recommendations. Nutritional status should be optimized to provide increased substrate availability to meet the demands of tissue repair and immune function and to prevent wounds from succumbing to infection and delayed healing [2].

Wound healing typically occurs in 14 days [1]. In children, tissue defects are reported to close faster, because fibroblasts are more abundant, collagen and elastin are more rapidly produced and granulation tissue forms faster than it does in adults [2]. Another important thing is to optimize the local healing environment by necrotic and eschar debridement, washing away slough, correct moisture balance and appropriate dressings. The theoretical benefits of cleansing and debridement are that the remaining tissue is well vascularized and devitalized tissue that might support microbial growth and prevent access to leukocytes is removed [2].

Pseudomonas aeruginosa is resistant to most first-line antibiotics [8]. So it is recommended to choose the best antibiotic against these bacteria. Antipseudomonal penicillins such as ticarcillin and piperacillin, third and fourth generation cephalosporins, aminoglycosides, carbapenems such as imipenem and meropenem, fluoroquinolones such as ciprofloxacin and levofloxacin are the treatment options available. There is insufficient data from clinical trials to establish the superiority of any of those ones [9]. The antibiotic choices can be led by the results of culture examination. According to the bacterial culture and sensitivity results, in this case, we gave ceftazidime. Ceftazidime, a third-generation cephalosporin, is primarily intended for infections which are proved or strongly suspected to be caused by *P. aeruginosa* or other related bacteria [9]. The patient's condition had significant improvement, after comprehensively improving the general condition, caring the wound along with administration of systemic antibiotics; however, the residual defect was still present and required a long time and further interventions.

The gold standard for confirming the diagnosis of pyoderma gangrenosum is a biopsy, which should be followed by corticosteroid treatment. Nutritional intervention, proper wound care, and antibiotic treatment alone, however, improved the patient's condition. As a result, the diagnosis of pyoderma gangrenosum can be ruled out.

The malnutrition–environment–infection axis is complex and not easily addressed by individual interventions.

Key Points

- Malnutrition is a risk factor for the increased severity of infection in children.
- The proper holistic management can speed up the healing of the infection.

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Chapter 31
Unusual Pigmentation of Skin and Hair

Mohamed Saeed Mohamed, Shady M. Ibrahim, and Mohamed L. Elsaie

Case Presentation

Male patient 6 years old referred from pediatric clinic to dermatologic clinic for evaluation of unusual hypopigmentation of his skin and hair (Fig. 31.1). The lesion had history of 4 years duration with gradual progression and no family history. Medical history revealed mild delayed development, intellectual disability and seizures. On physical examination, the patient presented with discoloration of his hair compared to his parents, his hair color appeared lighter with normal length. His scalp hair appeared thin and pale. Dermoscopic examination of hairs revealed hypopigmented longitudinal short hair in a linear distribution (Fig. 31.2).

Examination of hair shafts under the light microscope showed unevenly distributed melanin, with a "road-dividing line"-like appearance (Fig. 31.3)

Based on the case description, clinical and dermoscopic and light microscopy photographs, what is your diagnosis?

1. Loose anagen hair syndrome.
2. Griscelli syndrome.
3. Mucopolysaccharidosis.
4. Hyper Ig syndrome.

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Fig. 31.1 Unusual hypopigmentation of skin and hair in male patient 6 years old

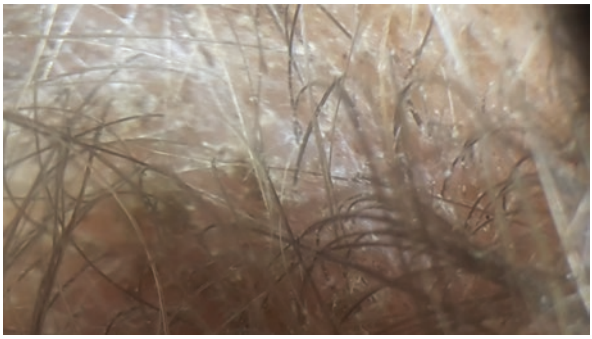

Fig. 31.2 Dermoscopy of the hair shafts showed hypopigmented longitudinal short hair in a linear distribution (Dermoscopy 3gen DermLite 3, magnification 10×)

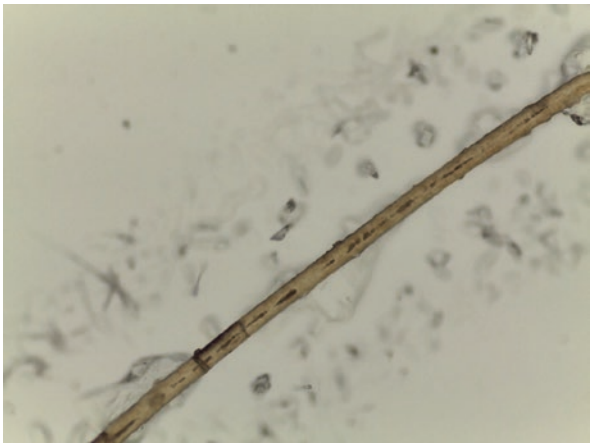

Fig. 31.3 Light microscope examination of epilated hair showed unevenly distributed melanin, with a "road-dividing line"-like appearance

Diagnosis

Griscelli syndrome

Discussion

Griscelli syndrome (GS) is a rare autosomal recessive disorder, characterized by unusual hypopigmentation of skin and hair. Its onset appears soon after birth between 4 months and 7 years of age with no sex predilection [1, 2].

GS is classified into three types. Type I is due to MYO5A gene mutations and manifests with hypomelanosis associated with a primary neurological deficit. Type II is caused by mutation in the RAB27A gene and presents with hypopigmentation combined with variable cellular immunodeficiency. Type III is characterized by hypomelanosis, but without neurological or immunological manifestations. This type may result from a mutation in melanophilin (MLPH) or the MYO5A gene [3, 4].

The combination of medical history, clinical manifestations, dermoscopic and microscopic of hair examination led to the diagnosis of Griscelli syndrome. The presence of "road-dividing line" on hair microscopic and dermoscopic examination which indicate unevenly distributed melanin inside the hair shaft is highly characteristic for Griscelli syndrome, emphasizing the important role of dermatologic examination in such cases for the diagnosis [1].

In Griscelli syndrome type 1, no specific treatment exists because the defect is in the brain rather than in the blood cells, as in Griscelli syndrome type 2. The severe neurologic impairment and retarded psychomotor development are permanent. In Griscelli syndrome type 2, bone marrow (or stem cell) transplantation is the only real treatment option for the hemophagocytic lymphohistiocytosis (HLH) syndromes in Griscelli syndrome type 2 [5].

Key Points

- Griscelli syndrome (GS) is a rare autosomal recessive disorder affect both males and females, its onset usually soon after delivery.
- Dermoscopic and microscopic examination in Griscelli syndrome have great importance for diagnosis.

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Chapter 32
Vaginal Bleeding and "Café Au Lait" Patches

Alfonso Delgado Rubio and Fabio Arcangeli

A 6-year-old girl was referred to our clinic because of vaginal bleeding that has lasted for 8 days. About 6 months ago, her breasts started growing. On physical examination, "café-au-lait" patches with irregular edges on the left periocular region and on the back are found (Figs. 32.1 and 32.2). Breast development occurs in Tanner stage three, there is no underarm and pubic hair. The right lower limb is 2 cm shorter than the left.

Radiology Bone age was 7 years and 10 months, according to Greulich and Pyle and 8 years according to Tanner-Rus (advanced in both evaluations) (Figs. 32.3 and 32.4).

An X-ray examination of the long bones of the lower limbs shows areas of dysplasia in the right femur, tibia and fibula.

Abdominal ultrasound shows large cyst with an important reduction of the parenchyma in the right ovary and microcysts in the left ovary.

Based on the clinical history, photographs and X-rays, what is your diagnosis ?

1. True precocious puberty
2. Type 1 Neurofibromatosis
3. Polyostotic fibrous dysplasia (McCune-Albright Syndrome)
4. Cystic fibrous osteitis
5. Ollier's enchondromatosis

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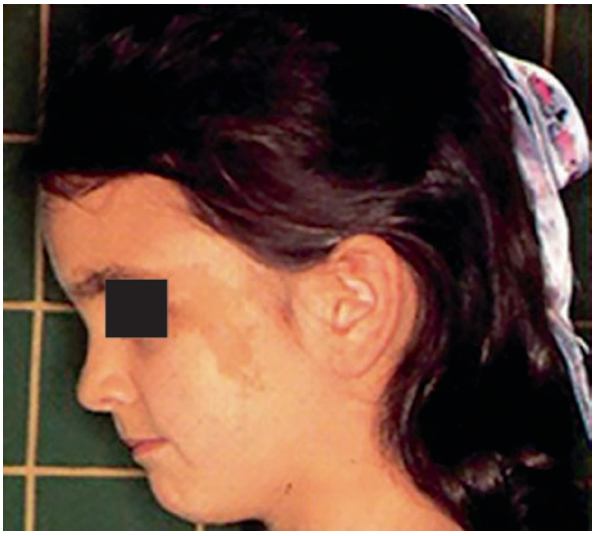

Fig. 32.1 "Café-au-lait" patches with irregular edges in the left periocular region

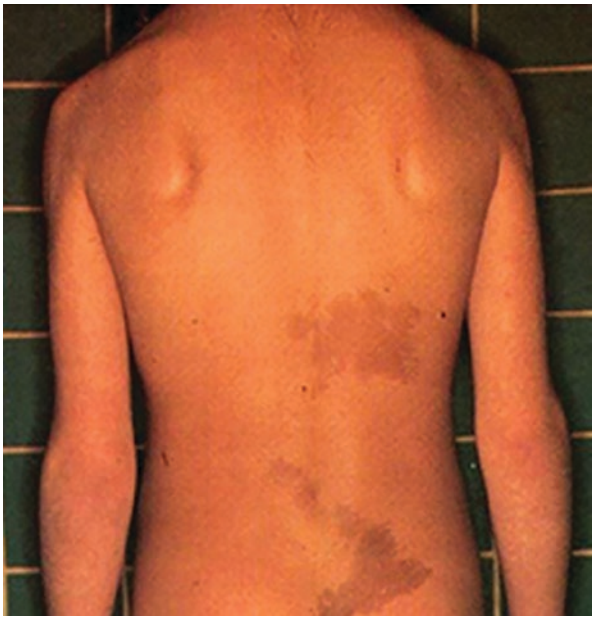

Fig. 32.2 "Café-au-lait" patches with irregular edges on the back and in the left sacral region

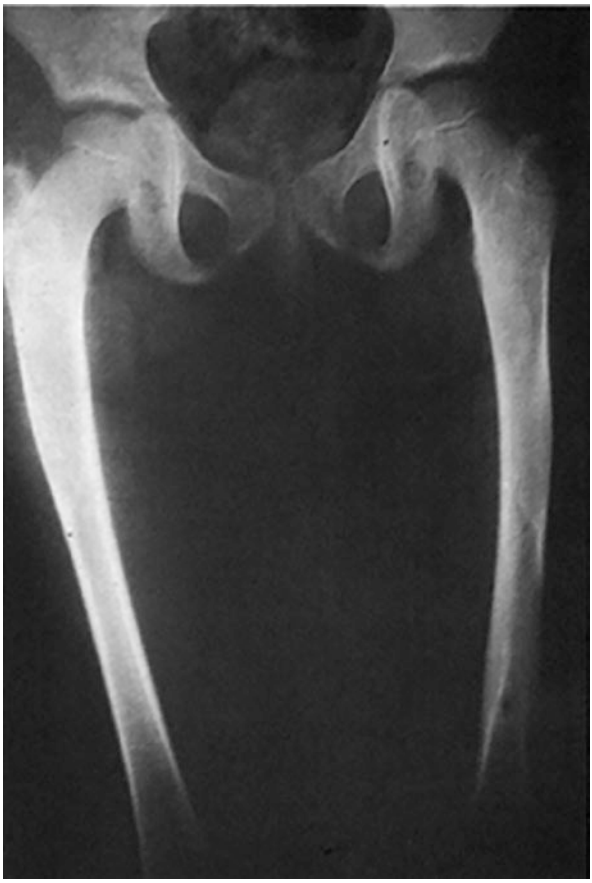

Fig. 32.3 X-ray examination of the long bones shows areas of dysplasia in the right femur

Diagnosis

McCune-Albright syndrome

Discussion

The presence in a patient, especially if female, of “café-au-lait” patches and polyostotic bone dysplasia in the lower limbs long bones, together with precocious puberty suggest the diagnosis of McCune-Albright syndrome [1]. Unlike Type 1 Neurofibromatosis “café-au-lait” patches, whose margins recall the California coast, McCune Albright syndrome patches have irregular borders like the Main coast [1].

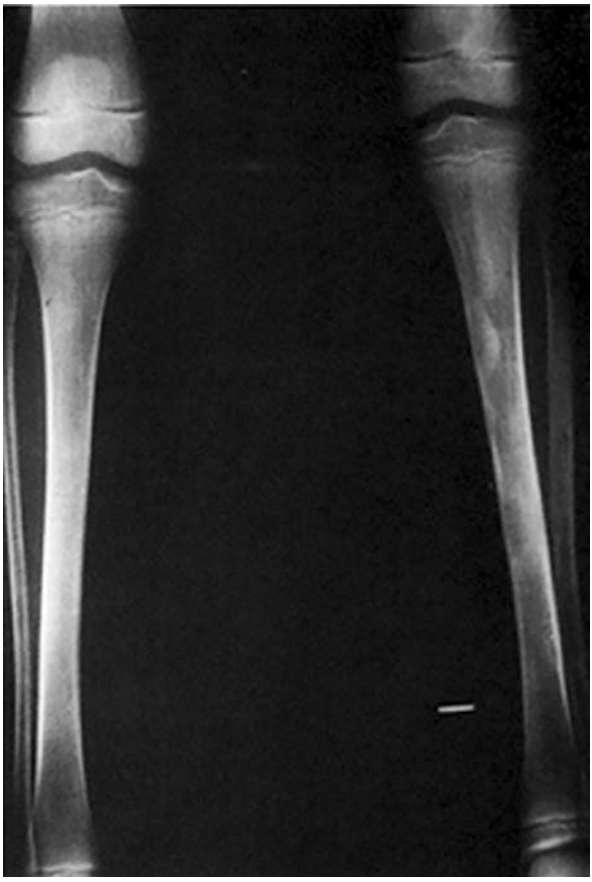

Fig. 32.4 X-ray examination of the long bones shows areas of dysplasia in the tibia and fibula

McCune-Albright syndrome is a rare disorder defined by the triad of polyostotic fibrous dysplasia, "café au lait" skin hyperpigmentation and hyperfunctioning endocrinopathies, such as precocious puberty.

The treatment of the disease is symptomatic both from an endocrinological and orthopedic point of view.

– True precocious puberty is usually associated with brain lesions due to tumor processes, hydrocephalus, hypothalamic hamartomas, post-encephalitis sequelae, tuberous sclerosis, etc. [2]. All these conditions present with neurological manifestations and intracranial hypertension, were absent in our case. Even true precocious puberty due to the Silver-Russell syndrome can be excluded, despite the asymmetry of the lower limbs. In fact this syndrome presents with intrauterine and postnatal growth retardation, particular faces, megacephalia, absent in our patient.

  Tumors producing gonadotropins (hepatomas, hepatoblastomas, teratomas, etc.) can cause precocious puberty, but in such situations abdominal ultrasound

shows images of tumor lesions and are not accompanied by “café au lait” patches nor polyostotic bone lesions.
– The presence of bone manifestations and “café au lait” patches may suggest a Type 1 Neurofibromatosis, however we have already commented on the clinical differences with this disease.
– A fibrous cystic osteitis can show bone radiological images similar to those observed in this patient, but in our patient the endocrinological and radiological manifestations are missing. For the same reasons, we can exclude Ollier enchondromatosis, as well as benign bone tumor of a different nature [3, 4].

Key Points
- Signs of precocious puberty associated with large “café-au-lait” patches should lead to a suspected McCune-Albright syndrome.
- McCune-Albright syndrome “café-au-lait” patches are generally large, and always have irregular and jagged edges, like the Maine coast.
- The association of suspected precocious puberty and “café-au-lait” patches suggests that polyostotic bone dysplasia should be investigated by radiographic examination of long bones

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Chapter 33
Well Defined Round Nodules of the Thigh

Mohamed Saeed Mohamed, Shady M. Ibrahim, and Mohamed L. Elsaie

Case Presentation

An 11-year-old-girl was seen in the dermatologic clinic due to asymptomatic slightly erythematous nodule on her left frontal aspect of the thigh (Fig. 33.1). The lesion had a history of a 1 month onset following trauma. The patient and her parents reported no other associated symptoms or significant medical history.

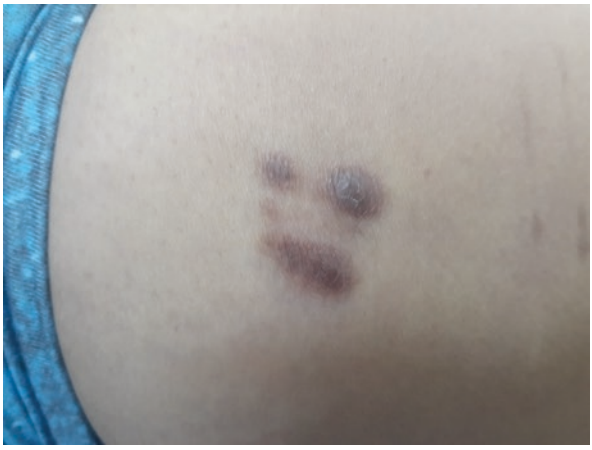

Fig. 33.1 Asymptomatic slightly erythematous nodules on the left frontal aspect of the thigh

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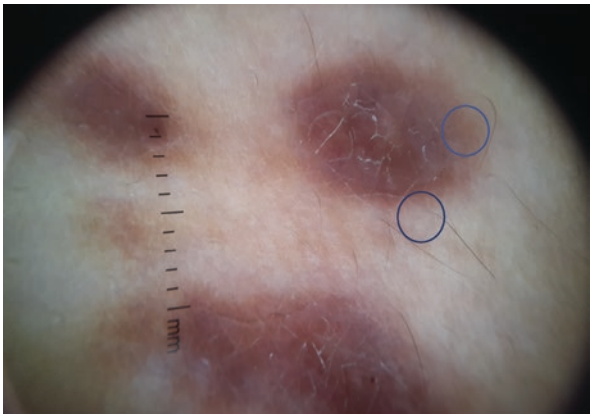

Fig. 33.2 Dermoscopic examination of the nodule revealed erythematous homogenous area with linear and arborizing vessels (circle) arranged in scattered manner (Dermoscopy 3gen DermLite 4, magnification 10×)

On physical examination of the skin, he had three erythematous well-defined round nodules on her left frontal aspect of the thigh. Examination of other areas of the skin, hair, nail and oral cavity showed no abnormality. There is no evidence of apparent lymph node enlargement.

Dermoscopic examination of the nodule revealed erythematous homogenous area with linear and arborizing vessels arranged in scattered manner (Fig. 33.2).

Based on the case description, clinical and dermoscopic photographs, what is your diagnosis?

1. Dermatofibroma.
2. Atrophic scar
3. Hypertrophic scar.
4. Keloid.

Diagnosis

Keloid

Discussion

Keloids and hypertrophic scars are abnormal wound responses characterized by the overgrowth of fibroblastic tissues during skin healing. They not only cause esthetic problems, but also symptomatic problems such as pruritus and pain [1]. Keloids and hypertrophic scars are thought to have the same clinical course [2, 3].

Dermoscopic examination with immersion fluid (e.g. alcohol) on the lesion eliminates surface reflection and renders the stratum corneum translucent, thus allowing the examiner to observe the size and shape of the blood vessels of the superficial vascular plexus [4, 5]

The dermoscopic features of keloids based on their characteristic features and they could demonstrate distinctive dermoscopic features especially in keloids where vascular structures including arborizing, linear irregular and comma shaped vessels were frequently seen [6].

Key Points

- Keloids and hypertrophic scars are abnormal wound responses characterized by the overgrowth of fibroblastic tissues during skin healing.
- Dermoscopy can differentiate between keloid and hypertrophic scar based on vascular patterns.

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Chapter 34
Widespread Itchy Dermatitis in an 8-Year-Old Girl

Antonio Iannone, Laura Ciulli, and Giuseppe Ruggiero

The patient an 8-year-old girl was found to have itchy erythematous-papular and crusted lesions on the extensor and flexor surface of the limbs, lumbar sacral region, buttocks and trunk. They had appeared about 1 month before and were grouped in ovalar or rounded patches with clear signs of scratching (Figs. 34.1 and 34.2). They were symmetric and in some areas showed a "cluster" arrangement. The patient has been affected by atopic dermatitis since her first month of life.

The dermoscopic examination shows white scales, yellow crusts and red dots with a patches pattern (Fig. 34.3).

Based on the case description and the photographs what is your diagnosis ?

1. Nummular eczema (nummular dermatitis)
2. Dermatitis herpetiformis
3. Scabies

Diagnosis

Nummular eczema (nummular dermatitis)

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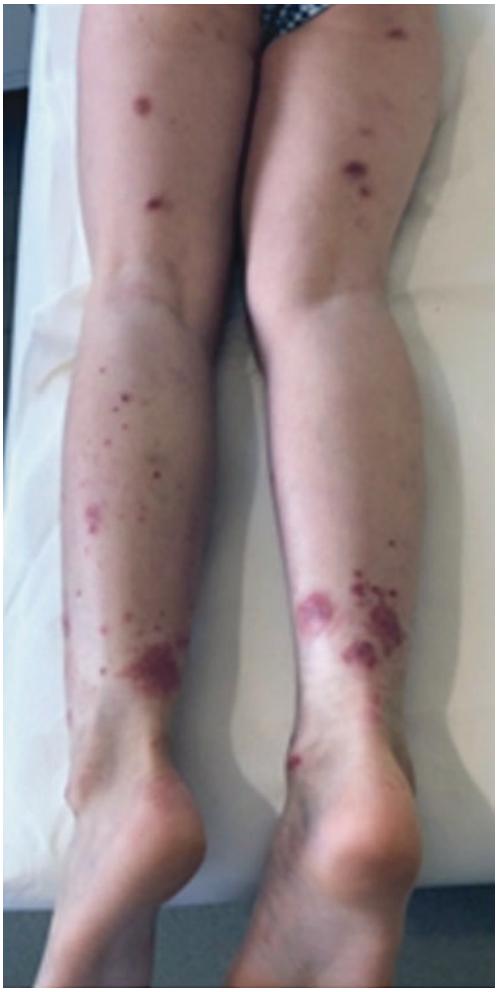

Fig. 34.1 Crusted lesions in rounded patches on the lower limbs

Discussion

Nummular eczema is one of the possible clinical presentation of Atopic Dermatitis. The term nummular derives from Latin "nummulus" (coin). It appears as discoid lesions characterized by erythema, scales and an intense crusting most frequently found on the limbs. The lesions are intensely itchy and relatively insensitive to therapies [1]. The dermoscopic examination shows red dot vessels with a pattern of patches and yellowish scales-crusts [2]. Despite the infiltrative and crusty nature of the lesions the intense itching and the abrasions, the healing generally occurs without scarring.

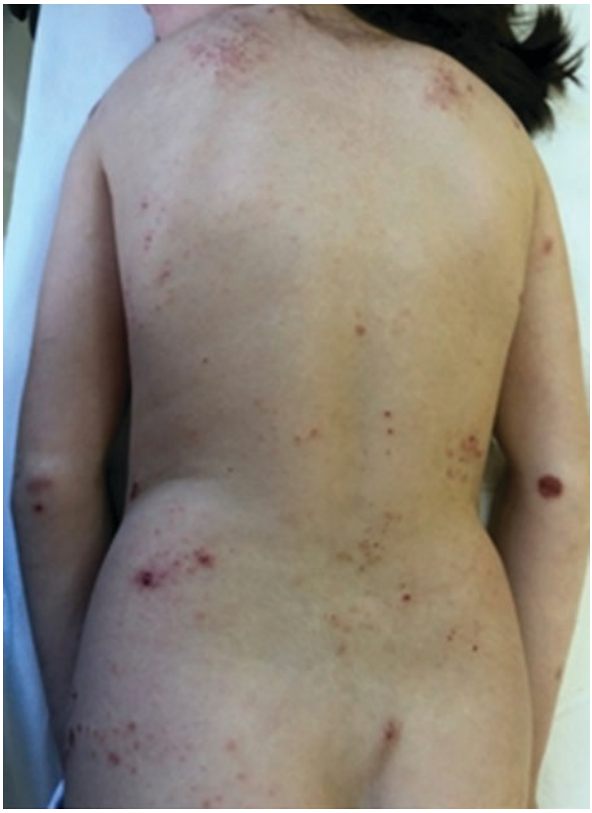

Fig. 34.2 Same lesions, with a clear cluster arrangement, on the upper limbs and trunk

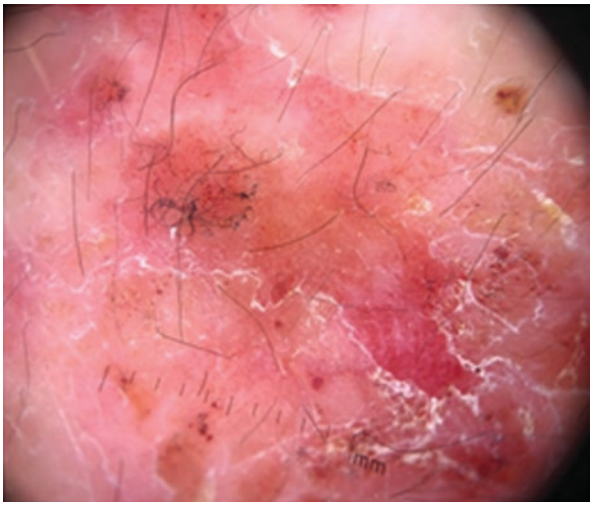

Fig. 34.3 Dermoscopy shows yellow crusts and clusters of red dots and white scales.

Differential Diagnosis

Dermatitis Herpetiformis Given the characteristics and topography of the lesions, the suspicion of dermatitis herpetiformis was justified, but the absence of specific anti-transglutaminase and antiendomysium antibodies make it possible to exclude this diagnosis. It is a sensitive gluten enteropathy, even in the absence of gastrointestinal symptoms. The lesions tend to collect characteristically in clusters with symmetrical distribution and typical involvement of the extensor surface of the limbs, sacral region, buttocks, upper back. A direct immunofluorescence on healthy periwound skin shows a pathognomonic picture that highlights IgA granular deposits at the dermal-epidermal junction of the dermal papillae [3].

Scabies It is an itchy dermatitis which presents erythematous, crusted and excoriated lesions. They are often spread or involve characteristic sites such as hands, wrists, axillae, genital regions and trunk. The itching is intense and persistent, especially during the night. Normally in pediatric age, other members of the family present the same symptoms. Specific lesions such as burrows and pearly vesicles—are not always visible. A dermoscopic examination, which is non-invasive and very easy to perform, can be a valuable aid in diagnosis when the "triangle sign", the "delta wing jet", burrow, eggs etc. are shown [4, 5].

Key Points

- Nummular dermatitis is the most common childhood itchy skin disease
- It is a regular wide spread example of atopic dermatitis
- Dermatitis herpetiformis can present very similar clinical pictures
- Serology for celiac disease is very useful for differential diagnosis between atopic dermatitis with nummular expression and dermatitis herpetiformis
- The knowledge of dermoscopy, as a new reliable diagnostic tool, is useful.

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Chapter 35
Xeroderma Pigmentosum

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A 6-year-old boy came to the Dermatology and Venereology outpatient clinic with a chief complaint of black spots on his face, neck, arms, and lower legs. The patient complained of black spots on his skin since 4 years ago. At first, the spots appear on the face, then aggravated and extended into the neck, hands, and feet. He also complained had red and watering eyes.

The patient is the fourth child in the family, the first brother suffered the same complaint, then died when he was 12 years old. The second brother also had the same complaint, currently 18 years old, while the patient's third brother does not have the same complaint. Both patient's older siblings were diagnosed with xeroderma pigmentosum. The social history showed that there was an inbreeding, which is the grandfather of his mother and the grandfather of his father are siblings (Fig. 35.1).

Based on the case description and the photographs, what is your diagnosis?

1. Xeroderma pigmentosum
2. Acute Cutaneous Lupus Erythematosus
3. LEOPARD syndrome
4. Rothmund-Thomson Syndrome
5. Cockayne Syndrome

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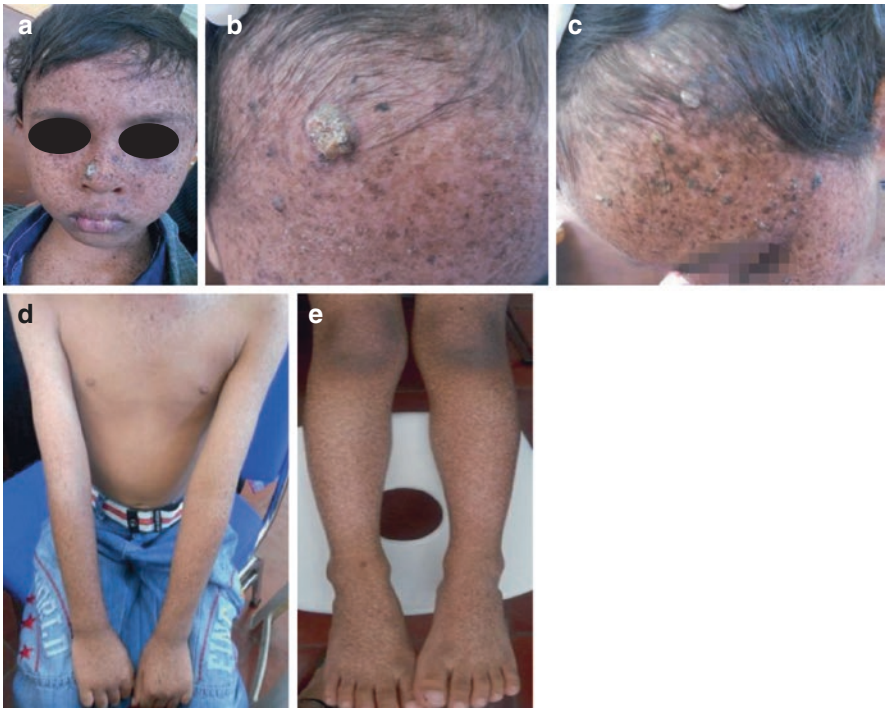

Fig. 35.1 (**a**–**e**) Hyperpigmentation and hypopigmentation macule on face, extremities, and multiple nodules on the face

On physical examination, we observed multiple hyperpigmentation and hypopigmentation macule, well-defined margin, vary in size, certain area followed by thin scale. On the face there are multiple nodules, varying in size.

From histopathological examination showed skin tissue consisting of the epidermis and superficial dermis (papillary dermis). No subcutaneous fat appears. The acanthotic epidermis with a surface showed hyperkeratosis. In the basal part seen an addition of melanin pigment with an increase in the number of melanocytes. On the papillary dermis, there is a mild perivascular lymphocytic infiltrate, tortuous blood vessels and there are also many melanophages and extracellular melanin pigments. Conclusion: punch biopsy of left upper arm skin, xeroderma pigmentosum (Fig. 35.2).

Diagnosis

Final diagnosis for this patient is Xeroderma Pigmentosum.

The treatment consists of sunscreen SPF 33 (for the face area), sunscreen lotion SPF 30 (for the body area), emollients, patient is advised to avoid exposure to ultraviolet radiation and reduce activity during the day, and using closed clothes if leaving the house, a wide hat and protective glasses, and avoid exposure to cigarette smoke. Patients also were referred to the Pediatrics, Neurology, Ophthalmology, and ENT department.

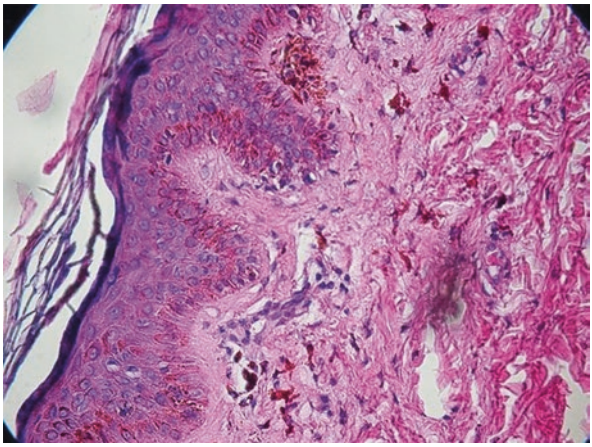

Fig. 35.2 Histopathological xeroderma pigmentosum

Discussion

Xeroderma pigmentosum is an autosomal recessive inherited skin disease. Patients have hypersensitivity to the ultraviolet (UV) rays especially at a wavelength of 290–340 nm. The abnormalities that occur include skin damage due to sun exposure (photodamage), premature photoaging, skin malignancy in sun-exposed areas, eye disorders (80%), neurological disorders (30%), and often cause death due to malignancy [1, 2].

The skin in XP patients is usually normal at birth, 75% of initial symptoms are seen between 6 months and 3 years old. There are acute reactions after minimal sun exposure, such as erythema, edema, bullae then the patient's skin becomes dry and parchment-like followed by scales or peeling of the skin. The accumulation of actinic triggers the occurrence of poikiloderma, which is characterized by skin atrophy, telangiectasis, hyperpigmented and hypopigmented patches. XP patients have a higher risk of developing basal cell carcinoma, squamous cell carcinoma, or melanoma [1, 3]. 80% of XP patients are found to have eye abnormalities. Eye abnormalities are important in XP and often present with skin disorders. Abnormalities are seen clearly on the anterior surface which is exposed to sunlight. Photophobia and conjunctivitis are often present as early symptoms. There are also often seen abnormalities in the form of ectropion, entropion, symblepharon, ulcers, pigmentation of the conjunctiva, pterygium, and corneal opacities. Neurological abnormalities have been reported to occur in 30% of patients. Its onset may be early, or in some patients, it may appear late into the second decade. Abnormalities such as mental retardation, areflexia or hyporeflexia, seizures, sensorineural deafness (starting with high-frequency hearing loss), and electroencephalographic abnormalities [1, 2].

Routine clinical laboratory examination did not show consistent abnormalities. There was no specific feature on histopathological examination. There are variable amounts of epidermal melanin, telangiectasia of the superficial vessels, and mild

perivascular inflammatory reactions. In pigmented areas, there was an irregular accumulation of melanin, increased melanin in the basal layer, Malpighi, and chronic inflammatory infiltration in the top layer of the dermis. In hypopigmented areas, epidermal atrophy is often seen [1, 4].

In this case, from physical examination, family history, and histopathological examination, the patient was diagnosed with Xeroderma pigmentosum. The absence of a family history of XP does not preclude the diagnosis. Molecular testing approaches can include serial single-gene testing use of the multigene panel and more comprehensive genomic testing [5].

Treatment is based on avoiding sun exposure and may include avoidance of being outdoors and the use of sunscreen. Treatment with anticancer drugs, including isotretinoin or fluorouracil can reduce the incidence of skin cancer in XP. However, these drugs have obvious adverse reactions and are not recommended for children. Routine skin examinations every 3–12 months; periodic routine eye and neurologic examinations and audiograms are required.

Patients with XP have a poor prognosis, with about two-thirds dying before age 20 years. Many of these patients will develop multiple malignant tumors within 3–4 years of diagnosis, with widespread metastasis of these tumors resulting in death [5]. The most common causes of death were skin cancer (34%); neurologic degeneration (31%); and internal cancer (17%) [6].

Key Points

- Xeroderma pigmentosum (XP) is a rare autosomal recessive disease
- Patients have hypersensitivity to the ultraviolet (UV) rays
- The abnormalities that occur include premature photoaging, skin malignancy in sun exposed areas, eye disorders, neurological disorders, and often cause death due to malignancy

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