Clinical Cases in Dermatology
Series Editor: Robert A. Norman

Fabio Arcangeli
Torello M. Lotti *Editors*

Clinical Cases in Early-Years Pediatric Dermatology

Springer

Clinical Cases in Dermatology

Series Editor
Robert A. Norman
Tampa, FL, USA

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Clinical Cases in Early-Years Pediatric Dermatology

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Chapter 1
5 Year Old with Fever and Perioral and Periorbital Erythema

Kenan Barut, Defne Özkoca, and Zekayi Kutlubay

A 5 years old male patient applied to the pediatric emergency department with the complaint of fever that has been persistent for 5 days and abdominal colic pain with a resultant diarrhea. The patient's past medical history and family history were unremarkable except for a SARS-CoV-2 infection in the family one-month ago. The patient was asymptomatic during the infectious period. At the time that the patient applied to the emergency department he was lethargic, tachycardic and tachypneic. His vitals were as follows: a body temperature of 38.9 degrees Celsius, a heart rate of 128 beats per minute, a respiratory rate of 30 per minute and a blood pressure of 60/40 mm mercury. Upon oscultation, the first and the second heart sounds were normal but a 2/6 systolic murmur was present. A hepatomegaly of 2 cm was present and the traube space was closed. Submandibular lymphadenopathies were palpated. A cutaneous rash, bilateral conjunctivitis and red strawberry tongue were observed. The laboratory examination revealed decreased white blood cells (5000/mm^3), decreased hemoglobulin (9.1 g/dL), decreased platelets (160,000/mm^3), decreased albumin (2.3 g/dL), increased ferritin (922 ng/mL), increased c-reactive protein (122.5 mg/L) and increased brain-natriuretic peptide (1894 pg/mL). The rest were normal. Echocardiography and cardiac enzymes were unremarkable.

The patient was consulted to dermatology due to the cutaneous findings. Upon the dermatologic examination the patient had a diffuse polymorphous eruption; perioral and periorbital erythema; and fissured lips (shown in Fig. 1.1). The tongue

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Fig. 1.1 Diffuse polymorphous eruption; perioral and periorbital erythema; and fissured lips

was bright red and edematous with the accentuation of the fungiform papillae (red strawberry tongue, shown in Fig. 1.2).

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

- Kawasaki Disease (MIS-C)
- Scarlet Fever
- Viral Exanthema
- Toxic Shock Syndrome

Diagnosis: Kawasaki Disease (MIS-C)

Given the increased C-reactive protein, leukopenia, increased ferritin, red strawberry tongue, bilateral conjunctival injection, submandibular lymphadenopathies, perioral erythema, fissured lips and polymorphous rash, the patient was diagnosed as Kawasaki Disease due to multisystem inflammatory syndrome in children (MIS-C), related to SARS-CoV-2 infection. Intravenous immunoglobulin, pulse corticosteroid, anakinra and wide spectrum intravenous antibiotherapy were initiated immediately upon the diagnosis. The fever and other symptoms subsided on



Fig. 1.2 Bright red and edematous tongue with the accentuation of the fungiform papillae (red strawberry tongue)

the fifth day of treatment and the patient was discharged on the 10th day of treatment.

Discussion

Kawasaki disease is a medium vessel vasculitis that is diagnosed by its cutaneous findings. It is the most common cause of acquired heart disease in children due to its affinity towards the coronary arteries. A typical patient has a persistent fever for at least 5 days and four of the following five principle manifestations [1]:

- Acute edema of hands and feet, acute erythema of palms and soles, subacute periungal peeling of the finger and the toes
- A polymorphous eruption (maculopapular, urticarial, erythema multiforme-like or erythrodermic, but not vesicular)
- Bilateral conjunctival injection
- Oral changes: Perioral erythema, fissured lips, strawberry tongue, diffuse injection of the oral mucosa
- Cervical lymphadenopaties (>1.5 cm, usually unilateral)

The main differential diagnoses of the disease are scarlet fever, toxic shock syndrome, measles, adenoviral infection or Steven Johnson's Syndrome [1].

The disease is important for its predilection to coronary arteries and may lead to coronary artery dilations or aneurysms. Rarely, macrophage activation syndrome can occur as a result of Kawasaki Disease [1, 2]. Recently, Kawasaki Disease has been linked with the MIS-C due to SARS-CoV-2 infection as well. The main treatment modality used in Kawasaki Disease is intravenous immunoglobulins. The aim of treatment is to overcome the acute systemic inflammatory process and its resultant possible coronary artery damage. In refractory cases, systemic corticosteroids, cyclosporine and anti-interleukin-1 biologic drugs may be used. Furthermore, aspirin is added to the regimen for its anti-platelet effects [2].

Strawberry tongue is the distinctive clinical picture that is observed due to the accentuation of the inflamed fungiform papillae on an erythematous background located on the dorsum of the tongue. It is a diagnostic criterion for the Kawasaki Disease and Scarlet Fever. This specific enanthema occurs due to the desquamation of the keratinized epithelium of the filliform papillae. Other diseases that may present with strawberry tongue are the toxic shock syndrome, group-A streptococcal pharyngitis, recurrent toxin-mediated perianal erythema, recalcitrant erythematous desquamating disorder, yellow fever and Yersinia pseudotuberculosis infection. This enanthema resolves with the treatment of the underlying disease [3].

Scarlet fever is an acute febrile exanthematous and respiratory infection that is caused by the Group-A streptococci (GAS), peaking during the winter and the spring. Streptococcal phayringitis is caused by the erythrogenic toxin of the GAS, which leads to vasodilation. Along with fever and pharyngitis, white followed by red strawberry tongue, a sand paper like rash most prominent at the flexures, circumoral pallor, pastia lines and the peeling desquamation of the hands and the feet are observed. The mainstay of treatment is antibiotherapy with penicillins [3, 4].

Key Points

- Kawasaki disease is a medium vessel vasculitis that is diagnosed by its cutaneous findings. A typical patient has a persistent fever for at least 5 days and four of the five principle manifestations.
- Kawasaki Disease can be seen along with the MIS-C due to SARS-CoV-2 infection.
- The main treatment modality used in Kawasaki Disease is intravenous immunoglobulins; and the aim of treatment is to overcome the acute systemic inflammatory process and its resultant possible coronary artery damage.
- Strawberry tongue is a distinctive clinical picture that is observed due to the accentuation of the inflamed fungiform papillae on an erythematous background; and it is a diagnostic criterion for the Kawasaki Disease and Scarlet Fever.
- Scarlet fever is an acute febrile exanthematous and respiratory infection that is caused by the Group-A streptococci; and it presents fever, pharyngitis, white followed by red strawberry tongue, a sand paper like rash most prominent at the flexures, circumoral pallor, pastia lines and the peeling desquamation of the hands and the feet.

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Chapter 2
A Child with High Fever, Rash, Chapped Lips and Conjunctival Injection

Alfonso Delgado Rubio and Fabio Arcangeli

A Child with High Fever, Rash, Chapped Lips and Conjunctival Injection

A three-year-old child came to our observation because he had had a high fever, intermittent vomiting and cough, for seven days. Five days earlier he had recived the diagnosis of acute bronchitis in an other clinic and was treated with amoxicillin, without any effect.

On clinical examination the child presented very irritable, with fever at 39.5 °C, mild rigidity of the neck and bilateral cervical lymphadenopathy. He also had a diffuse asymptomatic maculo-papular rash (Fig. 2.1), bilateral conjunctival injection without secretion (Fig. 2.2), dry, chapped lips (Fig. 2.3) and strawberry tongue.

Laboratory tests show several abnormal values: Hb 11.9 g/dl, leukocytes 19,820/mm^3. Erythrocyte Sedimentation Rate 95, C-reactive protein 17.5 mg/dl. The blood cultures were negative. The CSF examination showed a clear appearance with 55 cells/mm^3 (94% lymphocytes). The throat swab was negative. The serologies for CMV, EBV, Parvovirus B19, Toxoplasmosis and Rickettsia were negative. The ECG and the echocardiogram resulted normal.

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Fig. 2.1 Diffuse maculo-papular rash

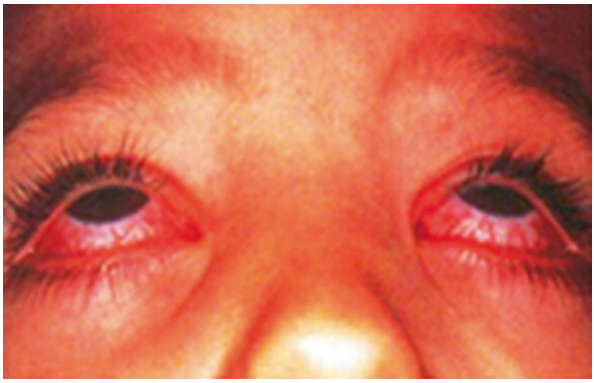


Fig. 2.2 Conjunctival injection without exudate



Fig. 2.3 Erythematous, dry, chapped lips

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

1. Classic exanthematous rash
2. Infectious Mononucleosis and Mononucleosis-like pictures
3. Drug reaction
4. Kawasaki disease

Diagnosis

Kawasaki disease (KD).

Discussion

Based on the clinical pictures and laboratory tests, we diagnosed Kawasaki disease and started therapy with aspirin (80 mg/kg/day) and intravenous gamma globulin (IVIG) at 2 g/kg in a single dose to prevent coronary artery abnormalities.

A few days later, hard edema of the hands and feet appeared, the exanthema improved, and after one week lamellar desquamation of the hands and the perineal region became evident.

The presence of a febrile syndrome and an exanthematous rash could have suggested a diagnosis of a classic exanthematous disease. However, our patient received the normal vaccinations. Furthermore, the clinical presentation did not point to any of the classic exanthematous diseases.

It should have been possible to think of a diagnosis of Mononucleosis or a Mononucleosis-like syndrome or even of an amoxicillin rash during of Mononucleosis. The clinical features and the serological investigations ruled out these diagnoses.

In the case of a drug hypersensitivity reaction, itching is frequently present, the fever is moderate and short. In addition, the laboratory tests show eosinophilia and an increase in transaminases, absent in our patient.

The appearance of an exanthematic eruption associated with a prolonged febrile syndrome that does not respond to antibiotics or antipyretics in a young child suggests a diagnosis of Kawasaki disease [1]. If bilateral conjunctival injection, oropharyngeal erythema, induration of the hands and feet, dry lips are present this diagnosis is more probable [1, 2].

In these cases, treatment with aspirin and IVIG should begin as soon as possible.

The diagnosis of Kawasaki disease is clinical and is based on defined criteria (Table 2.1) [3, 4].

In some cases, patients do not fulfill the classic criteria for Kawasaki disease and are classified as having incomplete (atypical) disease. This occurs in about 9.6% of cases. Atypical disease is suspected when patients have a fever for at least five days with only two or three of the principal clinical features [5, 6]. It is important to consider the diagnosis of Kawasaki disease and perform echocardiography in all infants younger than six months who have an unexplained fever lasting at least seven days with laboratory evidence of systemic inflammation [7] (Table 2.1).

Key Points

- Classic Kawasaki disease must be suspected in children with high fever of five days or more with at least four of five features: bilateral conjunctival injection, changes in the lips and oral cavity, cervical lymphadenopathy, polymorphous rash and extremity changes
- An early diagnosis of Kawasaki disease is very important because the treatment with aspirin and IVIG should begin as soon as possible.to prevent coronary abnormalities.

Table 2.1 Diagnostic criteria for classic Kawasaki disease [3, 4]

Diagnostic criteria for classic Kawasaki disease	Frequency (%)
Fever for at least five days and at least four of five principal clinical features listed below	100
1. Changes of the oral cavity and lips: cracked and erythematous lips, strawberry tongue	96.5
2. Polymorphous rash: maculo-papular, erythema multiforme–like or scarlatiniform rash, involving extremities, trunk, and perineal regions	96.0
3. Bilateral conjunctivitis, nonpurulent	89.0
4. Changes in the extremities (erythema of the hands and feet, desquamation of the hands and toes in weeks 2 and 3)	75.6
5. Cervical lymphadenopathy (> 1.5 cm in diameter), generally unilateral	62.7
Alternative diagnostic criteria for classic Kawasaki disease	
Fever for at least five days and two or three principal features; coronary artery abnormalities on transthoracic echocardiography	

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Chapter 3
A Five-Year-Old Girl with Erythematous Papules on Cheek

Xiangjin Song, Lihong Zhao, and Songmei Geng

A 5-year-old Chinese girl presented with erythematous papules located in her right cheek for six months. Her parents complained of tiny stab and wound occurred in site before. The papules gradually expanded and fused into erythematous plaques (Fig. 3.1).

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

1. Cutaneous Rosai-Dorfman disease
2. Cutaneous lupus erythematosus
3. Tinea Faciei
4. Majocchi granuloma

Physical examination demonstrated good general condition and no obvious abnormality of other internal organs. Routine laboratory work-up showed negative results or within normality limits. Biopsy from erythema papules on her cheek found that the epidermis was almost normal while granuloma consisting of histiocytes and plasma cells infiltrated around hair follicles in the dermis (Fig. 3.2). Gomori's methenamine silver staining (GMS) showed positive spores in granuloma. Based on clinical features and positive staining for fungi spores in tissues, the diagnosis of Majocchi granuloma was made. Oral terbinafine, 0.25 g per day, was prescribed to the patient. After 1 month follow up, the skin lesions were significantly improved and faded away.

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Fig. 3.1 Clinical manifestation of the patient. Red maculopapules on her right cheek

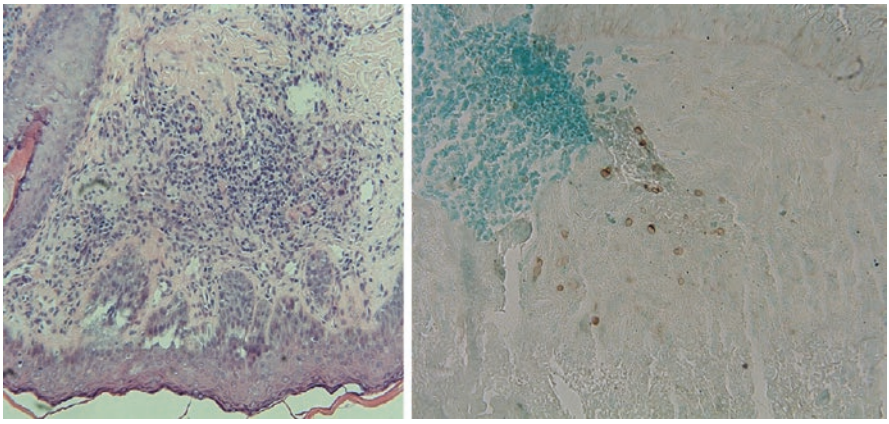


Fig. 3.2 Histopathology (HE×10) and GMS staining of the papules

Diagnosis

Mycotic Granuloma (Majocchi granuloma).

Discussion

Majocchi granuloma is a rare dermatophyte infection of dermis and subcutaneous tissues characterized by erythematous papules, stiffened plaques or granulomatous nodules [1]. Skin lesions usually presents on the extension of the thigh, some cases have reported groin, facial area and vulva may also be involved. The disease is

caused by infection of dermatophyte, especially trichophyton rubrum. Its risk factors include friction, trauma, and use of systemic immunosuppressants [2].

There are two clinical types of Majocchi granuloma: (i) small perifollicular popular form and (ii) deep subcutaneous nodular form [3]. The first type is usually secondary to trauma or the use of topical potent steroid, mainly occurs in healthy individuals. It's defined as a perifollicular infiltration and deep pustular type, erythema, papules and pustules on the smooth skin. Similarly to kerion celsii, scabs and purulent secretions can be seen in the local region. The second type is granulomatous secondary to immunosuppression and characterized by purplish red subcutaneous nodules, which can be fused into plaques. Nodules can be firm or fluctuant and usually present on the upper limbs.

Histopathological examination usually shows as a granulomatous folliculitis, ruptured hair follicle, hyphae and spores can be seen in the dermis. A large number of acute inflammatory cells on the basis of chronic inflammatory cell infiltration and podocytomatous granuloma are a characteristic change. Periodic acid-Schiff (PAS)and GMS methods are generally used to stain fungi. Molecular-base techniques fungal culture are helpful to define fungus [4].

For treatment, topical antifungal therapy usually is ineffective and systemic antifungal therapy is necessary. Terbinafine (250 mg/day), itraconazole (100–200 mg/day), griseofulvin (250–500 mg/day), voriconazole, and posaconazole were proved effective in treatment of Majocchi granuloma. [4] Treatment duration should be sufficient, usually takes 1–2 months, sometimes even as long as 6 months or longer until the lesions are completely cured and no mycology was detected repeatedly.

Cutaneous Rosai-Dorfman disease (CRDD) is an uncommon histiocytic disease with unknown etiology, often presents as solitary or numerous papules or plaques, mostly on facial area which is easily misdiagnosed as tinea. Histopathological examination is characterized by dense cellular infiltration in dermis. The infiltrating cells appear as large, eosinophilic histiocytes with abundant cytoplasm, lymphocytes and a few plasma cells can also be detected. Histiocyte's phagocytosis of lymphocytes and the proliferation of vasculars are features of CRDD. Histiocytes usually stained positive for S-100 protein and negative for CD1a.

Cutaneous lupus erythematosus (CLE) is a chronic autoimmune disease that encompasses a great variety of dermatologic manifestations such as discoid lupus or erythema annulare with papules and scales. The face is easily affected site and should be differentiated from infectious disease. The characteristic of histopathological examination are the presence of interface dermatitis with vacuolar changes of the basal keratinocytes and necrotic keratinocytes, mucin deposition, and variably dense lymphoid cell infiltrations [5].

Tinea Faciei is common superficial fungal infection manifested with red papules and scales. Most patients may have a history of using topical corticosteroids. Mycology direct microscopic examination is frequently used to make diagnose.

In this case, she presents with erythematous plaques and papules on her right cheek for 6 months after tiny stab. Histopathological examination with GMS stains confirmed the diagnosis and antifungal therapy was effective.

Key Points

- There are two forms of Majocchi granuloma: (i) small perifollicular popular form and (ii) deep subcutaneous nodular form [3].
- The diagnosis of Majocchi granuloma is usually based on history, clinical manifestations and histopathology. Molecular-base techniques and fungal culture is also required sometimes.
- The treatment of Majocchi granuloma need to be sufficient.

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Chapter 4
A Four- Year- Old Girl with Otalgia

Domenico Di Maria, Giuseppe Ruggiero, and Fabio Arcangeli

A four-year-old caucasian girl complained of ear fullness, mild otalgia and poor otorrhea. The symptoms began in September and the girl came to ENT observation two months later.

At symptoms onset, the patient was seen by a pediatrician who prescribed oral antibiotic and cortisone therapy. After about two weeks the symptoms showed up again and therefore the same pediatrician prescribed intramuscular antibiotic and oral cortisone. Despite this the therapy was not effective and for this reason the mother took her daughter to the otolaryngologist.

The video-otoscopy showed the skin of the external auditory canal and the outer face of the left tympanic membrane hyperemic and covered with a whitish "patchy" cloth (Fig. 4.1). The tympanometry was normal for both ears. The latter therefore excluded middle ear involvement (effusive otitis media, acute otitis media).

The clinical history documented that the symptoms began in September after a few days returning from a beach holiday.

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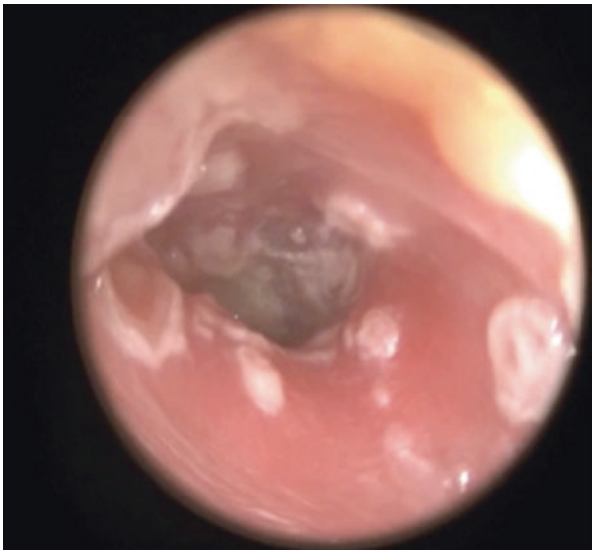


Fig. 4.1 Oto-endoscopic aspect of the left ear

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

1. Acute bacterial external otitis
2. Otomycosis
3. Acute otitis media

Diagnosis

Otomycosis.

Discussion

The clinical history and the failure to respond to antibiotics and steroids therapy suggested the non-bacterial genesis of the disease. The normal tympanometry of both ears excluded a middle ear involvement.

The skin of the external auditory canal and the outer face of the left tympanic membrane appeared hyperemic and covered with a whitish "patchy" cloth. This is a typical feature of otomycosis.

Otomycosis is a fungal infection of the external ear canal. It has been estimated that otitis externa make up 5 to 20% of ear-related visits to ENTs. Most of these infections are caused by bacteria but 9 to 25% are caused by fungi (otomycosis)

[1–3]. The otomycosis involves the external ear canal squamous epithelium and is characterized by pruritus, occasional otalgia and sometimes hypoacusia [3, 4]. Predisposing factors are considered: a failure in the ear's defense mechanisms (changes in the coating epithelium, changes in pH, quantitative and qualitative changes in ear wax), bacterial infections, hearing prosthesis, self-inflicted trauma (use of q-tips to clean the ear), swimming, broad spectrum antibiotic agents, steroids and cytostatic medications, neoplasia and immune disorders. All of these can cause an highest susceptibility to the fungal infections [4, 5].

In pediatric age, a relevant aspect of antifungal therapy is the handling and safety of the chosen drug. Our patient was treated with Ozoile (Stable Ozonides with Vitamin E acetate) which acts as a biological inducer and regulates the main metabolic pathways. Ozoile also stimulates the endogenous defense system and through the regulation of gene transcription promotes tissue regeneration and damage-injury repair.

It has a broad-spectrum microbicide activity due to its high affinity for the lipoprotein components of the bacterial and fungal wall and for the oxidizing action (Table 4.1).

After three weeks of therapy, complete clinical recovery was achieved without any complications (Fig. 4.2).

Table 4.1 The broad-spectrum activity of Ozoile

Time (h)	4	12	24	48
Sample	OZOILE	OZOILE	OZOILE	OZOILE
E. coil	99,40	99,99	98,20	98,20
P. aeruginosa	99,99	99,99	98,99	98,99
S. aureus	99,07	99,99	99,70	99,70
C. abicans	99,99	99,99	99,99	99,99
C. glabrata	99,99	99,99	99,99	99,99
A. niger	–	–	–	–
G. vaginalis	87,50	99,99	99,99	99,99
P. mirabilis	96,47	99,99	99,99	99,99
P. acnes	99,99	99,99	99,99	99,99
E. faecalis	82,77	99,99	99,99	99,99
S. epidermidis	99,99	99,99	99,99	99,99
T. mentacrophytes	99,99	99,99	99,99	99,99
S. agalactiae	48,75	99,99	99,99	99,99
E. cloacae	68,60	99,99	99,99	99,99
B. cepacia	90,00	99,99	99,99	99,99
k. oxytoca	99,99	99,99	99,99	99,99
A. baumannii	99,99	99,99	99,99	99,99
C. pseudodiphterticum	99,99	99,99	99,99	99,99



Fig. 4.2 Complete clinical recovery after three weeks of Ozoile therapy

Key Points

- Otomycosis is an infection that involves the external ear canal epithelium, characterized by itching and occasionally otalgia
- Otomycosis is more frequent in the hot or humid seasons and climates
- A careful consideration of the clinical history, the video-otoscopy and a normal tympanometry are useful for the diagnosis
- The topical therapy is gold standard and Ozoile is a safe and effective treatment

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Chapter 5
A Little Boy with Facial Erythema

Zhen-Ting Lin, Hao Guo, Jing Lan, Xing-Hua Gao, and Jiu-Hong Li

A 5-year-8-month-old boy presented to the office with his parents complaining of his purplish erythema on his face for 6 months after birth, which was obvious after heat stimulation (Fig. 5.1a). One year ago, the boy developed generalized erythema with scaling and itching, evident in both upper limbs and shoulders (Fig. 5.1b). Generalized erythematous scaling with papular nodules were noticed on his lower limbs for 1 year, which were hard and non-tender. There was an egg-sized nodular mass under the left iliac spine, with brown pigmentation on the surface of the mass and central ulceration and scab (Fig. 5.1c, d).

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

1. Juvenile dermatomyositis
2. Systemic lupus erythematosus
3. Systemic sclerosis
4. Mixed connective tissue disease

He was hospitalized, the boy had symptom relief with oral hormones and immunomodulatory drugs for around 10 days, and the patient's skin lesions were relieved and the condition was improved.

Biopsies for muscle pathology were obtained from one nodule on the buttocks. Pathological examinations showed abundant lymphocytes with scattered plasma cell infiltration and massive calcification deposited in the subcutaneous fat lobule (Fig. 5.2).

The little boy's serum muscle enzyme tests showed elevated such as CK, LDH, AST, the little boy's AST was 46 U/L, with a normal range of 15 ~ 40 U/L. LDH was 299 U/L, and a normal range of 120 ~ 250 U/L.

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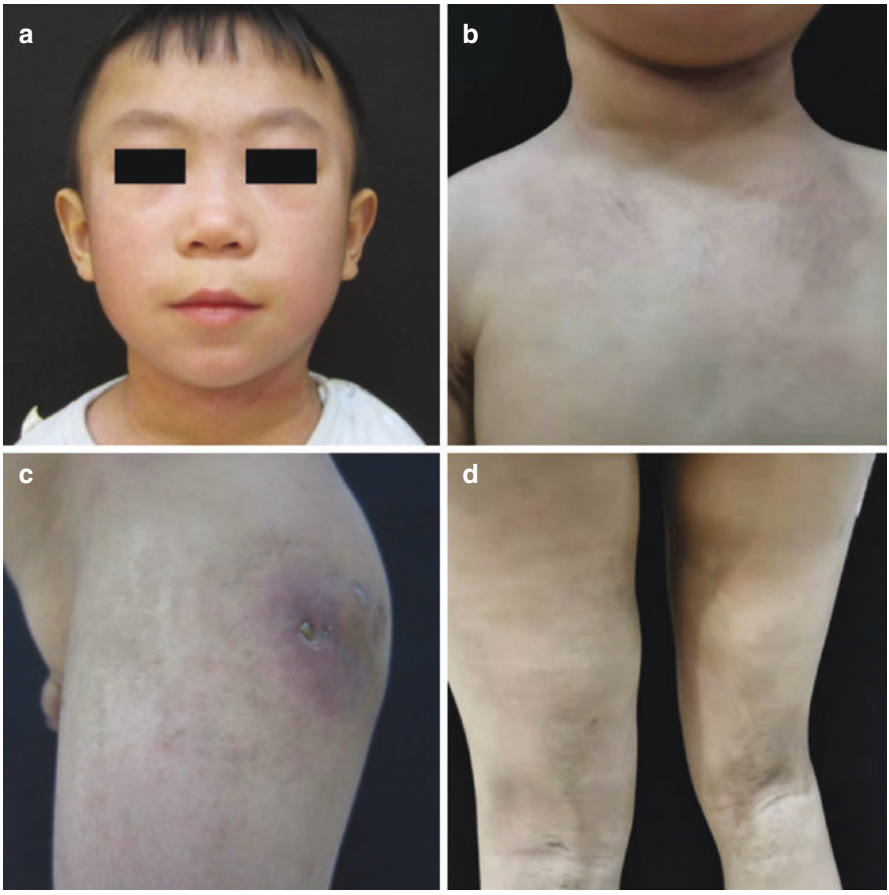


Fig. 5.1 Clinical manifestation of the patient. (**a**) purplish erythema on his face; (**b**) generalized erythema on the trunk; (**c**) an egg-sized nodular on the thigh with pigmentation and central ulceration; (**d**) pigmentations on the lower limbs

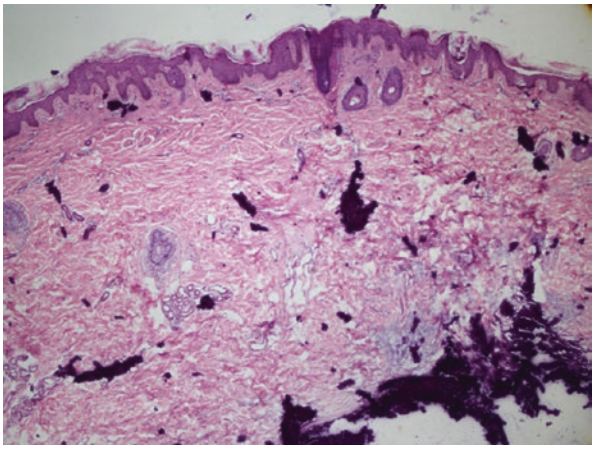


Fig. 5.2 Lymphocyte and scattered plasma cell infiltration in subcutaneous fat lobule, massive calcification (HE×10)

Diagnosis

Juvenile dermatomyositis (Dermatomyositis with calcinosis).

Discussion

Dermatomyositis is an autoimmune connective tissue disease involving the skin and striated muscle, usually including both skin and muscle changes, which can also present as a single aspect of changes. Dermatomyositis can be seen as a spectrum of disease, ranging from isolated inflammatory myositis to dermatomyositis with only cutaneous manifestations, which can be divided into dermatomyositis, polymyositis, and inclusion body myositis. Dermatomyositis can be divided into adult dermatomyositis, dermatomyositis associated with malignancy, juvenile dermatomyositis, dermatomyositis in overlap syndrome, and no myopathic dermatomyositis. Polymyositis is divided into single polymyositis and polymyositis in overlap syndrome.

The early symptoms of patients with dermatomyositis are often generalized weakness and muscle pain, followed by Raynaud's phenomenon and arthralgia. Lesions are unrelated to the degree of clinical manifestations of myositis. Typical lesions of dermatomyositis present as dark violaceous edematous macules on both upper eyelids and the face as well as Gottron's papules and Gottron's sign, that is, red or dark violaceous milia to mung bean large polygonal, irregular flat, or spiked papules with clusters of integrated groups or coalescing into small plaques on the extensor surfaces of the metacarpophalangeal, proximal, and distal interphalangeal joints. Gottron's sign or Gottron's papules rarely appear alone, mostly accompanied by dark violaceous edematous macules on the upper eyelid, and other dermatomyositis rashes such as poikiloderma-like rashes appear simultaneously. Meanwhile, symmetrical proximal muscle weakness is the main clinical manifestation of myositis. The commonly invaded muscle groups are the shoulder girdle muscles, proximal limb muscle groups, neck muscle groups, and throat muscle groups, with corresponding movement difficulties such as raising hands, squatting, stepping up, raising head, swallowing and hoarseness or nasal sounds.

Juvenile dermatomyositis is a symmetrical inflammatory myositis primarily affecting proximal muscles with characteristic skin lesions. Heliotrope rash, Gottron's papules and Gottron's sign tend to be related with disease activity, although Gottron's sign may persist after achieving remission. Juvenile dermatomyositis is divided into two types, Banker type and brunsting type. The brunsting type is generally a relatively benign type, often accompanied by calcinosis phenomenon. The most significant feature of the banker type is extensive vasculitis involving arterioles, capillaries, and veins of the skin, muscles, subcutaneous tissue, and digestive tract. Compared with the brunsting type, the banker type is generally more acute, has more dysphagia, intestinal ulcers, will present with fever and

leukocytosis and the results of ineffective glucocorticoid therapy. Calcification is the abnormal deposition of insoluble calcium salts in tissues including skin, subcutaneous tissue, tendons, fascia, and muscles. Calcinosis is more common in juvenile dermatomyositis, in contrast to dermatomyositis, which is a unique cutaneous sign [1].

A variety of specific autoantibodies can be detected in patients, and the most common positive autoantibodies are anti-Jo-1, anti-Mi-2, and anti-MDA5 antibodies. Perivascular lymphocytic infiltration in the diseased muscle with IgG, IgM, C3, and C5b-9 deposits in the vessel wall. Besides in contrast to adult dermatomyositis, there is no association of the autoantibody with malignancies in childhood. Anti-MJ/NXP2 antibodies are associated with severe muscle weakness and calcinosis [2].

Electron microscopic observation revealed interstitial inclusions in endothelial cells. Muscle fiber size, necrotic and regenerating fibers, and inflammatory cell infiltration in perivascular and perimysial regions all differ in a large number of patients, but none of these are specific to juvenile dermatomyositis.

Perifascicular atrophy and microinfarction are unique to dermatomyositis. Others specific to juvenile dermatomyositis include expression of type I interferon-inducible molecules, such as myxoma resistance protein (MxA), and deposition of membrane attack complex (MAC) with or without immunoglobulin association in immunohistochemical studies [2].

Systemic sclerosis (SSc), also known as scleroderma, is a chronic and complex connective tissue disease characterized by microvascular endothelial cell injury and progressive fibrosis of the skin and internal organs (lungs, heart, and kidneys). Compared with dermatomyositis, patients suffering from systemic sclerosis mostly have clinical manifestations such as Raynaud's phenomenon, finger sclerosis, pulp atrophy, and fingertip ulcers. Systemic sclerosis occurs predominantly in young and middle-aged women. The ratio of male to female incidence is approximately 1:4.7, and conversely, juvenile dermatomyositis patients are generally younger than 16 years of age [3].

The skin lesions of Systemic lupus erythematosus occur between the dorsal joints of the fingers, pulp of the fingers and toes, while the skin lesions of dermatomyositis tend to occur on the extended surface of the joints, and the erythema color is peculiar, which are dark violaceous or red wine-colored edematous macules [3].

Mixed connective tissue disease also has facial edematous erythema and severe myositis, but the patient's face and fingers are mostly swollen, almost all have Raynaud's phenomenon, have high anti-U1-RNP antibodies, and lack the specific rash of dermatomyositis, and the patient's age is generally 40 years [4].

Based on the patient history, clinical mafifestations and histological examinations, the diagnosis of juvenile dermatomyositis was made.

The patient was instructed to avoid sun exposure and keep the leisions dry and clean. After discharge, the patient continued to take oral triamcinolone 8 mg qd and oral pidotimod oral solution 0.4 mg qd, followed by outpatient reexamination 2 weeks later.

Key Points

- Dermatomyositis is an autoimmune connective tissue disease involving the skin and striated muscle, usually including both skin and muscle changes.
- Calcinosis is more common in juvenile dermatomyositis, in contrast to dermatomyositis, which is a unique cutaneous sign.

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Chapter 6
A Pediatric Case with Erythematous Plaques and Palmoplantar Keratoderma

Githa Rahmayunita, Rahadi Rihatmadja, Triana Agustin, and Rinadewi Astriningrum

A 4-year-old boy was referred to a pediatric dermatology outpatient clinic with mildly pruritic, widespread erythematous plaques and thickened skin of palms and soles of one-month duration. Previous treatment with topical corticosteroids and emollient failed to give satisfactory results. He suffered from mild pruritus that did not impair his daily activity. The patient was born to non-consanguineous parents, without a history of serious illness. Physical examination revealed multiple erythematous papules and plaques on the scalp, forehead, chest, abdomen, and back with sparing on some areas. Waxy palmoplantar hyperkeratosis was also observed (Figs. 6.1, 6.2, and 6.3). No abnormality of the internal organ was detected.

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Fig. 6.1 Coalescing erythematous papules and plaques leaving scattered islands of normal skin within on the trunk



Fig. 6.2 Characteristic salmon hue was difficult to assess in darker skin



Fig. 6.3 Waxy, palmoplantar keratoderma

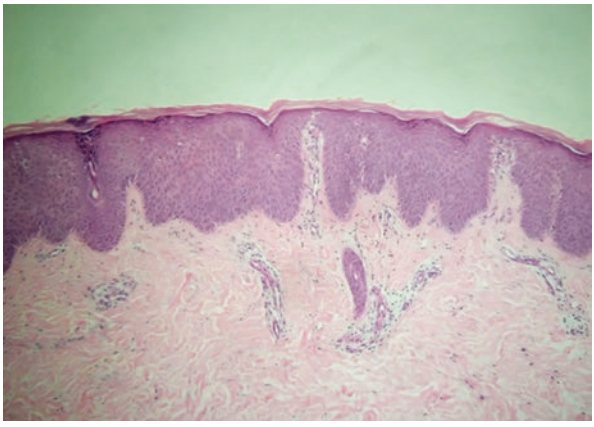


Fig. 6.4 Psoriasiform hyperplasia, suprapapillary thinning (HE, 50X)

Histopathology showed psoriasiform hyperplasia, suprapapillary thinning, papillomatosis, alternating parakeratosis, but lacking spongiform pustules (Figs. 6.4 and 6.5). Laboratory results were unremarkable.

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

A. Psoriasis vulgaris
B. Pityriasis rubra pilaris
C. Lichen planus
D. Pityriasis lichenoides
E. Mycosis fungoides

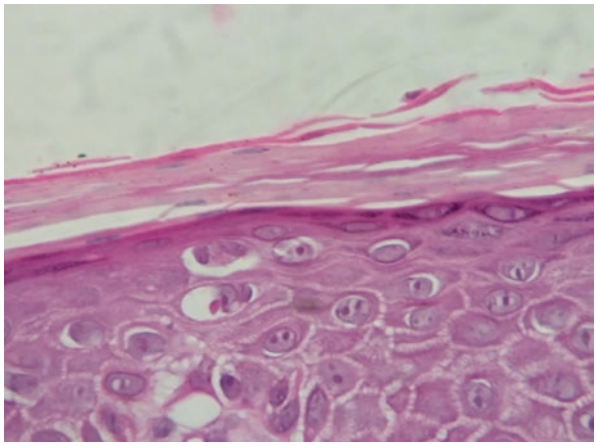


Fig. 6.5 Checkerboard parakeratosis (HE, 400X)

Treatment with methotrexate yielded no significant improvement so that we switched to narrow-band ultraviolet B phototherapy (NBUVB) with starting dose 200 mJ/cm^2 twice weekly. After 6 weeks there was significant reduction (highest dose was 1125 mJ/cm^2) and we had started to decrease the dose and frequency. Currently phototherapy still continues with a dose of 390 mJ/cm^2 fortnightly. He has been receiving phototherapy for 17 months without side effects.

Discussion

Pityriasis rubra pilaris (PRP) is a chronic dermatitis characterized by follicular papules, yellowish-pink plaques surrounding islands of normal skin and palmoplantar hyperkeratosis. Most pediatric cases are acquired. Sporadic forms show CARD14 activation without mutation and indistinguishable from hereditary form [1]. The most characteristic clinical feature is follicular papule surrounded by salmon-colored erythema. Papules usually coalesce leaving islands of normal skin. It distributed from the head, down from the face to the neck and onto the trunk. PRP must be distinguished from psoriasis. Salmon color is helpful to distinguish the two [1–3]. However, in Indonesian patients, it is not always observed except in lighter skin individuals. Pruritus is only occasional. Histopathology features acanthosis with checkerboard pattern of alternating ortho- and parakeratosis, focal hypergranulosis and broad thickening of the rete ridges [1–3]. According to PRP classification by Griffiths, our patient was categorized as type III (classic juvenile) based on clinical manifestation [1, 2].

Patients with PRP) may respond to emollient, topical corticosteroids, and keratolytic agents (urea, salicylic acids, alpha hydroxy acid). Unresponsive cases may be treated with systemic retinoids, perhaps more effective in children than UVB, methotrexate, cyclosporine, or azathioprine [1–3]. However due to isotretinoin

unavailability in our country, we had tried first with methotrexate. When it failed, NBUVB seemed the next plausible choice. The treatment gave marked reduction in erythema and the extent of the disease after 6 weeks. Its effectiveness in type IV juvenile PRP was also reported by Bragg et al. [4] who observed decrease in erythema and thickness after 10 sessions.

Key Points

- Pityriasis rubra pilaris is characterized clinically by coalescing, salmon-colored papules leaving islands of normal skin in their midst.
- Histologically, parakeratosis in checkerboard patterns is the hallmark. Other features closely resemble psoriasis vulgaris
- There are currently six known types of pityriasis rubra pilaris in the adult and pediatric population, each with its own pattern of distribution and medical background.

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Chapter 7
A Rare Case of Xeroderma Pigmentosum in 3 Years Old Child with Squamous Cell Carcinoma

Rina Gustia, Ennesta Asri, and Jessica Herlianez Saiful

A 3 years-old female child was referred from pediatric department to dermatology outpatient clinic, with chief complaint blackish patches on all over the body since two years ago, and reddish bumps below the left eye that was increased in size progressively since 2 months ago, Parents also notice that patient skin became dry, red, and scaly. One year ago, the blackish patches started to spread to the trunk, and patient started to complaint that whenever her eye exposed to sunlight, it became painful and red. From family history, there was cosanguine marriage pattern from the grandfather and grandmother side. There was no family history with skin malignancy, patient is the oldest among three siblings (Fig. 7.1).

On examination we observed hyperpigmented plaque and macule on all over the body, freckles, with tumour size 7 x 5 x 3 cm, easily bled (Fig. 7.1). Dermoscopy examination showed brown pigmented network that is suitable to actinic keratosis (Fig. 7.2). Histopathology examination done on eye tumour by pediatric oncologist showed cell proliferation with pleomorphic nuclei, vesicular, crude chromatin, nuclei, mitosis can be found. These cells are arranged solidly, some are in the form of islands with horn pearls, including solid cells lymphocytes and PMN leukocytes. This findings is suitable for squamous cell carcinoma.

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Fig. 7.1 Hyperpigmented plaque and macule all over the body, with tumour near the eye with size 7 x 5 x 3 cm

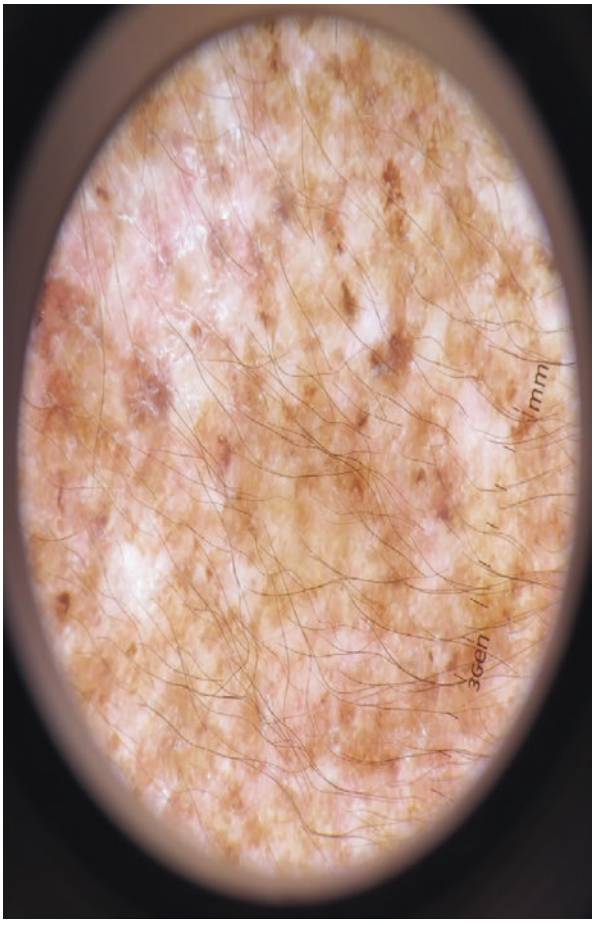


Fig. 7.2 Dermoscopy examination showed Delicate Brown Pigmented Network

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

1. Xeroderma pigmentosum with Squamous cell carcinoma
2. Dyschromatosis symmetrica hereditaria
3. Erythropoietic protoporphyria
4. Freckles
5. Impetigo Contagiosa

Based on medical history, physical examination, dermoscopy and histhopatology examination, **diagnosis** for this patient is Xeroderma pigmentosum with Squamous cell carcinoma.

Treatment: The patient was treated with sunblock SPF 30 and was advised to avoid sun exposure by wearing protective clothing and sunglasses. One of treatment modality for squamous cell carcinoma is wide excision, however, since the lesion located near the eye, this patient was then referred to ophthalmology department for further treatment.

Discussion

Xeroderma Pigmentosum is a hereditary photosensitive disease governed by autosomal recessive inheritance. History of consanguinity is common found in In XP patients. Their skin is extremely vulnerable to UV because of a congenital defect of repair ability for UV-induced DNA damage. Incidence of XP is one in a million in the United States and Europe. XP is more common in Japan and parts of the Middle East. People with XP are unable to repair ultraviolet radiation (UV)-induced DNA damage resulting in an increased risk for skin cancer (10,000-fold) and ocular cancer (1000-fold) in patients under 20 years of age) [1].

Symptoms of XP include hyper- or hypopigmented macules (freckling/lentigos) on UV-exposed skin surfaces, severe sun burns (often with blistering) after minimal sun exposure (50% of patients), photophobia, and corneal clouding. These ocular and cutaneous symptoms often occur before 2 years of age [2].

Xeroderma Pigmentosum occurs because the repair system (nucleotide excision repair [NER] and translesion synthesis) for UV-induced DNA damage (cyclobutane-type pyrimidine dimer, 6-4 photoproduct) does not act properly as a result of genetic abnormality.) [3].

Diagnosis of XP is made based on anamnesis, where there is history of photosensitivity, photophobia, and malignancy that is caused by sun exposure, physical examination, where we could find skin, eye, and neurology abnormalities, and DNA analysis. Family history with consanguinity is also common. Dermoscopy can be additional tool to differentiate benign and malignancy lesion, especially to distinguish generally actinic lentigo on the skin [4].

As this is an inherited disease, a radical cure cannot be expected. The basics of patient care are complete defense from UV and symptomatic treatments for complications [5].

Key Point

Xeroderma Pigmentosum is autosomal resessive inheritance disease in which there is defect of repair ability of UV-induced DNA damage that lead to vulnerability to skin malignancy. The main treatment is complete avoidance from sunlight and total cure cannot be expected.

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Chapter 8
A Young Boy with Fever and Rash

Pierangela Rana and Fabio Arcangeli

A five-year-old boy presented with a low fever and a modestly itchy skin rash. One week before a flu-like syndrome with fever, headache, myalgia and sore throat occurred. He was treated with oral paracetamol 250 mg twice a days. He was vaccinated and had always been in good health prior to the rash.

On physical examination the exanthema consisted of erythematous maculopapules started on the trunk and the limb roots (Figs. 8.1 and 8.2), with an appearance of goose bumps of the skin. Hyperemic and hypertrophic tonsils and modest hepatomegaly were also present. The day after the exanthema spread to the face and to the latero-cervical and retro-auricular regions (Fig. 8.3).

After two days the lesions took on a purpuric colour and the so-called "yellow hand sign" appeared (Fig. 8.4). By exerting a light hand pressure on the abdomen, where the exanthema was clear, a fleeting shape of the hand with a sub-jaundiced

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Fig. 8.1 Erythematous maculo-papular rash on the trunk



Fig. 8.2 Erythematous maculo-papular rash on the lower limb roots



Fig. 8.3 The spreading of erythema to the latero-cervical and retro-auricular regions



Fig. 8.4 The "yellow hand sign"

colour was evident. This was an expression of hepatic suffering and a consequent increase in bilirubinemia.

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

1. Measles
2. Rubella
3. Scarlet fever
4. Fifth disease (Erythema infectiosum)

Diagnosis

Fifth disease (Erythema infectiosum).

Discussion

Measles and rubella could be excluded not only for the different appearance but mainly because the child had received specific vaccines.

The causing agent of scarlet fever is group A beta-haemolytic Streptococcus (S. Pyogenes) producer of the erythrogenic toxin that induces the vasodilation responsible for the scarlet color of the exanthema.

It appears from five years old with an abrupt onset, high fever, pharyngodynia, hypertrophic tonsils with exudate and latero-cervical and sub-mandibular adenomegaly.

The rash begins after one to five days from a feverish onset and starts at the limb roots, then spreads over the entire body surface, typically sparing the perioral region (the Filatov mask). The tongue is white and patinated with red margins, then peels and takes on a raspberry appearance [1].

In our case, the clinical picture was very different due to the onset of the exanthema (although identical in morphology and distribution) at the same time as the onset of fever. Additionally the fever was low fever, there were good general conditions and an absence of lymphadenomegaly and tonsillar exudate. The throat swab for streptococcus was negative and laboratory tests showed a IgM and IgG positivity for Parvovirus B19.

The etiological agent of the fifth disease is Parvovirus B 19, a DNA virus which target is the nucleated cells of the erythroid series of the hematopoietic marrow. This binds through the P receptor and has a marked affinity for the hepatocytes [2].

The Fifth disease is transmitted by airways and has a biphasic clinical course. After a week of incubation there is the viraemic phase which produces an arrest of hematopoiesis and is expressed with flu-like symptoms. After a free interval of seven to eight days the rash appears with a resumption of fever [3].

Typically, the exanthema involves the face with an intense red colour. It is mainly on the cheeks with a "slapped cheek"appearance and spares the perioral area that remains pale (Fig. 8.5).

Subsequently, the erythema spreads to the trunk and limbs. Maculo-papular lesions are often itchy and tend to merge into larger patches (megalo-erythema), pale in the center creating a reticulated or cockade appearance (Fig. 8.6).



Fig. 8.5 Typical "slapped cheek" mark

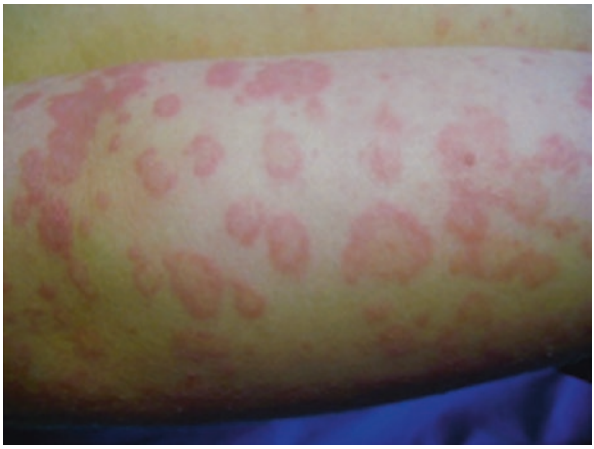


Fig. 8.6 Annular, reticulated and cockade appearance of the erythema

Key Points

- Fifth disease is a viral illness that causes a bright red rash on the cheeks. The rash can then spread to the body, arms, and legs.
- Sistemic symptoms can include sore throat and low fever.
- Slapped cheek and yellow hand sign are more suggestive for the diagnosis

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Chapter 9
A Young Child with Vesiculopustular Eruptions and Mucosal Erosion

Ru-Hong Cheng, Hong Yu, Zhi-Rong Yao, and Ming Li

A 3-year-old boy presented to the outpatient clinic with his mother complaining of vesiculopustular eruptions over scalp, bilateral retro-auricular regions, the trunk and lower extremities, as well as mucosal erosion for 4 months. The child initially had red patches over of bilateral retroauricular skin, mucosa of glans and inner prepuce plate, accompanied by conjunctival hyperemia. Afterward, blisters of uniform size occurred on retroauricular skin, transforming into pustules with a notch sign. Small pustules gathered and fused into erosive and exudation surface (Fig. 9.1). Similar eruptions shortly spread to the scalp, trunk (Fig. 9.2), and lower extremities. A string of pustules arranged along lower eyelid margin (Fig. 9.3). Meanwhile, mucosa erythema of glans and inner prepuce plate changed into erosion and exudation. Oral and throat mucosa was also affected severely (Fig. 9.4). During the course, the child didn't have a fever or itch. The acupuncture reaction was negative. Laboratory examination showed normal white blood cell count (7.92×10^9/L), decreased neutrophil ratio (44.7%) and elevated percentage of eosinophils (10.9%) in peripheral blood. The C-reactive protein was elevated (10 mg/L). Serum albumin was decreased at 30.5 g/L.

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Fig. 9.1 Small pustules gathered and fused into erosive and exudation surface on retroauricular skin



Fig. 9.2 A sunflower-like pustule on the back of the child



Fig. 9.3 A string of pustules arranged along the right lower eyelid margin



Fig. 9.4 Mucosa erythema of glans and inner prepuce plate changed into erosion and exudation. Oral and throat mucosa was also severely affected

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

1. Behcet's disease
2. Linear IgA bullous dermatosis
3. Pemphigoid in children
4. Pemphigus
5. Dermatitis herpetiformis

Histopathological examination of a fresh pustule on the skin showed spinous intercellular edema and an intraepidermal blister. Inside the blister, amount of neutrophils and eosinophils gathered with a few Tzanck cells. Lymphocytes, histiocytes, neutrophils and eosinophils infiltrated around the superficial dermal vessels and hair follicles (Fig. 9.5). Direct immunofluorescence showed weak deposition of IgA in the intercellular space between keratinocytes in the middle and lower layers of the epidermis (Fig. 9.6). Other proteins such as IgG, C3, C4, F, C1q, and IgM were all negative. After 3-week-use of oral sulfasalazine, the patient's symptoms improved significantly (Fig. 9.7).

Diagnosis

IgA Pemphigus (Intra-Epidermal Neutrophilic type).

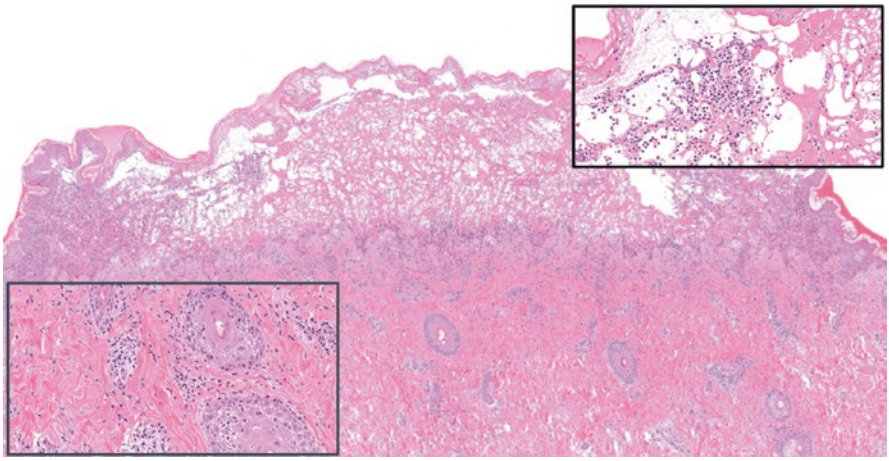


Fig. 9.5 HE staining showed an intraepidermal blister(×200) with amount of neutrophils and eosinophils inside it (×400). Around the superficial dermal vessels and hair follicles, there were lymphocytes, histiocytes, neutrophils and eosinophils infiltrated

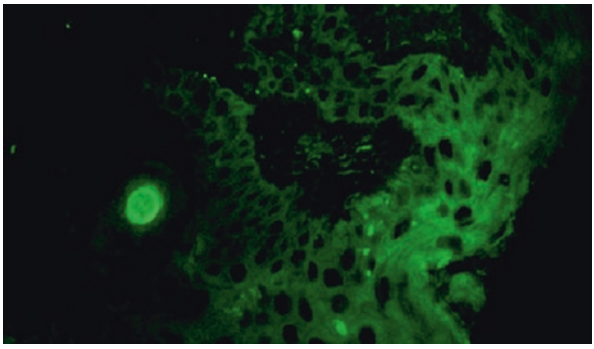


Fig. 9.6 Direct immunofluorescence showed weak deposition of IgA in the intercellular space between keratinocytes in the epidermis (×400)



Fig. 9.7 Both the mucosa and skin lesions improved significantly

Discussion

This case is a male child. The basic lesions are characterized by blisters, pustules and mucosal erosion. On the diagnosis, the first impression is apt to be Behcet's disease according to the affected sites, including conjunctiva, oral and genital mucosa, and skin. However, the ocular manifestation of the children was neither uveitis nor retinal vasculitis. An erosive surface with exudation of the oral and genital mucosa rather than ulcer, which is often seen in Behcet's disease, was found in the child. The main manifestations of skin lesions for the child were blisters and pustules, with a sunflower-like appearance due to the existence of umbilical depression. However, pustules in Behcet's disease were non-follicular sterile pustules, without umbilical depression. No other lesions such as erythema nodosum, acne-like or folliculitis-like papules were presented. In addition, the child's acupuncture reaction is negative. Therefore, the diagnosis of Behcet's disease is not considered. Besides, the diagnosis should be mainly considered in the scope of children's Blister disease. According to this case, the possibility of autoimmune bullous disease is more likely than Blister diseases caused by infection, allergy and genetic factors. Among autoimmune bullous disease in children, linear IgA bullous disease is relatively more common, followed by pemphigoid, pemphigus, and herpetic dermatitis is rarer. Finally, histopathology and direct immunofluorescence revealed that the child is suffered from IgA pemphigus. Oral use of sulfasalazine is demonstrated to be effective.

IgA pemphigus is rare in both children and adults. A systematic review on IgA pemphigus was published in the J AM ACAD DERMATOL of 2020. A total of 137 patients from 26 different countries were enrolled, about 22% of whom were from the Japanese population. The proportion of female patients is slightly higher, the average age of patients is 51 years old, with the age span from 1 month to 90 years old. In our case, he is a child. In literature, two major subtypes of IgA pemphigus are intraepidermal neutrophilic dermatosis (IEN) and subcorneal pustular dermatosis (SPD). The definitive diagnosis of IgA pemphigus is made via immunofluorescence. Almost all patients showed IgA deposits or circulating anti-IgA antibodies. Typical cutaneous lesions were vesiculopustular eruptions. The trunk and limbs were more often to be affected, followed by intertriginous sites. The head and neck, as well as mucosa, is even less frequently to be involved. In histopathology, the cells in infiltrate can be dominated by neutrophils or mixed with eosinophils in most cases. Indirect immunofluorescence, the concurrent deposition of IgA and IgG was observed in 10.6% of patients, and the deposition of IgA, IgG, and C3 was observed in 9.8% of patients. Less frequently, a combined deposition of IgA along with C3 and IgM was observed in 3.0% and 0.8% of patients [1, 2]. It is noted that among patients with IgA pemphigus, remarkable comorbidities exist including hematologic malignancies or lymphoproliferative disorders, solid malignancies, autoimmune disease, which need to be investigated closely [3, 4].

Key Points

- IgA pemphigus is a rare autoimmune bullous skin disease, which can also occur in children.
- Typical cutaneous lesions of IgA Pemphigus should be vesicles and sunflower-like pustules.
- Anatomic distribution of lesions in IgA Pemphigus is not only confined to the intertriginous site, but more often over the trunk, extremities, head and neck, with mucosal evolvement.
- In the majority of cases, predominant cells in infiltrate of IgA Pemphigus can be neutrophils, or mixed neutrophils and eosinophils.

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Chapter 10
A Young Girl with an Erythematous Lesion on the Lower Eyelid Region

Fabio Arcangeli, Elisa Sama, and Giuseppe Ruggiero

We present the case of a three-year-old girl, referred to our service for a single erythematous lesion on the lower left eyelid region, present for about eight months.

The lesion was asymptomatic and appeared the previous month of June. After about three weeks it spontaneously resolved (only an emollient cream was applied), then relapsed to a bigger size after about two months. At the time of our visit it persisted with the same apperance for about 5 months.

In recent months, repeated local cortisone and systemic antibiotic therapies had been performed with slight and temporary improvement.

On physical examination, the lesion appeared as a triangular-shaped patch of a purplish-red colour (Fig. 10.1) partially attenuated by finger pressure. Palpation revealed a discrete infiltration into the subcutis.

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

1. Previous impetigo
2. Cutaneous leishmaniasis
3. Lupus tumidus
4. Chalazion

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Fig. 10.1 The erythematous purplish-red lesion on the lower left eyelid

Diagnosis

Chalazion.

Discussion

Some diagnostic hypotheses could be excluded on the basis of clinical presentation and course. A previous pyoderma should have reported a history of crusty or exudative manifestations and could not be regressed with a persistent outcome for so long. A skin lesion due to leishmania could not have improved until it disappeared spontaneously and then reappeared after 2 months in the same location. A lupus tumidus could not have evolved recurrently in the absence of therapy, not even improving in the summer.

Due to the lack of a certain clinical diagnosis, a punch biopsy was taken for an histological examination (Fig. 10.2).

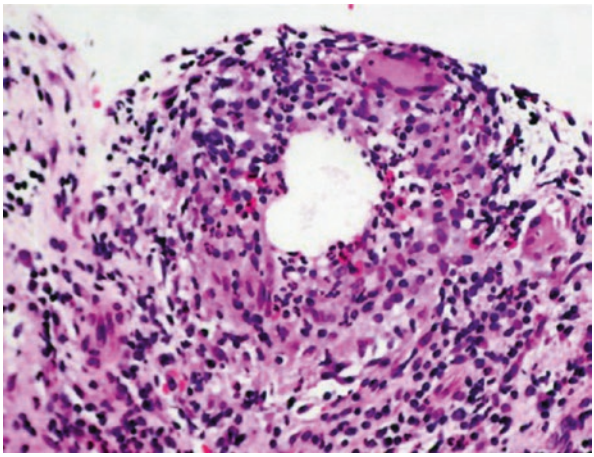


Fig. 10.2 Histological examination revealed a large area of granulomatous inflammation

The Histological examination showed a large area of chronic granulomatous inflammation with plurinuclear giant cells in the context of which there were adipose globules. The immunohistochemical evaluation excluded an epithelial origin of the lesion, resulting negative for CK pool. The PAS, Ziehl Nielsen and Giemsa were negative and ruled out a possible bacterial or protozoal nature of the lesion. The diagnosis was chalazion.

Our patient recovered spontaneously after about two months of local and systemic anti-inflammatory therapy.

The chalazion is a granulomatous inflammation of a gland producing oil that lubrificates the eye's surface, whose excretory duct is blocked by an excessive amount of sebum.

Chalazia can be categorized as either superficial or deep, depending on the glands that are blocked. Inflammation of a tarsal meibomian gland leads to a deep chalazion, whereas inflammation of a Zeis gland leads to a superficial chalazion [1].

Chalazion is different from hordeolum (stye) which indicates an infected oil gland or hair follicle and is usually very painful and may cause the eye to feel sore and scratchy [1]. Chalazion is not usually painful, larger than hordeolum and the prognosis is generally favorable, with spontaneous recovery in a few weeks.

In the event of non-regression the therapy involves [2]:

1. applying a warm compress to help the ducts open and drain more effectively
2. gently massaging the lesion for several minutes each day to help the oil ducts drain
3. using anti-inflammatory eye drops or ointment to reduce discomfort and speed healing
4. a course of oral antibiotics if there are signs of a bacterial infection
5. a steroid injection to reduce swelling
6. surgical removal

The frequent recurrence of chalazion, as well as of blepharoconjunctivitis, can represent a form of ocular rosacea, especially in the pediatric age. Ocular involvement can precede the skin manifestations, arise later or simultaneously [3].

Key Points

- Chalazion can involve not only the eyelid rim but rarely also the lower eyelid region
- Recurrent chalazia may be a sign of rosacea
- Chalazion itself is an inflammatory not infectious process
- The majority of chalazia respond very well to conservative management
- Antibiotics are indicated only when there is evidence of an associated infection

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Chapter 11
Chronic Bullous Disease of Childhood with Hypertrophic Scars Complications

Luh Made Mas Rusyati, I. G. N. Darmaputra, and Prima Sanjiwani Saraswati Sudarsa

A 3-year-old Balinese girl is referred to the Dermatology and Venereology clinic at Sanglah Hospital Denpasar with hypertrophic scars accompanied by wounds in both legs, left leg, left hand and left elbow since 5 months ago. The wound on her leg initiated with easily broken bubbles. It burst then started to widen and leave scar tissue. She consulted the Surgery Department in Singaraja and received surgery to remove the bubbles on her leg. Later, the patient developed extensive scarring that made her have difficulty to walk and straighten her left leg. Before the onset of this illness, the patient often suffered from coughs that had flared up for more than 1 month earlier.

On physical examination of the thigh to the left lower leg and right thigh, a geographic hypertrophic plaque was seen, sized 5 × 10 cm to 15 × 60 cm, well-defined, with multiple superficial ulcers and a round shape sized 1–2 cm on the edge and blackish crust on it (Fig. 11.1). There were multiple hyperpigmented plaques with round shapes sized 4–5 cm in diameter on the shoulder, upper arm and left elbow (Fig. 11.2).

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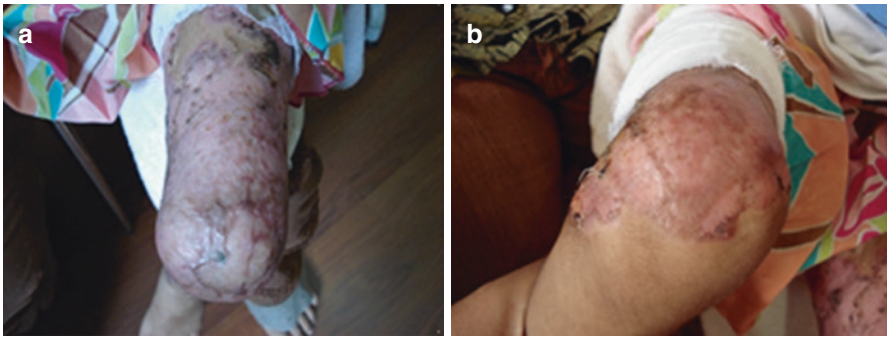


Fig. 11.1 (**a**, **b**) Lesions on the thigh to the left lower leg and right thigh



Fig. 11.2 (**a–c**) Lesions on the shoulder, upper arm and left elbow

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

1. Chronic Bullous Dermatosis of Childhood (CBDC)
2. Epidermolysis bullosa acquisita
3. Intercellular IgA Dermatosis

No bacteria found in gram examination. Skin biopsy results in the form of the epidermis with acanthosis and spongiosis. Among them, there is a distribution of neutrophil cells, some of which appear to be clustered to the epithelial surface. In

the dermis, there is a dense infiltrate of neutrophil inflammatory cells, a lot of nuclear debris, by forming clefts in the papillae of the dermis filled with densely packed neutrophils. There is also a slight lymphocyte infiltrating the vessel wall. The morphology is suitable for Chronic Bullous Dermatosis of Childhood. The results of the Mantoux test showed negative induration and negative hyperemia. Complete blood count results showed leukocytosis (40), neutrophilia (34.3), and ESR 10/55.

The final diagnosis for this patient is chronic bullous dermatosis of childhood.

The treatment consists of Methylprednisolone 3 × 4 mg (7 days), Cotrimoxazole 2 × 480 mg (7 days), Mebhydrolin Napadisylate 2 × ½ mg (7 days), Hydrocortisone 2.5% combine with Chloramphenicol 2% cream on erosion.

During the observation for 1 week, new blisters still appeared so that dapsone started with a dose of 2 × 50 mg/day. The response was great and no new bullae were found. In the last laboratory examination, there were no abnormalities in the blood components.

Discussion

Chronic Bullous Disease of Childhood (CBDC) is a rare blistering disease that occurs primarily in children under 5 years of age and is characterized by a homogeneous linear deposit of IgA along the basement membrane [1]. This disease is said to be non-familial and more common in girls. The clinical manifestation is the presence of clustered papules, vesicles, and bullae with symmetrical distribution on the extensor surfaces, including the elbows, knees, and buttocks. The lesions are often pruritic and cause crusted papules. In some cases, it is sometimes preceded by an upper respiratory tract infection [2].

The histopathological features consist of subepidermal bullae with a collection of neutrophils along the basement membrane and often accumulates in the dermal papillae. There is also a lymphocyte infiltrate around the blood vessels. We can also find blisters in the lamina lucida and on the sub lamina lucida using electron examination to the blisters of CBDC patients. Meanwhile, laboratory examination with direct immunofluorescence will show homogeneous IgA deposition formed along the dermal-epidermal junction [3]. Therefore, the histopathological features of the case further support the diagnosis of CBDC. Direct immunofluorescence examination and electron microscopy were expected to reveal linear IgA along the basement membrane yet could not be performed due to its unavailability [4].

CBDC patients respond very well to dapsone or sulfapyridine. The response usually occurs within 24–48 h. In most patients, the disease can be controlled with dapsone or sulfapyridine alone, but sometimes it requires treatment with low doses of prednisone to suppress the onset of new bullae [1]. Dapsone can start at an initial dose of less than 0.5 mg/kg body weight per day, and if it does not improve, it can increase to 1 mg/kgBW per day [1, 5, 6]. In this case, the patient is initially given cotrimoxazole (sulfonamide) at a dose of 2 × 480 mg/day. During the observation

for 1 week, new blisters still appeared so then we replaced the treatment and started dapsone with a dose of 2 × 50 mg/day [6]. Subsequently, no new bullae observed was found and no abnormalities in the blood components.

CBDC is a self-limited disease and can undergo spontaneous remission within 2 years of the onset. However, the prognosis of this case was worse since the hypertrophic scar occurred and causing a contracture [1].

Key Points

- Chronic Bullous Disease of Childhood (CBDC) is a rare blistering disease that occurs primarily in children under 5 years of age and is characterized by a homogeneous linear deposit of IgA along the basement membrane
- Clinical manifestation is the presence of clustered papules, vesicles, and bullae with symmetrical distribution
- A self-limited disease and can undergo spontaneous remission within 2 years of the onset

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Chapter 12
Chronic Cutaneous Lesions of Unknown Origin

Filiz Topaloğlu Demir, Nazlı Caf, Zafer Türkoğlu, Ayşegül Ak, and Zekayi Kutlubay

Case

A 5-year-old Syrian girl was admitted to our outpatient clinic with a non-healing wound on her right eyebrow and eye contour. There was no significance in the patient's medical history. It was stated by her family that her complaints were present approximately for 12 months. It has also been noted in her story that the wound started as a small pimple and slowly grew and crusted over. She was not feeling any pain, itch, or discomfort except for deterioration in her physical appearance. The patient was applied to different medical centers for the same complaint several times and oral amoxicillin clavulanic acid, topical antibacterial, and cicatrization creams were given for the wound. No response was observed to the given medications and the wound continued to grow despite the treatments. The dermatological examination of the patient revealed 3 × 2.5 cm sized, unpainful, red-colored plaque with a tightly attached adherent crust to the base on her right medial eyebrow extending to the right upper eyelid in addition to the erythema and edema (Fig. 12.1). Besides, nail-like protrusions were observed on the lower surface of the removed crust. In the dermoscopic examination generalized erythema and hyperkeratosis with yellow tears were present (Fig. 12.2). Submental, submandibular, cervical,

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Fig. 12.1 Erythematous infiltrated lesion with a yellow crust on the right eyebrow area and orbita

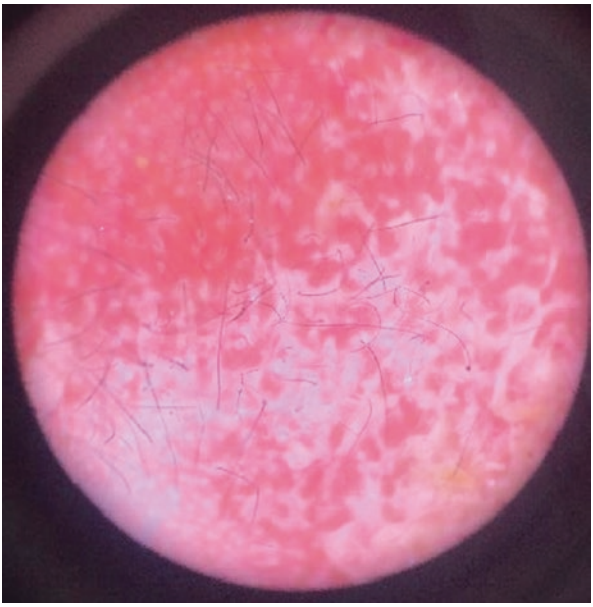


Fig. 12.2 In the dermoscopic examination generalized erythema and hyperkeratosis with yellow tears were present

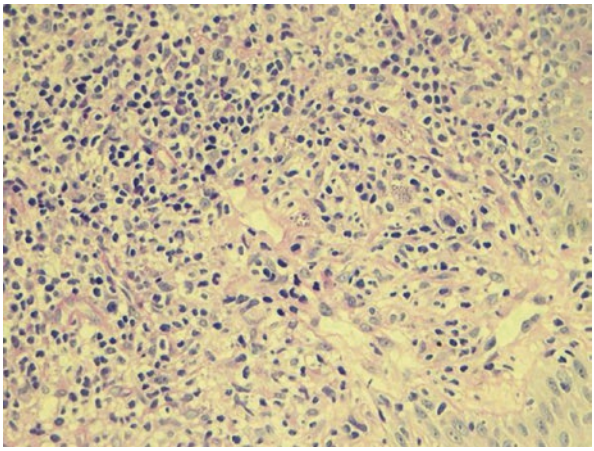


Fig. 12.3 Numerous amastigotes in histiocyte cytoplasm and tissue (PAS; ×400)

preauricular, postauricular, and occipital lymph nodes were examined with palpation and no regional lymphadenopathy was present. Hepatosplenomegaly was not detected in her physical examination.

Complete blood count and biochemical tests were completely normal. Viral Hepatitis and HIV serology were negative.

May-Grünwald Giemsa stained smear of the lesion was performed due to the suspected diagnosis and investigated directly with a microscope.

Amastigotes were not seen in direct microscopic examination. 4 mm Punch biopsy was taken from the patient for histopathological investigation and Polymerase Chain Reaction (PCR) assay. Amastigotes were detected in the histopathological examination and the agent was determined with PCR (Fig. 12.3).

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

- Sporotrichosis
- Exaggerated arthropod bite
- Cutaneous Leishmaniasis
- *Mycobacterium marinum* infection
- Foreign body reaction

Diagnosis

The patient was diagnosed with cutaneous leishmaniasis according to such clinical and histopathological findings. The agent was specified with PCR as *L. tropica*. Numerous amastigotes (Leishmania-Donovan bodies) were observed in histiocyte cytoplasm and tissue (PAS; ×400).

Discussion

Leishmaniasis is a notable health problem in many countries around the world as well as in Turkey. The disease is vectorborne and carried by female sandflies and seen mostly in childhood but it may affect individuals in every age group. Depending on the parasite type and immune response of the host, three clinical forms may develop: Cutaneous, mucocutaneous, and visceral type leishmaniasis [1]. The clinical severity and response to treatment depend on the type of parasite, so genuine treatments should be planned according to the type of parasite in the current treatment approach [2].

With the bite of the vector, some people may remain asymptomatic. Typical lesion formation begins with the occurrence of erythema and papule following the bite of the causative sandfly. Then a nodular structure is formed between 2 weeks and 6 months, and it becomes ulcerated. It should be kept in mind that lymphadenomegaly may accompany the skin lesions [3].

Cutaneous leishmaniasis is not a life-threatening disease and generally presented with an ulcer that self-heals within 3–18 months. However, it may cause morbidity by scarring, deformation, and stigmatization by the community. Cutaneous Lesions caused by *L. tropica* and *L. major* are generally self-limiting but tend to leave everlasting scars [4].

The most important step in the diagnosis of CL is the "clinical suspect". There is no gold-standard parasitologic diagnostic test for CL up to date. Dermatoscopy, histopathology, direct smear from the wound, Polymerase chain reaction (PCR), culture (NNN culture medium) may be used for the diagnosis [4].

Systemic or local treatment options are available for CL. The CL may self-heal but the treatment may provide more cosmetic results such as decreasing the scar formation. The World Health Organization(WHO) recommends the use of pentavalent antimonial drugs in the treatment of cutaneous leishmaniasis (i.e., sodium stibogluconate or meglumine antimoniate).

Musculoskeletal pains, renal failure, hepatotoxicity, and cardiotoxicity may occur during the use of pentavalent antimonial drugs so patients should be monitored. Parenteral Amphotericin B and Pentamidine isetionate are the other systemic treatment options. Besides, oral miltefosine, allopurinol, paromomycin, fluconazole, itraconazole, ketoconazole, and zinc may be used. Thermotherapy, topical paromomycin, cryotherapy, intralesional Interferon γ, topical or intralesional zinc sulfate solution are the local treatment options [5].

Intramuscular meglumine antimoniate was applied for 20 days with a 16 mg/kg/day dose to our patient and no side effects were detected during the treatment. One month after meglumine antimoniate treatment, no regression was present in re-examination and the patient was evaluated as unresponsive to meglumine antimoniate; 6 mg/kg/day fluconazole suspension was applied for 6 weeks. A dermatological examination was performed 6 weeks later and no response was observed in the lesion. Intravenous liposomal amphotericin B was applied for 7 days with a dose of



Fig. 12.4 After intravenous liposomal amphotericin B treatment

5 mg/kg/day. The existing lesion was completely regressed except erythema during follow up (Fig. 12.4).

Sporotrichosis is a chronic granulomatous fungal disease caused by *Sporothrix schenckii* and the causative agent is found naturally in the soil. The exact inoculation time of sporotrichosis is unknown but the average is 3 weeks. With the inoculation of the agent, infection occurs primarily in the skin and surrounding lymphatics. A small, indurated, progressive papulonodular structure forms in this area and may ulcerate (sporotrichotic chancre). Systemic symptoms are not observed during this period. The formed lesion may remain solitary or multiple lesions may develop. In the pediatric population, the clinical course is similar but facial involvement is more common (40–97%). Consequently, sporotrichosis is presented in three main clinical types: lymphocutaneous, fixed cutaneous, and multifocal or disseminated cutaneous sporotrichosis. In fixed cutaneous sporotrichosis the lesions are usually asymptomatic: Patients do not complain from pain or itch. Erythematous papules, nodules, plaques, and persistent ulcers may be observed [6].

Arthropod bites may be asymptomatic or manifest with mild, self-limiting itchy pink papules. Rarely, larger and intense reactions due to immunologic reactions may occur after the bite and are described as exaggerated arthropod bites. These lesions are large, erythematous, itchy, and edematous, and sometimes even appear

as hemorrhagic or necrotic wounds. If secondary infection, which is a possible complication that may develop after an arthropod bite, does not develop, the patient may not have pain. Serum IgE levels may be found to be increased in patients with exaggerated arthropod bites. Histopathologically, a large number of eosinophils and rich accumulation of B and T lymphocytes are observed [7].

Mycobacterial infections, whether painful or painless, can be life-threatening. *M. marinum* infections are theoretically divided into four subgroups in order to facilitate and guide treatment options (I–IV). Disease severity and dissemination increase towards type IV. In this case, type I *M. marinum* infection is included in the differential diagnosis and this condition is usually seen in individuals with normal immune systems. These patients have solitary or few (1–3) superficial skin infections and are characterized by crusted or ulcerated nodules or verrucous plaques. The most common skin involvement of the disease is fingers and hands due to contact with infected fish or water. These lesions may occur within weeks to months after the injured skin is contacted with infected fish and it is not transmissible from human to human [8]. The clinical suspect and different approaches for diagnosis are required. Granulomas with multinucleated giant cells, hyperkeratosis with focal parakeratosis, hyperplasia, and liquefaction degeneration of the basal layer may be detected but none of these findings are pathognomic. EZN staining and bacterial culture can be used for definitive diagnosis. Different antibiotic regimens may be used for the treatment and surgery is possible in selected cases [9].

The entrance of foreign bodies into the body may be due to voluntary reasons (i.e. tattoo, cosmetics) or unintentional basis (i.e. accidental skin trauma and foreign bodies entering the body). Regardless of the reason, granuloma formation can be triggered by the entry of foreign bodies into the body and may appear in the clinic in many different ways. Moreover, the development of hypersensitivity reactions can be expected. Papules, erythematous subcutaneous or cutaneous ulcerated nodules, or an exophytic tumor may be observed. Besides, sarcoidal granulomas, granuloma annulare-like reaction, necrobiosis lipoidica, perforating granulomatous dermatitis, vasculitis, a pseudolymphomatous pattern, panniculitis, and a morphea-like reaction are also reported in the literature. Medical and/or interventional treatment modalities may be applied [10].

Key Points

- CL should be considered especially in non-healing wounds in areas not covered by clothing.
- Clinical suspicion, and then direct staining, histopathology and PCR are important in the diagnosis of CL.
- Painless lesions should be warning for cutaneous leishmaniasis
- Unresponsiveness to both systemic and topical antibiotics, cicatrising creams should raise suspicion for CL
- The appearance of crust which is tightly attached to the erythematous base is typical. Nail-like processes (Tin-Tack' sign of Hulusi Behçet) may be observed on the inner surface with the removal of the crust [3].

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Chapter 13
Diffuse Pruritic Lesions in a 3-Years-Old Child

Giuseppe Ruggiero, Cosimo Ruggiero, and Luca Ruggiero

A 3-year-old Caucasian girl presented with intensely pruritic lesions appeared 1 year before. Physical examination revealed erythematous-symmetrical papules, vesicles and excoriations, localized on the lower and upper limbs, trunk and buttocks (Fig. 13.1a–c).

The dermoscopic examination showed small and erythematous vesicles and brown dots (old petechial lesions) (Fig. 13.2).

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

1. Dermatitis Herpetiformis
2. Atopic dermatitis
3. Scabies

Diagnosis

Dermatitis herpetiformis

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Fig. 13.1 Papules and excoriations on the right forearm (**a**), left thigh (**b**) and buttocks (**c**)

Discussion

Dermatitis herpetiformis (DH) is an autoimmune blistering skin disease characterized by granular IgA deposits along the dermal-epidermal junction in patients with celiac disease. The term dermatitis herpetiformis (DH) was first proposed by Louis Duhring (1884) [1], described a chronic disorder characterized by intense pruritus and pleomorphic skin lesions.

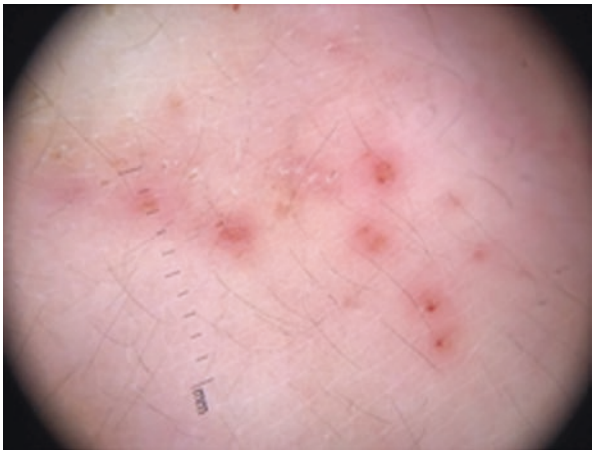


Fig. 13.2 Dermoscopic examination: small vesicles and brown dots

There is growing evidence that dermatitis herpetiformis should be considered the specific phenotypic cutaneous expression of a gluten-sensitive enteropathy indistinguishable from celiac disease. DH usually is diagnosed during adulthood, and the incidence is highest in females and males aged 50–69 years [2, 3]. Diagnostic age of DH has increased and a possible explanation could be changes in dietary habits [2]. Even though childhood diagnosis is rare in northern Europe [2–4], it seems to be more common in Italy and Hungary [5, 6].

It is characterized by diffuse, symmetrical, grouped polymorphic lesions consisting of erythema, papules and herpetiform vesicles or blisters followed by erosions and hyperpigmentation. Itching and burning sensation immediately precede the development of lesions, usually localized on extensor surfaces of the elbows (90%), knees (30%), shoulders, and sacral region.

In the suspicion of herpetiform dermatitis, serological tests evidenced IgA anti-tTG positivity (> 20 times of normal values) and IgA anti-endomysial antibodies (EMA).

Accordingly, small bowel biopsy would be unnecessary in dermatitis herpetiformis patients, and diet adherence would be monitored by serological testing and skin lesions observation [7, 8].

A gluten-free diet is the treatment of choice for patients with celiac disease/dermatitis herpetiformis since both the enteropathy and the cutaneous rash strictly correlated to gluten ingestion [8].

Differential Diagnosis

1. **Atopic dermatitis** is an itchy and inflammatory skin condition. It is characterized by erythema with edema, vesicles, and skin thickening (lichenification) in the chronic stage. It is difficult to define due to its variable morphology and distribution. The diagnosis is based on clinical features and requires generally

evidence of itchy and dry skin. Biopsy is often not necessary, while the dermoscopic examination can make a significant contribution to the diagnosis.

2. **The scabies** usually presents with pruritus in children. The lesions are characterized by small and erythematous papules and often with erosions and excoriantoins due to intense itching. Family members generally have similar symptoms. Signs and symptoms often mimicking other conditions. Dermoscopy has proven to be a non-traumatic method of diagnosis: serpiginous burrows and a small dark structure called triangle or "delta wing jet" are the most common signs of scabies [9, 10].

Key Points

- Clinical history, distribution of lesions and laboratory tests can guide the diagnosis
- Dermoscopic evaluation can exclude scabies for its pathognomonic lesions.

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Chapter 14
Fournier Gangrene in 3 Years Old Patient with B Cell Acute Lymphoblastic Leukemia

Eliza Miranda and Triana Agustin

A 3-year-old female patient was referred to the Department of Dermatovenereology because of a tender ulcer on the perianal area and black eschar for 3 days (Fig. 14.1). She had diarrhea six times per day and fever. She also suffered from B-cell acute lymphoblastic leukemia and started taking vincristine chemotherapy and corticosteroid. Blood culture revealed *Pseudomonas aeruginosa*. Therefore, initial diagnosis of ecthyma gangrenosum and diaper dermatitis were established. Ceftazidime was then given intravenously with gentamicin ointment and acetic acid 5% wet dressing for ecthyma gangrenosum along with zinc ointment for treating diaper dermatitis. Four days after admission, the violaceous-erythematous patch was enlarged. Physical examination on the perineum, gluteus, major labia, medial femoral, and inguinal fold showed tenderness with a necrotic-based ulcer, black eschar surrounded with a violaceous-erythematous patch, and edema (Fig. 14.2).

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

1. Fournier gangrene
2. Necrotizing cellulitis
3. Ecthyma gangrenosum

Laboratory results demonstrated anemia (7.9 g/dL), leucopenia (180/mm^3), thrombocytopenia (5000/mm^3), high C-reactive protein (263.5 mg/L), sodium 136 mmol/L, creatinine 0.4 mg/dL, glucose level 100 mg/dL and laboratory risk for necrotizing fasciitis (LRINEC) score was 6 (moderate risk), which is predictive for Fournier gangrene. Culture from the wound base revealed *Escherichia coli,* and the

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Fig. 14.1 Black eschar ulcer



Fig. 14.2 Violaceous-erythematous patch and black eschar were progressively enlarged in 4 days after initial consultation

antibiotic regimen was changed into Meropenem and Metronidazole. Consultation to the Department of Plastic Surgery was carried out, which proceeds to the explorative debridement. Six days after admission, the necrotic tissue was worsening and the LRINEC score became 9 (Fig. 14.3). An abdominal computed tomography (CT) scan showed that there was ascended infection to the sigmoid. The debridement would be conducted by a collaboration of plastic and pediatric surgeons when platelet >50,000/mm^3 to avoid bleeding. The patient was then transfused gradually with thrombocyte concentrate 4 L, pack red cells 2 L, and fresh frozen plasma 200 mL. Intraoperatively, there was grey fascia in some areas. The colostomy was also performed to reduce possible bacterial contamination in surgical wounds from the feces (Fig. 14.4). Tissue culture revealed *Acinetobacter sp.* that was resistant to all antibiotics except indeterminately resistant to tigecycline. The split-thickness skin graft was then planned to close the defect.



Fig. 14.3 Worsening of the lesion 6 days after initial consultation



Fig. 14.4 Two days after debridement surgery and colostomy

Discussion

Fournier gangrene occurs most frequently in patients aged 50–60 years with significant comorbidities, particularly diabetes mellitus or an underlying immunocompromised status [1]. Fournier gangrene is rarely noted in the pediatric population, particularly before the age of 3 years, which is thought to be secondary to the introduction of infection through underlying diaper dermatitis [2]. This patient suffered from diarrhea that makes the diaper area compromised and vulnerable to infection. However, the patient was in an immunosuppressed condition caused by chemotherapy and corticosteroid for treating B-cell acute lymphoblastic leukemia she suffered from.

Fournier gangrene is confirmed based on history, physical examination, and laboratory examination. The infection progress rapidly and the constellation of signs and symptoms include edema, black eschar, violaceous erythematous patches, and pain involving the genitalia and surrounding skin [1]. In 2004, a laboratory scoring system, LRINEC), was proposed to help detect early cases of necrotizing fasciitis and allow for prompt surgical intervention [3]. Early clinical suspicion for necrotizing fasciitis with a LRINEC score of 6 points or more has a positive predictive value of 92% and a negative predictive value of 96% as this patient had score 6 at initial diagnosis [3]. The gold standard for diagnosis is exploration surgery as we can directly observe necrotic grey, dusky, and edematous fascial plane as seen in this case [1]. To support a diagnosis of necrotizing fasciitis, CT scan or magnetic resonance imaging (MRI) should be conducted as there are thickening and inflammation of fascial planes and fat stranding caused by infection [4]. In our case, we found thickening of wall descending colon, sigmoid, and rectum. The fat stranding was also noted which was probably caused by infection. It is important for patients with extensive rectal involvement that diverting colostomy can help minimize fecal contamination of surgical wounds and assist wound healing following surgical debridement [5].

Early diagnosis, prompt initiation of appropriate antibiotics, hospitalization, and rapid surgical intervention remains the mainstay of therapy for patients with necrotizing fasciitis [1, 6]. Late in the establishing diagnosis and surgical intervention can increase mortality rate due to rapidly progressive infection causing septic shock and or multiorgan failure [1, 4]. In our case, there was a delay in surgical intervention caused by severe thrombocytopenia and some efforts to improve the general condition to avoid intra or postoperative hemorrhage.

Key Points

- Fournier gangrene is a life-threatening infection, rarely noted in the pediatric population, particularly before the age of 3 years.
- The diagnosis should be suspected when there is an immunosuppressed condition and the infection progress rapidly in a few days.
- Clinically, it is characterized by edema, black eschar, violaceous erythematous patch, and pain involving the genitalia and surrounding skin.

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Chapter 15
Itching Eyelids in a Child with Atopic Dermatitis

Giulia Veronesi, Miriam Leuzzi, Alba Guglielmo, Annalucia Virdi, and Iria Neri

A 4-year-old female presented in emergency department with complaints of itchiness and irritation in both eyes of 1 month duration. Medical history included atopic dermatitis and the parents referred that she had been treated for atopic blepharitis with slight response. However, the itchiness continued to exacerbate after treatment with topical glucocorticoids and systemic antihistamines. The child was in good health and the physical examination was otherwise normal. Dermatological clinical examination showed xerosis cutis with the Dennie-Morgan fold under the eye (Fig. 15.1). There was no hyperemia, swelling or redness of the upper eyelid margin. A careful dermoscopic examination revealed the presence of translucent oval structures attached at the base of the eyelashes (Fig. 15.2). General examination did not reveal similar lesions anywhere else on the body.

Based upon history, clinical and dermoscopic appearance, what is your diagnosis?

1. Atopic blepharitis
2. Trichorrhexis nodosa of the eyelashes
3. Allergic contact dermatitis
4. Phthiriasis palpebrarum
5. Bacterial conjunctivitis

Diagnosis: Phthiriasis palpebrarum

Based on the dermoscopic observation of small translucent oval nits coating the eyelashes (Fig. 15.2) a diagnosis of bilateral phthiriasis palpebrarum was made. General examination did not reveal lice anywhere else on the body. There was no history of symptoms suggesting pediculosis or phthiriasis in family members. She

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Fig. 15.1 Clinical examination showed xerosis cutis with the Dennie-Morgan fold under the eye. There was no hyperemia, swelling or redness of the upper eyelid margin



Fig. 15.2 Dermoscopy underlined translucent oval structures attached at the base of the eyelashes

was treated with topical vaseline application over the eyelashes of both the eyes, followed by the mechanical removal of all the parasites. Vaseline is adequate to stifle the parasites, facilitating the removal of the nits by pulling with a fine forceps. Vaseline was continued twice daily at home with no episode of recurrences.

Discussion

Pediculosis can be caused by three different organisms, the head louse, the body louse and the pubic louse [1–5]. Although differing in the anatomical areas involved, they produce equivalent disorders such as itching, burning and skin irritation with erythema, crusts and swelling more or less evident [1–5]. Phthiriasis palpebrarum is uncommon in children and might be confused with other forms of blepharitis or eczema [1, 2, 5]. Moreover, itching of the eyelid margins could be complicated by conjunctivitis [1]. Pthiriasis palpebrarum is generally caused by direct or indirect contact with Pthiriasis pubis and in children is secondary to the contact with an

adult carrier [1–5]. For this reason, phthiriasis in children requires the question of the method of contamination and the suspicion of sexual abuse must be raised [2–5].

The diagnosis is improved by dermoscopic examination with high magnification that can aid in the detection of nits and in monitoring treatment response. There are various options of treatment for phthiriasis palpebrarum including cryotherapy, pilocarpine gel, physostigmine eye ointment, petrolatum ointment, fluorescein drops, yellow mercuric oxide ointment, permethrin cream, malathion shampoo and oral ivermectin [1–5]. However, the simplest technique reported in leterature consist in topical application of vaseline and mechanical removal of the lice and the nits with fine forceps [4]. The vaseline acts as a suffocating agent to kill the lice and newly hatched lice from the nits and could be used for long time without ocular irritation [3].

Key Points

- Phthiriasis palpebrarum is uncommon in children and might be confused with other forms of blepharitis or eczema.
- The diagnosis is improved by dermoscopic examination with the detection of lice and nits
- The first-line treatment consists of topical vaseline and mechanical removal of nits and parasites.

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Chapter 16
Liver Involvement in Langerhans Cell Histiocytosis

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A-3-year-old male patient, was referred to the department of dermatology and venereology because of asymptomatic reddish skin rashes over the scalp, face, trunk and extremities for the past 10 months (Fig. 16.1). He also presented nail destruction and discoloration, hepatomegaly, jaundice for the past 4 months, weight loss, which he was admitted to another general hospital and evaluated for the cause (Fig. 16.2).

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

1. Langerhans Cell Histiocytosis
2. Seborrhoeic dermatitis
3. Generalized eruptive histiocytosis
4. Leukemia cutis

On examination, the patient was malnourished and stunted. The vital signs were normal. He had pallor, jaundice, ascites, ulceration of the oral mucosa and lips, with loss of teeth and hepatomegaly. Cutaneous involvement was found mostly on the scalp, trunk, hands and soles as a brown petechial rash with crusting or purpura. Nail examination revealed nail hemorrhage, discoloration, subungual hyperkeratosis and onycholysis in all fingers and toes.

Laboratory investigations: hemoglobin 5.4 g/dL, leukocytes 21.790/mm^3, albumin 0.92 g/dL, bilirubin total 13.909 mg/dL, direct 11.764 mg/dL, creatinine

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Fig. 16.1 (**a**, **b**) Asymptomatic reddish skin rashes over scalp, face, trunk and extremities

0.17 mg/dL. Magnetic resonance cholangiopancreatography (MRCP) showed multiple cyst communication with bile ducts. Skin biopsy showed infiltration of Langerhans's cells in the dermis which staining were positive for S100 and CD1a.

Diagnosis: Langerhans cell histiocytosis

Treatment: Topical moisturizer was given and showed significant improvement within 1 month. Orally, the patient was treated with prednisone, spironolactone and furosemide. He also received a packed red cell (PRC) transfusion, intravenous albumin and one cycle of chemotherapy with methotrexate and leucovorin. Continuing chemotherapy was planned but the general condition deteriorated. The patient was lost to follow-up within 3.5 month with further worsening of symptoms and he died due to acute bronchopneumonia.

Discussion

Langerhans cell histiocytosis (LCH), defined as a rare, heterogeneous neoplasm of dendritic cells [1]. LCH can occur at any age, however, peak occurrence occurs between 1 and 4 years of age. The nature of the disease still raises doubts, focused on functional dysregulation versus malignant transformation of epidermal dendritic cells [2]. The disease has different clinical courses from a mild form, which limited to one organ to a severe form with involvement of many organs and systems that progresses very quickly may lead to death [2, 3]. The organs more frequently involved are the skeleton (80% of cases), the skin (30–60%), and the pituitary (25%). Other organ involved are liver, spleen, the hematopoietic system and the



Fig. 16.2 (**a–d**) Cutaneous involvement on hands and soles, brown petechial rash with crusting or purpura. Nail hemorrhage, discoloration, subungual hyperkeratosis and onycholysis in all fingers and toes

lungs (15% each) [3]. In the youngest children, the disease is often a multisystem type (MS) disease with fever and symptoms of failure in various organs. The incidence of cutaneous involvement is significantly higher in children with MS disease. The eruption may be extensive and involve the scalp, face, trunk, buttock and intertriginous area, varying from crusted or scaly nodules and papules, purpuric macules

and petechiae [2]. Involvement of the oral mucosa include gingival swelling, or ulceration of the palatum, buccal mucosa or lips and loss of teeth [2, 4, 5]. The nail affliction may manifest as onycholysis, paronychia, hyperkeratosis, subungual thickening, deformity and loss of nail plate, which mostly represent in patients with multisystem disease with involvement of high risk organs [3]. A typical symptom in LCH causes hepatomegaly, tumor like nodular lesions and cystic lesions, it may be accompanied by organ dysfunction, an increase of transaminases, hypoalbuminemia, secondary oedema or ascites and jaundice [2, 3, 5].

In this case, the diagnosis of LCH was established by histological examination of the skin biopsy which showed dermal infiltration of histiocytes, then confirmed by positive S100 and CD1a and negative CD 68 immunostaining [6]. Liver involvement was diagnosed with the significant findings on MRCP [3]. Liver involvement drastically changes patients prognosis and treatment [2]. Treatment options vary depending on the extent of the disease and the severity at onset [3]. The prognosis for LCH varies depending on the form of the disease (MS-LCH), location, and response to chemotherapy. The prognosis worsen significantly with the involvement of "risk organs" (bone marrow, liver, spleen and the lungs) and the G.I Tract [2, 5], because high risk disease is less responsive to therapy and requires more aggressive treatment [5]. All liver LCH patients are recommended to received systemic chemotherapy early once the diagnosis was made [7]. In this case, the patient had received one cycle of chemotherapy with methotrexate and leucovorin. Continuing chemotherapy was planned but the general condition of the patient deteriorated. The patient was lost to follow-up with further worsening of symptoms and he died due to acute bronchopneumonia.

Key Points

- Langerhans cell histiocytosis (LCH), defined as a rare, heterogeneous neoplasm of dendritic cells.
- Liver involvement usually presents as a part of the disseminated process of disease which is difficult to treat.
- Early diagnosis is important for better response to therapy.

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Chapter 17
Localized Scaly Hair Loss

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

Case Presentation

Male patient 4 years old presented in the dermatologic clinic with scaly localized area of hair loss on the occipital region of the scalp (Fig. 17.1) with no family history of the same lesion. He gave a history of living in a rural area with frequent exposure to pets and animals. Four weeks prior, the lesions appeared initially as pruritic follicular pustules, which increased gradually in size and localized hair loss. Before appearance of these lesions, he had experienced scalp itch. He initially treated with combination of antibiotic, steroid and antifungal with little response.

Physical examination revealed a 3 × 4 cm ulcer with several peripheral follicular pustules on the occipital area of the scalp with slight erythema. Left cervical lymphadenopathy was present. The hairs around the lesion were plucked easily.

Dermoscopic examination revealed absence of hair follicles in some areas, follicular pustules (yellowish) with comma and morse code hairs. Also, there appeared yellowish amphorous (waxy) material around hair follicles (Fig. 17.2).

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

1. Lichen planopilaris.
2. Alopecia areata.
3. Tinea capitis.
4. Trichotillomania.

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Fig. 17.1 Localized area of hair loss on the occipital region of the scalp in male patient 4 years old

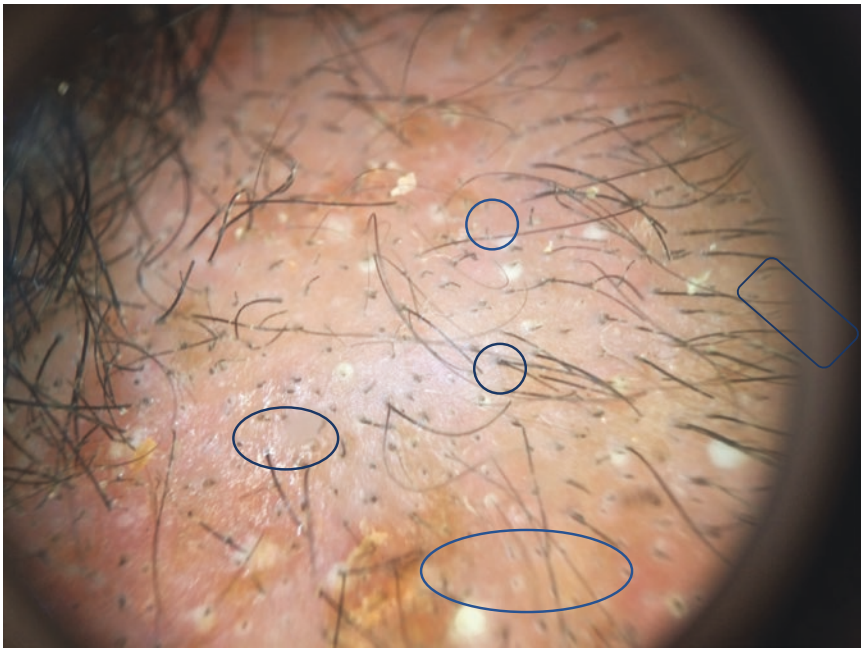


Fig. 17.2 Dermoscopic examination of localized area of hair loss revealed absence of hair follicles in some areas, follicular pustules (circles) with comma and morse code hairs (rectangle). Also, there is yellowish amphorous (waxy) material around hair follicles (oval) (Dermoscopy 3gen DermLite 4, magnification ×10)

Diagnosis

Tinea capitis (favus type)

Discussion

Tinea capitis (TC) is a widespread scalp infection caused by dermatophytes, occurring predominantly in children. The distribution of causative species of dermatophytes varies greatly in different geographic regions [1, 2]. Kerion is an inflammatory type of TC and occurs commonly in rural areas with poor hygienic conditions and prepubertal children are more easily affected. The incidence of kerion is increasing in urban areas and pets are likely to be the most important sources of infection [3].

At the beginning of the nineteenth century, Alibert [4] used the term 'favus' to describe the honey-like exudate in some scalp infections. Favus is a chronic inflammatory dermatophyte infection of the scalp [5]. The classic favus lesion is the 'scutulum', a concave, cup-shaped yellow crust on the scalp and glabrous skin that is associated with severe alopecia. These keratotic crusts contain fungal hyphae and can be highly infectious [6].

Dermoscopy is an aided non-invasive tool for diagnosis of tinea capitis and its subtypes [7] dermoscopy is an important in differential diagnosis between TC and alopecia areata [8]. "Comma" and "corkscrew" hairs are the classic findings of TC under dermoscopy [9]. Comma-shaped hairs are characterized by a sharp slanting end, homogeneous thickness, and pigmentation of hair shaft, which represent an intermediate stage before formation of dystrophic hairs. Mycological examinations are considered to be the gold standard for diagnostic method of TC [3]. However, fungal culture is time-consuming and usually requires 2 weeks for results [10].

Key Points

- Tinea capitis (TC) is a widespread scalp infection caused by dermatophytes.
- Tinea capitis (TC) occurs commonly in rural areas with poor hygienic conditions and prepubertal children are more easily affected.
- Dermoscopy is very helpful tool in diagnosis of tinea capitis and its subtypes.

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Chapter 18
Papular Lesions Arranged in Annular Configuration in Children

Nooshin Bagherani and Bruce R. Smoller

Two children were referred with relatively similar lesions with clearly defined borders. The first was a 2.5-year-old female baby with the appearance of an annular lesion superimposed with papules on the back of the left hand for 2 months. The second case was a 5-year-old male child with papular and tiny nodular lesions in an annular configuration on the right wrist for 1.5 months. The children were healthy without any other significant medical histories (Figs. 18.1 and 18.2).

Based upon the clinical feature, what is your diagnosis?

- Insect bite
- Dermatophytosis
- Granuloma annulare
- Lichen nitidus
- Child abuse
- Nummular eczema
- Hypertrophic lichen planus

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Fig. 18.1 A 2.5-year-old baby with appearance of an annular lesion superimposed with papules



Fig. 18.2 A 5-year-old child with papular and tiny nodular lesions arranged in annular shape

Diagnosis: Granuloma annulare

On pathological examination, in both of the cases there were foci of degenerating collagen bundles in the upper and middle dermis along with infiltrates of histiocytes, lymphocytes and giant cells palisading in the periphery (Fig. 18.3).

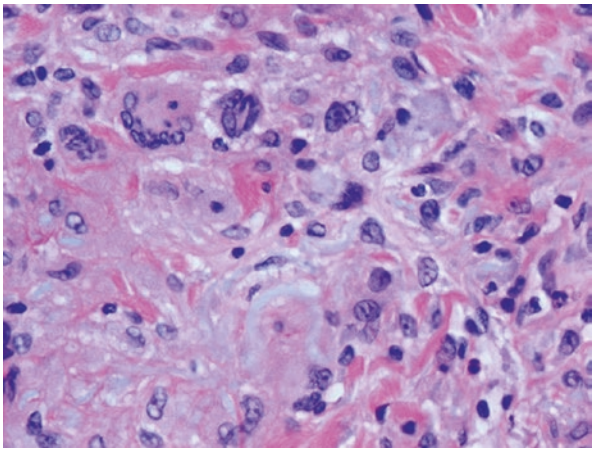


Fig. 18.3 Pathological view of the lesion from the first patient

Discussion

Granuloma annulare is a benign granulomatous condition that presents as asymptomatic cutaneous polymorphic lesions affecting children and adults. Its etiology is unknown [1–3]. Some patients complains of pruritus and tenderness [1]. Its clinical variants include localized, generalized or disseminated, subcutaneous or deep, perforating [1, 2], giant, and targetoid. The lesions are characterized by closely-set, flesh-colored, skin-colored, violaceous or erythematous-brown papules or tiny nodules measuring 1–5 cm in diameter arranged in a ring or annular configuration [3].

The localized variant comprises 75% of all granuloma annulare cases, most commonly affecting patients under the age of 30 with a female preponderance (female to male ratio 2:1). This variant mainly involves the dorsal aspects of hands and feet and resolves spontaneously within 2 years. Its pathological findings include foci of collagen bundles degeneration in the upper to middle dermis along with infiltrates of histiocytes, lymphocytes and occasional giant cells palisading in the periphery [1].

The generalized variant comprises 15% of cases of granuloma annulare and is characterized by appearance of 10 or more lesions or widespread annular plaques. Its age distribution follows a bimodal pattern affecting patients over 40 years of age or under 10 years [1]. The lesions involve the trunk, extremities, and neck, in a symmetrical pattern [3]. On pathologic examination, collagen degeneration is less apparent [1].

In the subcutaneous variant, deep dermal or subcutaneous nodules are detectable [1]. Its spontaneous regression has been reported over months to years. In its pathological aspects, the findings can include central necrobiosis in the deep dermis and subcutaneous fat along with increased deposition of mucin surrounded by palisading histiocytes, lymphocytes, eosinophils, and fibroblasts [2].

The perforating variant is the rarest type of granuloma annulare which presents as papules with a central crust or scale that can occasionally be umbilicated [1].

The lesions frequently regress spontaneously; hence, they can be managed conservatively. Topical and intralesional corticosteroids, antibiotics, [1], topical imiquimod, topical calcineurin inhibitors, pulsed dye laser [3], cryotherapy [1–3], and surgical removal [2, 3] can be used in their treatments.

Key Points

- Granuloma annulare is benign granulomatous condition with unknown etiology, presented as skin polymorphic lesions affecting children and adults.
- In pathological view, necrobiosis along with palisading peripheral infiltrates of histiocytes, lymphocytes and giant cells are seen in granuloma annulare.
- Spontaneous regression is expected in granuloma annulare.

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Chapter 19
Pustular Plaque on a Girl's Scalp

Rossella Lacava, Miriam Leuzzi, Marco Adriano Chessa, Valeria Evangelista, and Iria Neri

A 4-year-old girl presented with an inflammatory alopecic plaque located on the vertex of the scalp (Fig. 19.1). The lesion had appeared spontaneously 2 months earlier and the patient reported mild itching as the only associated symptom. Antibiotic therapy with oral amoxicillin-clavulanate was ineffective, so it was stopped and skin swabs for bacteria and fungi were performed.

Based upon history and clinical appearance what is your diagnosis?

1. Kerion
2. Dissecting cellulitis of the scalp
3. Erosive pustular dermatosis of the scalp
4. Pyoderma gangrenosum
5. Pemphigus foliaceus

Diagnosis: Erosive pustular dermatosis of the scalp (EPDS).

We observed an alopecic inflammatory erythematous plaque of the scalp surmounted by pustules and yellow-brown crusts.

Trichoscopy showed yellow brown crusts, yellow glubules corresponding to pustules, absence of follicular ostia on the pink-red background (Fig. 19.2).

Skin swabs for bacteria and fungi: negative.

Based on the clinical appearance, dermoscopic findings and exclusion of infections, a diagnosis of EPDS was made. A treatment with clobetasol propionate 0.05%

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Fig. 19.1 Alopecic inflammatory plaque of the scalp

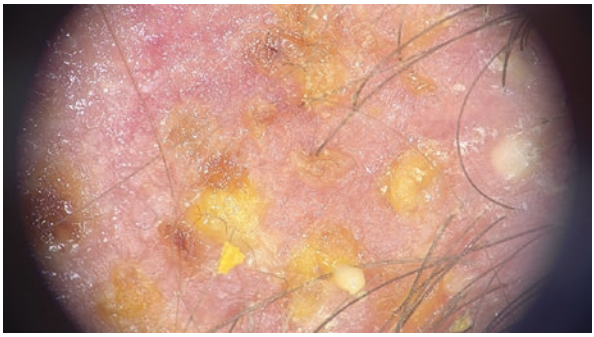


Fig. 19.2 Trichoscopy showed yellow brown crusts, yellow glubules corresponding to pustules, absence of follicular ostia on the pink-red background

cream once daily was started and the clinical evaluation after 1 month revealed absence of pustules and no more signs of inflammation. The patient is continuing with clobetasol cream 2 days a week associated with tacrolimus 0.1% ointment 5 days a week.

Discussion

Erosive pustular dermatosis of the scalp (EPDS) is a rare inflammatory condition that usually affects the elderly, while very few cases have been reported in childhood [1–4]. It presents as a chronic eruption of erosions, pustules and crusts that leads to increasing fibrosis and scarring alopecia if not treated promptly. The lesions are often sterile or secondarily colonized. The aetiology is unknown but local trauma, UV radiation and topical medications (including retinoids, imiquimod, and 5-fluorouracil) [4], seem to induce an autoimmune damage in predisposed patients. The reported pediatric cases are infants with necrotic caput succedaneum, intrauterine scalp injury and aplasia congenita and adolescents with previous surgery trauma and third-degree burn [1]. The diagnosis is based on the exclusion of other conditions and the combination of clinical appearance, trichoscopy, histopathology and response to topical steroids or calcineurin inhibitors.

Histopathology is nonspecific and varies depending on the stage of the disease: the early stage is characterized by psoriasiform hyperplasia, a mixed inflammatory infiltrate consisting of neutrophils, lymphocytes, plasma cells, giant cells with foreign body granulomatous reaction against hair shafts, erosions and subcorneal pustules; while progressive atrophy, fibrosis and reduction of follicles are typical of the intermediate-late stages.

In this case the diagnosis is made without skin biopsy on the basis of the clinical appearance, the negativity of bacterial and fungi cultures, the dermoscopy findings and the good response to topical steroids. Trichoscopic findings have been recently described and include: lack of follicular ostia, atrophic skin, follicular crusts and perifollicular pustules [3].

Differential diagnosis includes bacterial and mycobacterial infections, kerion, folliculitis decalvans, pemphigus foliaceus, pustular psoriasis and pyoderma gangrenosum. Topical high-potency steroids and calcineurin inhibitors are the most effective treatments, but recurrence may occur.

EPDS is often misdiagnosed, but it is important to recognize it, especially in paediatric population, because a prompt treatment may result in reduced scarring alopecia. In our case we emphasize that the prompt treatment has allowed hair regrowth.

Key Points

- EPDS is a diagnosis of exclusion
- High-potency steroids and tacrolimus are the first-line treatment
- Early recognition and treatment reduce scarring alopecia

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Chapter 20
Red and Swelling Scrotum as an Early Clue for Diagnosis

Miriam Leuzzi, Giulia Veronesi, Alba Guglielmo, Annalucia Virdi, and Iria Neri

A 3-year-old boy presented in emergency department with acute redness and swelling of the scrotum, painful on palpation, with tense-elastic consistency, not associated with fever or signs of infection. There was no trauma on history. The parents referred an upper respiratory tract infection (URTI) in the week preceding the described symptoms.

After 1 day, a palpable purpura appeared symmetrically on lower limbs and buttocks, less on upper limbs and trunk (Figs. 20.1 and 20.2).

Based upon history and clinical appearance, what is your diagnosis?

1. Leukocytoclastic vasculitis
2. Acute testicular torsion
3. Swelling and erythema of the scrotum in Henoch-Schönlein purpura (HSP)
4. Paraviral erythematous rash
5. Idiopathic thrombocytopaenic purpura

Diagnosis: Swelling and erythema of the scrotum in Henoch-Schönlein purpura (HSP).

Laboratory investigations showed only increase in inflammation parameters. Urinalysis showed no hematuria, proteinuria or nitrites.

Ultrasound sonography of the scrotum revealed acute left epididymitis with moderate hydrocele and thickening of the scrotal wall. The testes were normal-sized, with normal vascularity, thus excluding testicular torsion. Ultrasound sonography of abdomen was normal.

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Fig. 20.1 Swelling and redness of the scrotum with purpuric manifestations of the limbs



Fig. 20.2 Symmetric purpura on buttocks and lower limbs

Blood pressure, heart rate and temperature were within normal age-specific parameters.

No other signs or symptoms were detected.

The patient was treated with a 2-week course of oral amoxicilline-clavulanic acid and paracetamol, with resolution of the swelling of the scrotum and slow recovery of the purpuric rash.

Discussion

Henoch-Schönlein purpura (HSP) is the most common vasculitis in children. It usually affects the small vessels of the skin, joints, gastrointestinal tract and, more rarely, kidneys, brain, lungs and genitalia. It is a systemic leukocytoclastic vasculitis of the small vessels with

perivascular deposition of immunoglobulin A (IgA) immune-complexes leading to inflammation and necrosis of arterioles, capillaries and postcapillary venules [1].

Scrotal swelling and tenderness is described as accompanying symptom in HSP, usually unilaterally and following the purpuric eruption, with a mean prevalence of 11.6% (ranging from 2 to 38%). Also penile involvement is described, with few reports in literature; it may be isolated or associated with scrotal involvement. This complication most likely results from the deposition of immune complexes in the small vessels of penis and testes, which have a complex microvascular structure [2].

Almost all the cases reported in literature describe the swelling of the scrotum after the appearance of the purpuric rash of the body; our case is unusual since the swelling of the scrotum appears before the purpura, making correct diagnosis more difficult.

Testicular torsion have been reported in association with HSP in some cases. History taking, clinical examination and ultrasonographic findings are often sufficient to exclude this fearful condition, thereby negating the need for surgical exploration [3].

Also bacterial epididymitis should be taking in consideration as a differential diagnosis. The concurrent clinical diagnosis of HSP made it unlikely; thus, the decision to start antibiotics is contentious [4].

HSP is usually treated conservatively, with the necessity of non-steroidal anti-inflammatory drugs, steroids and/or antibiotics in some cases.

Key Points

- Scrotal involvement is recognized in HSP and should not be mistaken for testicular torsion.
- When scrotal erythema and swelling occurs before the vasculitic rash, the diagnosis may be difficult
- HSP is usually self-remitting, needing in some cases anti-inflammatory drugs and/or oral antibiotics.

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