Clinical Cases in Dermatology
Series Editor: Robert A. Norman

Francesca Satolli
Michael Tirant
Uwe Wollina
Torello M. Lotti *Editors*

Clinical Cases in Pediatric Skin Cancers

Springer

Clinical Cases in Dermatology

Series Editor
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Clinical Cases in Pediatric Skin Cancers

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Contents

Chapter 1
A Baby with Uniform Papules

Le Huu Doanh, Nguyen Van Thuong, and Michael Tirant

A four-month male presents with uniform papules on his forehead and back, which had been evident for about 1 month. There was no history of similar lesions in family members. Cutaneous examinations revealed relatively equal papules, 1 mm in diameter with some depressed lesions. Lesions focus on forehead, back and a little bit lesions on abdomen (Figs 1.1 and 1.2). Fungal microscopy is negative.

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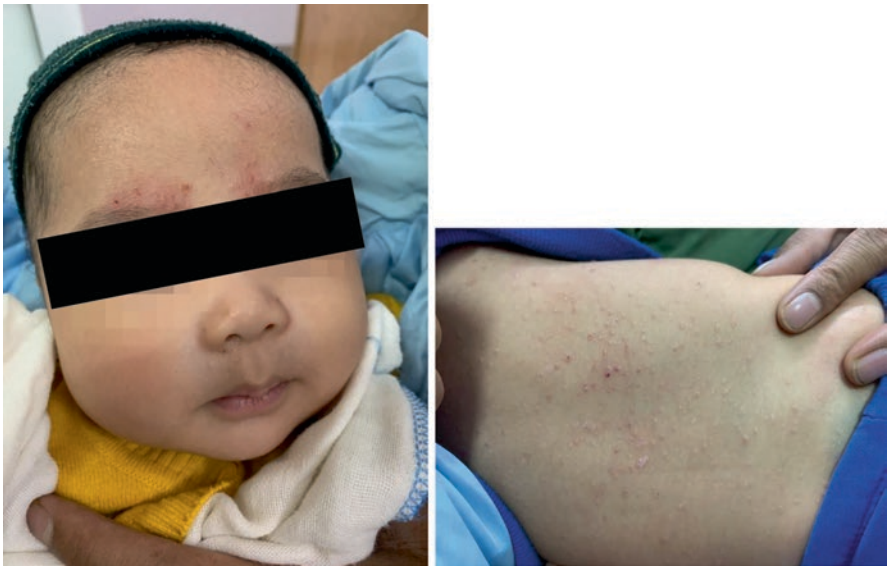

Fig. 1.1 Skin-colored uniform papules 1 mm in diameter. Some depressed lesions

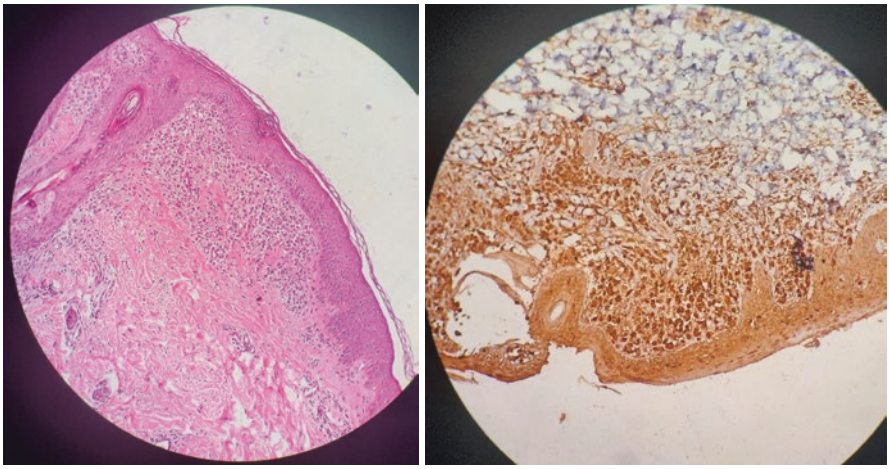

Fig. 1.2 Histopathology showing a dense histiocytes infiltrate of the superficial dermis

Histopathology revealed infiltrated histiocytes with cytoplasmic eosinophils in the superficial dermis, some monocytes and eosinophil in the dermis. Immunohistochemical markers show that S100, CD1a, CD68 are positive.

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

1. Atopic dermatitis
2. Langerhans cell histiocytosis
3. Benign cephalic histiocytosis
4. Seborrheic dermatitis

Diagnosis

Langerhans cell histiocytosis.

Discussion

Langerhans cell histiocytosis is a rare neoplasm of hematopoietic myeloid precursor cells that most commonly affects white male children, with a peak incidence of 1 to 3 years of age. Characteristically, CD1a/S100B/CD207-positive mononuclear cells with bean- shaped nuclei infiltrate single-organ systems, most commonly the bone, but also the skin, or multiple organ systems. Also, approximately 60% of LCH-cells bear a V600E mutation in the *BRAF* (v-Raf murine sarcoma viral oncogene homolog B) oncogene, and 33% of BRAF wild-type lesions harbor mutations in the *MAP 2K1* (mitogen-activated protein kinase 1) gene leading to universal MEK (mitogen activated protein/extracellular signal-related kinase kinase) and ERK (extracellular signal-regulated kinase) activation.

Significant risk factors for LCH include maternal urinary tract infection during pregnancy, feeding problems or blood transfusions during infancy [1] Hispanic ethnicity, crowding, low education level [2] neonatal infections, solvent exposure, family history of thyroid disease [3] and in vitro fertilization [4].

The most commonly affected organs overall are: bone (80%), skin (33%), pituitary (25%), liver (15%), spleen (15%), hematopoietic system (15%), lungs (15%), lymph nodes (5–10%), and the central nervous system excluding the pituitary (2–4%). Cutaneous involvement is typically representative of multisystem disease, because 87% to 93% of patients also have systemic involvement.

In Li et al's retrospective analysis of 918 cases of LCH in China (newborns to patients 65 years of age), 510 patients (56%) were reported to have skin lesions, of which 106 patients (12%) presented with cutaneous lesions as the initial disease manifestation. Cutaneous involvement typically presented as pinpoint erythematous or skin-colored papules or pustules. The morphology can mimic a seborrheic dermatitis or an eczematous erythematous, skin-colored, or brown petechial rash with or without scale, scabbing, crusting, or purpura [5]. This broad variety of skin and mucosal manifestations frequently leads to a delayed diagnosis as skin lesions are misinterpreted as eczema, miliaria, scabies, varicella, seborrheic dermatitis, folliculitis, or candidiasis.

LCH should be kept in mind as a rare, but important, differential diagnosis when the above-mentioned lesions are seen, especially if they are resistant to therapy and are spreading. In the case we presented, cutaneous lesions are also initial disease manifestation and it is difficult to make a right diagnosis in the first examination without histopathology [6].

Key Points

- LCH is an inflammatory myeloid neoplasia.
- Attributed to activating mutations of the MAPK pathway in all patients.
- The clinical course varies from single-system disease, often with spontaneous resolution, to life-threatening, treatment-refractory multisystem disease.
- When LCH affects the skin, it typically presents as pinpoint erythematous or skin-colored papules or pustules mimicking eczema or seborrheic dermatitis.
- Most patients presenting with cutaneous disease also have systemic involvement.
- Identification of patients with risk organ involvement is essential, because these patients need more aggressive treatment.

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Chapter 2
A Boy with a Slow, Painless Nodule and Blister on the Back of His Neck

Feifei Hu and Lujuan Gao

A 12-year-old boy came to our hospital complaining of a slow, painless nodule and blister on the back of his neck, which the patient had noticed three months ago (Fig. 2.1).

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

1. Degenerating fibroxanthoma
2. Foreign body reaction
3. Dermoid cyst
4. Pilomatrixoma
5. Sebaceous cyst

According to the patient's complaint, a nodule on the back of his neck appeared without obvious cause about three months ago. The diameter was about 0.5 cm, which was not taken seriously at that time. The nodules gradually increased since then, and later a blister appeared on the surface one month prior to consultation. The patient started to getting nervous, and told his father about the skin lesion. They referred to the department of dermatology in a nearby community hospital and the diagnose of epidermal cyst was made by ultrasound. However, they doubted the diagnosis.

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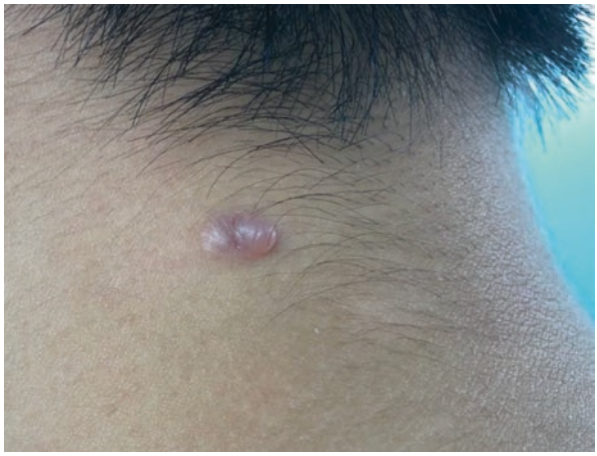

Fig. 2.1 A 12-year-old boy came to our hospital complaining of a slow, painless nodule and blister on the back of his neck, which the patient had noticed three months ago

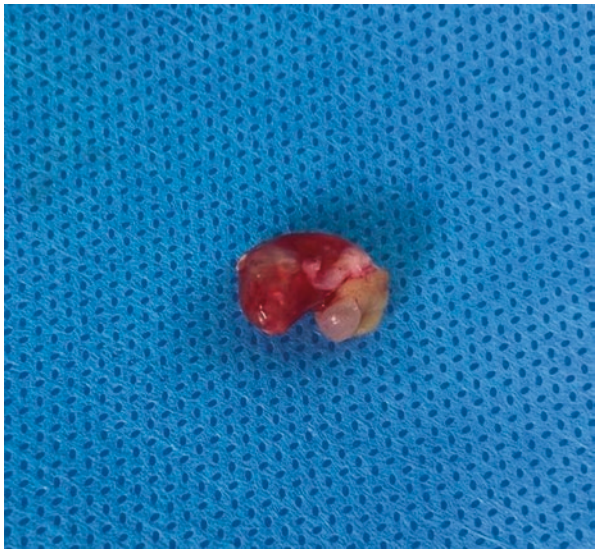

Fig. 2.2 The anatomic sample: well-defined, encapsulated, red-yellow tumor, measuring approximately 1.5 cm in greatest diameter

Upon physical examination, a 1.5-cm fixed, rock-hard and painless nodule with a 0.5-cm blister on the surface was observed on the back of the neck. No other relevant medical information was identified in the patient's clinical history.

We performed surgical resection with clear margins under local anesthesia. The anatomic sample obtained showed well-defined, encapsulated, red-yellow tumor, measuring approximately 1.5 cm in greatest diameter (Fig. 2.2). Histopathological evaluation of the resected tissue reported a benign pilomatrixoma (Fig. 2.3).

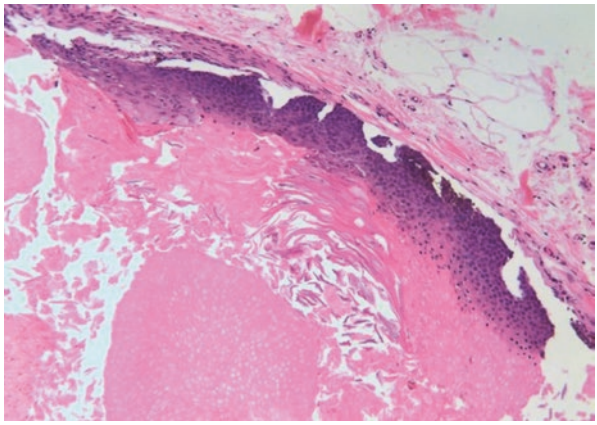

Fig. 2.3 Histopathological: a cyst with central matrical cornification. The cyst wall consists of basaloid matrical cells and shadow cells can be seen in the center. (HE×10)

Diagnosis

Pilomatrixoma (Perforating: Blister type).

Discussion

Pilomatrixoma, also known as pilomatrixoma, is a rare benign tumor arising from the hair follicle [1].

CTNNB1 mutations have been reported in a high percentage of pilomatrixomas. Expression of β-catenin, the protein encoded by CTNNB1, is also frequently observed. It is supposed to be a component of the key signaling pathway that influences cell differentiation and proliferation [1].

Pilomatrixoma is a benign adnexal tumor very common in pediatric age and in young adults that derives from follicular matrix cells. It can be induced by trauma or insect bites. It is most often located in the head and neck, followed by the extremities and upper trunk, while rarely in the lower extremities. So far, no cases of pilomatrixoma have been reported on the palms, soles, or in the genital region [2].

It usually presents as a solitary, slow-growing dermal or subcutaneous nodule without symptoms. The multiple lesions maybe relate to Gardner Syndrome, Turner Syndrome, Rubinstein–Taybi Syndrome, Churg–Strauss syndrome, xeroderma pigmentosum or sarcoidosis. Most masses measure less than 1.6 cm in diameter, while lesions with a diameter of 20 cm were also reported as characterized as “rock-hard” and plate-like on palpation [2].

Several clinical variants of pilomatricomas have been described which include perforating, anetoderma, proliferating, pigmented multiple and familial [3]. The clinical variants increased the difficulty of diagnosis. Perforating pilomatrixoma often displays as a bilster or ulceration appearance. Blister type in the case of our patient is very rare. The “rock-hard” nodule on palpation can assist the diagnosis.

Ultrasound is the most commonest modality used in diagnosis of pilomatrixoma. It is economical and convenient, but only 40–50% of lesions are correctly diagnosed. In our case, it was misdiagnosed as an epidermal cyst by ultrasound. Fine needle aspiration is also an important modality used in diagnosis of pilomatrixoma. The cytological triad of basaloid cells, ghost cells, and giant cells can be diagnosed, but only 44–45% of lesions are correctly diagnosed [2].

Pathology is the gold standard for diagnosis. Pilomatrixomas evolve with time and have been classified into three histopathologic stages: early, fully developed, and regressive.

The early pilomatrixoma frequently presents as a cyst with central matrical cornification. The cyst wall consists of basaloid matrical cells that show an abrupt transition to central eosinophilic cornified matrical cells in which barely discernible nuclear out-lines remain. Sometimes, pink trichohyalin granules, illustrative of matrical cornification, are identifiable at the transition point. The central anucleate cornified cells are commonly referred to as ghost or shadow cells. In fully developed pilomatricomas, a cystic configuration is commonly lost. Solid collections of basaloid matrical cells and matrical corneocytes are present in varying degrees. The cornified matrical cells elicit considerable fibrosis and a secondary granulomatous infiltrate, which can become predominant in longstanding lesions. Matrical cells have a proliferative capacity as high as any human tissue and can display numerous cells in mitosis. The designation proliferative pilomatricoma refers to a pilomatricoma with a high mitotic index. At times, such lesions can be misinterpreted as carcinoma. In late lesions, basaloid matrical cells may be lacking. An involutional pilomatricoma at times can presents with only a few shadow cells buried in a larger fibrosing granulomatous reaction. Calcification and ossification also ensue in late lesions [1].

Complete resection with clear margins is the preferred treatment and serves as diagnostic confirmation. In the case of multiple pilomatrixomas, all lesions should be removed. The pilomatricoma may recur after limited excision. While, in our case, there was no recurrence during a follow-up period of 20 months.

Key Points

- Pilomatrixoma is a rare benign tumor arising from the hair follicle, which is quite common in childhood and adolescence.
- Complete resection is the preferred method.

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Chapter 3
A Boy with Recurrent Erythema and Blisters

Yang-Yang Luo, Jian-Ping Tang, Zhu Wei, Jing Chang, and Bin Zhou

An 1-year old boy was admitted to our ward complaining of recurrent erythema, wheal and blisters for the past seven months. He had pigmented and hypopigmented spots on the trunk at 3 months after birth. Sometimes there were wheals or blisters (Fig. 3.1a) due to scratching, temperature rising or vaccination. Moreover the rash spreaded all over the body and was presented with peau d'orange appearance (Fig. 3.1b). Physical examination did not present alterations in other organs or systems.

Based on the History and the Photography, What Is Your Diagnosis?

1. Bullous pemphigoid
2. Papular urticaria
3. Urticarial pigmentosa
4. Diffuse cutaneous mastocytosis
5. Mycosis fungoides

Following his hospitalization, he received related auxiliary examinations including routine blood tests, abdominal ultrasonography, skin biopsy and bone marrow biopsy. The skin histological appearance showed numerous mast cells arranged diffusely in the dermis stained by Hematoxylin&eosin (Fig. 3.2a). And the CD117

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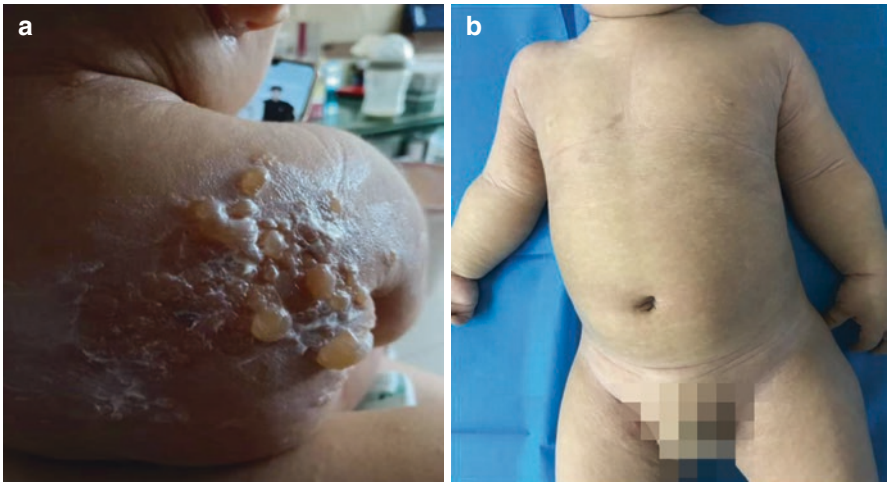

Fig. 3.1 Clinical manifestation of the patient. (**a**) recurrent blisters on erythema; (**b**) peau d'orange appearance

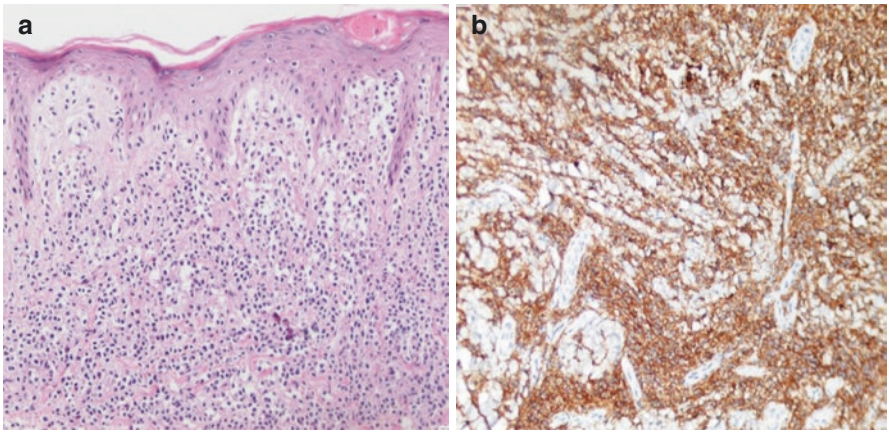

Fig. 3.2 The characteristics of histopathology. (**a**) numerous mast cells arranged diffusely in the dermis stained by Hematoxylin eosin; (**b**) the CD117 (c-kit) immunohistochemical reaction was diffusely positive in mast cells

(c-kit) immunohistochemical reaction was diffusely positive in mast cells (Fig. 3.2b). Other laboratory and imaging investigations did not show any evidence of systemic abnormalities.

Then he received the therapy of intravenous glucocorticoids and antihistamines. The pruritus was relieved and no new blister was discharged. After that, the glucocorticoids was gradually reduced to discontinuation. One year later, there is no pruritus or new rash in the patient.

Diagnosis

Diffuse cutaneous mastocytosis.

Discussion

Mastocytosis is a heterogeneous disorder characterized by the pathological increase and accumulation of mast cells (MCs) in different tissues and organs [1]. It is divided into 3 main forms: cutaneous mastocytosis (CM), systemic mastocytosis (SM) and mast cell sarcoma depending on the sites of organ involvement. And the CM includes 3 variants: urticarial pigmentosa, diffuse CM (DCM) and mastocytoma of skin [2]. DCM is an extremely rare and severe variant that usually manifests with erythema, dermal thickening and edema with a typical leather or peau d'orange appearance [3]. Cutaneous symptoms consist of widespread spontaneous blisters with erosions, erythroderma or thickening of the skin and positive Darier's sign (rubbing of skin lesions results in the reddening and urticarial swelling). Other MC mediator-related symptoms due to the widespread MC load in the entire skin includes simultaneous occurrence of pruritus, flushing, vomiting, diarrhea, abdominal pain, hypotension and anaphylactic shock [4].

Urticarial pigmentosa is the most common variant of cutaneous mastocytosis characterized by hyperpigmented, brownish macules and patched with positive Darier's sign. Unlike other forms of mastocytosis, there is rarely any internal organ involvement [5].

Bullous pemphigoid is an autoimmune bullous disease classically characterized by tense blisters over urticarial plaques on the body accompanied by intense pruritus. Diagnosis relies on the histopathological results demonstrating eosinophilic spongiosis, the detection of IgG and/or C3 deposition at the basement membrane zone using direct or indirect immunofluorescence assays and the quantification of circulating autoantibodies against BP180 and/or BP230 using ELISA [6].

Papular urticaria is a chronic inflammatory disease characterized by a hypersensitivity reaction to the bite of arthropods and manifested through papule-type skin lesions, wheals, vesicles, blisters or scabs. Occasionally, the patient might develop hypo-or hyperchromic pigmentations in the skin with intense pruritus [7]. While mycosis fungoides is the most common cutaneous T-cell lymphoma, a type of non-Hodgkin T-cell lymphoma presenting with cutaneous patches, plaques, and tumors [8].

The patient was presented with typical peau d'orange appearance and positive Darier's sign. The histopathology showed numerous mast cells in the dermis with positive CD117. Diffuse cutaneous mastocytosis is clearly diagnosed. The mainstay of treatment includes controlling mast cell mediator-related symptoms and avoiding triggers. Treatment options include antihistamines, systemic and topical corticosteroids, mast cell stabilizers and psoralen with UVA [9].

Key Points

- Diffuse cutaneous mastocytosis is rare diease characterized by erythema, dermal thickening and edema with a typical leather or peau d'orange appearance.
- Antihistamines, systemic and topical corticosteroids are the main treatment options for DCM.

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Chapter 4
A Case of Ulcerated Hemangioma

Neslihan Deniz and Ümit Türsen

Introduction

A 1-year-old infantile male patient presented to us with a 60-mm × 40-mm right ear and face superficial protuberant infantile hemangioma that appeared at birth growing steadily over time and then gradually developed 30-mm × 35-mm ulceration of the inferior half over the last 6 weeks. In his medical history, he had systemic steroid, oral propranolol and sirolimus drug medication. With the regression of the lesion that benefited from the treatment, discontinued the treatment and relapsed. Prednizolon 1.5 mg/kg/day and propranolol 3 mg/kg/day were restarted by our pediatric oncology department. Prednizolon was discontinued in outpatient clinic examinations 1 month later and then 2 drops of 0.5% betoxalol solution was applied thrice daily over the entire lesion by our dermatology department. The ulcer complete healing was achieved by 3 month, leaving a whitish scar. At monthly outpatient clinic reviews, the child's recorded vital signs ranged as follows: heart rate, 120–135 beats per minute; blood pressure, 80–96/55–73 mmHg; and random glucose, 5.6–8.2 mmol/l. Propranolol and 0.5% betoxolol solution were continued for 6 months, and there was no ulcer recurrence at 12 months after stopping treatment (Figs. 4.1 and 4.2).

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

- Capillary malformation
- Congenital hemangiomas and vascular malformations
- Kaposiform hemangioendothelioma and tufted hemangioma
- Subcutaneous tumors
- Pyogenic granuloma

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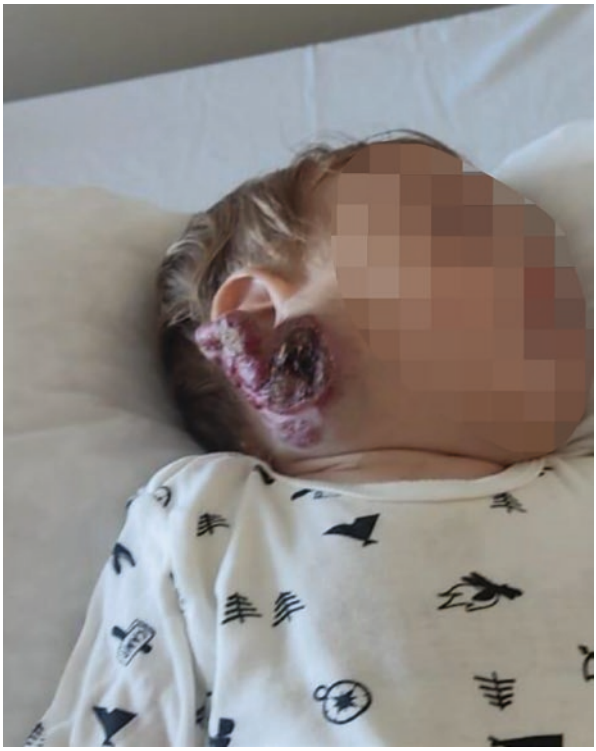

Fig. 4.1 Clinical evolution of an infantile ulcerated hemangioma on upper ear of patient

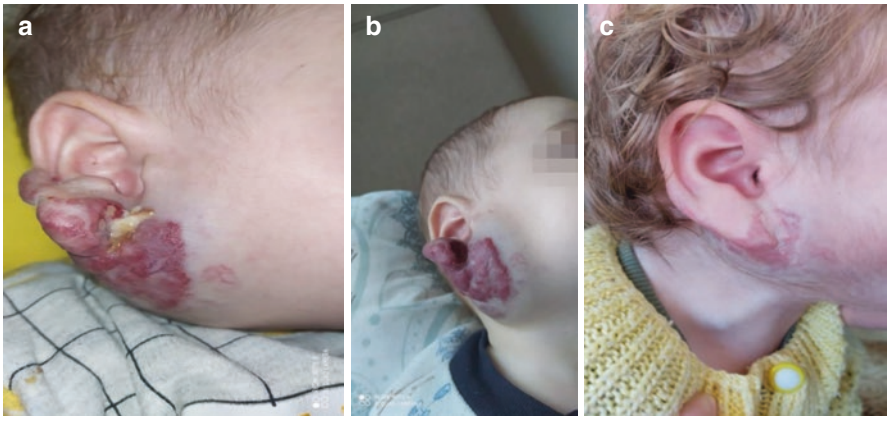

Fig. 4.2 (**a**) Right ear hemangioma (60 mm × 40 mm) with inferior-half ulceration (30 mm × 35 mm) in a 1 year-old child before starting propranolol and prednol. (**b**) 2 months after prednol and propranolol treatment (**c**) After 12 months of follow-up, complete healing after 6 month of propranolol, prednol, %0.5 betaxolol solution, leaving behind a whitish telangiectasic scar

Diagnosis

Ulcereted infantile hemangioma.

Discussion

Infantile hemangiomas are the most common tumors of childhood. Although most are benign and self-limited some hemangiomas can cause complications such as ulceration or life-altering disfigurement. Treatment of hemangiomas should be determined by the size, morphology, location of the lesion (s), the presence or possibility of complications, the potential for scarring or deformity, the age of the patient, and the rate of growth or curl [1] Ulceration is common complication of hemangiomas and is usually in the centrofacial and perineum regions. Propranolol is the first-line agent for hemangiomas with the potential to impair function or cause permanent disfigurement. Potential mechanisms of action for propranolol may include vasoconstriction, decreased expression of vascular endothelial growth factor and, triggering of apoptosis [2, 3]. The randomized controlled trial shows that propranolol is effective in the treatment of infantile hemangioma at a dose of 3 mg per kilogram for 6 months. 10% of patients in whom treatment with propranolol was successful required systemic retreatment during follow-up. Known adverse events associated with propranolol (hypoglycemia, hypotension, bradycardia and bronchospasm) were observed infrequently [4]. Rebound growth was observed in approximately 14–25% of children after propranolol was discontinued [5].

Topical beta blockers can be used to treat mild to moderate relapses. A retrospective study was conducted to evaluate the efficacy of timolol gel solution in the treatment of ulcerated infantile hemangioma. In this study, 30 children with ulcerated infantile hemangioma were identified. The results show that timolol can be well tolerated with oral propranolol [6]. Systemic corticosteroids were compared with propranolol in randomized trial. In this study, 34 children with facial hemangioma were assigned to treatment with propranolol 2 mg /kg/day and prednisolone 2 mg/kg/day for 16 weeks. Response to treatment, generally defined as cessation of progression and volume reduction, was achieved by 96 percent and 92 percent of patients in the propranolol and systemic corticosteroid groups, respectively. This study showed that propranolol is not inferior to steroid in terms of therapeutic effects in IH [7]. Abrupt withdrawal or rapid reduction of glucocorticoids should be discontinued slowly, as rebound proliferation may occur. Pulsed dye laser (PDL) 595 nm can be used as an alternative for medically resistant ulcerated hemangiomas. In summary, in our patient, positive results were obtained in the combined treatment of oral propranolol and low dose prednisolone. In our patient, topical beta blockers were used in the treatment and good results were obtained. No recurrence was experienced.

Key Points

- Oral propranolol and low dose oral prednisolone are very effective in the treatment of ulcerated infantile hemangioma.
- -Topical beta blockers are used as alternatives in the treatment of ulcerated infantile hemangioma.

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Chapter 5
A Case of Unusual Erythematous and Desquamative Skin Disorders in a Child

Gizem Aydın, Mustafa Anıl Yılmaz, Yasemin Yuyucu Karabulut, Ümit Türsen, and Kıymet İnan

A 9-year-old male patient presented with a non-itching red swelling on lower abdomen for two months. There is no medical illness in the medical history of his family and himself. It was initially treated as tinea by family physician then he was sent to our hospital for the persistence of the plaque.

In dermatological examination, there are two 4*2.5 and 4*1.5 cm annular erythema plaques in the suprapubic area, 3*2 cm in the right inguinal region and 4*2 cm in the left lumbar region (Fig. 5.1).

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

- Lichen scleroatrophicus
- Fixed Drug Eruption
- Morphea
- Mycosis Fungoides

Biopsy was performed and the histomorphological examination of the biopsy specimen, orthokeratosis and follicular plugging were observed in the epidermis. Lymphocytes that fill the papillary dermis also lie transepidermally. These lymphocytes have significant cytological atypia such as nuclear enlargement, nuclear contour irregularity and perinuclear halo. In the immunohistochemical study, strong cytoplasmic staining with CD3, CD4 and CD5 in lymphocytes which filling the dermal papillae, showing alignment at the dermoepidermal junction and spreading transepidermal in patches was observed. Some of these lymphocytes showed strong

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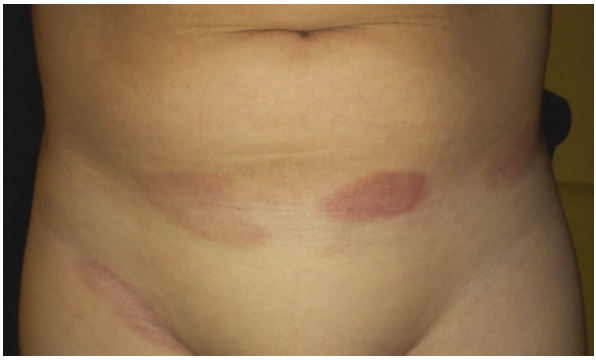

Fig. 5.1 There are annular erythema plaques in the suprapubic area, right inguinal region and left lumbar region

cytoplasmic staining with CD8 and the ratio CD4/CD8 was 4. Most of the lymphocytes that filling the dermal papillae showed loss of expression with CD7 (Fig. 5.2). Moderate cytoplasmic staining was seen in very few lymphocytes in the dermal papillae with CD30 and CD20 positivity was not observed.

Diagnosis

Mycosis Fungoides.

Discussion

Mycosis fungoides, which is characterized by infiltration of the skin with malignant T cells is the most common primary cutaneous lymphoma in adults and children. It comprises approximately %65 of all primary cutaneous lymphomas in pediatric patients. The mean age at diagnosis in pediatric patients is 10, and this condition is rare in infants. The male to female ratio of MF is 1.1: 1 in young patients under 30 years old [1].

Similar to adults, children may have an indolent clinical course that is difficult to distinguish from inflammatory skin conditions [2]. In adults, classic MF presents initially with scaly erythematous patches that may progress into infiltrated plaques and tumors in %35 and %20 of patients. The incidence of classic MF in children is approximately %41. Unlike adults, the majority of children with MF present with nonclassic variants of the disease, which include hypopigmented, hyperpigmented, folliculocentric, and poikilodermatous forms. Multiple variants are often present at the time of diagnosis. Hypopigmented form is >%50 of pediatric cases [1].

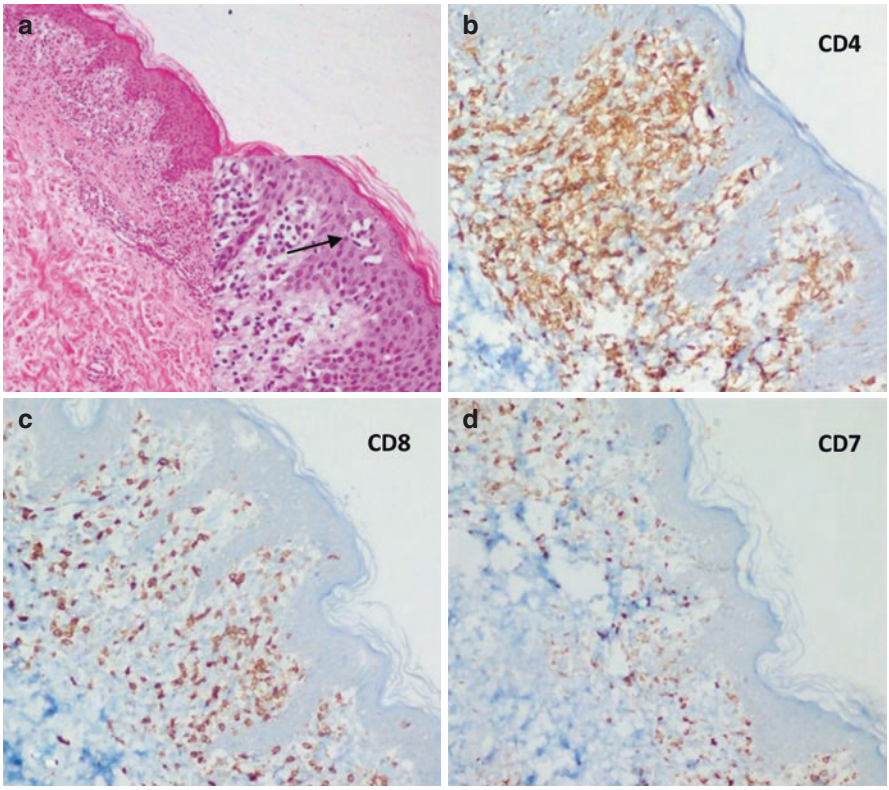

Fig. 5.2 (**a**) Lymphocytes that fill the dermal papillae and progress transepidermally were seen in the fibrotic papillary dermis (H&EX100), arrow; Pautrier's microabscess. (**b**) CD4 expression in a large number of lymphocytes (CD4X200), (**c**) CD8 expression in fewer lymphocytes (CD7X200). (**d**) CD7 expression in fewer lymphocytes (CD8X200)

Clinically, the cutaneous lesions of mycosis fungoides typically progress through three somewhat distinct stages, an inflammatory premycotic phase, a plaque phase and a tumor phase. The patch stage includes superficial lichenoid infiltrate, mainly lymphocytes and histiocytes and a few atypical cells infiltrating the epidermis without significant spongiosis. This stage may mimic other dermatoses such as eczema or lichenoid dermatoses, like our case. In pediatric cases the atypia may be difficult to appreciate and multiple biopsies are required to make the diagnosis and numerous studies may be needed to prove a clonal proliferation of T cells. In plaque stage, mycosis fungoides progresses, there is more obvious epidermotropism and a denser dermal infiltrate. There may be intraepidermal collections of atypical cells. When tumours start to form clinically, histomorphology shows a much denser dermal infiltrate. There may be no exocytosis of lymphocytes in this stage. Transformation to large cells may occur [3]. Sezary syndrome is the leukemic form of the disease in

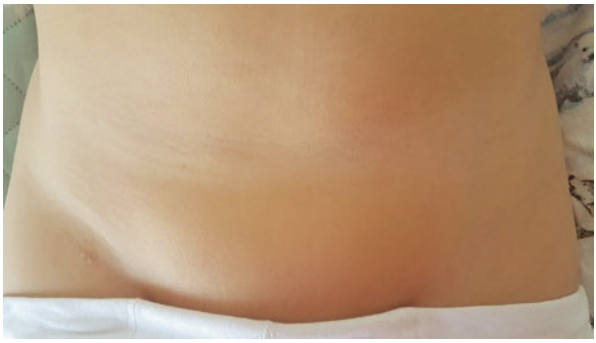

Fig. 5.3 Regression of the lesions after phototherapy

which erythroderma is accompanied by measurable blood involvement by malignant lymphocytes with hyperconvoluted, cerebriform nuclei known as Sezary cells [4].

The neoplastic lymphocytes typically have a T-helper phenotype (CD3+, CD4+, CD8−); less commonly, the neoplastic cells may express cytotoxic T-cell phenotype (CD3+, CD8+, CD4−). Studies have shown the cytotoxic T-cell phenotype is more frequently seen in children than adults and the CD8+ variant may be associated with an indolent course. The loss of CD7 expression can be observed even in the early phases of the disease [1]. Pautrier's microabscesses (lymphocytes with cerebriform nuclei and formation of intraepidermal aggregates) are pathognomonic for MF and are rarely seen in other lymphomas.

According to the studies conducted for the last 10 years, the most common treatment method is phototherapy. Topical corticosteroids were often used in combination with phototherapy and were used as the sole treatment in %9 of patients. Other topical agents such as retinoids (%6) and topical nitrogen mustard (%2) were used less frequently. Local radiation and surgical excision were used in a minority of cases (%1) [1].

The patient was threated with phototherapy (narrow-band UVB two times a week). Significant improvement was observed clinically after the current treatment (Fig. 5.3).

Key Points

1. Mycosis Fungoides is the most common primary cutaneous lymphoma in adults and children.
2. Children with MF often present with nonclassical variants of the disease. The most common of these are hypopigmented, hyperpigmented, folliculocentric and poikilodermic types.
3. Phototherapy is the most commonly used treatment method in pediatric patients.

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Chapter 6
A Complex Skin Disease

Tugba Kevser Uzuncakmak, Ayşe Mine Önenerk, and Zekayi Kutlubay

Case

A 16-year-old male presented to the dermatology department with a 2-month history of rapidly growing crusted and ulcerated lesions on his forehead. Clinical examination revealed several lentiginous proliferations on sun exposed areas on face, extansor surfaces of upper extremity and on pretibial region. He has widespread crusted erosions on scalp, frontal region, perioral region, periorbital edema, a 2 cm ulcerated lesion with oozing and a 4 mm pink nodular lesion beyond this ulcerated lesion. (Fig. 6.1). His lesions were first appeared when he was 2 years old and he was under follow up in the departments of dermatology, plastic surgery and pediatric oncology, routinely. On dermoscopic examination ulceration, telengiectatic vessels on a pink structureless basis and superficial scales were detected (Fig. 6.2). In his laboratory tests including complete blood counting, biochemistry, erythrocyte sedimentation rate and C-reactive protein, an increase in acute phase markers were detected.

He was referred to plastic surgery department for excisional biopsy. Histologically,well differiated infiltrated squamous cell carcinoma and basal cell cell carcinoma were detected (Fig. 6.3). No perivascular or perineural invasion was detected. He was referred to department of oncology for further investigation.

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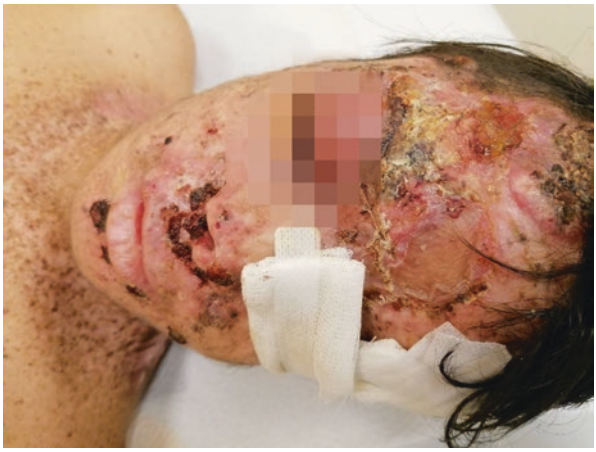

Fig. 6.1 Lentiginous proliferations, widespread crusted erosions on scalp, frontal region, perioral region, periorbital edema, a 2 cm ulcerated lesion with oozing and a 4 mm pink nodular lesion beyond the ulcerated lesion

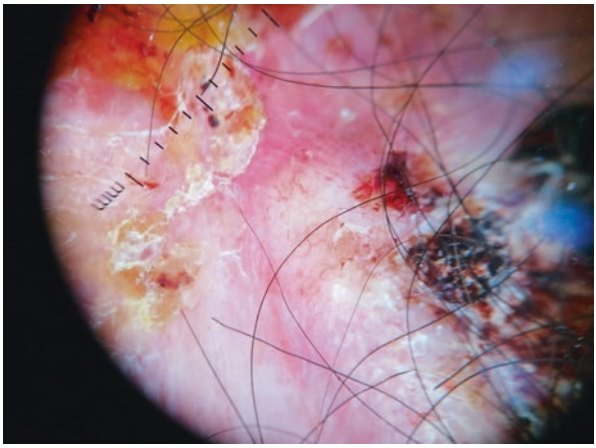

Fig. 6.2 A 4 mm pinkish structurelesss papular lesion with peripheral telengiectasias near crusted erosion on frontal region

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

- Hydroa vacciniforme
- LEOPARD syndrome
- Rothmund- Thompson syndrome
- Xeroderma pigmentosum

Diagnosis

Basal cell carcinoma and squamous cell carcinoma in xeroderma pigmentosum.

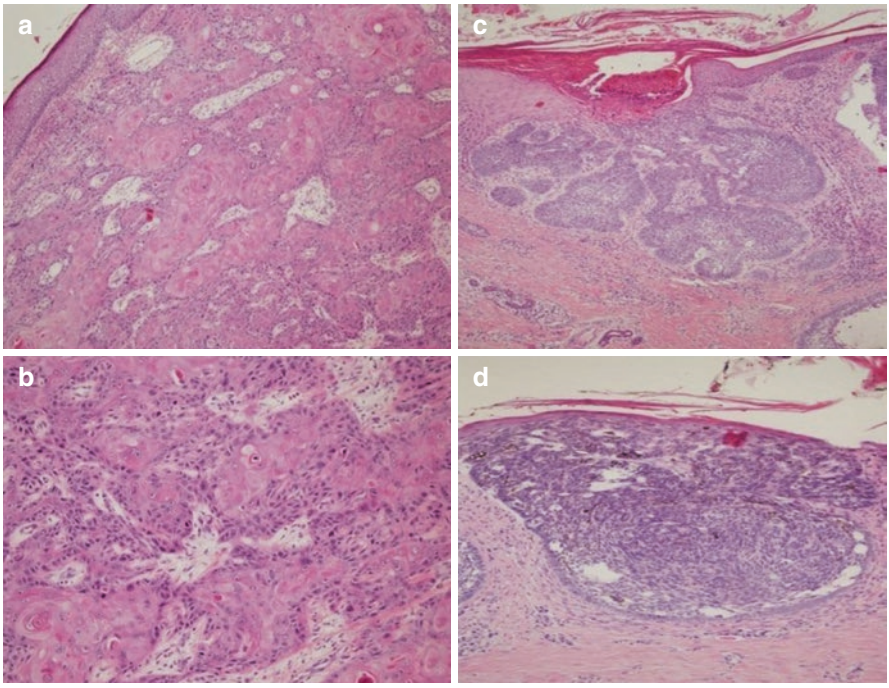

Fig. 6.3 (**a**) Well differentiated squamous cell carcinom (H&E, ×200), (**b**) Well differentiated squamous cell carcinom (H&E, ×400), (**c**) Basal cell carcinoma, nodular type (H&E, ×200), (**d**) Basal cell carcinoma, superficial type (H&E, ×200)

Discussion

Xeroderma pigmentosum (XP) is a rare hereditary disorder characterized by impaired DNA repair function [1]. This rare disorder was first defined by Hebra and Kaposi in 1874, afterwards, in 1882, the term xeroderma pigmentosum was used by Kaposi, referring to its characteristic dry and pigmented skin. It has been reported in all races with an equal prevalence in males and females. Characteristic lesions of XP usually begin at age of 1–2 years. The prevelance of XP is estimated to be approximately 1/250,000 population in United States. This ratio is approximately similar in Europe and reported to be higher (1 case per 40,000 population) in Japan [1, 2]. Similar with the other autosomal recessive disorders, the parents are heterozygotes and they are healthy. Usually there is no family history.

The disease is seen in 3 clinical phases according to clinical findings. Although the skin looks healthy at birth, typically, a diffuse erythema, lentiginous pigmentation and scaling occur after 6 months. These findings are commonly seen on sun-exposed areas, appearing especially on the face. Then these skin changes appear on the other parts of the body usually on lower legs, the neck, and even the trunk in

extreme cases by the time. Although these features may seem to diminish during the winter months due to the decreased sun exposure, they become permanent sooner [1].

In the second stage of the disease, poikiloderma, including mottled hyperpigmentation and hypopigmentation, telangiectasias and skin atrophy, is the main cutaneous finding. Telangiectasias may occur both in the sun-exposed areas and in unexposed skin and even on mucosal surfaces. In the third stage, different malignancies, including squamous cell carcinomas, malignant melanoma, basal cell carcinoma, and fibrosarcoma may occur. These malignancies may occur as early as age 4–5 years and are more prevalent in sun-exposed areas. Nearly half of the patients survive beyond age 20 years. Metastatic melanoma and squamous cell carcinoma are the important causes of mortality in these patients. It was reported that XP patients have an 10,000-fold greater risk of developing cutaneous malignancy when compared to the general population, median onset age of cutaneous cancers was also reported to be <10 years [3]. Actinic damage occurs very early, between ages 1 and 2 years. Education of the patient and the caregivers is the most important topic in the management of xeroderma pigmentosum. The need for adequate solar should be reinforced at every visit. Sunblocks should be used, even in winter months and during evening and early morning hours. The exposed surfaces of the skin should be shielded with protective, double-layered clothing and broad-brimmed hats. The eyes should be shielded with UV-absorbing sunglasses with side shields. Ocular problems may occur in aşmost 80% of individuals with xeroderma pigmentosum. The most common ocular problems are photophobia and conjunctivitis. The propensity for malignancies, such as squamous cell carcinoma, basal cell carcinoma, sebaceous cell carcinoma, and fibrosarcoma, can also involve the eyes of patients with xeroderma pigmentosum.

The main goal in the treatment of patients with XP is to protect the patients from sun exposure. Using waterproof sunscreens including both chemical and physical filters is a must do for these patients. Topical 5 fluorouracil, imiquimod 5%, systemic acitretin may be iniated for actinic keratosis. Using DNA repair enzyme into the skin via specially engineered liposomes is anew treatment modality in patients with XP. T4 endonuclease V has been shown to repair cyclobutane pyrimidine dimers resulting from DNA damage. No significant adverse effects were found among any of the patients. It was shown to lower the rate of development of basal cell carcinoma lesions after 1 year of treatment [4].

Gene therapy for xeroderma pigmentosum is still in experimental stage. Several pathways of correcting the defects in xeroderma pigmentosum have been shown in vitro and in animal studies using viral vectors (adenoviruses and retroviruses) carrying the gene replacement products. Skin grafting with the genetically corrected skin was also noted to be useful in xeroderma pigmentosum patients in the future.

Hydroa vacciniforme (HV) is a rare photodermatosis with an unknown etiology. It usually occurs in childhood accepted within the spectrum of *Epstein-Barr* virus–related lymphoproliferative disorders [5]. Clinically it is characterized by recurrent vesicles on sun-exposed skin that heal with vacciniform or varioliform scarring.

Histopathologically, intraepidermal reticular degeneration and cellular necrosis are commonly detected. Most of the patients heal spontaneously during late adolescence. Commonly, mild burning, itching, or stinging in exposed sites begins a few hours or days after sun exposure. Vesicles heal with varioliform scarring. The lesions often occurs in spring, with recurrences in summer [5].

LEOPARD syndrome is a complex dysmorphogenetic disorder of variable penetrance and expressivity. This syndrome is the acronym LEOPARD of the main features of the disorder, including eentigines (multiple), electrocardiographic conduction abnormalities, ocular hypertelorism, pulmonary stenosis, abnormalities of genitalia, retardation of growth and deafness [6]. The diagnosis of LEOPARD syndrome may be very difficult in early childhood and can be clinically suspected in patients who have 3 main features: characteristic facial features, hypertrophic cardiomyopathy, and café au lait spots in the first months of life.

Rothmund-Thomson syndrome, or poikiloderma congenitale, is another rare autosomal recessive disorder associated with mutations in the *RECQL4* helicase gene located on 8q24 [7]. It is characterized by early photosensitivity and poikilodermatous skin changes, juvenile cataracts, skeletal dysplasias, and a predisposition to osteosarcoma and skin cancer. Patients usually present with a rash (poikiloderma), small stature, and skeletal dysplasias. The acute phase begins in early infancy as red patches or edematous plaques, sometimes with blistering. Clinically lesions usually firstly begin from the cheeks then later occur on other areas of the face, the extremities, and the buttocks. In chronic stage cutaneous manifestations are characterized by poikiloderma (atrophy, telangiectasias, and pigmentary changes). These changes are typically seen on the face, extensor surfaces of extremities, and buttocks with sparing of the chest, abdomen, and back. Acral hyperkeratotic lesions can be seen on the elbows, knees, hands, and feet during puberty. Photosensitivity is a feature in more than 30% of cases. Gastrointestinal and hematological abnormalities have also been noted to occur.

Key Points

- Xeroderma pigmentosum is an autosomal recessive DNAa repair disorder h dermatological, ocular, and neurological manifestations with skin cancer predisposition.
- Clinically lesions usually appear by 2 years of age with increased number of lentigines (freckle-like pigmentation) in sun-exposed areas and extreme sensitivity to sunlight resulting in acute severe sunburns.
- Cutaneous malignancy risk is almost 10,000 folds higher in XP patients than the general population.
- The median age of cutaneous malignancies is <10 years in XP patients.
- Avoiding from sun exposure is the main therapeutic approach in XP patients.
- Topical 5 fluorouracil cream, imiquimod 5% cream and systemic acitetin can be offered for actinic daamage.

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Chapter 7
A Congenital Lesion of the Scalp with an Unusual Shape

Fabio Arcangeli and Elisa Sama

An 18-month-old male infant presented for evaluation of a well-defined linear and raised lesion on the right posterior parietal scalp (Fig. 7.1). His parents reported its presence from birth. The lesion had increased progressively in size until reaching 3.5 × 1 cm and had changed colour from intense red to yellowish pink.

The patient did not have any related symptoms except for sparse hair at the lesion. The medical history did not reveal any pregnancy or post-pregnancy trauma.

The physical examination showed a yellowish pink lesion, with a smooth and shiny surface, very similar to a scar lesion but with a soft consistency (Fig. 7.2).

A previous ultrasound examination excluded intracranial connection and brain abnormalities. This was subsequently confirmed by a nuclear magnetic resonance examination.

In the absence of a clinical diagnosis a surgical excision was planned.

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

1. Encephalocele
2. Lipoma
3. Meningothelial hamartoma
4. Keloid

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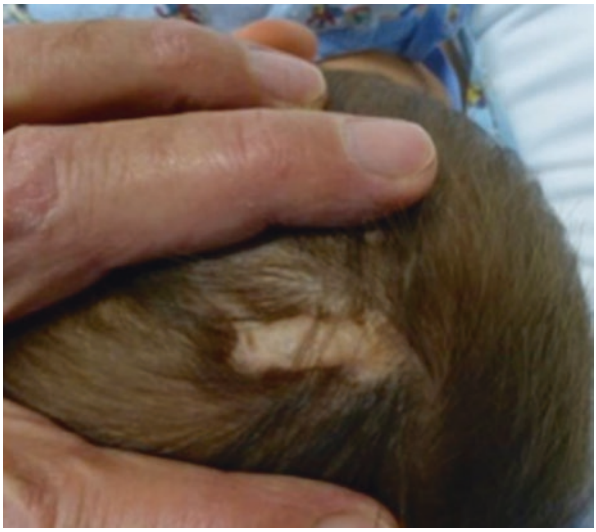

Fig. 7.1 A yellowish pink raised lesion on the posterior scalp

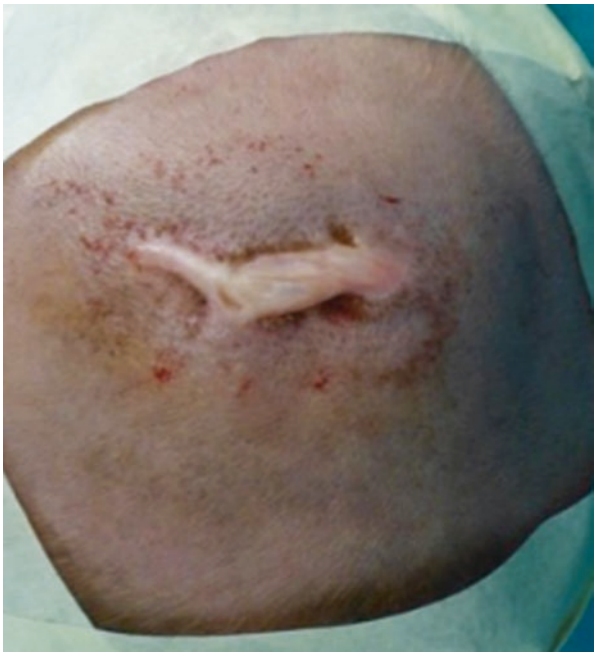

Fig. 7.2 The unusual shape lesion with a smooth and shiny surface

Diagnosis

Meningothelial hamartoma.

Discussion

A total excision was performed. The histological examination showed pseudo-vascular lumens bordered by often spindle shaped elements. They were devoid of significant atypia and sometimes multinucleated, arranged in a disordered way and mixed with connective fibers and adipose tissue (Fig. 7.3). Because the diagnosis was still unclear, to exclude other possible diagnosis such as angiosarcoma or melanoma, immunohistochemistry was performed. It resulted negative for HMB-45, S-100, cytokeratins, factor VIII and positive for epithelial membrane antigen (EMA), vimentine and focally for CD 68 (Fig. 7.4).

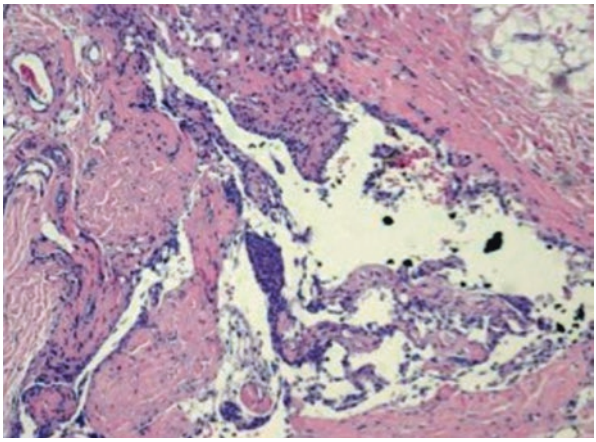

Fig. 7.3 The histologic examination showed a pseudo-vascular pattern, with disseminated epithelioid cells mixed up with connective fibers and adipose tissue, within deep dermis

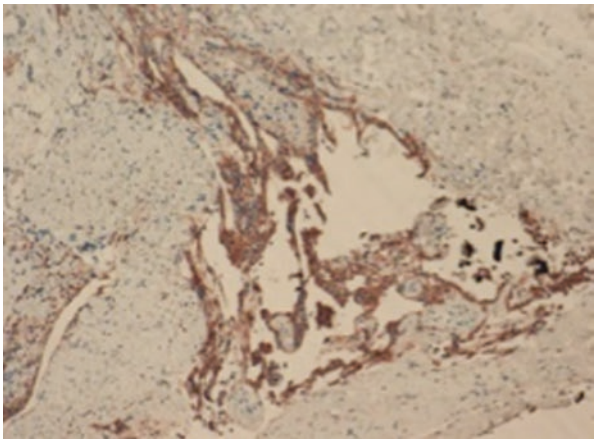

Fig. 7.4 The immunohistochemistry revealed a strong positivity for epithelial membrane antigen (EMA) and focally positivity for CD 68

On the basis of the histological examination and the immunohistochemical study a diagnosis of meningothelial hamartoma was made. One year after surgery no signs of recurrence were present.

Soft tissue masses of the scalp are commonly encountered in clinical practice. Pediatric scalp lesions are fortunately benign in most cases (98%) [1]. Congenital lesions include menigocele or encephalocele, due to a neural tube defect where part of the skull has not formed properly, dermoid cyst, lipoma and others [2]. It is important to consider that the lack of lesional mobility or a midline location should prompt an initial radiographic evaluation to rule out cranial meningo or encephaloceles.

Encephaloceles are usually found immediately after birth but sometimes a small encephalocele can go undetected. Encephalocele is a sac-like protrusion of the brain and the meninges through an opening in the skull. In our case the ultrasound examination excluded any communication with intracranial space.

Lipoma is the most common benign tumor of the scalp. It presents as a soft subcutaneous mass with an elastic or rubber consistency. In our case the shape and consistency did not suggest the diagnosis of lipoma.

Keloid is an abnormal proliferation of fibrous tissue that forms at the site of a cutaneous injury. It grows beyond the original margins of the scar, creating typical crab claw-like shapes. Keloid usually has an erythematous, smooth and shiny surface. Its consistency can range from soft to rubber but in most cases, it is very hard. In our case the shape of the lesion could suggest a diagnosis of keloid but no trauma was reported and the consistency was very soft.

Meningothelial hamartoma, first described by Suster and Rosai in 1990 is a collection of meningothelial elements in an ectopic location. It is characterized by a mixture of various components (among which meningothelial elements) of the connective tissue arranged in a disordered way in the dermis. It was also called "rudimentary meningocele", "sequestered meningocele" and "hamartoma of the scalp with ectopic meningothelial elements" [3]. It may have been associated with rare hair, as in our case, and the most common sites involved are the occipital midline and posterior regions of the scalp. Locations on parieto-occipital area, vertex and forehead regions have been reported too [3]. In general, it does not extend beyond the subcutis. The cause of meningothelial hamartoma is still unknown. Several hypotheses were proposed: some authors think it comes from ectopic meningothelial remains during embryologic development, some others have suggested the abortive migration of cells from the neural crest or that it is a form of meningocele with an obliterated intracranial communication [3].

The treatment of choice is surgical removal.

Key Points

- Soft tissue masses of the scalp are common and they are benign in most cases
- Congenital lesions include menigocele or encephalocele, dermoid cyst, lipoma and others
- The lack of lesional mobility or a midline location should prompt an initial radiographic evaluation to rule out cranial meningo or encephaloceles