Chapter 3
A Pediatric Patient with Photosensitivity

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

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

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

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

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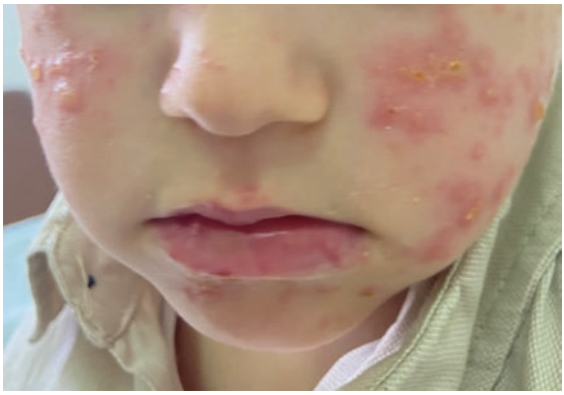

Fig. 3.1 Active lesions in various stages over the central part of the face

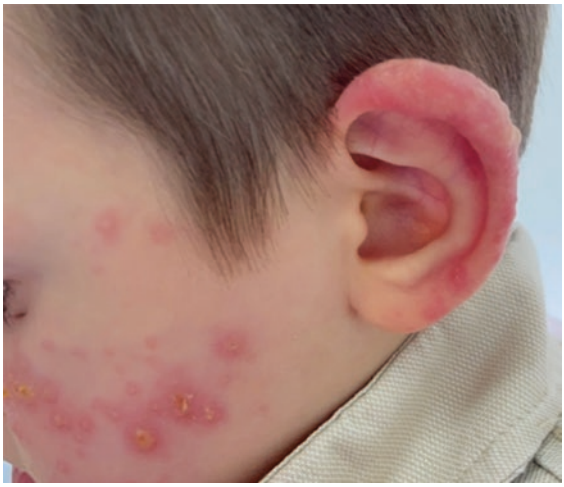

Fig. 3.2 Diffuse erythema and small vesicles on the auricular helix

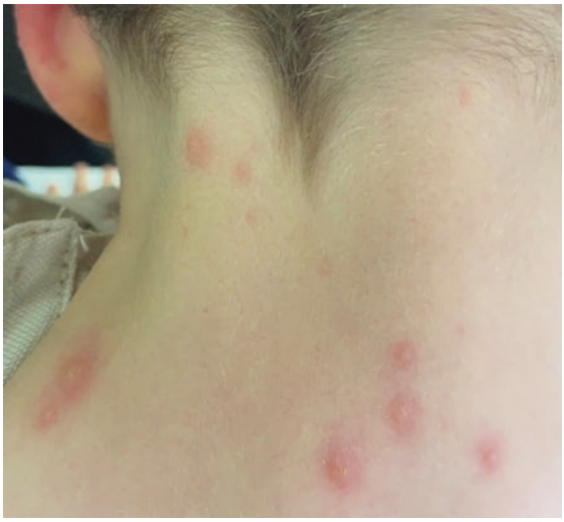

Fig. 3.3 Tense vesicles with surrounding erythema over the neck

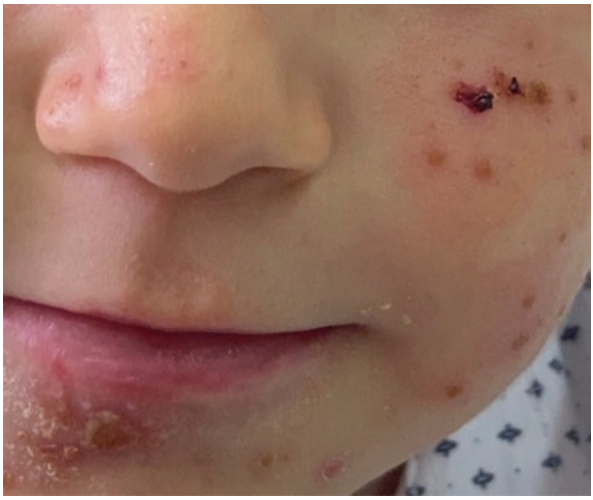

Fig. 3.4 Vesicles and papules were turning into necrotic crusts in the face a week after the first visit

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

Diagnosis

Classical hydroa vacciniforme.

Discussion

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

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

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

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

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

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

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

Key Points

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

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

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