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CASES FOR DIAGNOSIS

Intermittent Vesiculous Eruption in a Child

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Clinical History

The patient was a 10-year-old boy with no relevant personal or familial medical history, who attended our clinic for recurrent episodes of lesions on areas exposed to light. These lesions had started 3 years earlier and occurred mainly in the summer months.

Physical Examination

On physical examination, the patient presented polymorphic lesions on the face, ears, and forearms. Vesicles measuring about 2 mm diameter and filled with serous fluid were observed (Figure 1), together with scabs and a large number of depressed scars of vacciniform appearance (Figure 2).

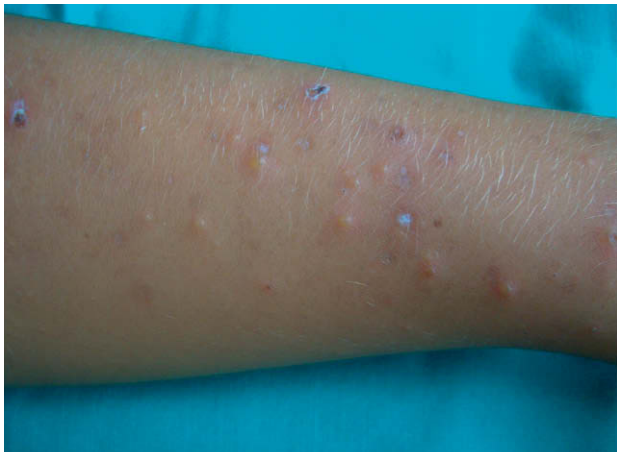

Figure 1.

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Complementary Tests

Blood tests were requested, including a complete blood count, biochemistry, antinuclear antibody, and extractable nuclear antibody, and measurement of porphyrins in blood, feces, and 24-hour urine; all the results were normal.

Histopathology

Biopsy of a vesicle was performed, revealing an intraepidermal blister with a few necrotic keratinocytes and an abundant dermal inflammatory infiltrate (Figure 3). Direct immunofluorescence was negative.

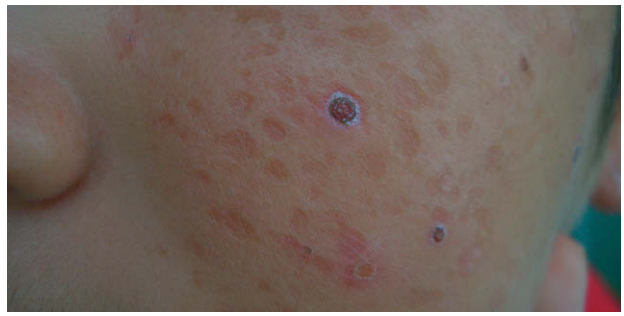

Figure 2.

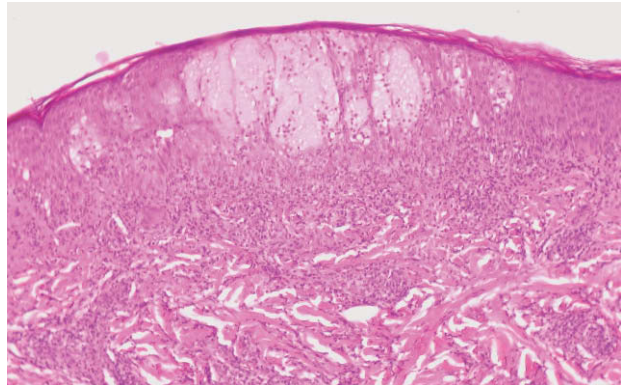

Figure 3. (Hematoxylin-eosin ×100)

What Was the Diagnosis?

Diagnosis

Hydroa vacciniforme

Clinical Course and Treatment

The patient and his parents were informed of the need to protect the skin from sunlight. At the first follow-up visit after diagnosis, the lesions had improved and there were no new lesions even though he was seen during the summer.

Discussion

Hydroa vacciniforme is an acquired, idiopathic photodermatosis described by Bazin in 1862. It is a rare disease, with a prevalence of 0.34 cases per 100 000 population. Onset usually occurs in childhood, with a bimodal peak incidence: one at 2 to 6 years and the other between 12 and 14 years. It is more common in males, in whom the onset occurs later and the disease is of longer duration.[1]

It is characterized by crops of vesicular lesions distributed symmetrically on areas exposed to light, preferentially affecting the face and dorsum of the hands, appearing a few hours after exposure. The vesicles tend to be umbilicated in the centre, progress to a crusting phase, and resolve over a period of 1 to 6 weeks, leaving a round, vacciniform scar. Disease of the mucosas is rare, although cases with oral and ocular involvement have been reported in the literature.[2,3]

This disease usually resolves spontaneously after adolescence, and complications are rare. Severe forms with systemic symptoms and disease in which scarring can lead to reabsorption of the nasal or auricular cartilages, with the consequent deformity, have been reported infrequently.[2]

Histological study is characteristic, revealing an intraepidermal vesicle with a dermal infiltrate of neutrophils and lymphocytes in the initial phases.[7] In more advanced stages, clusters of necrotic keratinocytes are also observed. There have even been reports of signs of vasculitis and panniculitis in severe cases. Direct immunofluorescence is negative, a datum that may be helpful in the differential diagnosis.

The exact etiology of hydroa vacciniforme is unknown. Familial cases have been reported, and so the existence of some genetic factor has been hypothesized.[4] Other authors consider that hydroa represents a scarring variant of polymorphous light eruption. Although the possibility of photosensitivity to UVB radiation has always been considered, the most widely accepted hypothesis on the pathogenesis of this disorder is currently that the causative radiation is of a longer wavelength.[5] This is supported by reappearance of the photo-induced lesions in patients exposed to radiation of 320 to 390 nm; some authors consider the absence of such photo-reproduction to be a good prognostic indicator.[6]

The differential diagnosis includes, on the one hand, other photodermatoses that can develop during childhood, such as erythropoietic protoporphyria, vesicular polymorphic light eruption, actinic prurigo, or bullous lupus erythematosus, and on the other, more common diseases such bullous impetigo and herpes simplex. The clinical history, laboratory findings (porphyrins in urine, blood, and feces, and autoimmune study), and the histology provide sufficient data for the diagnosis to be reached.

Treatment of this disease is difficult.[7,8] The principal element is protection against sunlight by the use of topical sunscreens and suitable clothing. In those cases in which this conservative treatment is not sufficient, systemic agents may be used; PUVA, antimalarials, azathioprine, cyclosporin A, β-carotenes, diets rich in polyunsaturated fats, and thalidomide have been used, all with variable results. In a study performed in 2000, 4 patients underwent treatment with narrowband UVB with encouraging results.[1]

Conflicts of Interest

The authors declare no conflicts of interest.

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