

Case Report

Hydroa Vacciniforme

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Abstract

Observations: Hydroa vacciniforme is rare photodermatitis usually beginning in the earlier years of life. We would like to report a 10-year-old male patient with complaints of depressed scars and blisters on the cheeks and ears. It was noted that the complaints increased in the summer and decreased in winter. Hydroa vacciniforme was diagnosed according to clinical and histopathological findings. The patient was prescribed wet dressings and topical antibiotics as local treatment and was instructed to use high factor sunscreen.

Introduction

Hydroa vacciniforme is rare photodermatitis usually beginning in the earlier years of life. We would like to report a 10-year-old male patient with complaints of depressed scars and blisters on the cheeks and ears.

Case

A 10-year-old male patient came to our clinic with complaints of depressed scars [Figure 1] and blisters with a long history on the cheeks and ears. His complaints had started 2 years ago as blisters on the face and ears. The blisters would later turn into hemorrhagic crusts [Figure 2]. The lesions would not appear elsewhere. He noted redness of the eyes during the summer. In winter, the complaints would resolve with scars. There was no family history for such complaints. Varioliform scars and vesicles on the cheeks, hemorrhagic crusts, vesicles and scars on the ears were observed upon examination. General physical examination was normal. Ophthalmological examination was normal. There was no

fluorescence of the urine or teeth with Wood's lamp. Blood erythrocyte porphyrin levels, urinary



Figure 1. Depressed scars on the patient's cheek



Figure 2. Blisters and hemorrhagic crusts on his ears

and fecal protoporphyrin, coproporphyrin, uroporphyrin levels were in normal ranges.

The lesions were biopsied and the results were reported as:

Throughout the whole epidermis; Full-thickness necrosis (reticular degeneration), exudate consisting of plasma, neutrophil polymorphonuclear cells and erythrocytes; *in the upper and partially mid-dermis;* necrotic appearance of the skin appendages, greatly increased fibroblastic activity, fibrinoid material in the walls and lumens of a few vessels, extravasated erythrocytes, *in the whole dermis;* presence of perivascular and interstitial inflammatory cells.

There was no finding of porphyria with routine stains and PAS staining.

Discussion

Hydroa vacciniforme is a rare, sporadic and idiopathic photodermatitis characterized by formation of vesicles and crusts after contact with sunlight that resolve with varioliform scars [1, 2, 3]. Symptoms usually begin in the earlier years of life. It is seen more frequently in girls [4]. Familial inheritance is rare [5]. The lesions are located especially on areas with more sun contact such as the ears, cheeks and nose [4]. Symptoms such as erythema and edema start hours after exposure to sunlight, symmetrically, and become vesicles [5]. Vesicles become hemorrhagic with time and turn into hemorrhagic crusts after a few weeks. These lesions resolve with varioliform scars. In severe cases there may be loss of tissue on the nose, ears and tips of the fingers [4]. Systemic symptoms such as headaches, fever and malaise may accompany severe cases [4, 5]. Eye involvement manifesting as conjunctivitis and keratitis may be observed. There may be scar formation on the

cornea, leading to visual disturbances. The lesions recur every spring until adolescence when they decrease in frequency [4]. Association with EBV infection has been reported in recent papers but some of these cases are associated with lymphoma and not classical cases of hydroa vacciniforme [6].

Differential diagnosis should include erythropoietic protoporphyria and congenital erythropoietic porphyria. Both these diseases are characterized by early development of photosensitivity and scars. Porphyrin metabolism is normal in hydroa vacciniforme [4]. Typical lesions may occur with photo-provocation tests. Photo-tests in the UVA ranges are abnormal [6].

Histopathological appearance is characteristic and diagnostic [7]. Immunofluorescent findings are nonspecific [5, 7]. Following epidermal spongiosis, local intraepidermal vesiculation, reticular keratinocyte degeneration, epidermal and upper dermal necrosis and sometimes ulceration may be seen in the early phase [7]. There is perivascular mononuclear cell and neutrophil infiltration in the dermis [5, 7].

The first step of treatment should be avoiding exposure to ultraviolet light. Appropriate clothing and especially high sun protection factor, wide spectrum sunscreens may help [7]. In resistant cases, prophylactic low dose, wide and sometimes narrow band UVB or PUVA may help [5, 7]. Antimalarials have been proposed but found to be insufficient [7]. Hydroxychloroquine may reduce symptoms and decrease sensitivity to UVA.⁵ A case treated with chloroquine and topical sunscreens has been reported [8]. Immunosuppressive treatment with intermittent oral steroids may be used [7]. There have been cases reportedly benefiting from carotenoids [9]. Diet rich in omega-3 unsaturated fatty acids has shown to decrease the erythema and polymorphous light eruption due to UV [10]. The omega-3 unsaturated fatty acids in fish oil decreases local inflammation due to UV exposure. Recent cases of hydroa vacciniforme treated successfully with fish oil diet have been reported [11].

When our patient first came to our clinic in March 2004, hemorrhagic crusts and varioliform scars besides active vesicles were ob-

served. The patient was prescribed wet dressings and topical antibiotics as local treatment and was instructed to use high factor sunscreen. In April, we observed that the lesions of the ears had resolved but new lesions had appeared on the cheeks. Upon questioning, it was found that the patient had not used his sunscreen appropriately. Wet dressings were prescribed again and the importance of sun protection was explained in detail.

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