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Case Report

Hydroa vacciniforme: a very rare photodermatosis

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ABSTRACT

Hydroa Vacciniforme (HV) is a rare, acquired and chronic paediatric disorder that is characterized by photosensitivity and recurrent crops of skin lesions on sun-exposed skin, such as the face, ears and hands that heal with vacciniforme scarring. The pathogenesis of HV is unknown. No chromosome abnormality has been identified so far. HV patients have no abnormal laboratory results. The histopathologic features are distinctive and demonstrate intraepidermal multilocular vesicles and cellular necrosis. Most cases remit spontaneously by late adolescence.

Keywords: Hydroa vacciniforme, Photodermatosis

INTRODUCTION

Hydroa Vacciniforme (HV) is a rare photodermatosis of unknown etiology, which predominantly affects children. Skin and mucous membranes are the primary sites affected by HV.[2] It is characterized by recurrent erythema and crops of vesicles on light-exposed areas, especially the malar areas, bridge of the nose, lips, ears, and the dorsa of the hands and forearms, a few hours after sun exposure. They may be accompanied by a mild keratoconjunctivitis, photophobia, or constitutional symptoms. The vesicles crust and heal over a period of 1-6 weeks, leaving varioliform scars.[2] HV was first described by Bazin in 1862.[3] The condition is seen equally in both sexes. Females have a higher incidence of HV than males and report earlier onset. Males who are affected have a longer course of disease than females. Presentation is usually during the first decade predominantly affects children aged 3-15 years, but there is a late-onset variety. The etiology of HV is not known. HV may be a distinct entity distinguished by scarring or may occur within the spectrum of polymorphous light eruption. No mortality is associated with typical HV.

CASE REPORT

A 13 year old boy came to our OPD with complaints of recurrent multiple papules, vesicles and crusts over his face (Figure 1), arms (Figure 2) and legs (Figure 3) for last 10 years. Symptoms were more profound in the summer months with regression in winters. These skin lesions developed several minutes to hours after sun exposure. Patient also complained of itching on the lesions. The lesions usually started over the face and then gradually progressed to involve the arms and legs. Patient stated a feeling of generalized weakness during fresh crops of eruptions. Each vesicle subsequently ruptured within 1 or 2 days, became crusted, and then gradually

healed with black scabs, leaving behind pale depressed scars. No photophobia. No constitutional symptoms associated. Physical examination showed crops of oedematous papules and clear filled discrete vesicles, few cloudy vesicles, surrounded by an erythematous halo. These lesions were interspersed with necrotic papules on an erythematous base along with crusting of lesions on the cheek and nose, which healed with hypopigmented depressed scars. Similar lesions with atrophic scars were also present on the elbow and dorsa of hands. Teeth, nails and eye[4] examination were unremarkable.

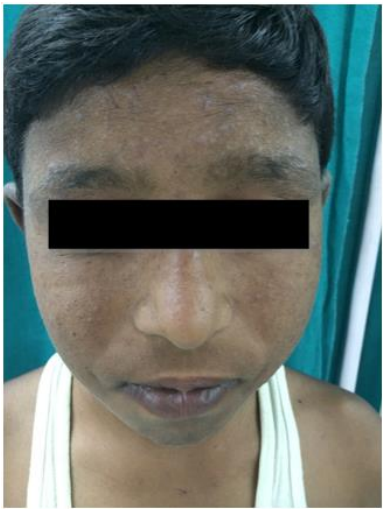

Figure 1: Papulo-vesicles with crusting and scarring.

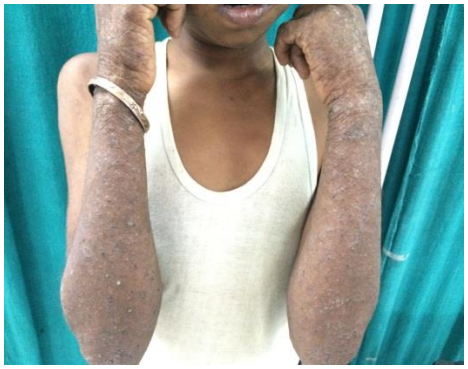

Figure 2: Papulo-vesicles with necrotic base over arms.

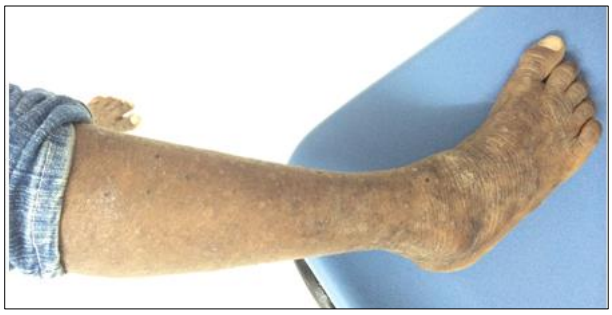

Figure 3: Papules & vesicles with scarring over right leg.

Investigations

Complete blood counts, ESR, liver and renal function tests, red blood cell porphyrin levels, 24-hour fecal porphyrin levels and 24-hour uroporphyrin levels and antinuclear antibody levels were within normal limits. Skin Biopsy revealed an epidermal, multilocular vesicle and epidermal necrosis dense, perivascular, lymphohistiocytic infiltrate (Figure 4). To rule out Epstein-Barr virus infection, immunostaining for EBV - determined nuclear antigens (EBNAs) and Latent Membrane Proteins (LMPs) was done. There was no evidence of EBV in the skin biopsy sample provided. Historically, clinically and with supportive histopathogial evidence, the case was diagnosed as hydroa vacciniforme. Patient was instructed to strictly avoid sun with full sleeves light colour clothing and frequent application of sunscreen with a high SPF. Patient was given tab hydroxychloroquin 250 mg twice a day. He was counselled about the disability, its course and resolution usually by adolescence.

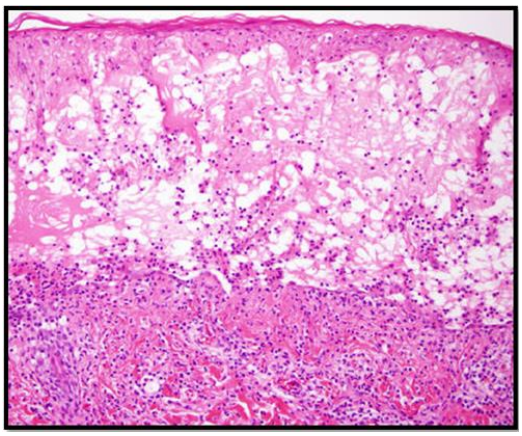

Figure 4: H & E (200x) showing epidermal, multilocular vesicles, perivascular, lymphohistiocytic infiltrate.

DISCUSSION

HV, initially described by Bazin in 1862, is a very rare photodermatosis of unknown etiology that principally starts in childhood.[5] It has several distinctive features, including the (1) uniform development of vesicles and crusts several hours to 1 or 2 days after sun exposure, (2) healing of lesions with varioliform scarring, (3) absence of laboratory abnormalities, including serologic and porphyrin studies, (4) characteristic histopathology with epidermal necrosis and intraepidermal vesiculation, and (5) demonstrable evocation of the typical lesions by exposure to light.[6] The development of lesions and their distribution suggest a causal relationship between HV and ultraviolet (UV) exposure, although the pathogenetic mechanism remains unknown.[7] Some reports have recently demonstrated that HV is associated with EBV infection and lymphoma, but some of these cases showed

atypical features, and these cases may not represent the usual form of HV.[8]

HV was at first infrequently diagnosed because of the terminological confusion and uncertainty concerning the role of porphyrin metabolism in its pathogenesis.[9,10] At that point in time, some of the cases classified as HV had been protoporphyria until erythropoietic protoporphyria (EPP) was defined clearly.[5]

The differential diagnosis of HV consists of several blistering disorders that are light induced, including EPP, vesicular polymorphous light eruption (PMLE), bullous lupus erythematosus, solar urticaria, hydroa aestivale and Porphyria Cutanea Tarda (PCT).[5,6] Clinicians can distinguish between these different illnesses in most cases by obtaining detailed historical, clinical, histopathologic and laboratory data. The clinical presentation of vesicular form of PMLE may be similar to HV but unlike HV, the lesions of PMLE almost always heal without scarring,[11] and the histological features differ from those of HV. The distinctive histologic changes of HV include initial intraepidermal vesicle formation with later focal epidermal keratinocyte necrosis and spongiosis in association with dermal perivascular neutrophil and lymphocyte infiltration. In vesicular PMLE, there is subepidermal vesicle formation, interspersed with mild to moderate epidermal spongiosis and there is no evidence of epidermal necrosis.[12]

Hydroa aestivale is considered by some investigators to be a childhood type of PMLE, while other researchers have postulated that it is a non-scarring form of HV.[13,14] The eruptions in EPP are typically an intensely edematous, urticarial reaction, and only its more severe purpuric and vesicular forms cause scarring. The histological features of EPP are deposition of a hyaline substance around the upper papillary blood vessels after repeated injury.[15] The urine, blood and stool porphyria laboratory results can help to exclude EPP and PCT. Bullous lupus erythematous can be differentiated by a positive serologic profile and characteristic histology. The ability to reproduce the typical lesions in HV patients by phototesting has made this procedure a valuable diagnostic aid. In 1960, Schiff and Jillson were the first to document induction of lesions by phototesting that were clinically identical to those of HV, and phototesting with repetitive irradiation using a large dose of UVA has recently been shown to be very important for confirming the diagnosis of HV. Interestingly, our case showed that skin lesions were induced even with the MED of UVA, not with repetitive irradiation using a large dose of UVA.

Two reports of HV in siblings have been documented, suggesting a genetic component to HV.[16]

To date, no oral therapy reliably prevents the appearance of HV lesions. Oral antimalarials[17] and beta-carotene[18-20] are most commonly used and are occasionally useful, especially when combined with a strict sun avoidance program. Other therapies that have been used with varying success include thalidomide, azathioprine, cyclosporine,[21] and fish oil supplementation.[22-24]

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