Varioliform Scarring in a Young Adult- Revisiting Hydroavacciniforme

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ABSTRACT
Hydroa Vacciniforme (HV) is a rare, idiopathic acquired photodermatosis, with onset in childhood characterised by recurrent crops of vesiculopapular eruption that heal with varioliform scarring. The pathogenesis of HV remains unknown. Herein, we presented a case to highlight the importance of this condition as a mimicker of other photodermatosis.

INTRODUCTION

Hydroa Vacciniforme is a rare idiopathic photodermatosis, predominantly affects children between 3 and 15 years.[1] The exact incidence of HV is not known. It was first described by Bazin in 1862. Both sexes are affected equally with males usually have a longer disease course than females.[2] It is characterized by crops of vesiculopapular eruption on sun exposed areas after few hours of sun exposure. The vesicles then crust and heal over a period of 1-6 weeks, leaving behind depressed varioliform scars.[3] They may be accompanied by a mild keratoconjunctivitis, photophobia, or constitutional symptoms.[4] The histopathological findings of HV were distinct with no laboratory abnormalities.

CASE REPORT

A 23 year old male patient came to our outpatient department with complaints of depressed scars over his face, hands and foot for a long history. His complaints had started at the age of 4 years as blisters on the face and ears, and then progressed to involve dorsum of hands and legs. The blisters later turned into hemorrhagic crusts and healed with varioliform scars. Patient also complained of mild burning on exposure to sunlight with aggravation of symptoms in summer months and regression in winter. There was no family history for such complaints. No constitutional symptoms were associated. Physical examination showed multiple hyper pigmented varioliform scars present over bilateral cheeks, ears (over anti helix), nose, lower lip, dorsal aspect of both the hands and foot (fig. 1). Dystrophic nails were present in all the digits and toes (fig. 2). Teeth and eye examination were unremarkable.

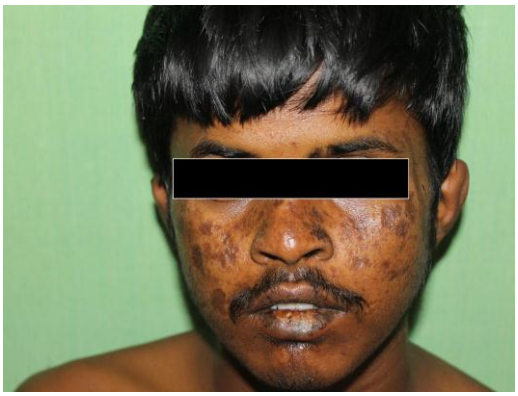

Fig. 1: Multiple depressed varioliform scars over both cheeks, nose and lower lips

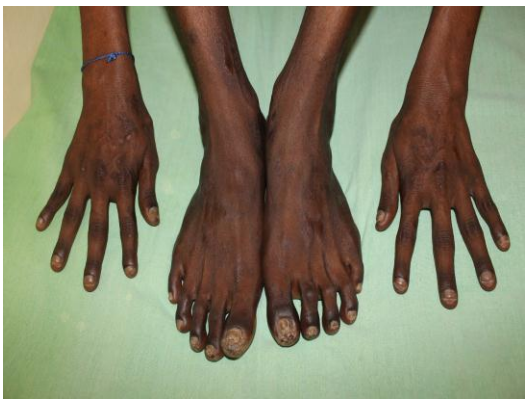

Fig. 2: Multiple scars over dorsum of both hands and foot with dystrophic nails

INVESTIGATIONS

Laboratory investigations including complete blood cell count, ESR, liver and renal function tests, red blood cell porphyrin levels, 24-hour fecal porphyrin levels, 24-hour uroporphyrin levels and serum antinuclear antibody levels were all within normal limits. Histopathology of the skin biopsy revealed an atrophic epidermis, dense collagen and elastic fibres in the dermis; and loss of capillaries and skin appendages (fig. 3). There was no evidence of porphyria with routine

stains. Based on detailed history, clinical examination and histopathological findings, the case was diagnosed as hydroa vacciniforme.

The patient was prescribed a topical sunscreen with oral beta carotene and told to have a strict sun protection with full sleeves clothing and wide brim hat. He was counselled about the nature of disease, disability, its course and resolution.

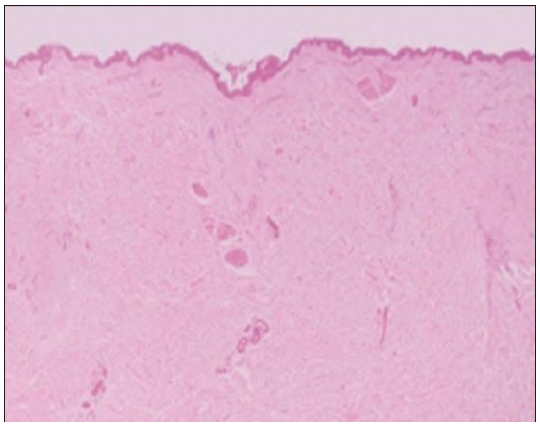

Fig. 3: H & E stain (40X): Showing atrophic epidermis, dense collagen and elastic fibres in the dermis with loss of appendageal structures

DISCUSSION

Hydroa vacciniforme is typically a disease of childhood but it may extend its spectrum to adulthood. The following diagnostic criteria are used to distinct HV from other photodermatoses, (1) development of vesiculopapular eruptions after sun exposure (usually 30mins to 2 hrs after sun exposure), (2) healing of lesions with varioliform scarring, (3) no laboratory abnormalities including serologic and porphyrin studies, (4) characteristic histopathologic features of reticulate epidermal necrosis and intra epidermal vesiculation in the acute stage of disease, (5) exclusion of other photodermatoses.[5] Systemic symptoms are usually not observed in HV. The exact mechanism by which ultra violet (UV) rays induce vesiculopustular eruption is still not known.[6] Some reports have recently demonstrated that severe HV is associated with Epstein-Bar virus (EBV) infection.[7]

The histological hallmark of HV is epidermal spongiosis, intra epidermal vesiculation, reticular keratinocyte degeneration followed by epidermal necrosis with dense lymphocytic and neutrophilic dermal infiltrate.[8]

Differential diagnosis includes polymorphic light eruption (PMLE), Erythropoietic Protoporphyria (EPP), bullous lupus erythematosus, porphyria cutanea tarda, hydroa aestivale, HV-like lymphoma (HVLL), herpes simplex eruption, and varicella.[5] Vesicular PMLE usually have a similar presentation as HV, but heals without scarring.[9] Histopathologically, epidermal necrosis which is typical of HV is lacking in PMLE. Urine, blood and stool porphyrin levels helps to rule out EPP.[10] Bullous lupus erythematosus can be differentiated by a positive serologic profile. HV-like lymphoma (HVLL) is a cutaneous T-cell lymphoma characterized by recurrent papulovesicles predominantly in the sun-exposed areas, often accompanied by fever, lymphadenopathy, hepatosplenomegaly and increased liver enzymes.[11] HVLL is distinct from HV by its severity, presence of systemic symptoms and involvement of sun protected areas.

Treatment options for HV are limited. Sun avoidance and use of broad-spectrum sunscreens have been reported to diminish and sometimes prevent skin eruptions. Systemic therapy, including anti-malarials, ß-carotene, azathioprine, cyclosporine, and fish oil supplementation have been attempted with limited success.[12-17]

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