Adult-onset Hydroa Vacciniforme: A Rare Occurrence or a Lymphoma Premonition?

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Sir,

Hydroa vacciniforme (HV) is a photodermatosis affecting children and adolescents, with papulovesicular eruptions and varioliform scars on face and other photoexposed parts.[1]

Classically, the presentation is during childhood, though very rarely late onset cases have been described.[2] A subset of cutaneous T cell lymphoma, designated as HV-like lymphoma (HVLL) has also been described, which closely mimics HV.[3]

A 42-year-old male patient, farmer by occupation, reported to dermatology outpatient department with history of severe burning sensation and swelling limited to face and scalp on sun exposure. This was followed by painful skin eruptions, which further progressed to become fluid filled within 3 to 5 days, followed by brown coloured scab formation and eventually depressed scars. The patient has been experiencing similar recurrent episodes since last 8 years, from February to August every year. The patient reported mild malaise and lethargy during vesicular phase of illness, but no other symptoms like fever or chills, burning in eyes, arthralgia, weight loss, easy fatiguability, bruising tendency, abdominal discomfort, or features suggestive of lymphoproliferative etiology. There was no family history of similar illness and patient denied intake of any photosensitizing drug.

On cutaneous examination, few erythematous papulovesicles 0.2 cm to 0.4 cm in size with overlying crust were noted on right cheek [Figure 1]. Both cheeks and scalp also showed multiple brown to black-colored scabs, 0.5 cm to 1.2 cm in size [Figure 2]. Multiple well-defined, depressed, and varioliform scars ranging from 0.2 cm to 1.5 cm [Figure 1] were present on forehead, cheeks, nose, and scalp. No cervical, axillary or inguinal lymphadenopathy, and hepatosplenomegaly was noted. Cytological smears and blood samples were negative for human immunodeficiency virus (HIV), Epstein-Barr virus (EBV), and herpes simplex virus (HSV). Urine samples showed no fluorescence on Wood's lamp examination and urinary and fecal porphyrin levels were within normal limits. The hemogram, peripheral blood smear, ultrasonography of abdomen and lymph nodes revealed no abnormality. A provisional diagnosis of discoid lupus erythematosus (DLE), adult-onset HV, and HVLL was arrived at and a lesional skin biopsy was obtained. Biopsy revealed intracellular edema, basal cell vacoulization, and areas of necrosis in epidermis along with mixed neutrophilic and lymphocytic dermal infiltrate [Figure 3]. There were no atypical lymphocytes or viral

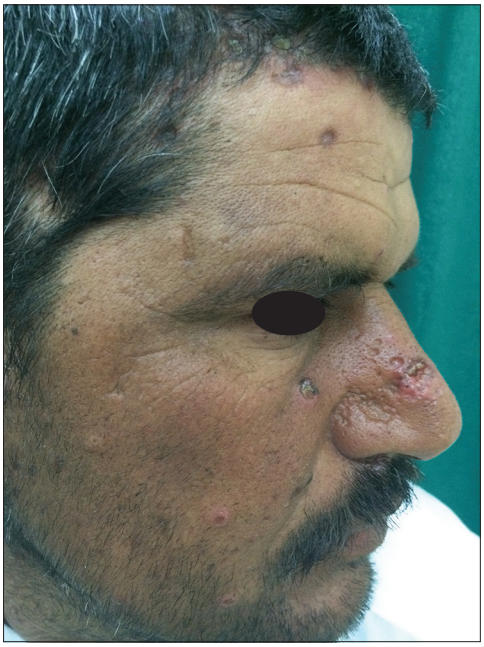

Figure 1: Erythematous papulovesicles and multiple depressed varioliform scars of varying size on right cheek

How to cite this article: Sangwan A, Aggarwal K, Kaur S, Jain VK. Adult-onset hydroa vacciniforme: A rare occurrence or a lymphoma premonition?. Indian Dermatol Online J 2017;8:490-1.

Received: October, 2016. **Accepted:** June, 2017.

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Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ_286_16

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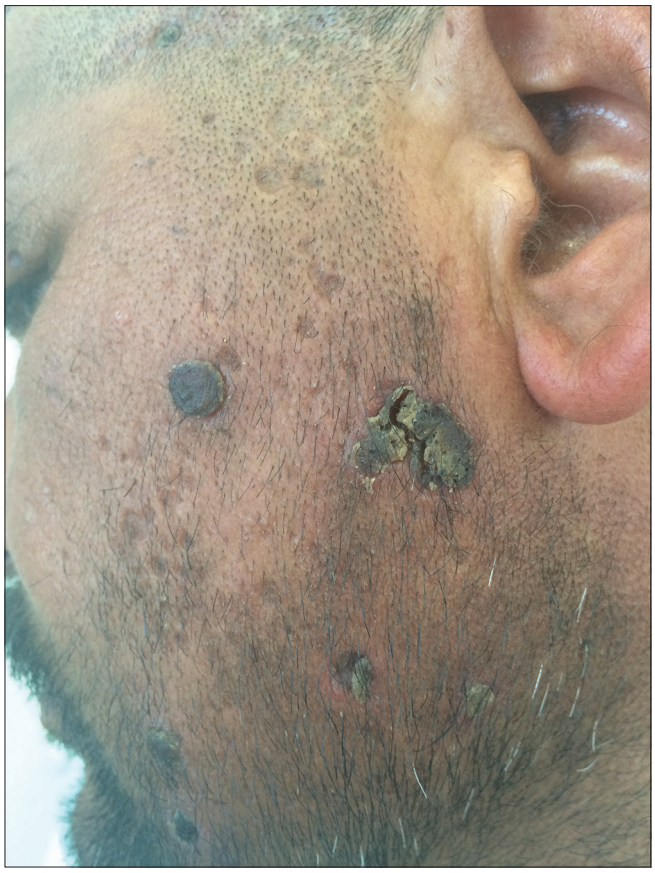

Figure 2: Brown-colored scabs with multiple varioliform scars on left cheek

inclusion bodies. The immunohistochemical (IHC) studies were negative for T-cell lymphoma (CD8 and CD56). These findings were consistent with the primary diagnosis of HV. Immunofluoresence for antinuclear antibody (ANA) was negative.

The patient was advised strict photoprotection and oral hydroxychloroquine (400 mg/day) with beta-carotene supplements. On follow-up visits at 3 and 6 weeks, patient was experiencing lesser number of episodes with decreased clinical severity.

The original description of HV has been credited to Bazin, though it was confused with erythropoietic protoporphyria in those reports.[4]

The disease typically affects children and adolescents as a phototoxic rash and remits by early adulthood. Typically exacerbations are noted in spring to summer season due to higher proportion of UVA rays reaching earth during this time of year.

HV is sometimes confused with HVLL due to similarities in clinical presentation and age group affected. However, HVLL is typically associated with EBV infection, higher clinical severity and disfigurement, involvement of non-photoexposed parts, e.g., scalp and trunk, as well as systemic complications.[5] Unlike HV, HVLL lesions are not induced by sun exposure and do not tend to resolve with age. Involvement of scalp in our patient did not correlate with HV because dense scalp hair normally provides considerable photoprotection, hence creating a suspicion of HVLL. This; however, could possibly be due to the occupation of patient which mandates prolonged sun exposure in open spaces, hence increasing the dose of photoexposure and clinical severity.

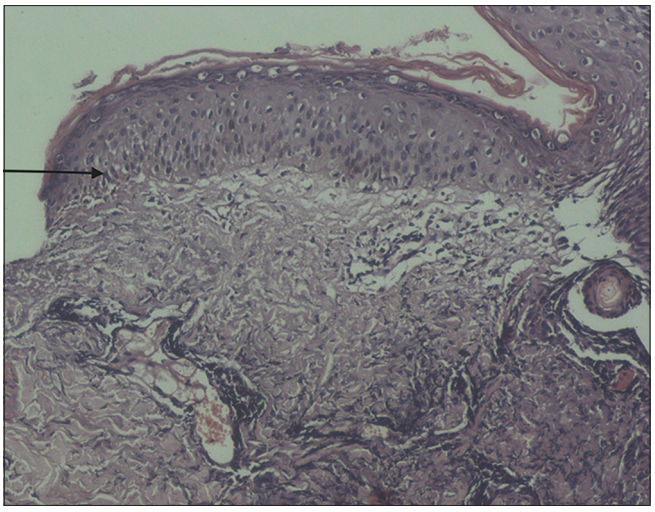

Figure 3: Hematoxylin and eosin (H and E) stain of biopsy from papulovesicle showing epidermal edema, basal and suprabasal cell vacuolization and keratinocyte necrosis (depicted by black arrow) (×10)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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