An Adult Case of Hydroa Vacciniforme

Reza Mahmoud Robati, MD
Marjan Saeedi, MD
Nima Sarrafi-rad, MD

Skin Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Corresponding author:
Reza Mahmoud Robati, MD
Skin Research Center, Shohada-e Tajrish Hospital,Shahid Beheshti University of Medical Sciences, Tehran, Iran
Email: rmrobati@gmail.com

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Abstract

Hydroa vacciniforme (HV) is a rare acquired photodermatosis, usually with onset in childhood and characterized by vesicles, crust and scar formation that follow exposure to sunlight.Vacciniform scars of face and dorsa of the hands are common features but oral ulcer and eye signs also rarely occur. It usually resolves before adult age. A rare manifestation of the disease would be persistence until adult age which is presented in this report. *(Iran J Dermatol 2009;12:64-66)*

Keywords: adult, hydroa vacciniforme, photodermatosis

Case Report

A 25-year-old man presented with a 15-year history of a recurrent, mildly pruritic, blistering eruptions affecting his nose and forehead which occurred throughout the year but were more severe in the summer. The eruptions began as small papules, which were later transformed into vesicles a few hours after exposure to sun. Some lesions developed several days after sun exposure. They burst within 24 hours, then became crusted over and eventually healed with fine varioliform scars. The eruption could be brought on by sun exposure directly or through window glass. The patient was on no medication and had no known contact allergies. He was otherwise well with no constitutional symptoms. He did not complain of photophobia and lacrimation after sun exposure. There was no known exposure to photosensitizers, and the family history revealed no photosensitivity diseases. Moreover, there was no history of similar cutaneous eruptions in the patient's family.

On physical examination, his nose had a burn-like scar and some crusts (Figure 1). No intact vesicles were seen but some erosion with crust formation were present on his nose. The crusts healed at times leaving scarring. Ear lesions especially on the mid third of helix area were also found with erosion and crusting on the upper surface (Figure 2).

The clinical laboratory tests including complete blood count, urinalysis, liver function tests, plasma and urine porphyrin, antinuclear antibody, chest X-ray and photopatch test reported negative or within normal limits. Direct immunoflorecense test was reported negative.

Biopsy specimens from the nose lesion showed focal epidermal necrosis, epidermal edema and a lymphocytic and neutrophilic infiltrates under the

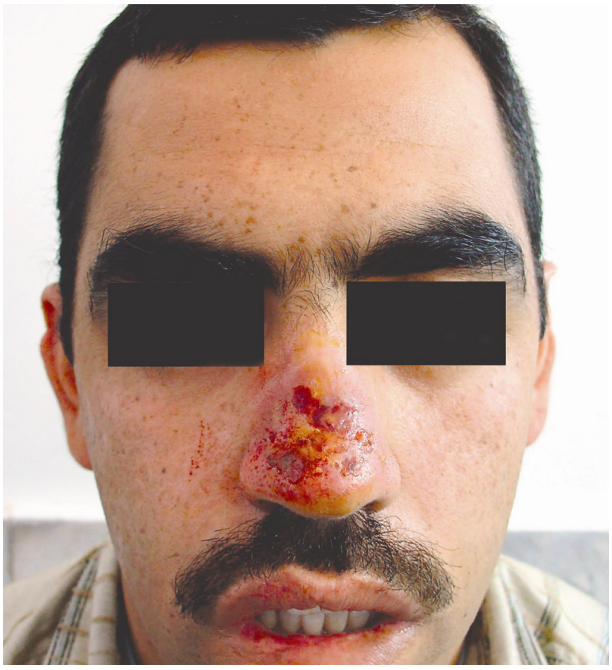

Figure 1. *Erosion, crusting and scar on the nose.*

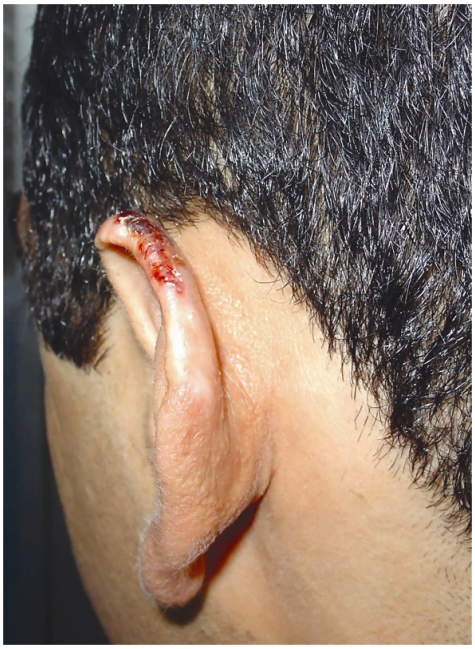

Figure 2. *Ear lesions on the mid third of helix area with erosion and crusting on the upper surface.*

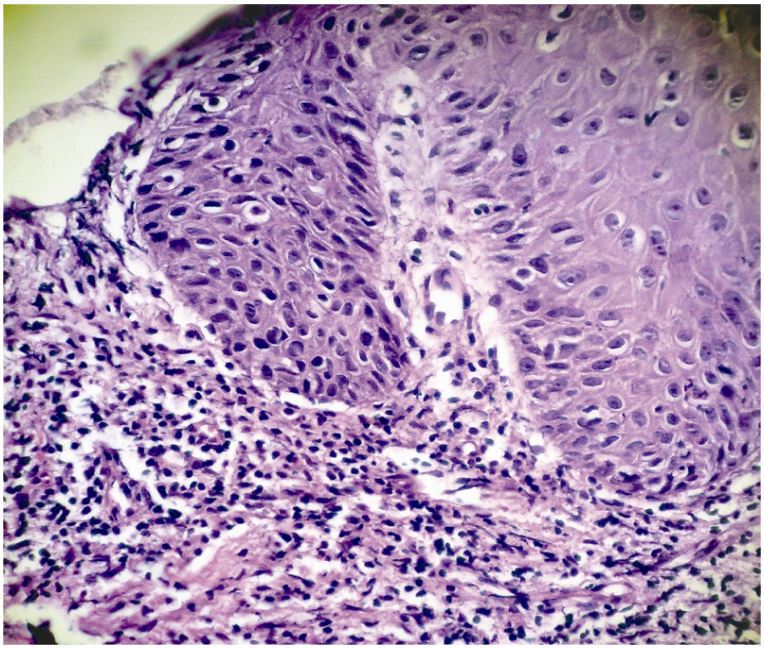

Figure 3. *Histopathologic view; focal epidermal necrosis , epidermal oedema and a lymphocytic and neutrophilic infiltrate under necrotic area. (H&E *40).*

necrotic area. Superficial and deep perivascular lymphocytes with a few neutrophil and eosinophil infiltrates were also observed. Periodic Acid Shift (PAS) staining was negative (Figure3).

The case was diagnosed as hydroa vacciniforme (HV) regarding these clinical and histopathological data.

Discussion

Hydroa vacciniforme (HV) is a rare, usually sporadic but occasionally familial condition occurring in both genders. The origin of this condition is unknown. Spring and summer sun exposure, particularly to UVA wavelength, appears to be responsible. Presentation is usually during the first decade. About 12 to 24 hours after exposure to direct or window glass transmitted sunlight, pruritic, sometimes haemorrhagic vesicles and papules arise on an erythematous background, typically affecting the cheeks, ears, nose and hands[1,2]. Early lesion histological changes include intraepidermal vesicle formation with later focal epidermal keratinocyte necrosis and spongiosis in association with dermal perivascular neutrophil and lymphocyte infiltration. Older lesions show necrosis, ulceration and scarring [3].

Complications of HV are rare. Eye involvement manifests as conjunctivitis, sometimes associated with severe chemosis. Corneal ulcerations have been described in a few cases and occur on the exposed areas of the cornea while the upper photoprotected surface spared [4]. Scarring of skin lesions can result in hand deformation including flexor deformities and malposition of the first, second, or third interphalangeal joints of the hands. Partial bone absorption of the fingers has also been noted [5]. Cicatricial contracture of the lower lip with incisor extrusion has been reported[6] . Ear mutilation seems to be one of the rare complications of hydroa vacciniforme that has been reported only in few cases [5,7].

Apart from usual manifestations, several rare clinical presentations have been reported. Late onset HV has been reported in two men during compulsory military service from Singapore. [8] Although the disease usually resolves in adult age, persistence of the disease until 20 and 60 have also been reported [9,10].

Our patient presented with some remarkable manifestations of HV including persistence of the disease up to the third decade of life and burn-like scars on the nose or ear mutilation. Although hydroa vaccinifirme usually manifests in childhood with few complications, we recommend including this entity in the differential diagnosis of unusual cutaneous scars or mutilations with erosion and crusting in sun exposed areas of the body especially in adults.

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