Hydroa Vacciniforme

—Recurrence at Adulthood and Confirmative Diagnosis by Repetitive Ultraviolet-A Phototesting—

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Hydroa vacciniforme is a very rare photosensitivity disorder. The primary skin lesion is a vesicle or bulla which then heals with vacciniform scarring. We report a case of hydroa vacciniforme recurred after 3 years period of quiescence in a 20 year-old man who had had history of the disease from the age of two. The duplication of the natural lesion, clinically and histologically, was successfully made by artificial UV-A irradiation on the patient's back.
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Key Words: Hydroa vacciniforme, Phototest

Hydroa vacciniforme is a rare, chronic photosensitivity disorder that is characterized by recurrent, discrete vesicles on sunlight exposed areas that heal with vacciniform scarring. Initially, it is manifested clinically by the development of itching erythema progressing to painful papules and vesicles within several hours to one or two days on sun exposed areas. The histologic finding of the vesicle is distinctive and is characterized by intraepidermal reticular degeneration and cellular necrosis. It usually begins in childhood and, in most instances, involutes spontaneously by the late teenage years and is not associated with other systemic disease.[1]

We describe a case of hydroa vacciniforme which recurred after summer military training in a young soldier who had had a 15 year history of the disease until 3 years ago and the vesicles were reproduced by repetitive UV-A exposure.

REPORT OF A CASE

A 20-year-old soldier was referred to the Depart-

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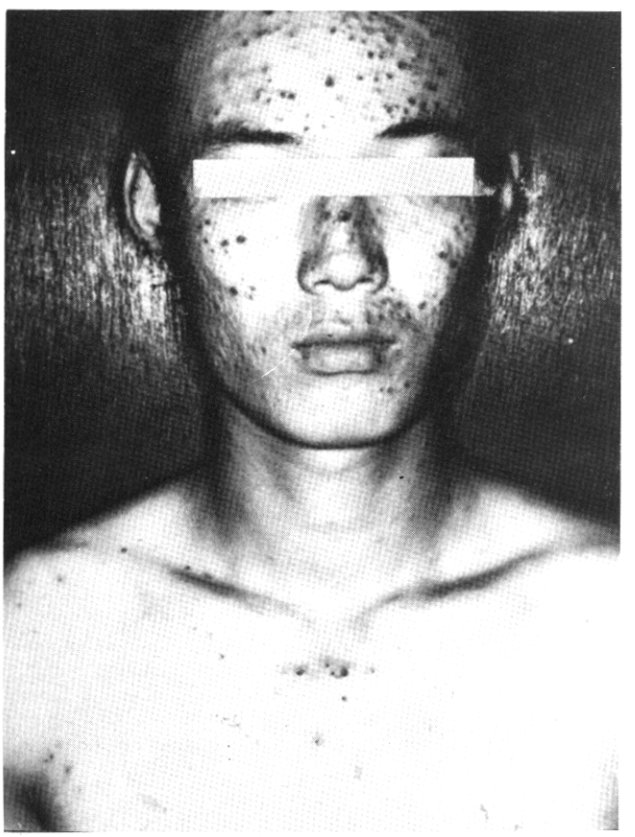

Fig. 1. Diffuse, uniform, hypopigmented, deep and umbilicated scarring on the face and V-area of the neck.

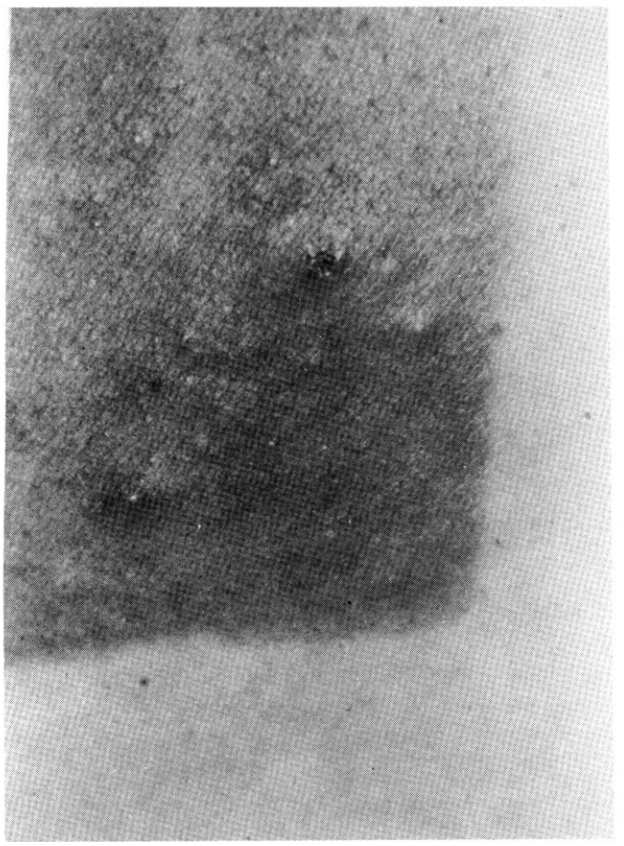

Fig. 2. The UV-A induced vesiculation and papular eruptions resemble the sun-induced lesions.

ment of Dermatology, Capital Armed Forces General Hospital in September 1988. He had a fifteen year history of recurrent, scattered, discrete vesicular lesions followed by characteristic scarring on sun-exposed areas during summer. For the last three years, he had been relatively well until vesicles developed on his face, dorsa of the hands, extensor surfaces of the forearms and V-area of the neck after summer military training (Fig. 1). There was no known exposure to photosensitizers or familial history of a photosensitivity disorder. On physical examination, there were diffuse, uniform, hypopigmented, deep and umbilicated scars on the face, ears, dorsa of the hands, extensor surfaces of the forearms and V-area of the neck but vesicles were not present.

Results of laboratory studies were within normal limits, including complete blood cell count with differential cell count, platelet count, erythrocyte sedimentation rate, electrolytes, liver and renal function test, serum uric acid, serum iron, total iron-binding capacity, lupus erythematosus cell preparation, antinuclear antibody concentration, chest roentgenogram, rheumatoid factor, VDRL, urinalysis and qualitative tests for blood, urinary and fecal porphyrin.

Phototest

Phototesting for the minimal erythemal dose

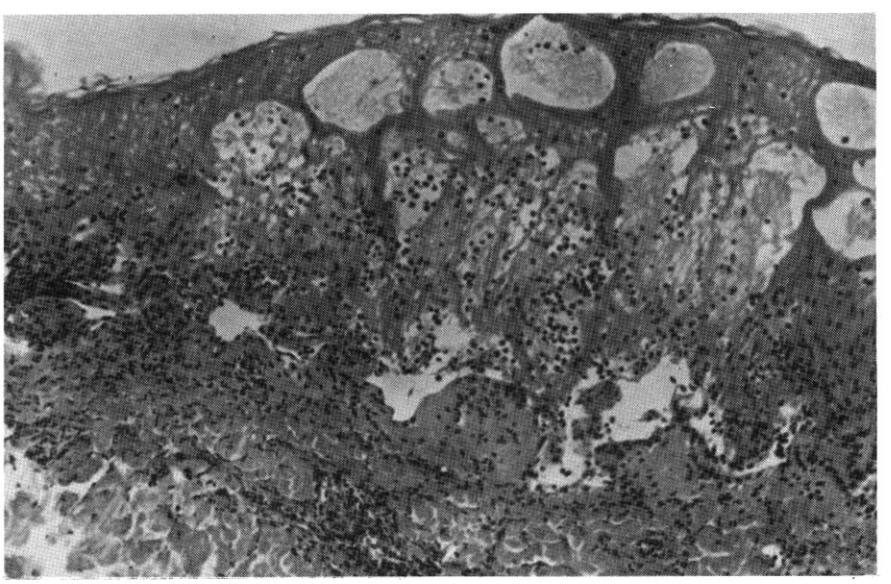

Fig. 3. The microscopic findings of an UV-A induced vesicle show intraepidermal vesiculation and diffuse epidermal necrosis (H & E stain, ×40).

(MED) of ultraviolet light in the B range (UV-B) was performed with a bank of fluorescent lamps (Elder Co. FS T 12-UVB-HO lamp) and with a strong UV-A lamp (Sellas Sunlight, Dr Sellmeier Co.) for the UV-A MED. The irradiance of the light sources was measured with appropriate radiometers and probes (International Light Inc). A UV-B MED of 30 to 40 mJ/cm^2 and a UV-A MED of 30 to 40 J/cm^2, both within normal limits for the patient's skin type, were demonstrated on the patient's back. There was no induction of skin lesions on the MED test sites.

To reproduce lesions on his back, the patient initially received 60 mJ/cm^2 of UV-B. An additional 60 mJ/cm^2 of UV-B was given to the initial exposure site within 24 hours and then again within 48 hours. Two sites on his back were irradiated with UV-A. The patient received 100J/cm^2 and 200J/cm^2 on each site initially and then again within 24 hours. The UV-B irradiated site developed only erythema, even after multiple exposures. Although the UV-A irradiated sites did not have a significant reaction after a single irradiation, repetitive irradiation to the same site caused a papular eruption and vesiculation accompanied by pruritus, in addition to erythema and mild hyperpigmentation (Fig. 2). The UV-A induced vesiculation resembled the sun-induced lesions and healed with scarring similar to that produced by sunlight. The histopathologic findings of UV-A induced vesicles showed intraepidermal vesiculation and epidermal necrosis (Fig. 3) which are typical findings of hydroa vacciniforme.

DISCUSSION

Hydroa vacciniforme described by Bazin[2] was at first infrequently diagnosed because of terminological confusion and uncertainty concerning the role of porphyrin metabolism in its pathogenesis. It is a rare, debilitating photodermatosis of children with unknown etiology.[3] In most instances, it involutes spontaneously by the late teenage years, but our patient's skin lesions recurred after summer military training, at 20 years of age. Interestingly, the recurred age of our patient was the adulthood when hydroa vacciniforme usually remits. At present, hydroa vacciniforme has several distinctive features, including 1) uniform development of vesicles and crusts several hours to one to two days after sun exposure; 2) healing of these lesions with vacciniform scarring; 3) absence of laboratory abnormalities, including serologic and porphyrin studies; 4) characteristic histopathology with epidermal necrosis and intraepidermal vesiculation; and 5) demonstrable evocation by exposure to light.[4] Our case also had the above distinctive features. There has been a suggestion of familial incidence in the literature,[5] but this has not been further substantiated. Hydroa vacciniforme has also been reported in association with Hartnup disease.[6] However, our patient did not have any familial history and associated disease.

The differential diagnosis of hydroa vacciniforme consists of several blistering disorders that are light-induced, including erythropoietic protoporphyria, vesicular polymorphous light eruption, bullous lupus erythematosus, and hydroa aestivale.[4]

Hydroa vacciniforme can be differentiated from erythropoietic protoporphyria by the latter's earlier age of onset of photosensitivity with more intense itching and burning occurring within minutes after sun exposure. The eruption in erythropoietic protoporphyria is typically an intensely edematous, urticarial reaction that only in its more severe pur-

Table 1. Reported cases of hydroa vacciniforme confirmed by phototesting

Source	Phototesting	Respones to Phototesting
Schiff and Jillson[10]	Carbon arc;Cold quartz	Erythema, papules, vesicles, no scarring
Ramsay[11]	300 nm	Papules, possible scarring
Bickers et al [1]	280-420 nm	Erythema, vesicles, no scarring
Bickers et al [1]	Cold quartz	Erythema, no scarring
Jaschke and Hönigsmann.[12]	Repetitive UV-A	Erythema, vesicles, scarring
Goldgeier et al [7]	Repetitive UV-A	Erythema, vesicles, scarring
Halasz et al [8]	Repetitive UV-A	Erythema, vesicles, scarring
Eramo et al [4]	Repetitive UV-A	Erythema, vesicles, scarring
Present case	Repetitive UV-A	Erythema, vesicles, scarring

puric and vesicular forms causes scarring. Microscopically, hydroa vacciniforme shows focal epidermal necrosis whereas erythropoietic protoporphyria shows hyaline substance deposition around upper papillary blood vessels after repeated injury.[7] Polymorphous light eruption usually has a somewhat later onset than does hydroa vacciniforme and may variably present papules, eczematous infiltrated plaque, excoriations, and lichenification. Vesiculation develops only occasionally and these vesicles arise from dermal edema with subepidermal bulla formation, and spongiosis is exceptional. Bullous lupus erythematosus can be differentiated by positive serologic profiles and a characteristic skin biopsy histology.[8] Hydroa aestivale is also a vesicular and bullous disease that tends to recur each summer during childhood on skin areas exposed to sunlight, but it does not produce scars.[9]

The ability to reproduce hydroa vacciniforme lesions in the patient by phototesting has made this procedure a valuable diagnostic aid. Recently, phototesting with repetitive irradiation of a large dose of UV-A has been revealed to be very important in confirming the diagnosis of hydroa vacciniforme as shown in Table 1.

We were also consistently able to produce vesicles with 200 J/cm^2 doses. UV-B was ineffective in producing vesicles, even after 5 MED were irradiated in repeated doses.

In summary, our case of hydroa vacciniforme recurred during adulthood when it usually remits and its diagnosis was confirmed by historical, clinical, histopathologic, and laboratory data and especially repetitive UV-A phototesting.

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