Hydroa Vacciniforme: Diagnosis by Repetitive UVA Phototesting

Wandandorj Narantsetseg[1], M.D., Hae-Shin Chung[2], M.D.,
Seung-Kyung Hann[2], M.D., Kwang Hoon Lee[2], M.D.

Department of Dermatology and Venerology[1],
Mongolian National Medical University, Ulaanbaatar, Mongolia, and
[2]Yonsei University College of Medicine, Seoul, Korea

Hydroa vacciniforme is a rare, chronic photosensitivity disorder manifested in childhood by recurrent vesicles that heal with scarring. We report a case of hydroa vacciniforme in which vesicles were induced with multiple exposures to UVA. The clinical features and appropriate laboratory evaluation of hydroa vacciniforme are reviewed.
(Ann Dermatol 8:(1)70~72, 1996).

Key Words : Hydroa vacciniforme, UVA phototesting

Hydroa vacciniforme is a rare, chronic photosensitivity disorder of unknown etiology[1]. The disease usually begins in childhood and remits spontaneously during adolescence[2].

The eruption can involve all sun-exposed areas, particularly the cheeks, ears, nose, part of the neck and dorsal surfaces of the hands.

The primary lesions are erythematous vesicles or vesiculopapules that develop several days after sun exposure, with a burning sensation. The vesicular lesions become umbilicated and can also become confluent or hemorrhagic. Subsequently there is necrosis and resultant vacciniform scarring.

The lesions of hydroa vacciniforme can be reproduced by phototesting[3,4]. This report presents hydroa vacciniforme with mild scarring that was confirmed by repetitive UVA phototesting.

REPORT OF A CASE

A 8-year-old Korean girl had recurrent eruption with scarring for 2 years. She had been well until the age of 6, when papular eruptions appeared on her face and both upper extremities during the summer season. The patient was treated with oral and topical corticosteroids.

There was no family history of cutaneous photosensitivity and she had had no known exposure to photosensitizers. Physical examination showed pinhead sized reddish papular and pitting scars on the face, especially on the cheeks (Fig. 1). The following clinical laboratory tests were negative or within normal limits: (1) Complete blood cell count (2) Antinuclear antibody (3) Free erythrocyte protoporphyrin (4) Plasma porphyrins (5) Urinary uroporphyrins, coproporphyrins, porphobilinogen (6) Free electroporphyrin. A skin biopsy specimen confirmed the eruption as consistent with hydroa vacciniforme.

Phototesting for the minimal erythemal doses (MEDs) of UVA and UVB was performed with a Sellas sunlight (Dr. Sellmeier Co., Düsseldorf, Germany) as the UVA light source and a bank of fluorescent lamps (FST 12-UVB-HO lamp, Elder, Bygan, Ohio) for the UVB light source. The irradiance of the light sources was determined by an IL 442 A radiometer with an SEE 015 probe for UVA and an SEE 240 probe for UVB. The MED was 40 joules/cm^2 for UVA and 30 millijcules/cm^2 for

Received September 15, 1995.
Accepted for publication November 27, 1995.
Reprint requests to : Seung-Kyung Hann, M.D., Department of Dermatology Yonsei University College of Medicine C.P.O. Box 8044 Seoul, Korea
FAX:82-2-393-9157

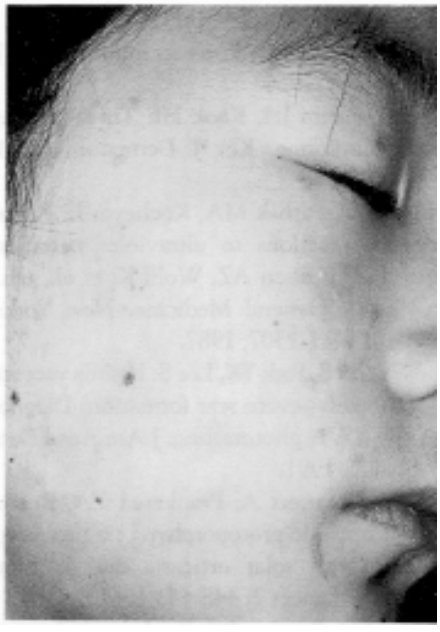

Fig.1. Erythematous, pitted and atrophic scars on face, especially on cheeks.

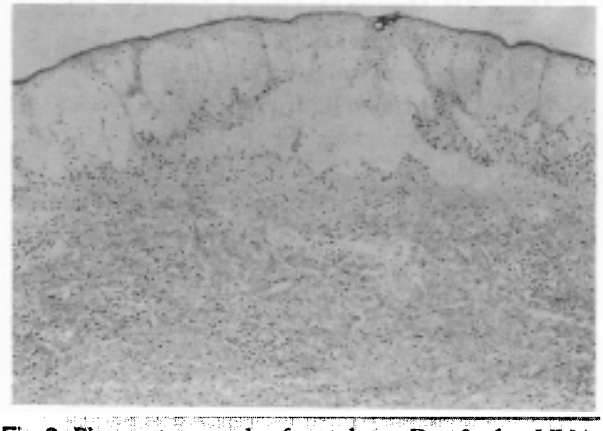

Fig.3. Photomicrograph of vesicle on Day 3 after UVA phototesting. Intraepidermal vesiculation with lymphohistiocytic dermal infiltration is consistent with histopathologic findings of vesicular lesions of hydora vacciniforme.

UVB. The back of the patient, which had not been exposed to sun, was irradiated with 80 joules/cm^2 of UVA on day 1, and the same site received the same dose of artificial UVA on day 2. Vesicles appeared on the UVA-irradiated back on day 3 (Fig. 2). Biopsy specimens from vesicles at the UVA test site on the back showed changes consistent with hydroa vacciniforme (Fig. 3.): multilocular intraepidermal vesiculation with reticular degeneration and lymphohistiocytic dermal

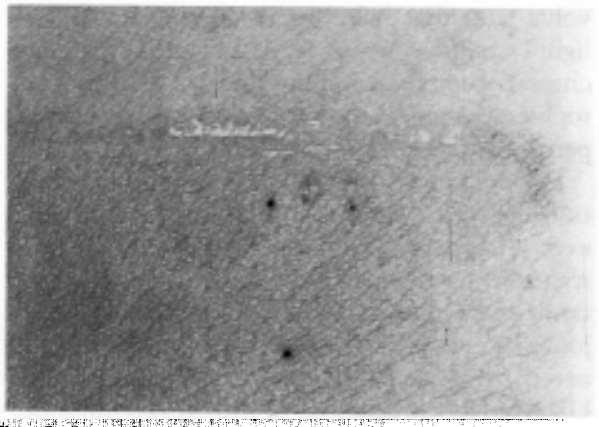

Fig. 2. Vesicle appearing on the UVA-irradiated back on day 3.

infiltration. Avoidance of sunlight and sunscreen application was recommended.

DISCUSSION

Hydroa vacciniforme is a rare, idiopathic photodermatosis with onset in early childhood and characterized by recurrent erythema with discrete vesiculopapular necrosis with vacciniform scarring appearing on sun-exposed skin. Lesions appear in the summer season[5] and vesicles and papular lesions with an erythematous base occur within 1 to 2 days after light exposure and then crust and heal with a tendency to scar formation[6]. The onset usually occurs in childhood (90%) and the ratio of affected males to females is 2 to 1. In most cases it appears to occur sporadically with no family history of photosensitivity. The characteristic histology includes spongiosis, epidermal necrosis and a perivascular round cell infiltrate in the dermis. The differential diagnosis of hydroa vacciniforme includes hydroa aestivale, erythropoietic protoporphyria (EPP), and bullous lupus erythematosus and actinic prurigo.

Hydroa aestivale may show similar clinical characteristics, but this leaves no scarring and is not induced by phototesting[1]. EPP is another type of cutaneous photosensitivity that is usually first manifested in childhood. It is characterized by edematous, urticarial skin lesions and more severe purpuric and vesicular forms, causes waxlike scarring on the sun-exposed areas. EPP has a more intense itching and burning sensation[7,8]. Actinic prurigo may persist into the winter months and may in-

volve sites that have not been exposed to sunlight[9]. Bullous lupus erythematosus manifests chronic cutaneous lesions that eventuate as atrophic scarring with hypopigmentation and hyperpigmentation[2].

The definite action spectrum for the reproduction of hydroa vacciniforme is unknown[10]. However, it has been previously reported that the action spectrum for hydroa vacciniforme is in the UVA range and can be elicited by three exposures of 35 joules/cm^2 each. In our case, photosensitivity with two consecutive daily exposure of 80 joules/cm^2 of UVA led to the diagnosis of hydroa vacciniforme.

The treatment of hydroa vacciniforme remains highly unsatisfactory. Oral hydroxychloroquine and β-carotene[2], topical sunscreens and antimalarials[10,11], and PUVA therapy[3] have been said to be helpful for selected patients. Our patient did not have vesiculation after avoidance of sun exposure and application of topical sunscreen.

REFERENCES

1. Harber LC, Bickers DR: Hydroa aestivale and Hydroa vacciniforme. In Harber LC, Bickers DR eds. Photosensitive diseases: Principles of diagnosis and treatment. Philadelphia: WB Saunders 235-240, 1989.
2. Bickers DR, Demar LK, DeLeo V, Poh-Fizpatrick MB, Aronberg JM, Harber LC: Hydroa vacciniforme. Arch Dermatol 114: 1193-1196, 1978.
3. Lim SD: A case of hydroa vacciniforme. Kor J Dermatol 1:65-67, 1960.
4. Paik SA, Hahm JH, Kook HI: Three cases of hydroa vacciniforme. Kor J Dermatol 15:341-345, 1977.
5. Bernhard JD, Pathak MA, Kochevar IE,Parrish JA: Abnormal reactions to ultraviolet radiation. In: Fitzpatrick TB,Eisen AZ, Wolff K, et al., eds. Dermatology in General Medicine. New York: McGraw-Hill 1481-1507, 1987.
6. Hann SK, Im S, Park YK, Lee S: Hydroa vacciniforme with unusually severe scar formation: Diagnosis by repetitive UVA phototesting. J Am Acad Dermatol 25: 401-403, 1991.
7. Magnus IA, Jarrett A, Prankered TA, Remington C: Erythropoietic protoporphyria : a new porphyria syndrome with solar urticaria due to protoporphyrinemia. Lancet 2: 448-551,1961.
8. Redeker AG, Bronow RS: Erythropoietic protoporphyria resenting as hydro aestivale. Arch Dermatol 73: 164-169,1964.
9. Meara RH, Magnus IA, Grice k, Jones EW, Johansson SG: Hutchinson's summer prurigo. Trans St Johns Hosp Dermatol Soc 57:87-97,1971.
10. Sonnex TS, Hawk JLM: Hydroa vacciniforme. A review of ten cases. Br J Dermatol 118:101-108,1988.
11. Annamalai R: Hydroa vacciniforme in three alternate siblings. Arch Dermatol 103; 224-51, 1971.