Hydroa Vacciniforme with Ocular Involvement

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Abstract

A 7-year-old Thai boy had several episodes of hydroa vacciniforme which were accompanied by an anterior uveitis with corneal clouding and stellate keratic precipitates. Wearing sunglasses prevented additional eye symptoms despite recurrence of the skin lesions. Repetitive UVA phototesting reproduced the typical skin lesions with fever and malaise. No reproduction of skin lesions was revealed by repetitive UVB phototesting. One should be aware of eye involvement in hydroa vacciniforme, and those who experience the eye involvement should be advised to wear protective sunglasses.

Hydroa vacciniforme (HV) was first described by Bazin in 1862. Over the years, increasing numbers of cases have been reported. It is a rare, chronic photodermatosis of unknown etiology, characterized by recurrent discrete vesicular eruptions, followed by progressive necrosis and healing with varioliform scarring on sun-exposed area. The disease usually begins in childhood and remits spontaneously by the late teenage years(1). Eye involvement is unusual in HV and is usually limited to mild keratoconjunctivitis or photophobia without objective findings(2-5). Many researchers(2,4, 6-12) have succeeded in reproducing the eruption by multiple exposures to UVA.

We describe a child with HV who developed severe keratitis and anterior uveitis during several episodes of the eruptions. Phototesting with repetitive exposures to UVA reproduced the typical skin lesions with fever and malaise.

CASE REPORT

A 7-year-old Thai boy was first seen in September 1994 with a two-year-history of recurrent vesicular eruption over the face, ears, chest, dorsal forearms and hands. The lesions started with pruritic erythema which became vesicles and healed with scars in a week. The eruption was usually preceded by fever and malaise. Photophobia, lacri-

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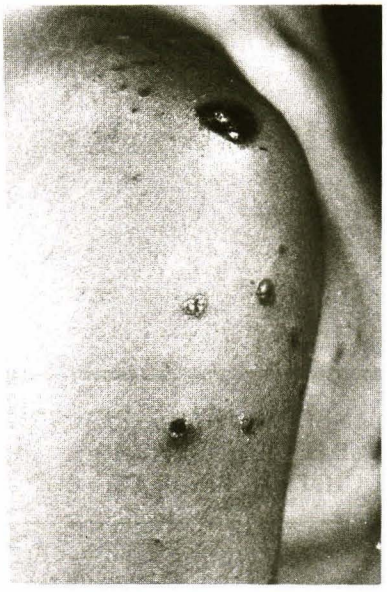

Fig. 1. Umbilicated vesicles with central crust on the shoulder.

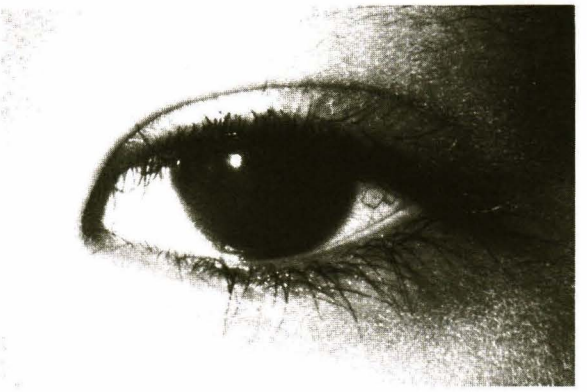

Fig. 2. Conjunctival injection and cloudy cornea.

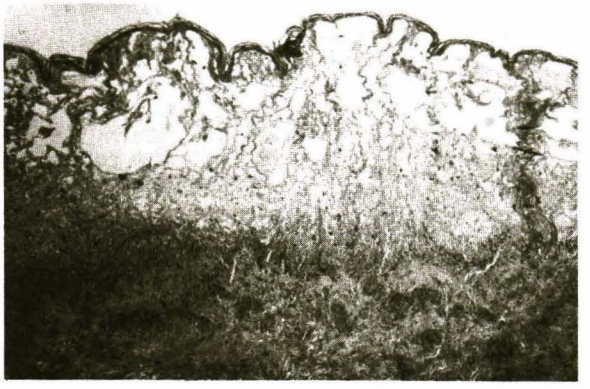

Fig. 3. Histopathological study of the lesion showed spongiosis, diffuse epidermal and upper dermal necrosis.

mation, eye tenderness, and visual disturbance usually accompanied the eruptions. During the first year, the eruption occurred periodically every 2-3 months for the last 6 months the lesions appeared almost every week. No relationship between the eruption and sun exposure was recognized. There was no known exposure to photosensitizers, and the family history revealed no photosensitivity diseases. Physical examination revealed multiple hemorrhagic vesicles and depressed scars on the earlobes, face, shoulders, chest, upper back, and extensor aspects of forearms (Fig. 1). Ophthalmologic examination revealed stellate keratic precipitates on the posterior surface of both corneas causing corneal clouding and decreasing of visual acuity (Fig. 2). There were a small number of cells in anterior chambers consistent with inflammatory keratitis and anterior uveitis of both eyes. The remainder of the physical examination was unremarkable. The results of laboratory studies including urine, stool and blood porphyrin were within normal limits.

The histopathological study of a vesicle from the arm showed spongiosis, diffuse epidermal and upper dermal necrosis. There was lymphohistiocytic infiltration with hemorrhage and thrombosis of small vessels in the dermis (Fig. 3).

Phototesting

Radiation Sources

Sunlamp. The source of polychromatic UVB was a bank of 10 fluorescent bulbs (Philips TL 20 W/12) (UV 800, Waldmann, Villingen-Schwenningen, F.R.G.). The emission spectrum extended from 285 to 350 nm with a maximum between 310 and 315 nm. The UVB irradiance was 2.5 mW/cm^2 at a target distance of 30 cm.

High intensity UVA. The source of polychromatic UVA was from a high-pressure metal halide lamp (UVASUN®3000, Mutzhas, Munich, F.R.G.) that emits wavelengths between 330 nm and 460 nm without any measurable UVB[13]. The UVA irradiance was 66 mW/cm^2 at a target distance of 30 cm.

Dosimetry. A UV-meter with separate UV-detectors for UVA and UVB (IL 1700A radiometer, International Light Inc., U.S.A.) served to determine the UV-irradiance of the UVASUN® and the UV 800.

Determination of Minimal Erythema Doses (MEDs) of UVB and UVA

UVB was applied to small skin fields (1x1 cm) on the patient's back in doses ranging from 0.05-0.3 J/cm^2, in increments of 0.05 J/cm^2. UVA was applied in doses of 5, 10,15, 20, 25, 30 J/cm^2. Test reactions were evaluated immediately and 24 h later.

MEDs of UVB and UVA were determined to be 200 mJ/cm^2 and more than 30 J/cm^2 respectively, which were within normal limits for the patient's skin type. There was no induction of skin lesions in the MED test sites.

Provocative Phototesting

A large test site (5x8 cm) on the patient's back was repeatedly irradiated with daily doses of 100 J/cm^2 UVA on three consecutive days. Another test area was irradiated with daily doses of 300 mJ/cm^2 UVB on three consecutive days.

Twenty four hours after the second irradiation 2- to 4- mm erythematous papules appeared at the UVA test site. Twenty four hours after the last irradiation, the lesions evolved into hemorrhagic vesicles and large bullous and then crusted, leaving hypopigmented scars on healing (Fig. 4). No lesions were reproduced using repeated daily applications of 1.5 MEDs of UVB.

The tissue specimens from UVA-induced lesions showed histopathology which was indistinguishable from those of natural occurrence. Direct immunofluorescence studies of the perilesional and lesional skin were negative for immunoglobulins M, G, A, and C'3 deposition. A serum sample for indirect immunofluorescence studies was also negative.

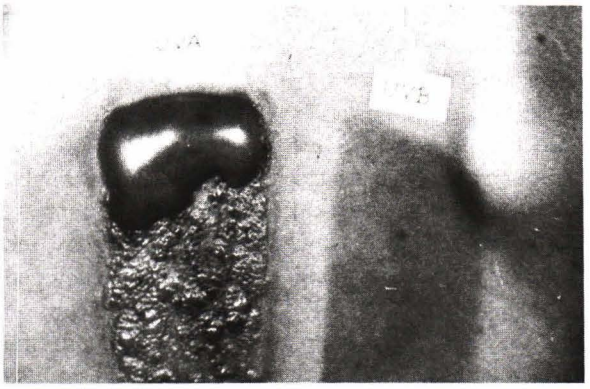

Fig. 4. Erythematous papules, vesicles and large bullous at the UVA test site.

Determination of the effectiveness of broad spectrum sunscreen

UVA was applied to small skin fields (1x1 cm) in daily doses of 5, 10, 15, 20, 25, and 30 J/cm^2 on two consecutive days on the extensor aspects of his forearms with and without broad spectrum sunscreen (Spectraban 28, Stiefel Laboratories Ltd., U.S.A.). Test reactions were evaluated 24 h after the last irradiation.

With daily doses of 20, 25 and 30 J/cm^2 on 2 consecutive days, the erythematous papules appeared on the area without sunscreen application while the area with sunscreen application showed normal appearance.

He was then advised to wear sunglasses and apply topical broad spectrum sunscreen while outdoors during the daytime. While wearing the sunglasses he developed no new eye lesions despite having a recurrence of his skin vesicles.

DISCUSSION

HV was diagnosed in this patient based upon a recurrent vesicular eruption on sunexposured skin that healed with scarring and demonstrated the histopathologic feature of epidermal necrosis. Moreover, no laboratory abnormalities including blood, urine and fecal porphyrin were demonstrated.

Reproduction of vesicles with repetitive UV phototesting was shown to be an important

diagnostic aid. Although there is controversy regarding the wavelengths of light responsible for producing skin lesions, several recent reports have specifically implicated UVA light[6-12]. As in our previous report,[12] we found that the skin lesions in our patient were caused by UVA. Interestingly, we found that constitutional symptoms such as fever and malaise which usually accompany the naturally occurring skin eruption also developed during reproducing HV lesions in our patient.

Eye involvement in hydroa vacciniforme is uncommon[3-5]. When it occurs, mild keratoconjunctivitis or photophobia without objective findings is usually found. Conjunctivitis is often of the congestive type with edema and chemosis associated with lacrimation, photophobia and blephalospasm[3-5]. Keratitis from corneal involvement is due to either the results of vesicle formation on the cornea or secondary to ectropion. It often occurs on the exposed part of the cornea with the upper photoprotected surface spare[5,12]. Sometimes the corneal vesicles heal and remain clear. If deep ulceration develops, the cornea becomes infiltrated and results in superficial keratitis, corneal opacity or complete leukoma[3-5,12]. Similar to a case reported by Bennion SD et al[2], our patient developed severe keratitis and anterior uveitis coincidentally with several outbreaks of the skin eruptions. Protective sunglasses prevented further damage in our patient as in other reported cases[2,12]. Although broad spectrum sunscreen seems to give some UV-protection in the phototesting, it is not efficient enough to prevent the patient from developing HV.

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โรคฮัยโดรอะ แวคซินิฟอร์เม ที่พบร่วมกับอาการทางตา

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รายงานผู้ป่วยเด็กชายไทย อายุ 7 ปี มีประวัติเป็นตุ่มน้ำคันเป็นๆหายๆ ตามใบหน้า ใบหู หน้าอก แขนด้านนอกและหลังมือมา 2 ปี ตุ่มแตกเป็นคราบน้ำเหลือง แล้วกลายเป็นแผลเป็นบุ๋ม มักมีไข้ อ่อนเพลียก่อนเกิดตุ่มน้ำ พร้อมกับมีตาแดง ไม่สู้แสง น้ำตาไหล ปวดตา และตามัวร่วมด้วย ไม่มีประวัติแพ้แสงแดดในครอบครัว การตรวจตาพบมีตาดำขุ่นจาก stellate keratitic precipitate ด้านหลังของแก้วตา และมี inflammatory keratitis และ anterior uveitis ของตาทั้งสองข้าง ตรวจไม่พบพอร์ไฟรินในเลือด ปัสสาวะ และ อุจจาระ การตรวจทางพยาธิวิทยาพบ spongiosis, diffuse epidermal และ upper dermal necrosis มี lymphohistiocytic infiltration, เลือดออก และมีการอุดตันของเส้นเลือดเล็กๆ ในชั้นผิวหนังแท้ ได้ทำการทดสอบด้วยแสง โดยแสง UVA และ UVB ในขนาดต่างๆ กัน ได้ผลบวกต่อแสง UVA ทำให้เกิดตุ่มน้ำ และมีเลือดออกภายในตุ่มน้ำ แต่ได้ผลลบเมื่อทดสอบด้วยแสง UVB การทำ direct immunofluorescence จากตุ่มน้ำ และรอบๆตุ่มน้ำ ไม่พบการฝังตัวของ immunoglobulin ทุกชนิดและ C3 รวมทั้ง indirect immunofluorescence ก็ไม่พบความผิดปกติ ซึ่งลักษณะดังกล่าว เข้าได้กับโรค Hydroa Vacciniforme หลังการรักษาโดยให้สวมแว่นตากันแดดและใช้ยาทาป้องกันแสงแดด ไม่มีรอยโรคเกิดที่ตาอีกเลย แต่ยังมีตุ่มน้ำเกิดขึ้นที่ผิวหนังอีกบ้างเป็นครั้งคราว

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