Recurrent scarring papulovesicular lesions on sun-exposed skin in a 22-year-old man

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CASE SUMMARY

History

A 22-year-old white man has been followed in the Clinical Center at the National Institutes of Health (NIH) for recurrent outbreaks of scarring herpetiform lesions on sun-exposed areas. The patient and/or parents signed consent forms and were followed under NIH protocols that were approved by the institutional review boards of the National Human Genome Research Institute, National Institute of Allergy and Infectious Diseases, and National Cancer Institute.

He reported itching and burning within 15 minutes of sun exposure that evolved into fluid-filled blisters over the next day, subsequently evolving into darkly colored crusts and healing over 1 to 2 weeks, sometimes with poxlike depressions (Fig 1, *A-E*).

While living in the northeastern part of the United States, he experienced outbreaks in the spring and summer with upward of 20 lesions and intermittently throughout the year. Sitting near windows (predominantly ultraviolet A) in school or riding in a car could precipitate lesions, but fireplaces or fluorescent bulbs did not. He continued to participate in outdoor activities.

His first outbreak occurred at age 3 years during the spring in the southeastern part of the United States, when he and 2 siblings had a varicella infection. The summer outbreaks continued but resolved in winter after the family moved to the Northeast. When he was 5 years old, his parents recognized that the lesions were precipitated by sun exposure and began protection with hats, long sleeves, and sunscreen. Outbreaks tended to occur at the beginning of spring or when the patient did not use photoprotection, and 2 were associated with fever and pharyngitis. Bacterial and viral cultures of active skin lesions for herpes simplex virus and varicella-zoster virus were negative. Some episodes were treated with antivirals, antibiotics, or prednisone, with inconsistent improvement. The results of ultraviolet A and ultraviolet B provocative testing were negative. The results of testing for porphyrins in his urine and blood during skin eruptions were negative, as were the results of direct immunofluorescence of a skin biopsy specimen.

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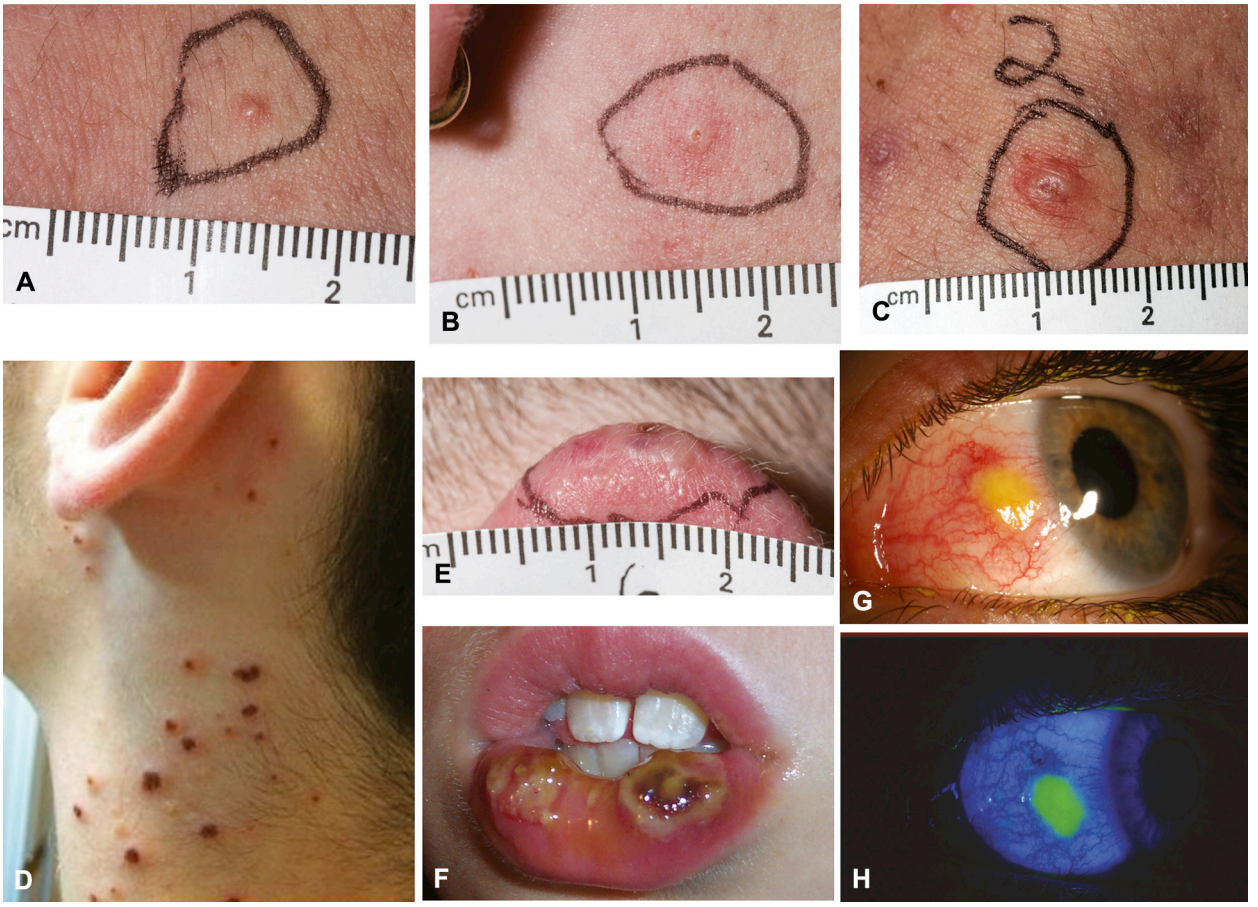

Fig 1. Hydroa vacciniforme. Lesion evolution begins as a vesicle on a red base (**A**), which umbilicates (**B**) and continues to erode (**C**), eventually leading to crusting (**D**). **E**, Larger vesicle (6 mm) on the helix. **F**, Ulcerations of the lower lip that were associated with sun exposure at 9 years of age. **G**, Conjunctival involvement at 10 years of age. The conjunctiva is injected. In the center of the injected conjunctiva is an area of fluorescein staining indicating an epithelial defect, which is also demonstrated visualized with cobalt blue light (**H**). The denuded epithelium is consistent with a ruptured conjunctival vesicle.

Severe inflammation and blistering of his lips (Fig 1, *F*) occurred in association with sun exposure. At 14 years of age, he had gingival inflammation treated with gingivectomies. Oral cultures were negative for adenovirus, cytomegalovirus, herpes simplex virus, and varicella-zoster virus. He had multiple episodes of bilateral follicular conjunctivitis associated with skin lesions and occasionally, nonpainful corneal lesions (Fig 1, *G* and *H*) but denied photophobia.

He denied fatigue, weight loss, night sweats, joint symptoms, skin infections, hypersensitivity to mosquito bites, abnormal reactions to vaccinations, and cold sores. He did not have evidence of immunodeficiency.

The patient's mother had pruritic sun-induced rash without blisters or scarring, and several family members had autoimmune-related abnormalities, including Raynaud syndrome, Sjogren syndrome, gestational lupus, Hashimoto thyroiditis, gluten enteropathy, multiple sclerosis, and positive results of testing for antinuclear antibody.

Physical examination

At age 22 years, examination of the patient at the NIH Clinical Center showed active lesions in various stages (small vesicles, umbilicated papules, and papules with crusts) over the neck, superior helices of the ear, and dorsal surface of the hands (see Fig 1, *A-E*). There were hypopigmented scars (1 mm to 2 cm) on his cheeks, nose, neck, dorsum of his hands, and upper part of the trunk, as well as larger scars on his shoulders. He had a few facial lentigines and 3 café au lait macules (2 to 4 cm) on the right side of the chest and upper part of the right arm but no oral or ocular lesions. A small (.5-cm) right