

CLINICAL IMAGES

Hydroa vacciniforme

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A 10-year-old boy presented with a three-year history of recurrent lesions on photoexposed areas that worsened each summer. The boy had no family history of photosensitivity. Some vesicles and tiny depressed scars resembling chickenpox scars were present on his arms, face and ears. He had freckles on his face and a deep ulcer covered with a crust on his left cheek (Figure 1).

Our initial differential diagnosis included erythropoietic protoporphyria, lupus erythematosus, polymorphic light eruption, actinic prurigo and hydroa vacciniforme. Results of tests for routine laboratory parameters, anti-extractable nuclear antigen antibodies, anti-double stranded DNA antibodies and porphyrins were normal, and antinuclear antibodies were present at low titre (1:40). Histologic examination showed epidermal necrosis and a dense inflammatory suppurative infiltrate that was diffusely distributed (Appendix 1, available at www.cmaj.ca/cgi/content/full/cmaj.092088/DC1). We made a provisional diagnosis of hydroa vacciniforme, which we confirmed by a provocation test for ultraviolet A.

Hydroa vacciniforme is a rare idiopathic photodermatosis characterized by an itchy and stinging sensation followed by the appearance of an erythematous rash within a few hours after exposure to sun. The rash progresses to papules, which undergo vesiculation. Lesions are symmetrically localized in photoexposed areas. The vesicles tend to become umbilicated, then covered by crusts and, within one to six weeks, heal with a depressed vacciniiform scar.¹

Hydroa vacciniforme generally begins in childhood and regresses spontaneously after adolescence, but variants that persist into adulthood have been described. One estimated prevalence of hydroa vacciniforme was at least 0.34 instances per 100 000 patients.¹

Currently, the most accepted pathogenetic hypothesis suggests ultraviolet radiation with wavelengths between 320 and 390 nm as the causal agent of hydroa vacciniforme, but the chro-



Figure 1: (A) Scarring, vesicles and freckles on the face of a 10-year-old boy and (B) a deep ulcer on his cheek.

mophore leading to ultraviolet-induced damage is still unknown.²

The differential diagnosis includes the diagnoses we considered and other more common diseases such as bullous impetigo and herpes simplex.^{1,3} Diagnosis is based on history, clinical findings and histology, and confirmed with photoprovocation.

Therapy consists of topical photoprotection and avoidance of the sun. In patients who do not respond to conservative treatment, use of systemic agents has been reported (β -carotene, diet rich in polyunsaturated fatty acids, psoralen with exposure to ultraviolet A [PUVA], ultraviolet B TL-01 phototherapy, antimalarial agents and immunosuppressive medication). These treatments may be useful in reducing outbreaks but do not reliably prevent lesions.¹

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3. Jacyk WK, Moosa Y. Crusts and vacciniiform scars on sun-exposed skin. *Clin Exp Dermatol* 2010;35:97-8.

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