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CASE REPORT

Progressive scarring facial lesions in a boy

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Key Clinical Message

Hydroa vacciniforme is a photosensitivity disorder characterized by the childhood onset of necrotic vesiculopapules on exposed areas. We present a case of a 10-year-old boy with a 1-year history of papular lesions over his face. In this report, we discuss the in-depth histology of hydroa vacciniforme, and the management options.

Keywords

Hydroxychloroquine, paediatric dermatology, photosensitivity, scarring conditions, vesicular eruptions.

Introduction

A 10-year-old Malay boy presented with a 1-year history of papular lesions over his face, described as blisters which subsequently healed with depressed scarring. The lesions typically occurred after playing soccer outdoors. He was otherwise well and denied any oral medications.

On examination, an erythematous lesion was seen on his left cheek. There were also multiple small, atrophic varioliform scars over his cheeks and nose (Fig. 1, 2, 3 and 4).

Preliminary investigations revealed normal complete blood count and an erythrocyte sedimentation rate of 15. Anti-nuclear antibodies, anti-double-stranded DNA, and plasma porphyrins were negative.

Photopatch test showed a decreased minimal erythema dose (MED) to ultraviolent B irradiation.

A skin biopsy was taken from the erythematous papule.

What is the Diagnosis?

Diagnosis: Hydroa vacciniforme.

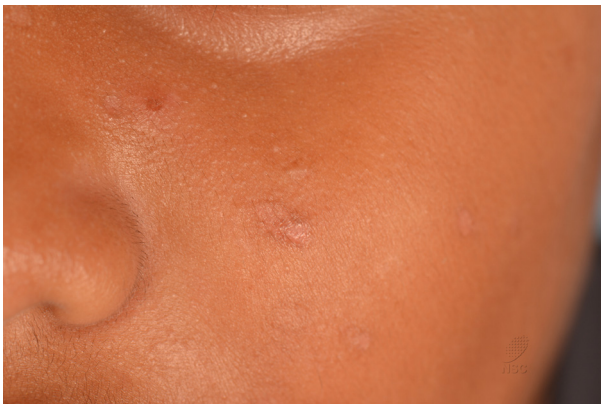

Figure 1. Multiple small, atrophic scars over left cheek.

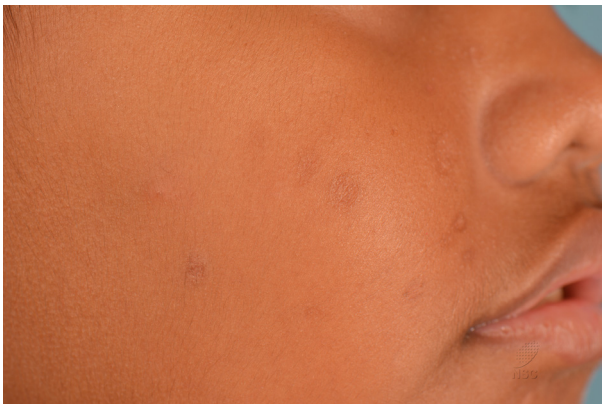

Figure 2. Similar scars over right cheek.

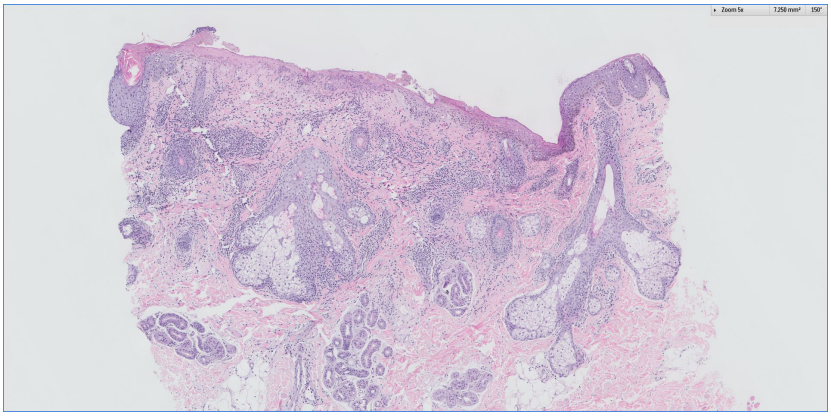

Figure 3. H&E staining at 5× magnification.

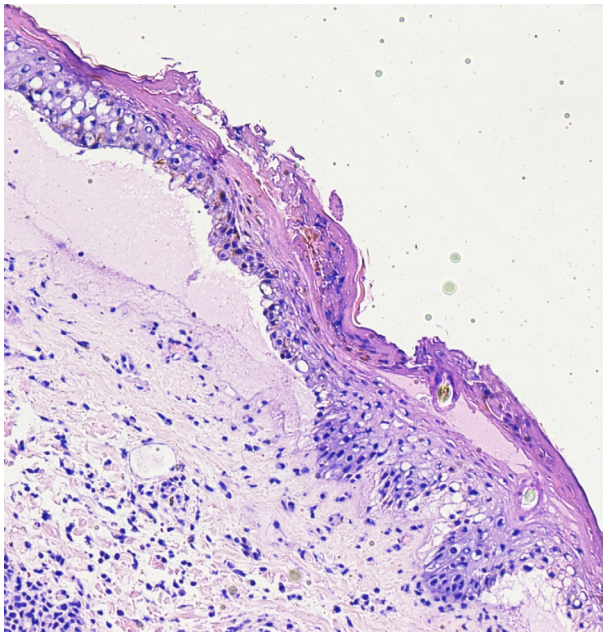

Figure 4. H&E staining at 20× magnification.

Histological Findings and Clinical Course

Histology revealed overlying mild hyperkeratosis, focal parakeratosis, and serous crusting. The underlying epidermis was spongiotic, associated with pallor of the keratinocyte. There was a subepidermal blister filled with serous fluid. There was a moderately dense superficial and mid-perivascular and periadnexal lymphohistiocytic infiltrate. The lymphocytes stained positively for CD3, with a CD4:CD8 ratio of 4:1, with admixed CD20+ B-lymphocytes. Epstein–Barr Encoded RNA In-Situ Hybridisation test (EBER ISH) was negative.

Our patient was advised on daily sun protection with donning of protective clothing and regular application of sunscreens. His parents declined treatment with Hydroxychloroquine for fear of potential side effects. Our patient was compliant with judicious sunscreen application, and his condition remained well controlled. No new lesions were report at the 3-month review.

Discussion

Hydroa vacciniforme is a photosensitivity disorder of unknown etiology, characterized by the childhood onset of necrotic vesiculopapules on exposed areas [1, 2]. The average age of onset is 6 years [3]. Lesions usually appear 24–48 h after exposure to sunlight, appearing as itchy or tender papules and vesicles on a background of edema and erythema. This is followed by a phase of impetiginized crusting. Lesions eventually heal with characteristic varioliform scars [2].

Uncommon presentations of HV include eye signs such as conjunctival injection, photophobia, lacrimation, corneal ulceration, scarring deformities of the ear, nose, and contractures of the fingers [4].

Spontaneous improvement usually occurs at puberty or by the late teenage years [2].

The characteristic histology includes spongiosis and epidermal necrosis, leading to vesicle formation and a perivascular mononuclear cell infiltrate. The upper dermis may also demonstrate necrosis, with infiltration of polymorphonuclear cells [2, 3].

No other abnormalities are usually found on investigation [5]. Differential diagnoses of photosensitive dermatoses in childhood, which may mimic hydroa vacciniforme include bullous lupus erythematosus, erythropoeitic protoporphyria, pseudoporphyria, and hydroa vacciniforme-like T-cell lymphoma.

Phototesting should be carried out. Monochromator tests essentially demonstrate photosentivity in the UVA spectrum, and the UVA provocation tests trigger similar changes and result in scarring [5]. Narrowband UVB is usually normal in patients with HV. However, UVB sensitivity in HV has also been suggested [2, 6]. The wavelengths implicated vary between 320 and 390 nm [1, 5].

The mainstay of treatment for patients with HV is the frequent use of broad-spectrum sunscreens with advice on protective clothing [1]. Besides the use of sunscreens, hydroxychloroquine has also been shown to be of benefit [2, 3, 6], associated with an increase in the UVA MED and in the number and magnitude of UVA doses required to reproduce clinical lesions [5].

Learning Points

Hydroa vacciniforme is a photosensitivity disorder characterized by the childhood onset of necrotic vesiculopapules on exposed areas. Spontaneous improvement usually occurs at puberty or by the late teenage years. No abnormalities are usually found on investigation.

The characteristic histology includes spongiosis and epidermal necrosis, leading to vesicle formation and a perivascular mononuclear cell infiltrate.

Important differential diagnoses include bullous lupus erythematosus, erythropoeitic protoporphyria, pseudoporphyria, and hydroa vacciniforme-like T-cell lymphoma.

Frequent usage of broad-spectrum sunscreens and protective clothing is the predominant method of treatment of HV. Hydroxychloroquine has also been shown to be beneficial.

Conflict of interest

All authors have declared no conflicts of interests.

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