Extensive hydroa vacciniforme*

Hidroa vaciniforme extensa

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DOI: http://dx.doi.org/10.1590/abd1806-4841.20131867

Abstract: Hydroa Vaciniforme is a very rare photodermatosis that is mainly seen in childhood. An 18 year old female student reported that since the age of 5 she has been suffering necrotic lesions and vesicles lesions in exposed areas, leaving asymptomatic varioliform scars, which worsened in summer. Light microscopy showed epidermal necrosis with lymphocytic infiltration . Sunscreens were prescribed with light improvement.
Keywords: Hydroa vacciniforme; Necrosis; Photosensitivity disorders

Resumo: O Hidroa Vaciniforme é uma fotodermatose muito rara vista geralmente na infância. Uma paciente de 18 anos foi examinada, a qual apresenta lesões vesiculosas e necróticas varioliformes nas áreas fotoexpostas, desde os 5 anos de idade, que evoluem para cicatrizes atróficas, piorando no verão. A microscopia óptica mostrou necrose epidérmica com infiltrado linfocítico. Houve pouca melhora com uso de filtros solares.
Palavras-chave: Hidroa vaciniforme; Necrose; Transtornos de fotossensibilidade

INTRODUCTION

Hydroa Vaciniforme (HV) is a rare photodermatosis that is mainly seen in childhood. It is characterized by necrohemorrhagic lesions that appear on uncovered areas of skin, which became crusted then gradually heal leaving varioliform scars, hence the denomination.[1] The course is chronic, characterized by periods of activity and remission.[2,3]

The etiopathogeny is unknown, and there are no genetic or laboratory abnormalities. Therefore, the diagnosis of HV is based on clinical and histological changes.[3] Recently, an association has been reported in some cases between latent Epstein-Barr virus (EBV) infections.[1,4] There is only one report of this condition in the Brazilian literature.[2]

CASE REPORT

An 18-year-old female student reported that since the age of 5 she has been suffering from necrotic lesions and vesicles in exposed areas that leave asymptomatic varioliform scars, which worsen in summer.

Skin examination showed that the nose and ears were covered with hemorrhagic/yellow crusts and erosions (Figure 1). The dorsal surfaces of hands were also strongly involved with hemorrhagic crusts and vesicles (Figure 2). There were also lesions on other areas that were unprotected from the sunlight, such as the ankles and the presternal region (Figure 1).

Light microscopy showed epidermal necrosis with lymphocytic infiltration (Figure 3). Laboratory

Received on 10.05.2012.
Approved by the Advisory Board and accepted for publication on 01.08.2012.
* Study carried out at the Federal University of Pelotas (UFPel) – Pelotas (RS), Brazil.
Conflict of interest: None
Financial funding: None

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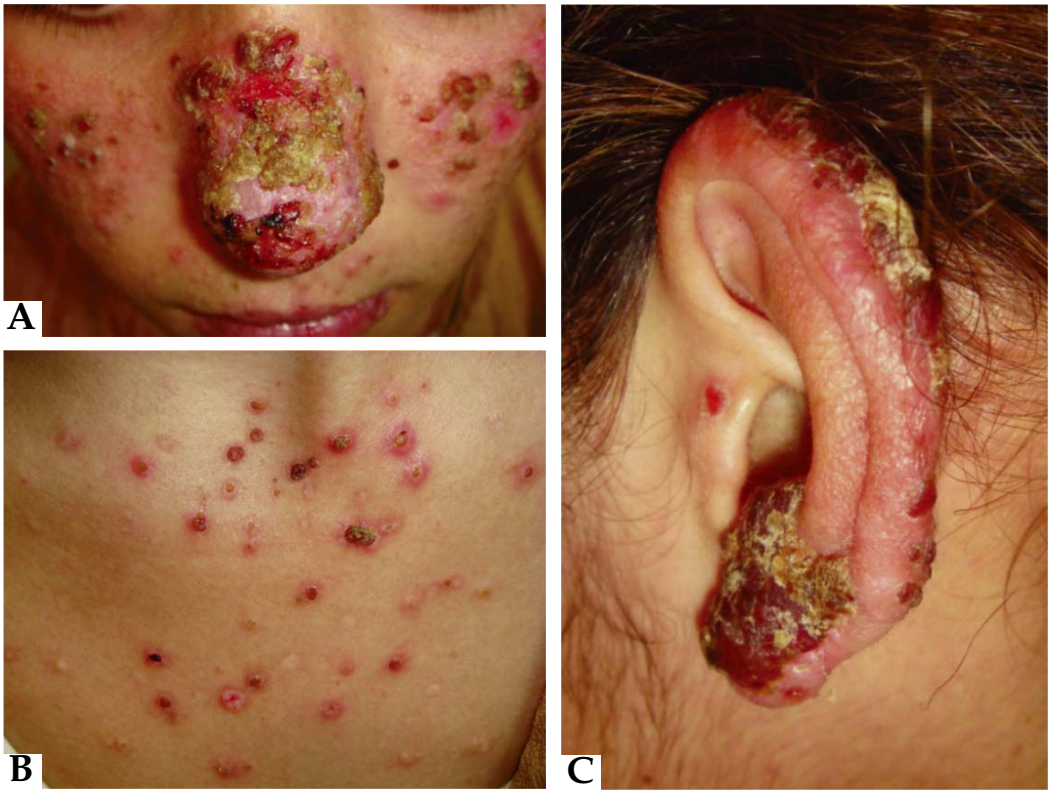

Figure 1: A. and **C**. Serohemorrhagic crusts on the nose and ear. **B**. varioliform pre-sternal lesions

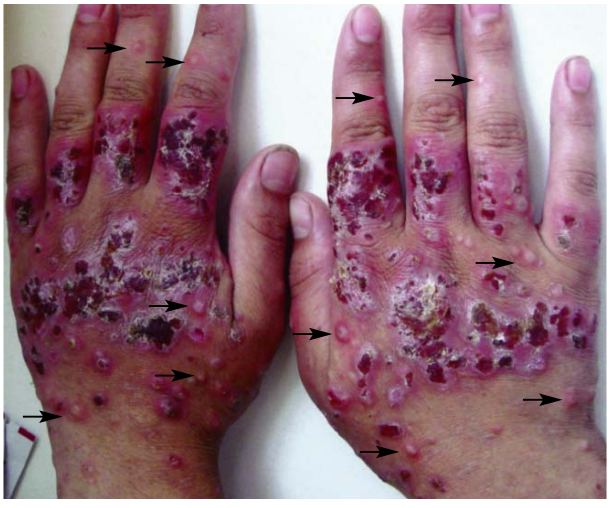

Figure 2: Hemorrhagic crusts on dorsal surface of hands. Note numerous vesicles (arrows), which are initial lesions

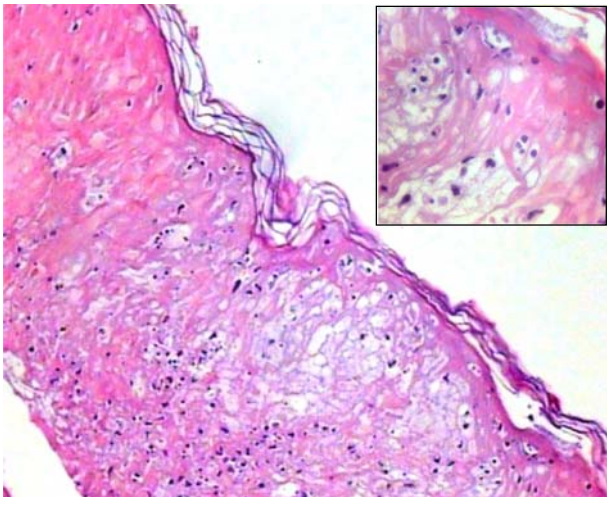

Figure 3: Light microscopy with necrotic epidermis (x 100), inset with detail of the vesicle formation with intraepidermal lymphocytes (x 400).

tests for lupus erythematosus (ANA) and uroporphyrines were normal. There are no similar cases in her family. There was little improvement with the use of highly protective sunscreens (SPF 45), with onset of new lesions. Topical antibiotics were prescribed for secondary infection.

DISCUSSION

The HV differential diagnosis considers several bullous disorders that are caused by exposure to sunlight, including Erythropoietic protoporphyria (EPP), Polymorphic Light Eruption (PLE), Bullous Lupus Erythematosus, Solar Urticaria and Porphyria Cutanea

Tarda. They can be distinguished by detailed clinical history, laboratory findings and histopathology.[1,3]

Similarly to the case here reported, the disease begins in childhood with ages ranging from 1 to 15 years. It affects mostly females and white individuals.[4] It usually subsides spontaneously during adolescence, but there are case reports where it persisted into adulthood.[2,5] The reported prevalence is 0.34 cases per 100,000 people.[2,5]

The initial prodromal manifestation is a sensation similar to an insect bite, followed by itching, and in a few hours, recurrent erythema, vesicles or tense blisters, which can be purulent or hemorrhagic. They gradually become umbilicated, and evolve into crusts. The blisters appear 8-12 hours after sunlight exposure. A vacciniform scar appears within a six-week period. Depending on the case, it may be accompanied by fever, headache and gastrointestinal symptoms.

Histologically, the HV lesions are characterized by skin necrosis and dense suppurative inflammatory infiltrate, as found in the case reported, which helps to differentiate it from other photodermatoses.[5]

It mainly involves sun-exposed sites, particularly the face, ears and the backs of hands. Interestingly, this patient has an extensive presentation with lesions in the presternal region and ankles.[1,2,5] According to some reports, the oral mucosa can also be affected.[6] The disease may relapse in outbreaks; however, after puberty, these outbreaks become sparser.

The treatment is based on solar protection, and in severe cases, systemic corticoids can be used. The use of PUVA proved to be beneficial when used before sun exposure.[1,5,7]

Recent studies described the treatment of hydroa vacciniforme associated with Epstein-Barr viral (EBV) infection with acyclovir/valacyclovir therapy, with a good clinical response. After treatment, the patients reported having less fatigue, rashes, and scars. They were able to spend more time outdoors without causing new eruptions. The risk of lymphoproliferative malignancy in children with HV and chronic infection by EBV should be observed carefully and follow-up is recommended.[8] ❑

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How to cite this article: de Almeida HL Jr, Sartori DS, Kopp J, Velloso CD, Jorge VM. Extensive hydroa vacciniforme. An Bras Dermatol. 2013;88(4):620-2.