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Hailey-Hailey disease associated with herpetic eczema—the value of the Tzanck smear test

Thomás de Aquino Paulo Filho[1], Yara Kelly Rodrigues deFreitas[1], Mylenne Torres Andrade da Nóbrega[1], Carlos Bruno Fernandes Lima[1], Barbara Luiza Medeiros Francelino Carriço[1], Maria Aurora Pinto Leite e Silva[1], Filipe Lauria Paulo[1], Pedro Bezerra da Trindade Neto[1]

1 Department of Clinical Medicine, Dermatology Center, Rio Grande do Norte University, Natal, Brazil

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Corresponding author: Thomas de Aquino Paulo Filho, MD, 240, Deodoro Avenue, 59012600, Natal, RN, Brazil. Email: thomasfi@uol.com.br

ABSTRACT

Background: Herpetic eczema is a herpetic superinfection of a preexisting skin disease. Hailey-Hailey disease is an autosomal dominant dermatosis that is clinically characterized by flaccid vesicles and rashes in intertriginous areas. The coexistence of those findings is a rare condition; only five cases have been published in literature.

Purpose: To report the rare coexistence between Hailey-Hailey disease and herpetic eczema and to highlight the importance of cytology for a quick diagnosis.

Case report: A 38-year-old man had been diagnosed with Hailey-Hailey disease for 13 years. His condition evolved into what could be herpetic eczema, which was later confirmed by skin cytology and histopathology. The man showed remission in the infection after 10 days under treatment with acyclovir.

Conclusion: Research on the concomitance of infection by the herpes virus must be performed in the exacerbations of Hailey-Hailey disease, and, in those situations, the quick diagnosis through skin cytology makes the early treatment possible.

Introduction

Herpetic eczema (HE), also known as Kaposi varicelliform eruption, is the dissemination of the herpes simplex virus in the same area of a pre-existing skin disease. Hailey-Hailey disease (HHD), or benign familial pemphigus, is autosomal dominant and manifests through erythematous plaques with flaccid blisters that rupture easily. It results in superficial linear erosions with crusts and maceration in intertriginous areas. The coexistence between HE and HHD is a rare condi-

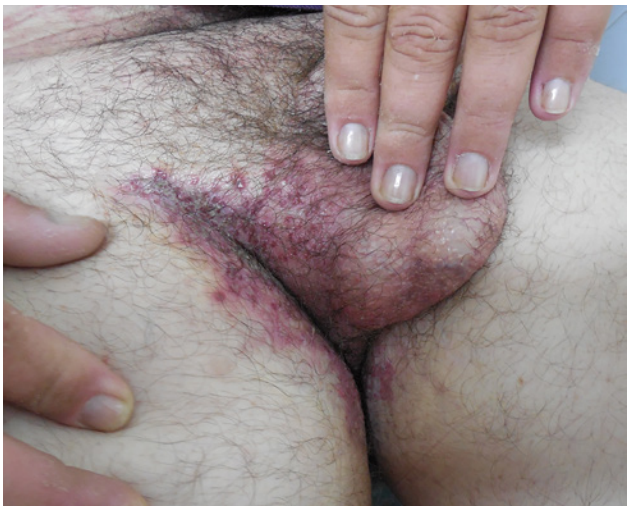

Figure 1. Presence of vesicopustules and ulcerations in an erythematosus base in the inguinal region. (Copyright: ©2014 Paulo Filho et al.)

tion with only five cases that have been described in literature [1,2].

The diagnosis should be clinically suspected as the showing up of monomorphic eruption of vesiculopustules in the areas that had previously been affected by the underlying disease. The diagnosis is confirmed by viral culture, direct immunofluorescence, polymerase chain reaction (PCR) procedure or by skin cytology (Tzanck test); the last one is a rapid and practical method that shows giant viral multinucleated cells, which is an indirect piece of evidence of the presence of herpetic infection [1,2,3,6].

Case report

A 38-year-old male single driver has been diagnosed with Hailey-Hailey disease, which has affected his axillary and inguinal regions for 13 years. Three months prior, he was being treated with acitretin 50 mg/day and desonide 0.05% alternating with tacrolimus 0.1% topically in the areas of exacerbation of lesions. A burning sensation, followed by redness, vesicles and subsequent ulcers in the inguinal region for about two days was reported. Physical examination showed vesiculopustules and areas with ulcerations in the inguinal region (Figure 1). Skin cytology showed giant viral multinucleated cells and characteristic changes of the underlying disease—acantholytic cells, not only parabasal ones, but also those from upper layers of the epidermis, giving them the aspect of "crumbling wall" (Figure 2). Histopathology confirmed the diagnosis of Hailey-Hailey disease and herpetic eczema (Figure 3). The treatment with acyclovir 400 mg three times daily for ten days was recommended with complete remission of the herpetic condition (Figure 4).

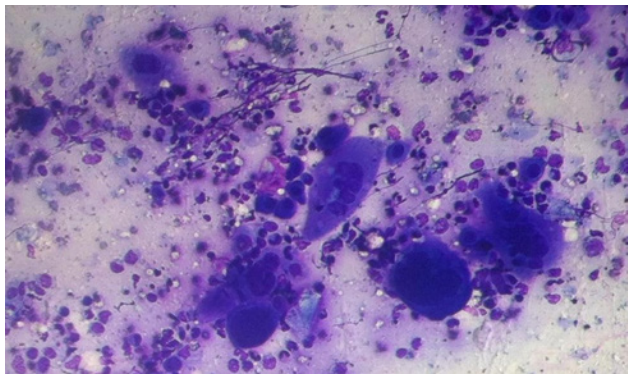

Figure 2. Giant multinucleated cells and acantholytic cells in the same slide. Giemsa stain, at 400x. (Copyright: ©2014 Paulo Filho et al.)

Discussion

HHD was first described in 1939. It normally appears in the third or fourth decade of a person's life, but it may occur at any age. It is a rare entity, and a person may have multiple

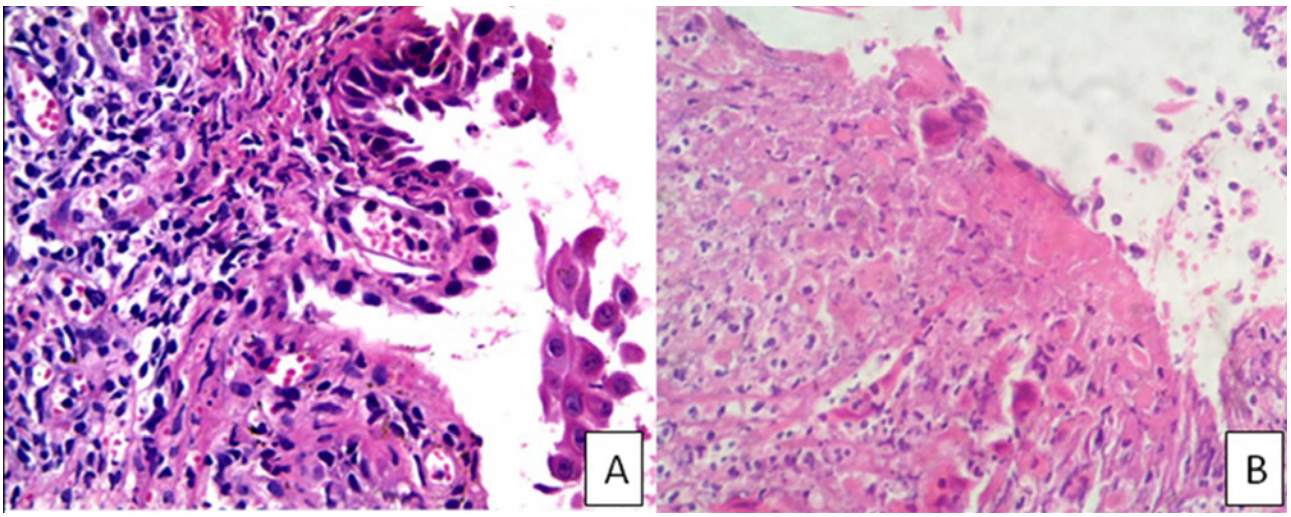

Figure 3. (A) Intraepidermal suprabasal acantholytic blisters with dyskeratosis of keratinocytes. (B) Giant multinucleated cells. H&E, at 400x. (Copyright: ©2014 Paulo Filho et al.)

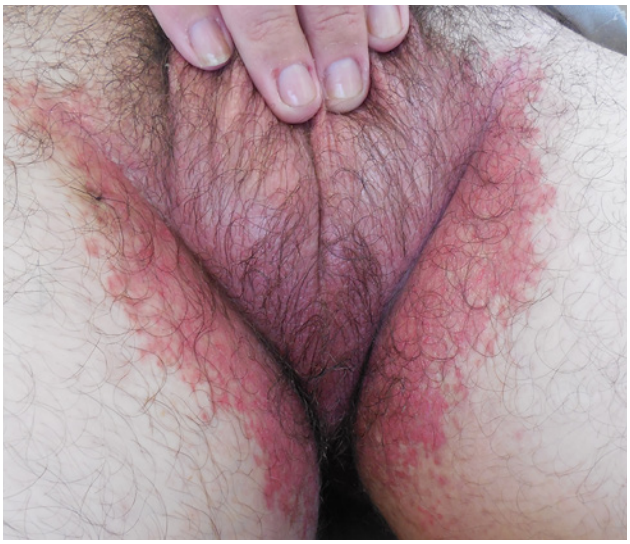

Figure 4. Post-treatment (presence of erythema with no signs of vesicopustules). (Copyright: ©2014 Paulo Filho et al.)

relapses and remissions of the disease [1,4]. It occurs due to mutations on the ATPC21 gene that causes failure in keratinocyte adhesion. The epidermal defect leads to spontaneous acantholysis or it might occur as a result of friction or infections [1]. Infections caused by herpes simplex virus may lead to exacerbation of the lesions [2].

HE is a viral infection caused by herpes simplex (potentially fatal) that appears in preexistent conditions such as atopic and contact dermatitis, Darier disease, pemphigus, pityriasis rubra pilaris, skin lymphoma, and benign familial pemphigus among others. The risk factors in the pathogenesis include rupture of the epidermal barrier and the use of topical calcineurin inhibitors; some authors also describe the topical use of corticosteroids [2,5]. Our patient appeared to be susceptible to the development of the condition presented since he had the main risk factors.

The diagnosis of coexistence of those findings may be reached in a reliable way through skin cytology, which is a simple, quick and viable method that shows sensitivity higher than 80%, making the establishment of early treatment possible. To confirm HE, the presence of giant viral multinucleated cells that might contain inclusion bodies is necessary. On the other hand, the parabasal and upper layers of the skin acantholytic cells may be observed, having the aspect of "crumbling wall" [1,3].

The current therapeutic strategies for HHD aim to reduce outbreaks and improve the patients' quality of life. Oral retinoids, antibiotics, corticosteroids, cyclosporine, topical vitamin D analogues, calcineurin inhibitors, botulinum toxin, surgical excision and CO2 lasers may be utilized [4]. Upon the occurrence of HE, the treatment must be promptly provided. The early use of antiviral drugs—acyclovir, famciclovir and valacyclovir—is extremely important in order to avoid potentially fatal complications [1,2].

Thus, it is important to stress the possibility of concomitant infection by the herpes virus in the frequent exacerbations of Hailey-Hailey disease and, in those cases, the quick diagnosis through skin cytology will allow the initiation of early treatment in order to avoid complications that can be potentially fatal.

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