BRIEF ARTICLES

Hailey-Hailey Disease Complicated by Herpes Simplex Viral Infection

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ABSTRACT

A 51-year-old man with a history of Hailey-Hailey disease presented to clinic due to worsening discomfort from a rash in his groin. Physical examination of the inguinal area and scrotum revealed grouped, punched-out ulcers on a base of macerated, erythematous plaques. Viral culture from the ulcers was positive for HSV-2. The patient had no known history of Herpesvirus infection. Treatment with oral valacyclovir resulted in marked clinical improvement. Although cases of Hailey-Hailey herpeticum are much less common than the better-studied eczema herpeticum, it is important for physicians to be aware that any disease that disrupts the stratum corneum increases the risk of superimposed HSV infection, which constitutes a medical emergency. Prompt treatment with antiviral agents can hasten patient recovery and optimize outcomes.

INTRODUCTION

Eczema herpeticum refers to a disseminated HSV infection in areas of impaired skin barrier, most commonly in the setting of atopic dermatitis. It has rarely been reported in the literature in Hailey-Hailey disease patients. When recognized early, eczema herpeticum is easily and effectively treated with antiviral medications.

CASE PRESENTATION

A 51-year-old man with a history of Hailey-Hailey disease and recurrent MRSA cutaneous infections presented with a rash in his groin persisting for two weeks. This flare was more painful than usual and felt “like ants under (his) skin”. He had been treated without improvement at an outside facility for presumed staphylococcal and yeast infections with low-dose prednisone, IV daptomycin, and topical nystatin. Gold Bond powder was also unhelpful. On physical exam, macerated erythematous thin plaques were present with superimposed grouped punched-out ulcers in the inguinal folds and on the scrotum. There were also crusted small red papules scattered over the central chest, upper back, axillary vaults and posterior neck. Biopsy from the upper back showed extensive suprabasal acantholysis consistent with Hailey-Hailey disease. Secondary infection in the groin with herpes simplex virus (HSV) was suspected; a viral culture from the

inguinal ulcers was positive for HSV type 2. A pemphigus antibody panel was negative and CBC showed mild leukocytosis. The patient denied a history of genital or oral ulcers and had no known history of HSV infection. The patient experienced significant improvement with oral valacyclovir.

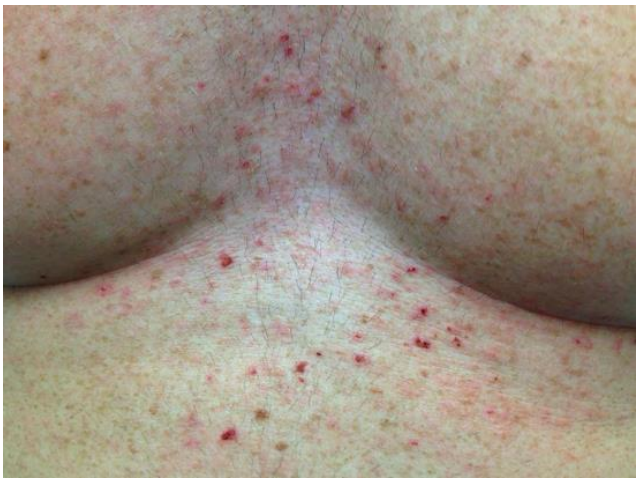

FIGURE 1: Crusted erythematous papules scattered over the central chest.

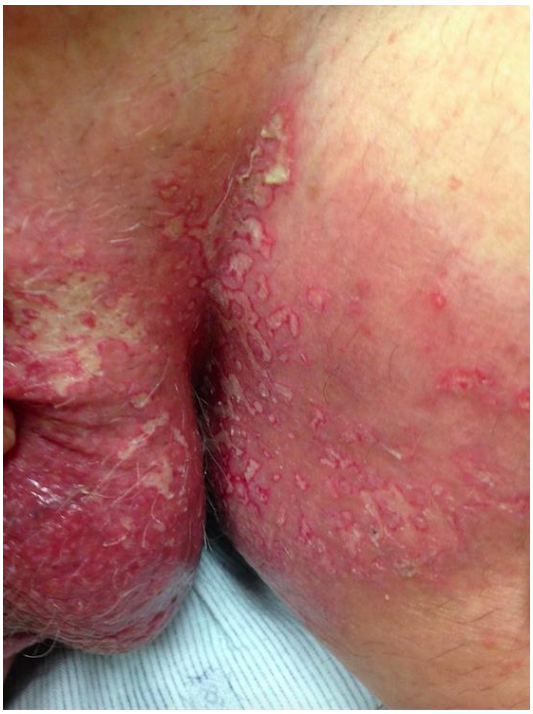

FIGURE 2: Macerated erythematous thin plaques with superimposed crusted punched-out ulcers in the inguinal fold and scrotum.

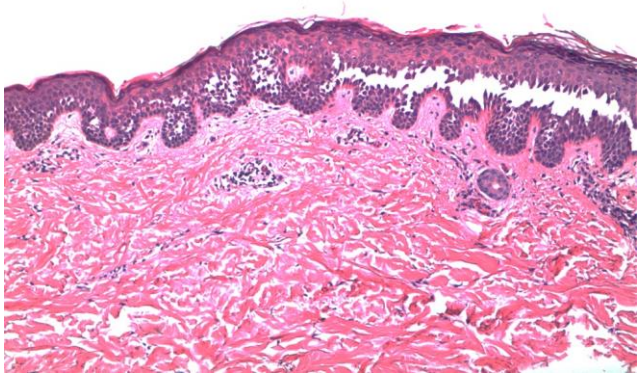

FIGURE 3: H&E (40X). Suprabasal acantholysis throughout the epidermis consistent with Hailey-Hailey disease

DISCUSSION

Eczema herpeticum (EH), also known as Kaposi varicelliform eruption, was initially described by Kaposi in 1887, who thought it resembled chickenpox[1]. EH is the dissemination of a viral infection in the setting of a preexisting skin disease. Hailey-Hailey disease, also known as familial benign pemphigus, is a chronic autosomal dominant blistering dermatosis which usually presents around the third or fourth decades. The disease is a rare genetic disorder that is caused by a lack of protein hSPCA1 produced by gene ATP2C1 located on chromosome 3. This protein is essential for the proper health of skin. The lack of the normal protein causes cells of the skin not to adhere together properly due to malformation of intercellular desmosomes. Patients with such disease will develop blisters and erosions most often affecting the neck, armpits, skin folds and genitals.

EH in the setting of Hailey-Hailey disease is an uncommon occurrence and only a handful of cases have been reported in the literature[2,3]. Most cases of EH are due to *Herpes simplex virus* type 1 or 2. Other viruses may rarely be responsible, such as coxsackievirus A16[1]. This potentially fatal infection appears in areas of compromised skin barrier, including those affected by atopic and contact dermatitis, thermal burns, pemphigus, Darier disease, cutaneous T cell lymphoma, seborrheic dermatitis, and Hailey-Hailey disease among others[2,3]. It usually begins as clusters of vesiculopustules on affected skin and may be accompanied by flu-like symptoms such as fever, chills, and malaise. EH can progress to have severe sequelae, such as herpes keratitis, secondary bacterial infection, disseminated infection with multi-organ involvement, and death, with mortality ranging from 1 to 9 percent[4]. Diagnostic methods of herpes infection include viral culture, direct fluorescent antibody staining, skin biopsy, Tzanck smear exhibiting epithelial multinucleated giant cells and acantholysis, and polymerase chain reaction[1,2,3,5]. Early initiation of antiviral therapy in suspected cases should not be delayed while awaiting test results. The nucleoside analogs, which inhibit viral DNA polymerase, are most commonly used and are very effective[4] as was seen in our patient.

CONCLUSION

Eczema herpeticum can be life-threatening and is considered a dermatologic emergency[4,6]. This case demonstrates the importance for physicians to consider superinfection with herpes in patients with a breakdown of skin barrier, especially in particularly painful and treatment-resistant flares of their skin disease. Early recognition and treatment of eczema herpeticum with an antiviral can rapidly mitigate the morbidity associated with the infection.

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