Relapsing Kaposi's varicelliform eruption in a patient with Darier's disease: A case report

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Darier's disease results from abnormal keratinization of skin and is characterized by numerous keratotic papules that preferentially involve seborrheic areas. Kaposi's varicelliform eruption (KVE) occurs by viral infections such as herpes simplex virus (HSV) in some inflammatory skin disorders like Darier's disease.

Herein, we describe a 68-year-old man, a known case of Darier's disease, who presented with a sudden appearance of umbilicated vesiculopustular lesions on the face, trunk, and extremities. Diagnosis of KVE was confirmed by Tzanck smear and skin biopsy. Although the lesions resolved after oral acyclovir administration, the patient had recurrent episodes of KVE less than one month later.

Keywords: Kaposi's varicelliform eruption, Darier's disease, viral infections

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INTRODUCTION

Darier's disease or keratosis follicularis is an autosomal dominant genodermatosis. This disease results from abnormal keratinization of the skin, nails, and mucous membranes. It is characterized by numerous yellowish–brown keratotic papules and plaques that preferentially involve seborrheic areas. Darier's disease usually appears at puberty with exacerbation by trauma, heat, sweating, and ultraviolet exposure [1]. Herpes simplex virus (HSV) can compromise Darier's disease [2,3]. In this article, we describe a patient with Darier's disease compromised by Kaposi's varicelliform eruption (KVE).

CASE REPORT

A 68-year-old man with a confirmed diagnosis of Darier's disease was admitted with fever, malaise, and sudden appearance of generalized painful skin lesions on his face, trunk, and extremities since 3 days prior to admission (Figure 1). In his family history, he mentioned that five of his children had Darier's disease. One of his daughters also had KVE one week later. He used acitretin for one year, which he discontinued during the previous 8 months. Physical examination revealed the presence of vesiculopustular lesions with central umbilication and crusted erosions in flexural areas that rapidly extended throughout his body. He

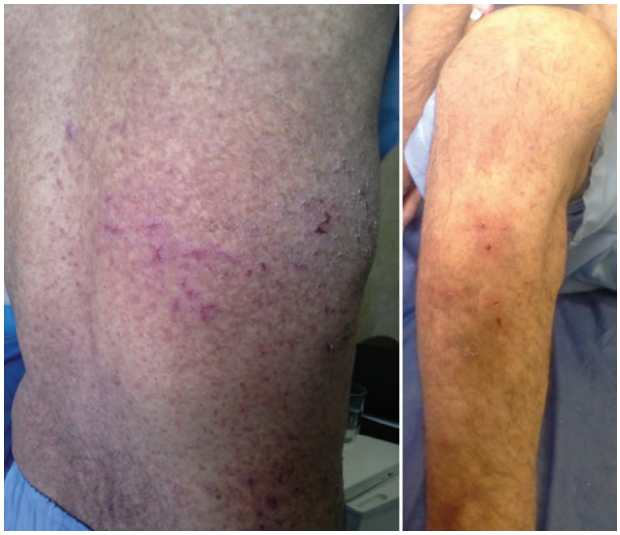

Figure 1. Vesicular lesions and crusted erosions on the trunk and extremities.

also had keratotic papules with yellowish crust on his neck and upper chest. A Tzanck smear was positive for multinucleated epithelial giant cells (Figure 2).

Skin biopsy demonstrated hyperkeratosis, papillomatosis, suprabasilar cleft, and corps ronds in a granular layer (Figure 3). Laboratory test results included a white blood cell count of 10800/mm^3 with 68.6% neutrophils; erythrocyte sedimentation rate (ESR) of 38 mm at 1 hour; and glomerular infiltration rate (GFR) of 68 ml/min. Other laboratory tests were within normal limits. The patient was treated with intravenous

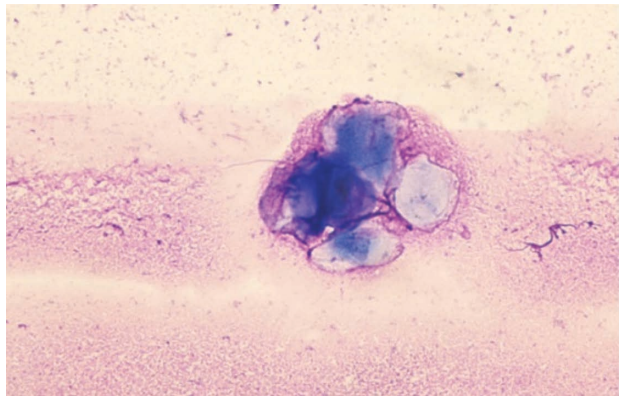

Figure 2. Tzanck smear. Multinucleated giant epithelial cell (Giemsa, 400×).

acyclovir at the same dosage used to treat herpes zoster (10 mg/kg) for 5 days and continued with oral acyclovir (800 mg) for 5 more days, 5 times per day. Direct smear and culture from the lesions revealed *Staphylococcus aureus*. We added ceftriaxone to his treatment regimen for 5 days and after discharging the patient, this medication was changed to cephalexin for 8 more days. After one month, the patient returned with recurrence of similar vesiculopustular lesions of less severity for which he received parenteral acyclovir.

DISCUSSION

KVE eruption or eczema herpeticum occurs in chronic inflammatory skin diseases such as atopic dermatitis, congenital ichthyosiform erythroderma,

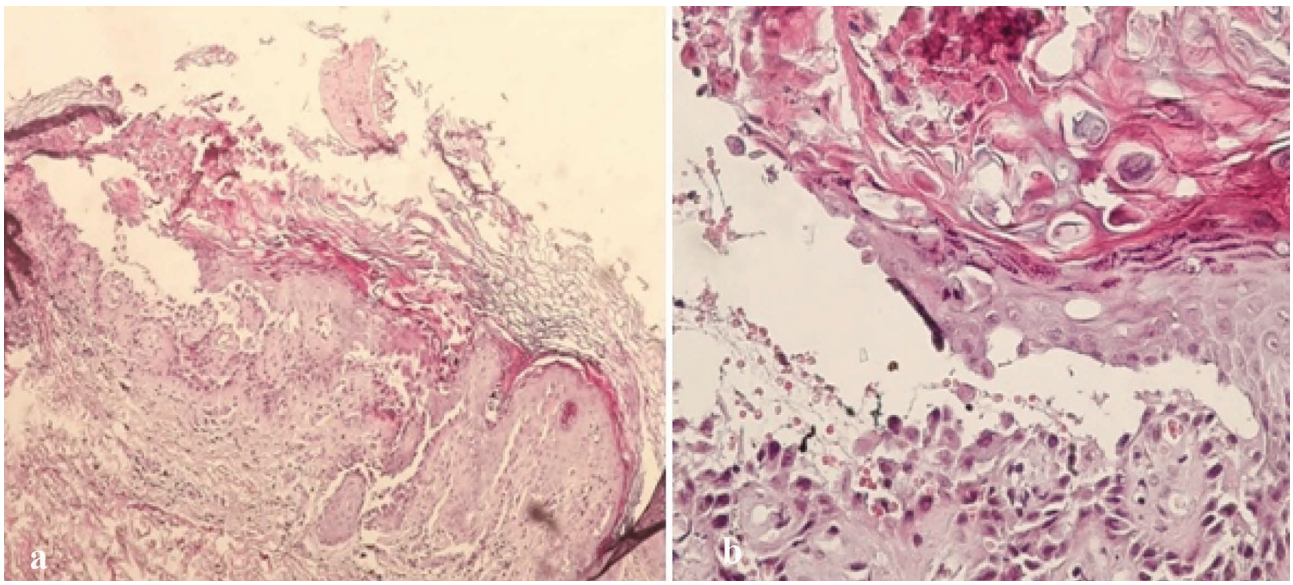

Figure 3. Suprabasilar cleft with multiple characteristic corps ronds, grains (H&E, ×40 **(3.a)** ×400 **(3.b)**)

Darier's disease, cutaneous T-cell lymphoma, Hailey-Hailey disease, seborrheic dermatitis, Wiskott-Aldrich syndrome, pemphigus vulgaris, lupus erythematous, pityriasis rubra pilaris, and psoriasis [3-5]. It is caused by viral infections such as HSV, varicella zoster virus, coxsackie virus, and vaccinia in the setting of cell-mediated immune dysregulation [6-8]. Skin barrier disruption such as second degree burns, dermabrasion, and skin grafts have a role in spread of the lesions [9,10]. Studies have demonstrated that decreased circulating natural killer cells and IL2 receptors have a possible effect in eczema herpeticum eruption in atopic dermatitis [11]. Unresponsiveness (anergy) to skin antigen tests, absence of lymphokine production by peripheral lymphocytes, and immune system dysregulation have been proposed to contribute to the susceptibility of Darier's patients to viral infections [12,13]. It has been shown that severe Darier's disease commonly predisposes patients to KVE. Therefore, control of Darier's disease by topical steroids, keratolytics, and topical and systemic retinoids can reduce possibility of KVE development [13].

Tzanck smear, direct fluorescent antibody, skin biopsy and PCR can be performed for diagnosis confirmation. Treatment is based on antiviral drugs such as acyclovir, valacyclovir, and famciclovir with similar dosages used for varicella zoster. The drug is usually administered orally unless the patient has immunodeficiency or the inability to consume oral medications. In case of resistance to acyclovir, foscarnet can be prescribed [7-9,14]. Recurrence of this disease with less severity is possible due to the decreased protection by neutralizing antibodies [15].

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