

SIGNIFICANCE OF A SIMPLE BEDSIDE TEST IN THE EARLY DIAGNOSIS OF A RARE CASE OF KAPOSI'S VARICELLIFORM ERUPTION

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Sir,

Kaposi's varicelliform eruption (KVE) is a disseminated Herpes simplex virus (HSV) infection which is superimposed on a pre-existing dermatosis. Hailey-Hailey disease (HHD) is a rare genetic dermatosis which is characterized by chronic, recurrent vesicles, erosions and fissures in flexural areas. It is an autosomal dominant condition due to mutations in ATP2C1 gene, leading to defects in keratinocyte adhesion and intraepidermal acantholysis.^{1,2} The occurrence of KVE along with HHD is very uncommon. We hereby report a case of Hailey-Hailey disease who developed KVE and an early diagnosis was arrived at with a simple bedside test, i.e. Tzanck smear.

A 49-year-old male, a known case of HHD, with seasonal recurrences and remissions since 9 years was referred to our hospital with an acute exacerbation of lesions in the form of macerated lesions with oozing and crusting involving neck, both armpits and groins and multiple red raised lesions over the chest and back of one month duration. On examination, he had macerated and crusted plaques on the neck, both axilla and genitocrural folds and multiple erythematous macules and papules over the chest and back. (Figure 1, Figure 2). 10% potassium hydroxide (KOH) mount of skin scraping from axilla was positive for Candida. In view of the widespread involvement, he was started on oral corticosteroids and antihistamines along with oral and topical antifungal agents for the axilla and groins. However, while on therapy, after about a month, he developed fever and sudden eruption of multiple red raised and fluid filled lesions on the

body. On examination, he was febrile and had multiple, discrete vesicles over on an erythematous base with central umbilication and multiple pustules distributed primarily on the chest, abdomen and back (Figure 3) along with increase in oozing and crusting of pre-existing lesions over the flexures (Figure 4). Tzanck smear showed multinucleated giant cells (Figure 5). His total leukocyte count was raised along with increase in the neutrophils but other hematological and biochemical parameters were within normal limits. Serology for IgM and IgG antibodies to HSV was positive. Polymerase chain reaction (PCR) for HSV was also positive. Skin biopsy from one of the lesions showed intraepidermal acantholytic keratinocyte separation with perivascular lymphocytic infiltrate in the dermis. (Figure 6). Based on the clinical features and Tzanck smear he was diagnosed as a case of KVE which was confirmed later with serology and PCR for HSV infection. He was managed with intravenous Acyclovir initially for four days followed by oral Acyclovir along with Injection Amoxycillin with Clavulanic acid and nursing care. Systemic corticosteroids were continued and gradually tapered and stopped. After ten days of treatment, there was complete resolution of the herpetic lesions and other lesions also started improving.

KVE is a serious life-threatening HSV infection that arises in pre-existing skin disorders. In some cases, it may progress to a life-threatening condition in the form of disseminated infection with visceral involvement and death. The most common predisposing factor is the breach in the stratum corneum secondary to skin disease.



Figure 1: Macerated and crusted plaques involving axilla



Figure 2: Macerated and crusted plaques involving genitocrural folds



Figure 3: Discrete vesicles over on an erythematous base with central umbilication and multiple pustules



Figure 4: Discrete vesicles over on an erythematous base with central umbilication and multiple pustules

It has been reported to occur in various pre-existing dermatoses namely, Darier's disease, mycosis fungoides, Sézary syndrome, atopic dermatitis, seborrheic dermatitis, pemphigus foliaceus, ichthyosis vulgaris, and Hailey-Hailey disease.¹ It begins as clusters of umbilicated vesicles and pustules in the areas where the skin has been affected by a pre-existing dermatosis. Tzanck smear from scraping of vesicle base showing multinucleated giant cells supports the diagnosis. Importance to this bedside test cannot be stressed enough as in our case it helped us for an immediate bedside diagnosis when he was examined for this eruption. Diagnosis can be confirmed by polymerase chain reaction for viral DNA, electron microscopic detection of herpes virus from blister fluid or immunofluorescence tests for cells affected by HSV.³

Treatment with intravenous acyclovir should be started without delay in case of high suspicion of KVE or a positive Tzanck smear as was done in our case. The other antiviral drugs which can be used are valacyclovir and famciclovir.

The occurrence of KVE with HHD has rarely been reported. We came across a few such cases in literature.⁴ Our case emphasizes the fact that a simple bedside test like Tzanck smear can be

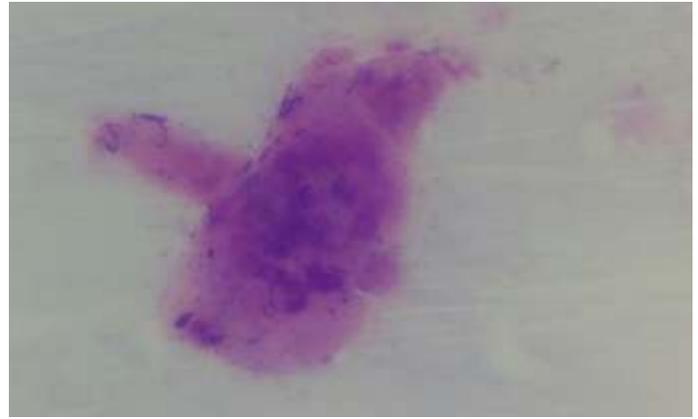


Figure 5: Tzanck smear (1000 X) showing multinucleated giant cell

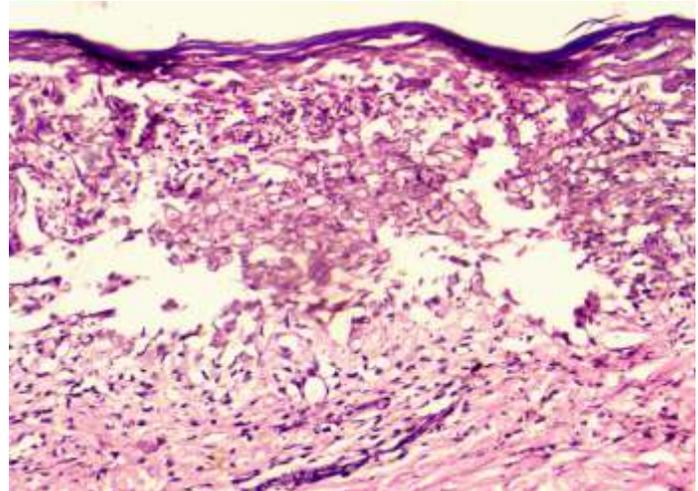


Figure 6: Histopathology (H & E 100X) showing intraepidermal acantholytic keratinocyte separation with perivascular lymphocytic infiltrate in dermis.

crucial in early diagnosis and also, if there is a high index of suspicion of HSV infection, systemic antiviral treatment should be started without delay in order to prevent serious complications and achieve clinical cure.

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