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Case Report

Pathology Section

Kaposi's Varicelliform Eruption in an Adult

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ABSTRACT

Kaposi's Varicelliform Eruption (KVE) or Eczema herpeticum (EH) refers to a widespread cutaneous infection which is caused by a virus that normally causes localized vesicular eruptions, in a patient with a pre-existing skin disease. Children are much more commonly affected and the primary episode is more severe than the recurrence. We are describing here, a case of KVE in an 18- year old atopic male, who presented with an abrupt onset of umblicated vesicular eruptions with a haemorrhagic crusting over the face, neck, trunk and the arms, which was associated with fever. Punch biopsy revealed an intra-epidermal blister with marked acantholysis and a viral cytopathic effect in the form of ground glass nuclear inclusions, giant cell formation and ballooning degeneration of the keratinocytes. Eczema herpecticum is now being seen with increasing frequency in adults. Since the mortality during the primary episode is approximately 10% in children and adults with impaired cellular immunity, it is important to recognize this lesion early.

Key Words: Kaposi's Varicelliform Eruption, Adult, Histopathology

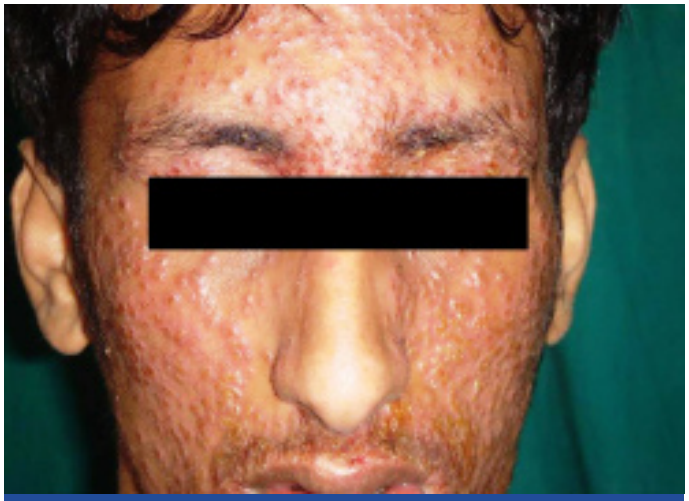
INTRODUCTION

Kaposi's Varicelliform Eruption is a distinct cutaneous eruption caused by Herpes simplex viruses (HSVs) type 1 and 2 and rarely by Coxsackie A16 virus and the Vaccinia virus over pre-existing dermatoses [1]. It is most commonly seen in patients with atopic dermatitis and for this reason, it is also referred to as Eczema herpeticum [2]. KVE has also been described in various dermatoses like pemphigus foliaceous, chronic benign familial pemphigus, Darier's disease, Grover's disease, multiple myeloma, psoriasis, lupus vulgaris and allergic contact dermatitis [1]. When it is recognized early, it can be easily and effectively treated with antiviral agents [3].

CASE REPORT

An 18-year old male presented with an abrupt eruption of reddish, itchy fluid filled lesions over the dorsum of the feet, of five days duration, which was associated with fever and malaise. The lesions rapidly spread to involve the bilateral lower and the upper limbs, the trunk and the face. There was no history of any drug intake or any similar blistering episodes in the past. However, the patient was a known case of atopic dermatitis with a strong family history of atopy. The patient had suffered from chicken pox ten months back. His examination revealed clusters of umblicated fluid filled vesicles with haemorrhagic crusting over both the upper and the lower limbs, the trunk and the face [Table/Fig-1]. Generalized xerosis was present and there was tender lymphadenopathy in the cervical and the axillary regions. The oral cavity showed punctate haemorrhagic lesions on the hard palate. His palms, soles, genitalia and scalp were normal.

Investigations revealed neutrophilic leukocytosis and an elevated ESR of 44 mm (at the end of the first hour). His serum IgE level was markedly raised to 7210 IU/ml. His pus culture revealed the growth of Staphylococcus aureus and the Tzanck smear showed multi-nucleated giant cells.

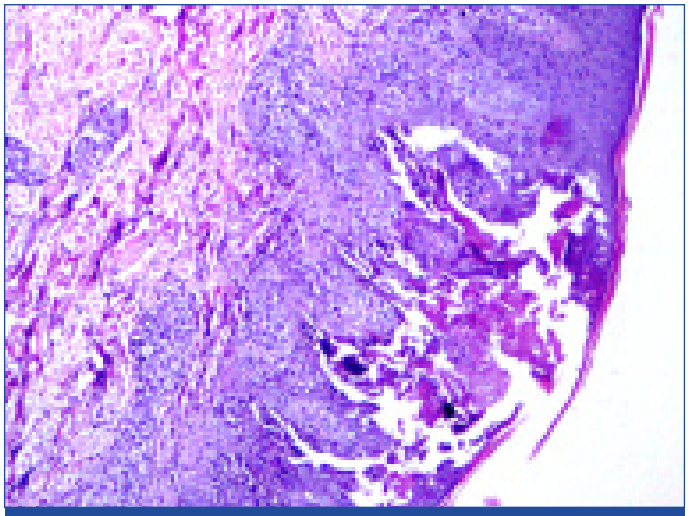


[Table/Fig-1]: Clusters of umblicated vesicles with haemorrhagic crusting distributed all over the face

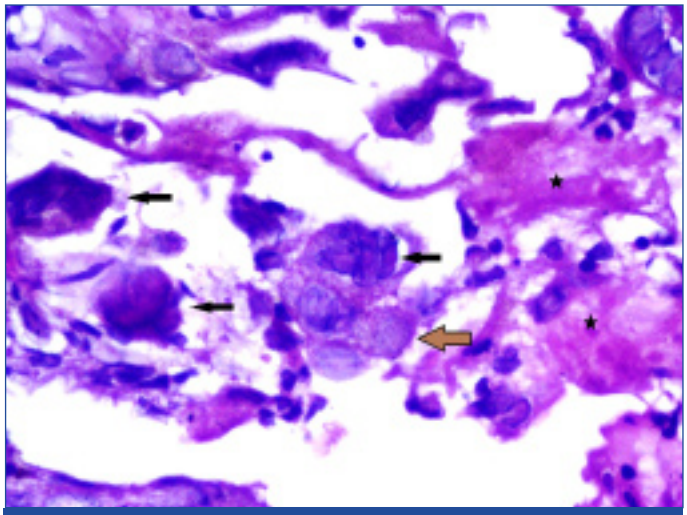
A 3 mm punch biopsy from an umblicated vesicle showed epidermal hyperkeratosis, psoriasiform hyperplasia and an intra-epidermal blister [Table/Fig-2]. The lumen of the blister was filled with acantholytic cells and fibrinoid necrotic material. The blister was lined by intact to acantholytic keratinocytes which demonstrated ground glass hazy nuclei with a peripheral margination of chromatin [Table/Fig-3]. Multi-nucleated cells which harboured eosinophilic inclusions as well as cells which exhibited ballooning degeneration were also seen. The dermis showed mild, perivascular, mixed inflammatory infiltrate. Thus, keeping in view the clinical history and the characteristic histomorphology, a final diagnosis of Kaposi's Varicelliform Eruption was made. The patient was treated with oral acyclovir and he responded dramatically to the treatment. No recurrence has occurred so far.

DISCUSSION

KVE was first described in 1887 by Moritz Kaposi [4]. KVE or EH is a potentially life threatening viral infection that arises in a pre-



[Table/Fig-2]: Scanner view of the lesion showing epidermal hyperkeratosis and psoriasiform hyperplasia along with an intra-epidermal blister (H&E x 40)



[Table/Fig-3]: Another view showing multi-nucleated cells harbouring eosinophilic inclusions (black arrow) and few cells showing ballooning degeneration (brown arrow). Fibrinoid necrosis in the lumen is also seen (black star). (H&E x400)

existing skin condition [2]. These infections which are otherwise mild and localized, present in a florid and disseminated manner on the background of such dermatoses which have been caused due to some unknown reasons [2]. Disruption of the stratum corneum secondary to the skin disease is the most common predisposing factor and it is a commonality among all the diseases which are associated with KVE [3].

The true incidence of KVE is not accurately known, because of its rarity and because of the lack of large scale studies. KVE was originally considered as a disease of infants, and it remains to be more common in children, but it can occur at any age [5]. EH can occur as either a primary or a recurrent type of infection. A majority of the patients with the primary type of EH are infants or children. The primary type of EH can be a serious disease with viraemia and potential internal organ involvement which results in death, while the recurrent type generally shows no viraemia, except in immunologically compromised patients [6].

The exact pathogenesis of KVE is yet to be ascertained. It has been speculated that an impaired barrier function of the epidermis and a defective host immune response are the factors which are responsible for an increased susceptibility to KVE [1]. In a retrospective review of 100 KVE patients, Wollenberg et al [7] found that a high serum IgE level and an early onset of atopic dermatitis were both risk factors. Both these factors were present in our patient also. T-cell mediated immunity is important in the control of the primary and the recurrent infections and an HSV specific cell mediated immune defect could be the reason for the development of KVE. A recent study by Howell et al [8] concluded that the cathelicidin peptide, LL-37 might be deficient in patients with atopic dermatitis, which could explain the increased susceptibility to KVE. The cathelicidin family of inducible antimicrobial peptides is an integral component of an innate immune response that has activity against bacteria, fungal and viral pathogens [8].

The disease begins as clusters of umblicated vesiculopustules which are accompanied by a flu-like syndrome [2,5]. These pustules progress to painful, haemorrhagic, crusted, punched out erosions that coalesce to form denuded areas which are prone for secondary bacterial colonization [5].

Though HSV spreads by droplet infection or by direct contact, there has been no mention of strict isolation of the KVE cases in literature. There have been reports of KVE presenting as mini outbreaks in dermatological wards, resulting in life threatening nosocomial infections [1,9].

The diagnosis of KVE is mainly made on the basis of clinical examination, although several laboratory tests can be helpful [5]. A Tzanck smear of an opened vesicle can provide a rapid diagnosis when it shows the characteristic multinucleated giant cells and acantholysis [3]. Though it is the fastest test, the Tzanck smear is neither sensitive nor specific for the HSV infection [5]. Viral culture of fresh vesicular fluid and direct observation of the infected cells scraped from the ulcerative lesions by direct fluorescence antibody staining, are the most useful and reliable diagnostic tests available. If the lesions are atypical, old or equivocal, a biopsy or PCR should be considered [3].

EH is a self-limiting disorder but it carries a minor risk for severe and bilateral herpetic ocular disease. Systemic viraemia with multiple organ involvement is the major cause of morbidity and mortality [10]. An early use of both anti-viral drugs and antibiotics is extremely important; their use should not be delayed pending laboratory tests. The most commonly used antiviral drugs are the nucleoside analogues, which inhibit the viral DNA polymerase. The initial treatment is generally high dose IV acyclovir which is the most widely studied and used drug for KVE [5].

REFERENCES

[1] Rao GRR, Chalam KV, Prasad GP, Sarnathan M, Kumar HKY. Mini outbreak of Kaposi's varicelliform eruption in the skin ward: A study of five cases. *Indian J Dermatol Venereol Leprol* 2007;73:33-35.

[2] Shenoy MM, Suchitra U. Kaposi's varicelliform eruption. *Indian J Dermatol* 2007; 73(1): 65.

[3] Lee JS, Park YL, Whang KU. A case of eczema herpeticum with the Hailey-Hailey disease. *Ann Dermatol* 2009; 21(3): 311-14.

[4] Wheeler CE Jr, Abele DC. Eczema herpeticum, primary and recurrent. *Arch Dermatol* 1966; 93:162-73.

[5] Olson J, Robles DT, Kirby P, Colven R. Kaposi varicelliform eruption (eczema herpeticum). *Dermatol Online Jour*. 14(2): 18.

[6] Xu X, Erickson L, Chen L, Elder DE. Diseases caused by Viruses. *In*: Elder DE, Elenitsas R, Johnson BL, Murphy GF, Xu G, editors. Histopathology of the skin. 10th ed. Philadelphia: Lippincott William and Wilkins; 2008; 641-42.

[7] Wollenberg A, Zoch C, Wetzel S, Plewig G, Przybilla B. The predisposing factors and the clinical features of eczema herpeticum-a retrospective analysis of 100 cases. *J Amer Acad Dermatol* 2003; 49:198-205.

[8] Howell MD, Jones JF, Kisich KO, Streib JE, Gallo RL, Leung DY. Selective killing of the vaccinia virus by LL-37: Implications of eczema vaccinatum. *J Immunol* 2004; 172:1763-67.
[9] Sepkowitz KA. How contagious is vaccinia? *N Eng J Med* 2003;348: 439-46.
[10] George M, Pakran J, Rajan U, George S, Thomas S. Localised psoriasis herpeticum: Case report and review of literature. *Indian Dermatol Online J* 2011;2;16-18.

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