



CASE STUDY

An Unusual Facial Rash



 **Pre-test CME Quiz**

ABSTRACT

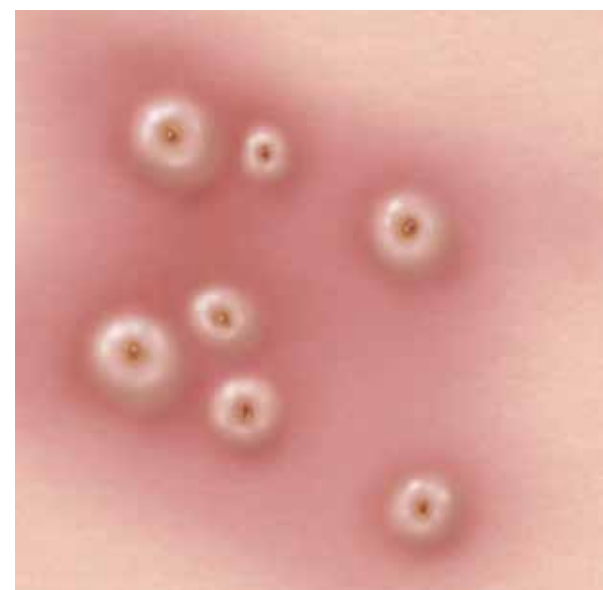
Kaposi varicelliform eruption (KVE) is an infection of a dermatosis by pathogens such as herpes simplex virus (HSV) type 1, HSV-2, coxsackievirus A16, or vaccinia virus. KVE begins as a sudden eruption of painful and crusted or hemorrhagic vesicles, pustules, or erosions in areas of a preexisting dermatosis. Transmission occurs through contact with an infected individual or by dissemination of primary or recurrent herpes. Viral cultures of fresh vesicular fluid or direct observation of infected cells scraped from ulcerated lesions by direct fluorescent antibody staining are the most reliable diagnostic tests for KVE. Antivirals, such as acyclovir and valacyclovir, are used in the treatment of KVE.

KEYWORDS: Kaposi varicelliform eruption, eczema herpeticum, herpes simplex virus, atopic dermatitis, antivirals

A 44-year-old female presents with a 1 month history of umbilicated vesiculopustules on the lower half of the face. She has been treated with multiple topical and systemic corticosteroids, antibiotics, and antifungals with only worsening symptoms. She has a history of chronic dermatitis and herpes labialis.

What is your diagnosis?

Kaposi varicelliform eruption (KVE) is an infection of a dermatosis by pathogens such as herpes simplex virus (HSV) type 1, HSV-2, coxsackievirus A16, or vaccinia virus. This condition is also referred to as eczema herpeticum because this condition commonly occurs in patients with atopic dermatitis (AD) and disseminated HSV infection.¹



Gross appearance of umbilicated vesiculopustules



ABOUT THE AUTHOR

Francesca Cheung, MD CCFP, is a family physician with a special interest in dermatology. She received the Diploma in Practical Dermatology from the Department of Dermatology at Cardiff University in Wales, UK. She is practising at the Lynde Centre for Dermatology in Markham, Ontario and works closely with **Dr. Charles Lynde, MD FRCPC**, an experienced dermatologist. In addition to providing direct patient care, she acts as a sub-investigator in multiple clinical studies involving psoriasis, onychomycosis, and acne.

The pathophysiology of KVE remains unclear. Preexisting cutaneous conditions that have been associated with KVE include AD, Hailey-Hailey disease, ichthyosis vulgaris, pemphigus foliaceus, Darier disease, seborrheic dermatitis, lupus erythematosus, psoriasis, Wiskott-Aldrich syndrome, congenital ichthyosiform erythroderma, mycosis fungoides, and Sézary syndrome.²

It has been postulated that individuals with AD are more likely to develop KVE is due to immune defects involving both cell-mediated and humoral immunity, especially the Th-2 cytokine milieu found in AD, as well as the defective mechanical barrier of affected skin in an individual with AD.³

KVE has also been reported after epidermal disruption caused by scratching, burns, and skin grafts.⁴ Filaggrin is a critical protein involved in formation of an effective skin barrier. Filaggrin mutations, most commonly R501X,

Figure 2: Kaposi varicelliform eruption

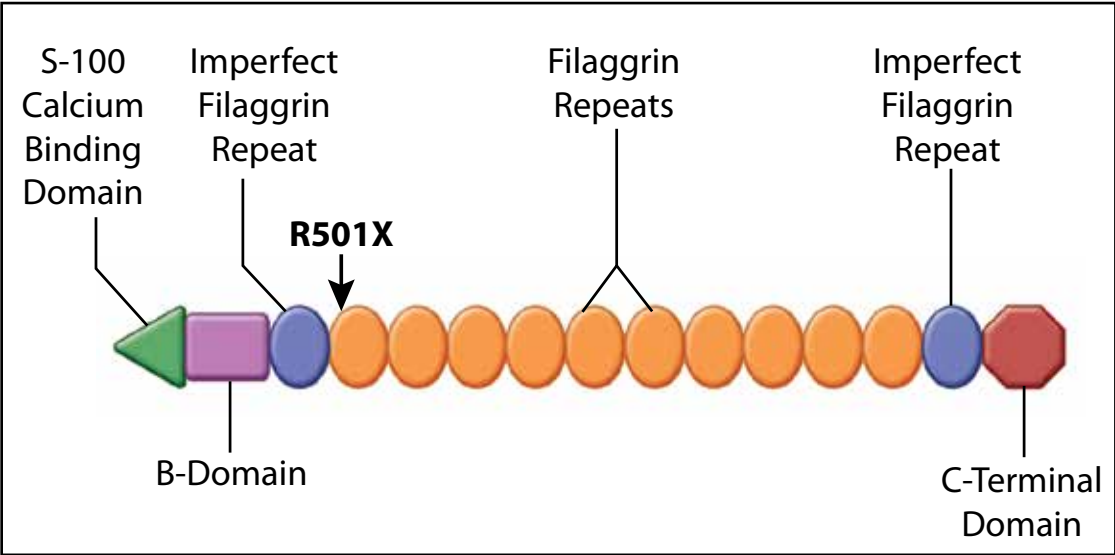


significantly increase the risk of developing KVE in patients with AD (Figure 1).⁵

KVE begins as a sudden eruption of painful and crusted or hemorrhagic vesicles, pustules, or erosions in areas of a preexisting dermatosis (Figure 2). Transmission occurs through contact with an infected individual or by dissemination of primary or recurrent herpes. Associated symptoms include fever, malaise, and lymphadenopathy. The lesions tend to spread over 7-10 days and heal in 2-6 weeks in primary episode. The average duration of illness is 16 days.¹ Recurrent episodes may occur, but are usually milder and not associated with systemic symptoms. Lesions may heal with scarring in severe cases.

Viral cultures of fresh vesicular fluid or direct observation of infected cells scraped from ulcerated lesions by direct fluorescent antibody staining are the most reliable diagnostic tests for KVE. A biopsy can establish a diagnosis in cases that are atypical or equivocal. Bacterial culture is used to assess

Figure 1: Filaggrin protein and location of mutation (R501X)





SUMMARY OF KEY POINTS

Kaposi varicelliform eruption (KVE) is an infection of a dermatosis by pathogens such as herpes simplex virus (HSV) type 1, HSV-2, coxsackievirus A16, or vaccinia virus.

KVE is also referred to as eczema herpeticum because this condition commonly occurs in patients with atopic dermatitis (AD) and disseminated HSV infection.

KVE begins as a sudden eruption of painful and crusted or hemorrhagic vesicles, pustules, or erosions in areas of a preexisting dermatosis.

Preexisting cutaneous conditions that have been associated with KVE include AD, Hailey-Hailey disease, ichthyosis vulgaris, pemphigus foliaceus, Darier disease, seborrheic dermatitis, lupus erythematosus, psoriasis, Wiskott-Aldrich syndrome, congenital ichthyosiform erythroderma, mycosis fungoides, and Sézary syndrome.

With the introduction of intravenous acyclovir, in addition to systemic/topical antibiotic treatment, the mortality rate from KVE has decreased from as high as 50% to less than 10%.



Post-test CME Quiz

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for secondary infection by Staphylococcus or Streptococcus.¹

Antivirals, such as acyclovir and valacyclovir, are used in the treatment of KVE. Systemic viremia with multi-organ involvement or septicemia from secondary bacterial infection is the major cause of morbidity and mortality. With the introduction of intravenous acyclovir, in addition to systemic/topical antibiotic treatment, the mortality rate from KVE has decreased from as high as 50% to less than 10%. Patients should generally be reassessed in 2 weeks to assess for therapeutic response. Significant complications may arise from keratoconjunctivitis or uveitis and consultation with an ophthalmologist is indicated when ocular involvement is suspected.⁶

All of the tables and photos are original.

No competing financial interests exist in preparation of this case study.

References

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