Kaposi's varicelliform eruption: A case series

Bruno Ferrari[1], Vanina Taliercio[1], Paula Luna[1,2], María Eugenia Abad[1,2], Margarita Larralde[1,2]

[1]Pediatric Dermatology Section, Hospital Ramos Mejía, [2]Dermatology Service, Hospital Alemán, Buenos Aires, Argentina

ABSTRACT

Kaposi's varicelliform eruption is a rare and potentially fatal viral infection caused mainly by reactivation of herpes simplex virus. It concomitantly occurs with pre-existing skin conditions, mostly atopic dermatitis, so it is predominately found in children. We present a case series that includes four adults, familial cases, and previously healthy patients. We also highlight clinical features, associations and therapeutic options.

Key words: Atopic dermatitis, eczema herpeticum, herpes simplex virus

INTRODUCTION

Kaposi's varicelliform eruption (KVE), also known as eczema herpeticum,[1] is a rare and potentially fatal viral infection, caused mainly by reactivation of herpes simplex virus (HSV), predominantly type I. Other viruses such as Coxsackie A 16, vaccinia,[1] and varicella zoster[2] have also been implicated in its pathogenesis. It concomitantly occurs with preexisting skin conditions,[3] mostly atopic dermatitis (AD), with a higher prevalence in childhood,[1] although it can occur at any age. It is characterized by widespread clusters of umbilicated vesicopustules and eroded vesicles, alternating with punched-out ulcers covered by hematic crusts usually located over the head, neck, and trunk. It may affect general health and can be fatal. The diagnosis is clinical[1]. Antiviral treatment proved to be effective and should be instituted with no delay to prevent significant morbidity and mortality.[3]

CASE REPORTS

We conducted a retrospective observational study that included 19 patients [Table 1] between 0 and 55 years who were diagnosed with KVE between 01 March 2008 and 01 March 2014. Fifteen were children and only four were adults, with a similar distribution in both sexes (9 females and 10 males).

Fourteen patients had a history of AD, one had psoriasis, one pemphigus foliaceus, one Sézary syndrome, and two previously healthy patients developed their debut of AD immediately before KVE. Fifteen cases were sporadic, whereas four were familial (two cousins on one side and two siblings on the other). The head was the most commonly involved site, followed by trunk and limbs. Four patients had generalized skin involvement [Figure 1]. The most severe forms of KVE were seen in patients with AD (three of them had widespread lesions).

Clinical diagnosis was performed in all cases, supported by the positivity of cytodiagnosis of Tzanck in all patients in whom it was performed. In five patients a skin biopsy was taken, showing intraepidermal vesicle and peripheral ballooning degeneration further confirming the diagnosis. Two patients showed oral involvement with feeding difficulties, two had significant scarring on their face [Figure 2], whereas two patients had severe involvement of conjunctival and genital [Figure 3] mucosa. All patients were treated with oral or parenteral acyclovir (with a dose ranging from 800 to 1600 mg per day until complete resolution), whereas in 10 patients, systemic antibiotic was added. Five patients required acyclovir suppressive therapy due to relapses (acyclovir 800 mg per day for 6–12 months).

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.169714

Quick Response Code:

Address for correspondence:
Dr. Bruno Ferrari,
Pasaje Rauch 3974 9° D CP - 1177, Buenos Aires, Argentina.
E-mail: brunoferraripiel@gmail.com

Cite this article as: Ferrari B, Taliercio V, Luna P, Abad ME, Larralde M. Kaposi's varicelliform eruption: A case series. Indian Dermatol Online J 2015;6:399-402.

DISCUSSION

KVE was first described by Moritz Kaposi in the 19th century, who assumed it was a fungal infection, whereas Juliysber argued that it was caused by bacterias. In the 20th century Freun showed cytoplasmic inclusions confirming the viral etiology.[4]

KVE usually develops in patients with preexisting chronic dermatoses, especially atopic dermatitis. It has also been reported in association with psoriasis, pityriasis rubra pilaris, Darier's, Grover's, and Hailey–Hailey diseases,[1,5] and contact dermatitis (both irritant and allergic),[6] among others.[1,4,7-10] Of note, one of our patients developed KVE while being treated with prednisone 60 mg per day and cyclophosphamide 150 mg per day for pemphigus foliaceus [Figure 4]. Another developed it while on extracorporeal photopheresis for Sezary syndrome.

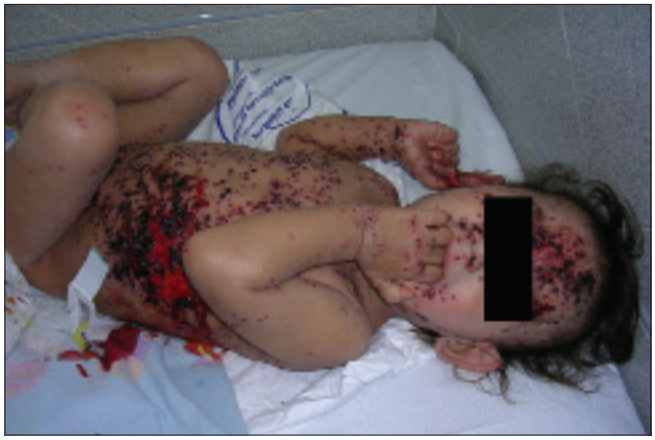

Figure 1: Widespread clusters of vesicles and erosions covered by hemorrhagic crusts

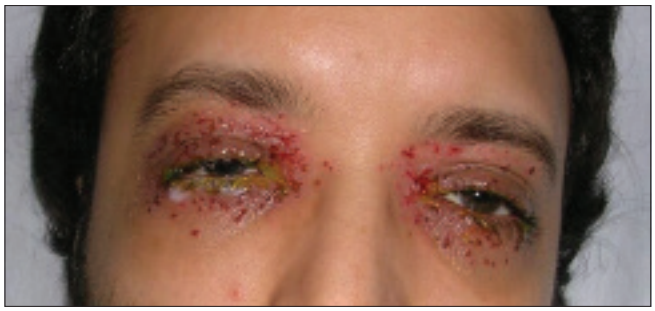

Figure 3: Kaposi's varicelliform eruption affecting conjunctival mucosa and periocular area

Although the pathogenesis of KVE is not completely elucidated, it is well recognized that the patients with diseases with underlying skin barrier damage and those with immune deficiencies are the most susceptible.[1] It is believed to be due to humoral and cellular immunity dysfunction.[5,7] In a study that assessed the growth of HSV I in cultured skin of patients with atopic dermatitis and psoriasis, faster replication than in normal skin[8] was observed. Decreased production of some cytokines, such as interferon or CXCL 10/IP B 10, or elevated levels of serum IgE have also been associated with a higher risk of developing KVE.[11] Atopic patients would have an impaired immunity against the virus mediated by T cells, a defect in the specific antibody to the virus and a decrease in NK cells and IL-2 receptors. They would also show inhibition of the Th1 response due to increased level of IL4, decreased

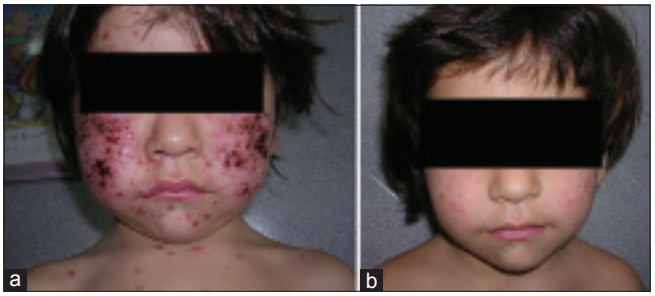

Figure 2: (a) Kaposi's varicelliform eruption involving a child face. (b) Significant scars after the acute period

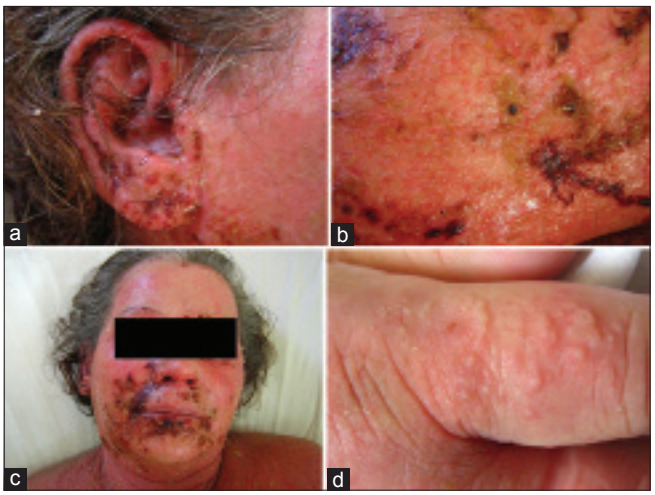

Figure 4: Kaposi's varicelliform eruption over pemphigus foliaceus. In a clockwise direction: Erosions covered by hematic crusts on (a) Right ear, (b) abdomen, and (c) face; (d) clusters of vesicles on right index finger

Table 1: Clinical features of patients with Kaposi's varicelliform eruption

Age	Sex	Associated dermatosis	Mucosal involvement	Treatment	Complications
0-6 years: 12 patients	Male 10 patients	Atopic dermatitis: 16 patients	Oral: 1 patient	Acyclovir: 19 patients	Scars: 2 patients
11-15 years: 3 patients		Psoriasis: 1 patient			
34-55 years: 4 patients	Female 9 patients	Sezáry syndrome: 1 patient	Conjunctival and genital: 2 patients	Systemic antibiotics: 10 patients	Relapses: 5 patients
		Pemphigus foliaceus: 1 patient			

levels of cathelicidins and plasmacytoid dendritic cells producing interferon-gamma, a cytokine with known antiviral effects.[1,5,8] The source of KVE would be HSV I infection in family members or close contacts and/or endogenous recurrent infection.[9] It has been speculated that HSV may enter due to the overexpression of adhesion molecules in the affected skin of KVE.[12]

It is more common in children, mainly because of its relationship with AD, although cases in healthy adults have also been reported.[1,13] Sixteen of our patients had AD.

Most cases are sporadic,[11] although an outbreak in a dermatology ward has been recently described.[14] In our study we included four contemporary cases within two families: Two were siblings with severe AD, both under treatment with cyclosporine and two were previously healthy cousins who made their debut of AD immediately before their KVE.

There has been a significant increase in the communications of KVE cases since 1980 due to the drastic increase in HSV infection in the world.[8] This phenomenon took particular effect in Japan in 2007, with yet no known cause.[11] Some authors relate this increase to the use of corticosteroids and topical tacrolimus in AD.[8,15]

Lately, KVE has been related to the use of methotrexate and retinoids in patients with psoriasis, Darier, and Hailey–Hailey.[8] The term psoriasis herpeticum has been proposed to describe KVE seated on psoriasis.[8] Our series included a 15 year old girl with a previous diagnosis of psoriasis with lesions located only on the buttocks.

Recently, Mathes *et al*. proposed the term eczema coxsackium to refer to a KVE caused by Coxsackie virus A 16.[16]

KVE is characterized by vesicopustules, some umbilicated and other eroded, and extended in clusters.[3] It may also show hemorrhagic crusts with an erythematous base. The most commonly affected sites are head, neck, and trunk. The lesions are painful[7] and they are often associated with fever, malaise, and regional lymphadenopathy.[1,7,8] The average disease duration is 16 days and most of the lesions heal without scarring within 2–6 weeks.[8]

The diagnosis is mainly clinical. In case of doubt, the Tzanck smear allows a diagnostic approach, that is rapid and economical,[1,4] though not specific. It is and its positivity rules out smallpox and its vacccine, booming because of bioterrorism.[8] Detection of viral DNA by PCR, electron microscopy, and immunofluorescence[1] as well as sampling of vesicular fluid for culture and viral serology[3,4] and histopathological biopsy[7,9] could also be performed.

Differential diagnoses are chickenpox, impetigo, contact dermatitis,[1] and smallpox vaccine could also be done.[8]

The most common complications are both bacterial infection that can lead to sepsis and viremia, with involvement of other organs. Therefore, prophylactic antibiotic therapy is recommended.[1] Viremia could affect lungs, liver, brain, gastrointestinal tract, and adrenal glands.[4] Ocular involvement may lead to loss of vision.[1] The bacterial agent most commonly found in secondary infected erosions is *Staphylococcus aureus*, although beta-hemolytic streptococci and pseudomonas may also be isolated.[5]

Disseminated cutaneous herpes simplex in childhood, often seen in association with AD may be associated with bone marrow suppression and disseminated intravascular coagulation, presumably caused by viremia.[5] When general health is affected, a complete laboratory study including blood count, metabolic profile, and liver function studies should be made to rule out systemic involvement.[3] At the present time, there is no standard therapeutic regimen for KVE. Patients treated with acyclovir (10–15 mg/kg/day), intravenous or oral, resolved the lesions faster than patients treated with placebo. For outpatients the administration of oral valacyclovir had a higher bioavailability.[8] This treatment is very effective and should be instituted without delay to prevent significant morbidity and mortality.[3] Some authors add vidarabine ointment to the oral acyclovir therapy.[11,15] In recalcitrant cases, foscarnet is the agent of choice.[8] After healing, suppressive antiherpes treatment is recommended.[3] Vaccinia immunoglobulin is recommended in cases of eczema vaccinatum.[8] Systemic antibiotics are used to control the bacterial colonization.[9] Relapses may occur, which should be recognized.[5,9]

The highlights of our case series is the low frequency of the disease and the unusual presentation of cases in the same family. We believe it is important to recognize this entity and to establish early treatment in order to avoid complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

The authors declare that there are no conflicts of interest, that the manuscript has been read and approved by all the authors, that the requirements for authorship have been met, and that each author believes that the manuscript represents honest work.

REFERENCES

1. Ugalde MG, López-Cepeda LD. Erupción variceliforme de Kaposi en un paciente adulto. Piel (Barc) 2012;27:304-6.
2. Garg G, Thami GP. Psoriasis herpeticum due to varicella zoster Virus: A Kaposi's varicelliform eruption in erythrodermic psoriasis. Indian J Dermatol 2012;57:213-4.
3. Olson J, Robles DT, Kirby P, Colven R. Kaposi varicelliform eruption (eczema herpeticum). Dermatol Online J 2008;14:18.
4. Bestue M, Cordero A. Kaposi's Varicelliform eruption in a patient with healing peribucal dermabrasion. Dermatol Surg 2000;26:939-40.
5. Kosann MK, Fogelman JP, Stern RL. Kaposi's varicelliform eruption in a patient with Grover's disease. J Am Acad Dermatol 2003;49:914-5.
6. Paradisi A, Capizzi R, Guerriero G, Rotoli M, Bussoletti C, Amerio PL. Kaposi's varicelliform eruption complicating allergic contact dermatitis. J Am Acad Dermatol 2006;54:732-3.
7. Kucukyilmaz I, Alpsoy E, Yazar S. Kaposi's varicelliform eruption in association with rosacea. J Am Acad Dermatol 2004;51(Suppl):S169-72.
8. Santmyire-Rosenberger BR, Nigra TP. Psoriasis herpeticum: Three cases of Kaposi's varicelliform eruption in psoriasis. J Am Acad Dermatol 2005;53:52-6.
9. Nath AK, Sori T, Thappa DM. A case series of Kaposi's varicelliform eruption in dermatology in-patients in a tertiary care centre. Indian J Dermatol 2011;56:110-5.
10. Gupta M, Huang V, Linette G, Cornelius L. Unusual complication of vemurafenib treatment of metastatic melanoma: Exacerbation of acantholytic dyskeratosis complicated by Kaposi varicelliform eruption. Arch Dermatol 2012;148:966-8.
11. Kimata H. Rapidly increasing incidence of Kaposi's varicelliform eruption in patients with atopic dermatitis. Indian J Dermatol Venereol Leprol 2008;74:260-1.
12. Abreu-Velez AM, Klein AD 3rd, Howard MS. Junctional adhesion molecule overexpression in Kaposi varicelliform eruption skin lesions - as a possible herpes virus entry site. N Am J Med Sci 2010;2:433-7.
13. Joshi A, Sah SP, Agrawal S. Kaposi's varicelliform eruption or atypical chickenpox in a normal individual. Autralas J Dermatol 2000;41:126-7.
14. Rao GF, Chalam KV, Prasad GP, Sarnathan M, Kumar H. Mini outbreak of Kaposi's varicelliform eruption in skin ward: A study of five cases. Indian J Dermatol Venereol Leprol 2007;73:33-5.
15. Kimata H. Kaposi's varicelliform eruption associated with the use of tacrolimus ointment in two neonates. Indian J Dermatol Venereol Leprol 2008;74:262-3.
16. Mathes EF, Oza V, Frieden IJ, Cordoro KM, Yagi S, Howard R, *et al*. "'Eczema coxsackium" and unusual cutaneous findings in an enterovirus outbreak. Pediatrics 2013;132:e149-57.