Why is it So Important to Recognize Kaposi's Varicelliform Eruption Early? FT Demir¹, D Demir¹, N Dalgıc², I Kivanc Altunay I¹, Yalcın O³

ABSTRACT

Kaposi's varicelliform eruption or eczema herpeticum is a viral infection developed on the grounds of a present skin disease. It is frequently seen on grounds of atopic dermatitis. Although it is generally related to herpes simplex virus type 1 and 2, rarely coxsackievirus or vaccinia virus play a role. In some patients, it threatens life by causing serious complications such as herpetic keratitis, encephalitis or sepsis. This case report presents a 4-month-old infant patient who was consulted by infant infection clinic for umblical vesicular eruption and who was diagnosed with Kaposi's varicelliform eruption and in whom encephalitis was detected. Following treatment, the patient was evaluated in terms of dermatologic diseases which might cause KVE and diagnosed with atopic dermatitis. Our purpose is to remind the Kaposi's varicelliform eruption which threatens life and might cause serious complications and in which early diagnosis and treatment are quite important and to emphasize the importance of investigating the underlying dermatologic disease. In addition, our case is the Kaposi's Varicelliform Eruption case which occurred at the youngest age which is reported in the literature.

Keywords: Kaposi's varicelliform eruption, herpes simplex encephalitis

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INTRODUCTION

A case of Kaposi's varicelliform eruption (KVE) in a patient who was complicated by encephalitis was reported, characterized by erythematous, partly haemorrhagic, umblical vesicular lesions on face and front side of the body. Herpes simplex virus type 1 (HSV-1) DNA in cerebrospinal fluid using PCR assay was detected and was diagnosed as herpes encephalitis. The acyclovir dosage was adjusted for pediatric dose for HSV encephalitis. The diagnosis of atopic dermatitis was also made after the diagnoisis of Kaposi's varicelliform eruption, in terms of evaluating the underlying disease. Our aim is to remind that Kaposi's varicelliform eruptions is a life-threatening disease leading to fatal complications such as encephalitis and sepsis and to emphasize the importance of evaluation in terms of the underlying disease.

CASE REPORT

A 4-month-old male infant patient was consulted by infant infection clinic for blisters on his face, inside his mouth and upper parts of his body. In his history, it was found out that eruption occurred one week ago along with complaints such as fever, unrest and reduced breastfeeding. In the background of the patient, it was found out that he was born mature following a pregnancy without complications with caesarean section weighing 4000 gr and no pathology was detected in postnatal period follow-up examinations. He had no previous history of another diagnosed dermatologic disease. The mother stated that the baby has no skin related problems other than pruritus and skin drying. In his family history investigation, his mother had allergic rhinitis and asthma history. In the physical examination of the patient; fever: 38^oC, respiratory rate: 30, heart rate: 98 detected, there was servikal LAP and he was found to be prone to sleep. In his dermatologic examination, there were erythematous, partly

haemorrhagic, umblical vesicular lesions which are most common on perioral area but localized on face, lips, both buccal mucosa and front side of the body and grouped especially on perioral area (Figure 1a and 1b). In Tzanck test conducted on lesions, multinuclear giant cells were detected along with achantosis. With these findings, the patient was diagnosed with Kaposi's Varicelliform Eruption. In laboratory examinations of the patient, leukocyte number was 13.51/mm³, AST: 64.4 and C reactive protein: 29. The patient, who had been considered to have encephalitis due to complaints such as reduced breastfeeding, tendency to sleep and unrest, was found out to have HSV-1 DNA in cerebrospinal fluid in PCR assay, he was diagnosed with herpes encephalitis. Intravenous (IV) acyclovir 3x10 mg/kg/day was administered for 21 days. Herpes encephalitis of the patient was healed without leaving any sequel and lesions regressed by leaving post-inflammatory hyperpigmentation (Figure 2). When the patient was evaluated in terms of dermatologic diseases which may cause KVE, it was detected that total IgE level was 956 IU/mL. Specific milk and egg IgE levels were high. The patient who was monitored together with infant allergy clinic was evaluated as atopic dermatitis.

DISCUSSION

KVE was first defined by Moritz Kaposi in 1887 (1). Although it is frequently related to herpes simplex virus (HSV) type 1 and 2, it is an acute viral disease in which coxsackievirus or vaccinia virus are effective rarely (1). The most important risk factor in the occurrence of the disease is the impairment of the epidermal barrier (1). Although it is most frequently seen on the grounds of atopic dermatitis, it is correlated in literature with Darier's Disease, psoriasis, tinea cruris, allergic contactdermatitis, ichtyosis vulgaris, pemphigus vulgaris, pemphigus foliaceous, pityriasis rubra pilaris, Hailey-Hailey Disease, topical tacrolimus and pimecrolimus use (2,3). The virus is transmitted with contact with infected lesions or a latent infection in host (2). Following an incubation period of 5-19 days, ventricose or punched looking, ventricose vesiculospustules which might me haemorrhagic start to be seen on head and upper part of the body. Systemic symptoms such as fever, unrest, lack of appetite and vomiting might accompany. It rarely threatens life by causing serious complications such as herpetic keratitis, hepatitis, encephalitis, meningitis and sepsis. Mortality rate is reported as 6-10% (4). Starting treatment within 24-48 hours reduces the development of complications (5). Tzanck test from skin ground or direct fluorescent antibody test in which viral culture made of fresh vesicular fluid, swab cultures taken from ulcerative lesions are used may be used in diagnosis in addition to clinical findings. In final diagnosis, the most reliable method is to produce virus in the sample taken from skin lesions (1). In our case, we applied Tzanck test to verify the diagnosis as it is a fast, economic and simple method.

The age of HSV infection is between 2 and 5 years of age (4). It is most frequently presented on children with skin and mucosa infections such as gingivostomatitis, herpes labialise, recurrentstomatitis and keratoconjunctivitis. Rare viremia and systemic organ and central nervous system involvement might be observed. It might cause systemic complications such as sepsis, hepatitis, encephalitis, meningitis. Incidence of encephalitis is 2.5-4 cases out of a million and it does not exhibit any gender and seasonal difference (6). It is generally developed in children and young adults depending on primary disease (7). It has a prodromal period of 2-3 days which starts with fever and headache. Later it might end up with psychotic behavioural disorders, epileptic seizures, hemiplegia, speech disorders, amnesia, stupor and coma (8). Mortality and neurological sequel development rate is reduced significantly with early treatment (9). While mortality rate is 70% in untreated cases, it is possible to have 92% success when early treatment is initiated (7).

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KVE is generally seen between ages 10 and 30. Our case was a 4-month-old infant. In the literature, it is the youngest case diagnosed with KVE. Few cases were reported to develop herpes encephalitis. In order to prevent complications, the most important step in treatment is to start systemic antiviral treatment in the first days of infection (1, 10). It is necessary to increase treatment duration and dose in cases in which central nervous system involvement is observed.

As a conclusion, KVE is a disease which may cause serious complications such as blindness secondary to herpetic keratitis, encephalitis, sepsis and death, which is accompanied by an underlying dermatosis and responds to systemic antivirals well. To know the characteristic dermatologic manifestations of the disease is very important for early diagnosis. Early diagnosis is extremely important to start treatment early to prevent occurrence of fatal complications. It is also important to examine the patients for underlying dermatological diseases. In our case, atopic dermatitis diagnosis was made after KVE diagnosis.

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Fig 1: Erythematous, partly haemorrhagic, umblical vesicular lesions on face and front side of the body.



Fig 2: A photo of the patient after the treatment