Kaposi varicelliform eruption in a patient with epidermolysis bullosa simplex generalized severe

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INTRODUCTION

Inherited epidermolysis bullosa (EB) are genodermatoses characterized by the formation of blisters after minor trauma. There are 4 major types of inherited EB[1]: EB simplex, junctional EB, dystrophic EB, and Kindler syndrome. Classification is performed according to the mode of inheritance, the phenotype, the level of skin cleavage found by immunohistochemical analysis, and the type of mutation.

EB simplex generalized severe, which was named Dowling-Meara EB, is the most serious form of EB simplex.

The first lesions occur at birth as superficial arciform blistering over the entire body surface, typically after trauma or heat exposure. Oral lesions can also occur. The clinical manifestations are worse in childhood and decrease in adulthood. Onychodystrophy is often associated with the disease, and keratoderma of the palms and soles can appear after the age of 6.

This rare genodermatosis is characterized by autosomal dominant transmission. Complications may involve the gastrointestinal tract, with consequences for growth and nutrition, and the ocular, genitourinary, and respiratory tracts. Skin complications include pruritus and superinfection.

Bacterial and viral infections are frequently reported. Nevertheless, to our knowledge, no case of Kaposi varicelliform eruption in the context of EB has been published. Kaposi varicelliform eruption is a diffuse herpes simplex virus (HSV) infection of preexisting skin lesions. The most common association is with atopic dermatitis. This is a serious complication that can be life threatening.[2] Here, we present a case of a child with persistent fever of unclear origin resulting from a herpetic infection of generalized EB simplex.

Abbreviations used:

EB: epidermolysis bullosa
HSV: herpes simplex virus

CASE REPORT

An 11-month-old child with generalized EB simplex was hospitalized for hyperthermia. The child and his family had no history of atopic dermatitis. Since birth, he presented with blisters of the oral cavity, hands, and feet followed by blisters on the whole body, thickened nails, and trumpet nails deformity. He had a family history of EB simplex generalized severe; his father also had the condition diagnosed at birth.[3] Histopathologic examination found intraepidermal cleavage within basal keratinocytes. Immunofluorescence antigen mapping found a normal positivity for keratin 5, laminin 332, and collagen 7. Laminin 332 and collagen 7 were located on the base of the blister, and keratin 5 was expressed both on the roof and base of it, leading to the diagnosis of EB simplex generalized severe. As he grew, he usually presented with 20 to 30 blisters every 2 days on the hands, feet, and trunk, with mucosal involvement (tongue and palate) and onychodystrophy.

Fever (39°C) was present for 5 days. The patient was hypotonic. He also had diarrhea. Clinical

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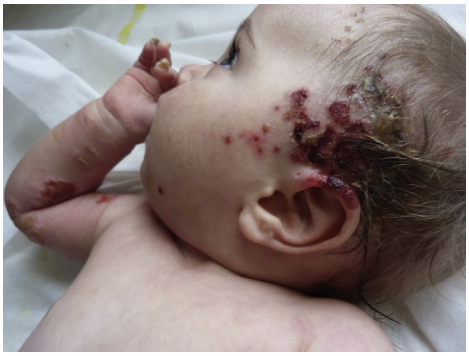

Fig 1. Kaposi varicelliform eruption in a patient with generalized EB simplex. A dome-chapped blister on the left cheek and hemorrhagic crusting of typical blisters.

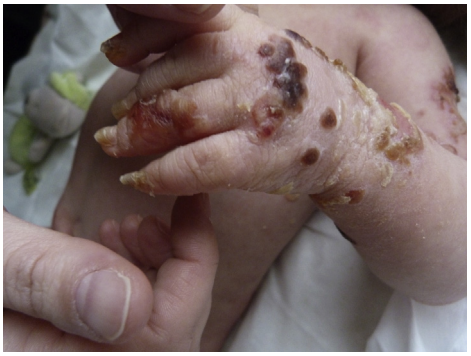

Fig 2. Kaposi varicelliform eruption in a patient with generalized EB simplex. Previous blisters with a hemorrhagic transformation and yellow crusts on the left hand and onychodystrophy (thickened nails and trumpet nails deformity).

examination excluded otitis media or any other type of ear, nose, or throat infection; pulmonary auscultation was unremarkable (Figs 1 and 2). Findings on cytobacteriologic examination of the urine were normal, and blood cultures were negative for infection. A blood test found an inflammatory reaction, with a high C-reactive protein level of 40 g/L, but no other abnormalities. Viral stool cultures were negative, and an abdominal ultrasound scan was normal. The patient was initially treated with ciprofloxacin and amoxicillin clavulanic acid for bacterial superinfection.

The depression in the center of the blisters on the left cheek and left hand suggested that a herpetic superinfection was possible. These blisters were not located exactly on the same site of EB lesions. Systemic antiviral treatment with acyclovir, 10 mg/kg every 8 hours, was started in association with amoxicillin-clavulanic acid. The diagnosis was confirmed by polymerase chain reaction, which indicated the presence of herpes simplex virus type 1 in the lesions on the left cheek. Local bacteriologic samples showed the presence of *Staphylococcus aureus* and *Streptococcus dysgalactiae*. Treatment was successful and was accompanied by apyrexia within 24 hours of initiation.

Because of the child's diagnosis, the close relatives were examined, and the mother was clinically positive for HSV. She was treated with valacyclovir to prevent subsequent recurrences or other complications.

DISCUSSION

This case report is an original presentation of Kaposi varicelliform infection in a patient with generalized EB. HSV is highly prevalent and endemic throughout the world.[4] Kaposi varicelliform eruptions are potentially life-threatening infections caused by HSV infection over a preexisting dermatosis. These eruptions often manifest as febrile eruptions of dome-chapped blisters and hemorrhagic crusting but may also present as erosions without vesicles or pustules and worsening of a preexisting dermatosis. The upper body is the most common site of infection, with a predilection for the head and neck. In some cases, the infection may progress to a fulminating, life-threatening stage and can have severe sequelae, including herpes keratitis, disseminated infection with visceral involvement, and death. The most common form is eczema herpeticum on atopic dermatitis. Although eczema herpeticum is rare, a subset of patients with atopic dermatitis seems to have a predisposition for HSV infection because of defects in specific proteins critical for skin barrier function[5] and innate immunity.[6] Other cases have been described in conjunction with Grover's disease, bullous pemphigoid, Darier disease, Hailey-Hailey disease, burns, and Staphylococcal scalded skin syndrome.[7]

To our knowledge, only 1 other article reported a case of herpetic infection in a child affected with EB.[8] The patient presented a gingival-stomatitis in a dystrophic EB.

In a multicenter observational study of hospitalized children with eczema herpeticum,[9] the delayed initiation of acyclovir therapy was associated with increased length of stay. This study's findings emphasize the importance of early acyclovir therapy for children with eczema herpeticum. Our patient was hospitalized for 12 days, and acyclovir therapy was initiated after 5 days of hospitalization. The case highlights the importance of the diagnosis of herpetic infection of EB lesions in the case of an unclear source of fever because the best prognosis is correlated with early diagnosis and prompt treatment. However, a prompt diagnosis of an

infectious blistering disease on top of a mechanical blistering disease can be tricky to discern.

Our case also highlights the importance of preventing herpetic superinfection in children with EB. Like in atopic dermatitis, we have to prevent these infections. Informing parents of the risk of contagion must become routine. Prophylactic treatment of the parents should also be discussed to avoid transmission. In recent guidelines for the management of inherited EB,[10] chicken pox vaccination is recommended, but currently there is no recommendation for the prevention of herpetic infections. Our case highlights the importance of diagnosing and preventing herpetic superinfection in children with EB, a probably underestimated complication of this disease.

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