Pemphigus Foliaceus Complicated by Kaposi Varicelliform Eruption

A Kaposi varicelliform eruption (KVE) refers to a widespread cutaneous infection with a virus that often causes local vesicular eruptions over a pre-existing skin disease. Pemphigus foliaceus complicated by KVE is rare. We report the case of a 68-year-old Chinese woman with pemphigus foliaceus complicated by KVE.

A 68-year-old woman was admitted to our hospital with a 14-month history of erythema and bullae distributed over the entire body. Fourteen months prior to evaluation in our hospital she was diagnosed with pemphigus and was prescribed prednisone as primary therapy. One month prior to evaluation in our hospital more severe lesions recurred because of the sudden discontinuation of the prednisone treatment. She denied any systemic diseases. On physical examination, multiple erythematous erosions, crusts, and scales were noted on the face, trunk, and upper limbs (Figure 1, a). No bullae were present and the oral mucosa was not involved. Routine blood and urine testing, liver and kidney function, and blood glucose and lipid levels were normal. A biopsy of thoracic lesions revealed acantholysis on the upper spinous layer (Figure 1, b). Direct immunofluorescence (DIF) demonstrated immunoglobulin G (IgG) and immunoglobulin M (IgM) deposits in the epidermal intercellular space (Figure 1,c). Indirect immunofluorescence (IIF) showed circulating antibodies directed against the intercellular substance (titer=1:1280). A diagnosis of pemphigus foliaceus was made. Intravenous methylprednisolone (80 mg qd) combined with intravenous immune globulin 20g qd (0.4 g/kg/d for the first 3 days) was administered. After ten days, the lesions gradually resolved.

On the 11th day, however, the patient reported malaise and complained that the facial lesions had suddenly worsened. On physical examination, a widespread eruption of several closely-grouped, painful blisters and crusts on the erythematous skin of the face and chest typical of a herpetic infection

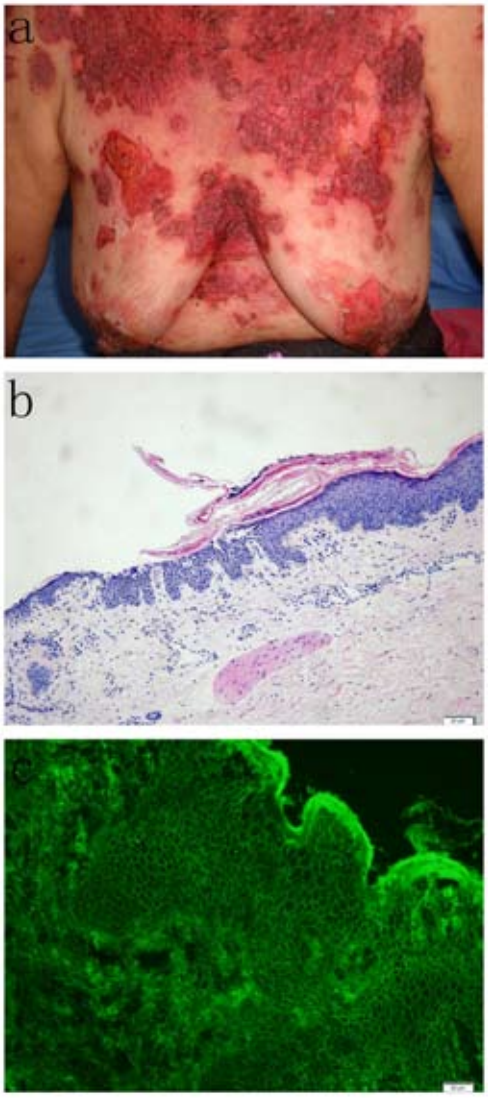

Figure 1. a) Erythema, erosions, crusts on the chest. b) Dermatopathology revealed acantholysis on the upper spinous layer (hematoxylin and eosin ×100), c) Direct immunofluorescence (DIF) demonstrated immunoglobulin G (IgG) and immunoglobulin M (IgM) deposits in the epidermal intercellular space.

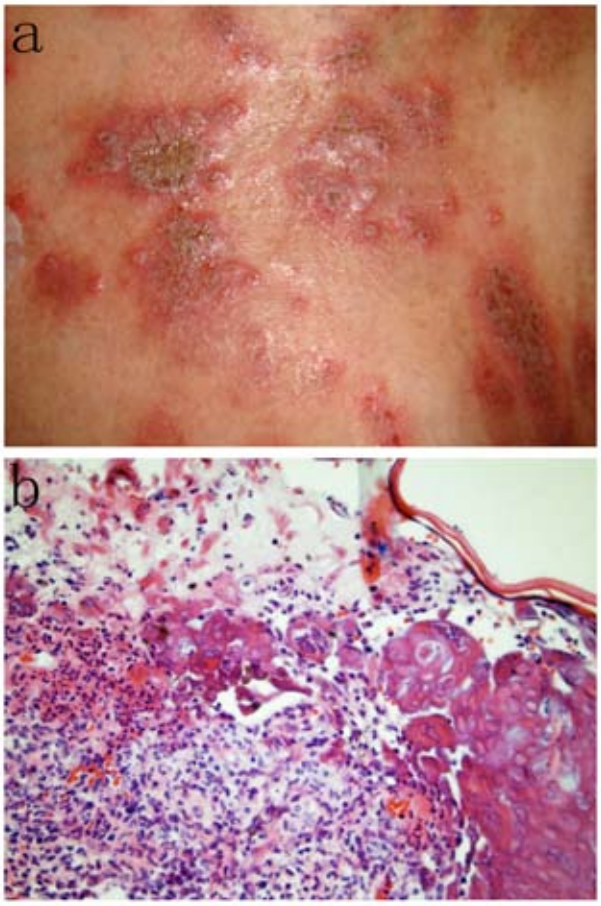

Figure 2. a) A widespread eruption of several grouped blisters on the erythema on the chest. b) A biopsy from blisters on the chest revealed marked acantholysis with an intraepidermal vesicle, ballooning, and several multinucleated epithelial giant cells (hematoxylin and eosin ×200).

was noted (Figure 2, a). A biopsy of the chest revealed marked acantholysis with an intraepidermal vesicle, ballooning, and several multinucleated epithelial giant cells (Figure 2, b), which was ascribed to a herpes virus infection. Viral polymerase chain reaction (PCR) of the swab revealed herpes simplex virus type 1 (HSV-1). A diagnosis of KVE was established. In addition to methylprednisolone, oral valacyclovir (300 mg bid for 6 days) and intravenous immune globulin (10 g qd for the first 3 days) were administered. The KVE lesions subsided ten days later.

KVE is a vesicular dermatosis caused by a viral infection in patients with a pre-existing skin disease. These viruses include HSV-1 and HSV-2, and rarely Coxsackie A16 and vaccinia viruses. The most common pre-existing dermatosis for KVE is atopic dermatitis, but KVE has been reported in many other dermatoses (1), such as autoimmune bullous dermatoses. The typical manifestation of KVE is the sudden appearance of monomorphic, umbilicated, and grouped vesicular lesions on the face, neck, axillae, chest, and upper extremities, usually accompanied by fever and malaise.

The pathogenesis of KVE is unclear. Impaired skin barrier function in the above-mentioned diseases facilitates the spread of the viral infection (2). Furthermore, these patients, in whom the immune system is greatly suppressed due to the extensive application of immunosuppressive drugs, are more susceptible to different microbial infections, including viruses.

In 1963, Marton and Angyal (3) reported the first case of KVE concurrent with pemphigus foliaceus. In fact, only a few cases of pemphigus foliaceus complicated by KVE have been reported (2,4-6). Among the reported cases, one case had a poor prognosis because of multiple organ failure (6). The diagnosis of KVE is sometimes difficult to make when the pre-existing disease is pemphigus since the new lesions may be interpreted as deterioration or relapse of pemphigus, which is when immunosuppression should be enhanced.

In summary, early diagnosis and prompt systemic antiviral treatment for KVE before lab confirmation is crucial, especially in elderly patients with autoimmune bullous diseases, so as to limit disease duration and prevent further complications. Dermatologists should make a correct judgment and manage this condition appropriately, based on clinical manifestations.

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