

Eczema Herpeticum in Darier's Disease

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Eczema herpeticum is a herpes simplex virus infection with disseminated skin involvement superimposed on a pre-existing dermatosis. Darier's disease has been reported to be among the dermatoses susceptible to the sudden onset of a widespread vesicular eruption accompanied by high fever known as Kaposi's varicelliform eruption.

We report a case of eczema herpeticum in a 46-year-old woman associated with Darier's disease. (Ann Dermatol 10:(1) 32~34, 1998).

Key Words : Eczema herpeticum, Darier's disease

Eczema herpeticum is the clinical picture of widespread cutaneous infection by the herpes simplex virus, usually in patients with pre-existing skin disorders such as atopic dermatitis, neurodermatitis, ichthyosiform erythroderma, Wiskott-Aldrich syndrome, seborrheic dermatitis, irritant contact dermatitis, pemphigus foliaceus, cutaneous T-cell lymphoma and Darier's disease. The occurrence of eczema herpeticum in patients with Darier's disease is reported relatively rarely. To our knowledge, only two cases have been reported in Korea^{1,2}.

It is not known why patients with Darier's disease are predisposed to the development of eczema herpeticum, but disturbances in cellular immunity, photoactivation of the virus or the provision of a suitable medium for viral growth in active lesions have been suggested as doubtful mechanisms^{3,4}.

This report describes a case of eczema herpeticum due to the herpes virus in a patient with Darier's disease confirmed by skin biopsy and clinical features.

CASE REPORT

A 46 year-old woman, who had brown hyperkeratotic papular skin lesions present for over 10

years on the neck, anterior chest and lower extremities, was admitted to our department of dermatology. Four days before admission, the patient noted multiple umbilicated vesicles on the anterior chest where Darier's lesions had been previously present. The lesions extended to her cheek and entire face. The patient developed a chill, fever, myalgia and cervical lymphadenopathy.

A physical examination revealed a grouped umbilicated vesicular eruption on her face (Fig. 1) and follicular hyperkeratotic papules on her neck, anterior chest and lower extremities (Fig. 2). Family history revealed that none of her family members had the similar skin eruptions.

A microscopic examination of a Tzanck preparation taken from the base of a fresh vesicle showed multinucleated giant cells. Results of laboratory studies, including a complete blood count, electrolytes and blood chemistry, were within normal ranges. The result of a cell mediated immunity test (multi-CMI test kit) showed decreased immunity. A chest roentgenography was insignificant.

A biopsy specimen from an active skin lesion on the cheek showed a circumscribed area of ballooning degenerating epidermal cells, the formation of multinucleated giant cells, acantholysis and necrosis with ulceration (Fig. 3). These histologic alterations were considered to be characteristics of a herpetic vesicle. Also a biopsy specimen taken from an old papular lesion on the right knee revealed typical characteristics of Darier's disease such as hyperkeratosis, focal parakeratosis and

Received September 1, 1997.

Accepted for publication December 2, 1997.

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Fig. 1. erosive papules and grouped, umbilicated vesicular eruptions on the right cheek which extended from anterior chest lesions.

Fig. 2. erythematous papules on the right knee.

Fig. 3. A high power view of a biopsy specimen of the vesicular lesion on the face shows ballooning degeneration of epidermal cells and infiltration of multinucleated giant cells. (H & E stain, $\times 100$).

suprabasal lacunae. There were dyskeratotic cells with formation of corps ronds and grains(Fig. 4).

The patient was admitted to the hospital for systemic antiviral therapy. After admission intravenous acyclovir was given to the patient at a dosage of 750 mg/day, administered over a two-hour period three times a day. The patient was discharged in good condition after eight days, with slight improvement of her lesions of Darier disease.

DISCUSSION

Darier's disease is a papulosquamous disorder with slow progression in severity with age and frequently involves seborrheic areas such as the face,

Fig. 4. A high power view of a biopsy specimen of papular lesions on the lower extremities shows suprabasal lacunae and dyskeratotic cells such as corps ronds and grains. (H & E stain, $\times 100$).

anterior chest, back, and over the scrotum. Patients with Darier's disease frequently suffer from secondary skin infections of bacterial or occasionally viral origin and have been rarely reported to be susceptible to eczema herpeticum⁵. The pathophysiological mechanism of Darier's disease has not been completely understood, but decreased delayed hypersensitivity is found after in vivo skin testing and in vitro transformation studies. These results may explain the infectious complications of

Darier's disease^{6,7}.

Eczema herpeticum is the dissemination of the herpes simplex virus over the surface of the body which complicates pre-existing cutaneous diseases. This widespread dissemination occurs following direct inoculation of the herpes simplex virus into damaged stratified squamous epithelium, resulting in large numbers of primary colonizations. The commonest pre-existing cutaneous diseases are atopic dermatitis and neurodermatitis. However, other cutaneous diseases such as Darier's disease may serve as a background⁸.

The reason why eczema herpeticum occurs in patients with Darier's disease is not exactly known. However, it may be considered that patients with immunodeficiency are more susceptible to herpes simplex infections, possibly due to defective cell-mediated immunity and disturbances in cell mediated immunity are occasionally found in patients with Darier's disease^{9,10,11}. Our case revealed defective cell mediated immunity compared to cell mediated immunity of a normal person.

The symptoms of eczema herpeticum consist of a sudden onset of grouped vesicles, accompanied by moderate to severe malaise and fever. The vesicles are primarily localized on the diseased skin, but may spread to the intact skin. It happens quite often that only the head or at least the upper body regions are affected, whereas only rarely can an isolated involvement of the lower parts of the body be observed. In milder cases the skin symptoms are often restricted to the upper half of the body. Normally, the lower half is involved only in generalized cases^{12,13}. On admission, the symptoms of our patient were mild in that vesicular eruption preceded by prodromal symptoms such as fever, chilling and myalgia, were restricted to the upper half of the body.

The therapeutic efficacy of acyclovir in eczema herpeticum is well known^{14,15}. Also, compared with other antiviral drugs, acyclovir seems to be remarkably free of side effects to date. Transient impairment of renal function, transient rises in transaminase levels and delirium have been reported. Therefore, acyclovir was chosen for treatment in our patient because of continued cutaneous dissemination of the herpetic lesions, and the rapid cessation in the spread of the eruption. In addition, an overall clinical improvement in the patient's condition was observed.

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