

Infantile Erosive Pustular Dermatitis of the Scalp Associated with Klippel-Feil Syndrome

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Sir,

Erosive pustular dermatosis of the scalp (EPDS) is a disease characterized by eroded, crusted and pustular lesions resulting in scarring alopecia. It occurs chiefly in elderly persons, and trauma has been implicated as one of the predisposing factors (1). We report here a case of EPDS associated with Klippel-Feil syndrome, which is a congenital anomaly of the cervical vertebrae (2).

CASE REPORT

A 6-month-old girl was brought to our hospital by her parents, who were concerned about an ulceration of the scalp. She was born at 41 weeks of gestation with no problems. At birth, she was noted to have a short neck, deafness, and defect of the left kidney. X-ray examination showed fusion of the cervical vertebrae, which led to the diagnosis of Klippel-Feil syndrome. The hair was sparser on the frontal and parietal scalp than in other regions. The lesion was associated with faint erythema and mild desquamation of the scalp skin, but not with atrophy. At 3 months of age, a skin ulcer had appeared on the parietal scalp and developed gradually. Her parents were not consanguineous or symptomatic. On physical examination, she had a superficial ulcer with pustules and crusts extending from the frontal scalp to the parietal scalp (Fig. 1a). Routine laboratory tests were normal, including the serum zinc concentration. A culture from the ulcer was positive for *Serratia marcescens*. The ulcer was treated with an ointment of fusidate sodium and local wound care, which resulted in a partial improvement. However, the girl continued to develop recurrent pustules and persistent crusts in the same area. We made a diagnosis of EPDS. Due to prophylactic therapy the bacterial infection was only secondary colonization, she was started on beclometasone dipropionate ointment (twice a day) for 2 weeks, leading to a decrease in the ulcers and pustules. After 6 weeks of treatment, the scalp lesion had healed completely, leaving a mild scar with sparse hair growth (Fig. 1b). Thereafter, 0.25% hydrocortisone ointment was applied twice daily for 4 months, because the ulcer recurred when glucocorticoid treatment was discontinued.

DISCUSSION

EPDS represents a distinct cutaneous disease despite the lack of specific clinical, histopathological and laboratory findings. As EPDS responds to glucocorticoids, some

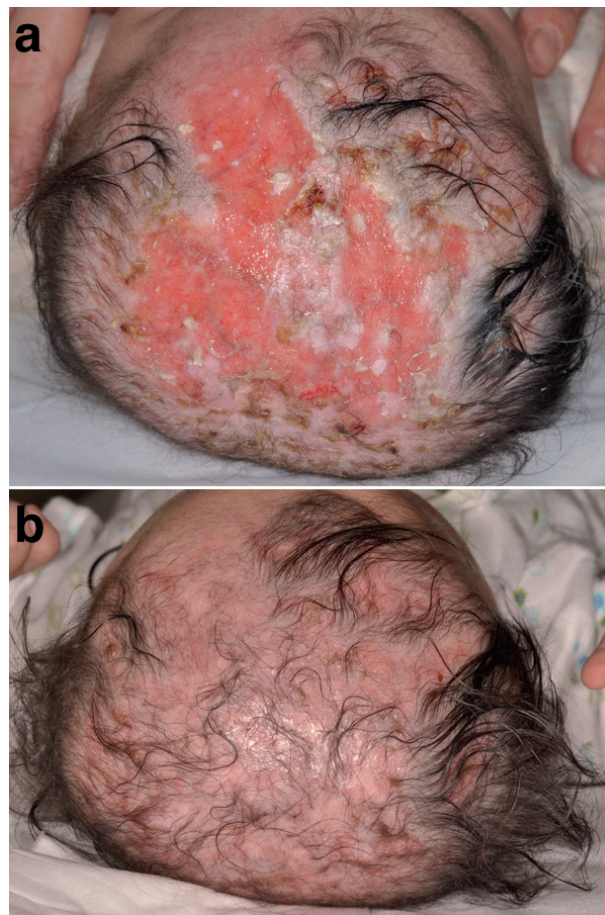


Fig. 1. Clinical feature. (a) An extensive skin ulcer on the frontal and parietal scalp at the age of 6 months. (b) Scalp skin after epithelization.

immunological dysfunctions seem to be related to its pathomechanism. Based on the clinical appearance and response to topical glucocorticoids in the present patient, we made a diagnosis of EPDS. The pathogenesis of EPDS has not been elucidated, but the skin ulcers responded to topical anti-inflammatory therapy, but not antibiotics, which suggests that the ulceration was based chiefly on inflammation (1, 3). In addition, trauma may play a role in the pathogenesis, because some patients with EPDS have a history of scalp trauma (4). Histopathologically, EPDS shows chronic inflammatory changes and foreign body giant cells around broken hair shafts. Trauma-induced damage may cause the chronic inflammation due to a foreign body reaction to the broken hairs.

EPDS occurs chiefly in elderly women. Interestingly, Siegel et al. (5) reported four cases of infantile EPDS who had necrotic caput succedaneum at birth. The

infantile cases seem to have developed in association with perinatal scalp injury. Our case did not have any history of scalp trauma, but some injury may have occurred in relation to Klippel-Feil syndrome during intrauterine life. Klippel-Feil syndrome is a rare disorder characterized by the congenital fusion of the cervical vertebrae (2). The most common signs of the disorder are short neck, lower posterior hairline, and limited neck movement. Associated abnormalities may include hearing loss, anomalies of the kidneys and ribs, respiratory problems and heart malformation, while skin anomalies have not been reported. In our case, the anomaly of the vertebrae resulted in neck stiffness, which may have contributed to injury of the scalp skin due to engagement with the uterine wall during foetal development. Alternatively, some gene mutations may cause inflammation specifically in the scalp skin. In any event, some skin anomaly had already developed prenatally, because our case showed an alteration of scalp hair, as well as erythematous skin at birth.

Infantile EPDS is uncommon and difficult to treat. Topical glucocorticoids treatment should be commenced

when bacterial and mycological organisms represent nothing more than a secondary colonization, because persistent ulceration can lead to scarring alopecia. Furthermore, EPDS has been treated with topical tacrolimus and calcipotriol, as well as oral nimesulide and isotretinoin (3).

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