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Kindler's Syndrome

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ABSTRACT

A 3-year-old girl with congenital poikiloderma had episodic blistering spontaneously or after trauma. Growth and development had been normal. Family history did not show any evidence of cutaneous disease. We believe that this case best fits the designation of Kindler's syndrome.

Key words: *Kindler's syndrome, Poikiloderma Bulla*

In 1954, Kindler[6] reported a patient who had poikilodermatous changes and episodic blistering. Since there have been very few reports of similar cases, controversy continued as to whether these cases can represent a distinct clinical syndrome. We recently encountered a 3-year-old girl with a bizarre poikiloderma and subsequent episodes of bullae.

CASE REPORT

A 3-year-old girl was the product of an apparently normal pregnancy. At birth, red patches were noted over the entire body. At about 1 month of age, she developed blisters of the thighs, feet and trunk, sometimes after trauma and sometimes spontaneously. From about 2 months of age, the red patches have been gradually replaced by a mottled and reticulated pigmentation. At approximately 8 months of age, she exhibited a most remarkable poikiloderma involving large areas of the skin. New blisters continued to appear at intervals. Each episode lasted about two weeks. Bullae ranged in size from about 0.5 to 4 cm (Fig. 1). Nikolsky's sign was negative. No scars were evident that could be attributed to previous bullae. The hair, nails, teeth, mucous membranes and eyes were unaffected. There was no abnormality of sweating. No acrokeratotic lesions were observed. The patient's general health was good. The patient's developmental milestones were normal. Various routine laboratory examinations revealed no abnormalities. Urinary porphyrin levels were normal.

A skin biopsy of the bulla from the right thigh showed epidermal atrophy, vacuolated degeneration of the basal layer, colloid bodies, telangiectasia, mild inflammation, and subepidermal bulla (Fig. 2, 3). In sections stained with the PAS technique, the PAS-positive basement membrane was located on the dermal side of the bulla. The floor of the bulla showed several basal cells adherent to the dermis (Fig. 4). Direct immunofluorescence testing revealed no deposits of immunoglobulins along the basement membrane.

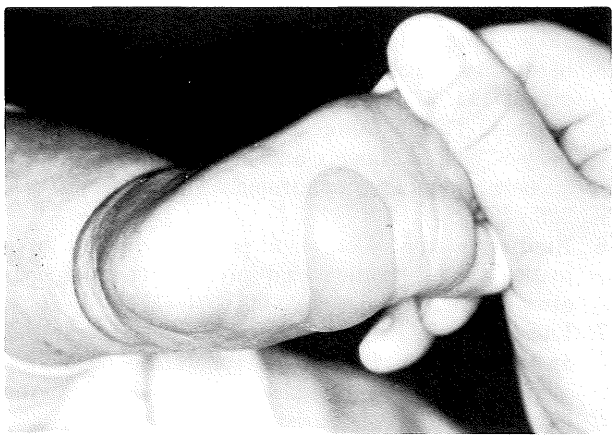

Fig. 1. Blister on the sole.

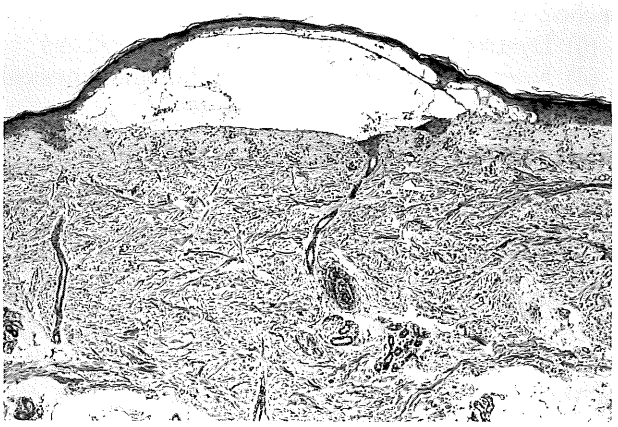

Fig. 2. Skin from thigh showing an intact bulla (hematoxylin-eosin, original magnification ×20).

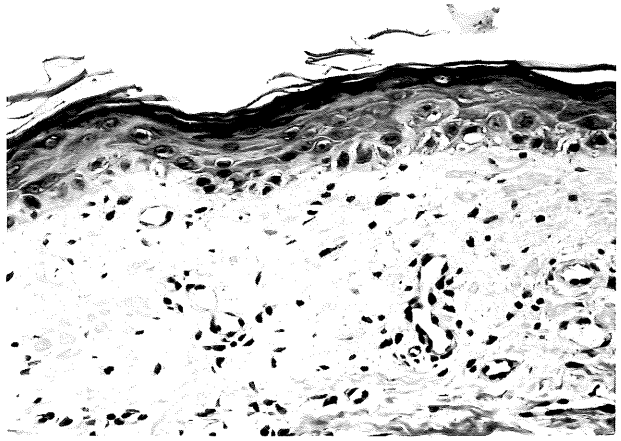

Fig. 3. Atrophy of epidermis, considerable proliferation of capillaries and mild inflammation (hematoxylin-eosin, original magnification ×100).

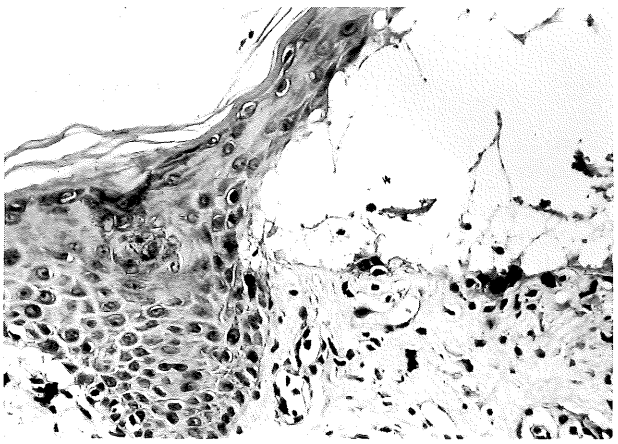

Fig. 4. The floor of the bulla showing several basal cells adherent to the dermis (hematoxylin-eosin, original magnification ×100).

There was no consanguinity among parents or grandparents.

COMMENT

In 1954, Kindler[6] described a 14-year-old girl who had cutaneous manifestations of both epidermolysis bullosa and poikiloderma congenitale. Since then, similar cases have been reported[1,2,8,10]. On the basis of the clinical manifestations, Kindler's syndrome is characterized by the following features[1,2,6,8,10]:

(1) Predominantly male

(2) Lack of evidence for an inherited condition

(3) Onset of the eruption at birth or in infancy : The eruption consisted of poikiloderma almost over the entire body.

(4) Onset of bullae at birth or in infancy : Bullae occurred especially in the hands, feet and extremities sometimes after trauma and sometimes spontaneously. Blistering was unrelated heat and sunlight, and have lessened in frequency and severity with increasing age. Histopathology of the bullae showed subepidermal bullae. Electron microscopy performed revealed anchoring fibrils attached to the basal lamina.

(5) Dystrophic nails and defective dentitions

(6) Normal hair, eyes, and mental and physical development

We believe that our case fits well to the concept of Kindler's syndorme.

There have been some other reports with similar poikilodermatous skin changes and episodic blistering[3,4,5,9,11,12]. While some cases were hereditary by an autosomal dominant or recessive factor, some had no evidence suggesting they were the results of heredity. The problem of the occurrence of these disorders in an isolated case poses the question of whether he has a nonheritable disorder or whether he should be considered either as mutations or as recessively inherited. As far as the name of the syndrome is concerned, many reporters had different ideas, depending on which aspects of the syndrome were most marked in their own cases. It is proposed that the term 'bullous and hereditary Weary-Kindler's acrokeratotic poikiloderma' may be an inclusive one applied to all these patients with similar cutaneous changes[7]. We recognize, however, that more cases need to be documented before this syndrome is completely defined.

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