Kallin's Syndrome: Two More Cases

Sir,

Kallin's syndrome was first described in 1985 and was characterized by epidermolysis bullosa simplex localisata, associated with anodontia, hair and nail disorders (1). In 1989 two siblings in a new family were admitted to the Department of Dermatology, Central Hospital, Boden, Sweden.

A boy, born in 1977, and his sister, born in 1981, had the localized epidermolysis bullosa simplex type. Their parents and family members were healthy and without inherited disorders. No relationship between this family and the one previously described could be traced. According to the parents neither inbreeding nor consanguineous marriages were found on either side of the families. The pedigree of the family is shown in Fig. 1a.

The father and mother had no history or signs of blisters, hair or nail disorders. A panoramic radiograph of the father's jaws showed that the following permanent teeth were missing: 15, 46, 47 and 28, 38. Radiograph of the mother's jaws was normal.

The boy developed at the age of 3 months poor, scanty growth of hair, which during the following years became dry and brittle with areas of non-scarring alopecia (Fig. 1b). At the age of 4 years blisters occurred spontaneously on hands and feet, leaving no scars after healing. Generally blisters occurred in the spring and summer and almost every year. Blisters were monolocular and sometimes hemorrhagic. Traumatic blisters were also observed. However, at the age of 10 years blistering decreased but did not completely disappear (Fig. 1c). Myopy and hyperhidrosis were observed when he was 4 years old, and at his first dental examination anodontia was established. A panoramic

radiograph of the jaws showed aplasia of 14, 15, 17, 24, 25, 27, 34, 35, 37, 44, 47 and 18, 28, 38, 48 of the permanent teeth (Fig. 1d). At that time some of the nails showed onychogryphosis.

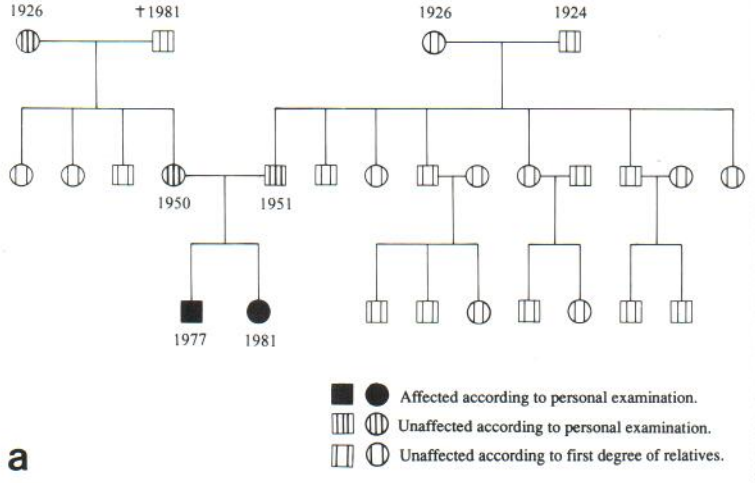
The girl was at the age of 4 years suspected to suffer from gluten intolerance. She was given a diet without gluten and lactose. Two years later, she was considered cured. At the age of 4 years, blisters occurred on hands and feet of the same type as those of her brother. Hair and nails were normal. At the same age a serious hypermetropy was found and anodontia was established in connection with the visit of her elder brother. A panoramic radiograph of the jaws showed that the following permanent teeth were missing: 14, 15, 24, 25, 27, 34, 35, 37, 44, 45, 47 and 18, 28, 38, 48.

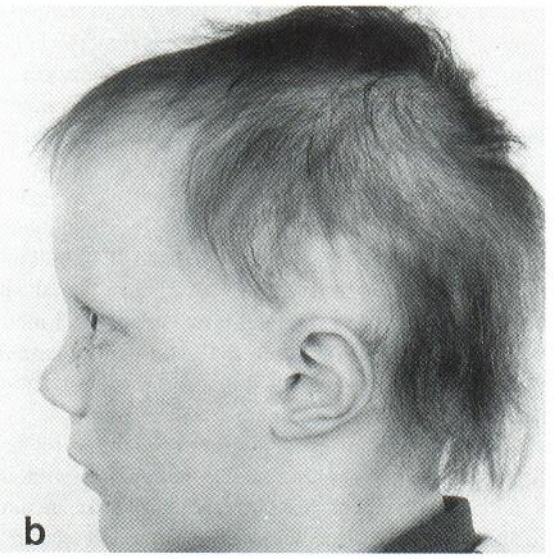
COMMENTS

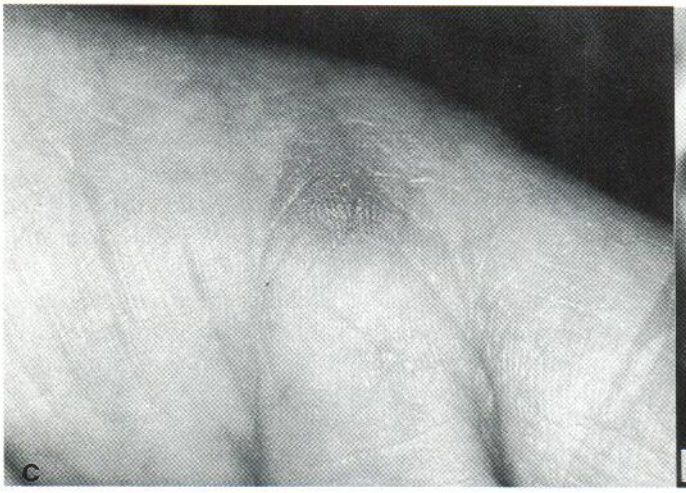
Light microscopic examination of biopsies from blisters showed in both cases a cleavage in epidermis associated with dyskeratosis. These findings suggested an epidermolysis bullosa of the dominant simplex type, first reported by Weber & Cockayne (2, 3). However, ultrastructural examination of all four cases from the two families with Kallin's syndrome showed a type of recessive intraepidermal pseudojunctional epidermolysis. This type differed from epidermolysis bullosa simplex, Weber-Cockayne variety, by the initial blister formation, localized deeper in the basal cells, and by the morphology of full-blown blisters (4).

Associated features to epidermolysis bullosa are rare. However, myopy and hyperhidrosis have been mentioned together with the Weber-Cockayne variety (5). Hair and nail disorders and total or partial anodontia are generally considered to possess a dominant mode of inheritance.

According to the pedigrees of both families, an autosomal recessive mode of inheritance is obviously the most reliable genetic interpretation of Kallin's syndrome. The new ultrastructural characteristics also support a recessive genetic trait, even though features generally considered to possess a dominant mode of inheritance are associated with the syndrome. Therefore, epidermolysis bullosa of Kallin's syndrome should not be considered the common dominant localized simplex type, but according to the ultrastructural findings a recessive intra-







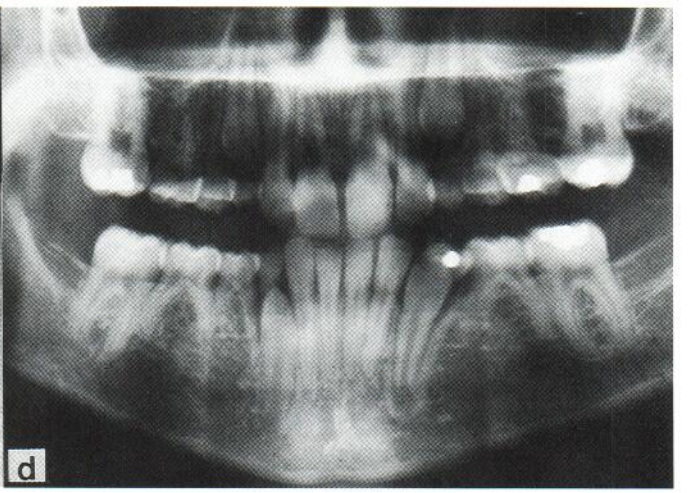


Fig. 1. (*a*) Pedigree of the family. (*b*) Scanty growth of hair with non-scarring alopecia. (*c*) Blister on the right hand. (*d*) Panoramic radiograph of the jaws.

epidermal pseudojunctional epidermolysis bullosa associated with anodontia, hair and nail disorders.

Modern gene finding technique and more cases are still needed to clarify the clinical entity and the final genetic trait.

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