thighs, recalling the "baboon syndrome" (3). Severe cases show pustules or purpura. Systemic symptoms and fever may be present. Histopathology shows spongiosis and a superficial perivascular infiltrate of lymphocytes and neutrophils or a subcorneal pustule.

In our patient, the exanthem was clinically and histologically similar to the one reported by Nakayama et al. The Japanese authors consider it as a systemic contact dermatitis caused by inhalation of mercury vapours. Indeed, our patient had an allergic contact dermatitis, as the positive results of patch tests suggest. However, two points differ from Nakayama et al.'s report: the particular severity of lesions where the amulet had been applied and, especially, the widespread eruption that followed patch-testing. They suggest that the transepidermal absorption is also important to explain generalization.

REFERENCES

G. F. Muzio, M. Guarerra and F. Rongioletti
Clinica Dermatologica dell'Università, Viale Benedetto XV, 7, 16132 Genova, Italy.

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**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 consanguinous 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 hypermetropia 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-

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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.
Localized Pachydermodactyly in a Woman

Sir,

The term pachydermodactyly (pachyderma, digital fibromatosis, discrete keratoderma, knuckle pad disease, acanthohyperkeratosis) describes a rare and not yet well defined condition characterized by a dense, bulbous swelling around the proximal phalanges or interphalangeal joints of the fingers (1). Histological features include hyperkeratosis, acanthosis or normal epidermis with underlying dermis showing an increased content of irregular collagen bundles, occasionally an augmented number of fibroblasts and mucin deposition (2).

We here report the case of a 23-year-old woman who consulted us for a fusiform swelling of the proximal interphalangeal joint of the fourth finger of the right hand. This bulbous swelling was localized on the ventral, dorsal and lateral sites of the affected finger like a ring and it had appeared 3 months earlier without history of trauma to the hand or osteoarticular symptoms. The patient complained that she was not able to move the finger because of intense pain.

A roentgenogram as well as an echography of her affected finger showed a swelling of the soft tissue without bone or articular involvement. Routine laboratory tests, including serum rheumatoid factor and antinuclear antibodies, were negative. General examination revealed no other systemic manifestations of disease.

The patient was submitted to an incisional biopsy of the affected area. The specimen was routinely processed and paraffin-embedded.

The formalin-fixed specimens were stained with hematoxylin and eosin, PAS with and without diastase digestion, Giemsa, Weigert stain for elastic tissue, Alcian blue at pH 2.5, and submitted to hyaluronidase digestion.

The epidermis showed slight hyperkeratosis and acanthosis. The papillary dermis was normal while the reticular dermis was characterized by the presence of thickened, tortuous, haphazardly arranged collagen bundles. An increased number of plump fibroblasts were present between collagen bundles. No inflammatory infiltrate was present but numerous mast cells were detected. The eccrine sweat glands were embedded in an abundant loose mucinous stroma, where also nerve fibres were seen.

These mucin deposits were PAS negative, Alcian blue positive at pH 2.5, hyaluronidase sensitive and metachromatically stained with the Giemsa stain. A Weigert stain for elastic tissue showed sparse thinned and elongated elastic fibres.

The clinical and pathological features of our case were consistent with a superficial fibromatosis.

Fibromatosis consists of benign fibrous tissue proliferations that generally occur in certain areas and at certain ages, in some cases characterizing clinically and histologically well-defined diseases or syndromes. Sometimes, however, it may be hard to define the differences between simple overgrowth of fibrous tissue and benign tumours as well as to distinguish between the different diseases characterized by the presence of fibromatosis.

Pachydermodactyly is a benign superficial fibromatosis of the fingers, first described in 1975 by Verbov (3). It usually occurs in young adult males, causing dense bulbous fusiform swelling around the proximal interphalangeal joint. The condition is generally asymptomatic and not associated with systemic disease. Until now, 13 cases have been published in the literature, only one of them being a woman (4); 11 of these were submitted to histological examination and all cases showed comparable features: overgrowth of fibrous tissue with cytologically typical fibroblasts and collagen deposits in the dermis with unremarkable overlying epidermis. In 12 patients this form of digital fibromatosis was apparently idiopathic and asymptomatic, apart from the association in two cases with carpal tunnel syndrome and tuberculosis, respectively (5); one case was preceded by repetitive trauma (6) and only one patient complained that the condition was painful for 2–3 years (7). Although our case presented many of the clinical and pathological features previously described, it differed in that the patient was female and the disease was painful and limited to only one finger.

A possible explanation for the pain experienced by our patient is the recent onset of her condition, whereas all cases reported in the literature were examined much later. Furthermore the abundant mucin deposition observed around eccrine glands may have caused nerve fibre compression.

The diagnosis of pachydermodactyly limited to one digit is not as obvious as in cases involving several digits. The differential diagnosis of pachydermodactyly limited to one finger in-