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Nail Changes of Punctate Keratoderma: A Clinical and Pathological Study of Two Patients

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The pathological study of the nail changes of 2 patients affected by punctate keratoderma is described. Both patients presented nail abnormalities that were clinically suggestive of a nail psoriasis. Subungual hyperkeratosis was a prominent feature but onycholysis, splinter haemorrhages and pitting were also present. The pathology of the nail bed revealed sharply limited columns of hyperkeratosis associated with hypergranulosis and depression of the underlying nail bed epidermis. Etretinate therapy produced a significant improvement in the palmo-plantar keratoderma, but it was of no apparent value in treating nail keratoderma. ***Key words: Palmo-plantar keratoderma; Nail bed hyperkeratosis; Etretinate.***

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Patients with palmo-plantar keratoderma may exhibit nail changes that are more commonly associated with diffuse palmo-plantar keratoderma. We present here the clinical and pathological study of the nail changes of 2 patients affected by punctate keratoderma. The pathology of the nails confirmed that the nail abnormalities had been caused by a localization of the disease in the nail apparatus.

CASE REPORT

Case 1

A 48-year-old hairdresser consulted us in February 1990 with a 1-year history of nail abnormalities involving the toenails. She had been previously treated with several antimycotic agents without success. On clinical examination the toenails were thickened, yellow-brown in colour and showed marked subungual hyperkeratosis. In addition, the 2nd and 3rd right fingernail showed irregular pitting, distal splinter haemorrhages and mild subungual hyperkeratosis (Fig. 1). The overall clinical appearance of the nail changes suggested psoriasis as a possible diagnosis. Examination of the skin revealed a mild hyperkeratosis on the pressure areas of the soles. The keratosis was clinically characterized by multiple, discrete, yellowish-white, slightly elevated, conical keratotic plugs, 2 mm in diameter. Removal of the horny epithelial plugs resulted in the formation of small crateriform pits. The lesions were distributed symmetrically on the soles, extending to the plantar surface of the great toes. Palms were less severely affected, with only isolated hyperkeratotic lesions. The fingertips and the toetips were not affected. The patient did not complain of subjective symptoms except for occasional pain when walking. She stated that the hyperkeratotic lesions on her palms and soles had first appeared 2 years before and had since then gradually increased in size and number. The onset of the keratoderma corresponded to the beginning of the menopause. No other members of her family were affected.

The patient was treated with etretinate at initial dosages of 1 mg/kg/day followed by gradual tapering to 0.5 mg/kg/day. After 6 months of therapy the palmo-plantar keratoderma had considerably improved, but the nail abnormalities persisted unchanged.

Case 2

A 46-year-old barman consulted us in April 1991 due to a severe onychodystrophy affecting the 2nd, 4th and 5th right fingernail. The patient had first noticed his nail abnormalities 1 year before and had previously been treated with systemic and topical antifungal drugs without result. At the clinical examination the nails of the affected fingers were yellow-brown in colour and showed marked nail bed hyperkeratosis associated with onycholysis. The lunula of the 2nd finger was markedly enlarged. Distal splinter haemorrhages were also evident (Fig. 2).

A mild punctate keratoderma was present on the palm of the right hand.

Pathology

The patients' skin and nail pathology were studied.

Skin biopsies were taken from the palmar lesions of keratoderma. In both cases, the pathology showed sharply limited areas of massive hyperkeratosis associated with hypergranulosis and depression of the underlying malpighian layer. A Weigert elastic tissue stain revealed normal elastic fibers. No inflammatory infiltrates were detected.

The 48-year-old woman was submitted to a longitudinal nail biopsy of the 2nd toenail, whereas the 46-year-old man was submitted to a nail bed biopsy of the 5th right finger. In both cases the nail pathology revealed lesions of punctate keratoderma in the nail apparatus. The nail bed showed diffuse hyperkeratosis that lifted up the nail plate. Sharply defined limited columns of massive hyperkeratosis associated with hypergranulosis and depression of the underlying nail bed epidermis were clearly evident within the nail bed hyperkeratosis (Figs. 3–4). Similar changes were present in the distal nail matrix of the first patient. Elastic fibers were normal and no inflammatory infiltrates were detected. PAS stain was negative.

DISCUSSION

In these 2 patients identical pathological changes were found in the palmo-plantar skin and in the nail bed and matrix: a massive localized horny mass associated with depression of the underlying malpighian layer. This indicates that the nail

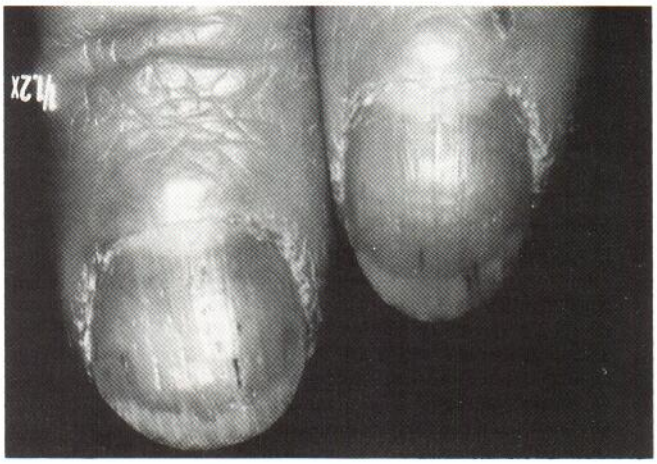

Fig. 1. Patient 1. Irregular pitting, distal splinter haemorrhages and mild subungual hyperkeratosis of the 2nd and 3rd right fingernail.

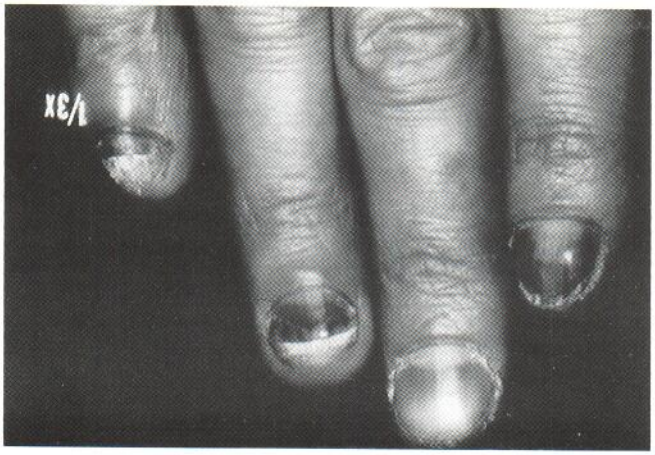

Fig. 2. Patient 2. Marked nail bed hyperkeratosis and onycholysis of the 2nd, 4th and 5th right fingernail.

changes were due to a localization of the dermatosis in the nail apparatus.

Papular keratotic lesions of the palms and soles can present different pathological pictures. In punctate keratoderma (1), palmo-plantar keratotic pits (2) and focal acral hyperkeratosis (3), a localized massive hyperkeratosis with hypergranulosis is

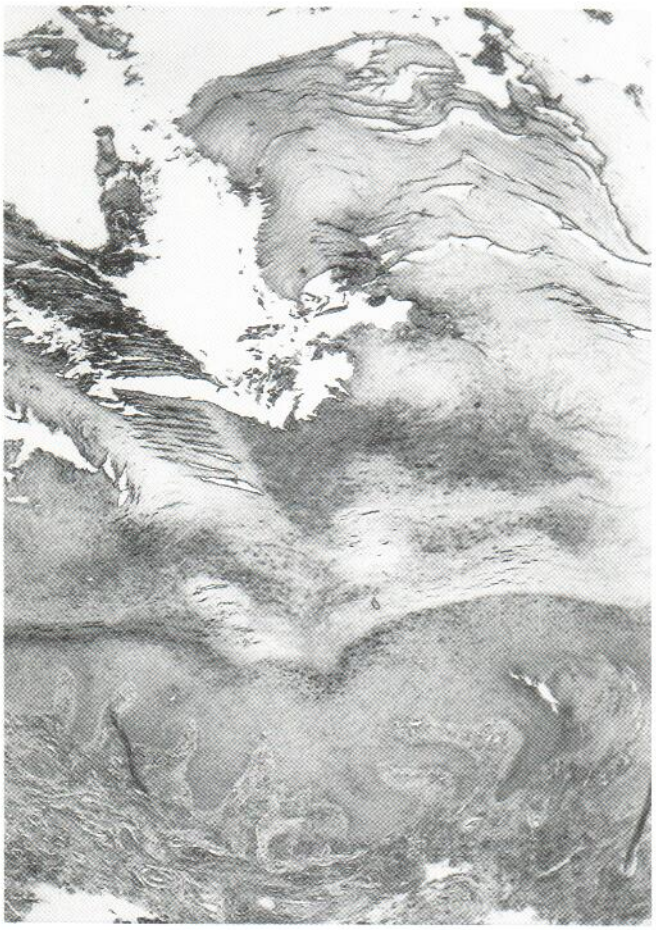

Fig. 3. Longitudinal nail biopsy of the 48-year-old woman. Sharply limited columns of hyperkeratosis associated with depression and hypergranulosis of underlying epidermis are detectable in the nail bed. HE 100×.

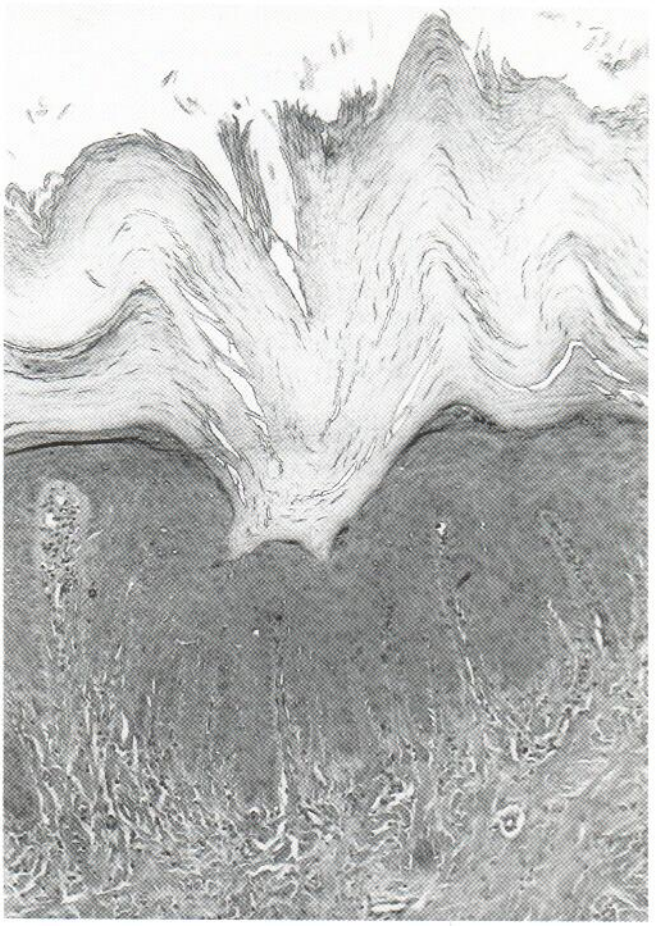

Fig. 4. Nail bed biopsy of the 46-year-old male. High-power view showing a typical lesion of punctate keratoderma consisting of sharply limited hyperkeratosis and depression of the underlying nail bed epidermis. HE 200×.

observed. The underlying malpighian layer is often located below the general level of the epidermis. Acrokeratoelastoidosis (4) shows similar features but is further characterized by fragmentation or absence of the elastic fibers. Punctate porokeratotic keratoderma (5) or porokeratosis punctata palmaris et plantaris (6) show columnar parakeratosis and absence of the granular layer. Focal acantholytic dyskeratosis has also been reported in a patient with punctate keratoderma (7). The clinical and pathological features of our patients were consistent with a diagnosis of acquired punctate palmo-plantar keratoderma.

The classification of punctate palmo-plantar keratoderma is confused by the presence of innumerable clinical variants. Nail involvement has occasionally been described in punctate keratoderma as well as in closely related or identical conditions such as Brauer-Buschke-Fisher's keratoderma and keratosis palmo-plantaris papulosa. Onychogryphosis, nail thickening, subungual hyperkeratosis, longitudinal fissures and onychomadesis have all been reported (8–11).

The development of keratotic lesions in the nail bed, which is devoid of eccrine sweat glands, rules out the hypothesis that an abnormal keratinization of the sweat ducts may play a role in the pathogenesis of the disease. Although etretinate therapy

produced a significant improvement in the palmo-plantar keratoderma, it was of no benefit in the nail keratoderma. Retinoids, however, are in general of little value for the treatment of nail keratinization disorders. This is, to the best of our knowledge, the first pathological evidence of nail bed involvement in punctate keratoderma.

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