one of exclusion, after discarding other possibilities such as onychomycosis, psoriasis, trauma and drug eruption, in patients with proved sarcoidosis elsewhere.

A punch biopsy in the proximal fold is enough for diagnostic purposes; yet to establish an appropriate study of the nail unit pathology, a longitudinal biopsy would be more adequate, since – as our cases have demonstrated – all their structures were affected. The most remarkable finding is the strong granulomatous dermal infiltrate, from the proximal nail fold to the hyponychium, particularly dense in the nail bed. The epidermal changes were also prominent at this level, and both facts explain the colour changes and contributed to the thickening, irregularity and roughening of the nail plate. The atrophy in the matrix (significantly more intense in the case 2, associated with clinical fissuring) and the distortion of the rete ridges cause longitudinal grooves, and their progression could lead to pterygium formation and nail loss. Splinter haemorrhages are in relation to minimal trauma in a matrix in close contact with dermal capillaries. All these findings can be explained on the basis of the microcompressing effect by sarcoid granulomas in a dermal compartment limited by two rigid structures: the phalanx and the nail plate. Furthermore, disorders of keratinization were observed, expressed as parakeratosis, coarse keratohyalin granules, loss of cohesion, staining disturbance and distal hyperkeratosis and detachment, which clinically manifested as thickening, fragility, onycholysis, and subungual hyperkeratosis. Since specific disorders of keratinization are not attributable to sarcoidosis, these will also be secondary to granulomatous compression.

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Epidermolytic Hyperkeratosis of the Nails in Keratosis Palmoplantaris Nummularis

Sir,

Patients with localized palmoplantar keratoderma (PPK) may exhibit nail changes that are more commonly associated with diffuse PPK (1, 2). We report here the pathological evidence of epidermolytic hyperkeratosis (EH) of the nails in a patient affected with localized epidermolytic PPK.

CASE REPORT

A 25-year-old male carpenter consulted us because of painful palmoplantar hyperkeratosis associated with psoriasiform nail abnormalities involving both fingernails and toenails.

The clinical history revealed that the patient had been affected with plantar keratoderma since the age of 3 and palmar keratoderma since the age of 18. The patient had first noticed thickening and discoloration of his fingernails and toenails 1 year before our examination. During the last 3 years plantar and palmar lesions had considerably enlarged and become painful. The family history revealed that his mother and sister were also affected by nummular painful PPK.

Clinical examination revealed numerous nummular thick yellow keratotic patches over the pressure areas of the patient's palms and soles. His toenails were dark-yellow in colour and markedly thickened because of subungual hyperkeratosis. His fingernails showed mild subungual hyperkeratosis associated with onycholysis and distal splinter haemorrhages. The cuticles of fingernails and toenails were markedly hyperkeratotic (Fig. 1). The rest of the skin was normal, including the dorsal aspect of the hands and feet. The teeth and hair were also normal.

The patient had no local hyperhidrosis of the palms and soles. KOH preparations and cultures of nail and skin scrapings were negative.

A biopsy from a keratotic plantar lesion revealed massive orthohyperkeratosis, with irregular hypergranulosis, acanthosis and vacuolization of the upper malpighian layers.

A longitudinal nail biopsy from the fifth right toenail revealed marked hyperkeratosis of the ventral portion of the proximal nail fold, distal nail bed and hyponychium. The ventral portion of the proximal nail fold (Fig. 2), the distal nail bed and the hyponychium showed perinuclear vacuolization of suprabasal keratinocytes and a thickened granular layer containing irregular basophilic keratohyalin granules. The nail matrix was not involved.

Since the patient refused oral retinoids, a local treatment with keratolytic agents was prescribed, with mild improvement of the keratoderma.

DISCUSSION

The clinical and pathological features of our patient are consistent with a diagnosis of keratosis palmoplantaris nummularis (KPPN). This name was coined by Wachters et al. (3) to describe a dominant inherited circumscribed type of PPK associated with the pathological features of EH. Clinically, KPPN is characterized by focal keratoses located on the plantar pressure

areas. Pain is a major complaint and some patients may be almost invalids. Palmar keratoses are not a common feature of the disease, having only been occasionally observed in hard manual workers. Nail involvement with periungual and subungual hyperkeratosis and/or thickening of the toenails is commonly found.

In our patient painful nummular keratotic lesions had been present on the soles since infancy and had gradually appeared on the palms a few years after he started his work as a carpenter. Repeated trauma may also have induced the development of the nail lesions, which had only recently been noticed by the patient.

Although hereditary epidermolytic PPK (4) is pathologically indistinguishable from KPPN, the restriction of the keratotic lesions to the pressure areas and the presence of intense pain rule out this diagnosis in our patient.

The finding of EH in the nail biopsy of our patient indicates that the nail lesions are due to the disease and should be included among the clinical symptoms of KPPN.

EH was more prominent in the ventral portion of the proximal nail fold, which is very similar to skin interfollicular epidermis. Nail matrix was unaffected. This finding may be explained by the different mechanism of keratinization taking place in the nail matrix epithelium, where onychocytes keratinize without the presence of keratohyalin granules (5).

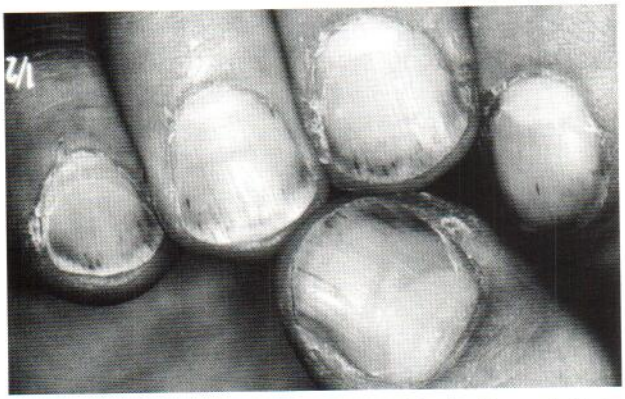

Fig. 1. Mild subungual hyperkeratosis associated with onycholysis and distal splinter haemorrhages. Cuticles are hyperkeratotic.

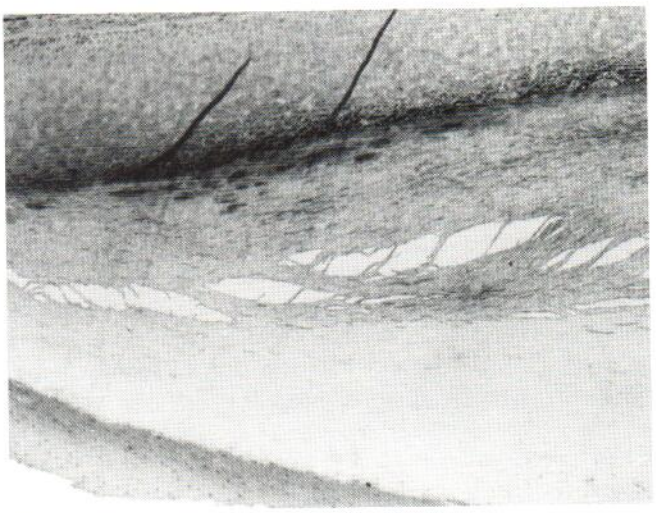

Fig. 2. The ventral portion of the proximal nail fold shows perinuclear vacuolization of suprabasal keratinocytes and a thickened granular layer containing irregular basophilic keratohyalin granules.

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