## 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 affects several fingers of young adult males, causing dense bulbous fusiform swelling around the proximal interphalangeal joints. 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 tuberous sclerosis, respectively (5); one case was preceded by repetitive traumas (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-

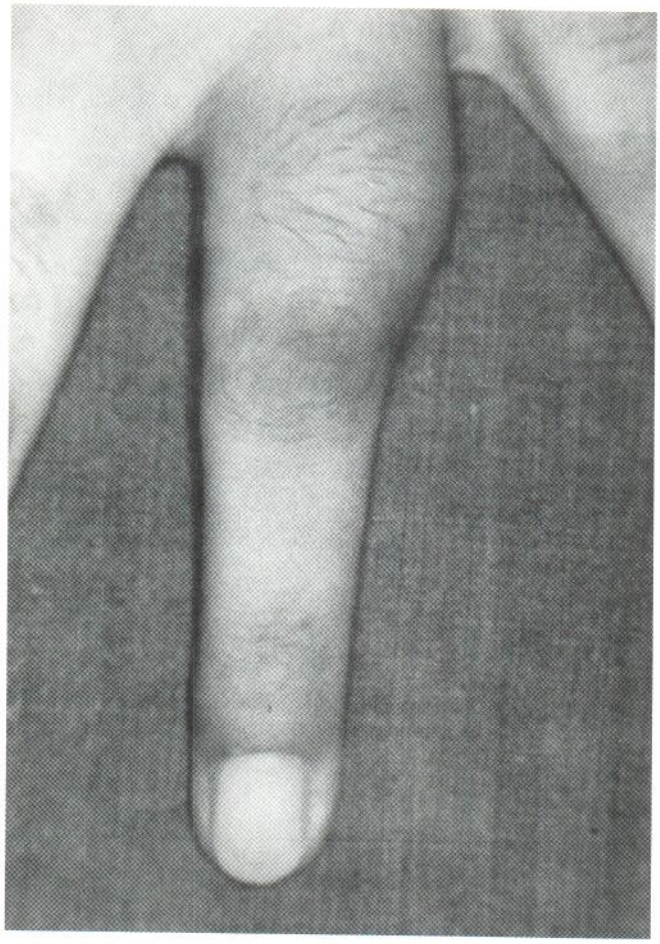

*Fig. 1.* Bulbous fusiform ring-like swelling of the proximal interphalangeal joint of the fourth finger of the right hand.

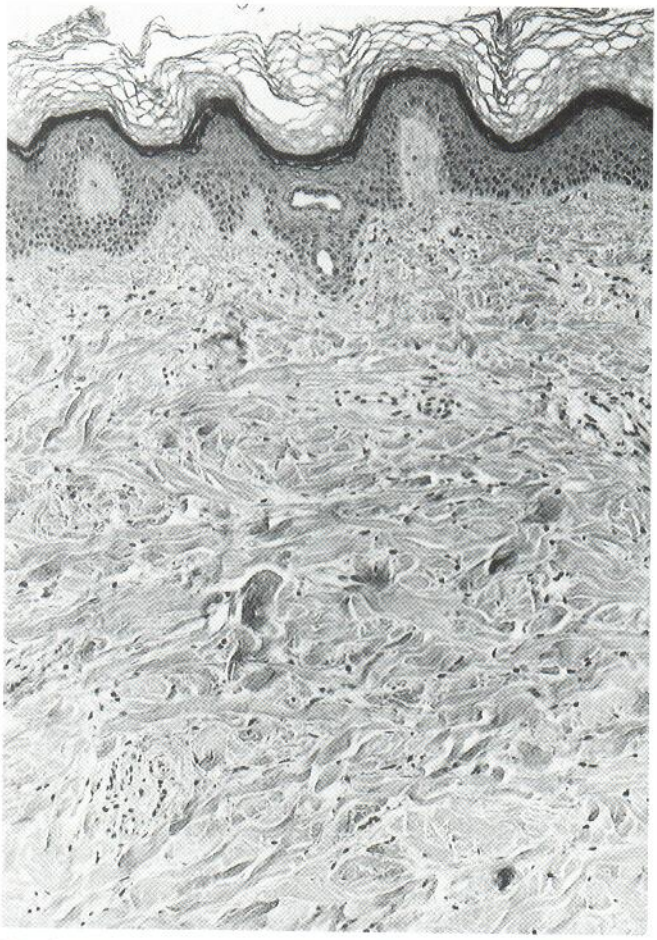

*Fig. 2.* Moderate hyperkeratosis of the epidermis. The thickened reticular dermis shows the presence of thick, tortuous and haphazardly arranged collagen bundles (H&E 80×).

cludes tendon ganglion, a giant cell tumour of tendon sheaths, an epidermal cyst or a foreign body reaction. It is advisable to carry out a skin biopsy in order to distinguish between these benign conditions and pachydermodactyly, since the latter condition may also be limited to one finger. Intralesional triamcinolone may be helpful in improving the clinical appearance (7).

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