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Pachydermodactyly: A Rare Form of Acquired Digital Fibromatosis

Paquidermodactilia: una forma poco frecuente de fibromatosis digital adquirida

To the Editor:

Pachydermodactyly is a rare, benign form of acquired digital fibromatosis, characterized by soft tissue swelling that affects the skin of the fingers, specifically on the lateral aspect of the proximal interphalangeal (PIP) joints, mainly of the second, third, and fourth fingers. It particularly affects adolescents with no family history and is occasionally confused with rheumatological disease. To date, few cases have been reported in the medical literature; we describe a new case.

The patient was a 14-year-old boy with no past history of interest who was referred to the dermatology clinic for evaluation of diffuse thickening that had developed some months earlier on the second, third, and fourth fingers of each hand. A point of interest is that the patient was a competition-level climber who practiced for several hours each day.

Physical examination revealed bilateral and symmetric swelling of the lateral aspect of the PIP joints of the second, third, and fourth fingers of each hand (Figures 1 and 2), more marked on the right side (Figure 2). There were no other relevant alterations of the skin.

A skin biopsy of one of the affected areas revealed a thickened dermis with an increase in the number of collagen fibers and a slight increase in the number of fibroblasts. There was also a slight increase in the quantity of mucin, with no significant inflammatory infiltrate. The overlying epidermis was hyperkeratotic with compact orthokeratosis (Figure 3).

Blood tests were requested, including kidney, liver, and thyroid function, rheumatoid factor, antinuclear antibodies, and other autoimmune studies, all with normal or negative results.

Plain radiography of the hands revealed thickening of the soft tissues around the PIP joints, with no bone or joint abnormalities (Figure 4).

Pachydermodactyly was first described by Bazex in 1973 and it was named by Verbov 2 years later.¹ It is a rare disease, though its true incidence could be underestimated. This benign fibromatosis mainly affects adolescent men and the diagnosis is questionable if made in adults.² In many cases there is a history of manual sporting practice such

as martial arts, weight-lifting, or climbing (as in our case). The etiology is unknown. Rai and Zaphiropoulos suggested that it might represent an incomplete presentation of pachydermoperiostosis.³ Pachydermoperiostosis or Touraine-Solente-Golé syndrome is the primary or idiopathic form of hypertrophic osteoarthropathy.⁴ The secondary form is more common and usually follows lung or heart disease, which may be of a neoplastic nature. Pachydermoperiostosis is defined by 3 major criteria: pachyderma, periostosis, and clubbing of the fingers. There are also minor criteria (seborrhea, sebaceous hyperplasia, folliculitis, acne, and more). The primary form is considered to be a hereditary disease, although a family history is found in only 25% to 38% of cases. In addition, it has a variable degree of penetration, and complete forms of the syndrome are therefore uncommon.⁵

Clinically, pachydermodactyly is characterized by asymptomatic symmetric swelling of the PIP joints, predominantly on their lateral aspect, and particularly affecting the second, third, and fourth fingers. Occasionally it also affects the dorsum of the hand, then receiving the name pachydermodactyly transgrediens.⁶

Histopathology study reveals epidermal hyperplasia with compact orthokeratosis and thickening of the dermis with an increase in the number of collagen fibers and a mild proliferation of fibroblasts, with no significant inflammatory infiltrate.

The differential diagnosis should include certain forms of polyarthritis that can affect the PIP joints, such as the polyarticular form of juvenile chronic arthritis, psoriatic arthritis, and rheumatoid arthritis. However, all those disorders have characteristic joint involvement, whereas pachydermodactyly is an asymptomatic fibrous swelling of the PIP joints of the second, third, and fourth fingers with no bone or joint abnormalities.

Pachydermodactyly must also be differentiated from juvenile hyaline fibromatosis, although there are some authors who, as occurs with pachydermoperiostosis, consider that pachydermodactyly is a localized form of juvenile hyaline fibromatosis. Juvenile hyaline fibromatosis is a mesenchymal dysplasia of autosomal recessive inheritance that appears in early childhood or in adolescence; only about 65 cases have been reported. It is characterized clinically by skin lesions, gingival hypertrophy, flexion contractures of the large joints, and bone lesions. The skin lesions consist of multiple tumors, often located on the scalp and around the nose, associated with small pearly papules and plaques on the trunk, chin, and ears and around the nares. Juvenile hyaline fibromatosis is due to synthesis of abnormal collagen, which is deposited as



Figures 1 and 2 Swelling of the lateral aspect of the proximal interphalangeal joints of the second, third, and fourth fingers of each hand, more visible on the right hand (Figure 2).

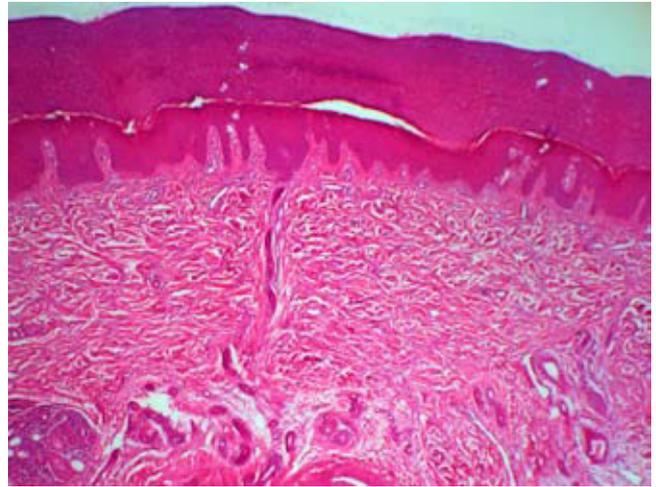


Figure 3 Hyperkeratosis with compact orthokeratosis. Thickening of the dermis, with an increase in the number of collagen fibers and fibroblasts. Slight increase in the quantity of mucin, with no significant inflammatory infiltrates. Hematoxylin-eosin, original magnification $\times 40$).



Figure 4 Thickening of soft tissues around the proximal interphalangeal joints, with no bone or joint abnormalities.

hyaline material in the connective tissue of the skin, gums, and, less frequently, of the bones and joints.⁷

Knuckle pads or Garrod nodes are a subtype of superficial fibromatosis. They basically consist of very well-circumscribed papules or plaques that are most commonly situated on the dorsum of the metacarpophalangeal or PIP joints of the fourth finger. They may be idiopathic (true) or secondary to repetitive injury (false). The fundamental difference between pachydermodactyly and knuckle pads lies in the fact that the thickening in knuckle pads is on the dorsum of the joints, whereas in pachydermodactyly it is on the lateral aspect. However, there are authors who believe pachydermodactyly to be a rare variant of knuckle pads.⁸⁻¹⁰

Other conditions should also be considered in the differential diagnosis, including joint tophi, xanthomatous deposits, and paraneoplastic acrodactyly.¹¹

The histopathological findings of pachydermodactyly include a thickened dermis, with an increase in the number of collagen fibers accompanied by a discreet increase in the number of fibroblasts. There is also a slight increase in the quantity of mucin, with no significant inflammatory infiltrate. The overlying epidermis often shows hyperkeratosis with compact orthokeratosis.

Local infiltration of triamcinolone hexacetonide and resection of the fibrotic subcutaneous tissue can improve the outward appearance. Treatment with topical corticosteroids has not been shown to be effective.¹²

To date, few cases of pachydermodactyly have been reported in the medical literature, probably because its prevalence is underestimated due to its asymptomatic nature and the fact that it does not limit joint mobility. We believe it is necessary to consider other disorders in the differential diagnosis in order to avoid unnecessary additional tests and treatments, as pachydermodactyly is a completely benign disorder and does not require treatment.

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Subungual Squamous Cell Carcinoma. Presentation of two Cases

Carcinoma escamoso subungueal. Presentación de dos casos

To the Editor:

Primary malignant subungual tumors are rare, although, of these, squamous cell carcinoma (SCC) is the most frequent. Initial diagnosis is frequently delayed by the relatively nonspecific clinical presentation, easily confused with benign inflammatory conditions such as paronychia, pyogenic granuloma, or a common wart. Diagnosis of this kind of lesion is only possible by pathology study of a biopsy. Treatment requires complete excision of the lesion, often leading in turn, to amputation of the distal phalanx

of the affected finger. We describe 2 new cases of difficult-to-diagnose subungual SCC.

The first patient was a man aged 73 years, with a history of hypertensive-ischemic heart disease, who consulted with an excrescent lesion of the nail bed of the first finger of the right hand which had developed over the previous 4 years (Figure 1). The patient reported no possible triggering factors such as repeated injury or viral warts at this site. Surgery was performed twice on the patient, and the pathology diagnosis was digital fibrokeratoma. The patient had responded poorly to treatment with different topical antibiotic and antifungal agents. Examination revealed a suppurating tumoral lesion, adherent to the nail plate of the first finger of the right hand and causing painful onychodystrophy. An ingrown nail with excessive granulation tissue was suspected. A biopsy confirmed the diagnosis of subungual SCC, and the lesion was excised. No bone involvement was evident on an x-ray of the first finger of