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Symmetrical fusiform swellings around the proximal interphalangeal joints

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The patient

A 15-year-old, otherwise healthy boy presented with 2-year-history of swelling and thickening localized to the lateral aspects of his fingers. Dermatological examination revealed fusiform soft tissue swelling on the lateral and medial aspects of the second to fourth proximal interphalangeal (PIP) joints of both hands (Figure 1). There was no arthralgia or limited range of motion. Rheumatological examination was otherwise normal. There was no history of repetitive trauma, tick-like habits or any mental disorders.

Laboratory tests, including routine biochemistry, thyroid function tests, and rheumatologic markers, disclosed no abnormalities. An MRI and X-ray examination was normal except for soft tissue hypertrophy around the affected joints (Figure 2).

What is your diagnosis?

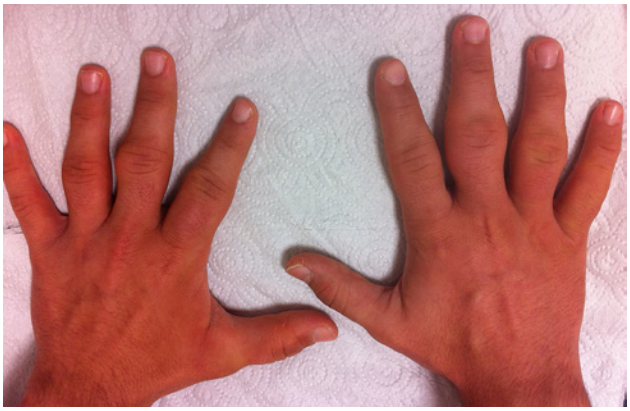

Figure 1. Fusiform, soft tissue swelling on the lateral and medial aspects the proximal interphalangeal joints of the second to fourth fingers. [Copyright: ©2015 Saricam et al.]

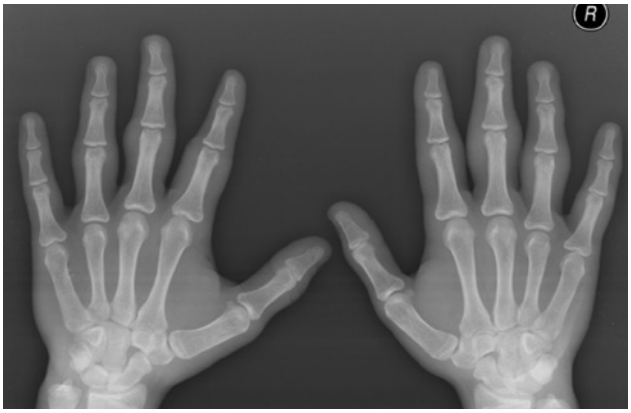

Figure 2. X-ray examination shows soft tissue swelling around the affected joints without any osseous changes. [Copyright: ©2015 Saricam et al.]

Answer

Pachydermodactyly.

Clinical course

Although a treatment with intralesional corticosteroids was planned, the patient was lost to follow-up.

Discussion

Pachydermodactyly (PDD), is an uncommon, benign form of superficial digital fibromatosis [1]. It is characterized by asymptomatic, periarticular soft tissue swelling of the PIP joints. The disease is most commonly seen in young, otherwise healthy males [1-3]. While PDD usually affects the PIP joints symmetrically, involvement of distal interphalangeal (DIP) joints and metacarpophalangeal (MCP) joints or unilateral disease can also be seen [4]. In the transgradient form of PDD, both MCP and PIP joints are affected [3].

The etiology of PDD is not fully understood but repetitive minor traumas (occupational, habitual or compulsive) are thought to be the major contributing factor [1,2]. PDD was reported to be associated with obsessive-compulsive disorder, Asperger syndrome, Ehlers-Danlos syndrome and tuberous sclerosis [3].

PDD may resemble juvenile idiopathic arthritis, rheumatoid arthritis, knuckle pads, pachydermoperiostosis or acromegaly [3]. The diagnosis of PDD is usually made with typical clinical findings. Laboratory tests and radiographic examination may be helpful in exclusion of the differential diagnosis. X-ray imaging shows soft tissue swelling without any articular or osseous changes. Ultrasonography also confirms the presence of soft tissue hypertrophy without hypervascularization and the absence of joint disease. Similarly, magnetic resonance imaging (MRI) reveals only soft tissue swelling without synovitis or tendinitis [3]. Though histopathology is not routinely necessary in clinical practice, it shows epidermal acanthosis, increase in dermal collagen and mucin deposits in dermis [1-3].

There is no effective treatment for PDD. Avoidance of mechanical trauma may result in improvement. Intralesional corticosteroid injections and localized resection of subcutaneous tissue were reported to be effective in some cases [2-5]. However, given to its benign nature, non-invasive treatment options are recommended for PDD [2].

In conclusion, recognition of this rare, or under-reported, benign condition is crucial to prevent patients from unnecessary or expensive laboratory tests and improper treatment with immunosuppresive agents.

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