Pachydermodactyly may mimic juvenile idiopathic arthritis

Sir,

A 16-year old boy presented with a 3-year history of a painless, progressive, symmetrical swelling of the fingers, recently causing impairment while playing the guitar; there was a similar swelling on his chin since 6 months. General health was good. Family history was negative for similar changes or rheumatic disorders.

On clinical examination (Fig. 1) there were fusiform swellings limited to the ulnar and radial sides of the proximal interphalangeal joints and proximal phalanges of fingers II to IV and, less important of the radial side of finger V of both hands. The overlying skin was thickened, slightly erythematous and scaly. A similar swelling was present on the chin.

An ultrasound and X-rays of hands and chin showed soft tissue swelling with normal underlying bones and joints. Blood tests did not show inflammation nor the presence of antinuclear antibodies. A skin biopsy revealed hyperorthokeratosis, hypergranulosis and mild acanthosis of the epidermis. In the thickened reticular dermis there was an increase of irregular collagen bundles and mucine, but a loss of elastic fibres.

Based on these findings the diagnosis pachydermodactyly was made. After further questioning it became clear that the boy had a habit of crossing and rubbing his fingers followed by touching his chin when playing computer games. There was no evidence for an underlying psychiatric disorder. Three months after stopping this movement an obvious improvement was apparent (Fig. 2).

Pachydermodactyly is a benign superficial fibromatosis of the fingers, mainly affecting young males, first reported by Verbov (1) in 1975. About 60 cases are published until now, but it may be underreported.

The condition is characterized by symmetrical painless swellings over the radial and ulnar aspects of the proximal interphalangeal joints of the fingers II to IV, although atypical forms have been described.

Bardazzi *et al*. (2) proposed a classification with five different forms, including: classical pachydermodactyly, localised or monopachydermodactyly, pachydermodactyly transgrediens (extension to the palms or proximal fingers), familial pachydermodactyly, and pachydermodactyly associated with tuberous sclerosis.

The cause of pachydermodactyly remains unknown and may vary among patients. Repeated mechanical trauma (as in our patient) has been proposed in some children (2,3). As far as we know, regression after cessation of the mechanical trauma has been reported only once up till now (4). A compulsive neurotic personality (3) such as in Asperger syndrome (5) has been associated with pachydermodactyly, although this remains exceptional. Moreover pachydermodactyly seems exceptional in children with comparable compulsive disorders.

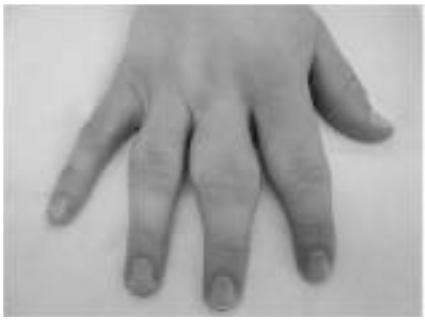

Fig. 1. Swelling of the fingers at the first visit.

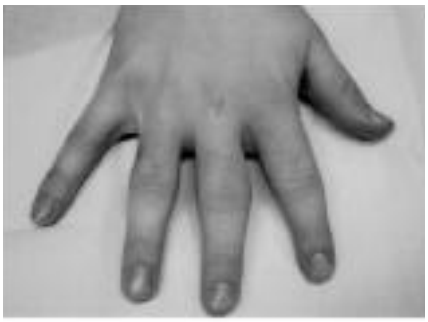

Fig. 2. Obvious improvement 3 months after stopping compulsive movements.

As an association with knuckle pads in the father has been described, it has been suggested that pachydermodactyly presents a variant of this disease, an underlying defect in proliferation of collagen being a predisposing factor (6).

Pachydermodactyly may be misdiagnosed as juvenile idiopathic arthritis (7). The absence of pain and morning stiffness, of synovitis on clinical examination, of inflammatory blood changes, together with the typical thickening of the skin are the most important differences with JIA. The differential diagnosis should also include true knuckle pads (6) which are fibromatous thickenings on the dorsum of the fingers joints; pachydermoperiostosis (8), a familial disease where bone and soft tissue proliferation produce clubbing and a spade-like enlargement of hands and feet, associated with thickening and furrowing of facial skin and juvenile digital fibromatosis, mostly occurring in infants as solitary or multiple soft tissue tumors at the digits of hands and feet.

No satisfactory treatment for pachydermodactyly has been described in the literature. Intralesional corticosteroids (9) and excision (10) are possible. Correction of a triggering compulsive habit may lead to remarkable improvement as illustrated by our case.

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Comparative analysis of hexosaminidase and cathepsin D expression in synovial fluid of patients with rheumatoid arthritis and traumatized joints

Sirs,

Recently, significant advances have been made in understanding the mechanism of articular cartilage destruction. Matrix metalloproteinases and cysteine proteases (cathepsins) are claimed to be the major enzymes implicated in this destructive process (1, 2). However growing data have been published on exoglycosidases as participants in the pathogenesis of joint damage (2-4). In the present paper we compare the activity of N-acetyl-β- hexosaminidase (HEX) and cathepsin D in the synovial fluid (SF) of patients with rheumatoid arthritis (RA) and juvenile idiopathic arthritis (JIA).

We examined patients with two types of rheumatoid diseases. 11 patients had RA (6 female, 5 male; 27-76 years old), with