

Magnetic Resonance Imaging in Pachydermodactyly

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ABSTRACT. The magnetic resonance imaging (MRI) findings of a teenage boy with pachydermodactyly are described. The findings include fusiform soft tissue swelling around the proximal interphalangeal joints of the hands, with sparing of the tendons and ligaments. There was no capsular involvement and no evidence of periosteal reaction. (*J Rheumatol* 2005;32:2239–41)

Key Indexing Terms:

PACHYDERMODACTYLY

PACHYDERMOPERIOSTOSIS

MAGNETIC RESONANCE IMAGING

Pachydermodactyly is an acquired, benign digital fibromatosis of unknown etiology. It is characterized by painless fusiform swelling affecting the skin overlying the radial and ulnar aspects of the proximal interphalangeal (PIP) joints and often poses a diagnostic problem¹. Most cases have been described in young male adults, although there are increasing reports of affected women.

This is not to be confused with pachydermoperiostosis. Readers may recall that both the British poet W.H. Auden and the French playwright Jean Racine shared this rare condition, also known as Touraine-Solente-Golé syndrome. This is an inherited defect occurring mainly in males, who also have clubbing of the fingers, oily skin, and a lugubrious expression. Affected individuals may also display thickening of the legs and forearms resulting primarily from periosteal new bone formation at the distal ends of long bones².

Plain radiological and histopathological findings in pachydermodactyly have been reported^{3,4}. We report the first description of magnetic resonance imaging (MRI) findings seen in pachydermodactyly and review the literature.

CASE REPORT

A 15-year-old boy was referred with a 2 year history of painless swelling around the PIP joints. He had no history of early morning stiffness or psoriasis. He was a keen basketball player and his mother attributed the finger swelling to trauma. There was no relevant family history.

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Accepted for publication June 7, 2005.

On examination, there was pronounced fusiform, doughy swelling at the level of the PIP joints. This was noted in the second to the fifth digits of both hands, especially the third and fourth digits (Figure 1). There was no synovitis, psoriatic rash, or nail changes of psoriasis. He had no finger clubbing. The range of movement and function in both hands was normal. There was no involvement of other joints. Rheumatoid factor and autoantibody screen were negative. Radiographs of both hands showed soft tissue swelling around the PIP joints and normal underlying bone.

MRI (1.5 T Siemens Vision-plus system) of the left hand was performed to delineate the soft tissue changes. Coronal and axial T1 and STIR sequences were performed through the left hand. Fusiform swelling was seen at the PIP joints of the second through to the fifth digits, more prominent in the third and fourth digits (Figures 2A and 2B). The thickening predominantly involved the soft tissues around the PIP joints. The collateral ligaments and bony structures were otherwise normal, with no evidence of periosteal cortical thickening or capsular hypertrophy. The tendons all had a normal appearance.

A skin biopsy was taken from the area of swelling at the right middle finger. The epidermis showed compact orthokeratosis and mild acanthosis. There were mildly thickened collagen fibers in the dermis. The mucin stain was negative for acid mucopolysaccharide ground substance. These findings were in keeping with pachydermodactyly. Two years after the initial consultation, he remains asymptomatic on no specific therapy with no progression of his signs.

DISCUSSION

Pachydermodactyly was described in 1975 by Verbov⁵. The fusiform swellings around the PIP joints are typically asymptomatic and usually affect the second through to the fourth, and occasionally the fifth, fingers. Unilateral involvement is more common. Reported associations include type 3 Ehlers-Danlos syndromes and tuberous sclerosis. A classification system has been proposed by Bardazzi, *et al*⁶. They suggested classifying pachydermodactyly into 5 types: (1) classic (frequently associated with mechanical trauma), (2) mono or localized, (3) transgrediens (where the cutaneous thickness extends to the metacarpophalangeal areas), (4) familial (can be transgrediens), and (5) pachydermodactyly associated with tuberous sclerosis. Most cases, described as classic pachydermodactyly, appear to be attributable to trauma of some description.



Figure 1. Soft tissue swelling around the PIP joints.



Figure 2. Coronal T1 weighted MRI showing fusiform swelling at the PIP joints of the second through to the fifth digits of the left hand.

Although there has been at least one familial case of pachydermodactyly, most described cases are sporadic⁷. In contrast, pachydermoperiostosis is inherited in an autosomal fashion¹. Whereas pachydermodactyly is manifested only by cutaneous thickening affecting the hands, pachydermoperiostosis is distinct, with characteristic clubbing, seborrhea, and scalp and bony changes¹.

The histopathological findings previously described include thickening of the dermis by deposition of collagen

with varying degrees of cellularity. The collagen is predominantly type III and electron microscopy has shown the collagen fibers to be of smaller diameter and less uniform³. Ultrasonography has revealed thickening of the subcutaneous tissues without evidence of synovitis¹. Bone scintigraphy has been reported to be normal⁴. In our case, the MRI demonstrated fusiform soft tissue swelling of the fingers with minor edema in the subcutaneous tissues. There was no inflammation of the ligamentous and tendinous structures of

the fingers. Importantly, there was no bony abnormality. The MRI findings confirm that the condition is confined to changes in the soft tissue.

Our patient has remained well 2 years after the onset of the symptoms with persistent periarticular swelling. This is in keeping with the natural history observed with other case reports.

ACKNOWLEDGMENT

The authors acknowledge the contribution of Dr. David Spencer and Dr. Graydon Howe, Rheumatology Department, Westmead Hospital, for helpful discussion.

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