

CLINICAL IMAGES

The many symptoms of pachydermoperiostosis

Bilal Abdool-Gafoor

A 26-year-old black man with a 3-year history of persistent joint pain and swelling of the wrists and ankles and enlargement of his hands and feet, also reported 'deformity' of his fingernails (clubbing) since childhood. His father had similar symptoms, but no siblings were affected. The subject had marked soft-tissue enlargement of the hands and feet and gross clubbing with increased curvature of the nail angle (Lovibond's angle), boggiess of the nailbed and typical 'drumsticking' (Fig. 1). There was bony overgrowth and tenderness at the wrists and ankles with marked limitation of movement, and hyperhidrosis of the face, hands and feet was noted. No other physical signs or abnormal biochemistry were noted.

Radiographs of the hands and wrists revealed thickened cortical bone at the distal ends of the radius and ulna, subperiosteal reaction along the metacarpal shafts and distal ends of the radius and ulna, and tufting at the distal phalanges (Fig. 2). Similar changes were present in radiographs of the ankle and proximal tibia.

The absence of an underlying cause and the genetic history prompted a diagnosis of pachydermoperiostosis or primary hypertrophic osteoarthropathy.

Discussion

Hypertrophic osteoarthropathy (HOA) includes: pachydermoperiostosis, the primary form of which accounts for 5% of cases; and secondary HOA, 80% of which cases are associated with primary or metastatic pulmonary malignancies (and therefore previously called hypertrophic pulmonary osteoarthropathy). Other associations include intrathoracic lymphoma, rheumatic diseases such as systemic vasculitis, congenital cardiac disease, nasopharyngeal carcinoma, inflammatory bowel disease, and infections such as tuberculosis, subacute bacterial endocarditis and human immunodeficiency virus (HIV).

Primary HOA is rare and often familial, transmitted as autosomal dominant with variable penetrance. Autosomal recessive cases have been reported. There is a male:female predominance of 7:1. First recognised by Touraine, Solente and Gole in 1935, it is also referred to as Touraine-Solente-Gole syndrome.

Idiopathic HOA is characterised by periosteal new bone formation, clubbing, swelling of joints and thickened,



Fig. 1. The patient's right hand and wrists.



Fig. 2. X-rays of the hands and wrists.

furrowed skin. Skin manifestations include coarse facial features reminiscent of acromegaly. Bony changes consist of symmetrical irregular periosteal hypertrophy with new bone formation. Acro-osteolysis is also reported.

The differential diagnosis includes secondary HOA, acromegaly, thyroid acropachy and syphilitic periostitis.

Treatment includes the following: non-steroidal anti-inflammatory drugs; bisphosphonates have been used with success; pamidronate; tamoxifen; raloxifene with synovectomy; vagotomy may improve articular pain and swelling; and plastic surgery may improve cosmesis for the face and digital clubbing. The patient should be reassured that the condition follows an otherwise benign course.

The patient provided written consent for publication.

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