MONOSTOTIC LOCALIZATION OF PAGET DISEASE IN THE HAND

L. DE SMET, P. ROOSEN, B. ZACHEE, G. FABRY

Paget disease rarely affects the hand and when it does, there is usually a polyostotic involvement. Only a few cases have been described in the literature on monostotic localization in the hand. This paper adds two cases with solitary involvement of a metacarpal.

Keywords: Paget disease; hand; metacarpal; radiology.
Mots-clés: maladie de Paget; main; métacarpien; radiologie.

CASE 1

A 69-year-old man sustained a minor injury to his left hand. Physical examination revealed a mallet-finger-type deformity. Roentgenograms of the hand showed a small avulsion fracture at the base of the distal phalanx of the little finger. In addition there was widening with coarse trabeculation, cortical thickening and loss of cortical medullary definition in the third metacarpal (fig. 1). Serum alkaline phosphatase, calcium and inorganic phosphate were within normal limits.

Roentgenograms of the skull, lumbar spine, pelvis and femora did not identify any abnormality. Bone scanning, performed with $^{99m}\text{Te-EHDP}$, demonstrated hyperactivity over the third metacarpal of the left hand, with no other hot spots in the rest of the skeleton (fig. 2). CT-scanning of the hands revealed the same radiological features and confirmed the diagnosis of Paget disease (fig. 3). These findings were sufficient to obviate biopsy. One and a half years later, the patient is still pain free, and physical examination and x rays have remained unchanged.

Fig. 1. — Radiographs of case 1.

CASE 2

A 70-year-old man observed a painless swelling on the dorsal side of the second metacarpal of the right hand. This swelling has progressed slowly over the last 5 years. Recently, the swelling became

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mildly painful, and flexion of the metacarpophalangeal joint decreased to 70°. There were no other areas of bone pain and/or deformity.

The radiographs (fig. 4) demonstrated a grossly enlarged metacarpal with irregular thickened cortices and coarsened trabeculae. There were no obvious radiological arguments for a pathological fracture or sarcomatous degeneration. The head of the metacarpal was deformed, which could explain the decreased mobility. Serum alkaline phosphatase was within normal levels.

Bone scintigraphy with 99m Te-EHDP showed greatly increased activity over the second metacarpal, without other hot spots in the skeleton.

**DISCUSSION**

The incidence of Paget disease has been estimated to be about 3% (5, 6). Involvement of the hands has been reported infrequently, although studies with bone scintigraphy revealed a hand localization varying from 6.4% to 11.6% (1), and even up to 20% (5) of all patients with Paget disease. Those studies however only reported cases with polyostotic involvement. Monostotic localization seems to be far less frequent, probably because most of these lesions remain asymptomatic and are detected coincidentally, as in our case. The mechanical forces acting on the bones in the hand are minimal, and obvious deviation does not occur.
The typical radiological picture (1, 2, 3, 4, 6) should be diagnostic for Paget’s disease (4).

In these cases the three radiological characteristics were present: trabecular coarsening, cortical thickening and lysis. There was no homogeneous sclerosis, as described by Friedman (1) as a feature often found in monostotic hand disease.

Although biopsy may be necessary in some cases, bone scanning is a reliable method for analysis of skeletal distribution. However, it has no value for monostotic lesions. The lytic phase may weaken the bone and predispose it to pathological fracture (5).

Sarcomatous degeneration has never been described in the monostotic hand localization. Nevertheless, it was reported in the hand in a polyostotic form (1). Even in the absence of pain, a yearly x ray seems prudent. The recognition of Paget’s disease may prevent unnecessary surgery.

REFERENCES


SAMENVATTING

L. DE SMET, P. ROOSEN, B. ZACHEE, G. FABRY
Monostotische lokalisatie van de Ziekte van Paget in de hand.

De auteurs beschrijven 2 nieuwe gevallen van Ziekte van Paget in een metacarpaal van de hand. In beide gevallen was het radiologisch beeld voldoende voor de diagnose.

RÉSUMÉ

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Localisation isolée de la maladie de Paget dans la main.

Les auteurs rapportent 2 cas de maladie de Paget localisée à un métacarpien. L’aspect radiographique était dans ces cas typique et permettant de poser le diagnostic.