Paget Disease of the Hand: Radiographic Spectrum

Paget disease uncommonly affects the hand. When it does, radiographic findings are the same as when the disease occurs elsewhere. Eleven new cases that demonstrate four predominant radiographic patterns are described: homogeneous sclerosis, trabecular coarsening, cortical thickening, and lysis. The homogeneous sclerosis pattern was associated with monostotic hand disease. One patient developed bilateral phalangeal osteosarcomas. In every case, the disease process extended to the subarticular region and the affected bone was enlarged. With one exception, both ends of the bone were involved.

Paget disease, originally described by Sir James Paget in 1877 [1], is a bone disease of unknown etiology characterized by osteolysis followed by excessive attempts at repair. It is generally considered when there is radiographic evidence of cortical destruction, expansion of bone, coarsened trabeculae, and/or bone sclerosis. Paget disease usually affects bones of the skull, spine, pelvis, and lower extremities, and is distinctly unusual in the hand [2–7]. As late as 1950, it was thought that Paget disease did not occur in the hand, and that hand involvement served to differentiate Paget disease from fibrous dysplasia [3]. Our experience with a recent case of Paget disease of the hand prompted a review of the literature and a search for additional cases. We present 11 new cases and discuss the radiologic manifestations of Paget disease of the hand.

Materials and Methods

A survey of the files at Walter Reed Army Medical Center and the Armed Forces Institute of Pathology yielded 11 patients with Paget disease of the hand involving 13 hands; two patients had both hands involved. All but two cases had pathologic proof; radiologic findings were sufficiently diagnostic in those two cases to obviate biopsy.

Results

The age range of our patients was 26–82 years; all but one were more than 40 years old. There was an 8:3 male:female ratio. Symptoms were: pain only, three hands; swelling only, two hands; and both pain and swelling in two hands. Four hands had no symptoms. These clinical data were unavailable in two instances. Five patients had involvement of only a solitary hand bone and five had hand bone involvement in conjunction with typical involvement of other usual sites. One patient had one bone in each hand affected. The distribution of disease in individual hands was: proximal phalanges, seven; metacarpals, two; and middle phalanges and carpal, (pisiform), one each; also one case had bilateral involvement of multiple phalanges, metacarpals, and carpals. This last patient was the only one who had more than one bone affected in a hand (the also had widespread skeletal disease). The distal phalanges were spared completely in this series.
Fig. 1—A, Typical radionuclide scan. Markedly increased uptake in enlarged third middle phalanx. B, Anteroposterior view of same bone. Homogeneous sclerosis with few visible trabeculae.

Fig. 2.—A, Anteroposterior view. Homogeneous sclerosis with thickened trabeculae shining through in fourth proximal phalanx. B, Another patient. Asymmetric cortical thickening and medullary canal narrowing in second metacarpal. Base is spared; head shows homogeneous sclerosis.

Fig. 3.—Anteroposterior (A) and oblique (B) views of fourth proximal phalanx. Lytic pattern. Cortex is split (arrows). Biopsy of area showed poorly mineralized osteoid. Homogeneous sclerosis at base and coarsened trabeculae at head: all three phases simultaneous in one bone.

Radionuclide scans were available in two instances; both showed markedly increased uptake in the entire bone with enlargement of the bone (fig. 1A).

We found four predominant radiographic patterns: homogeneous sclerosis, five cases (figs 1 and 2A); cortical thickening, two cases (fig. 2B); lysis, two cases (fig. 3); and trabecular coarsening, two cases. All five instances of monostotic hand disease manifested the homogeneous sclerosis pattern. One patient developed osteosarcomas 3 years apart in one phalanx in each hand (fig. 4). In all instances, the affected bone was enlarged and the disease process extended to involve subarticular bone. With the exception of one case (fig. 2B), all showed involvement of both ends of the bone.

Discussion

Paget disease affects middle-aged and elderly adults with male predominance [8]. It is particularly prevalent among Caucasians in northern temperate climates [8]. In most cases, the disease is an incidental finding on radiographs obtained for other reasons [8]. Common clinical presentations are bone enlargement or deformity and pain [2, 8]. Radiologic findings have been divided into three phases: osteolysis, a combined phase with both osteolysis and sclerosis, and an osteoblastic phase with a diffusely increased bone density [2, 8]. All three phases may occur together in the same patient or even in the same bone [8].
In previously reported series of Paget disease of the hand, most patients have had no hand symptoms or signs [4]. Two cases with pathologic fracture of a hand bone have been reported [7]; also one case who had tenderness and swelling [3]. Our series had a higher incidence of symptoms and signs, with only three cases (four hands) discovered as incidental findings. Previously, hand involvement has been primarily reported as part of generalized skeletal disease [4]. However, five of our 11 patients had monostotic involvement in the hand. We found, as did Barry [2], that usually only one hand bone is involved when Paget disease affects the hands. However, Grundy and Patton [4] described a series of 11 patients in whom nine had more than one hand bone affected. These patients were all asymptomatic and hand films were obtained as part of skeletal surveys for Paget disease found elsewhere, possibly explaining the lower incidence of symptoms and higher incidence of multiple hand bone involvement in this population.

We found, as have others, that Paget disease in the hand usually affects the phalanges and metacarpals, with carpal involvement rare [2, 4], and distal phalangeal involvement almost unknown [2, 4]. The most common radiographic pattern of Paget disease in the hand is homogeneous sclerosis [3, 4, 7], which occurred in five of our 11 cases. A curious association, previously unreported and possibly coincidental, is that all of our instances of homogeneous sclerosis occurred in symptomatic monostotic disease. However, all the patterns present elsewhere in the body (cortical destruction, bone expansion, coarsened trabeculae, and bone sclerosis) were found in the hand in our series. Our findings that Paget disease in the hand generally involves both bone ends with enlargement of the whole bone has not been emphasized previously [3, 4, 7]. Sarcomatous degeneration, which likely occurs in less than 1% of patients with Paget disease [2, 8], can occur in the hand as well. Our case may be the first reported instance of malignant degeneration of Paget disease in the phalanges.

Hand bone findings in Paget disease are the same as in Paget disease occurring elsewhere, with the addition that both ends of the bone are almost always affected. The initial phase of osteolysis is seen in figure 3. This may be followed by repair which is manifested primarily by cortical thickening (fig. 2B), either symmetrically or asymmetrically, which represents mineralization of previously unmineralized osteoid present in the lytic phase, or by trabecular coarsening or both. When trabecular coarsening occurs, the trabeculae are decreased in number but those remaining are thickened.

The disease may arrest at this point or progress to homogeneous sclerosis, the third phase (fig. 1B) which corresponds to the mineralization of excess, previously poorly mineralized osteoid. At this stage there is usually, but not always, residual trabecular thickening visible (fig. 2A) and loss of distinction between cortical and medullary bone is common. The disease may present radiographically in the hand at any stage. There may be visible progression from the lytic to the sclerotic phase [2, 8], and all phases may occur simultaneously (fig. 3).

REFERENCES
1. Paget J. On a form of chronic inflammation of bones (osteitis deformans). Med Chir Trans 1877;60:37–64