Case report 806

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Fig. 1A–C. Chronological radiographic sequences of the right fifth metacarpal bone during the different stages of disease. A lytic and expansile lesion within the medullary canal is seen (A, C). After treatment, a sclerosing reparative pattern is present (B)

Clinical information

A 43-year-old man presented in January 1989 with a 5-year history of a nontender enlargement of the right fifth metacarpal area. Physical examination showed a swelling in this region without cutaneous erythema. Results of routine laboratory tests, including tests for serum alkaline phosphatase, calcium, and phosphorus levels, were within normal limits.

A subchondral location was noted. Enchondroma was the clinical and radiographic diagnosis. Extensive curettage of the medullary canal was performed, after which the cavity was packed with a cancellous bone graft obtained from the right iliac crest. During the curettage a thick fluidlike material was observed, and the bone fragments obtained were submitted for histological analysis. A radiological study performed 3 months later revealed a trabecular, sclerosing reparative pattern (Fig. 1B). Three years later, in January 1992, a new roentgenogram disclosed an expansile lytic lesion in the same anatomical zone, very similar radiographically to the original lesion (Fig. 1C). Curettage was performed at that time. Periodic skeletal surveys were unremarkable except for the findings in the hand.

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Diagnosis: Monostotic Paget’s disease of the hand (fifth metacarpal)

Grossly, 1.0 × 0.8 × 0.4 and 1.0 × 0.9 × 0.6 cm bone fragments containing cortex and medulla were recovered after the first and second curetages, respectively. Histologically the lesions were similar. Most of the bone trabeculae were thin and irregular (Fig. 2) with prominent osteoblastic rimming and osteoid seams. Osteoclasts were also prominent and related to areas of resorption (Fig. 3). The bone matrix showed increased numbers of cement lines arranged in a mosaic pattern. Although most of the bone tissue was of the woven type, focal lamellar bone was also present (Fig. 3). Intertrabecular marrow spaces were replaced by fibrous tissue. No inflammatory cells were present.

Discussion

Paget’s disease (osteitis deformans) is a chronic disorder of unknown etiology characterized by excessive osteoclastic resorption followed by marked but inadequate bone formation. Immunohistochemical, ultrastructural, and in situ hybridization studies suggest that paramyxovirus is associated with this disorder [3, 9, 13, 14].

Paget’s disease usually affects middle-aged and elderly adults. Its incidence increases with age, affecting 3% of those persons over 40 years of age [2], especially males, in whom the disease predominates [10].

The polyostotic form of the disease is well known to involve the bones of the skull, spine, pelvis, and lower extremities. However, involvement of the hand has been infrequently reported [1, 2, 5, 6, 11, 18]. Of 136 and 382 cases of polyostotic Paget’s disease reviewed by Ravault et al. [12] and Louyot and Gaucher [7], respectively, 3 patients in each series displayed disease in the hand.

When polyostotic Paget’s disease affects the hand, it usually involves one bone [2, 4]. Nevertheless, Grundy and Patton [5], in a series of 11 cases of polyostotic Paget’s disease involving the hand, described 9 patients with more than one bone of

Fig. 2. Irregular and thin bone trabeculae with ossification of woven-type bone

Fig. 3. Bone trabeculae with woven and lamellar patterns. An increased number of osteoclasts is seen. Intertrabecular marrow space is occupied by fibrous tissue.