Chondrosarcoma in Paget’s disease of bone

Sirs,

A 60-year-old patient was diagnosed with Paget’s disease of bone (PDB) after having a fracture of the humerus. He had suffered another fracture at the same site when he was 30.

Radiographs showed a predominantly osteolytic pagetic lesion involving the left proximal humerus. Bone scintigraphy showed uptake on the left humerus, the seventh rib, the eighth dorsal vertebra, lumbar vertebrae and the sacrum (Fig. 1). The serum alkaline phosphatase concentration was 1026 IU/L (normal range, 98-270). He was treated first with alendronate 30 mg daily for 6 months and after that with risedronate 30 mg daily for 2 months, and the serum alkaline phosphatase decreased to almost normal values. Two years later a painful mass was observed on the proximal region of the left superior limb. The serum alkaline phosphatase concentration was 599 IU/L, slightly increased respect to previous analysis. Plain radiographs of the humerus were similar to the previous x-ray, except for an image of increased density out of the cortical bone near the former fracture. MRI showed an area of decreased signal intensity in both T1 and T2-weighted images with irregularity of the cortical and a soft tissue mass, which involved the surrounding muscles and subcutaneous cellular tissue. Histopathologically this mass was a grade 3 chondrosarcoma. The patient was treated with radical surgery in addition to chemotherapy and local radiotherapy. Fifteen months later he suffered pulmonary metastasis. Sarcomatous transformation in PDB is a very rare event nowadays (0.7% to 5%) (1, 2), probably due to a good control of the disease. The estimated 5-year survival rate is about 3-8% (1, 3-6), although a few patients enjoy a longer survival (6, 7). Treatment must be instituted as early as possible, and include radical surgery and subsequent radiotherapy and chemotherapy.

References