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CASE REPORT

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Benign tumefactive soft tissue extension from Paget's disease of bone simulating malignancy

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Abstract Osteosarcoma is a frequently fatal complication of Paget's disease of bone typically manifesting radiographically as a lytic lesion with soft tissue extension. A clinically worrisome, but benign manifestation of Paget's disease simulating malignancy because of an extraosseous mass is reported.

Keywords Paget's disease · Pseudotumor · Femur · Radiography · MRI

Introduction

Secondary tumors are rare but well-described complications that may develop in bone affected by Paget's disease [1]. The most common malignancy is osteosarcoma, with giant cell tumors and lymphomas also reported [1]. Development of osteosarcoma in Paget's disease may be heralded by increased, unremitting pain, or simply by a change in the pain pattern [2]. Radiographic studies frequently show a lytic lesion with cortical destruction and soft tissue extension [3]. This report describes a patient with a soft tissue mass arising from bone with Paget's disease which simulated a malignancy, but which was found histologically to be uncomplicated Paget's disease.

Case report

A 59-year-old man without prior diagnosis of Paget's disease was found on annual laboratory testing to have an elevated alkaline phosphatase level of 291 U/l (reference range 38–126 U/l). He reported mild but worsening right knee pain of 4 years' duration following a minor car accident. A bone scan revealed a nonspecific, focally intense, isolated area of increased uptake in the bone involving the epiphysis, metaphysis, diaphysis, and adjacent soft tissue of the right distal femur. Radiographs demonstrated Paget's disease of the distal femur, an eccentrically located periosteal reaction, and a soft tissue mass arising from the medial aspect of the distal femoral metaphysis (Fig. 1). A magnetic resonance imaging (MRI) study of the involved region showed abnormal intracortical signal through the area of thickened cortex involved by Paget's disease and the adjacent soft tissue mass overlying the medial cortex.

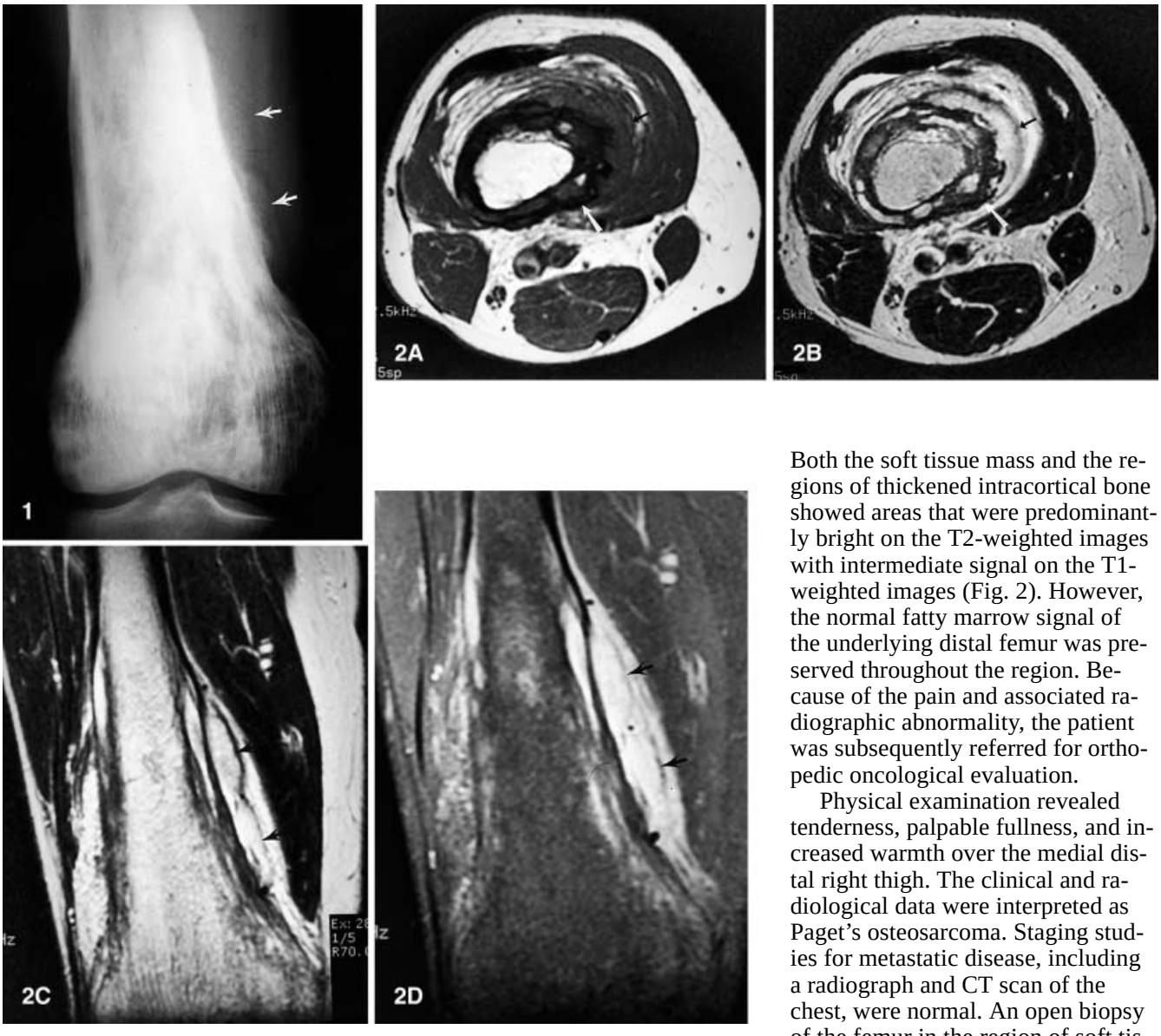

Fig. 1 Plain anteroposterior radiograph of the right distal femur shows typical sclerotic pagetoid bone involving the entire distal femur. Periosteal reaction and associated soft tissue extension is present in the medial distal femoral metaphysis arising from within a typical area of pagetoid bone (*arrows*)

Fig. 2A–D Magnetic resonance images of the distal femur. **A** Axial T1-weighted (TR533, TE18) spin echo sequence at the distal metaphyseal level shows the typical low signal coarsened trabeculae of the distal femoral cortex (*white arrow*) associated with irregularity of the medial cortical surface and an overlying intermediate-signal soft tissue mass (*black arrow*) beneath the quadriceps musculature. Note that while both the soft tissue mass and the thickened cortex are of low to intermediate signal, the normal bright fatty marrow signal of the medullary canal is preserved. **B** Axial T2-weighted (TR5600, TE110) fast spin echo sequence shows the bright signal characteristic of the soft tissue mass (*black arrow*). Within the thickened cortex (*white arrow*) are similar areas of bright signal. There is no evidence of hemorrhage. **C** Coronal T2-weighted (TR3300, TE110) fast spin echo sequence shows the bright signal of the periosteally based soft tissue extension (*arrows*). **D** Coronal T1-weighted (TR566, TE18) fast spin echo, fat suppression, with gadopentetate dimeglumine contrast shows high signal intensity due to increased vascularity and vascular permeability within the mass (*arrows*) but preservation of normal marrow signal within the medullary canal

Both the soft tissue mass and the regions of thickened intracortical bone showed areas that were predominantly bright on the T2-weighted images with intermediate signal on the T1-weighted images (Fig. 2). However, the normal fatty marrow signal of the underlying distal femur was preserved throughout the region. Because of the pain and associated radiographic abnormality, the patient was subsequently referred for orthopedic oncological evaluation.

Physical examination revealed tenderness, palpable fullness, and increased warmth over the medial distal right thigh. The clinical and radiological data were interpreted as Paget's osteosarcoma. Staging studies for metastatic disease, including a radiograph and CT scan of the chest, were normal. An open biopsy of the femur in the region of soft tissue extension showed bone fragments composed of haphazardly organized bone, the "mosaic pattern" characteristic of Paget's disease (Fig. 3). The regions of nonuniformity consisted of tumefactive collagenous soft tissue extension of the Paget's disease process. There was no evidence of osteosarcoma, giant cell tumor, or lymphoma.

The patient was treated with pamidronate for the Paget's disease. Within 1 year, the symptoms resolved. Serial radiological evaluation of the distal femur revealed progressive and complete ossification of the

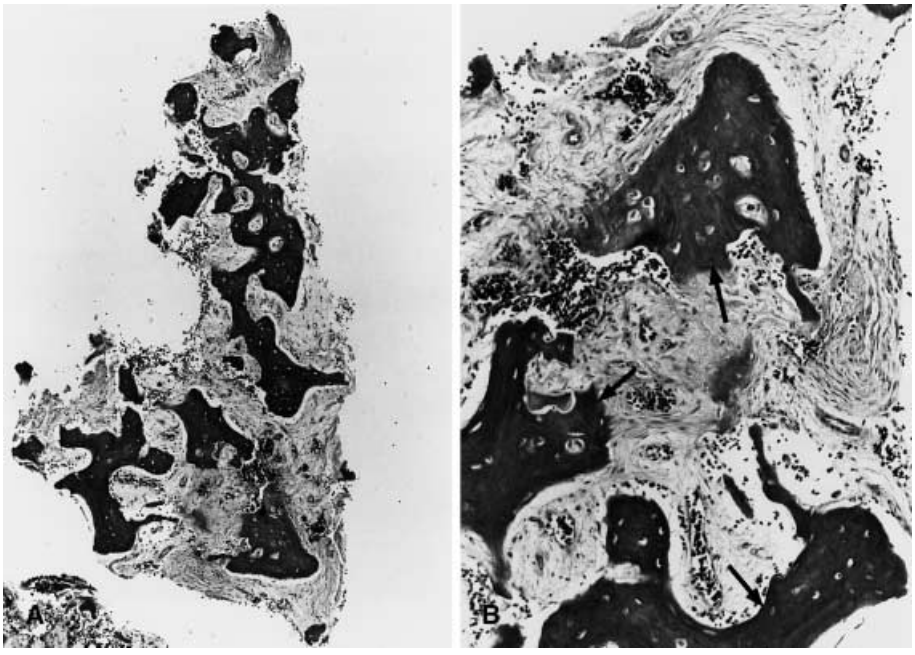

Fig. 3A, B Histological sections demonstrate bone composed of haphazardly organized, thickened trabeculae (*arrows*). The soft tissue component is moderately cellular but without atypia. There is no evidence of osteosarcoma, lymphoma or metastatic disease (hematoxylin and eosin; A ×40, B ×80)

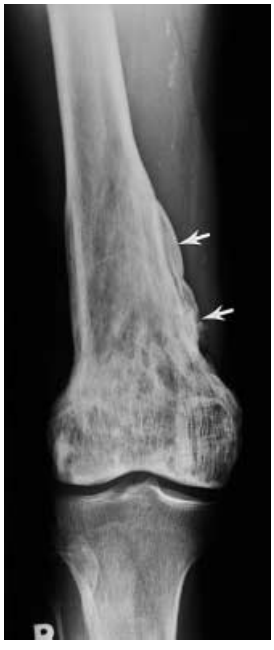

Fig. 4 Plain anteroposterior radiograph of the right distal femur following treatment shows complete ossification of the pagetoid soft tissue extension (*arrows*)

bone defect and adjacent soft tissue mass (Fig. 4). The alkaline phosphatase level normalized over the same time period. The patient has remained well, without recurrence of the pain, lytic lesion, or soft tissue mass, over the subsequent 3-year follow-up.

Discussion

The incidence of sarcomatous transformation in pagetoid bone is estimated to be approximately 0.7–0.95% [4, 5]. Despite the low transformation rate, the prevalence of Paget's disease in the population over 40 years of age makes sarcomatous transformation a significant health risk [6]. The development of increased pain or a change in the nature of pain in bone with Paget's disease may signal the development of osteosarcoma [2, 7]. Osteosarcoma in Paget's disease typically has a lytic appearance on radiography, with cortical breakthrough and an associated soft tissue mass, and is most likely to occur in the femur, humerus, and pelvis with nearly equal frequency, which generally follows the most common sites of Paget's disease [5, 7, 8, 9].

MRI is an important tool in demonstrating soft tissue extension superimposed upon Paget's disease [10]. Two important clues on MRI favoring a diagnosis of malignancy in Paget's disease are transformation of marrow fat signal showing a low signal on T1-weighted images and the presence of a soft tissue mass. A soft tissue mass associated with loss of normal marrow signal is highly suggestive of sarcoma and should always prompt biopsy [10]. In this case, the absence of familiarity with benign tumefactive masses in Paget's disease combined with failure to recognize the preservation of normal intramedullary marrow signal led to the erroneous conclusion that this patient might have a malignancy. Biopsy proved otherwise.

Pagetoid osteosarcoma is not the only entity associated with Paget's disease that may cause soft tissue extension. Benign soft tissue extension in Paget's disease has been reported rarely [10, 11, 12]. In one previously reported case, the soft tissue mass was attributed to fracture. The fracture also resulted in loss of normal marrow signal on MRI scans and multiple biopsies were needed to exclude a sarcoma in light of the MRI findings and clinical symptoms [10].

Only one previously reported case of pseudotumor in Paget's disease has been described in detail, but this was in the pre-MRI era. In that case, a 72-year-old woman presented with left knee and groin pain of several weeks' duration. Physical examination revealed warmth and erythema of the left knee. Laboratory values were remarkable for an elevated alkaline phosphatase level of 650 international milliunits (reference range 10–32 milliunits). A partial skeletal survey showed coarse trabeculae and increased density of the femur, pelvis, and lumbar spine. Some thoracic and cervical vertebrae were enlarged and sclerotic. The medial aspect of the distal femur showed a lobular, periosteal, nonmineralized soft tissue mass. The cortex beneath the lesion was intact. The surrounding femur was heavily involved by sclerotic Paget's disease. A presumptive diagnosis of sarcoma complicating Paget's disease was rendered, and a biopsy was performed. Histological examination of the biopsy tissue showed Paget's disease without evidence of neoplasia. A follow-up roentgenogram of the lesion showed no significant change in appearance of the lesion over a follow-up period of 22 months [12].

Giant cell tumors may also occur within Paget's disease and result in soft tissue extension. Giant cell tumors most often occur in the facial bones of elderly patients with severe Paget's disease. An expansive lytic lesion is observed, occasionally with a soft tissue mass [11].

Benign periosteal bone proliferation can produce a soft tissue mass in the setting of Paget's disease, as in this case [11]. The key to distinguishing between this benign pseudotumor and a sarcoma arising from Paget's disease is the recognition on MRI of preservation of normal fatty marrow signal in the benign process [10]. Close attention should be paid to the T1-weighted marrow signal on MRI studies to determine whether the fatty signal in the underlying marrow cavity involved by Paget's disease is preserved, as in the reported benign tumefactive pseudotumor, or replaced, as in secondary tumors arising within Paget's disease [10].

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