

## CASE REPORT

# Femoral Chondrosarcoma Complicating Paget's Disease of Bone

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Malignant transformation associated with Paget's disease of bone has an incidence of approximately 1%, which increases to 10% in long-term poliostotic Paget's disease. Approximately 75% of patients who develop Paget's sarcoma have the disease years before sarcoma onset and 90% have poliostotic involvement.<sup>1</sup>

This article reports a patient with Paget's disease who presented with extensive lysis in the left proximal femoral shaft and a vigorous palpable mass. Following incisional biopsy, a grade I chondrosarcoma associated with Paget's disease was diagnosed.

## CASE REPORT

A 72-year-old woman with a 12-year history of Paget's disease presented with occasional pain in the left thigh and knee of 8 months' duration. Gait and hip motion were impaired. Clinical examination revealed a rigid palpable mass in the left thigh with a notable reduction in range of movement in all planes.

Radiographs revealed a mixed osteoblastic and lytic lesion involving the proximal femur with visible extension into the soft tissues (Figure 1A). Radiographic changes of Paget's disease were sclerotic, representing a late phase of disease, having an accented trabecular "honeycomb" pattern with extensive widening and complete blurring of the corticomedullary interface. Destruction of the cortex was noted without periosteal reaction.

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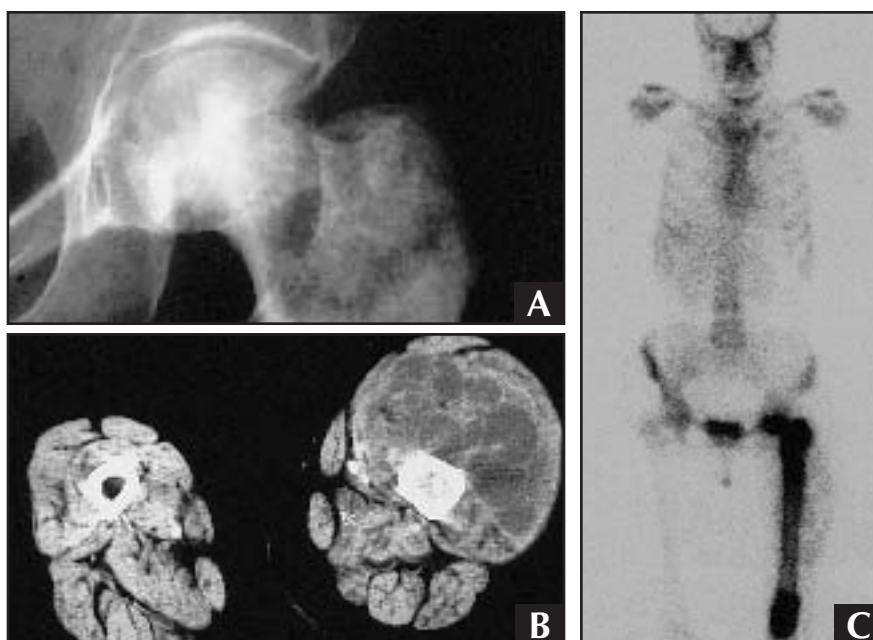


Figure 1: Radiograph showing a mixed osteoblastic and lytic lesion involving the proximal femur (A). CT of the femur shows a wide mass extending from the medullary to the cortical bone (B). Bone scan with 99mTc-MDP showing increased radionuclide uptake within both the tumor and Pagetic bone (C).

Computed tomography (CT) revealed a wide mass extending from the medullary to the cortical bone and a vigorous mass in the surrounding soft tissue (Figure 1B). Technetium-99m methylene diphosphonate (99mTc-MDP) bone scan showed increased radionuclide uptake within the tumor and Pagetic bone (Figure 1C). Hematochemical tests demonstrated elevated values for alkaline phosphatase and normal values for lactic dehydrogenase. Radiographs, bone scan, and CT of the chest showed no pulmonary involvement.

Incisional biopsy of the mass was performed, and the patient underwent total femur resection followed by reconstruction with a

total femoral prosthesis including the hip and knee (Figure 2).

## Histology

Gross examination revealed a thickened cortical bone and a large neoplastic mass covered with fibromuscular tissue. The tumor was located at the upper third of the femur (10-cm transverse diameter) and extended to the medullary cavity (30-cm longitudinal diameter). In addition, the tumor extended into the surrounding soft tissue with cortex destruction (Figure 3). The cut surface was gelatinous and gray to tan. The entire specimen was processed for gross examination according to standard guidelines.<sup>2</sup> One section was taken for each

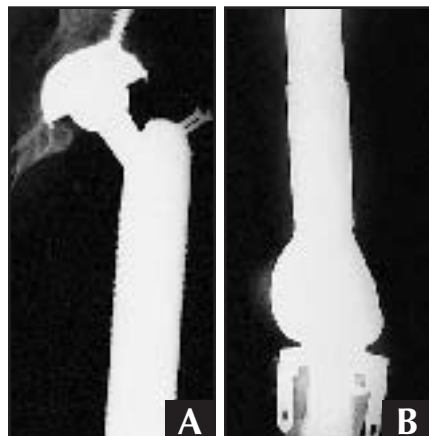


Figure 2: Femur reconstruction with a total femoral prosthesis including the hip (A) and knee (B).

centimeter of the tumor diameter and representative sections from the margins.

Microscopically, the tumor displayed a multinodular pattern. The nodules were well circumscribed and consisted of rounded or slightly elongated cells of uniform size and shape. These cells were embedded in mucoid material (Figure 3). The neoplastic cells exhibited chondroblastic features (small hyperchromatic nucleus and narrow eosinophilic cytoplasm). Rare cartilage cells with atypical features and rare mitotic figures were recorded. The mucoid material developed positive Alcian blue stain and the Periodic Acid Schiff stain revealed glycogen within tumor cells. The tumor extended to the soft tissue (Figure 3). Adjacent to the tumor, the bone displayed features characteristic of Paget's disease. These were comprised of a "mosaic" appearance of the entrapped bone trabeculae. No neoplastic bone or osteoid formation was present.

Extraskelatal myxoid chondrosarcoma was diagnosed. Because slides from the first biopsy were unavailable, the current slides were sent to the referral center; in both instances, pathologic examination displayed the same findings.

## DISCUSSION

Chondrosarcoma has rarely been associated with Paget's disease. More specifically, the range of chondrosarcomas reported in Paget's disease (3%-15%) may be related to the identification of minimal osteoid production by the tumor, which classifies the lesion as

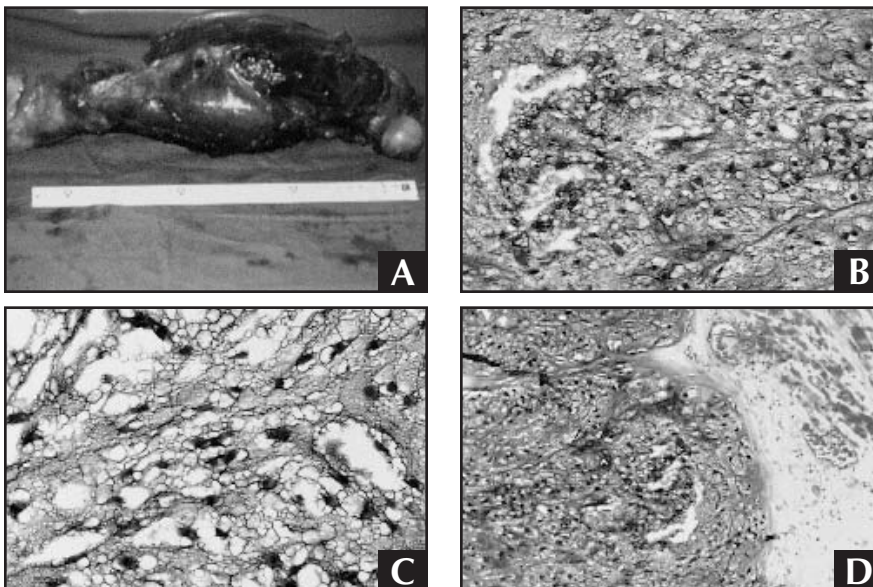


Figure 3: Macroscopic appearance of the tumor (A). Microscopic section from the tumor. Neoplastic cells embedded in mucoid material (hematoxylin-eosin, original magnification  $\times 250$ ) (B). A higher magnification of the previous microphotograph showed that neoplastic cells are characterized by small hyperchromatic nucleus and narrow eosinophilic cytoplasmic rim (hematoxylin-eosin, original magnification  $\times 400$ ) (C). Photomicrograph showing involvement of the quadriceps by tumor nodules (hematoxylin-eosin, original magnification  $\times 250$ ) (D).

osteogenic sarcoma, even if it is extensively chondroblastic. In the 653 Paget's sarcomas reported in the literature, only 19 cases of chondrosarcoma were reported in which the preferred locations (femur and humerus) were in agreement with the main anatomic sites of Paget's sarcoma distribution. Other reported tumor locations included the tibia, vertebra, scapula, and ribs.<sup>3,4</sup>

Painful, destructive lesions with cortical breakthrough and soft-tissue masses in patients with long-standing Paget's disease suggest the possibility of neoplastic changes.<sup>5</sup> Rarely, however, are coincidental metastases considered in the differential diagnosis. In addition, aggressive Paget's disease may simulate a sarcoma—the so-called pseudomalignant lesions in Paget's disease.<sup>6</sup> A needle or incisional biopsy confirms or excludes the diagnosis of sarcomatous changes.

Paget's sarcomas have been associated with an unfavorable prognosis; very few patients were long-term survivors in the Paget's sarcoma group and no prognostic factors have been identified.<sup>7,8</sup> Of the 19 cases of chon-

drosarcoma associated with Paget's disease in the literature, survival ranged from 9-66 months. Sixteen months postoperatively, our patient is alive.

## REFERENCES

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