Skeletal Radiol (1997) 26:497–500

CASE REPORT

F. Brandolini
P. Bacchini
M. Moscato
F. Bertoni

Chondrosarcoma as a complicating factor in Paget's disease of bone

F. Brandolini (✉)[1] · M. Moscato
Department of Orthopedics,
Rizzoli Orthopedic Institute,
I-40139 Bologna, Italy

P. Bacchini · F. Bertoni
Department of Surgical Pathology,
Rizzoli Orthopedic Institute,
Bologna, Italy

Present address:
[1] Clinica Ortopedica, Università di Ferrara,
Arcispedale S. Anna, Corso Giovecca 203
I-44100 Ferrara, Italy

Abstract A 65-year-old male patient with an 8-year history of polyostotic Paget's disease complained of shoulder pain that started 6 months prior to admission. An extensive lytic area was identified in the right proximal humerus along with Paget's disease. There was cortical destruction and a soft tissue mass. Following an incisional biopsy, a diagnosis of grade 2 chondrosarcoma associated with Paget's disease was made. The histologic identification of chondrosarcoma associated with Paget's disease is rare. However, the presence of a calcified matrix in a destructive lesion associated with Paget's disease should alert the radiologist and the pathologist to the possibility of a chondromatous differentiation taking place in the sarcoma associated with Paget's disease. The histologic evaluation of the lesion will form the basis for the diagnosis.

Key words Chondrosarcoma · Paget's chondrosarcoma · Sarcoma in Paget's disease

Case report

A 65-year-old man had an 8 year history of Paget's disease. He was admitted to our institute in February 1995. He complained of having suffered occasional pain in the right arm and shoulder in the 6 months prior to admission.

The patient had impaired shoulder motion and suffered pain principally at night. The clinical evaluation showed alteration in the right shoulder anatomical outline together with a notable reduction of the range of movement in all planes (abduction was 60°).

An X-ray examination showed a predominantly blastic lesion involving the entire humerus, which had a puffed, confluent, sclerotic appearance (Fig. 1). CT revealed a coarse osteolytic area in the epiphyseal-metadiaphyseal humerus with wide cortical erosion posteriorly. A huge soft tissue mass with a longitudinal extension of 10 cm involved the subspinate and deltoid muscles (Fig. 2).

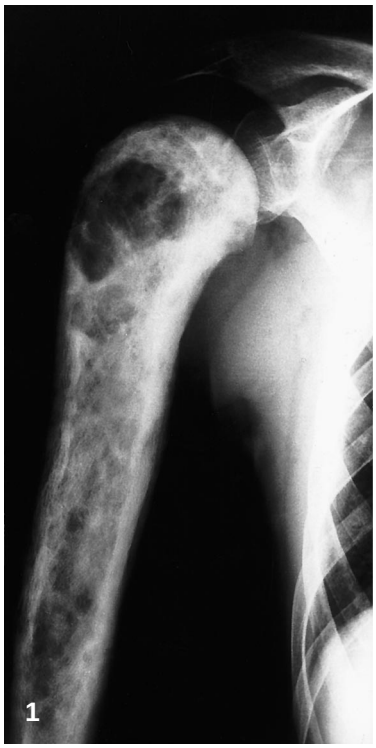
Radiographs and a bone scan showed involvement of the first thoracic vertebrae and the right hemipelvis. MR images showed a wide mass extending from the medullary to the cortical bone with a mass in the surrounding soft tissue.

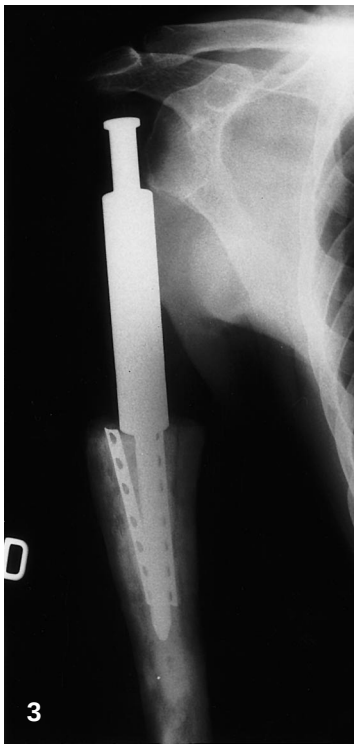
Hematochemical tests demonstrated elevated values for alkaline phosphatase and normal values for lactic dehydrogenase. CT of the chest showed no pulmonary lesions.

After performing an incisional biopsy, a wide extra-articular resection was done, followed by reconstruction with a cemented modular resection shoulder (MRS) prosthesis and two plates in order to reinforce the implant (Fig. 3).

Gross features

The gross appearance of the surgical specimen coronal cut showed thickening of the cortical bone (1 cm) and a huge mass completely covered with fibro-muscular tissue (wide surgical margins). The tumor had a multilobulated, destructive appearance. It was gelatinous and soft with a yellowish-white color. The largest part of the tumor was located at the upper end of the humerus (a transverse diameter of 10 cm). It extended down the medullary cavity (a longitudinal diameter of 9 cm) and expanded into the soft tissue with destruction of the cortex.





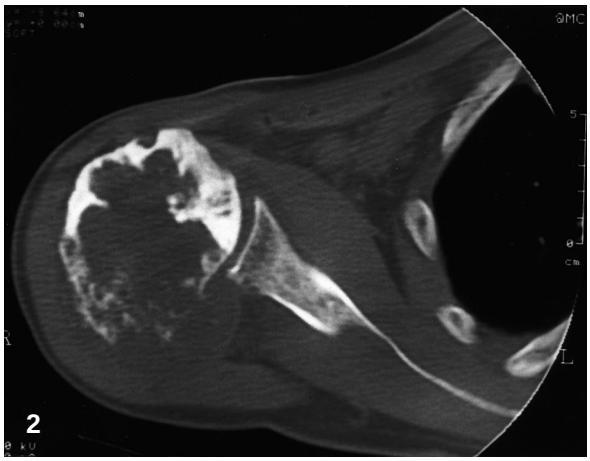


Fig. 1 AP view: pagetic bone ("cloudy aspect") with a large lytic area showing irregular borders

Fig. 2 CT scan showing the lytic area: there is destruction of the cortex and extension into the soft tissue, where a mass showing irregular calcified foci is detectable

Fig. 3 An extra-articular resection of the proximal humerus was performed. Plain roentgenograms show the proximal humerus reconstructed by a cemented prosthesis and plates

Table 1 Chondrosarcoma in Paget's disease: treatment and prognosis in 19 reported cases (*ns* not specified)

Author	Year	No. of cases	Site	Follow-up	Treatment	Metastasis
Mathey-Cornat (reported in Schajowicz) [11]	1930	1	Scapula	Dead months (ns)	ns	no
Wanke (reported in Schajowicz) [11]	1932	1	Femur	Dead months (ns)	ns	yes
Schajowicz [11]	1942	1	Proximal femur	Dead after 64 months	Radio-therapy	ns
Barry [12]	1961	1	Proximal humerus	Dead after 9 months	Amputation	yes
McKenna et al. [13]	1966	2	1 Rib, 1 tibia	Dead after less than 60 months	Not treated	ns
Serre et al. [16]	1975	1	Humerus	Dead but ns	ns	ns
Barry [14]	1980	3	2 Humerus 1 ns	Dead after less than 60 months	ns	ns
Schajowicz et al. [15]	1983	5	2 Femur, 1 vertebra, 2 tibia	Dead after less than 60 months	ns	ns
Smith et al. [4]	1984	1	Humerus	Dead after 66 months	Amputation	yes
Moore et al. [5]	1991	3	2 Femur, 1 vertebra	Dead after less than 60 months	ns	ns

Histologic features

The lesion had a biphase appearance. Irregular, coarse, deformed, bony trabeculae surrounded a huge, multilobulated neoplasm. It was centered in the metaphysis, and extended into the diaphysis. The lesion destroyed the host trabeculae of the medullary and the cortical bone and extended into the soft tissue (Fig. 4).

There was extensive fibrosis and vascularization of the host bone marrow. Osteoblasts and giant cells bor-

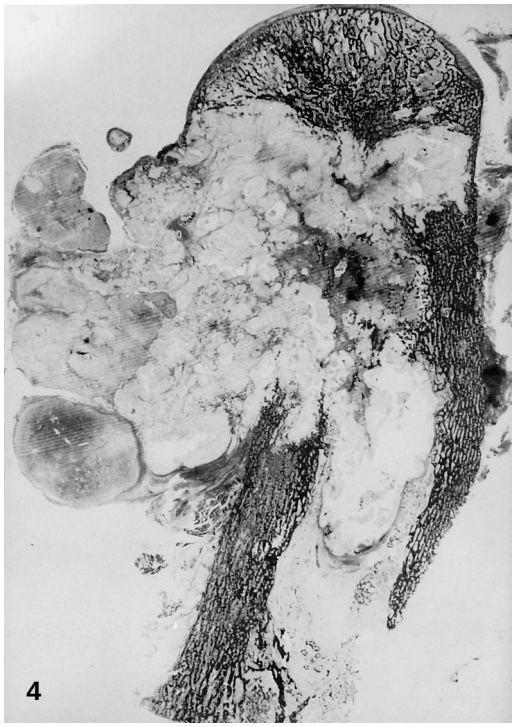


Fig. 4 View of the cut section: a large, multilobulated tumor is evident, which has destroyed the metaphysis of the proximal humerus and extends toward the diaphysis. Breaching of the cortex and extension into the soft tissue was also present (H & E ×1)

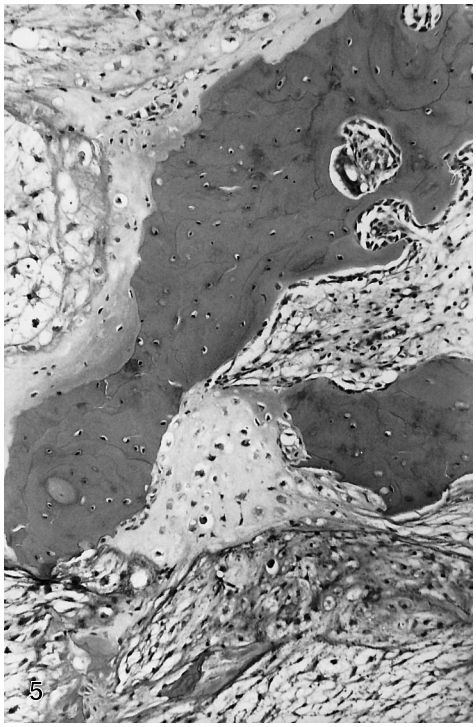


Fig. 5 Permeative growing pattern of the grade 2 chondrosarcoma. The entrapped bony trabeculae show a "mosaic" appearance (H & E ×160)

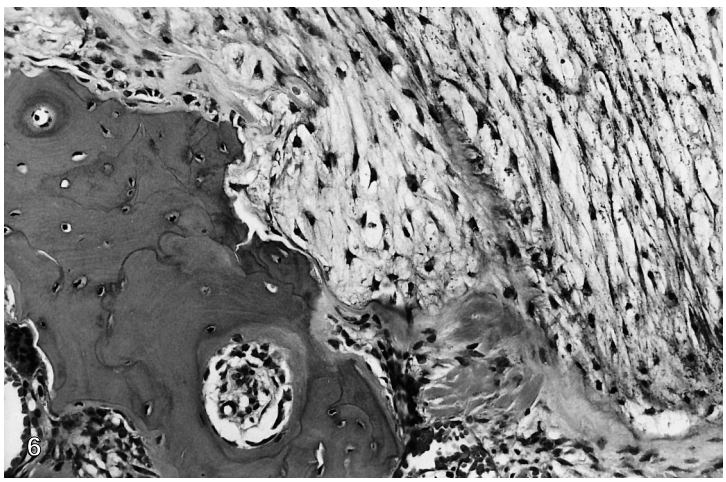


Fig. 6 Grade 2 chondrosarcoma side by side with pagetic bone. The chondrosarcoma is characterized by pleomorphic oval spindle cells with hyperchromatic nuclei. The bone shows "mosaics" and giant cells (H & E ×250)

dered the bony host trabeculae. Cement lines in a " mosaic arrangement" were detected in the trabeculae. Lying adjacent to these areas, grade 2 and 3 chondrosarcomas were detected that showed features characteristic of Paget's disease. Malignant cells were present in the hyaline cartilage, some of which had a myxoid and spindle-cell appearance. They grew in a diffuse manner between the host trabeculae (Fig. 5). There was neither osteoid nor bone production by the malignant cells; this was a useful hint in ruling out chondroblastic osteosarcoma (Fig. 6).

Discussion

The incidence of malignant transformation in Paget's disease is approximately 1%. An incidence of approximately 10% is reported in long-term, poliostotic Paget's disease [1]. For people over the age of 50, Paget's disease is the most common precursor of bone sarcomas (about 25% of cases). About 75% of the patients who develop Paget's sarcoma have documented Paget's disease years before the onset of the sarcoma, and 90% of these have polyostotic involvement [1].

The range of histologic types reported in the literature varies considerably [2]. More specifically, the range of chondrosarcomas reported in Paget's disease (3–15%) may be related to the identification of osteoid production. The identification of minimal osteoid production by the

undifferentiated malignant cells classifies the lesion as osteogenic sarcoma, even if it is extensively chondroblastic. Chondrosarcoma has rarely been associated with Paget's disease. In the Mayo Clinic files, Paget's disease was associated with osteogenic sarcoma in 54 patients, with fibrosarcoma in 6, with malignant fibrous histocytoma in 3, and with malignant lymphoma and giant cell tumors in 1 patient each.

No chondrosarcomas associated with Paget's disease were reported [3]. In the series of Paget's sarcomas (*n*=28) seen at the Rizzoli Institute, only one case (the present case) was detected. In the 653 Paget's sarcomas reported in the literature, there were 19 cases of chondrosarcoma (Table 1).

The preferred locations (femur 6 cases, humerus 4 cases) were in agreement with the main anatomic sites of distribution of Paget's sarcomas [2]. It is worth noting that no Paget's chondrosarcomas were reported, even though the pelvis is the preferred location of both Paget's sarcoma and chondrosarcoma.

Painful, destructive lesions with cortical breakthrough and soft tissue masses in pagetic bone suggest the possibility of neoplastic changes [4, 5]. However, rarely, coincidental metastases must be considered in the differential diagnosis [6]. Furthermore, occasionally, very aggressive Paget's disease may simulate a sarcoma [7, 8]: the so-called pseudomalignant lesions in Paget's disease [9]. A needle biopsy or incisional biopsy will confirm or exclude 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 were identified [10]. In particular, the three patients with chondrosarcoma and Paget's reported by Moore et al. were not universally high grade, but survival was poor [5]. Of the 19 cases of chondrosarcoma associated with Paget's disease in the literature, survival information was available for 16 patients, and varied from 9 to 66 months.

Of these 16 patients, the survival period was specified in three patients: 64, 9, 66 months, respectively [4, 11, 12]. In the remaining 13 patients, the exact survival period was not reported, but it was stated that it was less than 60 months [5, 13, 14, 15]. However, all the patients with chondrosarcoma associated with Paget's disease died (Table 1).

Our patient was alive and on chemotherapy 1 year after the surgical treatment.

References

1. Mirra JM, Brien EW, Tehranzadeh J. Paget's disease of bone: review with emphasis on radiologic features. II. Skeletal Radiol 1995; 24: 173–184.
2. Hadjipavlov AG, Lander PH. Paget's disease of the spine. J Bone Joint Surg Am 1991; 73: 1376–1381.
3. Unni KK. In: Dahlin's Bone Tumors: general aspects and data on 11.087 cases. Lippincott-Raven, 1996:162–163.
4. Smith J, Botet JF, Yeh SDJ. Bone sarcomas in Paget's disease: a study of 85 patients. Radiology 1984; 152: 583–590.
5. Moore TE, King AR, Kathol MH, El-Khoury GY, Palmer R, Downey PR. Sarcoma in Paget's disease of bone: clinical, radiologic, and pathologic features in 22 cases. AJR 1991; 156: 1199–1203.
6. Igou D, Sundaram M, Mc Donald DJ, Janney C, Chalk DE. Appendicular metastatic prostate cancer simulating osteosarcoma, Paget's disease, and Paget's sarcoma. Skeletal Radiol 1995; 24: 447–449.
7. Greditzer HG, McLeod RA, Unni KK, Beabout JW. Bone sarcomas in Paget's disease. Radiology 1983; 146: 327–333.
8. Monson DK, Finn HA, Dawson PJ, Simon MA. Pseudosarcoma in Paget's disease of bone. J Bone Joint Surg Am 1989; 71: 453–455.
9. Bowerman JW, Altman J, Hughes JL, Zadek RE. Pseudo-malignant lesions in Paget's disease of bone. AJR 1975; 124: 57–61.
10. Wick MR, Siegal GP, Unni KK, McLeod RA, Greditzer HG. Sarcomas of bone complicating osteitis deformans (Paget's disease).Fifty years' experience. Am J Surg Pathol 1981; 5: 47–59.
11. Schajowicz F. La degeneracion sarcomatosa de la osteitis deformante de Paget. Rev Ortop 1942; 12: 131–148.
12. Barry CH. Sarcoma in Paget's disease of bone in Australia. J Bone Joint Surg Am 1961; 43: 1122–1134.
13. McKenna RJ, Schwinn CP, Soong KY, Higinbotham NM. Sarcomata of the osteogenic series (osteosarcoma, fibrosarcoma, chondrosarcoma, parosteal osteogenic sarcoma, and sarcomata arising in abnormal bone). J Bone Joint Surg Am 1966; 48: 1–26.
14. Barry HC. Orthopedic aspects of Paget's disease of bone. Arthritis Rheum 1980; 23: 1128–1131.
15. Schajowicz F, Santini Araujo E, Berenstein M. Sarcoma complicating Paget's disease of bone. J Bone Joint Surg Br 1983; 65: 299–307.
16. Serre H, Morlock G, Sany J. La dégénérescence sarcomateuse de la maladie de Paget. In: La maladie de Paget, 1er Colloque Regional de Patologie Locomotrice, Montpellier, 1974. Paris: Galliena, 1975: 174–190.