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CASE REPORT

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Dedifferentiated parosteal osteosarcoma with rhabdomyosarcomatous differentiation

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Abstract Dedifferentiated parosteal osteosarcomas are characterized histologically by the combination of low-grade fibroblastic osteosarcoma admixed with a high-grade component that typically has the appearance of malignant fibrous histiocytoma or osteosarcoma. Herein we report a case of dedifferentiated parosteal osteosarcoma of the distal femur, in which the high-grade component consisted of rhabdomyosarcoma. To our knowledge, a rhabdomyosarcomatous component has not been described previously in a dedifferentiated parosteal osteosarcoma. The clinical, radiologic, and pathologic features of this rare type of surface osteosarcoma are described.

Key words Parosteal osteosarcoma, dedifferentiation, rhabdomyosarcoma · Parosteal osteosarcoma, femur, MRI

Introduction

Parosteal osteosarcoma is a low-grade neoplasm which represents the most common type of surface osteosarcoma. Between 10 and 20% of all parosteal osteosarcomas undergo "dedifferentiation" to a higher grade [1, 2]. Often, the dedifferentiated areas are recognizable on radiographic studies as lucent regions within an otherwise radiodense lesion. Most commonly, the high-grade component has the histologic appearance of malignant fibrous histiocytoma, fibrosarcoma, or osteosarcoma. Herein we report a case of a dedifferentiated parosteal osteosarcoma of the distal femur, in which the high-grade component was composed entirely of rhabdomyosarcoma. To our knowledge, a rhabdomyosarcomatous component in a dedifferentiated parosteal osteosarcoma has not been described previously.

Case report

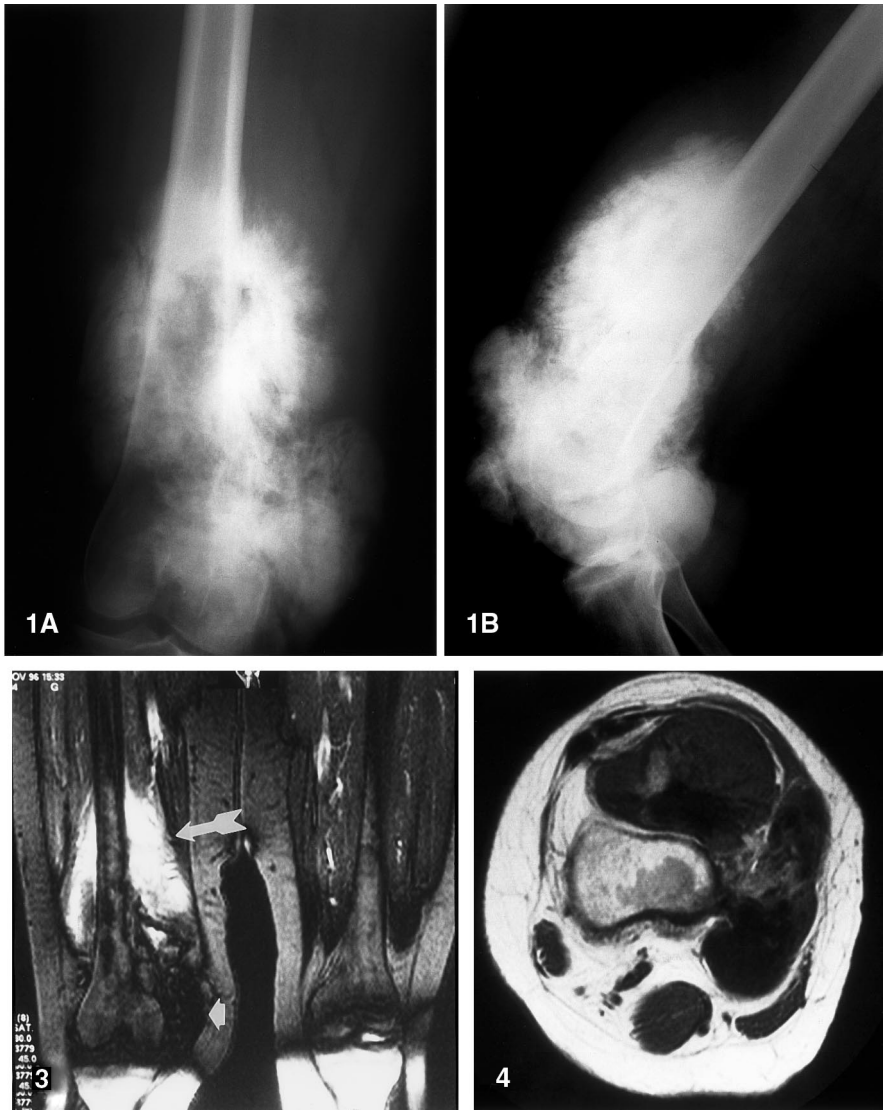
A 36-year-old pregnant woman presented with an 8-month history of increasing pain and swelling in the right knee. Physical examination revealed a palpable mass in the popliteal region associated with edema and warmth, and markedly decreased range of motion of the right knee. Radiographs showed a large, predominantly ossified mass arising from the surface of the distal femur (Fig. 1); areas of radiolucency were

Fig. 1 **A** Posteroanterior (PA) and **B** lateral radiographs of the knee obtained at the time of initial presentation demonstrate a large, vaguely lobulated, densely mineralized tumor encompassing the distal femur

Fig. 2 Axial CT scan demonstrates a lobulated, ossified lesion containing radiolucent foci arising from the surface of the femur

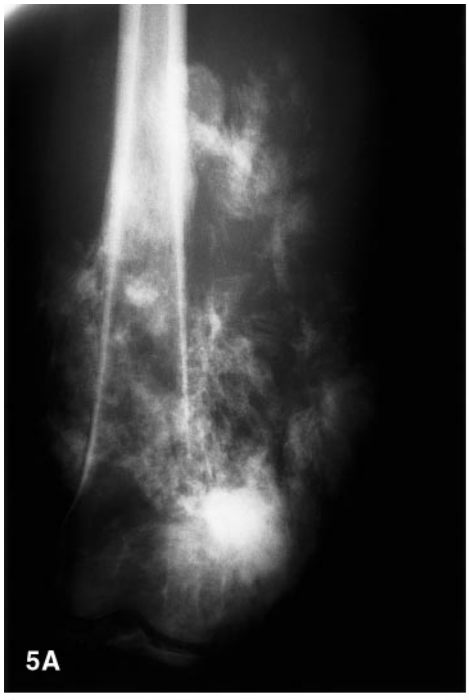
Fig. 3 Coronal plane T2-weighted fast spin echo/fat suppression MR images (TE=80, TR=3379) demonstrate the "biphasic" nature of the tumor with rounded, ossified areas distally (*arrowhead*) as well as areas of high signal intensity proximally (*long arrow*). Also note the mixed signal in the medullary canal, indicating involvement of this region as well

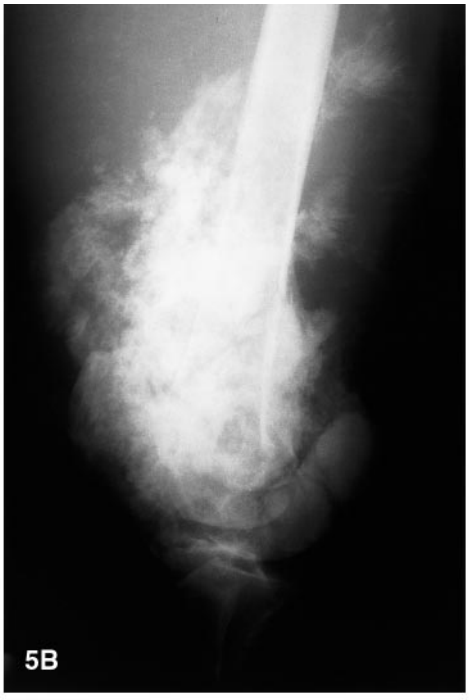
Fig. 4 Axial T1-weighted MR images (TE=14, TR=775) highlight the rounded, ossified low-grade areas as well as permeation of the medullary cavity by high-grade tumor



noted on CT scans (Fig. 2). MRI scans showed a lesion of mixed high and low signal intensity, with areas of reduced signal intensity corresponding to the mineralized regions seen on plain radiographs. MRI scans also highlighted areas of medullary cavity invasion (Figs. 3, 4). These radiographic findings were felt to represent a variant of surface osteosarcoma with secondary involvement of the medullary cavity. A CT scan of the chest showed no lung metastases. A CT-guided biopsy was interpreted as "osteosarcoma," and consisted entirely of fragments of low-grade, parosteal osteosarcoma.

The patient was non-compliant with her treatment plan and did not return for approximately 9 months following biopsy. At this time, additional imaging studies were obtained which showed interval progression of the tumor, including a marked increase in size and prominent medullary cavity invasion (Figs. 5, 6). CT scans of the chest revealed multiple pulmonary metastases bilaterally. Neoadjuvant chemotherapy was given, followed by a right hip disarticulation; limb salvage was not possible because of the size of the lesion and its proximity to neurovascular structures. Grossly, the tumor contained multiple dense bony nodules which partially encompassed the distal femur (Fig. 7). Proximally, a large, fleshy component with prominent cystic necrosis was identified, which invaded into the medullary cavity. The entire tumor measured approximately 28.0 cm in greatest dimension. Histologically, the tumor contained two distinct components. The distal-most aspect of the tumor was composed of interlacing trabeculae of woven and lamellar bone with intervening fibrous stroma (Fig. 8); no significant cytologic atypia or mitotic activity was noted in either the bone trabeculae or fibrous stroma.





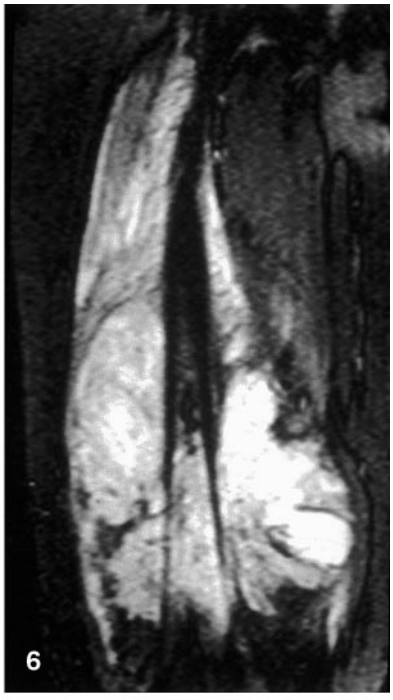
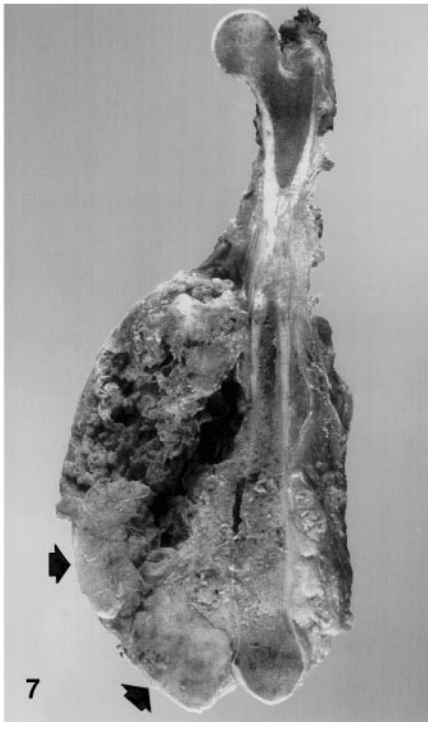


Fig. 5 PA (**A**) and lateral (**B**) plain radiographs obtained 9 months after presentation show interval progression of the high-grade areas, with more areas of radiolucency than seen previously

Fig. 6 Coronal inversion recovery MR images (TR=1900, TE=30, TI=125) show areas of ossified tumor, marked progression of the high-grade component (high signal intensity areas proximally), and involvement of the medullary cavity. The most proximal areas of high signal intensity represent reactive changes and edema (digital image)

Fig. 7 Gross photograph of the tumor, which consisted of large, oval, ossified nodules (*arrowheads*) and a large, fleshy, centrally necrotic mass



The histology of this component was typical of a parosteal osteosarcoma. The second component consisted of small round cells which, in areas, exhibited fibrillary pink cytoplasm, associated with abundant mitotic activity and extensive necrosis (Fig. 8). The cells from the high-grade component were strongly immunoreactive for desmin and myogenin (Fig. 9), and negative for keratin, S-100 protein, and CD99. The high-grade component extensively invaded the medullary cavity and permeated cancellous bone. A diagnosis of dedifferentiated parosteal osteosarcoma with a rhabdomyosarcomatous component was made. Postoperative chemotherapy was also given, but the patient died with widespread metastatic disease 9 months after hip disarticulation.

Discussion

Parosteal osteosarcomas represent approximately 5% of all osteosarcomas and are the most common type of surface osteosarcoma. The peak age incidence for parosteal osteosarcoma is in the third and fourth decades, and men and women are affected equally. The most common locations include the distal femur, proximal tibia, and proximal humerus. Patients typically present with localized swelling or a mass of long duration with or without pain. In the typical case, parosteal osteosarcomas are low grade and are composed of woven or lamellar bone embedded in a relatively bland fibrous stroma without obvious cytologic atypia or abundant mitotic activity. Dedifferentiation of low-grade osteosarcomas is a well-known phenomenon. Dwinnell et al. [3] and Unni et al. [4] described “transformation” of several parosteal osteosarcomas into higher-grade osteosarcomas. Similar cases were described by Dunham et

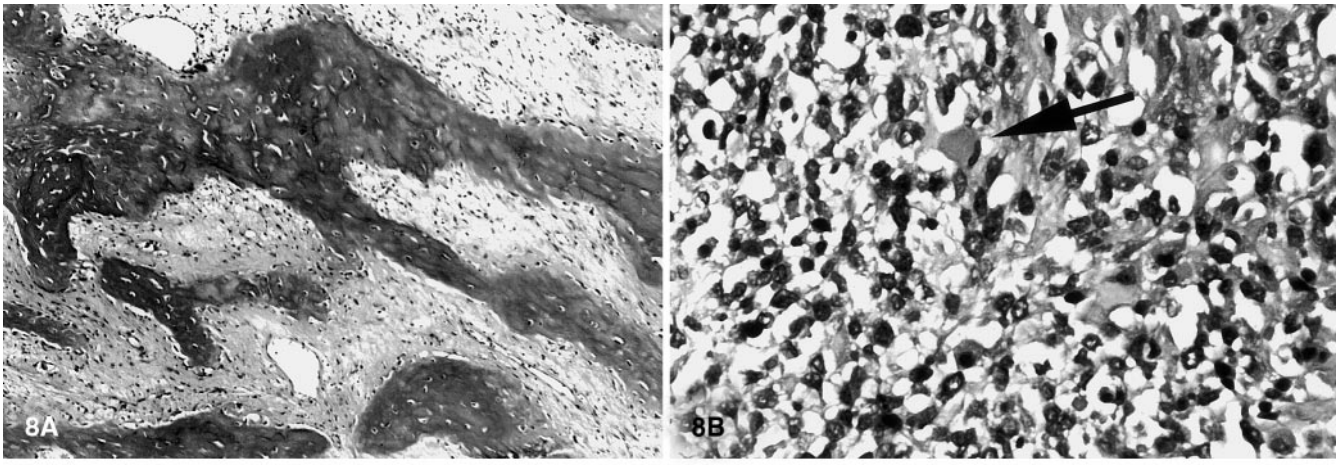


Fig. 8 Histologically, the tumor was composed of **A** areas typical of parosteal osteosarcoma, as well as **B** a high-grade component, which proved to be rhabdomyosarcoma (*arrow* indicates a rhabdomyoblast)

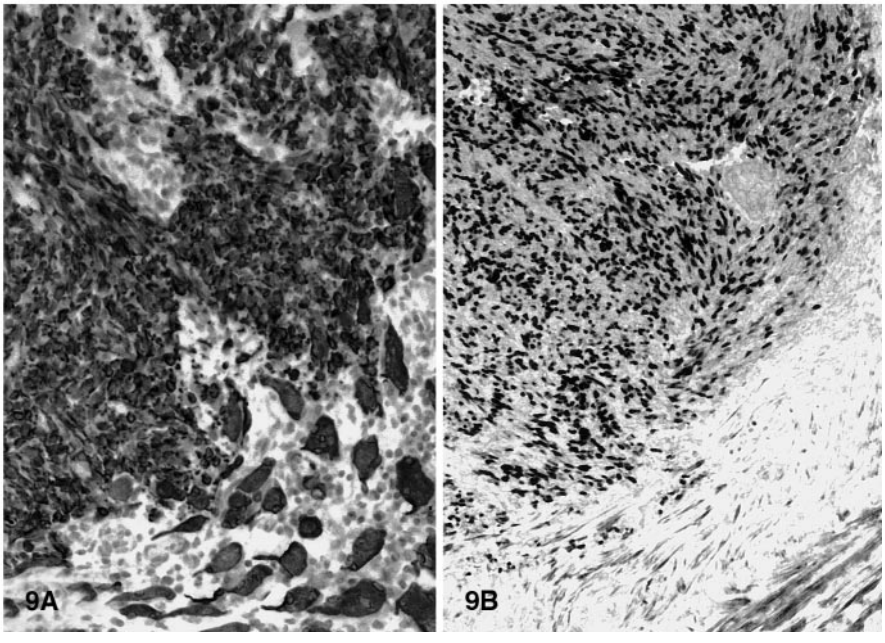


Fig. 9 Immunohistochemical stains for **A** desmin and **B** myogenin were strongly positive, confirming that the high-grade component was rhabdomyosarcoma

al. [5] and Pintado et al. [6]. A 1994 review of the Mayo Clinic experience with parosteal osteosarcomas by Okada et al. [1] revealed 37 cases of dedifferentiated parosteal osteosarcoma, representing 16% of all parosteal osteosarcomas seen there.

Parosteal osteosarcomas have a relatively typical radiographic appearance. Imaging studies demonstrate a lobulated, radiodense mass most commonly arising in the metaphyseal region of long bones, often the distal femur, proximal tibia, or proximal humerus. Less commonly, parosteal osteosarcomas arise from the diaphysis of a long bone. Occasionally, parosteal osteosarcomas invade the underlying medullary cavity, but are often separated from the underlying cortex by a thin lucent region. When higher-grade areas are present, they typically manifest radiographically as lucent regions which may destroy the mineralized, low-grade component or invade the medullary cavity [7, 8], as in the case presented here.

The unusual feature of the case presented herein is the histologic appearance of the high-grade component of the tumor. In most cases of dedifferentiated parosteal osteosarcoma, the morphology of the high-grade component is that of malignant fibrous histiocytoma, fibrosarcoma, or osteosarcoma [2, 5, 6, 9]. In the case presented here, the high-grade component was rhabdomyosarcoma, confirmed with positive desmin and myogenin immunostains. To our knowledge, this is the first reported case of a dedifferentiated parosteal osteosarcoma with rhabdomyosarcomatous differentiation. Other primary low-grade sarcomas have been known to dedifferentiate into rhabdomyosarcoma, including chondrosarcoma [10] and liposarcoma [11].

The presence of a high-grade, dedifferentiated component is associated with a more aggressive growth pattern and permeation of the underlying cortex and medullary cavity; medullary cavity involvement is more commonly seen in parosteal osteosarcomas with a dedifferentiated component. Metastases are also significantly more common when dedifferentiation occurs [1, 9]; approximately 50% of patients with dedifferentiated parosteal osteosarcomas develop distant metastases. The patient described here presented with

Musculoskeletal Tumor Society stage IIB disease, but developed pulmonary metastases in the interim between biopsy and definitive therapy. The prognosis of dedifferentiated parosteal osteosarcoma is similar to the prognosis for conventional intramedullary osteosarcoma.

Dedifferentiated areas are more frequently encountered in local recurrences than in de novo lesions. As in the case reported by Shuhaibar and Friedman [12], the patient described here likely had dedifferentiated areas at presentation based on the radiographic findings. However, the high-grade areas were not present in the initial biopsy material. The relatively poor prognosis of dedifferentiated parosteal osteosarcoma does not appear to change if the high-grade areas are identified at presentation or develop in a subsequent local recurrence.

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