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CASE REPORT

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Telangiectatic dedifferentiation of a parosteal osteosarcoma

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Abstract A unique case of parosteal osteosarcoma (POS) of the proximal femur, with areas of telangiectatic dedifferentiation, in a 28-year-old woman is reported. The patient had a 7-week history of pain and swelling in her right thigh. A biopsy diagnosis of POS was established. The patient was treated with two cycles of intra-arterial chemotherapy, followed by limb salvage surgery. Histological examination of the resected specimen showed POS with areas of dedifferentiation composed of high-grade telangiectatic osteosarcoma with associated secondary aneurysmal bone cyst change.

Keywords Parosteal osteosarcoma · Telangiectatic osteosarcoma · Dedifferentiation · Femur · Radiography · MRI

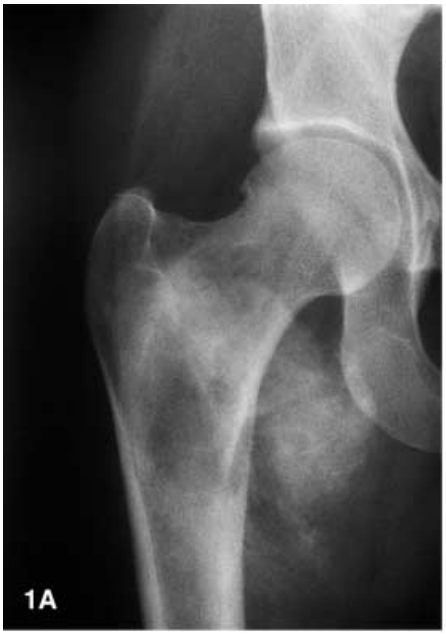
Introduction

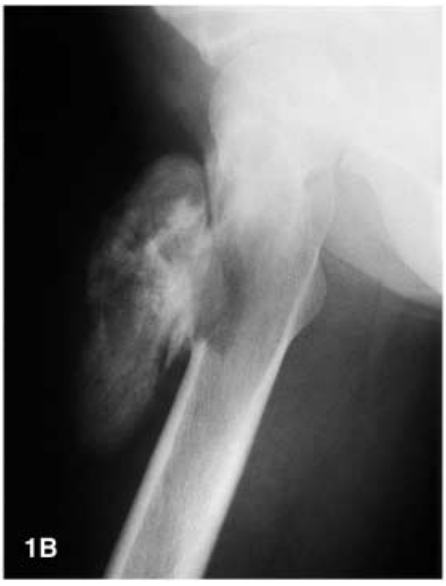
Parosteal osteosarcoma (POS) is an uncommon, predominantly fibro-osseous variant of osteosarcoma, arising almost exclusively from the juxtacortical areas of the long bones. It typically is a well-differentiated lesion that has a significantly better prognosis than conventional osteosarcoma. In contrast, telangiectatic osteosarcoma is an aggressive variant of osteosarcoma that carries approximately the same prognosis as conventional osteosarcoma. Infrequently, POS undergoes dedifferentiation, and in doing so the chances of metastatic disease increase significantly. Dedifferentiation is the process by which one type of tissue transforms into another, often more primitive, type of tissue. Secondary aneurysmal bone cysts are associated with a wide range of intraosseous neoplasms, including POS and telangiectatic osteosarcoma.

We report, for the first time in the world literature, the unique occurrence of a POS that contained areas of telangiectatic dedifferentiation, as well as areas of aneurysmal bone cyst.

Case report

A 28-year-old woman presented in April 1997 with a 7-week history of discomfort and swelling in the anterior aspect of her right thigh. Examination showed a large, hard, non-mobile mass at the level of the greater trochanter. Radiographs showed a large ossified mass attached to the proximal femur, with destruction of the adjacent bony cortex (Fig. 1 A, B). A computed tomographic (CT) scan demonstrated a bony, exophytic lesion, measuring 6×4×10 cm arising, by a broad base, from the an-





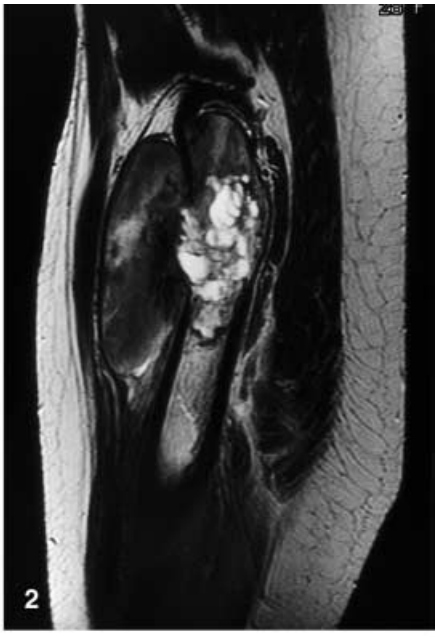
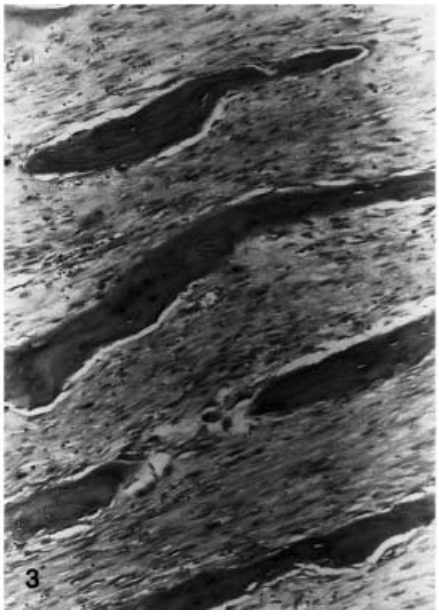


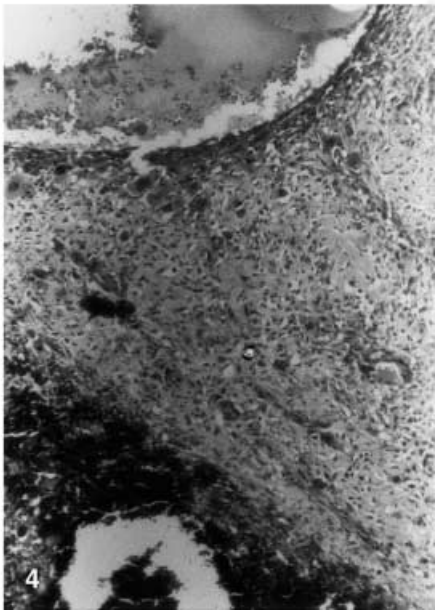
Fig. 1 **A** Anteroposterior and **B** lateral radiographs of a bony, exophytic lesion measuring 6×4×10 cm arising from the anterolateral femoral cortex at the intertrochanteric level of the right femur, with some destruction of the femoral cortex

Fig. 2 Sagittal T2-weighted MRI scan demonstrating a tumor of the proximal femur with intra- and extra-osseous parts. The extra-osseous component is a 5×5×10 cm well-defined, enhancing mass arising from the anterior femoral cortex. The intra-osseous component includes solid and cystic areas, with loculations and fluid levels

Fig. 3 Tissue from en bloc resection of the tumor showing mature but irregular bone trabeculae within a fibrous spindle cell stroma with cellular pleomorphism and mitotic figures consistent with parosteal osteosarcoma

Fig. 4 Parosteal osteosarcoma with areas of secondary aneurysmal bone cyst change





terolateral femoral cortex, at the level of the intertrochanteric region (not shown). A magnetic resonance imaging (MRI) scan showed a 5×5×10 cm well-defined, enhancing, bony mass in the anterior soft tissues of the thigh, arising from the anterior cortex of the femur (Fig. 2). The mass extended anteriorly, invading the vastus intermedius muscle, though the neurovascular bundle was not thought to be involved. The MRI scan also showed that the lesion extended into the intramedullary part of the proximal femur. The majority of the intramedullary portion of the lesion was cystic, and contained fluid levels. Chest radiographs and CT scans showed no evidence of metastatic disease.

Tissue from a Trucut needle biopsy showed a spindle cell, malignant tumor with moderate nuclear pleomorphism and occasional mitotic figures with embedded trabeculae of mixed woven and lamellar bone arranged randomly within the stroma. In one area, the tumor extended into adipose tissue adjacent to muscle. The specimen included islands of mature bone, most of which were randomly aligned. No areas of dedifferentiation or high-grade malignancy were seen. On the basis of the histological and radiological findings, a diagnosis of POS was made. The patient underwent two cycles of intra-arterial chemotherapy, consisting of cisplatin and adriamycin. A post-

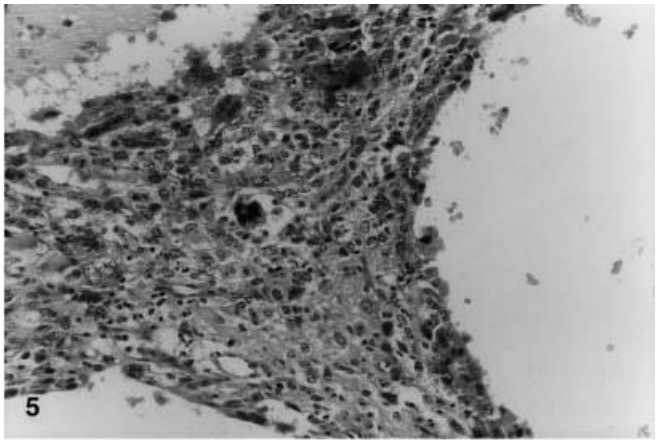
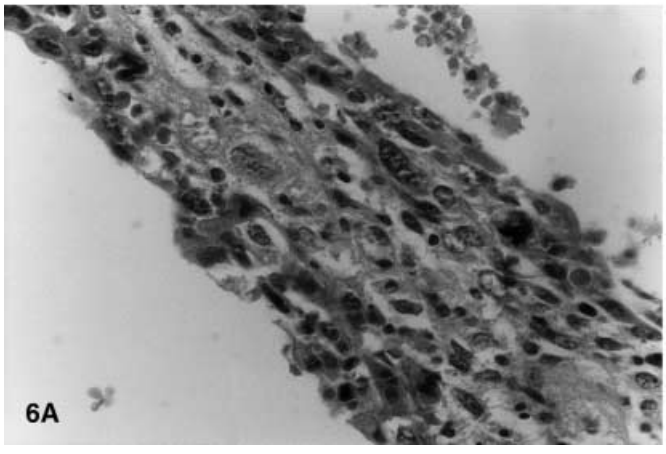


Fig. 5 Blood-filled spaces surrounded by septa including enlarged pleomorphic cells with atypical nuclei and abnormal mitoses, consistent with telangiectatic dedifferentiation



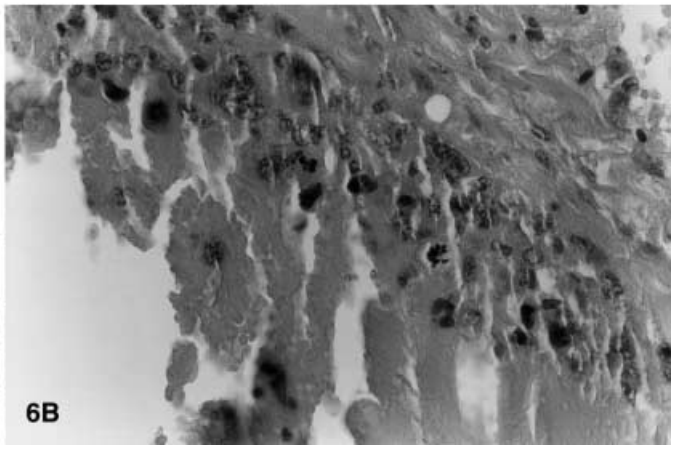


Fig. 6A, B Markedly atypical cells lining the blood-filled spaces, consistent with telangiectatic dedifferentiation

chemotherapy MRI scan did not show any appreciable changes in the size or appearance of the tumor.

The tumor was resected en bloc, in June 1997, along with 17 cm of proximal femur, most of the vastus intermedius and lateralis muscles and the femoral nerve. An MS 30 acetabular cup was cemented in position and the proximal femur replaced with a Kotz prosthesis.

The entire specimen measured 19×8.5×6 cm, made up of an extraosseous component (10×7.5×4.5 cm), a hard intramedullary component (3.5×2×2 cm) and a cystic intramedullary component (7×4×4 cm). Histologically, the majority of the tumor was made up of mature but irregular bone trabeculae within a fibrous spindle cell stroma, whose cells showed some pleomorphism and mitotic figures. A diagnosis of POS was made (Fig. 3). The tumor was considered to be a parosteal lesion rather than a well-differentiated osteosarcoma with extra-osseous extension because the majority of the solid tumor was outside of bone, and had a bulbous appearance projecting from the surface of the bone. This is in keeping with the findings of Campanacci et al. [1], who use the term POS for lesions projecting from the periosseous tissues with a mushroom-like radiographic appearance even when the radiographic and gross appearance suggest intramedullary involvement.

Most of the cystic areas in the lesion showed blood-filled spaces typical of aneurysmal bone cyst, with giant cells, bland stromal cells and the occasional foci of osteoid in their walls (Fig. 4). However, the wall of one cyst contained pleomorphic stromal cells with atypical nuclei, associated with osteoid production. A diagnosis of localized telangiectatic osteosarcomatous dedifferentiation was made (Figs. 5, 6A, B). The surgical margins were free of tumor.

The patient remained symptom-free for 12 months following surgery. However, in May 1998, chest CT scans showed two metastatic lesions in the right lung, and in the following month a mass was found in the patient's left gluteal region. Despite treatment with radiotherapy and adriamycin, the patient died 6 months later, approximately 18 months after the onset of her symptoms. These lesions were not biopsied, and an autopsy was not performed.

Discussion

Parosteal osteosarcoma is a well-differentiated, fibro-osseous variant of osteosarcoma arising from the juxtacortical region of the long bones [2], first described as a distinct

clinical entity by Geschickter and Copeland [3] in 1951. It is thought to comprise about 4% of all osteosarcomas [4] and most commonly occurs in the second and third decades of life [1]. Most cases tend to involve the postero-distal femoral metaphysis [5]. It most frequently presents as a painless swelling present for many months, but may also cause dull, aching pain or joint dysfunction [5]. The classic radiological appearance is that of a densely mineralized, amorphous mass attached to the cortical surface by a broad base [2]. Macroscopically, POS is a hard, white, lobulated mass [4] that rests upon the surface of bone. Microscopically, the tumor consists of regularly arranged osseous trabeculae or ill-defined islands of osteoid and woven bone separated by fibrous stroma. Cellular atypia is often sparse, and mitotic figures are rare. Cartilage may be present either within the lesion or as a cap. Although prone to local recurrence if incompletely excised, POS has a significantly better prognosis than conventional osteosarcoma. Sheth et al. [6] reported that 14 of their 15 patients (93%) with localized conventional POS were alive at a median survival of 77 months, with the one fatality due to unrelated causes. Okada et al. [7] evaluated survival for 39 patients treated for POS at the Mayo Clinic, and found a 5-year survival rate of 91%, with a 10-year survival of 80%.

POS may have intramedullary extension. Campanacci et al. [1] reported that 33% of grade I POS, 65% of grade II POS and 90% of grade III POS had medullary involvement. The prognostic relevance of medullary involvement in POS is controversial. Campanacci et al. [1] have not seen metastasis without involvement of the medullary cavity, and therefore believe that tumors with medullary involvement are inherently more aggressive. Others authors, for example Sheth et al. [6], have not corroborated this.

Telangiectatic osteosarcoma (TOS), which was described by Paget in 1854 [8], is a significantly different type of tumor from POS. It is usually an intra-osseous tumor and histologically consists of aneurysmally dilated blood spaces lined, or traversed, by highly malignant stromal cells that produce small amounts of osteoid [9]. There is often destruction of cortical bone, with soft tissue extension. Initially, the prognosis for TOS was thought to be poor, with the Mayo Clinic series reporting 23 deaths among their 25 cases [10]. However, this poor outcome was not substantiated by Huvos et al. [9], based on their experience at Memorial Hospital. The Mayo Clinic group later revised their view, based on stricter radiological criteria for the diagnosis, such that it is now recognized that TOS has a similar prognosis to conventional osteosarcoma [11].

On occasion, conventional POS may undergo transformation into a dedifferentiated tumor, exhibiting features of more poorly differentiated, high-grade sarcomata. In 1976, Unni et al. [12] described seven cases of POS in which there were coexistent areas of high-grade malignancy. Sheth and co-workers [6] reported a 43% dedifferentiation rate in a series of 28 POS, and Okada et al. [7], had 37 cases of dedifferentiation among 226 cases of POS (16%). Sheth et al. [6], Raymond [13] and Mirra [5] report cases of dedifferentiated POS containing high-grade elements of osteosarcoma, fibrosarcoma, chondrosarcoma and malignant fibrous histiocytoma. Dedifferentiation may be present initially, as in our case, or as a secondary event after local recurrence [13]. Once a POS undergoes dedifferentiation, the patient's prognosis is significantly worse. Of the 11 patients who died of POS in Okada et al.'s study, 10 had a dedifferentiated tumor [7].

This case is of significance because of the nature of the dedifferentiated component. A MEDLINE (OVID) search of the world medical literature since 1966 did not reveal any other reports of telangiectatic dedifferentiation of a POS. This case demonstrates the dedifferentiation of the relatively quiescent cells of a POS into an area of aggressive telangiectatic osteosarcoma.

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