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Cortical Bone Remodeling in Parosteal Osteosarcoma Mimicking Medullary Involvement: A Case with the Difficulty in Pre-Surgical Staging

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Parosteal osteosarcoma (POS) is a low-grade well-differentiated variant of osteosarcoma that affects the metaphyseal surface of a long bone. Although Grade-1 POS sometimes involve the medullary canal, such patients are not at a greater risk of local recurrence or metastases. In this report, we describe a rare case of POS in the right distal femur with an intramedullary sclerotic lesion mimicking medullary involvement caused by secondary remodeling of the underlying cortex of the tumor. A 34-year-old woman complained of having a painful hard mass in her right knee for six months. Imaging studies revealed a broad-based sclerotic mass attached to the cortex of the distal and lateral aspect of the femur, along with an intramedullary lesion. Histopathological examination of a biopsy specimen revealed Grade-1 POS. We diagnosed a medullary involvement and we performed a wide resection, including the intramedullary lesion. Histopathological examination of the resected specimen revealed that the intramedullary lesion only exhibited remodeling of the underlying tumor cortex without tumor cell invasion. To the best of our knowledge, this is the first report of such imaging features and pathological findings in a patient with POS. Our experience with the present patient indicates that good local control and overall prognosis of patients with medullary involvement in Grade-1 POS may be due to the remodeling of the underlying cortex mimicking "medullary involvement." This feature will add to the range of diagnostic difficulty experienced during the preoperative staging of POS.

Keywords: low-grade malignancy; medullary involvement; parosteal osteosarcoma; radiological study; surgical stage
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Introduction

Parosteal osteosarcoma (POS) is a low-grade, malignant osseous tumor that usually develops on the metaphyseal surface of long bones and accounts for 4%-6% of all osteosarcomas (Okada et al. 1994). Unlike conventional osteosarcoma, POS typically occurs in the 3rd and 4th decades of life and shows a slightly higher prevalence among females. The most common site of involvement is the posterior aspect of the distal femur, accounting for approximately two-thirds of all cases (Stevens et al. 1957; Smith et al. 1978; Campanacci et al. 1984). POS is a well-differentiated, low-grade variant of osteosarcoma (Grade-1 or -2) (Unni and Dahlin 1984; Okada et al. 1994); however, dedifferentiation of low-grade POS to high-grade disease reportedly occurs in approximately 15% cases (Okada et al. 1994; Azura et al. 2009). In addition, the rate of medullary involvement is reportedly 18% in Grade-1 lesions, 25% in Grade-2 lesions, and 43% in dedifferentiated lesions (Okada et al. 1994). Here we describe, to the best of our knowledge, the first case of POS exhibiting an intramedullary sclerotic lesion mimicking medullary involvement, which was eventually revealed as an ossifying reactive change caused by secondary remodeling of the underlying cortex.

Clinical Report

A 34-year-old woman presented with a moderately painful, hard mass located on the lateral aspect of the right knee that had gradually increased in size over 6 months. She could not recall any incidence of trauma associated with her present condition, and her family history was unremarkable. A radiograph of her right knee revealed a broad-based sclerotic mass on the lateral aspect of the distal femur (Fig. 1A and B). Computed tomography (CT) revealed a two-layer-sclerotic lesion composed of a coarse sclerotic lesion on the cortical surface overlaid by a homogeneous

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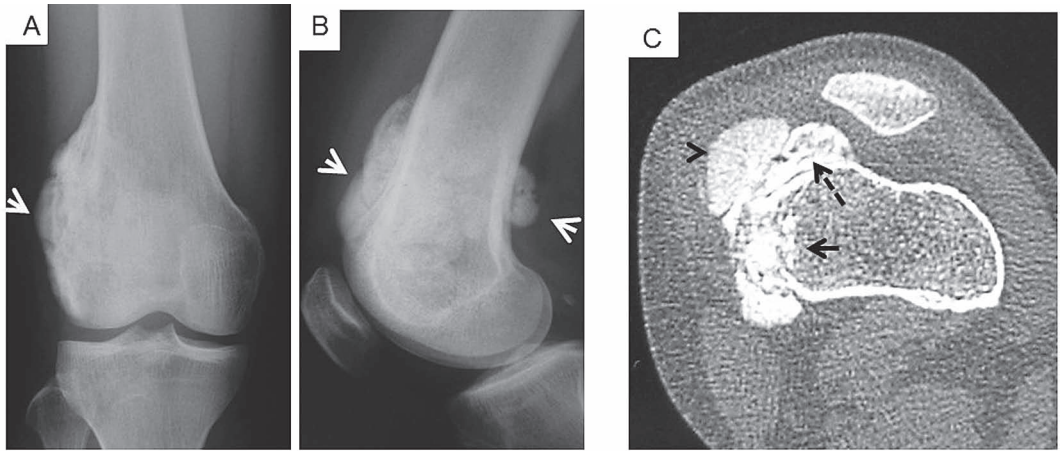

Fig. 1. Plain radiographs and computed tomography scan.
Anteroposterior (A) and lateral (B) plain radiographs demonstrate a dense and sclerotic mass on the lateral, anterior, and posterior surfaces of the right distal femur (white arrows). The computed tomography scan (C) reveals a dense and sclerotic mass (arrowhead), a coarse sclerotic lesion on the surface of the cortex (dotted arrow), and an intramedullary lesion (black arrow).

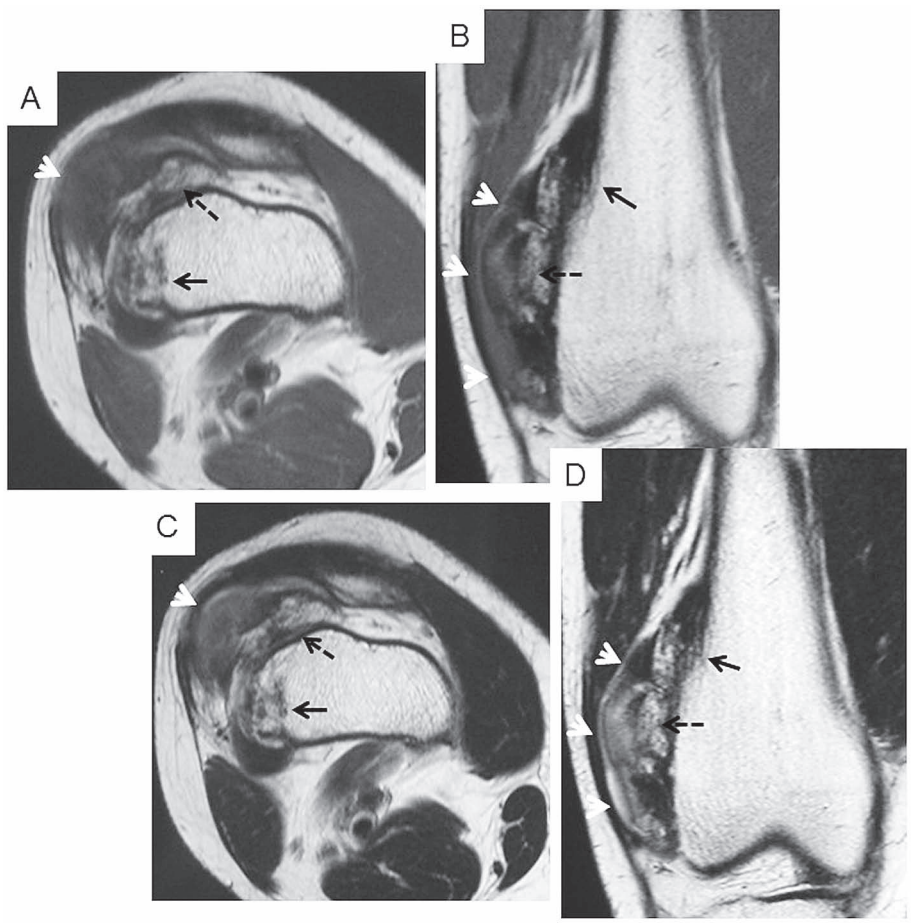

Fig. 2. Magnetic resonance images.
Axial (A) and coronal (B) T1-weighted images show a mass (white arrows) that appears with hypointense and isointense, and a lesion showing low to mixed hypointensity and hyperintensity on the cortical surface (dotted arrows). Axial (C) and coronal (D) T2-weighted images show the mass (white arrows) with low to slightly higher-signal intensity than that of muscle, and the lesion on the cortical surface (dotted arrows) showing low to mixed hypointensity and hyperintensity as well as T1-weighted images with an intramedullary lesion (black arrow) exhibiting a linear hypointensity on both T1-weighted and T2-weighted images.

lesion with dense sclerosis. An additional intramedullary sclerotic lesion was also identified (Fig. 1C). On both axial and coronal T1-weighted and T2-weighted magnetic resonance imaging (MRI), the coarse sclerotic lesion on the cortical surface showed low to high signal intensity suggesting that it contained fatty marrow. The dense sclerotic lesion overlying the mass was isointense, similar to muscle tissue on T1-weighted images, and showed slightly hyperintensity on T2-weighted images. The medullary lesion showed a linear hypointensity on both T1-weighted and T2-weighted images (Fig. 2A-D). A bone scan and chest CT scan confirmed that the tumor was solitary, with no pulmonary metastasis.

A biopsy specimen from the dense sclerotic lesion revealed densely fibrotic, osteosclerotic growth. The tumor cells exhibited mild nuclear atypia and pleomorphism. In contrast, the deeper, coarse sclerotic lesion on the cortex only showed a thickening of the trabeculae and fatty marrow, and there were no obvious tumor cells. Clinicopathologically, a preoperative diagnosis of Grade-1 POS with medullary involvement (surgical stage IB) was made.

Thus, we performed wide resection of both the cortical and medullary lesions followed by reconstruction with an allogenic bone graft and plate fixation.

A coronal slice of the gross specimen revealed an outer dense sclerotic lesion with peripheral cartilage and coarse, sclerotic lesion on the cortical surface contiguous with the medullary lesion. No portion of the cortex at the level of the lesion was normal (Fig. 3).

Histopathological examination of the outer lesion specimen revealed a primarily fibrous matrix with mixed lamellar and woven bone trabeculae, mimicking fibrous dysplasia, with the tumor cells displaying mild nuclear atypia. Mitotic figures were rare, and no necrosis was observed (Fig. 4A and B). Therefore, we made a histopathological diagnosis of Grade-1 POS. On the other hand, the inner coarse sclerotic lesion demonstrated continuity with the intramedullary sclerotic lesion, and both showed the same histological findings of thickened trabeculae and fatty marrow (Fig. 4C). There were no tumor cells or fibrous tissue in the medullary area in any section. On the basis of the above mentioned findings, we concluded that this condition may have resulted from secondary remodeling of the cortical bone adjacent to the tumor rather than a medullary involvement. Therefore, a final diagnosis of surgical stage IA (Grade-1 POS without medullary involvement) was made. Five years after surgery, the patient remains disease free.

Discussion

POS is a low-grade, malignant osseous tumor that usually develops on the metaphyseal surface of long bones and accounts for approximately 3%-5% of all osteosarcomas (Okada et al. 1994). Radiologically, conventional POS appears as a dense, lobulated mass with a broad-based attachment to the cortical bone. In the early stages, the

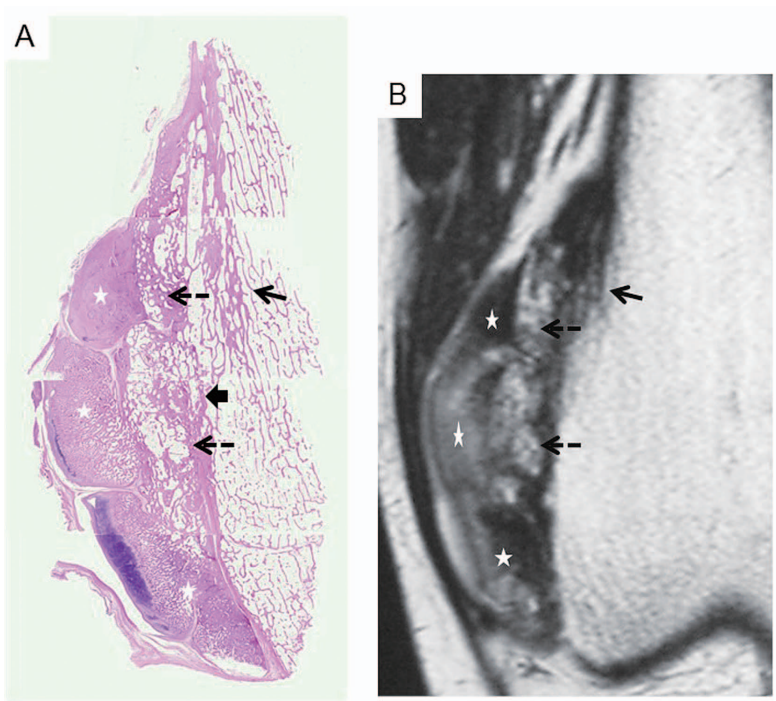

Fig. 3.
A photograph of the gross specimen (A) corresponding to a coronal T2-weighted image (B).
The gross coronal specimen consists of three parts: a lateral dense tumor mass corresponding to the potion of POS (asterisks), a coarse sclerotic lesion on the surface of the cortex (dotted arrows), and an intramedullary lesion (black arrow). The latter two lesions are contiguous. Cortical irregularities and disruption can be seen (short arrow).

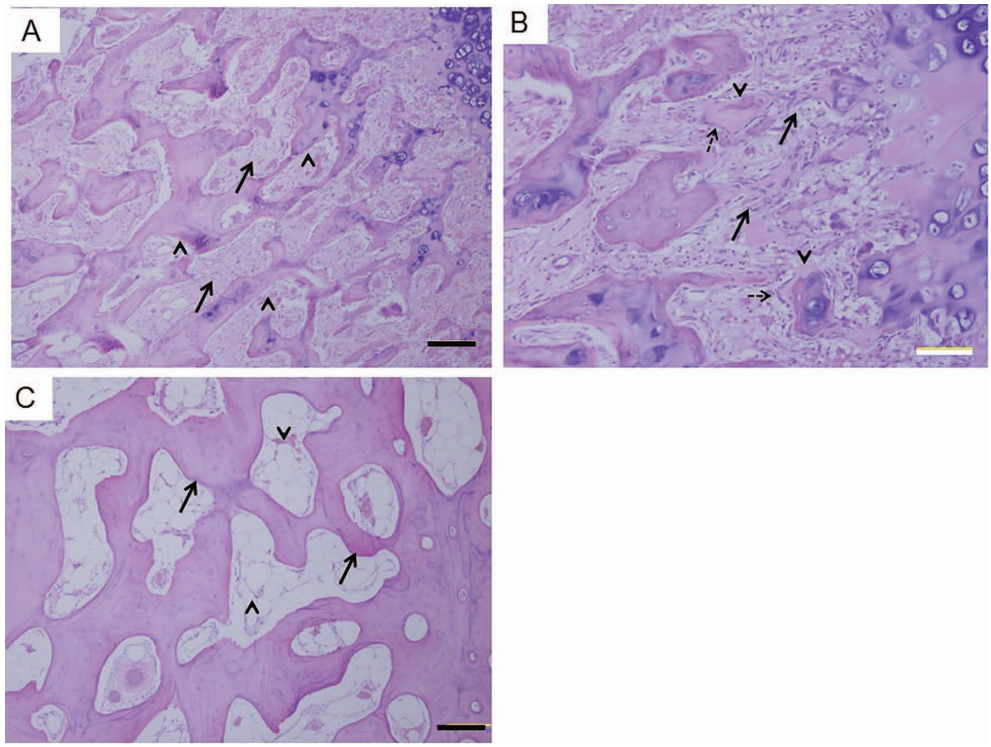

Fig. 4. Photomicrographs of the histologic sections.
(A) Low-power photomicrographs of the areas indicated by asterisks in Fig. 3 showing typical histopathological appearance of Grade-1 POS. The tumor contains well-differentiated osseous trabeculae (arrow heads) and spindle cellular stroma (arrows) (black bar, 200 μm; hematoxylin and eosin staining).
(B) A high-power photomicrograph showing proliferation of spindle cells (arrows) and woven bone (arrow heads) covered by benign osteoblastic rimming (dotted arrows) between the bone trabeculae (white bar, 100 μm; hematoxylin and eosin staining).
(C) Low-power photomicrographs in the coarse sclerotic lesion (dotted arrows) and intramedullary lesion (black arrow) shown in Fig. 3 with the same histological findings. Only thickened, irregular trabeculae (arrows) and fatty marrow (arrowheads) were observed, but no cellular stroma (black bar, 200 μm; hematoxylin and eosin staining).

tumor may only exhibit a small sessile attachment to the cortical surface (Stevens et al. 1957). Apart from the characteristic attachment of POS, the presence of a linear radiolucent zone is helpful for its diagnosis; however, in advanced stages, this radiolucent line may be absent (Lindell et al. 1987; Jelinek et al. 1996; Suresh and Saifuddin 2007). Okada and colleagues (1994) reported that 185 (82%) of 226 POS patients had a histological Grade-1 lesion, while 41 (18%) had a Grade-2 lesion. Thirty-seven patients (16%) were considered to have undergone dedifferentiation. The rate of medullary involvement was 18% for Grade-1 lesions, 25% for Grade-2 lesions, and 43% for dedifferentiated lesions. Campanacci and colleagues (1984), on the other hand, reported that 33%, 65%, and 90% of Grade-1, Grade-2, and Grade-3 lesions respectively exhibited medullary involvement. Although the prognostic significance of medullary involvement remains unclear (Ahuja et al. 1977; Picci et al. 1987), some authors have interpreted the presence of a tumor within the marrow cavity to be indicative of a more aggressive lesion that is invariably associated with pulmonary metastasis and a relatively poorer prognosis (Enneking et al. 1980; Campanacci et al. 1984). However, other authors have reported no similar prognostic implications of this feature (Ahuja et al. 1977; Kavanagh et al. 1990). Another study has reported that if the tumor involves the medullary canal, it usually does not involve more than 25% of the canal's diameter (Okada et al. 1994).

Okada and colleagues (1994) reported that in larger, broad-based tumors, deformity of the underlying bone was occasionally present secondary to remodeling, and that the cortex was expanded in 19% patients. Other conditions adjacent to the bone, such as periosteal chondroma (deSantos and Spjut 1981), periosteal ganglion (McCarthy et al. 1983), or soft tissue hemangioma (Murphey et al. 1995; Yuh et al. 1987; Ly et al. 2003), may induce osseous changes without direct invasion. Notably, these benign conditions often cause cortical deformity, erosion, thickening, destruction, and reactive new periosteal bone formation. Although the exact mechanisms leading to these changes remain unknown, several theories have been proposed. These include the effects of pressure caused by a mass lying in direct contact with bone, which may result in reactive new bone formation (Ly et al. 2003), or the presence of a mass that can result in the stretching or irritation of the adjacent periosteum (Yuh et al. 1987), which can also

cause new bone formation. Although the mechanisms underlying reactive changes in osseous structures with medullary involvement in POS remain unclear, similar theories have been proposed.

A remarkable feature of the present case is the existence of Grade-1 POS case, demonstrating the unique feature of mimicking medullary involvement by secondary remodeling of the underlying cortex of the tumor. Although medullary involvement reportedly correlated with pulmonary metastases and is an important predictor of overall prognosis in Grade-2 and Grade-3 cases, pulmonary metastasis has not been reported in Grade-1 cases regardless of medullary involvement (Campanacci et al. 1984). Moreover, Okada and colleagues (1994) found no association between local recurrence and medullary involvement in stage IB POS cases and Lewis and colleagues (2000) concluded that focal medullary involvement did not necessitate the radical resection of an entire segment of the lesion; instead, they recommended resection with a wide surgical margin. Good local control and overall prognosis of patients with stage IB POS may be associated with existence of stage IA tumors mimicking as stage IB tumors, as observed in the present case. Therefore, we propose that both radiological and histopathological findings of cases similar the present case should be carefully evaluated in the future.

In conclusion, we reported a case of stage IA POS that mimicked stage IB disease because of secondary cortical bone remodeling. This case report adds to the spectrum of radiological appearances of POS.

Conflict of Interest

The authors declare no conflict of interest.

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