Case report 715

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Imaging studies

Fig. 1. A Anteroposterior and B lateral radiographs of the knee reveal a lytic lesion in the distal end of the femur. No bony expansion or periosteal reaction is present. The proximal extent of the lesion (arrow) is ill-defined.

Fig. 2. Axial T2-weighted magnetic resonance (MR) image (SE 2300/80). The lesion in the metaphysis shows a heterogeneous pattern of high signal intensity. No fluid-fluid levels are identifiable.

Fig. 3. Axial T2-weighted MR image (SE 2300/80). The diaphyseal portion of the lesion demonstrates central hypointensity (straight arrow), surrounded by a thin peripheral zone of low signal intensity (curved arrow). Increased signal intensity in the surrounding marrow space is present.

Fig. 4. Coronal T1-weighted MR image (SE 600/20). The lesion extends into the femoral shaft. A low signal interface is present at the distal extent of the lesion (open arrow). The margins of the diaphyseal component are poorly defined. An intermediate signal intensity in a peripheral band (small arrows) is separated from the central lesion by a higher signal intensity in an outer band.

Fig. 5. Coronal T1-weighted MR image (SE 600/20), after administration of gadolinium diethylene-triaminepentaacetic acid. The intermediate signal band surrounding the diaphyseal component of the lesion has enhanced and is now isointense with the surrounding marrow.
Clinical information

A 27-year-old man presented with a 4–6-week history of pain in the right knee which was aggravated by walking. A history of trauma was not obtained. He was otherwise healthy and denied fevers, night sweats, or weight loss. Routine laboratory examination results, including leukocyte count, peripheral blood smear, and erythrocyte sedimentation rate, were normal.

Radiographic examination revealed a central lytic lesion of the distal end of the femur that extended to the subarticular portion of the intercondylar notch (Fig. 1). The proximal margin of the lesion was poorly demarcated. No evidence of a mineralized matrix, cortical expansion, or periosteal reaction was present.

Magnetic resonance imaging (MRI) at 1.5 T was performed (Figs. 2–5), which revealed extension of the lesion into the distal one-third of the femoral shaft. The distal metaphyseal and epiphyseal portions of the lesion displayed inhomogeneous intermediate signal intensity on T1-weighted images and inhomogeneous high signal intensity on T2-weighted images. Cortical permeation was noted, but the distal lesional margins were well-defined and surrounded by a rim of low signal intensity. Proximally, the lesion showed distinct hypointensity in its central portion on T2-weighted images. Lesional margins in the diaphysis were irregular and ill-defined. A thin, separate, peripheral zone of tissue in the proximal extent of the lesion preferentially enhanced after administration of gadolinium diethylenetriaminepenta-acetic acid. The remainder of the lesion showed no significant enhancement.

An open biopsy was performed.
Diagnosis: Necrotic giant cell tumor of the femur

Despite the somewhat uncharacteristic central location of the lesion, the radiographic findings were most typical of giant cell tumor. The unusual diaphyseal extension of the tumor and the irregular margins of the diaphyseal component on MRI prompted consideration of other diagnoses. These would include osteogenic sarcoma, lymphoma, and desmoplastic fibroma.

Extensive intralesional curettage with bone grafting proximally and cementation distally was performed. The specimen consisted of tan and mustard colored fragments of tissue with focal areas of hemorrhage. The vast majority of sections showed only extensive necrosis with foci of chronic inflammation, hematoidin deposition, and hemosiderosis. Small foci, however, revealed the remnants of a preexisting lesion. These foci consisted of ‘ghosts’ of giant cells and stromal remnants of giant cell tumor (Figs. 6, 7). No regional differences existed in the specimens obtained from the proximal segment as compared with distal aspects of the lesion. Only one specimen (from the proximal aspect of the lesion) showed a single viable region consistent with giant cell tumor (Fig. 8).

Discussion

The MR findings of this lesion presented diagnostic difficulty in two ways. First, the extensive diaphyseal extension is unusual for giant cell tumor and raised the suspicion of a more aggressive and/or invasive tumor. Secondly, MRI demonstrated a zonal heterogeneity in the tissue sig-

Pathological studies

Fig. 6. Most of the abundant necrosis evident at histological examination consists of granular debris, but selected areas demonstrate the ‘ghosts’ of former osteoclast-like giant cells and stromal cells. Faint nuclear remnants are noted in some of the giant cell ghosts (H & E, ×400)

Fig. 7. Granulation tissue at the periphery of a necrotic zone exhibits abundant deposits of hemosiderin pigment and moderate numbers of lymphocytes (H & E, ×400)

Fig. 8. Residual, degenerating, but still viable giant-cell tumor is found in only two microscopic foci in a single specimen obtained from the proximal portion of the lesion (H & E, ×400)
nal characteristics of the epiphyseal/metaphyseal component as compared with the diaphyseal portion. The epiphyseal and metaphyseal components showed heterogeneous intermediate to high signal intensity on proton density and T2-weighted images. The diaphyseal component exhibited relative hypointensity for these sequences. The basis for these zonal differences is unclear, as similar histological sections were obtained on separate biopsies of these regions. The hypointensity of tumor in the diaphysis on T2-weighted images is uncharacteristic of giant cell tumor as reported by Herman et al. [2], who found that 6 giant cell tumors were largely iso- or hyperintense to marrow on T2-weighted images.

Giant cell tumors are typically well-defined lesions on MR imaging [2, 5]. The single, peripheral, low signal intensity halo around the distal epiphyseal aspect of this lesion has been previously noted [5] and has been interpreted as a feature of benignity when assessing bony lesions by MRI [6]. The margins of the diaphyseal portion of the lesion in this report were, however, quite different. The diaphyseal portion demonstrated irregular margins with an intermediate signal intensity band peripherally, parallel to a second inner zone of high signal intensity on T1-weighted images. The peripheral intermediate signal band in the diaphysis exhibited preferential enhancement after administration of gadolinium. Perhaps the irregular margins of the diaphyseal component of the lesion and the enhancing, peripheral zone represent a reactive interface of granulation tissue between normal marrow and tumor as a result of the extensive tumor necrosis. Because en bloc resection was not performed, it is impossible to ascertain the precise pathological correlation for the zonal margins and to determine whether the hypointense peripheral zone in the diaphyses was a reflection of hemosiderin deposition.

Few data exist on the enhancement characteristics of giant cell tumor. It is said to exhibit more prominent and more rapid enhancement on static and dynamic enhanced MRI examination than most other benign neoplasms of bone [1]. Hudson et al. [3] reported striking, inhomogeneous contrast enhancement of giant cell tumors with CT, although two of the cases were hypodense lesions that exhibited poor contrast enhancement. It was not determined whether these two lesions were largely necrotic. The lack of central contrast enhancement on MRI examination in the present case is consistent with extensive necrosis. In general, however, the differential diagnostic value of contrast enhancement of solitary bony lesions on MRI remains largely undetermined.

It is not unexpected for giant cell tumor of bone to show variable amounts of hemorrhage, usually with minimal hemosiderin pigment [4]. Necrosis is more unusual but may be expected in the setting of pathological fracture. Some 5%-10% of conventional giant cell tumors exhibit prominent fibrohistiocytic features with xanthoma cells (fibrous histiocytoma-like variant) [4]. These features likely represent the sequelae of areas of intratumoral necrosis and hemorrhage, with subsequent organization by reparative tissues, lipoidization, and eventual replacement by fibrohistiocytic tissues. It is rare for a giant cell tumor to exhibit almost complete necrosis, as in this case, particularly in the absence of superimposed fracture. In those rare cases with almost total necrosis, the finding of ghost-like residues of giant cell tumor is essential to the recognition of this pathophysiologically altered in a giant cell tumor of bone. This finding may prevent diagnostic confusion when radiographic findings otherwise consistent with giant cell tumor are present. Massive necrosis may conceivably represent a stage in the evolution of some giant cell tumors to 'true' fibrous histiocytomas of bone.

In summary, we present a giant cell tumor of the distal end of the femur that exhibited unusual diaphyseal extension and atypical MRI features. MRI demonstrated differing zonal signal characteristics in the distal metaphyseal/epiphyseal versus the diaphyseal components of the tumor. It also depicted an irregular, proximal tumor margin with an unusual, enhancing, peripheral zone. The atypical MRI features may be related to the unusual finding at pathological examination of an almost entirely necrotic giant cell tumor. This massive necrosis may illustrate a stage in the evolution of some giant cell tumors to fibrous histiocytoma-like variants of giant cell tumor or to conventional, benign fibrous histiocytoma of bone.

References