Fibroblastic Type Osteosarcoma of the Ulna: a Case Report of a Tumor in a Rare Location with Atypical Imaging Findings

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The ulna is a rare site of origin for osteosarcoma, and purely osteolytic osteosarcomas are uncommonly noted on conventional radiographs. We present a patient with a lytic lesion of the distal ulna for which imaging findings suggested an aneurysmal bone cyst. The lesion was histologically confirmed to be a fibroblastic osteosarcoma.

Index terms:
Bone neoplasm, ulna
Osteosarcoma
Radiography
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Osteosarcoma is the second most common primary malignant bone tumor, accounting for approximately 15% of all primary bone tumors confirmed at biopsy (1). Osteosarcomas most frequently affect long bones, particularly around the knee, and rarely the ulna (1, 2). On radiographs, osteosarcomas classically appear as aggressive destructive lesions with a mixed osteolytic and sclerotic pattern, associated periosteal reaction, and soft-tissue mass. The purely osteolytic form of osteosarcoma occurs in about 10% of all cases (3). The differential diagnosis of lytic and expansive bone lesions includes solitary bone cysts, aneurysmal bone cysts, giant cell tumors, enchondromas, and telangiectatic osteosarcomas (4).

We present a case of fibroblastic osteosarcoma, with an accompanying pathologic fracture, which arose from the distal ulna and was initially suspected to be an aneurysmal bone cyst based on its features on conventional radiographs and magnetic resonance (MR) images.

CASE REPORT

A 37-year-old woman presented with a one-month-history of right wrist pain after lifting her baby. Three days before admission, the wrist pain increased when she felt a snap while lifting her baby again. Radiographs showed an eccentric radiolucent lesion with endosteal scalloping, cortical thinning, and a minimally bulging contour arising from the metaepiphyseal region of the distal right ulna (Fig. 1A). MR images demonstrated a lobulated, septated lesion with multiple locules containing fluid-fluid levels (Fig. 1B, C). In addition, focal cortical disruptions along the ventral aspect of the distal ulna suggested the presence of pathologic fractures (Fig. 1D). Contrast-enhanced T1-weighted, fat-suppressed images revealed peripheral, septal, solid enhancing portions within the tumor (Fig. 1E).

Concerning the differential diagnosis, we considered a primary aneurysmal bone cyst (ABC) and a solid tumor, such as a giant-cell tumor or chondroblastoma with a predominant secondary aneurysmal bone cyst component.

The orthopedic surgeon performed a massive curettage of the bone lesion and placed a calcium phosphate cement graft. Histologically, the neoplasm consisted of a

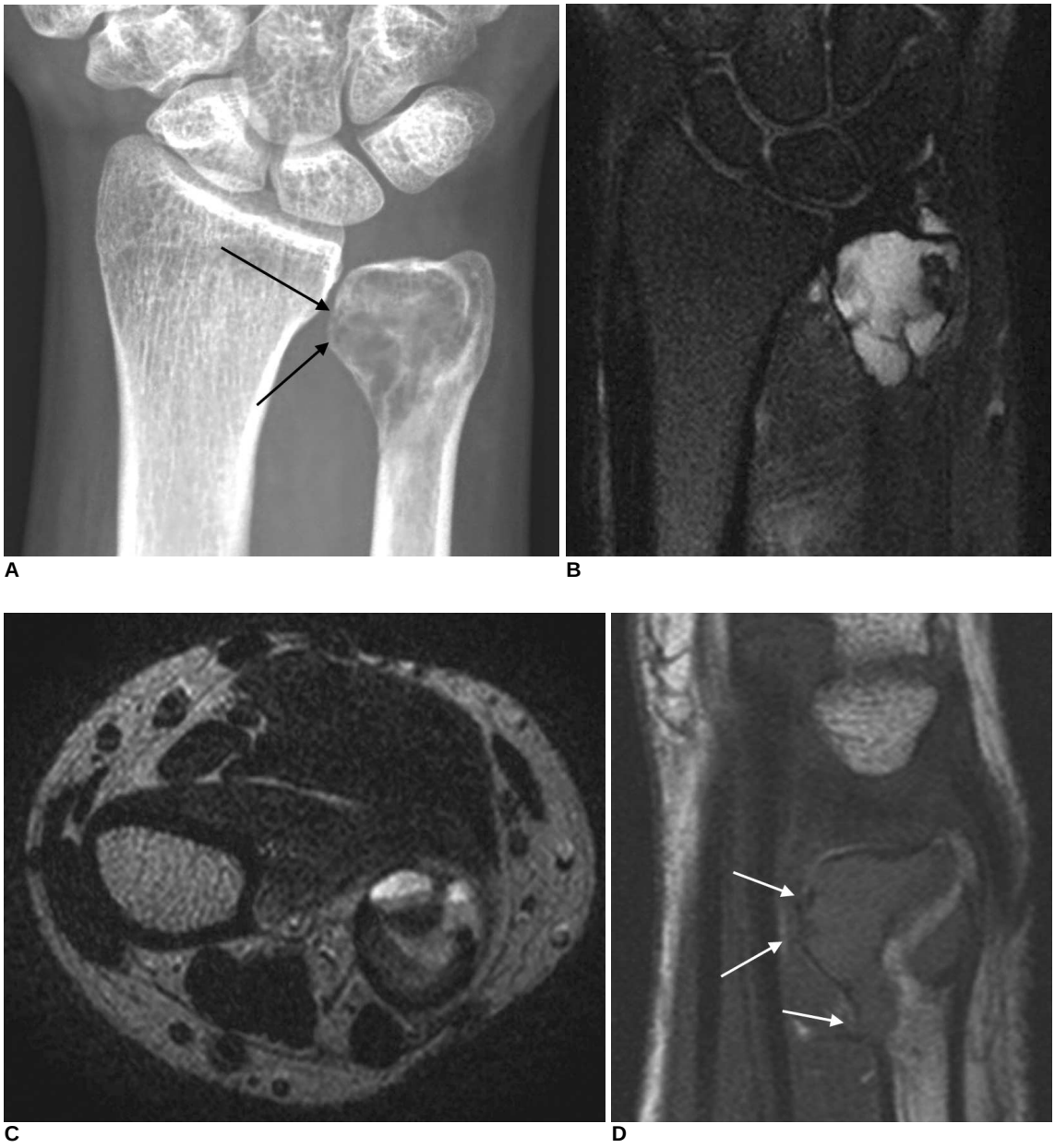
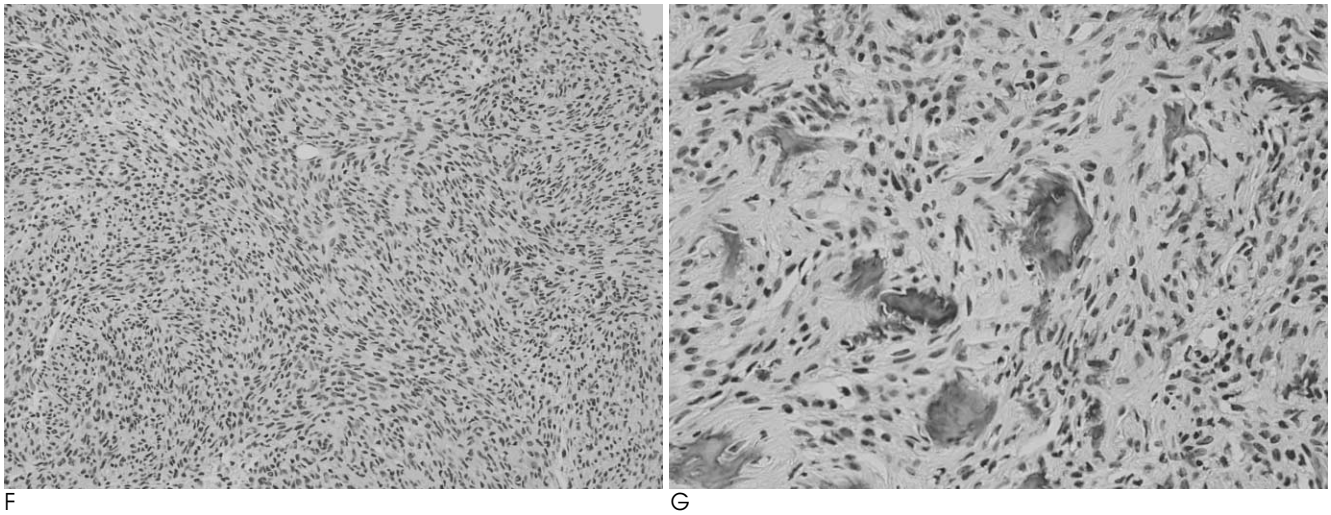


Fig. 1. Imaging findings of fibroblastic osteosarcoma in distal ulna of 37-year-old woman.
A. PA view of right wrist reveals well-marginated osteolytic lesion of slightly expansile nature with suspicious focal cortical disruptions (arrows) in distal ulna metaphysis and epiphysis.
B. Coronal T2-weighted (TR/TE = 2925/54), fat-suppressed MR imaging shows multi-loculated, high signal intensity lesion in distal ulna.
C. Axial T2-weighted (TR/TE = 4319/100) MR imaging shows fluid-fluid levels within cystic lesion.
D. Sagittal T1-weighted (400/18.46) MR imaging reveals cortical disruptions (white arrows).
E. On gadolinium-enhanced, fat-suppressed, T1-weighted (517.9/20) MR imaging, focal enhancement of lesion (white arrows) is noted along with largely non-enhancing portions.



F G

Fig. 1. Imaging findings of fibroblastic osteosarcoma in distal ulna of 37-year-old woman.
F. Large part of tumor is composed of spindle cells, often in "herringbone" pattern (Hematoxylin & Eosin staining, ×200).
G. Focal osteoid production by neoplastic stromal cells (Hematoxylin & Eosin staining, ×400).

spindle-cell proliferation that produced a focal "herringbone" pattern (Fig. 1F). The presence of focal osteoid production distinguished this fibroblastic variant of osteosarcoma from malignant fibrous histiocytoma (Fig. 1G). On immunohistochemical staining, the tumor cells showed increased Ki-67 expression and were negative for CK (cytokeratin) and EMA (epithelial membrane antigen). After initial and subsequent review of multiple specimens, pathologists at our institution and an outside referral institution agreed that the final histopathologic diagnosis was a fibroblastic-type osteosarcoma with a few giant cells and foam cells and malignant osteoid formation.

The patient received neoadjuvant chemotherapy followed by wide en bloc resection of the distal ulna as a definitive operation.

DISCUSSION

Osteosarcoma is second in frequency only to multiple myeloma when it comes to primary malignant neoplasms of bone. Osteosarcoma is characterized histologically by malignant tumor cells that directly produce osteoid or immature bone (5). High-grade intramedullary osteosarcoma accounts for about 75% of all lesions and is also referred to as conventional osteosarcoma (1). The microscopic pathology of conventional osteosarcoma has traditionally been subdivided into three categories: osteoblastic (50%), chondroblastic (25%), and fibroblastic (25%) (5).

Involvement of the forearm by conventional osteosarcoma is extremely rare (1). In most series, primary tumors of the ulna account for less than 1% of all lesions and less than 2% of all long bone lesions (6). Exner et al. (2) reported that the ulna was the primary site in 21 cases and the primary site for malignant bone tumors in three cases out of more than 2000 neoplasms and tumor-like bone lesions in the Balgrist tumor registry. Arndt et al. (7) reported four cases of ulnar involvement in 1649 osteosarcoma patients, with the distal ulna (the same location as in our case) involved in only two cases.

The radiologic features of osteosarcoma vary over a wide spectrum. In most cases, typical radiographic features clearly suggest the aggressive bone-forming nature of the lesion. Common features of osteosarcoma include tubular bone metaphyseal location, mixed pattern of osteolysis and osteosclerosis, cortical destruction, periostitis, and soft-tissue mass (1, 5). With the exception of metaphyseal localization in tubular bone, the commonly cited features were not seen in our case.

Purely osteolytic lesions are uncommonly seen on conventional radiographs, accounting for approximately 10% of all patients and usually indicating the presence of a fibroblastic, fibrohistiocytic, telangiectatic, or giant cell-rich type of osteosarcoma (3). The differential diagnosis of purely lytic osteosarcoma includes other bone malignancies that do not form osteoid, such as malignant fibrous histiocytoma, fibrosarcoma, metastatic disease, and Ewing's sarcoma (8). In cases of purely lytic osteosarcoma presenting with benign radiographic features, such as our case, the most likely tumors to be considered in the differential diagnosis are an aneurysmal bone cyst and a giant cell tumor of bone (8).

MR images characteristically reflect the predominant cellular component. Thus, a fibroblastic osteosarcoma is

generally characterized by short T1 and T2 relaxation times of fibrous tissue. A chondroblastic osteosarcoma typically has increased signal intensity on T2-weighted images in areas of uncalcified cartilage, with focal areas of low signal intensity corresponding to the extent of cartilage calcification. Osteoblastic osteosarcoma is characterized by cloudlike areas of low signal intensity on both T1- and T2-weighted images, depending on the degree of osteoblastic reaction (9).

In our case, the distal ulnar lesion presenting with imaging features of a lobulated, well-marginated osteolytic lesion with cortical thinning and multiple loculations containing fluid-fluid levels seemed to strongly suggest the presence of an aneurysmal bone cyst. Mahnken et al. (4) reported that reevaluation with a combination of conventional radiographs and MR images had a sensitivity of 83% and a specificity of 70% in all patients with initially suspected primary aneurysmal bone cysts. Considering the ABC component, telangiectatic osteosarcoma would have been another possible diagnosis. Telangiectatic osteosarcoma most frequently involves the femur, followed by the tibia and the humerus. It usually involves the metaphysis and may extend to the epiphysis. MR imaging is known to show fluid-fluid levels and inhomogeneous enhancement (5). In our case, we did not consider telangiectatic osteosarcoma in the differential diagnosis because of the patient's age and the lack of enhancement on contrast images. To our knowledge, the English literature has rarely reported the fibroblastic type of osteosarcoma to present with a predominant ABC component, especially on MR imaging.

We report a case of fibroblastic osteosarcoma arising in the distal ulnar meta-epiphysis with ABC-like imaging features both on radiography and MRI.

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