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Fibrous Dysplasia Associated with Cortical Bony Destruction: CT and MR Findings

Lawrence Yao, Jeffrey J. Eckardt, and Leanne L. Seeger

Objective: Fibrous dysplasia is a benign disorder of bone that can cause cortical thinning as well as bony expansion. Full-thickness cortical bony destruction, however, typically characterizes more aggressive or malignant lesions of bone. We present three cases of fibrous dysplasia that illustrate this more aggressive and potentially confusing feature.

Materials and Methods: The findings on radiography and cross-sectional imaging studies are reviewed in three surgically proven cases of fibrous dysplasia that exhibited full-thickness cortical bony destruction.

Results: Well-defined cortical perforations without associated periosteal reaction were clearly seen on CT. This finding was suspected on plain radiography in only one of the three cases. A small associated soft tissue mass was detected by MR imaging in this case. Many cortical perforations were seen in the one case of polyostotic fibrous dysplasia.

Conclusion: Full-thickness cortical bony destruction can be seen in fibrous dysplasia. This finding, which may only be evident on cross-sectional imaging studies, should not by itself alter a diagnostic impression of fibrous dysplasia supported by other radiographic and clinical features.

Index Terms: Bones, neoplasms—Bones, diseases—Computed tomography—Magnetic resonance imaging.

Fibrous dysplasia is a common benign disorder of bone in which proliferating fibrous tissue replaces the bony spongiosa. A characteristic, irregular pattern of "fibroosseous metaplasia" occurs in this proliferating tissue (1). Fibrous dysplasia commonly presents on radiography as an incidental lucent lesion with well-defined and occasionally sclerotic margins (2). These lesions may have a diffuse slightly increased internal density lending a "ground-glass" appearance. Fibrous dysplasia may cause cortical thinning as well as bony expansion. Focal full-thickness cortical bony destruction, however, typically characterizes more aggressive lesions of bone such as giant cell tumor, metastatic disease, and primary bone sarcomas. This report illustrates the variable manner in which fibrous dysplasia may focally destroy and extend through cortical bone.

CASE REPORTS

Case 1

A 23-year-old man had had vague pain in the distal femur for ~1 year. Radiography demonstrated a lytic lesion in the distal femoral diaphysis (Fig. 1a). The lesion exhibited benign features with the exception of full-thickness cortical destruction at the medial aspect of the metaphysis. Magnetic resonance better illustrated the focal full-thickness cortical destruction—without significant cortical thinning at the margins of the defect—and a small extraosseous tumor component (Fig. 1b). At open biopsy, a small soft tissue mass extruded through a 3 × 2 cm defect in the medial femoral metaphyseal cortex. After a final pathological diagnosis of fibrous dysplasia was made, definitive curettage of the right femoral lesion was performed, with iliac crest and allograft bone packing of the large femoral defect.

Case 2

A 38-year-old woman presented with a 6 month history of left shoulder and elbow pain, without antecedent trauma. On physical examination the patient lacked 15° of

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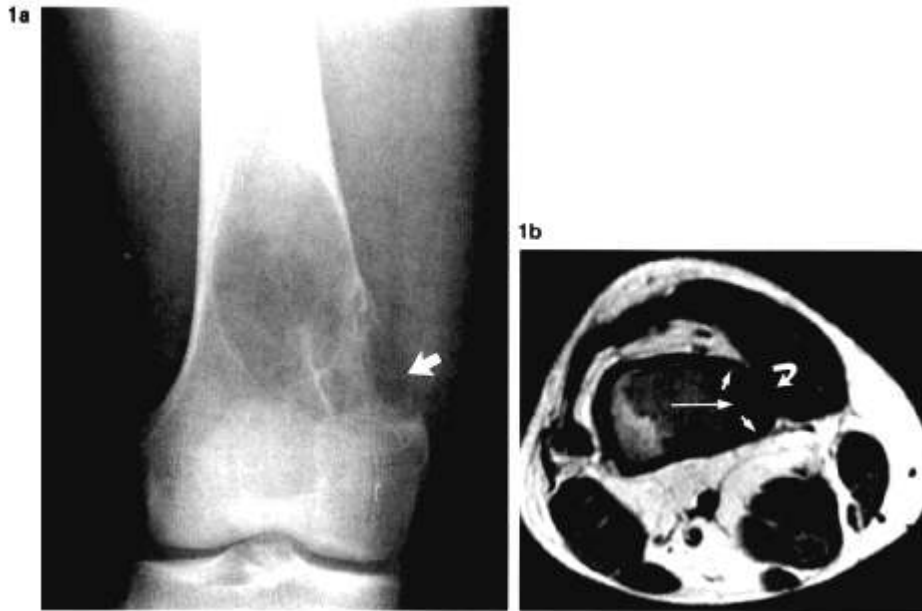


FIG. 1. Case 1. **a:** Anteroposterior radiograph of the femur demonstrates a well-defined lytic lesion of the distal femoral diaphysis, with sclerotic borders, ground-glass density, and some internal trabeculation. Full-thickness cortical destruction, however, is noted in the medial femoral metaphysis (arrow). **b:** Axial proton density MR image (1.5 T, TR 1,800, TE 20) depicts the area of marrow replacement in the distal femoral metaphysis, as well as the medial area of cortical destruction (short arrows). Areas of mineralization within the lesion exhibit lower signal intensity (long arrow). There is a small extraosseous component of tumor (curved arrow).

extension. Radiography of the shoulder was normal, but radiography of the elbow demonstrated an osteolytic lesion of the distal humerus (Fig. 2a) with well-defined and predominately sclerotic margins. The lesion exhibited endosteal scalloping, without significant bony expansion. Computed tomography demonstrated focal areas of full-thickness cortical destruction (Fig. 2b) that were not appreciated on radiography. At surgery, a firm reddish brown tumor protruded through an infarction of the anterior cortex of the distal humeral metaphysis. Open biopsy and intralesional curettage were performed, followed by iliac crest bone grafting. The pathological diagnosis

was fibrous dysplasia with abundant hemosiderin deposition.

Case 3

A 47-year-old man had a history of polyostotic fibrous dysplasia involving the left humerus and left radius. The patient had sustained a pathological fracture of the radius at age 14 which required iliac crest bone grafting. After the recent onset of increasing left elbow pain, the patient sought medical advice and his arm was placed in a sling

FIG. 2. Case 2. **a:** Anteroposterior radiograph of the left humerus demonstrates a lytic lesion of the distal left humeral metaphysis, with extension into the lateral condyle. The lesion exhibits well-defined margins, ground-glass density, and endosteal scalloping (arrows). On other projections, there was no definite evidence of full-thickness cortical penetration. **b:** Axial CT scan through the distal humeral condyles demonstrates an area of full-thickness cortical destruction (small white arrows) along the anterior aspect of the distal humeral metaphysis, without significant adjacent cortical thinning. There is a small extraosseous soft tissue component (curved arrow) without effacement of adjacent fat planes. More cephalad images better demonstrated a second area of full-thickness cortical destruction along the olecranon fossa (black arrow).



for possible "micro fracture" through the distal humerus. Radiography 3 months after the onset of symptoms demonstrated a left humerus widely affected by fibrous dysplasia. Sclerotic change was seen in the proximal and mid aspects, whereas lytic bony destruction predominated in the distal humerus (Fig. 3a). No definite fracture could be seen on radiography at this time. Computed tomography of the distal humerus (Fig. 3b) demonstrated diffuse cortical thinning and bony expansion, as well as multifocal areas of full-thickness cortical destruction. At surgery, the distal humeral cortex showed multiple perforations, which exuded fluid on exploration. The lytic focus in the distal humerus was found to be largely fluid filled and interspersed with fibrous appearing tissue. The patient underwent curettage of the distal left humerus followed by iliac crest bone grafting. Pathologic analysis of curettements revealed fibrous dysplasia with cyst formation.

DISCUSSION

Monostotic fibrous dysplasia typically appears on radiography as a well-defined lucent lesion, frequently with sclerotic margins. The lesions have a predilection for the diaphysis, and may be central or eccentric (2). The density of the lesions is quite variable, with many lesions exhibiting a diffuse mild increase in density referred to as a ground-glass appearance. Lesions in the facial bones and skull base tend to be densely sclerotic. Polyostotic fibrous dysplasia has some tendencies that differ from monostotic fibrous dysplasia; it tends to involve a larger portion of any individual bone, is more likely to exhibit a ground-glass appearance, is more commonly associated with face or skull lesions, and may cause bowing in cases of long bone involvement (2).

Fibrous dysplasia may cause bony expansion and pronounced cortical thinning. Frank cortical bony

destruction and associated soft tissue extension of tumor, however, are not traditionally associated with fibrous dysplasia. The incidence of these features is unknown, as many asymptomatic lesions of fibrous dysplasia are not thoroughly studied. Cortical violation may only be appreciated at CT or MR examination, as in Case 2. In such cases the imaging features may seem inconsistent with a preliminary radiographic diagnosis of fibrous dysplasia.

Cases of fibrous dysplasia in which there is dramatic bony expansion may be expected to exhibit marked cortical thinning, to the extent that the cortex is no longer visible (3,4). However, fibrous dysplasia can exhibit multifocal areas of cortical perforation with or without diffuse cortical thinning as illustrated by Cases 1 and 2. When the cortical perforations are large or multifocal, the risk of pathologic fracture must be regarded as high. The presence of focal cortical destruction in a mildly expansile bony lesion can be encountered in nonossifying fibroma (or fibrous cortical defect), which, unlike fibrous dysplasia, is an eccentric cortical-based process (1). The incidence of cortical perforation is probably higher in nonossifying fibroma than in fibrous dysplasia.

In general, when the imaging workup of a focal bony lesion reveals full thickness cortical destruction, an aggressive or malignant process must be suspected. When other signs point to the diagnosis of fibrous dysplasia, two other scenarios may deserve consideration. First, malignant transformation of fibrous dysplasia is rare but may occur in ~1% of cases (5). More cases of malignant transformation occur in polyostotic fibrous dysplasia than in monostotic disease. Secondary malignancies arising in fibrous dysplasia include osteogenic sarcoma, fibroblastic osteogenic sarcoma, fibrosarcoma, or, rarely, chondrosarcoma (1). Malignant

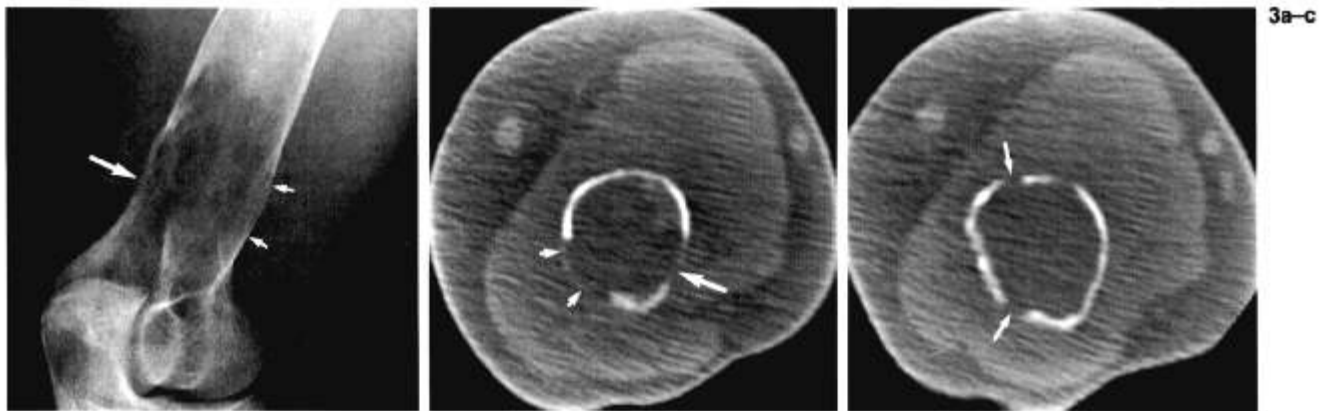


FIG. 3. Case 3. **a:** Oblique radiograph of the distal left humerus demonstrates lytic bony destruction with associated bony expansion, cortical thinning (smaller arrows), and focal cortical loss (bigger arrow). The more proximal aspects of the humerus demonstrated widespread replacement of the medulla by ground-glass density in this case of polyostotic fibrous dysplasia. **b and c:** Axial CT scans at two levels through the distal humeral lesion. In (b) there is diffuse cortical thinning that is focally pronounced in one area (small arrows) as well as complete focal cortical destruction in another area (large arrow). At a slightly more cephalad level (c) there are separate, focal areas of full-thickness cortical destruction (arrows) without evidence of periosteal reaction.

transformation may appear on radiography as an area of accelerated bony destruction and cortical perforation.

Second, aneurysmal bone cysts are frequently engrafted on other primary lesions of bone and can occur in association with fibrous dysplasia (6). An aneurysmal bone cyst complicating fibrous dysplasia may explain more aggressive features such as cortical penetration and associated soft tissue extension of tumor. This uncommon occurrence should be distinguished from the more common cystic degeneration noted in fibrous dysplasia, as illustrated in Case 3 (1). Cystic degeneration may be manifested on MRI by focal well-defined areas of fluid signal intensity, or fluid–fluid levels, within a lesion of fibrous dysplasia (7,8).

In summary, fibrous dysplasia may exhibit focal or multifocal full-thickness cortical bony destruction and, less commonly, soft tissue extension of tumor. The recognition of this feature will be greater in lesions studied by CT or MR. The finding of full-thickness cortical destruction is probably more typical of nonossifying fibroma but usually typifies more aggressive or malignant lesions of bone. However, when other radiographic findings suggest the diagnosis of fibrous dysplasia, the find-

ing of cortical violation should not by itself alter the diagnostic impression of benignity. This finding, however, does mark an increased risk of pathologic fracture and may warrant prophylactic curettage and bone grafting after confirmational biopsy.

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