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Case Report

MR Appearance of Cystic Fibrous Dysplasia

Andrew J. Fisher, William G. Totty, and Michael Kyriakos

Abstract: The radiologic and MR appearances of cystic fibrous dysplasia (FD) along with pathologic correlation are reported in a symptomatic patient. As a guide for operative intervention, MRI is helpful in both characterizing the lesion and its internal structure and determining the extent of osseous involvement. Cystic FD without an aneurysmal bone cyst component has not been previously reported in the radiologic literature. **Index Terms:** Bones, cysts—Bones, diseases—Magnetic resonance imaging.

Fibrous dysplasia (FD) is a common, benign osseous lesion with multiple, well documented radiologic appearances (1). It is usually asymptomatic and detected on routine radiographs or screening for unrelated trauma or pathology. Cystic degeneration is relatively uncommon and must be differentiated from malignant degeneration, especially in the setting of a symptomatic patient or a rapidly expanding lesion. We report a case of FD with the unique radiologic appearance of a large cystic component containing a fluid-fluid level, but without the histologic presence of a secondary aneurysmal bone cyst. Preoperatively, MRI demonstrated the extent of the lesion and accurately characterized its multiple components, providing valuable information for surgical planning that was not available from the radiographs alone.

CASE REPORT

A 24-year-old woman presented with an 8 week history of right leg pain that was exacerbated by increased activity. She had originally been treated by her local physician with muscle relaxants for 6 weeks without symptomatic relief. One week prior to admission, plain radiographs (Fig. 1) demonstrated a cystic lesion in the proximal diaphysis of the right femur with endosteal erosion along its proximal margin. The distal margin of the lesion was poorly defined and there was no detectable matrix. The lesion showed increased activity on bone scintigraphy

(Fig. 2). The patient was given crutches for partial weight bearing, and surgical intervention was scheduled. The differential diagnosis included a solitary bone cyst, an aneurysmal bone cyst (ABC), and FD.

Upon admission, the musculoskeletal examination was normal without right hip joint deformity, tenderness over the lesion, or significant reduction in the muscular strength of the leg. No abnormal cutaneous pigmentation was identified. Preoperative MRI (Fig. 3) using proton density- and T2-weighted sequences demonstrated a mildly expansile, 11 cm lesion longitudinally oriented along the right proximal femoral diaphysis, extending into the midshaft. On the supine sagittal images, the signal characteristics of the lesion indicated multiple components. Along cranial and caudal margins of the lesion, tissue with intermediate signal on T1-weighted images and inhomogeneous intermediate and high signal on T2-weighted images was present, indicating nonmarrow, cellular tissue. The margins of the cellular tissue were sharply defined, with no indication of invasion of the surrounding bone or marrow. Within the central portion of the lesion, between the cellular areas, a fluid-fluid level was noted. The dependent portion of this region contained material that was intermediate in signal intensity on both T1- and T2-weighted images. In the nondependent, anterior portion, the material was high in signal intensity on both T1- and T2-weighted images. This combination indicated a fluid-filled cystic cavity, with blood breakdown products layering within the cavity. Cortical thinning was noted anteriorly and laterally, although no disruption of the cortex was identified. The central fluid portion of the lesion on MR corresponded to the more expansile cranial portion of the lesion noted on the radiographs.

At surgery, an intralesional resection was performed, with bone allograft and intramedullary locked nail fixation. The initial resection was through the proximal margin of the lesion. The cyst contained brown, opaque liquid. Frozen section histology showed no malignant tissue

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FIG. 1. Anteroposterior (a) and frog leg (b) radiographs of the proximal femur show an expansile, lytic lesion of the proximal femoral diaphysis with a lucent matrix. The cortex is thinned in the proximal portion of the lesion. The margins of the lesion are indistinct.

and suggested a cartilage lesion. The resection was extended to include the proximal tumor. The patient did well postoperatively and was discharged with partial weight bearing on the 8th postoperative day.

The resected portion of the femur was 11.6 cm long with its contour distorted by a bulge in its proximal aspect. When bivalved along its long axis, there was an empty cyst in the proximal portion of the bone, corresponding to the bulge, and measuring 5.6 cm in length and 3.2 cm in width at its widest point (Fig. 4). The cortex in this area was intact but thinned. The cyst was lined by a 0.5 mm thick, tan-brown membrane that could be peeled from the endosteal surface. At its distal extent, the cyst was continuous with a firm, gray-white fibrous lesion that was 3.5 cm in length and 2.0 cm wide. Normal yellow marrow was present distal to this lesion.

Microscopic examination of sections from the solid area adjacent to the cyst showed FD. The stroma varied from compact areas composed of bland spindle cells to edematous zones where the stromal cells were stellate, round, or oval and in which areas of myxoid degeneration were present. The amount of metaplastic bone varied, with some areas consisting almost entirely of stroma without bone formation, while in other foci the bone and stroma appeared equal in amount. The bone consisted of irregular spicules and trabeculae without osteoblastic rimming; polarization showed it to be predominantly of woven type, although some lamellar change was noted at the periphery of some trabeculae. The cyst wall was composed of a nondescript fibrous stroma similar to some of the stroma present in the FD. Within the membrane, a few trabeculae of metaplastic woven bone were present, as well as focal regions of reactive new bone rimmed by plump osteoblasts. The overall impression was that the cyst had formed within the FD with remnants of the lesion still present in the cyst wall.

DISCUSSION

Originally described by Lichtenstein and Jaffe (2), FD is an abnormality of bone formation. Typi-

cally, patients present in adolescence or early adulthood, and symptoms usually are manifest as non-specific pain, swelling, and occasionally pathologic fracture. Fibrous dysplasia is seen in both a monostotic and a polyostotic form. Monostotic disease is six times as frequent as the polyostotic form and typically follows a less aggressive course. While most series do not note a sexual predilection, Kransdorf and colleagues (1) found males to be affected twice as frequently as females in monostotic FD and 1.3 times as often in the polyostotic form. Pregnancy and estrogen supplementation have been linked to de novo presentation or exacerbation of previously known FD (3).

Fibrous dysplasia may have a varied radiologic appearance. The most common finding is an isolated, predominately lytic, osseous lesion. The ribs (28%) are preferentially affected in monostotic FD, followed by the femur (23%), craniofacial bones (20%), tibia (8%), and the remaining bones to a lesser extent (1). The proximal femur is involved in the vast majority of patients with polyostotic FD. Polyostotic disease may present as McCune-Albright syndrome with associated endocrine abnormalities and cutaneous café-au-lait spots.

In FD, the normal intramedullary spaces are replaced by a fibroosseous matrix with variable amounts of stroma and bone. The stroma is moder-

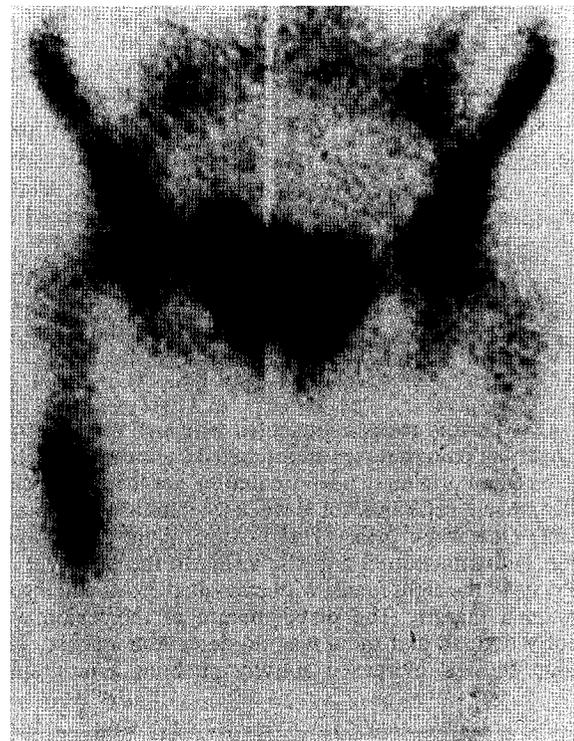


FIG. 2. Technetium-99m bone scintigram demonstrates a focal region of increased radiopharmaceutical uptake in the right proximal femur corresponding to the radiographic abnormality. No other abnormalities are noted.

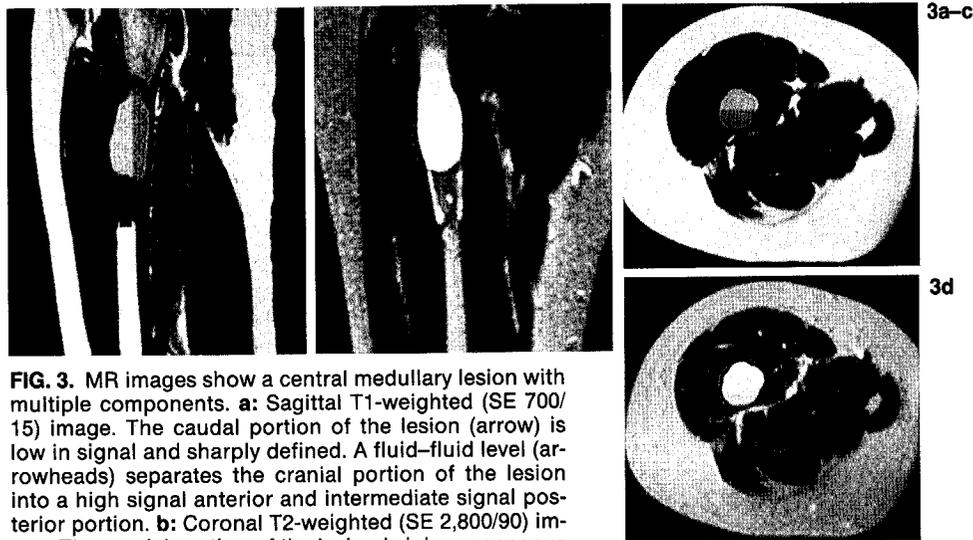


FIG. 3. MR images show a central medullary lesion with multiple components. **a:** Sagittal T1-weighted (SE 700/15) image. The caudal portion of the lesion (arrow) is low in signal and sharply defined. A fluid-fluid level (arrowheads) separates the cranial portion of the lesion into a high signal anterior and intermediate signal posterior portion. **b:** Coronal T2-weighted (SE 2,800/90) image. The caudal portion of the lesion is inhomogeneous with both intermediate and high signal components. The cranial portion, with the coronal plane through the anterior part of the lesion seen in (a), is high in signal. **c:** Axial T1-weighted (SE 700/15) image. **d:** Axial T2-weighted (SE 2,800/90) image. These images (c and d) through the cranial portion of the lesion show the fluid-fluid level with the anterior portion being relatively high in signal on T1- and very high signal on T2-weighted images. The posterior component is intermediate in signal on T1- and high in signal on T2-weighted images.

ately to loosely cellular and contains irregular trabeculae and spicules of metaplastic woven bone, characteristically without osteoblastic rimming. The radiographic density of the lesion depends upon both the amount of woven bone present and its degree of mineralization.

Magnetic resonance imaging is beneficial in both characterizing the internal architecture of FD and determining the extent of intraosseous and any extraosseous involvement. On MR images, FD typically has sharply demarcated margins, although the signal characteristics may vary. Norris et al. (4) found intermediate signal intensity of T1-weighted images in all 13 patients examined, while Utz et al. (5) noted FD to be of intermediate signal in all 11 FD lesions not affected by pathologic fracture. The T2 appearance varied greatly, although both groups noted frequent high signal intensity, 46% of the Norris population and 64% in the Utz study.

While cystic degeneration of FD with secondary ABCs has been reported, we know of no reports of primary cystic FD without pathologic detection of ABC elements. A cystic component is of diagnostic importance because its presence produces a more aggressive radiographic appearance that must be differentiated from malignant disease. Simpson and colleagues (6) have documented the difficulties in radiographically differentiating intraosseous sarcomas from FD with cystic components because both produce expansion of the lesion suggesting aggressive behavior.

Malignant transformation of FD is a rare, although well documented, complication of the dis-

ease (7,8). As of 1986, only 83 cases of malignant transformation of FD had been reported, with 57% in patients with monostotic FD and 43% in those with the polyostotic form (8). Approximately 0.4% of monostotic lesions undergo such transformation, while as many as 4% of cases of McCune-Albright syndrome do so. Malignant transformation to osteosarcoma is the most common pattern (54%), followed by fibrosarcoma (30%) and chondrosarcoma (16%) (7). Additionally, in one-third of the documented cases of FD complicated by malignancy, a history of antecedent radiation therapy was elicited. Although most patients with malignant tumors arising in FD lesions were initially diagnosed in childhood, the malignancy did not develop until early adulthood, the mean being 32.7 years (8). The radiographic findings of FD complicated by malignancy are not always obvious. Rapid expansion and locally aggressive changes including cortical infiltration, periosteal reaction, and pathologic fractures may be clues suggesting malignant change.

In the current case, radiographs demonstrated a lytic lesion with expansion and indistinct margins, characteristics that suggest an aggressive nature. Magnetic resonance imaging demonstrated a well defined margin with characteristics of a fluid-filled cyst. Magnetic resonance accurately defined the nature of the lesion as benign and nonaggressive, thereby distinguishing it from malignant degeneration. The presence of fluid-fluid levels on MR, as seen in our case, is a nonspecific finding. Although originally thought to be specific for ABCs, fluid-fluid levels simply represent blood and serous prod-

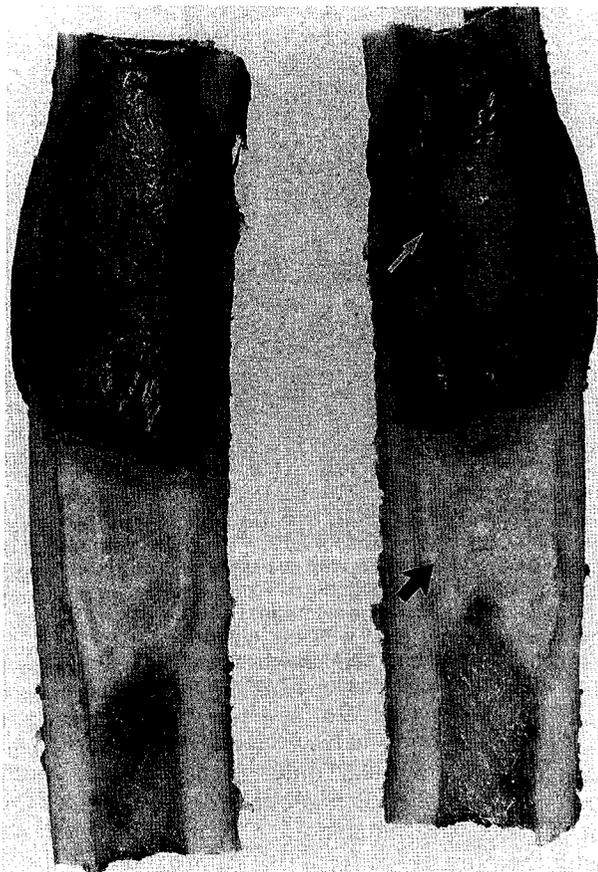


FIG. 4. Bivalved, resected portion of the proximal femur showing a proximal cystic cavity. The adjacent cortex is thinned. The distal end of the cyst is continuous with a solid area of fibrous dysplasia (black arrow). A ridge of residual bone is noted along the length of the cyst wall in one of the halves (white arrow).

ucts of mixed constituency within a lesion and also occur in solitary bone cysts, chondroblastomas, giant cell tumors, and telangiectatic osteosarcomas (9-13).

CONCLUSION

In conclusion, this case represents a unique presentation of FD not previously reported. The value

of MRI over plain radiography is clearly demonstrated. Magnetic resonance aids both in fully characterizing the lesion and its benign nature as well as in delineating the nonmarrow cellular components and intralesional structure. While FD with secondary ABC involvement has been noted, no authors have conclusively demonstrated cystic FD without the histologic presence of ABC elements. The presence of a fluid-fluid level, although a nonspecific finding, limits the differential possibilities to those osseous tumors previously mentioned.

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