Musculoskeletal case 16. Diagnosis

Fibrous dysplasia

The plain radiograph of the pelvis and proximal femora (Fig. 1, see presentation page 170) demonstrates well-defined lucent lesions involving the right ilium (supra-acetabular region) and the proximal femur, with a hazy "ground glass" type of matrix. There is bowing of the proximal femur with an associated pathologic fracture. This type of bowing is referred to as a "shepherd's crook" deformity. Computed tomography (Fig. 4) also showed multiple lytic lesions within both iliac bones and the right sacral ala. On T_1-weighted coronal magnetic resonance imaging (Fig. 2, see presentation), the lesions are of similar signal intensity to that of their surrounding muscles. There is mild expansion of the medullary cavity. On the coronal fSTIR sequence (Fig. 3, see presentation), the medullary cavity of the iliac bones demonstrates heterogeneous signal. Cystic-like lesions are present within the proximal right femur. No extraosseous soft-tissue mass is present. A pathologic fracture is again noted within the proximal right femur as evidenced by a low-signal transverse line at the lateral aspect of the proximal right femur. A diagnosis of fibrous dysplasia was suggested on imaging and confirmed by CT-guided core biopsy.

Fibrous dysplasia is a benign fibro-osseous disease that is neither familial nor hereditary but is considered a developmental abnormality involving excess proliferation and maturation of fibroblasts. Normal bone is replaced by a combination of these fibroblasts and abnormally arranged dysplastic immature woven bone. The amount of woven bone in the fibro-osseous tissue and the extent to which it is mineralized ultimately determine the radiographic density of the lesion.[1,2]

The process can affect a single bone (monostotic fibrous dysplasia) or many bones (polyostotic fibrous dysplasia). Monostotic disease seldom progresses to the polyostotic form. The disease is usually found in adolescents and young adults and is equally common in men and women. The monostotic form is approximately 6 times more common than the polyostotic form. The clinical course of fibrous dysplasia ranges from the more common asymptomatic form to more severe, and rarer, types with multiple pathologic fractures, rapidly progressive deformities, severe pain debilitation, and even death.[2] Generally the younger the patient is and the more extensive the lesions are at presentation, the more progressive the condition is.

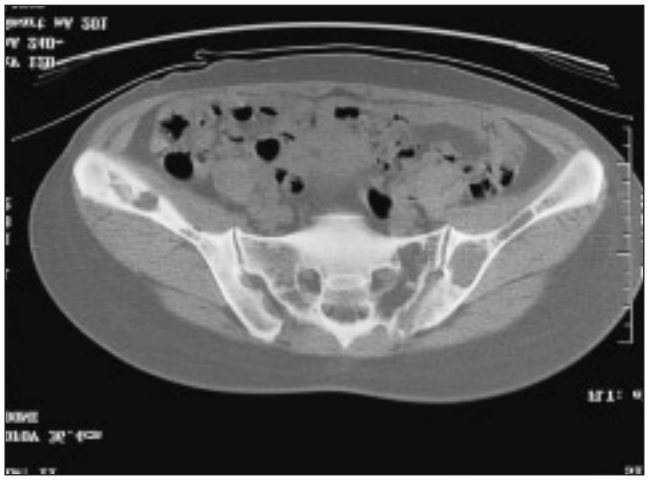

FIG. 4.

The frequency of malignant degeneration is reported to be less than 1%. The associated malignant diseases in descending order of occurrence are osteosarcoma, fibrosarcoma and chondrosarcoma.[3] The serum alkaline phosphatase level may be slightly elevated in fibrous dysplasia. Serum calcium and phosphate levels usually remain normal.[4]

The McCune–Albright syndrome is a form of polyostotic fibrous dysplasia typically found in women and associated with *café-au-lait* spots and sexual precocity.[5] Cherubism is an autosomal dominant form of fibrous dysplasia that affects the jaws. It is often recognized in babies between 18 and 24 months of age, and there is usually spontaneous regression during puberty. Mazabraud's syndrome is fibrous dysplasia with associated soft-tissue myxoma occurring in the vicinity of the affected bone. It is more often seen with the polyostotic form.

The differential diagnosis for fibrous dysplasia should include neurofibromatosis, osteitis fibrosa cystica and Paget's disease.

References

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4. Yochum TR, Haug JV, Rowe LJ. *Radiology study guide*. Philadelphia: Williams & Wilkins; 1998. p. 374-9.
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