An Unusual Case of Polyostotic Fibrous Dysplasia in a 53-Year-Old Woman With Low Back Pain

Federico E. Villafañe, DC^{a,*}, Alicia M. Yochum, RN, DC, DACBR^a,
Aimee R. Jokerst, DC^b, Norman W. Kettner, DC, DACBR^c

^a Diagnostic Imaging Resident, Department of Radiology, Logan University, Chesterfield, MO

^b Clinician, Department of Chiropractic Health Centers, Logan University, Chesterfield, MO

^c Chair, Department of Radiology, Logan University, Chesterfield, MO

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Abstract

Objective: The purpose of this case report is to describe the features of an unusual presentation of fibrous dysplasia.

Clinical Features: A 53-year-old woman had low back pain and numbness in the anterior upper left thigh that started 3 years earlier after a fall. She experienced pain during active lumbar flexion and extension range of motion. Radiographic examination demonstrated an oval geographic osteolytic lesion in the left ilium and abnormal trabecular architecture with variable-sized osteolytic lesions and both ill-defined and well-defined borders along the sacroiliac joint margin.

Intervention and Outcome: Because of the aggressive osteolytic appearance, magnetic resonance imaging of the pelvis with gadolinium contrast was obtained for additional characterization of the lesions. There were 3 additional mixed signal lesions located within the left femoral neck and extending to the greater trochanter that enhanced with contrast. Because of the suspicion of malignancy, needle biopsy was performed. The pathologic findings in combination with the radiographic appearance confirmed the diagnosis of polyostotic fibrous dysplasia.

Conclusion: This case demonstrated a rare aggressive appearance of polyostotic fibrous dysplasia located in the left innominate and the left proximal femur that prompted a diagnostic imaging workup and biopsy for suspected skeletal malignancy. These lesions may require careful evaluation by an experienced team of physicians, radiologists, and pathologists to ensure proper diagnosis and treatment.

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* Corresponding author at: 1851 Schoettler Rd, Chesterfield, MO 63017. Tel.: +1 636 230 1831; fax: +1 636 207 2429.

E-mail address: federico.villafane@logan.edu (F. E. Villafañe).

Introduction

Pain in the lower back is multifactorial and can be caused by many different conditions, most of which are benign, but some are life-threatening.¹ Estimates of the 1-year incidence of low back pain range between 6.3% and 15.4%,² and the lifetime prevalence of low back pain is reported to be as high as 84%.³ Diagnostic workup of a patient with lower back pain may include a variety of imaging modalities, usually initiated by radiography and, if inconclusive, followed by advanced imaging modalities.¹ The choice of which imaging modality to use generally depends on the patient's differential diagnosis.¹

Fibrous dysplasia (FD) of bone is a skeletal disorder characterized by extensive proliferation of fibrous tissue in bone marrow, leading to osteolytic lesion, fracture, and deformity.⁴ Fibrous dysplasia may affect a single bone (monostotic), numerous bones (polyostotic), or the skull and facial bones (craniofacial). The true incidence and prevalence of FD are difficult to estimate, but the lesions are not rare; they are reported to represent approximately 5% to 7% of benign bone tumors. The clinical presentation of pain occurs more in adults than in children.⁵ When bone deformities involve the facial or the cranial bones, neurological compromise may be present. Polyostotic FD most commonly involves the femur, skull, tibia, humerus, ribs, fibula, radius, and ulna.⁶ FD has great variability in appearance and has been referred to as one of the "great imitators of bone disease."⁶ This makes the potential list of radiographic differential diagnoses extensive. Malignant transformation occurs in 0.5% of cases.⁶ The purpose of this case study is to report the findings of a rare aggressive appearance of polyostotic FD located in the left innominate and the left proximal femur that prompted a diagnostic imaging workup and biopsy for suspected skeletal malignancy.

Case Report

A 53-year-old woman experienced low back pain and numbness in the anterior upper left thigh. The pain started 3 years earlier after a fall on ice. She localized her pain to the left sacroiliac joint and rated it a 4 on a numerical pain scale of 0-10 with 10 being the worst pain experienced. The pain was described as dull, getting worse, and present most of the day. She denied recent fever, infection, weight loss, or significant illness.

The physical examination produced pain on active lumbar range of motion while flexing and extending.

All lower extremity myotomes were 5 of 5, and heel walk and toe walk were negative. Standing Kemps elicited pain at the left sacroiliac region. The remainder of the orthopedic examination was negative. Light palpation of the lower lumbar spine and left sacroiliac region caused extreme discomfort.

Lumbar spine radiography (posterior-anterior, lateral, and Ferguson) was performed at the chiropractic clinic to evaluate for degenerative or inflammatory arthropathy of the left sacroiliac joint. The radiographic examination (Figs 1 and 2) demonstrated an oval geographic osteolytic lesion superior to the left acetabular cortex measuring 2.2 cm wide $\times$ 2.5 cm in the craniocaudal dimension. The left ilium, along the sacroiliac joint margin, demonstrated abnormal trabecular architecture with variable-sized osteolytic lesions with both ill-defined and well-defined borders. Because of the aggressive osteolytic appearance, the differential diagnosis included primary and metastatic infiltrative marrow lesions of the left innominate. Chiropractic manipulation was deferred until additional diagnostic workup was completed.

Magnetic resonance imaging (MRI) of the pelvis (Figs 3-5) with intravenous gadolinium contrast admin-

Fig 1. Anteroposterior lumbar radiograph demonstrating an oval geographic osteolytic lesion (white arrow) superior to the left acetabular cortex measuring 2.2 cm wide $\times$ 2.5 cm in the craniocaudal dimension.

Fig 2. Anteroposterior spot radiograph of the left sacroiliac joint. The left ilium, along the sacroiliac joint margin, had abnormal trabecular architecture with variable-sized osteolytic lesions (white arrow).

istration was obtained 4 days after the radiography at an outpatient imaging center. The examination demonstrated multiple nonspecific infiltrative lesions within the marrow of the left posterior superior iliac spine extending inferiorly to the superior acetabular region. The T-1 and T-2 signal intensity was mixed but predominantly high on T-2 and low on T-1 with some areas of high signal on T-1. The T-1 high signal likely represented areas of hemorrhage, protein, or melanin. These lesions demonstrated heterogeneous contrast enhancement. The lesion adjacent

Fig 3. Coronal T2-weighted MRI of the pelvis showing a lesion (white arrow) with mixed signal intensities, predominantly high at the left supra-acetabular zone.

Fig 4. Coronal T2-weighted MRI of the pelvis demonstrating a lesion (white arrow) with mixed signal intensities, predominantly high at the medial aspect of the left iliac bordering the left sacroiliac joint.

to the sacroiliac joint was the largest, measuring 7.7 cm in the craniocaudal dimension. The superior acetabular lesion measured 2.2 cm wide $\times$ 2.5 cm in the craniocaudal dimension. Slight osseous expansion was present in the affected areas, most pronounced in the posterior superior iliac spine. There was suspected cortical disruption at the anterior portion of the ilium near the iliac spine and the articular surface of the sacroiliac joint. There were 3 additional mixed signal lesions that also enhanced within the left femoral neck extending to the greater trochanter. These lesions were likely related to the innominate lesions, but an isolated process could not be excluded.

Needle biopsy of the left ilium was performed by an orthopedic surgeon in a hospital setting. The biopsy was obtained 6 days after MRI examination. The pathologic specimen revealed no mitotic activity and demonstrated a spindle cell lesion with extensive collagenous stroma. The cells were widely spaced within the collagenous stroma, very bland, and ovoid to fusiform in shape. Focally, the cells showed evidence of bone formation with small pieces of woven bone. It was determined that the pathologic findings in combination with the radiographic appearance confirmed the diagnosis of polyostotic FD. The patient was advised to follow up care with her oncologist every 6 months; however, the patient did not initiate treatment. The patient gave consent for the publication of this report.

Discussion

Fibrous dysplasia is primarily a pediatric disease, although it often progresses unnoticed until middle age.

Fig 5. Coronal T-1 postcontrast MRI of the pelvis showing a lesion (white arrow) with mixed signal intensities of the left proximal femur.

In this case, FD was subclinical until adulthood. This bone disease may be monostotic (affecting a single bone) or polyostotic (affecting many bones). When the process is polyostotic, it usually presents asymmetrically and often unilaterally.⁷ The polyostotic form is less common. Patients with polyostotic disease and large, painful lesions in weight-bearing long bones are at risk for pathologic fractures. These patients should be evaluated to determine the appropriateness of biopsy and prophylactic fixation of the involved bone. The risk for fracture is increased by the number of lesions and type, size, extent, and anatomical site of the lesions.⁸ McCune-Albright and Mazabraud syndromes can occur in patients with FD. Patients with McCune-Albright syndrome demonstrate café au lait spots and endocrine disorders, whereas patients with Mazabraud syndrome have soft tissue myxomas.⁹ Malignant degeneration complicates less than 1% of FD cases, most frequently resulting in osteosarcoma, fibrosarcoma, malignant fibrous histiocytoma (pleomorphic undifferentiated sarcoma), or chondrosarcoma.¹⁰

Radiographic findings of polyostotic FD frequently include osteolytic expansile lesions that may exhibit endosteal scalloping. The replacement of medullary bone by connective tissue and disseminated dysplastic spicules of bone leads to the presence of a sign described as the “ground glass” appearance. MRI is unhelpful in differentiating FD from other entities

because of variability in the appearance of the bone lesions, as they can often resemble more aggressive lesions, and it tends to demonstrate intermediate heterogeneous signal intensities in T-1 and low heterogeneous signal usually, but may have regions of higher signal on T-2.¹¹ The hypointense fibrous areas show moderate-to-marked enhancement following intravenous administration of gadolinium as a result of the multiple small vessels within this component of the lesion.⁷ This enhancement makes it more confusing to differentiate. There may be some heterogeneity due to secondary areas of degenerative cyst and/or areas of hemorrhage. Secondary aneurysmal bone cyst is not a true neoplasm and may be a benign vascular lesion resulting from hemodynamic changes. The exact etiology is unknown and controversial with an appearance of a blood-filled, expansile, spongelike tumor containing numerous giant cells.¹²

This case of polyostotic FD was unique and difficult to differentiate from disseminated bone metastasis because of its aggressive appearance. Typically, the appearance of FD is benign in nature and exhibits a thick, sclerotic border (rind of sclerosis) which includes a benign differential diagnosis (Paget disease, neurofibromatosis type 1, osteofibrous dysplasia, and adamantinoma). Although FD is considered a “great imitator of bone,”⁶ it tends to simulate benign lytic lesions, and yet in this particular case, FD mimicked an

aggressive neoplasm of bone. Even when combined with a patient's medical history and multiple imaging techniques, the clinical diagnosis of metastatic lesions may remain a challenge. The clinical characteristics of polyostotic FD and malignant bone metastasis in multiple sites are very similar because they can both produce pain. Consequently, differentiating between the 2 is important when determining the appropriate therapy.⁸ Very few cases of FD require biopsy to distinguish from other aggressive neoplasms unless the archetypal characteristics can be detected from plain films. Skeletal scintigraphy may assist with the diagnosis because it displays increased isotope uptake throughout life, but the uptake becomes less intense as the lesions mature.⁷ Some characteristic bone scan findings of fibrous dysplasia lesions are a bar-shaped pattern, whole bone involvement, and a close match between the lesion size on radiographs and the size of the area of uptake.⁵ In this case, the FD appearance did not follow the classical presentation.

Aggressive-looking osteolytic lesions located within both the innominate and proximal femur may simulate FD. These include metastatic bone disease of unknown origin, primary bone neoplasms such as multiple myeloma, and atypical nonmineralized chondrosarcoma.

Metastatic bone disease is highly variable and influenced by a number of factors, including the nature of the primary tumor, patient's age, location, and the timing of the radiographic examination. Osseous metastasis generally appears as osteolytic lesions in multiple locations, being most common in the vertebral column, ribs, and pelvis, in descending order.¹³

Multiple myeloma, a primary bone marrow neoplasm, demonstrates radiographic findings of osteolytic lesions with discrete margins that appear uniform in size known as "punched out lesions." Estimates indicate that 75% of multiple myeloma patients will have positive radiographic findings. Typically, these osteolytic lesions may be in multiple locations, affecting the spine, pelvis, skull, and proximal appendages.¹⁴

The most important radiographic characteristic of chondrosarcoma is matrix calcifications described as ring-and-arc or popcorn calcification and is seen in up to 60%-78%. Chondrosarcoma is also associated with large soft tissue masses, the majority being greater than 4 cm in size.¹⁵ It is well recognized that a relationship exists between the pattern and density of calcification in chondrosarcoma and degree of malignancy. Well-organized calcific rings within cartilage usually signify a low-grade tumor. High-grade chondrosarcomas, which may contain a greater amount of myxoid

material, are frequently associated with large areas of noncalcified tumor matrix.¹⁵

All entities that were included in our patient's differential diagnosis were difficult to rule out using radiography or MRI. Biopsy was necessary as the last diagnostic step because of the imaging similarities (low specificity) in this case. All physicians involved in the workup of this patient were considering a malignant diagnosis, but the biopsy results confirmed the diagnosis of polyostotic fibrous dysplasia. The patient was recommended to undergo evaluations at 6-month intervals to monitor complications such as acetabular protrusion (telescoping of the femur head into the acetabulum) and rare malignant transformation.

Limitations

The limitations of this case report are its inability to be generalized with respect to diagnosis or treatment across all FD patients. In a patient within this age group, fibrous dysplasia should be considered as a benign condition in the differential diagnosis. Unfortunately, the patient declined to follow up.

Conclusion

This case report illustrates several elements related to FD. Fibrous dysplasia could go unnoticed, become symptomatic at later age, be polyostotic, and on rare occasions simulate a malignant skeletal neoplasm. These lesions may therefore require careful evaluation by an experienced team of clinicians, radiologists, and pathologists to ensure proper diagnosis and optimal treatment.

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