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Left Upper Femur Fibrous Dysplasia

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Review Article

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Abstract: A 17 years old male patient came to Alzaytouna Specialist Hospital (Sudan) with vague complaints of left upper femur pain for ~2 years, difficulty with daily living activities. The doctor requested x-ray, CT, and then MRI.
Keywords: FD, X-RAY, CT, MRI.

INTRODUCTION

Fibrous dysplasia is a non-neoplastic developmental anomaly of bone in which normal bone marrow is replaced by fibro-osseous tissue [1, 2]. This condition was first described in 1942 by Lichtenstein and Jaffe [3]; hence, fibrous dysplasia is sometimes referred to as Lichtenstein-Jaffe disease. FD has a varied radiographic appearance. If they are asymptomatic, they do not require treatment.

The disease process may be localized to:

- A single bone (monostotic fibrous dysplasia)
- Multiple bones (polyostotic fibrous dysplasia) [4]

Polyostotic fibrous dysplasia can occur as a part of McCune-Albright syndrome (unilateral polyostotic fibrous dysplasia, ipsilateral café-au-lait spots on the skin, and endocrine disturbances such as precocious puberty) or Mazabraud syndrome (polyostotic fibrous dysplasia and soft-tissue myxomas).

Fibrous dysplasia has also been reported in association with other endocrine dysfunctions [4], such as hyperthyroidism, hyperparathyroidism, acromegaly, diabetes mellitus, and Cushing syndrome.

Epidemiology

Fibrous dysplasia represents about 5% of benign bone lesions [3]; however, the true incidence is unknown, as many patients are asymptomatic. Monostotic fibrous dysplasia accounts for 75-80% of the cases. Fibrous dysplasia is a slowly growing lesion that usually appears during periods of bone growth and is thus seen in those in early teen and adolescent years. Polyostotic fibrous dysplasia accounts for 20-25% of cases, and patients tend to present at a slightly earlier age (mean age, 8 yrs) [5].

Clinical Features

In the first type monostotic fibrous dysplasia the most common sites involvement are the proximal femur, and craniofacial bones, typically the posterior maxilla [4, 5]. The lesion may involve only a small segment of bone or it may occupy its entire length.

In the second type polyostotic fibrous dysplasia, the spectrum of involvement varies from two bones to more than 75% of the skeleton. Polyostotic fibrous dysplasia is most commonly found in the femur, tibia, pelvis, and foot. Other sites less commonly affected include the ribs, skull, and bones of the upper extremity. Uncommonly affected bones include the lumbar spine, clavicle, and the cervical spine.

Deformity and fracture

Fracture is the most common complication in fibrous dysplasia [6]. It is seen in more than half of the patients with the polyostotic form of the disease. Deformities in weight-bearing bones can occur. Almost 75% of patients with polyostotic fibrous dysplasia are symptomatic, with pain, deformity, or pathologic fractures [3].

Radiographic staging of bone involvement can be found in imaging studies below:
Fibrous Dysplasia on X-ray

On plain films, fibrous dysplasia is an intramedullary, expansile, and well-defined lesion in the diaphysis or metaphysis. The lesions can vary from completely radiolucent to completely sclerotic; however, most lesions have a characteristic hazy ground-glass appearance [8].

Fibrous Dysplasia on Computed Tomography CT.

The extent of the lesion is best demonstrated on Computed Tomography (CT) scans. This imaging modality is helpful in distinguishing fibrous dysplasia from other lesions in the differential diagnosis as follows:-

- Similar to findings on plain films
- Lesions can be radiolucent or radio opaque
- Confirms extent, specific dimensions and radio density of FD with a high degree of precision. So CT scanning is not optimal for the differentiation of fibrous dysplasia from other lesions that mimic it. CT findings complement plain radiographic findings .Usually, attenuation is in the range of 70-130 HU (Hounsfield unit) [10].

Fibrous Dysplasia on MRI

Magnetic Resonance Imaging (MRI) is a sensitive means of establishing the lesion's shape and content [7]. Because fibrous dysplasia is composed mainly of fibrous tissue and bone, T1-weighted images have a low-intensity signal [6]. T2-weighted images have a higher intensity signal that is not as bright as the signal of malignant tissue, fat, or fluid [9].

OnT1, lesions are largely isointense to skeletal muscle with areas of hypointensity. OnT2, lesions appear heterogeneously hyperintense with hypointense, isointense or markedly hyperintenseares within (Fig -3, 4, 5, 6).

Fibrous Dysplasia on Nuclear Medicine (NM)

In fibrous dysplasia, accumulation of isotope increases because of the lesion's hypervascularity. Hot spots or increased uptake of the radioisotope tracer technetium-99m methylene diphosphonate (^{99m}Tc MDP) occurs in the spine, pelvis, ribs, and appendicular skeleton. Pathologic or stress fractures also can increase isotopic activity in the lesions. The features on the bone scan are nonspecific for a conclusive diagnosis based solely on the distribution of the isotope [11, 12].

CASE REPORT

A 17 years old male patient came to Alzaytouna Specialist Hospital (SUDAN) with pain in the left hip.The doctor requested x-ray, CT, and then MRI.

Radiographic staging of bone involvement can be found in Imaging Studies below:

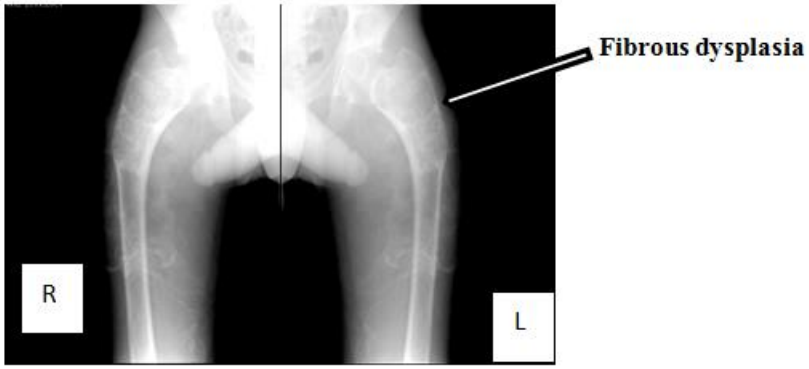

Fig-1: Image for both hip joints & upper femurs x-rays show fibrous dysplasia (arrow).

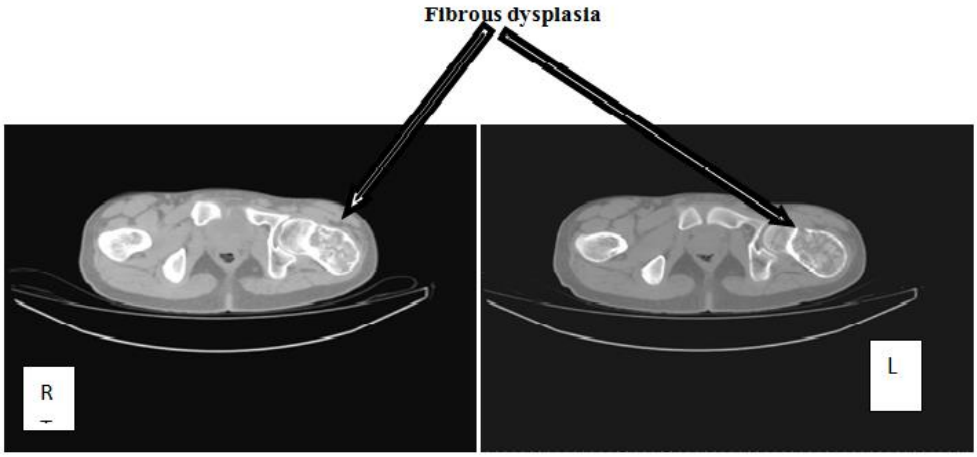

Fig-2: Two axial CT images of the upper left upper femur with different CT numbers (Dual window) show fibrous dysplasia (arrows).

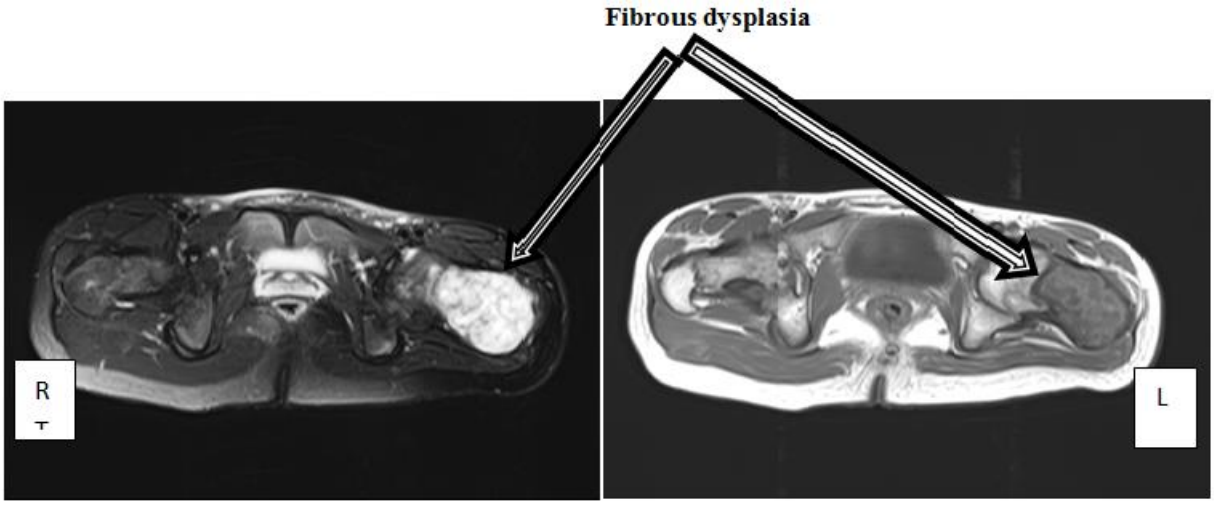

Fig-3: MRI axial T2 imagehave a higher intensity signal showsa heterogeneous high signal within a fibrous dysplasia in the proximal shaft of the left upper femur. (arrow)

Fig-4: MRI axial T1 imagehas low-intensity signal showsthe shaft of theleft upperfemur in a patient with fibrous dysplasia. (arrow)

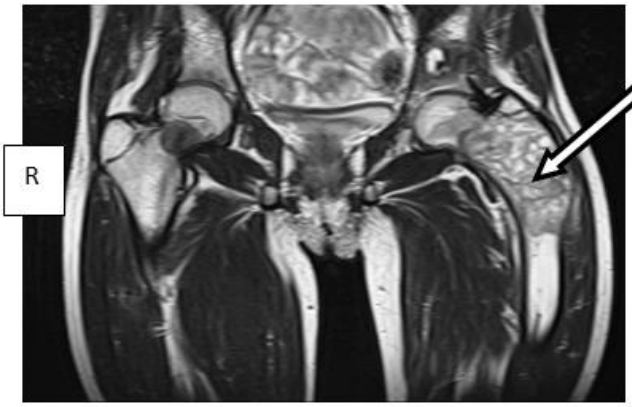

Fig-5: MRI coronal T1 imageofleft upperfemur shows fibrous dysplasia (arrow)

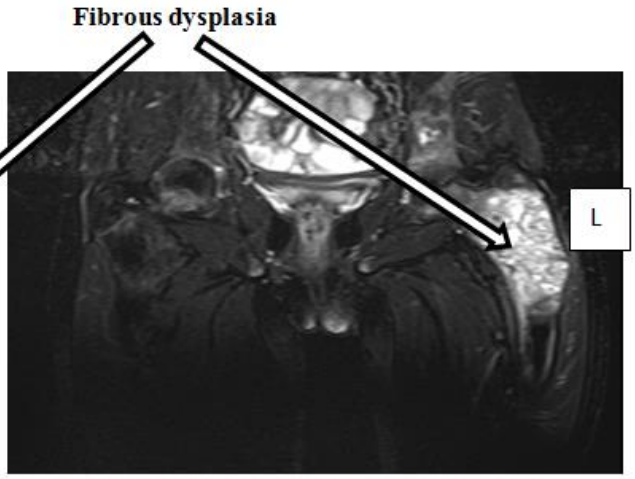

Fig-6: MRI coronal T2 image of theleft upperfemurhave a higher intensity signal shows fibrous dysplasia (arrow).

DISCUSSION

Plain radiography is the first-line study. Usually, the diagnosis is straight forward when typical features are present Fig-1.

Computed Tomography (CT) is extremely useful in evaluating the extent of disease in complex locations, such as the facial bones, pelvis, chest wall, and spine. The modality demonstrates the nature of the lesion better by characterizing the matrix of the lesion. It also depicts expansion of the affected bone and its subtle mineral contents Fig-2.

Magnetic Resonance Imaging features:-

On T1-weighted MRIs, the lesion has low-to-intermediate signal intensity equal to that of muscle. T2-weighted images also show low signal intensity owing to the high content of collagen and bone.

T2-weighted axial image showing a heterogeneous high signal within a fibrous dysplasia in the proximal shaft of the left upper femur.

MRI may be useful in assessing malignant change and demonstrating extension of the tumor into the surrounding soft tissues (Fig- 3, 4, 5, 6).

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