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Dr. Sachin Khanduri
Era's Lucknow Medical College
and Hospital, Era University,
Lucknow, India

Dr. Avani Kanojia
Era's Lucknow Medical College
and Hospital, Era University,
Lucknow, India

Dr. Aniket Chugh
Era's Lucknow Medical College
and Hospital, Era University,
Lucknow, India

Dr. K Prithvi Perumal
Era's Lucknow Medical College
and Hospital, Era University,
Lucknow, India

Corresponding Author:
Dr. Avani Kanojia
Era's Lucknow Medical College
and Hospital, Era University,
Lucknow, India

Polyostotic fibrous dysplasia: A case-based discussion

Sachin Khanduri, Avani Kanojia, Aniket Chugh and K Prithvi Perumal

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Abstract

Fibrous dysplasia (FD) is a rare, benign skeletal disorder characterized by the replacement of normal bone with fibrous tissue, leading to structural weakness, deformity, and potential complications. It results from a postzygotic activating mutation in the GNAS1 gene, affecting osteoblastic differentiation and bone remodeling. FD can present as monostotic (affecting a single bone) or polyostotic (involving multiple bones) and is often discovered incidentally on imaging. Radiologically, FD exhibits characteristic ground-glass opacity, cortical thinning, and bone expansion. In long bones, it may cause deformities such as the classic "shepherd's crook" deformity. Computed tomography (CT) and magnetic resonance imaging (MRI) aid in delineating lesion extent and assessing complications like fractures or malignant transformation, though the latter is rare. Clinically, FD remains asymptomatic in many cases but can present with pain, pathological fractures, or skeletal deformities. In syndromic forms, such as McCune-Albright syndrome, it is associated with endocrine dysfunction and café-au-lait skin pigmentation. Management is largely conservative, with bisphosphonates offering symptomatic relief, while surgical intervention is reserved for severe deformities or complications. This case report highlights the radiological findings and clinical implications of fibrous dysplasia, emphasizing the importance of imaging in diagnosis, differentiation from other bone lesions, and guiding appropriate management.

Keywords: Fibrous dysplasia, Polyostotic, ground-glass appearance, shepherd's crook deformity

Introduction

Fibrous dysplasia is a benign fibro-osseous disorder that can manifest as either a monostotic or polyostotic condition^[1, 2]. The monostotic form is the most common type of fibrous dysplasia, accounting for approximately 75% of cases. It primarily affects the craniofacial bones, followed by the ribs, femurs, tibias, and humerus in decreasing order of frequency. Patients, typically between 10 and 30 years of age, may present with pain or pathological fractures. Compared to the polyostotic form, bone deformities in monostotic FD tend to be less severe. There is no documented evidence supporting the progression of monostotic FD to the polyostotic form^[1, 3].

Fibrous dysplasia (FD) presents with a range of clinical manifestations, including persistent bone pain, an increased susceptibility to pathological fractures, and varying degrees of bone deformities. Pain may result from bone expansion, microfractures, or associated secondary changes such as cyst formation or nerve compression. Pathological fractures are common in weight-bearing bones, often occurring with minimal trauma due to the weakened bone structure. Bone deformities, which can range from mild asymmetry to severe skeletal abnormalities, are more pronounced in polyostotic cases and may lead to functional impairments or cosmetic concerns. The severity and presentation of symptoms depend on the extent and location of the affected bones^[4]. The radiological appearance of fibrous dysplasia (FD) is highly variable and is influenced by the relative composition of mineralized bone and fibrous tissue within the lesion. The imaging characteristics range from predominantly radiolucent areas, indicative of a higher fibrous tissue content, to more sclerotic regions where mineralized bone is more abundant. This variability in mineralization affects the lesion's density and overall radiographic presentation.

In its classical form, FD exhibits a "ground-glass" opacity on radiographs and CT scans, reflecting a mixture of woven bone and fibrous matrix. Lesions may also appear purely lytic, mixed, or sclerotic, depending on the stage of the disease and the degree of bone remodelling. Cortical thinning, bone expansion, and endosteal scalloping are frequently

observed, particularly in weight-bearing bones. Advanced imaging techniques, such as MRI, typically show lesions as hypointense on T1-weighted images and hyperintense on T2-weighted sequences, corresponding to the fibrous component. Additionally, bone scintigraphy can reveal increased radiotracer uptake, highlighting the metabolic activity of the lesion. These diverse radiological manifestations underscore the importance of imaging in diagnosing and differentiating FD from other bone lesions, guiding clinical management, and assessing disease progression

Case Report

A 19-year-old female presented to the orthopedic outpatient department with complaints of dull, intermittent pain in her bilateral femur for the past one year. There was no history of trauma, fever, weight loss, or systemic illness. On examination, there was mild tenderness over bilateral femur without swelling or deformity. No neurovascular deficits were noted. Radiographs of the bilateral femur revealed a multiple well-defined, ground-glass opacities involving the diaphysis. There was evidence of cortical breakthrough at the proximal diaphysis of left femur. A well-demarcated lesion with a thick sclerotic border is seen in the proximal left femur- "Rind Sign". The rind sign represents a reactive bone formation at the periphery of the lesion and helps differentiate fibrous dysplasia from other lytic bone lesions. It is a hallmark feature on radiographs and CT scans, signifying the slow-growing and benign nature of the disease. On MRI, the lesions appeared hypointense on T1-weighted sequences due to the replacement of normal bone marrow with fibrous tissue. On T2-weighted sequences, it demonstrated a hyperintense signal, reflecting the increased water content within the fibrous stroma. Post-contrast images showed variable enhancement, often mild to moderate, corresponding to fibro-osseous proliferation. CT confirmed a characteristic ground-glass matrix. Laboratory investigations, including serum calcium, phosphorus, and alkaline phosphatase, were within normal limits.



Fig 1: T2 Coronal Image of Bilateral Hip showing multiple hyperintense cystic lesions in bilateral femur.

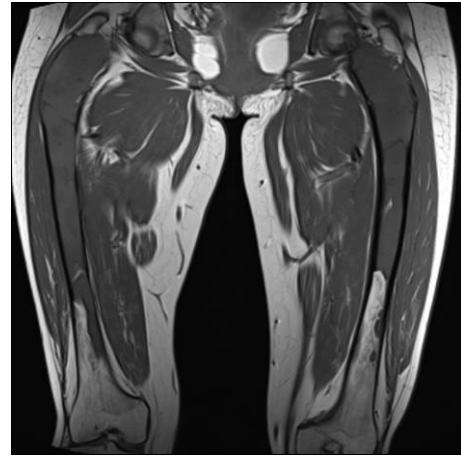


Fig 2: T1 Coronal Image of Bilateral Hip showing hypointense lesions in bilateral femur.



Fig 3: On Radiograph- AP view, A well-defined lytic lesion with a sclerotic rim - RIND SIGN (yellow arrow) is seen in the left femoral neck.

Discussion

PF is a complex disorder with significant clinical and radiological variability. Early diagnosis and appropriate management can help improve patient outcomes and quality of life. A multidisciplinary approach, involving radiologists, orthopedic surgeons, and endocrinologists, is essential for optimal care. Fibrous dysplasia should be considered in young patients presenting with localized bone pain and characteristic imaging findings. Conservative management is usually sufficient, but surgical intervention may be necessary in cases of functional impairment or fractures. Regular follow-up is crucial for monitoring disease progression.

Conflict of Interest

Not available

Financial Support

Not available

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