UNUSUAL RADIOLOGICAL PRESENTATION OF FIBROUS DYSPLASIA OF TIBIA – A CASE REPORT

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ABSTRACT: Fibrous dysplasia is a benign fibro-osseous lesion that can occur as an isolated skeletal lesion (monostotic form) or affect multiple skeletal sites (polyostotic form). The monostotic type is 30 times more common than the polyostotic type. [4] However, this is a very rare lesion, making a correct diagnosis difficult, especially since osteofibrous dysplasia and adamantinoma of the tibia have to be taken into account in the differential diagnosis of the disease. Here is an unusual case of aggressive monostotic fibrous dysplasia of tibia. We say this case is a rare presentation as the appearance was atypical and did not correspond to classical appearance of fibrous dysplasia and was subsequently proved histopathologically.

KEYWORDS: Fibrous dysplasia, fibro-osseous lesions, osteofibrous dysplasia

INTRODUCTION: Fibrous dysplasia and osteofibrous dysplasia are both benign fibro-osseous lesions of the bone and are generally seen during childhood or adolescence. Fibrous dysplasia (FD) is a benign skeletal disorder, first described by Lichtenstein, in which abnormal development of fibroblast replaces medullary bone with fibrocellular tissue [1, 2, 3]. The lesion usually involves the diaphyseal region and also may involve the metaphyseal region; the epiphyseal area is rarely involved. The lesion typically is well defined and has a “ground-glass” appearance caused by the finely scattered bone trabeculae within the tissue. There is medullary expansion of bone with surrounding cortical bone thickening, and a layer of reactive cortical bone on top of normal bone. Clinically, FD is most commonly found in long bones usually without signs and symptoms until the occurrence of pathological fracture [5]. In the long tubular bones, fibrous dysplasia may cause expansion of the bone contour with cortical thinning and endosteal scalloping. Fibrous dysplasia may present with radiographic features that mimic other benign fibro-osseous lesions, be associated with other lesions, or even be confused with certain types of malignancies. Although malignant degeneration of fibrous dysplasia is rare, the presence of cortical destruction and soft-tissue mass should raise suspicion to such a possibility. Many studies have reported the occurrence of malignant transformation of FD. However, it is more commonly found in the polyostotic than the monostotic type [7,8]. So a thorough evaluation of the different imaging modalities particularly plain radiograph and CT is helpful in precise diagnosis of the lesion and to differentiate from other benign fibro-osseous lesions. To confirm the diagnosis of FD, historical, clinical, radiographic and histopathological data are essential [6].

CASE REPORT: A 15 year old male patient presented with chief complaints of pain with swelling right leg since 4 years with past history of trauma. Plain radiograph, CT and MRI were performed. Plain radiograph showed well defined expansile lytic lesion of mixed density, thinned out cortex with narrow zone of transition seen involving proximal, meta-diaphyseal region of tibia, limited
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proximally by epiphysis and the lesion is seen scalloping adjacent proximal third of fibula. CT showed heterogeneous lesion in the proximal one third of tibia with finely scattered trabecular pattern and multiple areas of irregular cortical destruction. MRI showed large expansile heterogenous signal intensity lesion in proximal meta-diaphysis of left tibia involving both cortex and medulla with loss of differentiation. There was evidence of multiple areas of cortical breech, with minimal extension into adjacent soft tissues, with no fluid levels. The lesions were largely isointense with areas of hypointensity on T1 weighted images and appear heterogeneously hyperintense on T2 weighted images.

Considering the benign features of the swelling in spite of huge size it was decided to perform excision of the tumor mass and fill the gap with an autologous bone grafts supplemented by G bone. Histopathological examination revealed proliferating fibroblasts, curvilinear trabeculae of woven bone which lacks osteoblast rimming.

DISCUSSION: Fibrous dysplasia is a non-inherited developmental disorder in which normal bone marrow is replaced by fibro-osseous tissue. It is often asymptomatic and frequently incidentally detected on radiographs taken for unrelated clinical indications. However, it may be complicated by pathological fracture and, rarely, by malignant degeneration.

Radiographs show lesions that are medullary in origin. Sometimes, the lesions may be eccentric in location. Replacement of normal cancellous bone with abnormal tissue produces a purely lytic or denser lesion with ground-glass appearance [9]. The lesions are usually bound by a characteristic thick layer of reactive bone that has been described as a “rind”.

CT may be useful in clearly delineating the lesion [10]. The lesion typically is well defined and has a “ground-glass” appearance caused by the finely scattered bony trabeculae within the tissue. There is medullary expansion of bone with surrounding cortical bone thickening, and a layer of reactive cortical bone on top of normal bone.

The MRI appearance is variable, with a homogeneous intermediate to low signal intensity on T1-weighted images and an intermediate to high signal intensity on T2-weighted images [11]. Although some authors have stated a smooth cortical contour is always maintained [11]. Others classify fibrous dysplasia as a tibial lesion that may cause cortical destruction [12], which is seen in our present case.

Although malignant degeneration of fibrous dysplasia is rare, the presence of cortical destruction and soft-tissue mass should raise suspicion to such a possibility [11]. Fibrous dysplasia may present radiographic features consistent with other benign fibro-osseous lesions of the skeleton, and also may be confused with certain types of malignancies. In this aspect, fibrous dysplasia remains a radiological challenge.

It is believed fibrous dysplasia usually becomes quiescent at puberty and remains so throughout life [13]. Although the clinical presentation, laboratory findings, and imaging studies usually permit the diagnosis of fibrous dysplasia, in certain cases, such as with our patient, this entity may pose a diagnostic challenge.
**Image 1:** Plain radiograph AP and Lateral views showing well defined expansile lytic lesion of mixed density, thinned out cortex with narrow zone of transition involving proximal meta-diaphyseal region of tibia, limited proximally by epiphysis (E) and the lesion is seen scalloping adjacent proximal third of fibula.

**Images 2 and 3:** CT axial sections showing intramedullary, expansile lytic lesion showing varying degrees of hazy density giving a ground-glass appearance with fine trabecular pattern and irregular cortical destruction noted involving the proximal meta-diaphysial region of tibia.

**Image 4 and 5:** Depicting Coronal and sagittal 3D reconstruction CT images.

**Images 6 to 9:** T2WI axial, T2WI sagittal, T1WI sagittal and STIR coronal images respectively showing large expansile heterointense signal intensity lesion seen replacing medullary cavity of the proximal meta-diaphysis of tibia with loss of cortico medullary differentiation.
**Image 10**: Exposed tumour by closed soft tissue dissection

**Image 11**: Seen in the image the medullary portion of the bone was occupied with huge fibrocartilagenous mass without any cavities.

**Image 12**: Excised tumour mass.

**Image 13**: The tumour cavity is filled with fibular and iliac crest graft and hydroxyapatite bone crystals (G bone).

**Image 14**: Post operative radiograph.
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