Osteofibrous Dysplasia – an update

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Abstract

Introduction: Osteofibrous dysplasia (OFD) is a rare, benign, self-limiting, fibro-osseous lesion occurring in long bones especially of lower limbs. Patients typically present with painless swelling with or without anterior bowing of tibia. The diagnosis can be confirmed by peculiar radiological feature of well defined intracortical lytic lesion with variable degree of osteolysis and osteosclerosis. Admantinoma is close differential of this lesion. Most cases regress spontaneously by puberty, surgical intervention is required only for progressive lesions or in case of pathological fracture.

Keywords: Osteofibrous Dysplasia, Management

Introduction

Osteofibrous dysplasia (OFD) is a rare, benign, self-limiting, fibro-osseous lesion occurring in long bones especially of lower limbs. It is also called as Kempson-Campanacci lesion or cortical fibrous dysplasia. The prominence of the osteoblasts led Kempson in 1966 to describe the entity as ossifying fibroma of the long bones [2]. In 1976, Campanacci, gave the term “osteofibrous dysplasia of the tibia and fibula” in reference to its histological features, developmental origin and anatomic location [1].

Etiopathogenesis: Exact etiology is not known. A few of the cases are known to have occurred in families. It has also been reported that OFD may act as a precursor of adamantinoma which is supported by occurrence of OFD like adamantinomas. The evidence for this is limited and most of the cases are considered to be arising spontaneously.

Incidence: OFD is a rare benign self-limiting tumor, which accounts for about 0.2% of all primary bone tumors [3]. These lesions are mainly seen in the first two decades of life. It is very uncommon after skeletal maturity with any gender decades of life. It is very uncommon after skeletal maturity with any gender decades of life. It is very uncommon after skeletal maturity with any gender decades of life. It is very uncommon after skeletal maturity with any gender decades of life. It is very uncommon after skeletal maturity with any gender decades of life. It is very uncommon after skeletal maturity with any gender decades of life.

Site: It is invariably a disorder of the tibia and fibula. The lesion usually has its epicenter in anterior cortex of tibia. Tibial mid-diaphysis and proximal metaphysis are affected the most. Ipsilateral or contra lateral fibula may be involved. Even though most of the lesions are confined to a limited portion of the bone a few may grow rapidly and involve almost entire bone. Isolated fibular involvement is rare. Forearm bones (radius and ulna) are other uncommon sites of affection [5].

Clinical features: The typical presentation is painless swelling with or without anterior bowing of the tibia. Pain is only present in about one third of the cases and is usually due to pathological fracture. About a third of the cases are detected incidentally [6].

Radiological Features:

Radiograph: The lesion appears as a well defined intracortical lytic lesion, with variable degree of osteolysis and osteosclerosis located in the anterior cortex of the tibia. These lesions may present as a single focus or multiple elongated foci interspersed with reactive bone. The overlying cortical shell presents itself in a wavy pattern giving it a “saw tooth appearance”. Most of the lesions are associated with anterior tibial bowing and buttress type of benign periosteal reaction. Aggressive lesions may involve entire diaphysis and metaphysis and may have associated pathological fractures [7, 8].

Computed Tomography: It is helpful in assessing exact extent of the lesion, cortical involvement, periosteal reaction and pathological fractures and acts as an adjuvant to MRI in the overall assessment of the lesion.

Computed Tomography: It is helpful in delineating the cortical based lesion and to assess its medullary or soft tissue extension. The lesion demonstrates mixed signals on T1 and high intensity lesions on T2 weighted images. MRI is helpful in surgical planning and differentiating OFD from adamantinoma [9].

Pathology: On gross examination, a typical specimen appears as a whitish or yellowish solid lesion with surrounding gritty bony architecture. The cortex may be expanded and thinned out deficient at places with intact periosteum. Lesions may show medullary extension, which is usually demarcated by a sclerotic rim [10]. Microscopically, OFD demonstrates a zonal architecture with loose fibrous tissue containing spicules of woven bone in the centre which is lined by a layer of lamellar bone lined by prominent osteoblasts at the periphery. This shows a progressive maturation of the bone trabeculae from a central zone of delicate trabecular bone in a vascular fibrous stroma, to an outer zone of lamellar bone. The fibrous component in most cases contains cells which react positively for pan-cytokeratin. Desmosomes, tonofilaments, and...
microfilaments are seen on electron microscopy [2, 11].

**Differential Diagnosis:**
Several tumor and tumor like lesions can mimic Osteofibrous dysplasias on radiographs [12]. The differential diagnoses are that of a cortical, lytic, expansile lesion. Adamantinoma is the most closest differential diagnosis as both lesions are very similar clinico- radiologically and even on histopathology. Adamantinomas are more aggressive lesions and may lead to local and distant recurrences. These commonly involve the medullary cavity, but there is usually cortical infiltration, break and soft tissue component. Other differentials include Fibrous dysplasia, Nonossifying fibroma, Aneurysmal bone cyst, Chondromyxoid fibroma, Langerhans cell histiocytosis, Osteomyelitis and Hemangioendothelioma [13]. A thorough clinico-pathological correlation substantiated with characteristic radiological findings is very essential for a definitive diagnosis of OFD.

**Treatment:**
According to the case series on OFDs from the Rizzoli Institute in Milan and the Mayo Clinic, these lesions, owing to their benign nature, seldom progress during childhood and undergo spontaneous regression at puberty, thus can be carefully observed with serial plain radiographs and clinical evaluation at regular intervals. If associated with significant or progressive bowing then conservative treatment in the form of bracing may be helpful to minimize deformity and prevent pathological fracture [5, 14]. Surgical intervention is mainly required in extensive cases with progressive deformity or for pathologic fracture. Extraperiosteal “shark-bite” excision is the most widely considered surgical option for OFDs. The resultant defects may be reconstructed with auto or allo-strut grafts. Other surgical interventions may include curettage bone grafting and internal fixation after correction of deformity. [9].

**Prognosis –** OFD has a very good prognosis. Most of the lesion even though they grow in first decade of life get stabilised during the second decade and heal by spontaneous resolution. Deformities may

| Etiopathogenesis | • Incidence: 0.2% of primary bone tumors  • Age Distribution: 0-20 years  • Site: Tibia>Fibula, rarely radius and ulna |
| Clinical Presentation | • Pain  • Tibial bowing with or without pathologic fracture |
| Radiological Features | Radiograph:  • Well defined intracortical lytic lesion, with variable degree of osteolysis and osteosclerosis (“Saw tooth appearance”).  • Anterior tibial bowing /Pathological fractures  • MRI:  • Delineating the cortical based lesion with its medullary or soft tissue extension  • Helps in Surgical planning  • Pathology:  • Whitish or yellowish solid lesion with expanded and thinned out cortex.  • Loose fibrous background containing spicules of woven bony trabeculae lined by lamillated bone.  • Differential Diagnosis:  • Adamantinoma  • Fibrous dysplasia  • Nonossifying fibroma  • Aneurysmal bone cyst  • Chondromyxoid fibroma  • Langerhans cell histiocytosis  • Osteomyelitis  • Hemangioendothelioma  • Treatment:  • Spontaneous regression at puberty.  • Careful observation and plain radiography.  • Surgical: (Progressive lesions or Pathologic Fractures)  • Curettage bone grafting and internal fixation after deformity correction  • Extra-periosteal excision and appropriate reconstruction with auto or allo-strut grafting |
References


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