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Osteofibrous dysplasia of the tibia: A case report

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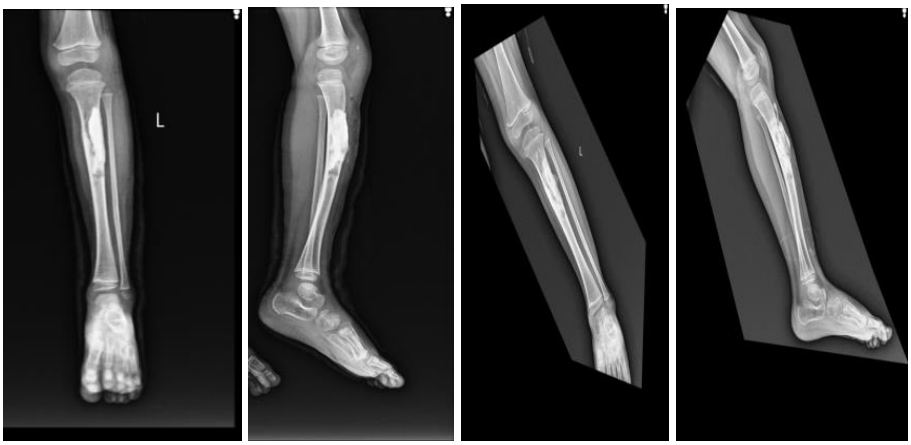
Abstract
Osteofibrous Dysplasia is an uncommon condition of childhood (< 10 years), most often affecting the tibial diaphysis confining to the cortices. Also known as Ossifying fibroma and Campanacci lesion. Histologically and radiologically OFD resembles fibrous dysplasia and adamantinoma respectively and differentiating the conditions might at times become challenging due to their overlapping features. Choosing a right way to manage, also becomes an important aspect in deciding the quality of life post operatively. We describe the case of a 4 year old male with an alleged history of fall which led to an incidental diagnosis of OFD of left Tibia. Radiographs were s/o OFD and we tried to manage it surgically by marginal excision along with application of synthetic bone graft under general anesthesia. Earlier literature and research on OFD was preoccupied with differentiating it from other conditions and whether or not it takes a benign course. But, we would here like to concentrate on various other important aspects like requirement of a high index of suspicion for diagnosis and its diverse management.

Keywords: OFD, tibia, marginal excision, fibrous dysplasia, adamantinoma, age, child

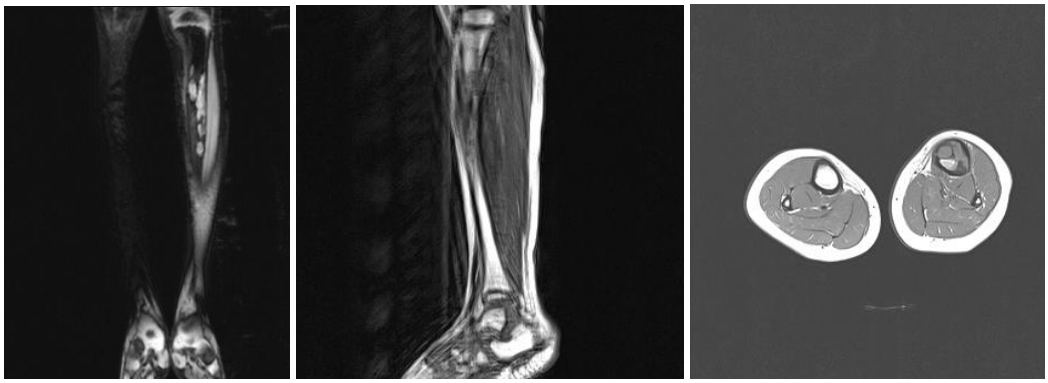
1. Introduction
OFD also known as Campanacci osteitis fibrosa [1]. Various other terms came into limelight which include congenital fibrous dysplasia, ossifying fibroma, congenital fibrous defect of tibia etc. OFD was proposed by Campanacci in 1976 (2,3). OFD is a benign, slow progressive lesion which most commonly localizes to middle 1/3rd of the tibial diaphysial region [4]. Nearly 2/3rd of the lesions are noted in boys before 5 years of age [3]. The pathogenesis of OFD remains unknown [4]. However, various theories have been proposed. OFD is mostly diagnosed incidentally on imaging which shows an extensive lesion, involving the anterior cortex of diaphysis. An eccentric intra cortical osteolysis is found with moderate/ marked expansion of the cortex. In some areas, bubbled/ ground glass appearance is noticed. Usually medial 1/3rd of tibia is involved which results in anterior bowing. However, cases involving distal third fibula have been reported. The two entities that need to be differentiated from OFD are Fibrous Dysplasia and Adamantinoma. However, both these entities are noticed after 10 years of age and our case is of a 4 year old male child which not completely rules out both the differentials but is not worrisome. Treatment generally depends on the course of the lesion. Our main aim should be to correct the deformity if present to increase the quality of life in infants and children.

Case Report
A 4 year old male child presented with complains of pain in the left lower limb since one month. Mother gives an alleged history of fall while running, following which the child developed pain in his left lower limb. On examination, child was playful, cooperative with stable vitals and no other abnormalities. Local examination of left lower limb showed normal gait with small swelling, tenderness and minimal anterior bowing of proximal leg. There was no local rise of temperature and range of movements of left knee, ankle and toe were active. The boy had an uneventful birth history with normal developmental milestones. Radiograph of his left leg showed an incidental tibial diaphyseal lesion with a large focal area of cortical thickening along the proximal aspect of the tibial shaft.



X-Ray Images showing lesion in the left tibial.

On MR imaging, a well defined lobulated eccentric expansile lesion was noted in the mid 1/3 rd shaft of left tibia. The lesion showed isointensity on T1 and heterogeneously hyperintensity on T2. Thin hypointense internal septations were noticed within the lesion on T2. The lesion was diaphysial in location involving the anterior cortex with cortical thickening. No bone destruction, periosteal elevation and marrow signal alterations.



MRI Images showing OFD in the left proximal tibial diaphysis.

On the basis of imaging features a diagnosis of OFD was made. Our aim was to correct the deformity and characterise the lesion histologically, so the child was surgically managed with marginal excision of tumour and filling the void with synthetic bone graft under general anesthesia. Post operative recovery was uneventful. The intra operative curetted sample was sent to histopathological examination, gram staining, acid fast staining, acid fast staining and gram staining showed no inflammatory cells. Histopathological examination revealed multiple fragments of fibro osseous tissue composed of irregular fragments of woven bone which are lined by osteoblasts. The fibrous component composed of spindle shaped cells with bland morphology, with focal areas of myxoid change and extensive areas of thickened collagen and few blood vessels. Histo pathological features confirmed the diagnosis. The child was further followed up regularly with regular imaging.

Discussion

The oldest description of this disease appears to date back to 1921 by Fragenheim [4] who termed it as congenital osteitisfibrosa. Many other terms were coined namely congentinal fibrous dysplasia, congenital defect of tibia and ossifying fibroma. It was in 1976 when Campanacci coined the term OFD, also called as Campanacci disease [3, 5]. OFD has always been described as a variant of fibrous dysplasia in the past [6]. Molecular investigations have depicted that in fibrous dysplasia Gs alpha mutation at Arg 201 is seen which is absent in OFD. This leads to a conclusion that both the diseases have different pathogenesis.

OFD is slightly more common in boys before 5 years of age [3]. Commonest site of occurrence is tibia and ipsilateral fibula. Our case is of a 4 year old male involving the left mid 1/3rd of tibia which is the most common site and classic presentation. However, the child presented to us after a fall and came with complains of leg pain. The diagnosis of OFD was incidental which was initially found as a lesion on Xray. It was later concluded as OFD on MR and histopathological evaluation. Though tibia is the commonest site, there were very few cases reported where OFD arose in other sites**.** Schinichirou *et al* study presented a case report of OFD arising in the humerus in a 34year old male [7]. Also, **Jun** Wen Wang *et al* study reported 4 OFD cases of which 2 had unusual radius and ulna involvement, whereas the 2 had usual tibial involvement [8].

The pathogenesis of OFD remains unknown; however various theories have been proposed- 1. Results from excess resorption of bone with fibrous repair of the defect. 2. It is a variant of fibrous dysplasia [6] 3. Due to abnormal blood circulation in the periosteum. 4. Has a genetic component.

MR imaging is sensitive for assessing the extent and also to asses recurrence post management. The characteristic MRI findings can help make an early non invasive diagnosis of OFD. The lesion is usually extensive, involving anterior cortex of diaphysis of tibia. Charactersistic eccentric intracortical osteolysis is found with expansion of cortex. In our case MRI classically showed a well defined lobulated eccentric expansile lesion in middle 1/3rd shaft of left tibial

diaphysis with no sign of bony defect, periosteal elevation/ no marrow signal alteration, transition zone. The lesion showed T1 isointense, T2 heterogenously hyperintense signal. There were thin T2 hypointense intetnal septations within the lesion. In Paola Simoni *et al* study CT scan confirmed the absence of a transitional zone and periosteal elevation (9). On MR the lesion showed round radiolucent lacunae, low intensity on T1 and T2 with no invasion of soft tissue and normal medullary bone marrow.

On confirmation of the diagnosis radiologically, the child was posted for marginal excision with filling of the void with synthetic bone graft (genex was used). There is no definitive treatment. Treatment usually depends on the course of the specific lesion and to correct the deformity. The most common clinical course is steady growth during the first 10years and then slowing down later with complete halt in expansion. In young children marginal sub periosteal resection/curettage has been reported to be successful but curettage is often followed by recurrence as explained in R.S Lee *et al* study which retrospectively reviewed 16 patients diagnosed with OFD of whom 6 patients presented with recurrence after initial curettage [10].

A wide extraperiosteal en bloc resection will cure but such a radical procedure is not indicated [4]. When deformity is mild-conservative management is preferred/ a minimally invasive osteotomy and plate fixation, an alternative to correct the deformity. In adolescents- conservative management is recommended if the radiograph appearance is unchanged, if any sign of lesion growth marginal examination followed by bone transport through distraction osteogenesis is reported to be successful [4].

In our case after necessary consents were taken, marginal excision was done with synthetic bone grafting under general anesthesia. Bone substitue was preferred to fill the bone to prevent donor site morbidity and to inhibit growing apophysis.

In R.S Lee *et al* study review of 16 patients with OFD- 5 of them underwent excision, 5 of them underwent excision and fibular autografting, 5 of them underwent excision and primary bone transport. 1 underwent proximal tibial replacement, 6 patients presented with recurrence after curettage of whom examination and fibular autografting was done for 3 post recurrence and excision and primary bone transport was done for 2 and proximal tibial replacement was done for 1 post curettage [10].

The squash was sent for HPE after operation. Grossly, on inspection periosteum was intact, cortex thinned, greyish brown fibrous osseous tissue bits. HPE showed fibro osseous tissue composed of irregular fragments of woven bone lined by osteoblasts. The fibrous component composed of spindle shaped cells with bland morphology with focal areas of myxoid change and extensive areas of thickened collagen and blood vessels. Similar histological features were seen in Paola Simoni *et al* study which showed the presence of osteoid tissue, fibrous tissue and small amount of epidermoid cells. The child's post operative status was uneventful with no complications and was followed regularly with repeated MRIs [9].

Differential Dianosis

OFD differs from the more common fibrous dysplasia with regard to age, site, radio features and clinical course. OFD also has histogenetic relationship with Adamantinoma. Considering the age of our patient, we had the differentials at the back of our mind, but primary aim was to correct the deformity and later send the specimen for HPE where we could rule out fibrous dysplasiaas there were osteoblasts-favouring OFD over fibrous dysplasia. Also no nest of epithelial cells ruling out Adamantinoma. However, Immunohisto chemical tests and molecular investigations were not done due to financial constraints.

Table 1: Differential Dianosis

	OFD	Fibrous Dysplsia	Adamantinoma
Age of presentation	Most common <5yrs , also seen in adolescents.	>10 yrs	>10 yrs
Histopathology	Irregular spicules of trabecular bone and fibrous stroma, lined by osteoblasts. Nests of epithelial cells NOT present.	Irregular spicules of trabecular bone and fibrous stroma, NOT lined by osteoblasts	Nests of epithelial cells present
Molecular	Absent Gs alpha mutation	Gs alpha mutation at Arg 201 is present.	Absent Gs alpha mutation
Immuno histo chemistry	Cytokeratin positive cells	Cytokeratin negative cells	Cytokeratin positive cells

Conclusions

OFD is a rare entity. Very few cases have been reported in literature so far. It is an important differential of tibial diaphysial lesion due to its incidental presentation. Marginal excision of lesion and filling with synthetic bone graft, histopathological confirmation of diagnosis and post operative regular followup shows better outcome in our case.

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