Osteofibrous Dysplasia managed with Extraperiosteal excision, Autologous free fibular graft and bone graft substitute

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What to Learn from this Article?

Treatment of Osteofibrous Dysplasia by extraperiosteal excision and autologous fibular bone graft with bone graft substitute.

Abstract

Introduction: Osteofibrous Dysplasia is a rare benign self-limiting fibro-osseous lesion most commonly seen in the diaphysis of the tibia. Its incidence is reported to be 0.2% of all primary bone tumors. It occurs in the first two decades of life with a slight male preponderance. Surgical options include extra periosteal resection, autologous graft, limb lengthening procedures etc. There are no case reports mentioning the use of synthetic bone graft to fill the defect following extraperiosteal excision.

Case Report: A 13 year old girl presented with pain and swelling of the (R) leg since 2 months following a trivial injury at school. Examination revealed a 5×3cm tender swelling on the anteromedial aspect of the middle third tibia. Radiographs and MRI, revealed an eccentric expansile lytic lesion, which was multilocular and was present at the junction of the metaphysis and diaphysis on the antero -medial aspect of tibia. The cortex had ballooned out and there was a possibility of an impending fracture. Biopsy was done which revealed osteofibrous dysplasia. We did an extraperiosteal excision of the lesion. To fill the cavity we harvested 10 cm of the contralateral fibula and since there was still space in the cavity, we packed bone graft substitute (hydroxyapatite crystals) into the defect. The surgical management of osteofibrous dysplasia is controversial. Various methods of treatment of such cases have been described in literature. The use of synthetic graft is an option in these patients as it reduces morbidity; and in our case we had good graft incorporation with this method.

Conclusion: Extraperiosteal Excision of Osteofibrous dysplasia combined with autologous free fibular graft and bone graft substitute is a good surgical option to prevent recurrence and mange bone defects in this rare lesion.

Keywords: Osteofibrous Dysplasia, Autologous free fibular graft, Bone graft substitute.

Author's Photo Gallery


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Introduction

Osteofibrous Dysplasia is a rare benign self-limiting fibro osseous lesion most commonly seen in the diaphysis of the tibia. Its incidence is reported to be 0.2% of all primary bone tumors. It occurs in the first two decades of life with a slight male preponderance. The etiology of osteofibrous dysplasia, as well as the cell of origin, is unknown. Osteofibrous dysplasia has been postulated to arise from a fibrovascular abnormality. It should be differentiated from fibrous dysplasia and adamantinoma of long bone[1]. Due to high incidence (60-100%) of recurrence, any surgical procedure is recommended only after skeletal maturity. Surgical options include extra periosteal resection, autologous graft, limb lengthening procedures etc. We present a surgical technique which we believe has low recurrence rate.

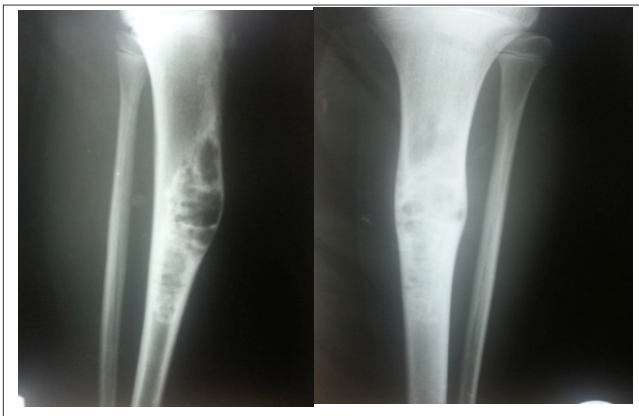

Figure 1: Radiographs of tibia revealed an eccentric expansile lytic lesion, which was multilocular and was present at the junction of the metaphysis and diaphysis on the antero -medial aspect of tibia

Case Report

A 13 year old girl presented with pain and swelling of the (R) leg since 2 months following a trivial injury at school. Pain was aggravated by running and standing for a long duration and was relieved with rest and medication. Swelling has increased over the past 2 months. Examination revealed a 5×3cm swelling on the anteromedial aspect of the middle third tibia. It was tender, non mobile and hard in consistency. Radiographs-revealed an eccentric expansile lytic lesion, which was multilocular and was present at the junction of the metaphysis and diaphysis on the antero -medial aspect of tibia. The cortex had ballooned out and there was a possibility of an impending fracture (Fig1). Blood Investigations reported hemogobin to be 9.3 g% and alkaline phosphatase to be 453U/L, Serum calcium was 8.0 mg/dl and Phosphorous was 5.2mg/dl.

First a biopsy from the lesion was done and greyish white bits of tissue were sent for histopathology. Biopsy reported bony trabeculae interspersed with fibrous tissue, some bony trabeculae showed curvilinear arrangement with osteoblastic rimming confirming the diagnosis of Osteofibrous dysplasia.

After about a month, since the patient still had pain, we planned to do a extraperiosteal excision of the lesion with autologous fibular graft (Fig 2a). Through an anteromedial approach we exposed the lesion, we did an extraperiosteal excision of the lesion (Fig 2b). To fill the cavity we harvested 10 cm of the contra lateral fibula and since there was still space in the cavity, we packed bone graft substitute (hydroxyapatite crystals) into the defect (Fig. 2c). We wanted to avoid harvesting iliac crest bone graft and increasing the morbidity. Post operatively the wound healed and good radiographic filling of the cavity was seen (Fig 2d). Patient was kept non-weight bearing for a month and then started on partial weight bearing mobilization. Two years follow up radiograph showed good healing with no recurrence (Fig 3) and patient was symptom free.

Discussion

Osteofibrous dysplasia (OFD) is a rare benign fibro-osseous lesion, the patient usually presents with swelling of the leg with or without pain, or with anterior bowing. Some authors mention it as a precursor to adamantinoma [2,3] while other authors conclude that it is a variant of fibrous dysplasia[4]. Campanacci in 1976 named this lesion as “osteofibrous dysplasia of the tibia and fibula “in reference to its anatomic location, developmental origin, and histologic resemblance to fibrous dysplasia”[1]. Sakamoto et al. found that GSα mutation at the Arg201 was seen in fibrous dysplasia but this did not occur in Osteofibrous dysplasia, and they concluded that this could be useful for distinguishing between the two lesions [5].

Radiologically the lesion usually involves the anterior cortex and may cause anterior bowing of the tibia [6,7]. It usually manifests

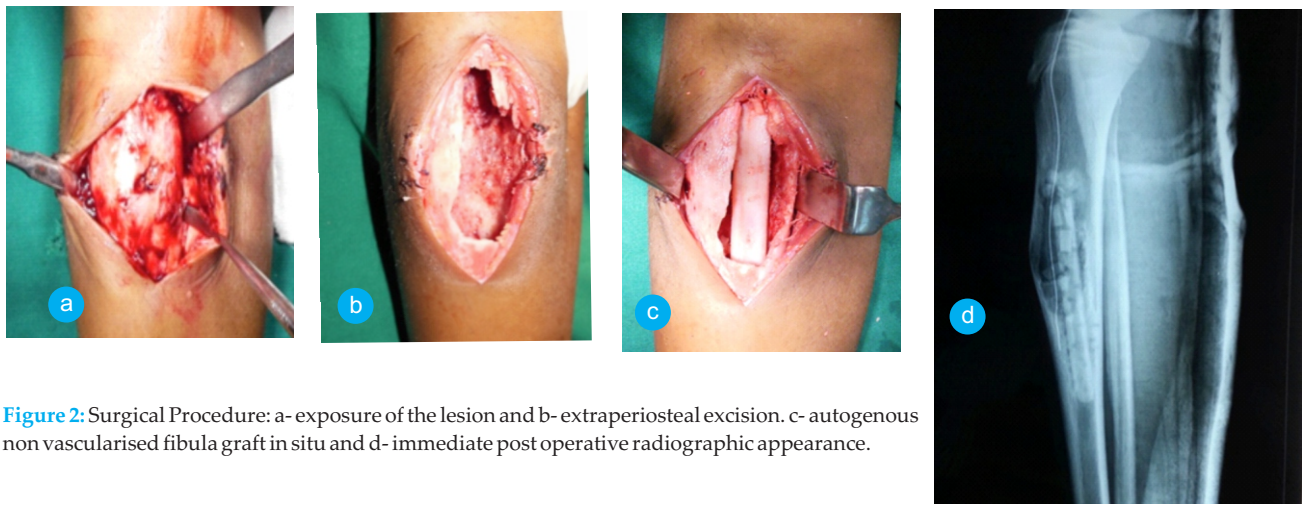

Figure 2: Surgical Procedure: a- exposure of the lesion and b- extraperiosteal excision. c- autogenous non vascularised fibula graft in situ and d- immediate post operative radiographic appearance.

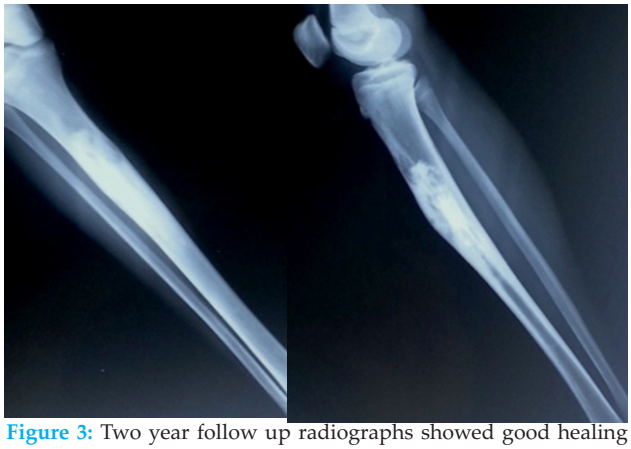

Figure 3: Two year follow up radiographs showed good healing and no recurrence

as an intracortical lytic lesion, which is generally well marginated and is often surrounded by a zone of sclerosis [6,7]. Also multiple lucencies may be present within the cortex, with intervening sclerotic areas. Our patient also had involvement of anterior cortex of the tibia with mild bowing, there were areas of multiple lucencies with intervening sclerotic areas (Fig 1).

Histologically OFD is characterized by a loose, often storiform fibrous background containing spicules of woven bony trabeculae that are lined by a layer of osteoblasts[8,9,10,11]. This histologic appearance is quite similar to that of fibrous dysplasia but fibrous dysplasia typically lacks the distinctive osteoblastic rimming of the bony trabeculae. Also OFD has a zonal architecture, in which more immature woven bony trabeculae are located centrally; however, as we move to the periphery of the lesion, the trabeculae become more numerous, larger, and more mature and lamellar [8,9,10]. A biopsy is very important to establish the diagnosis and differentiate it from early adamantinoma[5].

The surgical management of osteofibrous dysplasia is controversial. Various methods of treatment of such cases have been described in literature. The different options of treatment include, observation without surgical intervention, bracing to prevent fracture and minimize deformity, surgical option which includes en bloc resection, extraperiosteal resection and filling the defect with autogenous fibular graft, vascularized fibular graft or iliac crest bone graft. The high recurrence rate makes this benign tumor difficult to treat. Most authors advocate excision of the tumor after skeletal maturity as it reduces the chance of recurrence.

Campanacci et al.[1] studied 35 patients with this disease and have advocated surgery in patients with extensive disease, but it should be delayed as much as possible. They had good results with surgery even in patients with recurrence. Lee et al.[12] studied 16 patients with osteofibrous dysplasia and have advocated a more aggressive approach for this lesion. 5 patients underwent sharkbite extraperiosteal excision, 6 patients with larger lesions underwent segmental excision and fibula autograft. 4 patients underwent external fixation and bone transport. One patient underwent proximal tibial replacement. He recommended extraperiosteal resection in all cases of osteofibrous dysplasia.

In our patient we did an extraperiosteal shark bite excision of the tumor. Following the excision we had a large defect, so we packed the defect with contralateral autologous free fibular graft. We wanted to avoid harvesting graft from the iliac crest, so we used bone graft substitute to completely fill the defect. Now at 2 year follow up the graft has incorporated well, (Fig.11) the child has no pain and there is no evidence of recurrence. She is attending school and is able to do all her daily activities.

Autologous fibular graft, along with bone graft substitute is a good alternative in treating this benign lesion to fill up the defect that arises after extraperiosteal excision in large lesions. On doing a literature search we could not find any article using synthetic bone grafts along with autologous fibular graft as a mode of treatment of this lesion.

Conclusion

Extraperiosteal Excision of Osteofibrous dysplasia combined with autologous free fibular graft and bone graft substitute is a good surgical option to mange bone defects in this rare lesion. We believe this will reduce the chances of recurrence, however a larger series of cases will be needed to prove this, which would be difficult in this rare disorder.

Clinical Message

Osteofibrous Dysplasia is a rare fibro-osseous lesion. A biopsy must be done to differentiate it from adamantinoma and fibrous dysplasia. After skeletal maturity extraperiosteal excision of the tumor combined with autologous fibular graft and bone graft substitute may be done to prevent recurrence.

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