

RECURRENT FIBROUS DYSPLASIA ON RADIUS TREATED WITH VASCULARIZED FIBULAR GRAFT: A CASE REPORT

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Introduction: Fibrous Dysplasia is a benign intramedullary fibro-osseous lesion that are reported to represent approximately 5%-7% of benign bone tumor. Treatment of fibrous dysplasia include bisphosphonates therapy and surgical procedures.

Case Report: A 9-year-old girl presented with a mass on her left forearm 2 years ago. Primary resection by curettage was performed and patient did not and one year after the surgery, the mass recurred and patient complaint about lump and deformity on her left forearm. A limb salvage surgery by intercalary resection was performed to remove the tumor and the defect was filled with vascularized fibular graft from the right leg. Seven- month follow up show no sign of recurrence and patient underwent surgery to remove the implant.

Discussion: Fibrous dysplasia represents a dysplastic disorder of bone characterized by solitary or multifocal polyostotic intramedullary lesion composed of proliferation of fibroblastic spindle cells. Treatment of fibrous dysplasia include conservative treatment and surgical procedures may be required for correction of deformity, prevention of pathologic fracture and/or eradication of symptomatic lesion. The free vascularized fibular graft is particularly suited for reconstruction of large forearm defects because the fibula matches the radius and ulna in size and shape with studies that shown healing rate of 89%.

Conclusion: Fibrous Dysplasia is a benign intramedullary fibro-osseous lesion that are reported to represent approximately 5%-7% of benign bone tumor. The most common clinical symptom is swelling or deformity of the affected site. Treatment of fibrous dysplasia using vascularized fibular graft may be an effective option.

Keyword: Benign bone tumor, fibrous dysplasia, vascularized fibular graft

Introduction: Fibrous Dysplasia is a benign intramedullary fibro-osseous lesion that are reported to represent approximately 5%-7% of benign bone tumor. Fibrous dysplasia can present in one bone (monostotic) or multiple bones (polyostotic) and can be associated with other condition. Common site of skeletal involvements are long bone, ribs, craniofacial bone and the pelvis. In monostotic fibrous dysplasia, the lesions are usually asymptomatic that most of them are discovered incidentally on radiograph obtained for other reason. The most common clinical symptom is swelling or deformity of the affected site. Proximal femoral shaft is involved in approximately 505 of patients, followed by the tibia (15%), humerus (5%) and radius (5%).(DiCaprio, 2005)

Treatment of fibrous dysplasia include bisphosphonates therapy and surgical procedures may be required for correction of deformity, prevention of pathologic fracture and/or eradication of symptomatic lesion. In upper extremity, the proper treatment of monostotic fibrous dysplasia in long bone depends on presentation. Asymptomatic cases may simply be monitored. When symptomatic, treatment may involve closed fixation, curettage, curettage and bone grafting with more extensive cases may actually require vascularized bone grafting.(DiCaprio, 2005) With lesions in radius treated by closed fixation, recurrence was reported in 2 patients, and after revision surgery with addition of bone grafting recurrence rate of was not reported. In case series of 2 patients, 1 patient showed recurrence of lesions after surgical treatment by curettage.(Ozsen et al., 2018)

A simple curettage is associated with a high risk of recurrence, as is curettage with use of autogenous cancellous bone graft. Cortical graft, persist much longer than do cancellous bone graft. Recurrence of Fibrous Dysplasia was about 13.8% following curettage alone of lesion found in femur, maxilla, calcaneus and mandible. For upper extremity, 6 cases of recurrent lesion on humerus were reported after curettage and bone grafting, but no large scale study of recurrent lesion after surgical treatment on the distal radius was performed to date.(Kumta et al., 2000)

Case Report: A 9-year-old girl presented with a mass on her left forearm 2 years ago. The mass started off as large as marble and progressively enlarged to a size of an egg the following month. Two years ago, patient underwent examination and was found to have fibrous dysplasia. Primary resection by curettage was performed and patient did not have any complaint during the follow up.



Fig 1. Patient's initial clinical presentation.

One year after the surgery, the mass recurred and patient complaint about lump and deformity on her left forearm.



Fig 2. Patient's current clinical presentation

Patient readmitted for surgery after Xray showed lytic lesion on left Radius with dorsal and radial angulation of the bone



Fig 3. Forearm X-ray

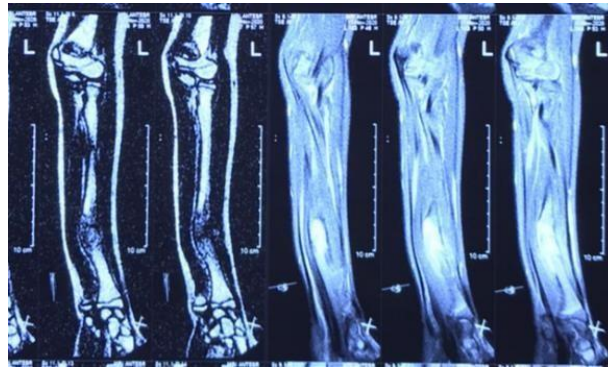


Fig 4. Forearm MRI

A limb salvage surgery by intercalary resection was performed to remove the tumor and the defect was filled with vascularized fibular graft from the right leg.



Fig 5. Resection of the tumor



Fig 6. Harvesting Fibular Graft



Fig 7. Fixation of the graft

Pathologic examination of the tumor was consistent with fibrous dysplasia with connective and osseous tissue. Connective tissue consisting of fibroblast, spindle shaped, soft chromatin and eosinophilic cytoplasm. Osseous component consisting of poorly organized bony trabeculae.



Fig 8. Gross appearance of the tumor.

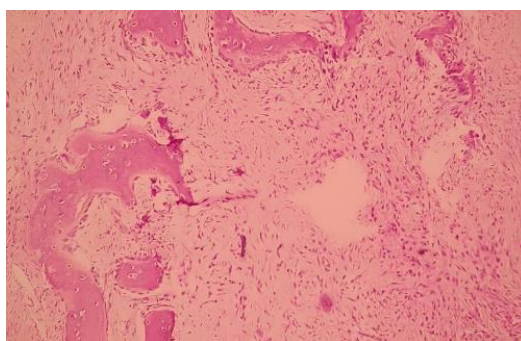


Fig 9. Histopathological examination of the tumor

Four months follow up show a sign of bone healing with callus formation and no sign of tumor recurrence.



Fig 10. 4-month Follow up Xray

The X-ray 3 month afterward show no sign of recurrence and diminished fracture line. Patient underwent surgery to remove the implant. The PEDS SQL score for this patient was 0 in total with no difficulties in activities, feelings, social life and school.



Fig 11. 7-month follow up xray



Fig 12. Xray after removal implant

Discussion: Fibrous dysplasia represents a dysplastic disorder of bone characterized by solitary or multifocal polyostotic intramedullary lesion composed of proliferation of fibroblastic spindle cells with a characteristic whorled pattern in which trabeculae of immature woven bone may be present.(Greenspan et al., 2007; Kokkalis et al., 2010)

On radiograph, fibrous dysplasia appears as a medullary lesion with the ground-glass appearance. The radiographic density of the lesion varies and depend on the relative proportion of bone and fibrous elements. In long bones, fibrous dysplasia may cause expansion of the bone contour with cortical thinning and endosteal scalloping.(Czerniak, 2016)

Fibrous dysplasia is composed of spindle cells that have a whorled or storiform arrangement with interspersed trabeculae of immature woven bone devoid of rimming osteoblast. The number and distribution of bone trabeculae and their level of maturation may vary among different lesion and in different areas of the same lesion(Czerniak, 2016)⁶

Treatment of fibrous dysplasia include conservative treatment, bisphosphonates therapy and surgical procedures may be required for correction of deformity, prevention of pathologic fracture and/or eradication of symptomatic lesion. A simple curettage is associated with a high risk of recurrence, as is curettage with use of autogenous cancellous bone graft. Cortical graft, persist much longer than do cancellous bone graft.(DiCaprio, 2005)

In upper extremity, an axial length>30mm, a circumferential cortical involvement>50% and presence of cystic degeneration was associated with increased risk of fracture and might be considered as indication for prophylactic treatment.(Liu et al., 2018)

In the normal repair of a cortical bone graft, only the bony portion is replaced by dysplastic host bone which involves necrosis of bone graft, resorption and new bone formation. Vascularized bone graft also provides safe and reliable means of ensuring good continuity of bone with little risk of recurrence or failure.(Liu et al., 2018)

The biologic advantages of a vascular bone graft are numerous, including bypasses the sequence of biologic events described above, maintained structural integrity and hypertrophies in response to mechanical load. The diaphyseal graft provides viable tissue and blood flow to incorporate as the graft-host junction by active osteogenesis.(Malizos et al., 2004)

Reconstruction of a segmental bone defect using vascularized fibular graft transfer was developed in the 1970s as microvascular surgical technique evolved. Historically, VFG has been the preferred choice for segmental defects > 10cm. The fibula can be removed as close as 4 cm to the fibular head without compromising the proximal tibiofibular joint and up to 6 cm proximal to the ankle joint without causing ankle instability.(Mauffrey et al., 2015)

The reconstruction of large skeletal defects in children following resection of a bone tumor presents a unique challenge to orthopaedic surgeon including remaining growth potential, the desire for biologic preservation of bone surface and the need for a long-term viable reconstruction in patients who are anticipated to survive for decades. Vascularized fibular grafts are necessary to reconstruct bony defects > 6cm or defects associated with poorly vascularized tissue. The use of free vascularized fibular grafts has been shown to provide biologic reconstruction that successfully addresses these issues in pediatric population.(Ghert et al., 2007)

The application of fibular graft in the reconstruction of bone defects caused by trauma, osteomyelitis or tumor resection represents an effective treatment option. Compared to vascularized fibular grafts, non-vascularized fibular grafts may need a longer time until union and should not be used when there is not a good cover with soft tissue and good blood supply or stabilization of the graft is difficult but non vascularized fibular graft is a simpler, less expensive and a shorter procedure than the use of vascularized graft.(Baghdadi & Arkader, 2020)

The free vascularized fibular graft is particularly suited for reconstruction of large forearm defects because the fibula matches the radius and ulna in size and shape with studies that shown healing rate of 89% and no stress fractures reported. Meta analysis of 110 articles by Feltri *et al* also showed union rate of 80.1%.(Feltri et al., 2021)

In this case, we choose the vascularized fibular graft due to segmental loss of bone after surgery and the length and size about 6 cm that we need, can be fulfilled by vascularized fibular graft.

The fibular graft in our patient was incorporated and no sign for recurrence after 7 months of follow up. This is consistent with a paper by Houdek *et al* which showed that the union of vascularized fibular graft was achieved in 3-14 month with mean of 10 month.(Houdek et al., 2017) Study by Eward *et al* also find that the primary union of vascularized bone graft was attained

in 77% of cases at a mean of 6 month. After treatment with vascularized bone graft on the upper extremity, there was no recurrence of fibrous dysplasia reported after 5.6 years of follow up.(Eward et al., 2010)

Outcome in this case was measured PEDS QL score as recommended by Javaid *et al* in their study and showed an excellent result with no decrease in function.(Javaid et al., 2019)

Conclusion: Fibrous Dysplasia is a benign intramedullary fibro-osseous lesion that are reported to represent approximately 5%-7% of benign bone tumor. The most common clinical symptom is swelling or deformity of the affected site. Treatment of fibrous dysplasia include bisphosphonates therapy and surgical procedures like simple curettage or resection with bone graft may be required for correction of deformity, prevention of pathologic fracture and/or eradication of symptomatic lesion.

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