Fibrous Dysplasia Of Long Bone: A Case Series

Dr. Supratim Roy¹, Dr. Aniket K Wankhede², Dr. Girish B Mote³, Dr. Chandrashekhar M Badole⁴
Dr Preetam Salunkhe⁵

¹M.B.B.S, M.S Orthopedics - Assistant Professor, Mahatma Gandhi Institute Of Medical Sciences Sevagram ,
²M.B.B.S, M.S. Orthopedics - Senior Resident Mahatma Gandhi Institute Of Medical Sciences Sevagram
³M.B.B.S, M.S., DNB Orthopedics - Assistant Professor, Mahatma Gandhi Institute Of Medical Sciences, Sevagram.
⁴M.D Community Medicine, M.S Orthopedics – Professor In Mahatma Gandhi Institute Of Medical Sciences Sevagram .
⁵MD, Assistant Professor of Medicine at Mahatma Gandhi Institute of Medical Sciences, Sewagram, Maharashtra.

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ABSTRACT

It is retrospective followup study of case series of 6 patients of fibrous dysplasia of long bone treated in a rural tertiary care center with bone grafting and internal fixation when required and their outcome was followed.

Here, we present a case series of 6 patients of Fibrous dysplasia affecting the long bones with the clinical, radiographical, and histopathological features. Diagnosis was confirmed by biopsy after all relevant investigations. The patients were treated surgically as well as conservatively. Patients were followed regularly to note the resolution of disease or progression or any complications.
INTRODUCTION:
The term fibrous dysplasia was first coined by lichenstein¹ and jaffe² in 1942 but the first description of fibrous dysplasia is attributed to Weil in 1922 ³. It’s a rare bony disorder in which normal bone is replaced by abnormal fibro-osseous tissue resulting in substantial loss of bone strength and subsequent pain, deformity and pathological fractures⁴. It’s a benign skeletal lesion and exists in three forms monostotic, polyostotic and polyostotic with endocrinopathies that is the Mccune Albright syndrome where there is hyper pigmentation and endocrinological disorder⁵.

Another rare form is the Mazabrounds syndrome characterized by association of single or multiple intramuscular myxomas with fibrous dysplasia which may be monostotic or polyostotic ⁶. FD is caused by somatic activating mutations of the gene GNAS, in a subunit of stimulatory G protein, located at 20q13.2-13.3 ⁴. Fibrous dysplasia’s represent 5-7% of all benign bone tumour⁴. Most common sites of FD are the maxillary bones ,proximal femur and tibia followed by humerus, ribs, radius and iliac bones⁷. Metaphyseal involvement is mostly seen in long bones. Monostotic fibrous dysplasia cease to grow after puberty but polyostotic forms continue after skeletal maturity and severe skeletal deformity may occur⁸. Generally the first symptoms appear between 5-20 years age.⁷ Diagnosis of fibrous dysplasia is based on physical, radiological and histopathological examinations and treatment approaches include observation, medical treatment and surgical treatment⁹. Management of fibrous dysplasia aims at increasing bone strength and correcting the deformity and preventing future fracture and deformity.⁴

Here we are discussing the outcome of management of 6 cases of fibrous dysplasia affecting the lower limb which presented in our rural hospital of central India.

CASE 1
20 year old male presented with a complaints of pain over right hip since 6 months and difficulty in walking since 6 months. There was no history of trauma or any history of fever. Patient appears well built and nourished.

On examination patient had tenderness over right hip around the greater trochanter. No obvious swelling and deformity of the hip joint, range of movement was complete but painful on weight bearing .Plain radiograph of the hip joint and MRI was done . Radiograph shows ground glass appearance of a clearly defined fibrosseous lesion involving the femoral neck. MRI shows well defined altered signal intensity area of approximate size 9.5 x 3 x 2.4 cm appearing isointense to adjacent muscles on T1W, slightly hyperintense on T2W and hyperintense on STIR sequence within the femoral head, neck and proximal shaft. The lesion shows moderate heterogeneous enhancement on post contrast images. There is no surrounding edema or evidence of vascular invasion.
Fig 1- Preop xray of pelvis with both hip showing lytic lesion on the right neck and proximal shaft of femur

Fig 2 – MRI images of the proximal femur showing the lesion in the neck and trochanteric region

Fig 3- MRI T1 weighted image of the proximal femur right side showing changes of fibrous dysplasia

Fig 4 – T2 weighted image of the proximal femur of the right side

Biopsy from the femoral neck was taken which suggested of fibrous dysplasia. The patient underwent autologous fibular strut grafting of the femoral neck through lateral approach.

Surgical technique - anesthesia spinal, Position supine. UAAP cleaning and draping done. Lesion confirmed on the C-arm. Incision given at right thigh around 5 cm starting just below the Greater trochanter, skin & subcutaneous tissue retracted, drilling at neck of femur right side done. Cavity exposed.

Curettage of the lesion done. 10 cm Fibular graft harvested from right leg and 8cm fibular graft was put in neck of femur right side. Position of the graft confirmed under C-arm. Wash given. Wound closed in layers. Sterile dressing done. Boot and bar applied to right lower limb. Postoperatively the patient was stable and was discharged after suture removal and completing the physiotherapy. Pain in the hip reduced significantly. Patient was pain-free in the follow-up period and the range of motion of hip joint was normal.
Fig 5 – immediate post op x-rays

Follow up x-rays

Fig 6 - Follow-up at 1 month

Fig 7 - Follow-up at 9 month

Fig 8 - Final follow up at 1.5 years showed fibular graft incorporation.
CASE 2
30 year old female presented with complaints of pain over the proximal part of left thigh since 3 years which was insidious in onset and gradually progressive, the pain increased on weight-bearing. No history of trauma, fever and weight loss.

The patient was averagely built. On local examination there was tenderness over the left greater trochanter, on hip movements, pain was present on the proximal thigh of the left side and no signs suggestive of any infection also, no crepitus.

Plain roentogram of proximal thigh with hip and MRI of left hip joint suggestive of fibrous dysplasia with pathological fracture of the greater trochanter.

Fig 9 – Preop AP and Lat view of left proximal femur fibrous dysplasia.

Fig 10 – MRI T1 and T2 coronal section

MRI revealed - Well defined homogeneously enhancing lesion with a narrow zone of transition in the left proximal part of femur involving part of the greater trochanter, femoral neck and intertrochanteric region mostly suggestive of fibrous dysplasia with a pathological fracture in the left subtrochanteric region.

HISTOPATHOLOGICAL suggestive of – fibrous dysplasia.
The patient underwent curettage of the lesion and grafting from the ipsilateral iliac crest and prophylactic fixation was done using a dynamic hip screw. Postoperatively the patient was pain-free no complications occurred and was discharged after suture removal with the advice of physiotherapy. At follow-up, the patient was pain-free with a normal range of motion.

*Postoperative radiograph*

**Fig 11 – Immediate post op (AP and LAT view)**

Follow-up radiographs at 1 month 3 month 6 month 1 year showed gradual graft incorporation and stable fixation.

**Fig 12 – follow-up at 1 month**

**Fig 13 – follow-up at 3 months**
CASE 3
41 yrs old male brought by relatives with a/h/o trauma to RT Leg 30 days back for which he did a radiograph and incidental finding of radiolytic cystic lesion over distal shaft tibia right side was found for which he was referred to our hospital
Now complaints of:- Pain in the right leg since trauma. L/E:- shows mild tenderness over right leg no gross swelling no crepitus no deformity no abnormal movement no distal neurovascular deficit.
MRI of the right leg was done which showed homogenously enhancing expansile lesion in distal shaft of right tibia suggestive of benign pathology likely to be fibrous dysplasia.
Operative procedure- UAAP C & D Done. Under c-arm guidance Radiolytic lesion identified over RT LEG. Incision given. Skin and S/C tissue retracted. Radiolytic lesion confirmed and a window of size 1cmx3cm made. Fibrous material from the lesion collected for biopsy and sent for histopathological examination. Curretage done proximally and distally in the window and washed with H2O2, Betadine and normal saline. Wound closed in layers. Sterile dressing done.

The patient was discharged on a long knee brace and advised ankle toe movements. The histopathological report showed fibrous dysplasia.
After 2 months the patient came with an alleged history of trauma sustaining an injury to the previously operated leg, he developed pain and swelling following trauma and difficulty in weight-bearing following trauma.

On examination, there was swelling and tenderness over the right leg on the operated scar. Radiograph of the limb showed a minimally displaced fracture of the right leg at the cortical window site.
The patient underwent stabilization of the fracture with titanium locking DCP and cancellous bonegrafting from the iliac crest.

**Operative Procedure** –  
**Procedure II** (For fracture shaft tibia RT side )UAAP C & D Done. Incision given over old scar. Skin and S/C tissue retracted .Fracture site exposed .The previous window exposed. Fibrous tissues removed and cavity washed with H2O2+Betadine. Open reduction and internal fixation with bonegrafting done for fracture distal 1/3 rd shaft tibia RT side by using 10 hole titanium dynamic compression plate and fixed with 4 proximal and 3 distal cortical screws with and the cavity filled with bone graft harvested from right iliac crest. Position of the graft and the implant fixation was checked under C-arm. Wound closed in layers under negative suction drain. Sterile dressing done.Long leg slab applied. Suture removal was done. Patient was discharged on long leg articast and advised non-weightbearing.  
No post operative complication seen.

Follow up x-rays showed stable fixation and graft incorporation. Patient was pain free and with the complete range of motion at the ankle joint.
Case 4
A 17 year old male presented with complaints of pain in the right hip since a week. Pain increased on walking.
On examination patient had of tenderness over right hip. No obvious swelling and deformity of the hip joint, range of movement was complete. Hip was painful on weight bearing. Radiograph shows ground glass appearance of a clearly defined fibroseous lesion involving the femoral neck. MRI findings were suggestive of fibrous dysplasia.
Biopsy from the femoral neck was taken which suggested of fibrous dysplasia. The patient underwent autologous fibular strut grafting of the femoral neck through lateral approach the fibula graft harvested from the ipsilateral side was 13.5 cm in size and they were grafted in two parts in the neck of size 6cm each. Boot and bar applied to right lower limb. Post operatively patient was stable and discharged after suture removal and physiotherapy. Pain in the hip reduced significantly. At the end of 3 months patient functionally improved and started with partial weight-bearing.

Fig 24 – Preop ap and lateral lytic lesion of the right hip

Fig 25 – Post op Ap view of the pelvis with both hips and Fibula bone

Followup radiographs

Fig 26 – follow up at 1 month
Case 5
35 years old female presented with COMPLAINTS OF Low backache Pain in left leg since 2 months.
On outside MRI patient was diagnosed to be having fibrous dysplasia of the proximal tibia of left side. Radograph of the left lower limb revealed osteolytic lesion in proximal tibia proximal fibula and distal femur. Blood investigations were done which showed increased Alkaline phosphatase and intact parathyroid levels and patient was diagnosed to be having polyostotic fibrous dysplasia with hyperparathyroidism. There were no other features of McCune Albright syndrome. We treated the patient conservatively on Long leg slab as there was no deformity or pathological fracture and medical management was done for hyperparathyroidism.

CASE 6
6 year old female came with pain in the right leg since 1 month. There was no history of trauma or fever or weight loss. Radiological investigation done which were suggestive of fibrous dysplasia.
Fig 29 – Xray proximal tibia ap and lateral view showing lytic region

Fig 30 – MRI T1 and T2

Biopsy was done and fibrous dysplasia was confirmed histopathologically. Patient was managed conservatively on long leg slab. On followup, patient was pain free and no deformity developed, radiograph showed signs of healing.

DISCUSSION

FD is a congenital, metabolic, nongenetic disturbance that represents 2.5% of all bone tumors and over 7% of all benign bone tumors. Although fibrous dysplasia is considered a benign condition, lesions involving the proximal femur can cause substantial morbidity. The bone is biomechanically abnormal and mechanically weakened due to the fibro-osseous replacement of normal bone. The differential diagnosis of FD includes simple bone cyst, non-ossifying fibroma, osteo-fibrous dysplasia, adamantinoma, low-grade intramedullary osteosarcoma, and Paget’s disease. The current gold standard for the diagnosis of FD is a histologically-proven fibro-osseous lesion with poorly defined margins which are confirmed by radiographic findings. Fibrous Dysplasia may also be seen with hypophosphatemic osteomalacia. Fibrous dysplasia can cause skeletal deformity. For example, shepherd’s crook deformity is seen in the proximal femur. A possible mechanism for this deformity is repeated microfracture and malunion, with the femur becoming laterally bowed and enlarged due to mechanical insufficiency. Bowing of the tibia may also be seen, and may due to the same reasons. Malignant changes in fibrous dysplasia are rare, occurring in less than 1% of all cases. The most common malignant tumor in this setting is osteosarcoma followed by fibrosarcoma and then chondrosarcoma. Although sarcomas in fibrous dysplasia can develop without prior irradiation, the role of radiation therapy in the occurrence of sarcoma in fibrous dysplasia is well known. For this reason, radiation therapy should be avoided in the treatment of fibrous dysplasia.

Treatment protocols for FD include observation, medical treatment, and surgery. Clinical observation is suggested for FD lesions that have no risk of pathologic fracture or deformity. Medical treatment with bisphosphonates may have benefits including improvement of function, pain relief, and lower fracture risk in appropriately selected FD patients.

Nonoperative treatment is usually recommended for dysplastic lesions involving the upper extremity. The standard treatment for symptomatic and asymptomatic lesions in fibrous dysplasia has been curettage and bone grafting. For patients with symptomatic dysplasia in the femur, surgical treatment alternatives include curettage and bone grafting, valgus osteotomy, plating, intramedullary nailing, and oblique wedge osteotomy. The modified Pauwels’ intertrochanteric osteotomy can be used in the treatment of pathologic femoral neck fractures due to extensive lesions of fibrous dysplasia. Osteotomies, intramedullary nailing, and neck cross-pinning are the preferred methods for the management of shepherd crook deformity. The osteotomies could be valgus with medial displacing osteotomy or multiple osteotomies. Yang et al. in treated 14 patients with...
shepherd crook deformity and obtained satisfactory results with valgus osteotomy curettage, massive impaction allograft and intramedullary nail with neck cross pinning. Valgus osteotomy with dynamic hip screw internal fixation was done by Lee et al with good result and improved functional outcome in fibrous dysplasia with shepherd’s crook deformity. Unsatisfactory results following curettage and cancellous bone grafting for lower extremity fibrous dysplasia was documented in several studies.

Graft resorption with replacement by fibro dysplastic bone may be the cause of failure as intralesional curettage does not necessarily remove all the dysplastic bone. Cortical bone grafts are superior to cancellous bone grafts or bone-graft substitutes because of the excellent quality of the remodelled cortical bone. Enneking and Gearen obtained satisfactory results in 15 patients of fibrous dysplasia (5 polyostotic and 10 monostotic) of proximal femur by using cortical bone graft. They used autogenous fibular and tibial graft, none of the patient developed severe Varus deformity of proximal femur resulting in osteotomy and intramedullary fixation.

In case of pathological fracture of proximal femur Gabriel and Campbell stated that the proximal fragment of the fracture should be reduced in a valgus position to provide stable fixation, compensate for any limb length discrepancy and prevent recurrent deformity. In our case series we had 2 male patients (17 yrs and 20 yrs) of fibrous dysplasia of proximal femur treated with curettage and autogenous cortical fibular strut grafting which were pain free in the follow-up period and also did not develop any progressive deformity or recurrence or pathological fracture in the follow-up period. Fixation was not done as there was no deformity requiring osteotomy or pathological fracture. One case of Fibrous dysplasia of proximal femur with pathological fracture of greater trochanter in a 30 year female was treated with curettage and cancellous bone grafting harvested from iliac crest and fixation was done using dynamic hip screw. Patient in the follow up period was pain free and there was no radiological signs of progressive deformity or recurrence and fixation was stable with graft incorporation. We also treated one patient (41 yr male) of fibrous dysplasia of tibia with curettage followed by H2O2 betadine wash alone but at the follow-up of 2 months the patient developed unicortical fracture at the pathological site which was treated by cancellous bone grafting of the cavity and fixation using locking plate. In the follow-up period was pain free showed fracture union and graft incorporation and no deformity. Curettage only without grafting or fixation resulted in weakening of the bone and pathological fracture hence, it is always better to go for a stable fixation in selected patient if impending pathological fracture is suspected.

RESULT
In our case series we found that cortical fibular grafting with the graft having properly anchored at the head and the lateral cortex had a good result and prophylactic fixation of the proximal femur is not necessary without pathological fracture or severe deformity. Cortical bone graft provides good structural support to the biomechanically weakened bone. In case of pathological fracture fixation along with curettage and bone grafting with cancellous bone healed with a good result. Though long term follow-up is necessary for the cases.

CONCLUSION
Prophylactic fixation of the fibrous dysplasia gives desirable outcome and also avoids future complications of fracture.

REFERENCES
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