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Original Article

# Clinical Outcome of Monostotic Fibrous Dysplasia Over Proximal Femur Treated with Intramedullary Nailing and Bisphosphonate Therapy

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## Abstract

**Introduction:** Fibrous dysplasia (FD) of bone is an enigma with no proper guideline. Treatment currently consists of curettage and bone grafting in an attempt to eradicate the lesion and to prevent progressive deformity. No definite criteria have been established to identify patients at high risk of presenting pathological fractures.

**Purpose:** The purpose of the study was to explore the effect of combination bisphosphonate therapy in diminishing pain, preventing fractures, lowering N-telopeptide values, and leading to partial resolution of FD lesions.

**Materials and Methods:** At Medical College, Kolkata, 10 patients with monostotic FD in lower extremities were treated between 2014 and 2018 and included in the study. All patients underwent full skeletal survey followed by core needle biopsy with the help of magnetic resonance imaging and C-arm guidance. After confirmation, closed intramedullary nail without reaming was used in all cases. Bone grafting was not performed. Zoledronic acid was given intravenously at the dose of 4 mg every 6 months. Patients were allowed full weight-bearing on the affected extremities on the 2nd post-operative day.

**Results:** Seven patients were female and three were male; their mean age was 26.9 years. The mean duration of follow-up was 30.5 months. We get good to average results. Clinicoradiological improvement of all cases was observed.

**Conclusion:** As a result of this study, we believe that intramedullary fixation can be performed successfully. Treatment of monoostotic fibrous dysplasia with adjuvant bisphosphonate therapy resulted in effective pain control and early return of functional activity. This will avoid problems that may occur following pathological fractures.

**Key words:** Intramedullary nailing, Monostotic fibrous dysplasia, Zoledronic acid

## INTRODUCTION

Fibrous dysplasia (FD) is a challenge to the orthopedic surgeon. FD, first identified by Lichtenstein in 1938,[1] is an anomaly characterized by widening of the affected bone with cortical thinning and presence of fibro-osseous tissue inside the bone. There may also be areas with islands of cartilage or cysts, and some lesions may be expansile. It may present under a monostotic or polyostotic form. FD of the proximal femur is difficult to treat due to the varied presentations such as pain, pathological fractures, severe deformity, and high chances of recurrence.[2] Lesions have a tendency to recur and may result in pathological fractures following curettage and grafting.[3] No definite criteria have been established to identify patients at high risk of presenting pathological fractures.[4] Clear guidelines for orthopedic management of FD in the proximal femur have not been established.

We reviewed published data on the treatment of FD with bisphosphonates, calcium, Vitamin D, and phosphorus. We present our results with intramedullary nailing along with intravenous zoledronic acid 4 mg in every 6 months, in 10 patients with monostotic FD, pain increasing with movement. To the best of our knowledge, no previous study has focused on intramedullary nailing of the proximal

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femur along with bisphosphonate therapy over monostotic FD in symptomatic patients.

The aim of our study was to analyze the various presentations of FD of the proximal femur such as pain, fractures, and shepherd crook deformity and describe the results of the various treatment modalities for the same.

## METERIALS AND METHODS

Ten patients with monostotic FD in their upper extremities operated between 2014 and 2018 at Medical College, Kolkata, were included in the study. Seven patients were female and three were male. The mean age of the patients was 26.9 years (range: 19–34). The mean duration of follow-up was 30.5 months. All patients underwent full skeletal survey followed by core needle biopsy with the help of magnetic resonance imaging and C-arm guidance. Patients who had been diagnosed with FD with functional pain or pathological fractures were included in the study and undergone biopsy confirmation and intramedullary nailing, whereas patients who had no functional pain and incidentally diagnosed were excluded from the study. After taking written consent from every patient and their family member, we performed core needle biopsy confirmation for all cases. Under C-arm guidance, we performed intramedullary nail titanium, preferably the third-generation long gamma nail for better fixation of proximal femur, as it is a weight-bearing bone. We put all patients under medical therapy of intravenous zoledronic acid 4 mg dissolved with 100 ml normal saline. The prepared solution was infused slowly over 30min half yearly. We avoid local curettage and bone grafting because FD is notorious for its recurrence and local fibrous tissue prevents the activity of bone grafting. As well as, it is an open procedure; hence, post-operative morbidity is higher. Resorption and recurrence secondary to grafting after curettage are other problems. Reaming was not used before nailing in our patients, as it was deemed unnecessary for nailing of non-fractured long bones, added to the fact that it might have contributed to weakening the bone to some extent.

We planned for adjuvant medical therapy as we did not use bone graft. Radiological follow up was done to see the effect of zoledronic acid over the pathological fibrous tissue. Functional pain, size of the lesion on radiographs, and stability of prophylactic fixation were evaluated in follow up visits every 6 months. A visual analog scale (VAS) was used in assessing functional pain.

## RESULTS

We have done an observational prospective study [Figure 1]. Mean VAS for functional pain was 5.33 ± 0.65 preoperatively and 2.26 ± 0.57 at final follow-up ($P < 0.05$). We present our results with intramedullary nailing along with intravenous zoledronic acid 4 mg in every 6 months, in 10 patients with monostotic FD, pain increasing with movement. For statistical analysis, Wilcoxon signed-rank test (with Bonferroni correction) was used and $P < 0.05$ was considered as statistically significant.

Seven patients were female and three were male; their mean age was 26.9 years. The mean duration of follow-up was 30.5 months. Recurrences, pathological fractures, and deformities secondary to the lesion occurred in none of the patients.

## DISCUSSION

FD is an orthopedic condition with a wide spectrum of presentation. The treatment of the dysplastic lesions in the proximal femur region is still somewhat unclear and varies widely.[5] FD of the bone can present as three clinical forms: Monostotic, polyostotic, and as a part of a McCune-Albright syndrome. Lichtenstein[1] is credited with having coined the term FD, in 1938; in 1942, Lichtenstein and Jaffe[6] reviewed all known cases of this entity. Those authors established that FD of bone was a distinct pathological and clinical condition. FD may occur due to a failure in remodeling of primitive bone into mature lamellar bone, which negatively affects the mechanical properties of the affected bone. Thus, pain, deformities, and pathological fractures may occur. It is generally accepted that monostotic lesions are easier to treat, are associated with better outcomes, necessitate fewer operations, and result in fewer fractures.[7]

Healing after pathological fractures in dysplastic bones is comparable with that of normal bone. However, the callus includes dysplastic bone tissue.[8] The lesion persists despite the healing of the fracture. The accepted principle in the treatment of lesions that are painful or at risk for fracture, even if asymptomatic, is curettage and grafting.[9] However, according to our review of literature, it is uncertain whether this form of treatment offers a definitive solution.[10] It has also been reported that curettage or biopsy of an isolated lesion may predispose the bone to pathological fracture or progression of the lesion.[3] There is no accurate indication of the rate of success of curettage and bone grafting. In their study on patients with FD localized in the neck of the femur, Guille *et al.*[9] have shown that the lesion was not eradicated with curettage and grafting, and the bone was further weakened due to deformation of the trabecular structure in dysplastic bone as a result of curettage. In the present


**Figure 1: (a) Pre-operative X-ray. (b) Magnetic resonance imaging. (c) Core needle biopsy. (d) Histopathology. (e) Post-operative. (f) Follow up post operative Xray. (g) Post operative rehab painless squatting. (h) Post zoledronic acid therapy 1 yr**

| Cases | Sex | Age (years) | Site | Indication of intervention | Follow-up (months) | Pre-operative VAS | Post-operative VAS |
|---|---|---|---|---|---|---|---|
| 1 | M | 34 | Proximal femur | Pain | 32 | 5.8 | 2.4 |
| 2 | F | 23 | Proximal femur | Pathological# | 30 | 4.9 | 1.7 |
| 3 | F | 19 | Proximal femur | Pain | 26 | 6.4 | 1.8 |
| 4 | F | 29 | Proximal femur | Pain | 32 | 5.7 | 2 |
| 5 | M | 23 | Proximal femur | Pain | 27 | 4.9 | 1.6 |
| 6 | F | 20 | Proximal femur | Pain | 29 | 6.1 | 2.5 |
| 7 | F | 30 | Proximal femur | Pathological# | 25 | 5.2 | 3 |
| 8 | M | 24 | Proximal femur | Pain | 25 | 5.3 | 2.5 |
| 9 | F | 25 | Proximal femur | Pain | 28 | 5.7 | 2.9 |
| 10 | M | 26 | Proximal femur | Pathological# | 29 | 4.5 | 1.8 |

M: Male, F: Female, VAS: Visual analog scale

series, we did not perform curettage and bone grafting. Resorption and recurrence secondary to grafting after curettage are other problems. Guille *et al.*[10] have shown in their study that all cancellous or cortical grafts they used, in addition to autogenous fibular strut grafts, were resorbed. In addition, according to DiCaprio and Enneking,[4] cortical grafts are more durable compared to cancellous grafts, as they are only partly replaced by dysplastic host bone: Only their osteonal portion (about 50% of the graft) is replaced by dysplastic bone, whereas the interstitial lamellae are not replaced and persist. The size of the lesion may change with skeletal growth; however, it is difficult to differentiate whether this is secondary to skeletal growth or to progression of the lesion.[7] Since FD is a genetic disorder which is not curable, the treatment modality should be long lasting. No definite criteria are available to state in which cases pathological fracture will occur.[4] In the multicentric study of the European Paediatric Orthopaedic Society, fractures had occurred in 47% of patients with monostotic FD.[11]

Therefore, we recommend prophylactic intramedullary fixation in patients with monostotic FD. This prophylactic therapy avoids complications such as delayed union and deformities following fracture. A vascularized fibula has been used in some cases following fracture.[12] It appears more reasonable to take the necessary steps to prevent fracture, considering the technical difficulty, delayed weight-bearing, risks of graft resorption, and refracture in addition to high costs, if the affected bone is not strengthened and fractures.

## CONCLUSION

As a result of this study, we believe that intramedullary fixation can be performed successfully in cases of monostotic FD with adjuvant bisphosphonate therapy proven to increase function activity and control of pain. This will avoid problems that may occur following pathological fractures.

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