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Management of Fibrous Dysplasia of Proximal Femur by Internal Fixation Without Grafting: A Retrospective Study of 19 Patients

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Abstract

Introduction: This paper assesses whether treating patients with fibrous dysplasia of proximal femur by internal fixation with correction of the deformity, if present, without grafting is enough or not.

Methods: This study is a retrospective study using patient's medical records, including analysis of 19 patients with fibrous dysplasia of proximal femur treated by internal fixation only and followed up between 2000 and 2017 for at least 2 years. Epidemiologic data, clinical manifestations, radiologic and histologic investigations, surgery, functional outcomes, and complications in these patients were analyzed.

Results: The study included 12 male patients and 7 female patients, with a mean age of 20.74 years. Of these, two patients presented with pathologic fractures, two presented with shepherd's crook deformity, and the remaining presented with hip pain and limping. Implants used for internal fixation were intramedullary nail in four patients, dynamic hip screw in eight patients, cannulated screws in four patients, broad dynamic compression plate in two patients, and narrow dynamic compression plate in one patient. The mean follow-up period was 53.58 months. Four patients had postoperative complications. The mean Musculoskeletal Tumor Society score was 27.63 points.

Discussion: The optimal management option for fibrous dysplasia of proximal femur is debatable. We suggest that internal fixation without grafting has a good local control and satisfactory functional long-term outcome.

Fibrous dysplasia (FD) of bone is a rare, benign bone lesion, which represents replacement of normal bone with disorganized, immature fibrous tissue resulting from the fail-

ure of remodeling of the primitive bone in response to the mechanical stresses and failure of mineralization of the immature matrix.^{1,2} FD may present as a single (monostotic) or

multiple (polyostotic) lesion or be a component of McCune-Albright syndrome.³⁻⁵

The proximal femur is the most common site of FD, which may be asymptomatic or present with pain, limping, limb shortening, deformity, or pathologic fracture.^{3,6} It may lead to hip varus deformity, which eventually leads to a shepherd's crook deformity because of weight bearing on weakened bone in the proximal femur.⁷

Benign bone lesions behave differently, and accordingly, their treatment methods differ from one type to another.⁸ The treatment is always directed to manage the present symptoms and prevent the future morbidities.⁹ For example, giant cell tumors cause pain and progressive bone destruction; hence, the treatment always aims at eradicating the active and aggressive nature of the tumor by extended curettage and filling the defect so as to restore the integrity of bone.^{9,10} However, latent lesions like FD cause pain or limping because of bone weakness, and they are not aggressive lesions.² Accordingly, management of FD always aims at increasing the bone strength and correcting the present deformity or preventing the future fracture and deformity.^{2,6}

Various treatment options have been described for managing FD of proximal femur.^{2,3,11} Nonsurgical treatment includes casting and pamidronate infusions.¹² Surgical options include curettage and bone grafting, valgus osteotomies, and internal fixation.^{11,13}

There is a debate whether restoring or augmenting the strength of bone in patients with FD of proximal femur requires just prophylactic internal fixation or does it require additional grafting.^{11,12,14}

Our study aims to evaluate the functional outcomes in patients with FD of proximal femur treated by internal fixation only without graft-

ing, aiming at alleviating their present symptoms and preventing future morbidities.

Methods

Our study is a retrospective study based on reviewing the clinical, radiologic, and pathologic data of patients. The study included 19 patients with FD of proximal femur treated by internal fixation only without grafting, who were followed up between 2000 and 2017 for a minimum of 2 years, were included in the study after approval of the ethical committee.

Records of the patients were assessed for the epidemiologic data, clinical manifestations, pathologic and radiologic investigations, number and type of surgical procedures, the choice of implant used for fixation, functional outcome, and complications.

Pre- and postoperative plain radiograph were reviewed; also, CT and MRI were reviewed when available (Figures 1 and 2).

Methods of Assessment

After treating the patients with FD by internal fixation only, they were followed up through clinical and radiologic evaluation, at 1.5, 3, 6, 12, and 24 months postoperatively. Postoperative complications were recorded. The Musculoskeletal Tumor Society (MSTS) score¹⁵ was used for the evaluation of functional outcomes.

Statistical Analysis

Statistical analysis of data focused on epidemiologic data and functional outcome. Continuous variables such as age and follow-up period were expressed as mean and range. Complications were described as a percentage rate.

Results

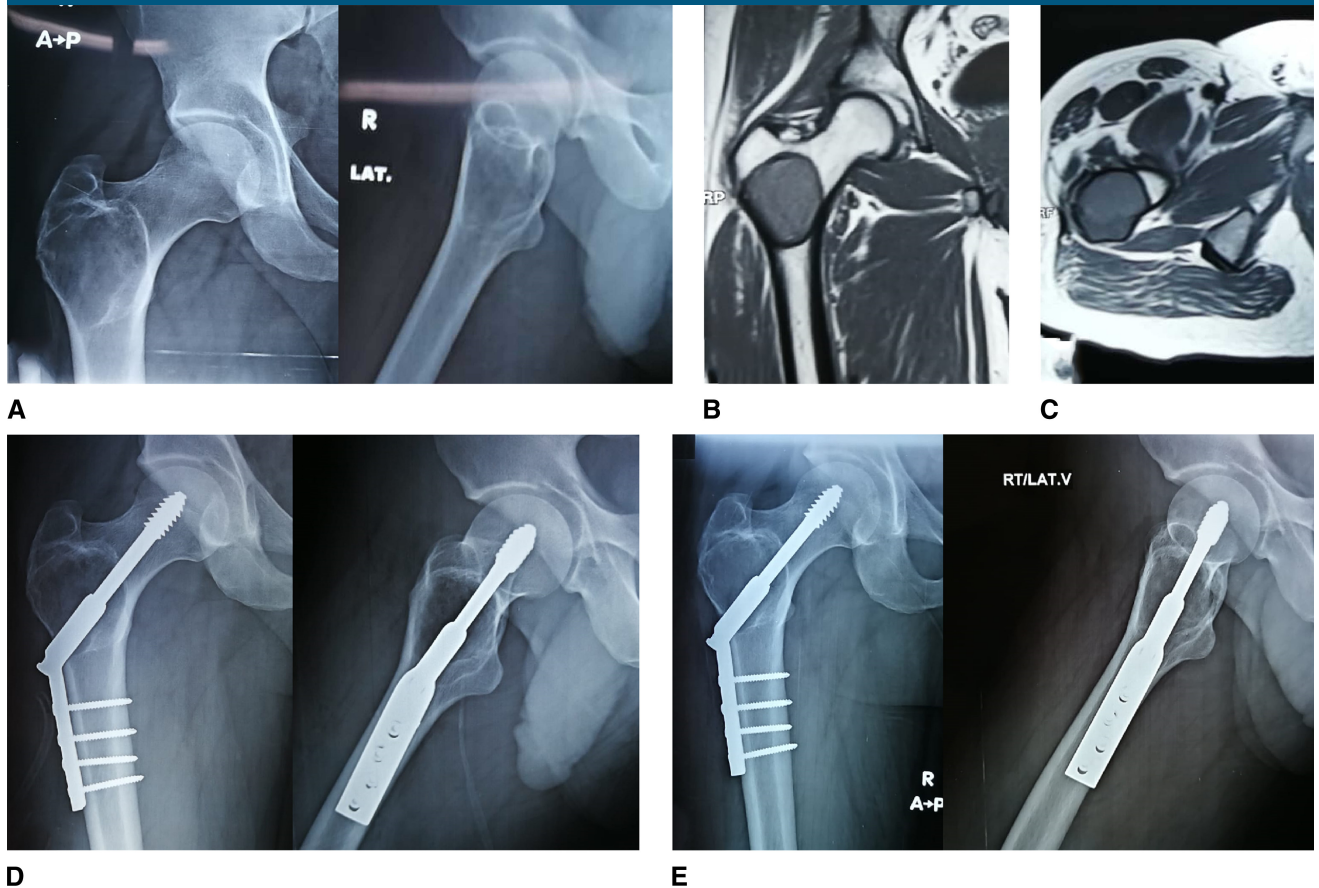
The study included 12 male patients (63.16%) and 7 female patients (36.84%). The mean age of the patients was 20.74 years (range, 7 to 40 years). Eighteen patients had a monostotic FD and one patient had a polyostotic disease. One patient had previous curettage and non-vascularized fibular grafting elsewhere and complicated with graft resorption (Figure 2). Eight patients (42.11%) presented with hip pain, two patients (10.53%) presented with limping, five patients (26.32%) presented with hip pain and limping, two patients (10.53%) presented with pathologic fractures, and two patients (10.53%) presented with shepherd's crook deformity (Table 1). Of the 19 patients, 2 patients (10.53%) underwent core biopsy for confirming diagnosis.

All the 19 patients with manifesting FD underwent internal fixation only without grafting. The choice of implant used for fixation depended on the location of the lesion and the bone quality. Fixation by intramedullary (IM) nail was done in four patients (21.05%), dynamic hip screw (DHS) in eight patients (42.11%), cannulated screws in four patients (21.05%), broad dynamic compression plate (DCP) in two patients (10.53%), and narrow DCP in one patient (5.26%). Valgus osteotomy was done in two patients (10.53%) with shepherd's crook deformity.

Functional Outcome

The mean follow-up period was 53.58 months (range, 24 to 159 months). The mean MSTS score was 27.63 points (range, 21 to 30 points) (Table 1). With the exclusion of the two patients with shepherd's crook deformity, who underwent valgus osteotomy and internal fixation, the mean MSTS score of the remaining 17 patients would be 28.41. At the final follow-up,

Figure 1



Thirty-year-old male patient with fibrous dysplasia of right proximal femur treated with internal fixation by dynamic hip screw. **A**, Preoperative radiograph AP and lateral views. **B**, Preoperative MRI coronal view. **C**, Preoperative MRI axial view. **D**, Postoperative radiograph AP and lateral views. **E**, Two-year follow-up radiograph AP and lateral views.

none of our patients had hip pain, refracture, or progression of deformity. All osteotomy sites showed complete union. No patients in our study developed internal fixation failure.

Complications

Four patients (21.05%) had postoperative complications. One patient (5.26%) developed mild limping, which required shoe lift of 1 cm. One patient (5.26%) experienced superficial wound infection, which was managed with antibiotics efficiently. One patient (5.26%) had residual varus deformity and mild limping, which required shoe lift of 1 cm. One patient (5.26%) had residual varus

deformity and 3 cm limb length discrepancy, which required also shoe lift.

Discussion

FD of bone is characterized by the presence of fibro-osseous tissue in the bone with cortical widening and thinning.^{2,16} FD of proximal femur has a wide spectrum of clinical presentations, ranging from hip pain, limping, to deformity, or may present as a pathologic fracture.^{2,7,11}

In our study of 19 patients with FD of proximal femur, 12 were male and 7 female, which coincided with the numbers in Tong et al,¹⁷ who reviewed 15 patients, of which 9 were

male and 6 female, and also in Kushare et al,¹⁸ who reviewed 23 patients, of which, 14 were male and 9 female. In our study, the mean age of the patients was 20.74 years, which was slightly lower than that of the patients in Tong et al¹⁷ (mean age of patients was 25 years) and Majoor et al¹⁹ (mean age of patients was 23 years).

The main problem in FD is the resultant weak bone that becomes so evident in the proximal femoral location. So, the treatment is always directed to augment or restore the strength and integrity of bone.^{2,6} The goal of surgery in FD of proximal femur is to eliminate the symptoms resulted from repeated fissures or fractures caused by the lesion, to

Figure 2



Thirty-year-old male patient with fibrous dysplasia of left proximal femur, with previous curettage and nonvascularized fibular grafting, treated with internal fixation by dynamic hip screw. **A**, Radiograph AP view showing osteolytic lesion in the neck femur. **B**, Radiograph AP view after curettage and nonvascularized fibula grafting. **C**, Preoperative radiograph AP view showing graft resorption and persistent lesion. **D**, Preoperative MRI coronal view. **E**, Postoperative radiograph AP view. **F**, Four-year follow-up radiograph AP view.

correct the deformity if present, and to prevent future morbidities such as pathologic fractures or deformities.⁶

Some authors believe that restoring the integrity of bone requires grafting.^{17,19} However, we do believe that

only internal fixation would achieve this goal without associated morbidities of bone grafting. So, this

Table 1

Patient Details								
No	Sex	Age (yr)	Presentation	Type of FD	Implant	Postoperative Complications	Follow-up Period (mo)	MSTS Score
1	Female	23	Hip pain	Monostotic	IM nail	None	195	27
2	Male	35	Pathologic fracture	Monostotic	DHS	None	78	30
3	Male	11	Pathologic fracture	Monostotic	DHS	Mild limp required shoe lift of 1 cm	79	27
4	Female	22	Hip pain	Monostotic	Three cannulated screws	Superficial infection healed with antibiotics	51	26
5	Female	16	Hip pain, mild limping	Monostotic	IM nail	None	54	30
6	Male	7	Hip pain	Monostotic	Two cannulated screws	None	58	30
7	Male	14	Hip pain, limping	Monostotic	Broad DCP and screws	None	59	30
8	Female	10	Shepherd's crook deformity	Monostotic	Valgus osteotomy + DHS	Residual varus deformity, mild limp required shoe lift of 1 cm	44	21
9	Male	20	Hip pain	Monostotic	DHS	None	40	30
10	Female	27	Hip pain	Monostotic	Two cannulated screws	None	39	30
11	Male	10	Hip pain, limping	Monostotic	DHS	None	41	30
12	Male	15	Occasional hip pain	Monostotic	Broad DCP	None	38	30
13	Male	40	Shepherd's crook deformity	Polyostotic	Valgus osteotomy and IM nail	Residual varus deformity, LLD 3 cm required shoe lift	27	21
14	Male	34	Hip pain, limping	Monostotic	Two cannulated screws	None	28	27
15	Female	15	Limping	Monostotic	DHS	None	25	26
16	Male	10	Limping	Monostotic	Narrow DCP	None	24	27
17	Male	30	Hip pain, previous curettage and NVF	Monostotic	DHS	None	65	25
18	Female	25	Hip pain	Monostotic	IM nail	None	49	30
19	Male	30	Hip pain, limping	Monostotic	DHS	None	24	28

DCP = dynamic compression plate, DHS = dynamic hip screw, FD = fibrous dysplasia, IM = intramedullary, LLD = limb length discrepancy, MSTS = Musculoskeletal Tumor Society, NVF = nonvascularized fibula

study assesses whether doing internal fixation for these patients would alleviate their symptoms and prevent future morbidities. Moreover, one might think that doing internal fixation without grafting would put the patient under the risk of fixation failure by time. However, in our study, none of the fixations failed with a mean follow-up period of 53.58 months (range, 24 to 159 months). The reason for this was

that bone integrity was already present and failure of fixation would happen if bone continuity was lost or ununited fractures were present, which was not the case. Moreover, the complications that occurred in our patients are not related to the nonuse of grafting.

In Nakashima et al²⁰ and Onoda et al,²¹ intralesional curettage and bone grafting was the treatment option. Majoor et al¹⁹ performed

cortical strut allografting for impending or actual proximal femoral fractures in 30 patients with FD. In Tong et al,¹⁷ 15 patients were treated by internal fixation with DHS and anatomic plates following curettage and bone grafting with valgus osteotomy in shepherd's crook deformity. In Nishida et al,²² eight patients with proximal femoral FD were treated with fibular strut grafting and compression hip screw fixation. Jung

et al²³ treated eight patients with shepherd's crook deformity by multiple osteotomies and IM nailing.

In our study, all the patients were treated by internal fixation only without grafting using variable implant options depending on the lesion site and bone quality. IM nailing was done in four patients (21.05%), which is a good fixation option for proximal femur, especially in subtrochanteric lesions.²⁴ Long IM nailing traversing the neck and having a firm purchase in the head can prevent loss of neck shaft angle.²³ In our study, DHS was used in eight patients (42.11%), cannulated screws in four patients (21.05%), broad DCP in two patients (10.53%), and narrow DCP in one patient (5.26%); and valgus osteotomy was performed in two patients with shepherd's crook deformity.

In our study, with the use of internal fixation implants, the mean MSTS score was 27.63 points, which was considered satisfactory. This score was slightly lower than that in Lang et al,²⁵ where the mean MSTS score was 28.42 points. Also, this score was worse than that in Rosario et al,²⁶ where the mean MSTS score was 29.6 points. This may be attributed to using a minimally invasive surgical approach by Rosario et al²⁶ and relatively low MSTS score, 21 of 30, in two patients with shepherd's crook deformity in our study in whom valgus osteotomy was performed but unlikely with under correction of the varus deformity.

Yang et al²⁷ obtained satisfactory results with valgus osteotomy, curettage, impaction allograft, and IM nail with neck cross-pinning. Li et al¹³ reported good results and improved functions with valgus osteotomy and DHS fixation. Stephenson et al¹² suggested that only curettage and bone grafting of proximal femoral FD is able to obtain a favorable outcome. Con-

trarily, Enneking and Gearen²⁸ reported that after curettage and bone grafting, lesions were likely to be recurrent and it was not possible to obtain a satisfactory result.

Regarding complications, four patients (21.05%) in our study had postoperative and late complications. One patient had superficial wound infection, which was healed with antibiotics. One patient had mild limping, which required shoe lift of 1 cm. And two patients had residual varus deformity, one of them with 3 cm limb length discrepancy and required also shoe lift. These complications would not be prevented if grafting was used. No internal fixation loosening occurred in any of the patients in our study. Complications in our study were slightly lesser than those in Kushare et al,¹⁸ with a complication rate of 21.7%. In Majoor et al,¹⁹ the complication rate was 3.3%, with only one patient having a refracture after surgery, which was treated and healed conservatively. In Rosario et al,²⁶ none of the patients developed complications.

In our study, follow-up loss of neck shaft angle did not occur, which was better than O'Sullivan and Zacharin²⁹ and Jung et al²³ who reported loss of neck shaft angle in five and two patients, respectively. Our better results were attributed to good stabilization of the femoral neck, which was crucial to prevent the progression of deformity.

The limitations of our study are its retrospective design and single-center site. Also, we had a relatively small number of patients with variable internal fixation implants.

Conclusion

The optimal management option for FD of proximal femur remains debatable. Internal fixation only without grafting has satisfactory

functional outcome that alleviates the patient's symptoms, improves the limb function, and prevents future morbidities such as deformities and pathologic fractures. This method eliminates the addition of extra morbidity, longer surgical time, and extra cost caused by bone grafting.

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