

Original Research Article

Functional outcome in surgical fixation of fibrous dysplasia of proximal femur

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ABSTRACT

Background: Fibrous dysplasia is a rare benign intramedullary fibro-osseous lesion. Surgical management is usually indicated in pathological fractures, symptomatic patients due to pain and due to deformity. The aim of this study was to summarize the functional results and to investigate surgical treatment methods by conducting a retrospective study of patients with fibrous dysplasia (FD) in the proximal femur.

Methods: This study is a retrospective study using patient's medical records, including analysis of 14 patients with fibrous dysplasia of proximal femur treated at Nizams Institute of Medical Sciences. The primary outcome was success of the procedure according to the Musculoskeletal tumour society score (MSTS). Secondary outcomes, which were assessed at one year and at the end of follow up in terms of revision free survival and change in femoral neck-shaft angle over time.

Results: Analysis of data showed that revision-free survival was 85% after a median follow-up of 2 years. Revision was necessary in one patient for structural failure due to a fracture distal to Dynamic hip screw and other patient due to progressive deformity after being operated with broad DCP. At the end of follow-up 85% of all patients had good walking ability and all patient were pain free.

Conclusions: Symptomatic fibrous dysplasia involving proximal femur needs to be operated to relieve pain due to biomechanical instability, to prevent and treat pathological fractures and to correct deformities.

Keywords: Fibrous dysplasia, Benign bone tumour, Deformity of proximal femur, McCune-Albright syndrome

INTRODUCTION

Fibrous dysplasia (FD) is a condition characterized by the presence of fibro-osseous tissue in the bone leading to widening and thinning of its cortex.¹ It begins in childhood and progress beyond puberty in to adulthood. It accounts for 5-7% of clinical benign tumors.²⁻⁴ According to its clinical characteristics, FD is divided into 4 types: monostotic type, polyostotic type, McCune-Albright syndrome and Mazabraud syndrome.⁵ McCune-Albright syndrome is fibrous dysplasia in association with distinct café-au-lait skin spots with a jagged (coast-of-Maine)

border, and/or in association with a number of hyperfunctioning endocrinopathies (including precocious puberty, hyperthyroidism, growth hormone excess, rickets/osteomalacia, and others) and Mazabraud syndrome is fibrous dysplasia in association with intramuscular myxomas.⁶⁻⁹ In general, the mean age of patients with monostotic FD presenting with clinical symptoms is 15 years old, and the mean age of patients with polyostotic FD is 30 years old.² Common invasive skeletal sites include the long bones, ribs, maxillofacial skeleton and pelvis. The proximal femur is the most commonly affected site and its main symptoms include

pain, deformity and lameness which seriously influence the functioning of affected limbs.¹⁰ Surgical management of fibrous dysplasia of proximal femur is challenging because of the high load of mechanical forces acting at this skeletal site, Severe deformity, Pathological fractures and high chances of recurrence. Aim of the study is to study the functional results of surgical management of fibrous dysplasia of proximal femur using outcome indicators as: revised musculoskeletal tumour score (MSTS), evision-free survival, change in femoral neck-shaft angle over time.

METHODS

This is a retrospective study using patient’s medical records, including analysis of 14 patients with fibrous dysplasia of proximal femur operated at Nizams Institute of Medical Sciences between May 2017 and March 2020. Inclusion criteria was: age between 10 years to 50 years, fibrous dysplasia of proximal femur: associated with pain, associated pathological fracture, associated with progressive deformity.

Exclusion criteria was: fibrous dysplasia affecting other bones except proximal femur, fibrous dysplasia of proximal femur treated conservatively, other bone tumours affecting proximal femur.

In all the patients, lesion is approached by lateral approach to hip, an Antero-lateral Bone window is made and intramedullary gravel-like lesion tissues were removed and sent for frozen. If the medial femoral calcar is destroyed by the disease, fibular graft harvested from ipsilateral leg was grafted at the femoral neck. Internal fixation done with Dynamic Hip Screw, Proximal femur nail or broad DCP. For patients with an open osteoepiphyseal line- damage to the epiphyseal line should be avoided. Plate should bypass the lesion and adequate number of screws should be placed in normal bone.

In the study, we have used DHS with fibular grafting in 6 cases, DHS alone in 3 cases, broad DCP in 2 cases and intramedullary nailing in 3 cases. A uniform postoperative rehabilitation protocol was employed for every patient. Partial weight bearing with walker support started 12 weeks after surgery. Complete weight bearing allowed at 4 months after surgery. In addition, postoperative follow-up and X-ray examination were conducted regularly every 3 months. All the patients are followed up for atleast 1 year.

The primary outcome was success of the procedure according to the revised Musculoskeletal Tumour Society (MSTS) functional score. Secondary outcomes, which were assessed at one year and at the end of follow-up included: revision free survival rate and change in femoral neck-shaft angle over time. Musculoskeletal Tumour Society (MSTS) functional score system measures outcomes in six categories, including pain, function, and emotional acceptance for all patients; use of walking aids,

walking ability, and gait. Each parameter is scored 0 to 5 and combined for a possible total score of 30. A score of 23 or greater is considered an excellent result; a score of 15 to 22 points, a good result; a score of 8 to 14 points, a fair result; and a score less than 8, a poor result.

RESULTS

Among the 14 cases, 7 patients were males and 7 were female patients. Mean age of all patients is 20years. 11 cases were monostotic type and 3 cases were polyostotic type. 2 cases were accompanied by shepherd’s crook deformity.

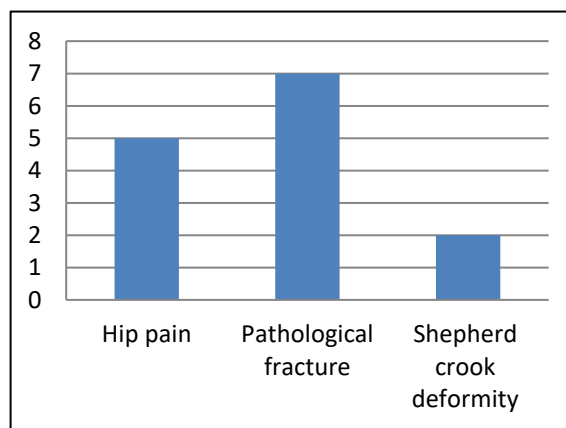


Figure 1: Presentation of fibrous dysplasia in proximal femur.

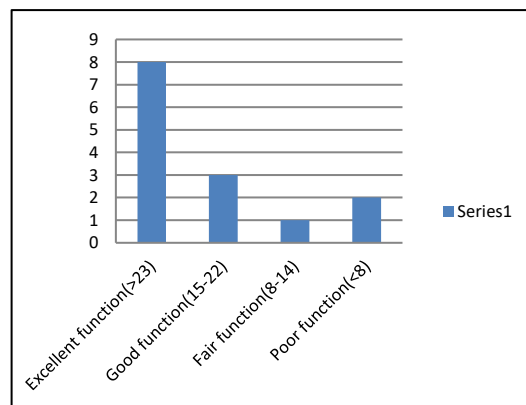


Figure 2: Functional outcome (MSTS score) at 1 year after surgery.

Out of 14 patients, 5 patients presented with hip pain, 7 patients presented with pathological fracture and 2 patients presented with shepherd crook deformity (Figure 1). All patients were followed up for 1 year. Prior to surgery only 3 of the 14 patients had a good function (walking a normal distance unaided and without complaints), 6 had moderate function (able to walk for a small distance) and 5 could only mobilize with the help of crutch. One year postoperatively 8 patients had excellent function, 3 patients had good function, 1 patient had fair function and

2 patients had poor function (Figure 2). Mean MSTS score in the study was 21.6.



Figure 3: Radiograph of 34 years male patient with fibrous dysplasia of left femur neck.

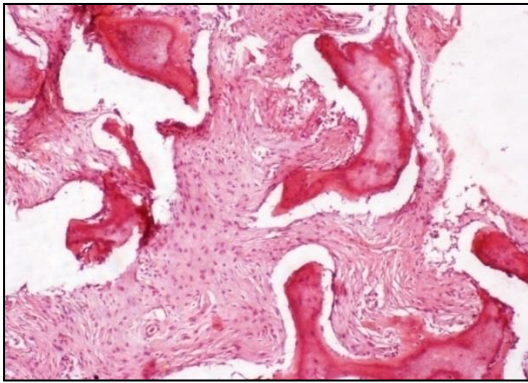


Figure 4: Histopathology showed dense fibrous stroma with irregular bony trabeculae with no osteoblastic rimming.

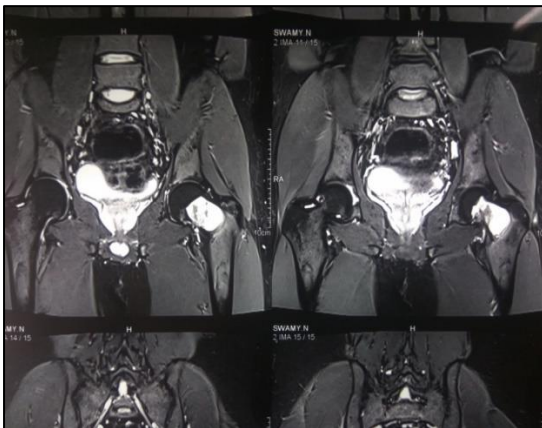


Figure 5: MRI of the patient.

One patient needed revision surgery 6 months after initial surgery due to structural failure of the implant (peri-implant fracture) operated with corrective osteotomy and IL-Nailing. One case operated with broad DCP showed increasing deformity and progressed to shepherd crook deformity which was reoperated with implant removal and corrective osteotomy and intramedullary nailing.

Revision-free survival was thus 85% for the whole cohort after 1 year.

Average neck shaft angle was increased from a preoperative 114 (92–130) to a postoperative 120.5 (100–130). FNSA did not significantly further change one year after surgery.



Figure 6: 18 months postoperative radiograph following curettage and fibular grafting and internal fixation with DHS.



Figure 7: Clinical photos of same patients at 18 months postoperative showing good functional outcome.

Complications

None of the 14 patients had neurovascular complications, complications related to the osteosynthesis or post-operative infections. There is no recurrence of the disease or postoperative infection in any of our cases in 1 year follow up. One patient operated with DHS needed revision surgery 6 months after initial surgery due to structural failure of the implant (peri-implant fracture) operated with intramedullary nailing. One case operated with broad DCP showed increasing deformity and progressed to shepherd crook deformity which was reoperated with implant removal and corrective osteotomy and intramedullary nailing at our institute.

DISCUSSION

FD of bone is characterized by the presence of fibro-osseous tissue in the bone with cortical widening and thinning.^{2,11} FD of proximal femur has a wide spectrum of clinical presentations, ranging from hip pain, limping due to deformity, or may present as a pathologic fracture.^{2,12,13}

In our study of 14 patients with Fibrous dysplasia of proximal femur, 7 were male and 7 female, similarly Tong et al, reviewed 15 patients, of which 9 were male and 6 female, and also in Kushare et al, who reviewed 23 patients, of which, 14 were male and 9 female.¹⁴ In our study, the mean age of the patients was 20 years, which was slightly lower than that of the patients in Tong et al¹⁴ (mean age of patients was 25 years) and Majoor et al (mean age of patients was 23 years).¹⁵

The mean MSTS score in our study was 21.6 compared to 27.6 in the study conducted by Ebeid et al. This difference is due to 3 polyostotic fibrous dysplasia cases in the study which have preoperatively poor function due to multiple bones involves bilaterally and also 2 cases has to undergo surgery twice due complication (refracture in one case and progressive deformity in other case).

Average neck shaft angle was increased from a preoperative 114 (92–130) to a postoperative 120.5 (100–130) where as average FNSA was corrected from $89^{\circ} \pm 20$ to $118^{\circ} \pm 13$ in the study conducted by Major et al.¹⁵ Revision-free survival was 85% for whole cohort compared to 97% in the study conducted by major et al at the end of 1 year.

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.^{2,16} 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 correct the deformity if present, and to prevent future morbidities such as pathologic fractures or deformities.¹⁶ Some authors believe that restoring the integrity of bone requires grafting.^{14,15} However, we have done fibular grafting only in cases where medial calcar was destroyed by the disease and done only internal fixation where calcar is intact.

In general, there is a fracture risk following lesion curettage and the initial structural support role may be obtained by fibular grafting at the defect site following proximal femur lesion curettage. However, the bone graft will lose its structural strength as the bone bonding process progresses. The additional application of internal fixation may provide a strong mechanical support for the reconstructed defect bone to obtain early stability, which facilitates maintenance of gravity line of lower limb and the early ambulation of patients (without load or with partial load). There are numerous optional internal fixation methods.^{17,18} For patients with an open osteoepiphyseal

line, the internal fixation of the anatomical plate was conducted in such a way as to avoid damaging the epiphyseal line.

In our case series, we have used DHS with fibular grafting in 6 cases, DHS alone in 3 cases, broad DCP in 2 cases and intramedullary nailing in 3 cases. In one case with shepherd crook deformity, proximal and distal realignment osteotomy done and proximal femur nailing was done. One patient operated with DHS needed revision surgery 6 months after initial surgery due to structural failure of the implant (peri-implant fracture) operated with intramedullary nailing.

In Nakashima et al and Onoda et al, intralesional curettage and bone grafting was the treatment option.^{19,20} Majoor et al performed cortical strut allografting for impending or actual proximal femoral fractures in 30 patients with FD.¹⁵ In Tong et al, 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 eight patients with proximal femoral FD were treated with fibular strut grafting and compression hip screw fixation.²² Jung et al treated eight patients with shepherd's crook deformity by multiple osteotomies and IM nailing. However, in our study, we have used fibular grafting in 6 cases with medial calcar deficiency.

Limitation of the study

This is being retrospective study, we do not have preoperative hip functional scores of the patients and the follow up is also 1 year, which may not be sufficient to interpret long-term results. These two are the limitations of the study.

CONCLUSION

Symptomatic fibrous dysplasia involving proximal femur needs to be operated to relieve pain due to biomechanical instability, to prevent and treat pathological fractures and to correct deformities. Internal fixation with Dynamic hip screw or cephalomedullary nail supplemented with fibular strut graft significantly improves functional outcome decreases pain and maintains neck shaft angle and was associated with lower revision rate.

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REFERENCES

1. Kushare IV, Colo D, Bakhshi H, Dormans JP. Fibrous dysplasia of the proximal femur: surgical management options and outcomes. *J Child Orthop.* 2014;8:505-1.
2. DiCaprio MR, Enneking WF. Fibrous dysplasia. Pathophysiology, evaluation, and treatment. *J Bone Joint Surg Am.* 2005;87:1848-64.
3. Coley BL. Neoplasms of Bone and Related Conditions: Etiology, Pathogenesis, Diagnosis and Treatment. 2nd edition. Paul B. Hober, Inc., New York. 1960;484.
4. Campanacci M. Bone and Soft Tissue Tumors: Clinical Features, Imaging, Pathology and Treatment. 2nd edition. Springer, New York. 1999;88-101.
5. Endres S, Wilke A. Fibrous dysplasia -differential diagnosis of cystic lesions in the proximal femur: a case report. *Cases J.* 2009;2:26-30.
6. Dorfman HD, Czerniak B. Fibroosseous lesions. In:Dorfman HD, Czerniak B (eds) *Bone tumors.* Mosby, St.Louis. 1998;441-91.
7. Collins MT, Bianco P. Fibrous dysplasia. In: Favus MJ (ed) *Primer on the metabolic bone diseases and disorders of mineral metabolism,* 5th edn. American Society for Bone and Mineral Research, Washington D.C. 2003;466-70.
8. Henschen F, Fallvon H. Osteitis fibrosa with multiple tumors in the musculature. *Verh Dtsch Ges Pathol.* 1926;21:93-7.
9. Mazabraud A, Girard J. A peculiar case of fibrous dysplasia with osseous and tendinous localizations. *Rev Rhum Mal Osteoartic.* 1957;24:652-9.
10. Stanton RP, Ippolito E, Springfield D, Lindaman L, Wientroub S, Leet A The surgical management of fibrous dysplasia of bone. *Orphanet J Rare Dis.* 2012;7(Suppl 1):S1-9.
11. Puls F, Niblett AJ, Mangham DC. Molecular pathology of bone tumours:Diagnostic implications. *Histopathology.* 2014;64:461-76.
12. Tsuchiya H, Tomita K, Matsumoto T, Watanabe S. Shepherd's crook deformity with an intracapsular femoral neck fracture in fibrous dysplasia. *Clin Orthop Relat Res.* 1995;310:160-4.
13. Keijsers LC, Van Tienen TG, Schreuder HW, Lemmens JA, Pruszczynski M, Veth RP. Fibrous dysplasia of bone: Management and outcome of 20 cases. *J Surg Oncol.* 2001;76:157-66.
14. Tong Z, Zhang W, Jiao N, Wang K, Chen B, Yang T. Surgical treatment of fibrous dysplasia in the proximal femur. *Exp Ther Med.* 2013;5:1355-8.
15. Majoor BC, Peeters-Boef MJ, van de Sande MA, Appelman-Dijkstra NM, Hamdy NA, Dijkstra PD. What is the role of allogeneic cortical strut grafts in the treatment of fibrous dysplasia of the proximal femur? *Clin Orthop Relat Res.* 2017;475:786-95.
16. Ippolito E, Bray EW, Corsi A. Natural history and treatment of fibrous dysplasia of bone: A multicenter clinicopathologic study promoted by the European Pediatric Orthopaedic Society. *J Pediatr Orthop B.* 2003;12:155-77.
17. Chapurlat RD, Huguency P, Delmas PD, Meunier PJ. Treatment of fibrous dysplasia of bone with intravenous pamidronate: long-term effectiveness and evaluation of predictors of response to treatment. *Bone.* 2004;35:235-42.
18. Jung ST, Chung JY, Seo HY, Bae BH, Lim KY. Multiple osteotomies and intramedullary nailing with neck cross-pinning for shepherd's crook deformity in polyostotic fibrous dysplasia: 7 femurs with a minimum of 2 years follow-up. *Acta Orthop.* 2006;77:469-73.
19. Nakashima Y, Kotoura Y, Nagashima T, Yamamuro T, Hamashima Y. Monostotic fibrous dysplasia in the femoral neck: A clinicopathologic study. *Clin Orthop Relat Res.* 1984;191:242-8.
20. Onoda S, Hatori M, Yamada N, Hosaka M, Kokubun S. A two-stage surgery for severe femoral neck deformity due to fibrous dysplasia: A case report. *Ups J Med Sci.* 2004;109:123-9.
21. Ebeid WA, Hasan BZ, Mesregah MK. Management of fibrous dysplasia of proximal femur by internal fixation without grafting: a retrospective study of 19 patients. *J Am Acad Orthop Surg Glob Res Rev.* 2018;2(10):e057.

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