Shepherd's Crook deformity with neck of femur fracture in fibrous dysplasia treated with valgus osteotomy, fibular strut graft and fixation with proximal femur locking plate – A Case Report.

Badaruddin Sahito[1], Dileep Kumar[2], Nauman Hussain[3], Noman Parekh[4], Asif Jatoi[5], Maratib Ali[6]

[1]Associate Professor
Dow University of health sciences/ Civil hospital Karachi
[2-4]Assistant Professor
Department of Orthopaedic Surgery, Dow University of health sciences/ Civil hospital Karachi
[5]Registrar Department of Orthopaedic Surgery
Civil hospital Karachi
[6]Chairman & Head of Department
Orthopaedic Surgery
Dow University of Health Sciences/ Civil Hospital Karachi

Authorship and contribution Declaration: Each author of this article fulfilled ALL Criteria of Authorship:
Conception of the case report, acquisition of data, Drafted the manuscript,
Revised the manuscript critically for important intellectual content, Final approval of the version for publication.

Correspondence Author:
Badaruddin Sahito
Email:
sahito.badar@hotmail.com

ABSTRACT

Fibrous dysplasia is a rare developmental abnormality of bone formation. The clinical presentation varies from asymptomatic disease to gross deformity and disability. Proximal femur bowing and coxa vara(Shepherd's Crook deformity) is frequently noted in polyostotic fibrous dysplasia. For treating Shepherd's Crook deformity but We treated our patient in a single stage surgery with valgus osteotomy, fibular strut grafting and proximal femur locking plate. At one year follow up she had excellent functional outcome. Our case report highlighted three important principles of managing fibrous dysplasia with pathological fracture neck of femur. Firstly, fixation of the fracture and accelerating bone healing with a fibular strut graft. Secondly, simultaneous restoration of the mechanical axis by correcting the deformity with valgus osteotomy. Thirdly, total hip replacement will be technically easy without needing further corrective surgery if the patient subsequently develops osteoarthritis or avascular necrosis of the femur head in future.

Key words: Fibrous dysplasia, Osteotomy, Shepherd's Crook deformity.

This article may be cited as: Sahito B, Kumar D, Hussain N, Parekh N, Jatoi A, Ali M. Shepherd's Crook deformity with neck of femur fracture in fibrous dysplasia treated with valgus osteotomy, fibular strut graft and fixation with proximal femur locking plate –A Case Report. J Pak Orthop Assoc. 2020;32(3)

INTRODUCTION

The term "Fibrous Dysplasia" was first introduced by Lichtenstein in 1938 while its pathological features were described by Jaffe in 1942.[1]It is a benign developmental pathology of bone and reported in 5-7% cases of benign bone tumours.[2] The normal bone and marrow is replaced by fibrous tissue and irregular woven bone.[3] Radiologically the affected bone reveals a lucent area surrounded by sclerotic rim with a granular ground glass appearance.[4] Although biopsy is rarely needed to confirm the diagnosis, MRI or CT scan are often needed to assess the extent of pathology and fatigue fractures.[5,6] The disease can involve any bone and my present in Monostotic (Solitary) or Polystotic (Multiple) forms with deformities and pathological fractures.[5] Proximal

femur is most frequently involved resulting in bowing, coxa vara and "Shepherd's Crook Deformity" in severe cases.[7,8] Rarely fibrous dysplasia can present as McCune-Albright syndrome and Mazabraud syndrome. McCune-Albright syndrome is a polyostotic fibrous dysplasia associated with skin pigmentation and endocrine pathologies,[9] while Mazabraud syndrome is associated with intramuscular Myxomas.[5] Malignant transformation to Osteosarcoma has been reported in 7% cases of polyostotic fibrous dysplasia.[10] Pathological fractures (mostly without any warning symptoms) have been reported in 20 to 40% cases of fibrous dysplasia.[11,12] The indications for surgery in fibrous dysplasia include sever pain, gross deformity and impending or pathological fracture.5 Various treatment options have been described in literature. These include curettage of the lesion and filling the defect with cancellous bone graft, fibular strut graft and allografts.[13,14] The goal of treatment in Shepherd's Crook deformity is to correct the deformity by restoring the mechanical axis, prevent the recurrence and achieve union at the fracture and osteotomy site.[5] We presented the management of an interesting case report of a 35 years old lady with monostotic fibrous dysplasia and Shepherd's Crook deformity with ipsilateral fracture neck of femur.

CASE REPORT

A 35 years old female, house wife presented to OPD of Civil Hospital Karachi with pain right hip for three and half months. Initially the pain was on and off but then persistent for the last one week. It was dull in character, not radiating but aggravated with walking and relived with rest and analgesics. There was no history of fever and weight loss. She had past history of fracture femur at age of 10 years which was treated conservatively at that time. On examination, she had a limping gait. Trendelenburg test positive on the right side and there was 3 cm true shortening o right side. Thomas test revealed 10 degree fixed flexion deformity of the right hip. Range of motion on right side was 10 to 120 degree, internal rotation 10 to 15 degrees, external rotation 20 -30 degrees, abduction 15-20 degrees and adduction 10 degrees. The opposite hip was normal. Distal neurovascular status was normal. Sensation and power was normal in both lower limbs. Spine was normal. Her X-ray right femur anterio-posterior and lateral view showed Shepherd's Crook deformity of proximal femur with neck shaft angle less than 90 degree. An osteolytic lesion in neck of femur with fracture was noted.(Fig. I) Magnetic Resonance Imaging(MRI) showed hyperintense lesion on T1 weighted image with neck of femur fracture.(Fig. II) Hematological investigations were normal.. Diagnosis of monostotic fibrous dysplasia with Shepahrd's Crook deformity made. The case was discussed at tumor board meeting of our hospital. The patient was counselled and consent taken for surgery and publication of her case report.

Under general anesthesia patient was placed on traction table and lateral incision was made from greater trochanter to mid shaft. Under C-arm control one guide wire was inserted into the neck to hold the fracture while another guidewire was used for triple reamer. After triple reaming the lesion in the neck was curated out of neck. Fibular strut graft harvested from the ipsilateral side was inserted into the neck to hold the fracture. Valgus osteotomy was made at the bowing site and proximal femur locking compression plate was used to fix the osteotomy and neck fracture.(Fig.III) Wound was closed in layers and dressing applied. Leg length discrepancy was corrected. Post operatively supervised physiotherapy was started in the hospital. Patient was called for follow up at 2 weeks and stitches were removed. Patient was allowed partial weight bearing at 6 weeks and full weight bearing at 10 weeks. At one year follow up the osteotomy site and fracture site was united (Fig.IV) and the patient was mobile without support. No recurrence or deformity was noted.We are regularly following this patient at monthly intervals till date. We reported our case report in accordance with CARE guidelines.[15]

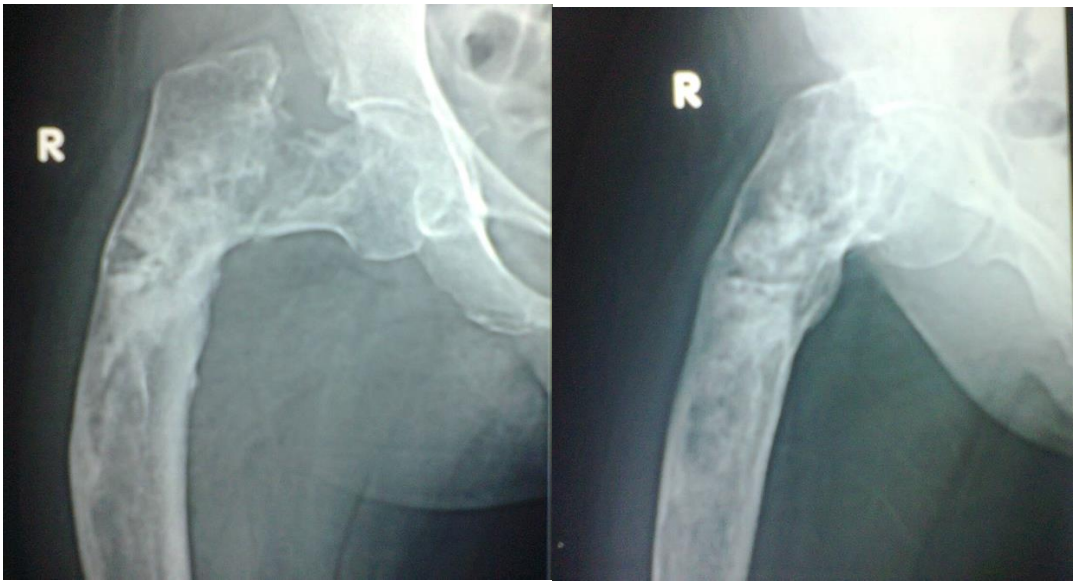

Fig. I: AP & Lateral radiographs showing Shepherd's Crook deformity and fracture neck of femur.

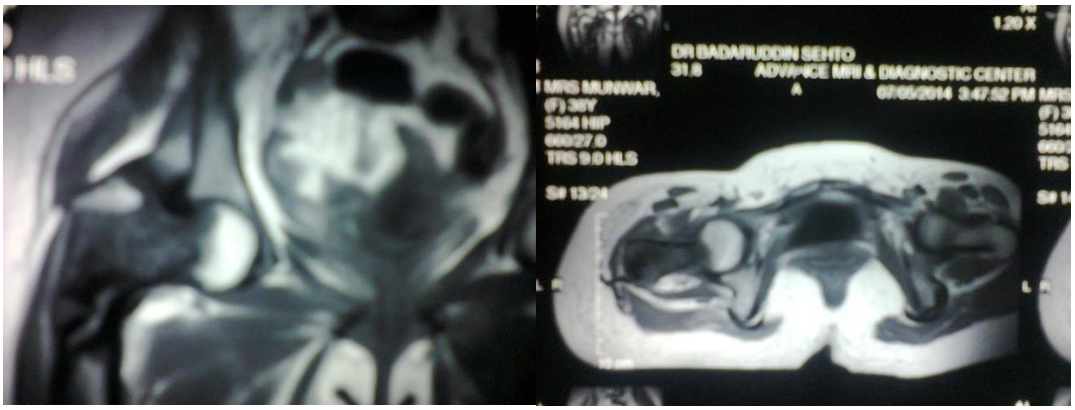

Fig. II: MRI: T1 weighted image coronal and axial image showing hypointense lesion in neck of femur with varus.

Fig. III: Immediate post op xray and xray at 3rd month post operatively.

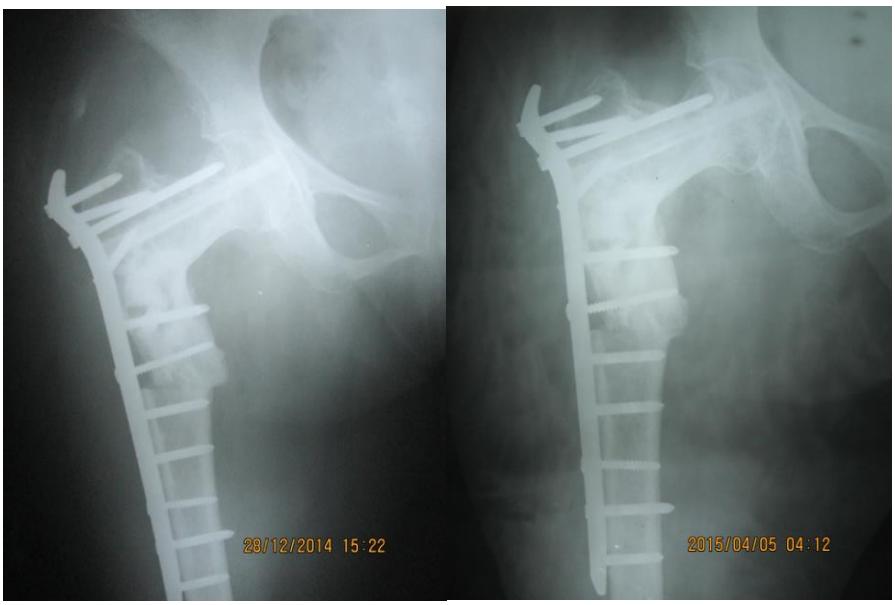

Fig IV: Xray at 6 months and one year follow up.

DISCUSSION

Although fibrous dysplasia is considered as a benign bone disease pathologically but clinically fibrous dysplasia involving lower limbs can cause significant morbidity resulting in decrease quality of life of the patient.[16]The Shepherd's Crook deformity may present at a different stage and in a different way thus warrants a "Customized" or "Tailored" approach to treat this deformity.[17] The paramount importance however should be given to achieve union with simultaneously correction of the deformity irrespective of the approach being used.[5]

We treated our patient of Shepherd's Crook deformity and ipsilateral pathological fracture neck of femur in a single stage surgery with valgus osteotomy, fibular strut grafting and proximal femur locking plate. Literature supports single stage fracture fixation and corrective surgery as it tend to decrease morbidity of the patient and helps in early rehabilitation.[18]

We used proximal femur locking plate because the quality of bone was poor and intramedullary nails were difficult to pass due to excessive bowing of the proximal femur.[17,18] Studies have shown that the pathology of fibrous dysplasia may persist after curettage but pathological fractures and osteotomies exhibited higher union rates.[8] In our case report the lesion of fibrous dysplasia extended into the neck of femur but spared the head providing us an opportunity to achieved excellent screw purchase in the head of femur. Similar observations were noted by Harris[19] and Freeman.[20]

In literature we could find very few case reports similar to our case report. Al-Mouazzen and colleague[6] reported a 23 years old man with Shepherd's Crook deformity and acute intra-capsular neck of femur secondary to fall. The authors fixed the fracture with proximal femur locking plate, allograft bone graft and sub trochanteric osteotomy. At one year follow up he had excellent functional outcome without any recurrence or complication. Tsuchiya and Tomita[21] successfully treated a 19 years old girl of Shepherd's Crook deformity and intracapsular fracture neck of femur with valgus osteotomy and fibular grafting. A very interesting case report of a 16 years old girl was presented by Kandhari and his colleagues[18] in 2015.The girl had bilateral Shepherd's Crook deformities and non union neck of femur fractures. They treated her with double osteotomies and dynamic hip screws. Right side was operated in a single stage followed by the left two months latter. At 18 months follow up the girl was fully weight bearing and without any recurrence or deformity. These authors recommended double osteotomies for the correction of deformities in patients of Shepherd's Crook deformity.

We presented our case report at the end of one year follow up and our patient was freely mobile and weight bearing without any support. There was no recurrence of the lesion or deformity and the fracture site and osteotomy site was completely healed. However, we recommend longer follow up of our patient to further confirm the effectiveness of our surgical approach.

CONCLUSION

Excellent functional and radiological outcome can be achieved in patients of fibrous dysplasia presented with Shepherd's Crook deformity and pathological fracture neck of femur when treated with valgus osteotomy, fibular strut graft and proximal femur locking plate. To prevent recurrence of the deformity the length of the locking plate must be adequate to span the lesion. Furthermore it is recommended to identify and treat sever coxa vara in patients of fibrous dysplasia to avoid pathological fracture neck of femur.

Conflict of Interests: None

Grants/Funding: None

REFERENCES

1. Nagda T V, Singh H, Kandoi M, Samant A, Patel BR. Two stage reconstruction for the Shepherd's crook deformity in a case of polyostotic fibrous dysplasia. J Postgrad Med. 1997;43:83
2. Lichtenstein L. Polyostotic fibrous dysplasia. Arch Surg.1938:36: 874-898.
3. Harris WH, Dudley HR, Barry RJ. The natural history of fibrous dysplasia. An orthopaedic, pathological, and roentgenographic study. J Bone Joint Surg Am. 1962;44:207-33
4. Ippolito E, Farsetti P, Boyce AM, Corsi A, DeMaio F, Collins MT. Radiographic classification of coronal plane femoral deformities in polyostotic fibrous dysplasia. Clin Orthop Relat Res. 2014;472(5):1558-67.
5. Harshavardhan JK, Uppalapati BR, Menon PG. A Case of Fibrous Dysplasia with Bilateral Shepherd Crook Deformity Treated with Dynamic Hip Screw Fixation. J Orthop Case Rep. 2018.8(3): 33-37.
6. Al-Mouazzen L, Rajakulendran K, Ahad N. Fibrous dysplasia, shepherd's crook deformity and an intra-capsular femoral neck fracture. Strategies Trauma Limb Reconstr.2013;8(3): 187–191.
7. Enneking WF, Gearen PF. Fibrous dysplasia of the femoral neck: Treatment by cortical bone-grafting. J Bone Joint Surg Am 1986; 68(9): 1415-1422.
8. Guille JT, Kumar SJ, MacEwen GD. Fibrous dysplasia of the proximal part of the femur. Long-term results of curettage and bone-grafting and mechanical realignment. J Bone Joint Surg Am. 1998; 80(5): 648-658.
9. Leet AI, Chebli C, Kushner H, Chen CC, Kelly MH, Brillante BA et al. Fracture incidence in polyostotic fibrous dysplasia and the McCune-Albright syndrome. J Bone Miner Res. 2004;19(4):571-7.
10. Ruggieri P, Sim FH, Bond JR, Unni LK. Malignancies in fibrous dysplasia. Cancer.1994;73:1411-24.
11. Saglik Y, Atalar H, Yildiz Y, Badarir K, Erekul S. Management of fibrous dysplasia A report on 36 cases. Acta Orthop. Belg.2007;73:96-101.
12. Kushare IV, Colo D, Bakhshi H, Dormans JP. Fibrous dysplasia of the proximal femur: surgical management options and outcomes. J Child Orthop. 2014;8(6):505–511.
13. Stanton RP. Surgery for fibrous dysplasia. J Bone Miner Res.2006;21(S2):105-109.
14. Wai EK, Davis AM, Griffin A. Pathologic fractures of the proximal femur secondary to benign bone tumors. Clin Orthop 2001; 393: 279-286.
15. David S. Riley DS, Barber MS, Kienle GS, Aronson JK, Schoen-Angerer TV, Tugwell P et al. CARE guidelines for case reports: Explanation and elaboration document. Journal of Clinical Epidemiology.2017;89:218-235.
16. Stewart MJ, Gilmer WS, Edmonson AS. Fibrous dysplasia of bone. J Bone Joint Surg Br.1962; 44:302-18.
17. Stephenson RB, London M, Hankin F, Kaufer H. 4Fibrous dysplasia. An analysis of options for treatment. J Bone Joint Surg Am. 1987; 69(3):400-9.
18. Kandhari VK,Bava SS, Desai MM, and Wade RN. Single Stage Treatment of Non-Union of Transcervical Neck Femur Fracture with Shepherd Crook Deformity of Proximal Femur in A Case of Fibrous Dysplasia using Dynamic Hip Screw Fixation. J Orthop Case Rep. 2015;5(3): 41-44.
19. Harris WH, Dudley HR Jr., Barry RJ. The Natural History of Fibrous Dysplasia - An Orthopaedic, Pathological and Radiographic Study. J Bone and Joint Surg.1962; 44A:207-233
20. Freeman BH, Bray EW, Meyer LC. Multiple Osteotomies with Zickel Nail Fixation for Polyostotic Fibrous Dysplasia Involving the Proximal Part of Femur. J Bone and Joint Surg 1987; 69A:691-698.
21. 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.