

# Shepherd's Crook Deformity with Pathological Femur Neck Fracture in a Patient with Fibrous Dysplasia Managed with Proximal Femoral Nailing and Valgus Osteotomy

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## ABSTRACT

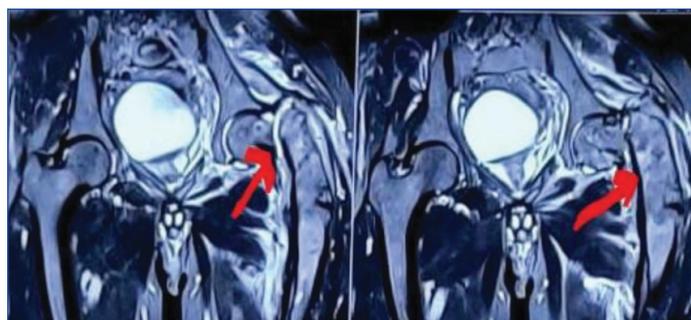
Fibrous dysplasia is a rare condition in which bone is replaced by fibroosseous tissue. It is a benign condition, divided into monostotic and polyostotic fibrous dysplasia, which accounts for about 5-7% of all benign bone tumours. Here, authors reported a case of a 38-year-old male that presented with left hip pain since 15 days and had a history of trauma. X-ray pelvis and Magnetic Resonance Imaging (MRI) showed shepherd's crook deformity in the left proximal femur with pathological femur neck fracture. It was managed with valgus osteotomy for deformity correction and fracture fixation using long proximal femur nail and cannulated cancellous screws. At one and half year follow-up, the patient showed good hip and knee range of motion with no complications.

**Keywords:** Benign bone tumour, Fibroosseous tissue, Metaphysis

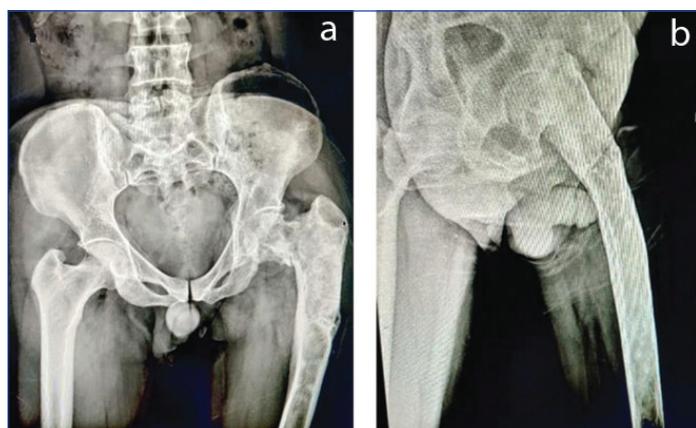
## CASE REPORT

A 38-year-old male reported with a complain of pain and swelling over his left hip since 15 days. The patient had a history of fall from height 15 days back following which he was not able to walk and sit cross legged. The patient visited a primary health center where radiographs were taken and traction was given. He was referred for further management. The patient had no co-morbidities like diabetes, hypertension, asthma or tuberculosis.

On physical examination, tenderness was present at the left hip region, left lower limb was externally rotated, shortened with no wound and distal neurovascular deficit. Plain radiographs of the left hip with femur anteroposterior and lateral views revealed pathological fracture of femur neck with shepherd's crook deformity with a lesion in proximal femur (expansile lytic, endosteal scalloping of the cortex and thick sclerotic reactive margin) suggestive of fibrous dysplasia [Table/Fig-1]. Lesions were also seen in the ipsilateral tibia and fibula. Findings were confirmed by Magnetic Resonance Imaging (MRI) [Table/Fig-2].



**[Table/Fig-2]:** MRI pelvis with both hips T2 weighted images showing ill-defined areas of altered marrow signal intensity in left femoral neck, proximal metaphysis and distal metaphysis. Associated with expansion in proximal femur with shepherd's crook deformity (varus angulation of proximal femur).



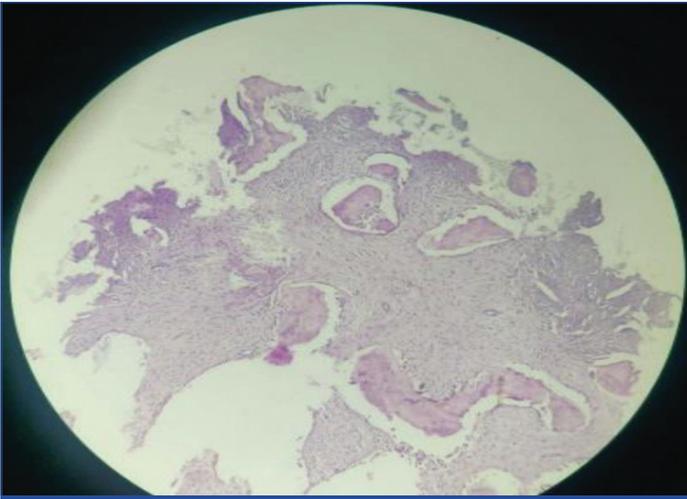
**[Table/Fig-1a-b]:** Preoperative radiographs showing femur neck fracture with shepherd's crook deformity, ground glass appearance and expansion of cortex.

The patient was young, requiring early mobilisation and weight-bearing, it was planned proximal femur nailing with cannulated cancellous screws and valgus osteotomy. The patient was operated under spinal anaesthesia, using a traction table in the supine position. Incision of around 5 cm was taken from the greater trochanter

extending proximally. The entry point was made over the tip of the greater trochanter after dissecting the soft tissues. The guide wire was passed in the medullary canal till the distal femur. The valgus osteotomy at the level of lesser trochanter was done to correct the varus deformity at the proximal femur. Gradual progressive reaming was done using cannulated reamers.

At the time of reaming, there was a lot of bleeding and difficulty in reaming. Long Proximal Femoral Nail (PFN) was inserted (size 11 mmx34 cm, 135° angle) with proximal and distal locking with two Cannulated Cancellous (CC) screws one with size of 6.5 mm in diameter and 85 mm in length and second with diameter of 6.5 mm and 65 mm in length. An intraoperative sample was taken from the medullary cavity and was sent for histopathological examination [Table/Fig-3]. The postoperative radiograph showed acceptable fracture reduction and correction of the shepherd's crook deformity [Table/Fig-4]. The patient was mobilised off the bed from the second postoperative day with nil weight-bearing. Histopathological examination confirmed the diagnosis of fibrous dysplasia with no evidence of malignancy. Partial weight-bearing was started at three weeks and full weight bearing at six weeks. The patient became completely symptomless by six months.

At one and half year's follow-up, the patient was comfortable with no pain and difficulty in walking and having good hip and knee range of motion. The follow-up radiographs showed union at the fracture site and osteotomy site [Table/Fig-5].



**[Table/Fig-3]:** Showing thin irregular trabeculae of bone in a fibrous background (H&E stain, 200x magnification).



**[Table/Fig-4]:** Immediate postoperative radiograph of left femur showing fixation of femur neck fracture with valgus osteotomy with bone grafting using long proximal femoral nail and cancellous cannulated screws.



**[Table/Fig-5]:** Follow-up radiograph at 1.5 years showing fracture union at the site of femur neck and osteotomy.

## DISCUSSION

Fibrous dysplasia is a rare condition in which bone is replaced by fibro-osseous tissue. It is a benign condition, divided into monostotic and polyostotic fibrous dysplasia, which accounts for about 5-7% of all benign bone tumours [1]. It equally occurs in males and females. It occurs throughout skeletal tissue but commonly occurs in long bones. Monostotic lesions are incidental findings, in some cases, only clinical observation is the treatment of choice. Polyostotic fibrous dysplasia most commonly involves the proximal femur, maxillofacial bone, and tibia [2]. The index patient also had lesions in ipsilateral tibia. The first symptom appears between the age of 5 to 20 years, extensive lesion is associated with the earliest symptoms. The pathological mechanism for fibrous dysplasia is not known but fibrous dysplasia has been linked to mutation in the *Gsa* gene located on chromosome 20q13.2-13.3 [3]. Polyostotic fibrous dysplasia commonly involves proximal femur and associated with shepherd's crook deformity.

Shepherd's crook deformity is a coxa varus deformity that is due to weight-bearing and stress exerted by the gluteal muscle. This varus deformity occurs in the first 4 to 5 years of age, particularly in the area of an intertrochanteric region, associated with limping, limb shortening, pathological fractures like the femur neck fracture, and chronic fatigue [4]. Shepherd's crook deformity with the neck of femur fracture can be managed with multiple osteotomies using a Proximal Femoral Nail (PFN), flexible nails, angulated blade plates with screws, and custom-made retrograde nail. Fixation using plates and screws has higher chances of refracture and deformities because stress shielding effect at the lower region of the plate. It also has difficulty to put plates due to deformities [5]. Long-term follow-up with intramedullary nails and flexible nails showed a high recurrence rate of deformity. Other adjuvant treatment options are allogenic cortical strut graft and bisphosphonate therapy [6]. Plating has higher chances of fracture at the lower site of plate because of stress shielding effect [4].

Kataria H et al., preferred the long PFN than multidirectional or reconstruction nails as the PFN had its entry point at the trochanter which was easily approached than piriformis fossa due to the varus deformity [6]. Also, the screws they used in fixation were of larger dimensions ensuring a better purchase in the femoral head. The PFN, having a long central lever arm, decreases the rate of recurrence. Gaski G et al., used intramedullary nailing without bisphosphonate for the pathological fracture with fibrous dysplasia in seven patients [7].

According to DiCaprio MR and Enneking WF cortical allograft or intramedullary fixation of the entire long bone is better when surgical correction is required to correct a deformity or in case of fracture [8]. Long proximal femur nail with subtrochanteric osteotomy which gives good functional outcome, provides stabilisation, has a low rate of deformity and refracture. Gupta V et al., reported their experience of treating a 21-year-old female with femur neck fracture with shepherd crook deformity using proximal femur nail and concluded that intramedullary implant for fixation is a better choice [9]. The intramedullary implant prevents occurrence of stress risers seen with an extra medullary implant at the bone implant interface.

Chaudhari T et al., reported a case of monostotic fibrous dysplasia of proximal femur treated with curettage and bone grafting and concluded that ideal treatment for lesions in proximal femur without shepherd's crook deformity and femur neck fracture is curettage and bone grafting [10]. George B et al., reported their experience of treating 17 patients with benign lesions of the proximal femur with non-vascularised, autologous fibular strut graft, without osteosynthesis [11]. They concluded that non vascularised fibular strut graft was safe and satisfactory method for benign lesions in proximal femur.

Connolly JF, reported the use of osteotomies with Zickel nail fixation [12]. Fibrous dysplasia should be distinguished from other proximal femur begin lesions, giant cell tumour, simple bone cyst, aneurysmal bone cyst and chondroblastoma [13]. In the present case, the deformity correction as well as fracture fixation was done in a single surgery, thus reducing the morbidity and risks associated with multiple-step procedure.

## CONCLUSION(S)

Femoral neck fractures in patients with fibrous dysplasia associated with shepherd's crook deformity are best managed by valgus osteotomy and intramedullary fixation using PFN. The procedure done in a single surgery decreases the morbidity and risks associated with anaesthesia in multiple surgeries, with good outcome.

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