

Fibrous Dysplasia of Proximal Femur: Management of the Complications

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Abstract

Fibrous dysplasia represents about 5% of benign bone lesions; however, the true incidence is unknown, as many patients are asymptomatic. Monostotic fibrous dysplasia accounts for 75-80% of the cases. It is caused by gene mutation [1,2]. Fibrous dysplasia is a slowly growing lesion that usually appears during periods of bone growth and is thus seen in those in early teen and adolescent years. Polyostotic fibrous dysplasia accounts for 20-25% of cases, and patients tend to present at a slightly earlier age (mean age, 8 y) [3]. Pregnancy can cause increased growth of the lesion as well as secondary changes of aneurysmal bone cyst formation. However, males and females are equally affected, although the polyostotic variant associated with McCune-Albright syndrome is seen more frequently in females [4].

Here we present the management and complications of a 23 years old female with complain of pain and swelling in Hip with pathological neck of femur fracture and shepherd's crook deformity due to fibrous dysplasia.

Introduction

The most common sites of skeletal involvement in monostotic fibrous dysplasia are the ribs, proximal femur, and craniofacial bones, typically the posterior maxilla. The lesion may involve only a small segment of bone or it may occupy its entire length. In polyostotic fibrous dysplasia, the spectrum of involvement varies from 2 bones to more than 75% of the skeleton. Polyostotic fibrous dysplasia is most commonly found in the femur, tibia, pelvis, and foot. Other sites less commonly affected include the ribs, skull, and bones of the upper extremity. Uncommonly affected bones include the lumbar spine, clavicle, and the cervical spine [2].

Incidentally discovered, asymptomatic, radiographically characteristic fibrous dysplasia lesions do not require further assessment and require only clinical observation. Follow-up radiographs every 6 months to look for progression has been recommended. In newly identified cases, a bone scan is needed to exclude a diagnosis of polyostotic disease. When polyostotic disease is found, referral to an endocrinologist for early detection of possible systemic abnormalities is warranted.

Bisphosphonates, primarily intravenous pamidronate, have been utilized to decrease bone pain in symptomatic patients with polyostotic disease [5,6]. Open biopsy may be indicated to confirm the diagnosis of fibrous dysplasia when there is a nonclassic presentation. Surgical procedures are required for correction of deformities, prevention of pathologic fractures, or eradication of symptomatic lesions [7,8].

Treatment of malignant transformation is based on the subtype of sarcoma, but the prognosis tends to be worse for patients with malignant transformation than it is for those with a similar primary sarcoma not associated with fibrous dysplasia (Figure 1).

Case Report

A 23 Year Old female reported to clinic of orthopaedics with complains of pain and swelling over left hip with inability to bear weight and restricted hip ROM. she was diagnosed with pathological fracture of neck of femur left side with shepherd's crook deformity due to fibrous dysplasia.

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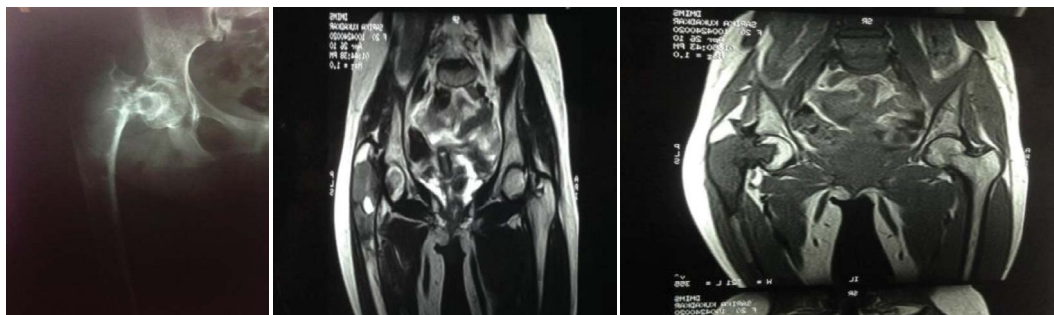


Figure 1: Pre-op X-ray Rt. Hip and MRI images.



Figure 2: Post-op X rays Rt. Hip shows valgus osteotomy with DHS plate osteosynthesis with healing of Fracture neck femur restoring neck shaft angle and limb length.

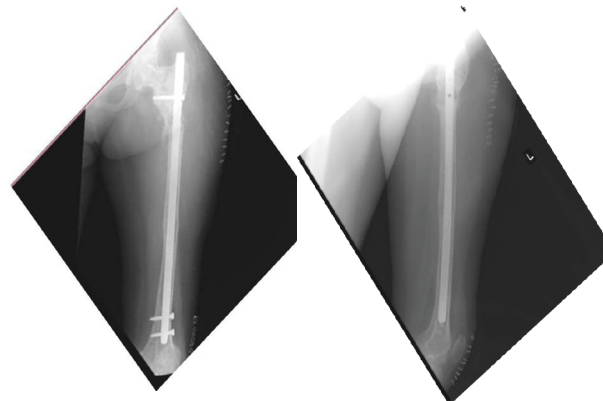


Figure 4: Interlock Nailing Done for proximal femur fracture.

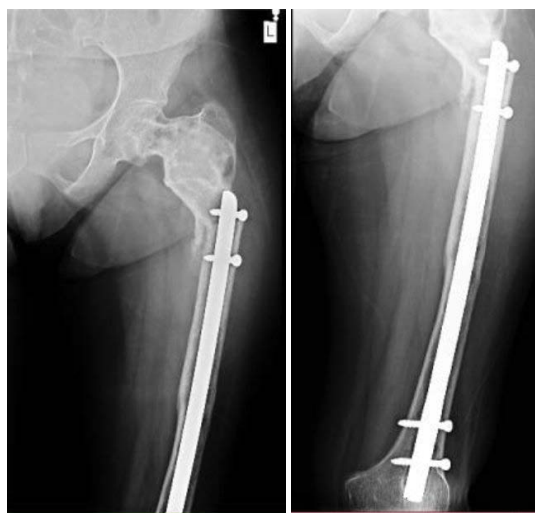


Figure 3: Fracture Proximal Femur Left Side.

Then patient reported in clinic of orthopaedics on 10/09/2014 with pain and swelling over left thigh. She was diagnosed with periimplant fracture femur left side and was managed with implant removal and open reduction and interlock nailing (retrograde) on 19/09/2014. Patient reported again on 03/08/2015 in orthopaedics clinic with complains of pain while walking and was diagnosed as fracture proximal femur left side (Figures 2 and 3). The patient was again operated, retrograde nail removed and interlock nailing done.

Discussion

Fibrous dysplasia is a rare bone disorder where normal medullary cavities of multiple bones are replaced by fibro-osseous tissue containing trabeculae of newly formed primitive bone [9]. Lesion of FD can affect the bones at all stages of growth process. Fibrous dysplasia lesions are more commonly found in proximal femur and tends to produce bowing and varus deformity due to constant muscle pull and body weight on the weakened bone [10]. The shepherd's crook deformity is a characteristics feature of FD presents with pain, limb shortening, limp and femoral neck fractures. Other common sites include the tibia, skull and ribs, although any bone can be affected [11]. Surgical treatment of FD has always been a challenge. The mainstay of surgical treatment is to restore normal alignment of the bone to attain normal walking ability and to provide pain relief secondary to pathological fractures. Several procedures have been advocated for treating proximal femoral lesions in fibrous dysplasia, including curettage and bone grafting, valgus osteotomy, plating and hip nailing, intramedullary nailing, and cortical bone grafting [12]. However, the type of intervention depends on many factors such as patient age, lesion characteristics (site, size and biological behaviour) and the presence of deformity [13]. Conservative modality such as second and third generation bisphosphonates plays a crucial role in maintaining the strength of the bone, pain relief and lowers the incidence of stress fractures which has been reserved for selected patients [14,15] (Figure 4).

The main cause for shepherd's crook deformity is the mechanical stress through the weak bone. So, it is paramount to

provide some mechanical support in form of internal fixation, although disease progression cannot be altered [16]. Breck [17] reported a case of fibrous dysplasia treated with total femoral plating and hip nailing, without further fracture or subsequent implant failure. Connolly [18] and Freeman et al reported the use of osteotomies with Zickel nail fixation [19]. There are various types of internal fixation for treating shepherd's crook deformity but none of them are superior to other. Moreover, deformity often spared the femoral head. So, firm purchase of implant in the femoral head provides sufficient mechanical support and prevent recurrence of the deformity [20]. The side plate should be long enough to provide adequate fixation of the mechanically deficient femur and to prevent the recurrence of deformity and implant failure. In our cases, fixation was achieved with an 8-hole side plate in 1, and a 6-hole side plate in the other. A gamma nail may be another option, because it can also provide good mechanical support over the femoral neck. In addition, it possesses a shorter level arm than a dynamic hip screw, and has a lower bending moment on the femoral neck. Initially, we tried to use a gamma nail to fix the osteotomy site in the second case. However, we found that the 12-mm diameter intramedullary nail and 2 distal screws were unable to provide adequate stability to the thinning and widened dysplastic bone. Moreover, a gamma nail is difficult to introduce, and may easily protrude from the canal because of the difficulty in locating a good entrance point and the deformed proximal femur [21]. As various studies have showed that fibrous dysplasia possesses a normal bone healing no bone grafting is not necessary for bony union. Therefore, we did not make any efforts to do an additional bone grafting in our present study [22,23]. In our case report the limitation of treating the patient was using extramedullary implant primarily for fixation rather we should have chosen intramedullary implant to prevent stress riser in dysplastic bone.

Conclusion

While treating fibrous dysplasia the condition of bone and stress riser should be considered and patient should be counselled about the consequences of the surgery. It's seen that intramedullary implant for fixation is better choice in such cases as they prevent the stress riser due to bone and implant interface on the cortex of the bone seen with extramedullary implant.

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