



OPERATIVE MANAGEMENT OF FIBROUS DYSPLASIA OF THE PROXIMAL FEMUR - A SINGLE CENTRE EXPERIENCE.

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Abstract : Introduction - Fibrous dysplasia is a benign condition of bone which can range from an innocuous lytic lesion to a complex deformity. The varied presentation necessitates an individualized approach. A number of surgical techniques and implants have been used in different series with satisfactory results however an ideal method of treatment is yet to be identified. Patients and Methods A retrospective study was conducted in the authors institution to review the management of fibrous dysplasia of the proximal femur. Over a 5 year period 6 males and 2 females with an average age of 28.6 years with biopsy proven fibrous dysplasia of the proximal femur were operated on for indications of (1) stabilization of impending or established pathological fracture, (2) deformity correction and (3) to arrest progression of a lytic lesion. The surgeries done were curettage and intra medullary nailing(3), corrective osteotomy and intra medullary nailing(2) and curettage and bone grafting(2). In one patient the surgery was abandoned due to intra operative hypotension. Results The average follow up was 21.8 months (6-50 months). The fractures and osteotomies united at a mean of 4.8 months. The complications encountered were one non union, sepsis and disseminated intravascular coagulation in one patient. One patient had incomplete correction of deformity while in the rest there was no recurrence of deformity. There were no wound related complications and all but one patient returned to work. Conclusions Curettage of the lesion followed by intra medullary nailing with appropriate osteotomies in case of a deformity is a satisfactory treatment for fibrous dysplasia of the proximal femur. It allows anatomical realignment and provides intra medullary splinting along the entire length of the femur decreasing stress shielding and recurrence of deformity. In view of the rarity of these cases and the perioperative issues a referral to a tertiary care centre with the necessary expertise and equipment would be prudent.

Keyword : Fibrous Dysplasia, Operative Management
INTRODUCTION

Fibrous dysplasia is a developmental anomaly of formation of bone, the hallmark of which is bone marrow replacement by fibrous tissue¹.

This results in thinning of the cortex threatening the structural integrity of the bone and can produce an isolated lytic lesion, a pathological fracture or a complex deformity². The varied presentation necessitates an individualized approach towards planning treatment. While the modalities of fixation may differ, the principles of anatomical realignment and internal splinting remain the same across the spectrum, of the disease.

PATIENTS AND METHODS

A retrospective study was undertaken to review the management fibrous dysplasia of the proximal femur. Between 2002 to 2007, 8 patients with biopsy proven fibrous dysplasia of the proximal femur were considered for the study. The medical records and operative notes were reviewed and status at latest follow up recorded. There were 6 males and 2 females with an average age of 28.6 years (19-52 years). Seven patients presented with pain in the affected limb while one presented with a chief complaint of deformity after having undergone deformity correction 16 years prior. Pathological fractures were present in 3 patients, coxa vara was present in 3 patients and the remaining 2 had lytic lesions in the neck of the femur. The co-morbid conditions present were idiopathic thrombocytopenic purpura, diabetes, hypertension and bronchial asthma. The procedures carried out were curettage and intra medullary nailing(3), corrective osteotomy and intra medullary nailing(2) and curettage and bone grafting(2). One patient with idiopathic thrombocytopenic purpura developed intra operative hypotension due to which the surgery was abandoned. The average blood required for the intra medullary nailing with or without corrective osteotomy was 4.5 pints. There was minimal blood loss when only a curettage and bone grafting was carried out. All patients had preoperative needle biopsies to confirm the diagnosis.

RESULTS

One patient underwent a prophylactic nailing and was subsequently asymptomatic. Of the remaining 7 patients one went into non union and the remaining united at an average of 4.8 months. The average follow up was 21.8 months (6-50 months). One patient who underwent a corrective osteotomy and intra medullary nailing required immobilization in a hip spica due to concerns regarding poor bone quality. He showed signs of union only at 20 months.

All except one patient returned to work. The complications encountered were sepsis and disseminated intravascular coagulation in one patient with underlying idiopathic thrombocytopenic purpura who developed intra operative hypotension. He experienced a stormy postoperative period requiring 20 transfusions of blood and blood products. He subsequently was managed non-operatively and was ambulant with crutches but was unable to return to his occupation. One patient had inadequate correction of the coxa vara but refused further surgical intervention. Of the rest who underwent surgery there was no recurrence of deformity. There were no wound related complications.

A number of techniques have been used in fibrous dysplasia including curettage and bone grafting, intra medullary nailing, plate fixation and external fixation¹⁻³. Ever since the first intra medullary fixation with a zickel nail by Connolly³ there has been a preference in the usage of intra medullary nail for its associated benefits^{1,6,7}. The use of blade plate devices for deformity correction has shown recurrence of deformity distal to the plate⁶. The varied picture at presentation accounts for the fact that no one standard surgical procedure could be adopted. The indications for surgery in this case series can be categorized into the

1. Stabilization for impending or established pathological fracture.
2. Correction of coxa vara and bowing deformity of the femur.
3. Curettage and bone grafting of lesion in structurally competent bone to arrest progression.

4. Combination of the above indications may be seen in some patients. Curettage of the pathological lesion is required^{1,5-7} prior to planning any further procedure. The general principles of anatomical realignment and intra medullary splinting can be applied to the first two indications acting as a broad guideline for management. In case of deformities appropriately planned and executed osteotomies will allow adequate correction. The type of intra medullary fixation will depend on the extent of the lesion and possible sites for interlock bolt placement. Intra medullary devices provide stability along the entire length of the femur, decrease stress concentration distally and in case of cephalomedullary nails also splint the neck^{1,6,7}. Tsuchiya et al⁵ reported the use of external fixation for deformity correction with minimal complications while preventing recurrent deformity and fractures. The Ilizarov ring fixator has the advantages of providing good stability and correction of deformities in multiple planes however it is technically demanding. The use of plate and screws is limited by the compromised hold provided by the pathological bone and recurrence of deformities distal to the fixation⁶. The plate also acts as stress riser and can result in fractures due to stress shielding. Operative management in fibrous dysplasia of the femur especially involving multiple osteotomies results in significant perioperative blood loss as have been shown in other series as well. The other operative issues faced are compromised fixation capacity of the bone¹, widened medullary canal^{1,2,7} and need for customized implants in extensive lesions⁸.

LIMITATIONS OF THE STUDY Given the retrospective nature of the study based on the available patient records and radiographs functional outcome assessment was not possible and the patients were not currently available for a detailed assessment of the functional outcome. The details on patient condition at follow up are based on the data available on the current preserved outpatient records.

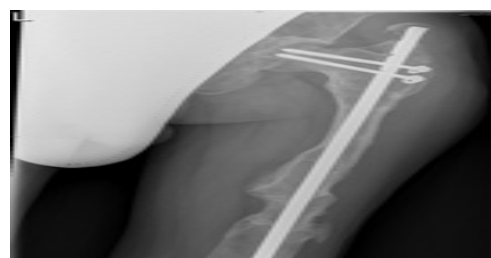
CONCLUSION Curettage followed by intra medullary nailing of proximal femur fractures is a reliable treatment modality for pathological fracture stabilization as well as deformity correction. The spectrum of clinical presentations allows only a general guideline for management and placement of osteotomies and implant specifications require individualized planning. The principles of anatomical realignment and intra medullary splinting holds good for most clinical situations. In view of the rarity of these cases and the perioperative issues a referral to a tertiary care centre with the necessary expertise and equipment would be prudent.

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Fibrous dysplasia left femur



Post Op



Fibrous dysplasia right femur



Post Op



Fibrous dysplasia left femur



Post Op



Post op



Fibrous dysplasia right femur



Fibrous dysplasia right femur with implant failure



Post implant exit and IM nailing