Intramedullary Nailing Of Fibrous Dysplasia Of Femur-A Case Report

Corresponding Author:
Dr. Ranat Vishnoi,
Resident, Padm .Dr. D.Y.Patil Medical College and Research Centre. Department Of Orthopaedics, 411018 - India

Submitting Author:
Dr. Prafulla G Herode,
Assistant Professor, Orthopaedics D Y Patil Medical College Pimpri Pune, 411028 - India

Article ID: WMC003882
Article Type: Case Report
Submitted on: 11-Dec-2012, 09:25:54 AM GMT  Published on: 11-Dec-2012, 01:41:53 PM GMT
Article URL: http://www.webmedcentral.com/article_view/3882
Subject Categories: ORTHOPAEDICS
Keywords: Fibrous Dysplasia, Multiple level ostetomy Kuntscher Nail.

How to cite the article: Joshi S , Herode PG, Vishnoi R, Bhamare D, Deokar BR. Intramedullary Nailing Of Fibrous Dysplasia Of Femur-A Case Report . WebmedCentral ORTHOPAEDICS 2012;3(12):WMC003882

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Source(s) of Funding:
Nil

Competing Interests:
The author(s) declare that they have no competing interests.
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**Author(s):** Joshi S, Herode PG, Vishnoi R, Bhamare D, Deokar BR

**Abstract**

**Introduction:** Fibrous dysplasia—a benign, nonfamilial disorder characterized by the presence of expanding intramedullary fibro-osseous tissue in one or more bones. The incidence of fibrous dysplasia is not known, but it is not an uncommon primary bone tumor. It occurs more frequently in girls than in boys.

**Case report:** A 14 year male who had experienced intermittent hip pain since childhood with progressive deformity in right thigh and limping. After a trivial fall he experienced severe pain with segmental fracture right femur, who responded well after osteotomy and internal fixation with kuntscher nail. Herein, we report this case for the rarity of its occurrence.

**Conclusion:** Bone curette and multiple level ostetomy internally fixed with kutscher nail can be kept in mind as a effective treatment modality for treating fibrous dysplasia of femur.

**Introduction**

The term fibrous dysplasia was originally proposed by Lichtenstein in 1938. He, along with Jaffe, McCune, and Albright, described this disorder of bone, as well as other extraskeletal abnormalities with which it is occasionally associated. Their descriptions remain among the best for fibrous dysplasia—a benign, nonfamilial disorder characterized by the presence of expanding intramedullary fibro-osseous tissue in one or more bones. The incidence of fibrous dysplasia is not known, but it is not an uncommon primary bone tumor. It occurs more frequently in girls than in boys, particularly the polyostotic form. Although most lesions are probably present in early childhood, they usually do not become evident before late childhood to adolescence. In general, fibrous dysplasia can be classified into one of three categories. Monostotic fibrous dysplasia involves only one bone, and many of these patients remain asymptomatic unless a fracture or swelling occurs. The polyostotic form is more severe, involving multiple bones. Nearly any bone in the body (in particular, one of the lower extremities) is more severely affected, resulting in deformity and limb length discrepancy. Craniofacial involvement occurs in nearly 50% of patients with polyostotic disease. The third category, polyostotic form with endocrine abnormalities, is the least common form. Precocious puberty, premature skeletal maturation, hyperthyroidism, hyperparathyroidism, acromegaly, and Cushing's syndrome can occur in these patients. The triad of precocious puberty (endocrinopathy), café-au-lait spots, and polyostotic bone involvement is commonly referred to as McCune-Albright (or Albright's) syndrome.

**Case Report(s)**

We present a case of 14 year male who had experienced intermittent bilateral hip pain since childhood with progressive deformities in both thigh and limping, presented to us with segmental fracture shaft femur right side following trivial trauma. X ray revealed segmental fracture of right femur with classical features of fibrous dysplasia in both femora[FIGURE 1], left tibia and right humerus[FIGURE 2]. After doing all the investigations patient was taken for surgery. On operation table supine position was given[FIGURE 3] and a Lateral Vastus splitting approach was taken after spinal anaesthesia. Incision was made upto whole length of the fracture level distally. Corrective osteotomy was done at two levels and retrograde guide wire was inserted and the deformity was correct to align the anatomical axis of femur. The medullary canal was found to be filled with gritty material which was curetted out leaving behind a thin rim of cortex. Retrograde reaming was done[FIGURE 4] and an intramedullary fixation with a kuntscher nail was done to maintain the reduction and to treat any further angular deformity. Material from fracture site was sent for histopathology study and it confirmed our diagnosis. Closure was done in layers and a cast was given after the removal of the sutures till G.T. for a month and patient was allowed to walk after with the support of Q brace[FIGURE 5]. Follow up x-ray after eight month show signs of union and good alignment.[FIGURE 6]
Discussion

Although most lesions are probably present in early childhood, they usually do not become evident before late childhood to adolescence or after a trivial trauma and tend to cease after puberty. After corrective osteotomy, the use of internal fixation devices such as plates that being a load bearing devices, place abnormal stresses at the end of the plate leading to failure. An intra medullary device could provide better stabilization. It prevents further deformity and protects the weak bone from failure as it is a load sharing device.

Conclusion

So this is a attempt to present that nailing as a safe, viable, stable alternative in the treatment of fibrous dysplasia of bone.

Abbreviations(s)

G.T – greater trochanter.

References

Illustrations

Illustration 1

Legend 1: Patient preoperative X-Ray.

![Patient preoperative X-Ray](image1)

Illustration 2

Legend 2: Patient Humerus X-Ray.

![Patient Humerus X-Ray](image2)
Illustration 3

Legend 3. Patient in OT in supine position.

Illustration 4

Legend 4. Intra operative retrograde reaming from osteotomy site.
Illustration 5

Legend 5. Patient walking with Brace.

Illustration 6

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