



FIBROUS DYSPLASIA WITH PATHOLOGICAL FRACTURE OF FEMUR - A CASE REPORT

Pranav Kothiyal., Puneet Gupta., Kunal Vij., Gaurav Menwal and Ankur Thakur

Department of Orthopaedics, SGRRIMHS Dehradun, Uttarakhand India

ARTICLE INFO

Article History:

Received 8 th, July, 2016,
Received in revised form 4 th,
August, 2016, Accepted 17th, September,
2016, Published online 21st, October, 2016

ABSTRACT

Background: fibrous dysplasia is a benign developmental abnormality of bone formation characterised by proliferation of spindle cells in bone and stroma. It can occur either in metaphysis or diaphysis and generally does not involve the epiphysis. If the lesion affects a single bone it is termed as monostotic and if it involves multiple bones it is termed as polyostotic. Here we present the case of a 17 year old female presenting with polyostotic fibrous dysplasia in bilateral femur and iliac bones with pathological fracture managed by intramedullary nail fixation.

Key words:

Fibrous Dysplasia", Pathological Fracture", "Intramedullary Nailing

Copyright © 2016 Pranav Kothiyal et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

INTRODUCTION

Fibrous dysplasia is a condition characterised by replacement of bone with fibrous tissue. It is a sporadic neoplastic lesion first described by Lichtenstein in 1938. Monostotic forms can often be asymptomatic but sometimes present with bony pains, pathological fractures of long bone or deformities.

Case report

A 17 years old female presented to us in the emergency department with the history of slip and fall on floor following which the patient had severe pain in her right thigh and was unable to bear weight on her right lower limb. The patient gave history of pain in her bilateral hip and right thigh since past one year. The range of motion was restricted in right hip and knee due to pain but was normal in left side. There was no history of endocrine dysfunction any signs of precocious puberty or any skin abnormalities. Clinically the patient was diagnosed to be having a fracture of right femoral shaft. X-rays of the pelvis and both hips and of the right thigh were done which showed fracture of the right femoral shaft through a expansile lytic lesion with ground glass appearance in the right femur (pathological). Also there were multiple expansile lytic lesions in both iliac bones and left proximal femur extending into the femoral neck. A MRI of the right thigh and pelvis with both hips showed multiple, expansile, multiloculated, multiseptate cystic lesions in bilateral iliac bones, neck and shaft of left femur and in the right femoral shaft. Other investigations were normal except for alkaline phosphatase that was raised with a value of 234 u/l and esr was raised with

a value of 53. The ultrasound of abdomen was insignificant except for a 3 mm calculus in the right renal cortex. The patient was managed operatively by closed reduction and internal fixation by intramedullary nail for the fracture of right femur. Intraoperatively the soft tissue was curetted from the fracture site by help of a reamer and collected for histopathological examination. Since the opposite femoral neck and proximal shaft also showed presence of similar expansile lytic lesion with cortical thinning a prophylactic fixation by a short intramedullary nail was done on the left side too. Curettage of the lesion in the left side was done from the greater trochanter and neck regions and the material was sent for histopathological examination. The histopathological examination showed immature bone formation in a background of fibrous stroma.

Postoperatively the patient was put on bisphosphonates 150 mg once a week along with high dose calcium and vitamin D3. Physiotherapy regimes for the knee and hip were started on third post day and the patient was allowed only partial weight bearing on the right lower limb for four weeks. The recovery was uneventful and at six months post op there are no fresh complaints of pain and the patient is ambulating freely without assistance.

DISCUSSION

Fibrous dysplasia is a benign well known condition affecting bones in which there is replacement of bone with fibrous tissue. The possible aetiology lies in developmental failure of immature bone to mature lamellar bone leading to the

formation of a mass of woven bone in abnormal fibrous tissue (1). This lesion accounts for approximately 7% of benign bony neoplasms and are seen in young patients with stoppage of growth after adolescence (2). However in case of McCune-Albright syndrome the lesions may continue to progress after adolescence.

The lesions in fibrous dysplasia render the bone morphologically and structurally fragile and the patients often present with bony pains and pathological fractures, hence it is important to follow the patients clinically and radiologically after the diagnosis is made even if no pathological fractures have occurred yet.



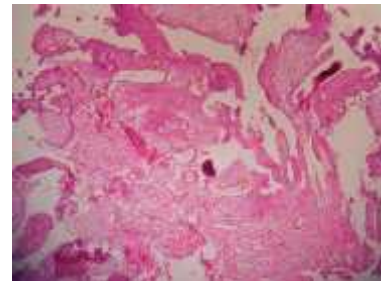
Preop X-rays and MRI showing pathological fracture and expansile lytic lesions



Fracture fixed operatively by intramedullary nail
On right side



Prophylactic nailing done on the other side



Histopathological specimen showing fibrous stroma with no mature bone

Conservative treatment is recommended for lesions that are present in the upper extremities and in patients with monostotic disease who are yet to achieve maturity. (3),(4),(5),(6)

Surgical management includes curettage and cancellous/corticocancellous bone grafting of large lesions and

internal fixation by a load sharing device like intramedullary nails or a load bearing device like a plate.

In the case reported here we fixed a pathological fracture of the femoral shaft that had occurred in a female patient who had almost achieved maturity with a intramedullary nail along with prophylactic nail in the opposite femur where a pathological fracture seemed imminent.

According to various studies conducted by Guille et al, Li et al, and Yang et al operative fixation done by intramedullary devices mostly without osteotomies, in patients with involvement of proximal femur having deformities and fractures were found to be satisfactorily managed. In case of severe shepherds crook deformities leading to marked coxa vara, osteotomy followed by plate fixation provided adequate treatment. (7), (8), (9)

In the case presented above fixation by intramedullary nails was done in both femurs, and it provided adequate treatment modality and the patient has had marked relief of pain and her recovery has been uneventful till now at one year postoperatively.

CONCLUSION

Thus by this case report we have attempted to show that in patients with symptomatic dysplastic lesions intramedullary nailing as a means of fixation provides adequate management and long term structural support to the weak bone.

References

1. W. F. Enneking, R. Rathe, and G. Cornwall, "Clinical musculoskeletal pathology," Office of Medical Informatics at the University of Florida, 1998, <http://www.medinfo.ufl.edu/cme/mspath>
2. M. R. DiCaprio and W. F. Enneking, "Fibrous dysplasia: pathophysiology, evaluation, and treatment," *Journal of Bone and Joint Surgery A*, vol. 87, no. 8, pp. 1848–1864, 2005. View at Publisher · View at Google Scholar · View at Scopus
3. Kusano T, Hirabayashi S, Eguchi T, Sugawara Y. Treatment strategies for fibrous dysplasia. *J Craniofac Surg* 2009; 20:768-70.
4. DiCaprio MR, Enneking WF. Fibrous dysplasia. Pathophysiology, evaluation, and treatment. *J Bone Joint Surg Am* 2005; 87:1848-64.
5. Parekh SG, Donthineni-Rao R, Ricchetti E, Lackman RD. Fibrous dysplasia. *J Am Acad Orthop Surg* 2004;12:305-13.
6. Chapurlat RD, Orcel P. Fibrous dysplasia of bone and McCune-Albright syndrome. *Best Pract Res Clin Rheumatol* 2008; 22:55-69.
7. 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;80:648-58
8. Li T, Xing B, Duan H, Liu Y, Pei F, Tu C. Surgical treatment for fibrous dysplasia of bone involving proximal femur. *Zhongguo Xiu Fu Chong Jian Wai Ke Za Zhi* 2009;23:261-4.
9. Yang L, Jing Y, Hong D, Chong-Qi T. Valgus osteotomy combined with intramedullary nail for Shepherd's crook deformity in fibrous dysplasia: 14 femurs with a minimum of 4 years follow-up. *Arch Orthop Trauma Surg* 2010;130:497-502. Back to cited text no. 8

