

## Treatment of Fibro Cartilagenous Dysplasia of Bilateral Proximal Femur: A Case Report

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### Abstract

*Fibro cartilagenous dysplasia is a variant of fibrous dysplasia in which extensive cartilage differentiation is seen. Fibrous dysplasia is an anomaly in skeletal development accounting approximately 5 to 7 percent of all benign bone tumors, resulting in defective bone forming mesenchyma leading to defective osteoblastic differentiation and maturation. It exists in two forms, monoostotic and polyostotic fibrous dysplasias whereas fibrocartilagenous dysplasia occurs commonly in polyostotic form. It commonly occurs in proximal femur at metaphyseal areas i.e. intertrochanteric regions, sub trochanteric region and neck. Due to repeated micro fractures at these sites various deformities like shepherd crook deformity can occur. It usually does not involve the epiphyseal region of head of femur. We are presenting a case of fibrocartilagenous dysplasia of bilateral proximal femora involving the neck and extending upto the articular surface of head of femur, which is an unusual site and bilateral total hip arthroplasty was performed in this case.*

**Keywords:** Fibrocartilagenous dysplasia, Fibrous dysplasia, monoostotic, polyostotic.

### INTRODUCTION:

We are presenting a case report of polyostotic fibrocartilagenous dysplasia involving left humerus and bilateral femora involving the neck and articular margin of both femoral heads which is an unusual site for fibrocartilagenous dysplasia, so the management of this case is different from the usual cases where the head is not involved and we only need to take care of neck and intertrochanteric regions by doing curatagge, bone grafting with or without prophylactic internal fixations. In some cases where the neck is highly deformed and various osteotomies may play some role. So we had to perform bilateral total hip arthroplasties in this case indicated to improve pain and function. To our knowledge this unusual presentation has not been reported in literature. There are also very few published reports regarding the arthroplasties in fibrous dysplasias of proximal femur and the indications of performing arthroplasties were also different.

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## **CASE REPORT:**

A 20 years old female patient came to us with history of progressive, continuous, dull aching pain in bilateral hip joints for last five years in right limb and for four years in left limb. The pain used to become severe after walking few steps and used to aggravate on prolonged standing or walking few steps. The relieving factors were taking medications and rest. The pain was associated with limp. The limp started two to three years after the start of pain in right hip. The patient was able to walk upto approximately fifty metres beyond which pain becomes so severe to restrict her from walking further. There was also history of pain in left shoulder for last two years. There was no history of precocious puberty or any menstrual dysfunction or any significant trauma.

On examination there was global decrease in the range of motion in bilateral hip joints with limb length inequality of 3 cms.

Plain radiograph of right hip showed typical shephard crook deformity with typical ground glass appearance involving neck and head of femur. The joint space was obliterated with loss of sphericity of head of right femur and there was acetabular dysplasia because of uncontainment of head of femur. Radiograph of left hip joint also showing ground glass appearance with loss of sphericity of head with decrease joint space. The head of left humerus also showed ground glass appearance. The radiograph of right knee showing genu valgum deformity of around 25 degrees which may be because of coxa vara of femur.

NCCT bilateral hip joint was done suggested either fibrous dysplasia or metabolic bone disease. Tc 99 MDP scan was done revealing hot margins at femoral and left humeral head with central cold spots. MRI bilateral hip joint suggested gross deformity and expansion b/l femora with associated hypo-intense region suggestive of fibrous dysplasia.

Core biopsy was taken from bilateral neck and head regions which was also suggestive of fibrous dysplasia.

All routine blood investigations were done. Additional investigations were done to rule out mc cune Albright syndrome like thyroid and parathyroid profile all serum hormone levels urine for free cortisol, ct and mri head to rule out pituitary adenoma.

So the diagnosis of bilateral fibrous dysplasia was made after the report of excisional biopsy revealing bony trabeculae without osteoblastic rimming with interspersed fibrous stroma, numerous mature hyaline cartilage nest area which is compatible with fibro cartilagenous dysplasia.

The patient was having pain, limp and limb length inequality of 3 cms so uncemented bilateral hip arthroplasty was performed, first in right hip followed by left after an interval of 6 months. Three weeks post operatively patient started walking with the aid of stick. There was minimal pain with good range of motion and with no limb length inequality. One year follow up has been done and the patient is walking without support and has no pain or limp. For fibrous dysplasia of right shoulder, we have kept the patient in routine follow up and she may require some surgical intervention like curratage with bone grafting later on.

## DISCUSSION:

The common site of involvement at proximal femora in case of fibrocartilagenous dysplasia is intertrochantricregion,subtrochantric region and neck of femur.The involvement of head of femur is very unusual that's why total hip arthroplasty is very rarely done in a case of fibrous dysplasia of proximal femur.The mainstay of treatment is curratage ,bone grafting ,internal fixations and osteotomies in most of the cases.In our case the head of femur was involved upto articular margins ,the patient was having pain ,decrease in range of motion and limp and the patient was young so we planned to perform total hip arthroplasty which was indicated to improve pain and function.

Total hip arthroplasty in fibrocartilagenous dysplasia is very challenging and various complications have been reported in literature.In case of fibrous dysplasia of proximal femur cemented total arthroplasty have preferred over non cemented total hip arthroplasty in literature because of high rate of stem loosening,but in our case the patient was only twenty years old that's why we performed non cemented total hip arthroplasty .the main difficulty we encountered in this case was the huge size of head and dysplastic acetabulum.peroperatively the capsule was very large so we had to remove the capsule,we created the acetabulum by reaming and fixed with screws to increase the stability of the acetabulum.we checked the limb length and stability preoperatively,which seemed to be good.

It is very difficult to say that after how many years, the revision surgery will be required but at present the patient is pain free with good functions.

Thus we concluded that the fibrocartilagenous dysplasia of proximal femur can involve any part from diaphysis to head of femur and if the head of femur is involved especially at weight bearing area,the functional disability and pain are inevitable .Thus the total hip arthroplasty is an excellent option to give the patient a better functional outcome and pain free hip joints.

## REFERENCES

1. Fletcher CDM, Unni KK, Mertens F : World health organization classification of tumors, Pathology and genetics of tumors of soft tissue and bone. IARC press, Lyon, 2002 : pg 341
2. Wagoner HA, Steinmetz R, Bethin KE, Eugster EA, Pescovitz OH, Hannon TS. GNAS mutation detection is related to disease severity in girls with McCune-Albright syndrome. *Pediatr Endocrinol Rev.* 2007;4 Suppl 4:395-400.
3. Vargas-Gonzalez R, Sanchez-Sosa S. Fibrocartilagenous dysplasia ( Fibrous dysplasia with extensive cartilagenous differentiation). *Pathology Oncology Research.* 2006;12(2):111-114
4. Ishida T, Dorfman HD. Massive chondroid differentiation in fibrous dysplasia of bone (fibrocartilagenous dysplasia). *Am J Surg Pathol.* 1993;17(9):924-30.
5. Kyriakos M, Mc Donald DJ, Sundaram M. Fibrous dysplasia with cartilagenous differentiation ("fibrocartilagenous dysplasia"): a review, with an illustrative case followed for 18 years. *Skeletal Radiol.* 2004;33(1):51-62.
6. Forest M, Tomeno B, Vanel D : *Orthopedic Surgical Pathology : diagnosis of tumors and pseudotumoral lesions of bone and joints.* Churchill Livingstone, Edinbourgh, pp. 595-612.
7. Hermann G, Klein M, Abdelwahab IF, Kenan S. Fibrocartilagenous dysplasia. *Skeletal Radiol.* 1996;25(5):509-11.
8. Mirra JM. Bone tumors. Clinical, radiologic and pathologic correlations. Philadelphia: Lea &Febiger, 1989.
9. Unni KK. Letter to the editor. *Am J Surg Pathol.* 1993;19:1447-48.
10. Ozaki T, Linder N, Blasius S. Dedifferentiated chondrosarcoma in albright syndrome. *JBJS.* 1997;79:1545-51.
11. Bhaduri A. Letter to the editor. Fibrocartilagenous mesenchymoma versus fibrocartilagenous dysplasia - are these a single entity? *Am J.SurgPathol.* 1995;19:1447.
12. Gedikoglu G, Aksoy MC, Ruacan S. Fibrocartilagenous mesenchymoma of the distal femur: case report and literature review. *Pathol Int.* 2001;51(8):638-42
13. Lichtenstein L. polyostotic fibrous dysplasia. *Arch Surg.* 1938;36:874-98
14. Coley BL. Neoplasms of bone and related conditions; etiology, pathogenesis, diagonosis and treatment. 2<sup>nd</sup> edition New York:Hoeber;1960

15. Cohen MM Jr ,Howell RE. Etiology of fibrous dysplasia and McCune Albright syndrome.Int j oral Maxillofac Surg.1999;28:366-71
16. Henry A. Monostotic fibrous dysplasia.J Bone Joint Surg Br.1969;51:300-6
17. HarrisWH, Dudley Jr., Barry RJ. The natural history of fibrous dysplasia.Anorthopaedic, pathological and roentgenographic study.Am J Orthop. 1962;44:207-33
18. Funk FJ Jr,Wells RE .Hip problems in fibrous dysplasia.ClinOrthopRelat Res. 1973;90:77-82.
19. EnnekingWF,Gearen PF .Fibrous dysplasia of femoral neck.Treatment by cortical bone grafting.J Bone Joint Surg Am.1986;68:1415-22
- 20.Guille JT ,kumarSJ,Macewen GD .Fibrous dysplasia of the proximal part of femur.Long term results of curettage and bone grafting and mechanical realignment.J Bone Joint Surg Am. 1998;80:648-58.
21. Huvos AG, Higinbotham NL, Miller TR. Bone sarcomas arising in fibrous dysplasia. J Bone Joint Surg Am.1972;54:1047-56.
22. Coley BL,Stewart FW. Bone sarcoma in polyostotic fibrous dysplasia.Ann Surg.1945;121:872-81
23. Chapurlat R, Meunier PJ. Fibrous dysplasia of bone. Baillieres Best Prac Res Clin Rheumatol.2000;14:385-98.
24. Lane JM, Khasn SN,O Connor WJ, NydickM,Hommel JP, Schneider R,Tomin E, Brand J, Curtin J. Bisphosphonate therapy in fibrous dysplasia. Clin OrthopRelat Res. 2001;382:6-12
25. Stephenson RB, London MD, Hankin FM, kaufner H. Fibrous dysplasia.An analysis of options for treatment. J Bone Joint Surg Am 1987; 69:400-9
26. Kumta SM, Leung PC, Griffith JF, Kew J, Chow LTVascularised bone grafting for fibrous dysplasia of upper limb. J Bone Joint Surg Br.2000;82:409-12
27. Freeman BH, Bray EW 3<sup>rd</sup>, Meyer LC. Multiple osteotomies with zickel nail fixation for polyostotic fibrous dysplasia involving the proximal femur .J Bone Joint Surg. Am.1987;69:691-8.
28. Ippolito E, Caterini R, Farsetti P, Potenza V. Surgical treatment of fibrous dysplasia of bone in McCune Albright syndrome.JPediatr Endocrinol Metab. 2002;15 Suppl 3:939-44.