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RESEARCH ARTICLE

MANAGEMENT OF MONOSTOTIC FIBROUS DYSPLASIA OF PROXIMAL FEMUR – A CASE REPORT

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ABSTRACT

Fibrous dysplasia is a developmental disorder of bone that can present in a monostotic or polyostotic form. Primarily affecting adolescents and young adults, it accounts for 7% of benign bone tumors. Many of the asymptomatic lesions are found incidentally; the remainder present with symptoms of swelling, deformity, or pain. Fibrous dysplasia has been associated with multiple endocrine and nonendocrinedisorders and with McCune-Albright and Mazabraud's syndromes. The etiology remains unclear, but molecular biology suggests a mutation in the Gsα subunit andactivation of c-fos and other proto-oncogenes. Fibrous dysplasia has characteristicradiographic appearance. Most cases do not require intervention, but those that dousually are managed surgically with curettage, bone grafting, and, in some cases, internal fixation. When some intervention is necessary but surgery is not practical, treatment is with bisphosphonates. The prognosis generally is good, although pooroutcomes are more frequent in younger patients and in those with polyostotic formsof the disease. The risk of malignant transformation is low. We report a case of unilateral monostotic fibrous dysplasia in a 19 year old female in a proximal femur with pathological neck fracture with Shepherd' crook deformity. The patient underwent a curettage of lesions combined with dynamic hip screw fixation and bone grafting. The patient was followed up in the outpatient department(opd),x rays was taken, signs of radiological healing was noted Partial weight-bearing was allowed at three months postoperatively and full weight-bearing at six months with no restriction in the activity. After six months, the patient was able to perform all activities without any difficulty. No evidence of recurrence was noted in the follow-up x-rays. After five years of follow up, implant exit done along with fibular strut graft done

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INTRODUCTION

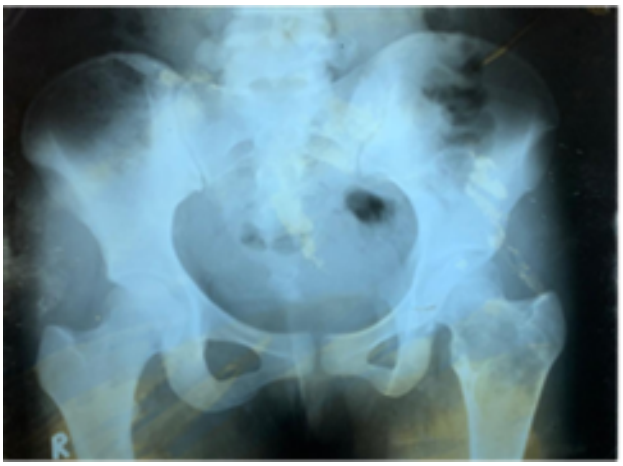
Fibrous dysplasia is a benign developmental disorder of the bone. Usually observed in adolescents and young adults, it accounts for 7% of benign bone tumors.3 In fibrous dysplasia, normal lamellar cancellous bone of the medullary canalis replaced with immature fibro-osseous tissue, resulting in poorly formed trabeculae of immature woven bone.4 Fibrous dysplasia may affect one (monostotic) or multiple (polyostotic) bones. Themonostotic form is more common, occurringin 75% to 80% ofcases. The polyostotic form can affect one (monomelic) or multiple (polymelic) extremities. Multiple endocrine disorders have been associated with fibrous dysplasia, typically with severe polyostotic fibrous dysplasia. These include hyperthyroidism, hypophosphatemia, acromegaly, hyperprolactinemia, and Cushing's disease.4,6-12 Nonendocrine abnormalities of the brain, thymus, heart, bone marrow, liver, spleen, and gastrointestinal tract are occasionally associated as well. Fibrous dysplasia is characteristic of McCune-Albright syndrome (polyostotic fibrous dysplasia, café-au-lait spots, and endocrine dysfunction) and Mazabraud's syndrome (polyostotic fibrous dysplasia and soft-tissue myxomas). The monostotic form is 7.6 times more frequent, but both variants show no predilection for gender . Pain is the most common clinical feature of the lesion if not associated with pathological fracture. The severity of pain was higher in patients with lesions of the lower extremities and ribs compared with upper extremity or craniofacial lesions. Other forms of FD (polyostotic/McCune-Albright syndrome) were more often associated with pain, often severe . The clinical spectrum of the disease ranges from simple bone enlargement or bone pain to pathological fractures and deformities of bone. Monostotic FD cases are often asymptomatic, which can be followed periodically with assessment for new symptoms and radiographs or sometimes may be detected as an incidental finding. Treatment is not required in asymptomatic cases. The mainstay of treatment for FD is surgical, but antiresorptive medication (bisphosphonates, anti-receptor activator of nuclear factor-kappaB ligand [anti-RANKL] antibody) aims at decreasing the local increase in bone turnover in the management of FD, thereby potentially decreasing or preventing the expansion of lesions, controlling the symptoms, and decreasing the risk for deformities and fractures if detected at an early stage.

Surgical management is as outlined by Enneking *et al*., which involved curettage of lesion and using bone graft to fill the defect for prevention of fracture and deformity. A detailed guide to treating FD with deformity with fracture of neck femur was published by Dheenadhayalan *et al*. Recurrence of lesions can occur if the lesion is managed only with curettage and bone grafting. We report a case of unilateral monostotic FD in a 19-year-old female with shepherd's crook deformity along with pathological neck of femur fracture managed with curettage with combined dynamic hip screw (DHS) and fibular strut graft.

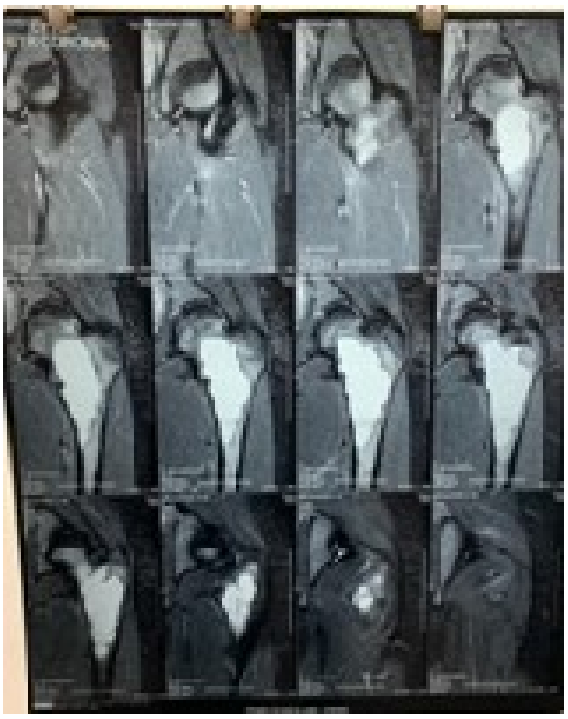
CASE PRESENTATION

19 year old female came to opd with c/o pain in left hip for past 3 months,c/o difficulty in using left lower limb.on examination, Tenderness + over greater trochanter, movements restricted. After a thorough clinico-radiological examination of the patient, the diagnosis of FD with Shepherd's crook deformity hip was established. There was no evidence of any endocrine disturbance on metabolic bone profile evaluation or altered pigmentation. The patient underwent a curettage of lesion and combined with Dynamic Hip Screw and bone grafting. After regular follow up period of 5 years, implant exit done with fibular strut graft.

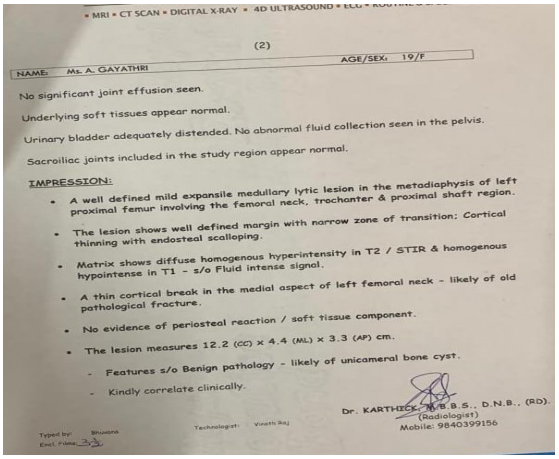
PREOP RADIOGRAPH



MRI IMAGES



MRI REPORT



= MRI = CT SCAN = DIGITAL X-RAY = 4D ULTRASOUND =

(2)

NAME: Ms. A. GAYATHRI AGE/SEX: 19/F

No significant joint effusion seen.

Underlying soft tissues appear normal.

Urinary bladder adequately distended. No abnormal fluid collection seen in the pelvis.

Sacroiliac joints included in the study region appear normal.

IMPRESSION:

- A well defined mild expansile medullary lytic lesion in the metadiaphysis of left proximal femur involving the femoral neck, trochanter & proximal shaft region.
- The lesion shows well defined margin with narrow zone of transition; Cortical thinning with endosteal scalloping.
- Matrix shows diffuse homogenous hyperintensity in T2 / STIR & homogenous hypointense in T1 - s/o Fluid intense signal.
- A thin cortical break in the medial aspect of left femoral neck - likely of old pathological fracture.
- No evidence of periosteal reaction / soft tissue component.
- The lesion measures 12.2 (CC) x 4.4 (ML) x 3.3 (AP) cm.
- Features s/o Benign pathology - likely of unicameral bone cyst.
- Kindly correlate clinically.

Dr. KARTHICK, M.B.B.S., D.N.B., (RD). (Radiologist) Mobile: 9840399156

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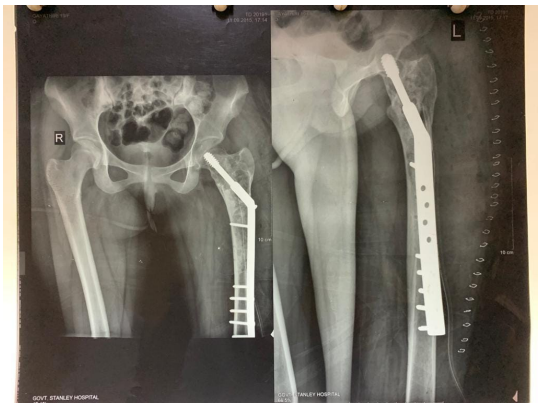
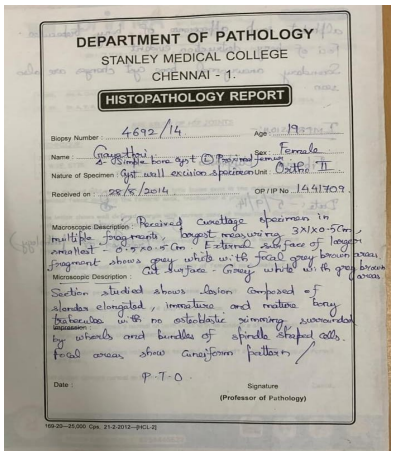
IMMEDIATE POST OP RADIOGRAPH



BIOPSY REPORT



DEPARTMENT OF PATHOLOGY
STANLEY MEDICAL COLLEGE
CHENNAI - 1.

HISTOPATHOLOGY REPORT

Biopsy Number: 4692/14 Age: 19

Name: Gayathri Sex: Female

Nature of Specimen: Cyst wall excision specimen Unit: Ortho II

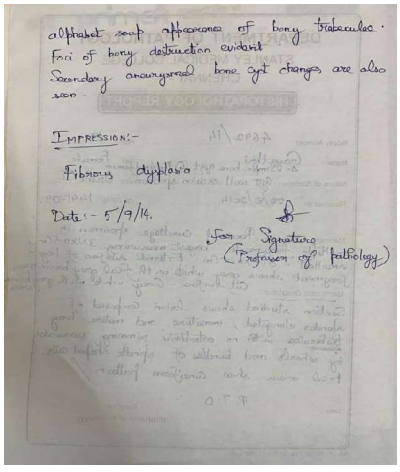
Received on: 28/8/2014 OP/IP No: 1441709

Macroscopic Description: Received curettage specimen in multiple fragments, largest measuring 3x1x0.5cm, smallest 2x2x0.5cm. External surface of largest fragment shows grey white with focal grey brown areas. Cut surface - Grey white with grey brown areas.

Microscopic Description: Section studied shows lesion composed of slender elongated, immature and mature bony trabeculae with no osteoblastic rimming surrounded by fibrous and bundles of spindle shaped cells. Focal areas show Giant form cells.

P.T.O.

Date: Signature (Professor of Pathology)



alphabet soup appearance of bony trabeculae. Foci of bony destruction evident. Secondary aneurysmal bone cyst changes are also seen.

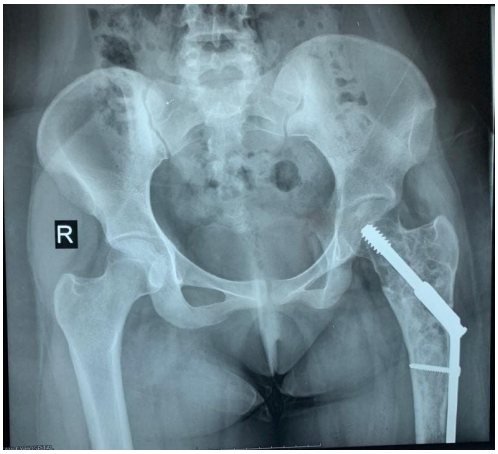
IMPRESSION:-

Fibrous dysplasia

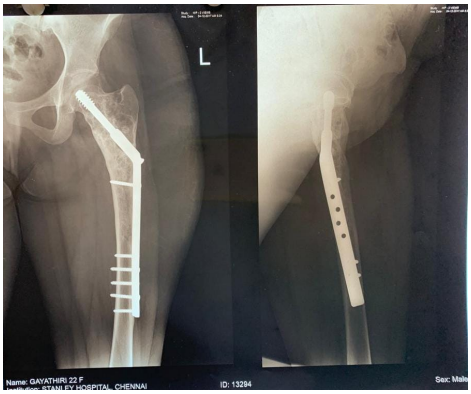
Date:- 5/9/14.

Signature (Professor of Pathology)

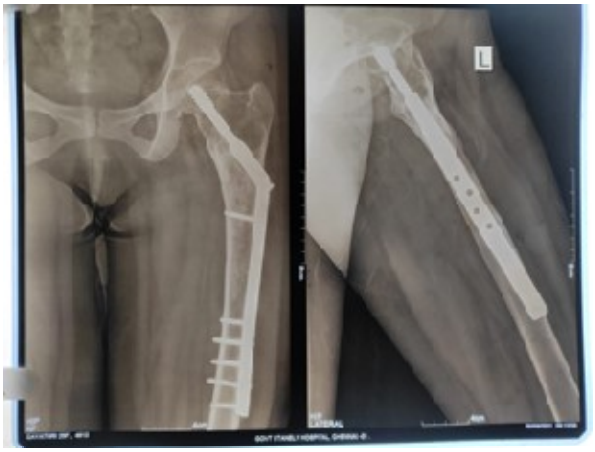
6 MONTH POST OP RADIOGRAPH



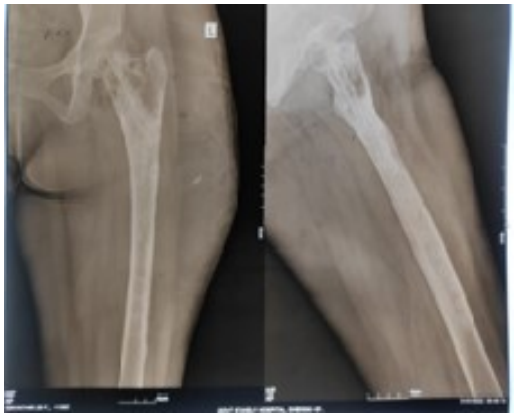
1 YEAR POST OP RADIOGRAPH



5 YEAR POST OP RADIOGRAPH



POST IMPLANT EXIT RADIOGRAPH



CLINICAL PICTURES

DISCUSSION

FD being a benign lesion may go undetected and may present as a deformity like a shepherd's crook as observed in our patient, which was affecting the quality of life and function of the joint. The presentation in our patient was in the third decade, and the patient had neglected his symptoms for six months. The patient was walking full weight-bearing in spite of his injury with a limp. Each Shepherd's crook deformity presents in a different way and with a variety of challenges, thus demanding patient-specific approaches. Customized approaches are needed to achieve correct alignment so as to obtain optimal functional outcomes and better quality of life for the patient.

In our case, Shepherd's crook deformity was associated with a pathological neck femur fracture, so it warranted the use of a dynamic implant like a DHS with a vascularized fibula graft. DHS provides controlled collapse of fracture at the proximal femur, which was adequately supported by the fibular strut graft. The single-stage correction of Shepherd's crook deformity helps in early rehabilitation and decreases patient morbidity.

CONCLUSION

The use of DHSs allows for collapse at the fracture site and spans the barrel plate according to the lesion. Cortical strut graft has also been used to additionally support the neck femur fracture. Single-stage surgery has the advantage of early rehabilitation and better patient outcomes. Good curettage with proper mechanical alignment also reduces the chances of recurrences.

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