

Case Report

Monostotic Fibrous Dysplasia of Proximal Femur with a Pathological Fracture: A Case Report

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ABSTRACT

Fibrous dysplasia is a congenital, non-hereditary anomaly of bone formation that may exist in monostotic or polyostotic form. It is characterized by replacement of normal bone with fibrous tissue which makes the bone weaker and prone for pathological fractures. We present a case of monostotic fibrous dysplasia of proximal femur with pathological fracture treated with curettage, bone grafting and dynamic hip screw for internal stabilization.

Key words: Fibrous dysplasia, Femur, Bone graft.

INTRODUCTION

Fibrous dysplasia is a benign fibro-osseous pathological condition characterized by the replacement of bone with fibrous tissue due to focal or generalized inability of bone forming tissue to form mature lamellar bone. The lesion was first described by Lichtenstein in 1938. [1] If fibrous dysplasia affects only one bone, it is called monostotic, but if multiple bones are affected then it is called polyostotic type. [2] In addition to these forms, Jones described hereditary familial form of localized mandibular fibrous dysplasia which is called cherubism. [3] Because fibrous dysplasia causes cortical thinning and expansion of the cortex concentrically or eccentrically, the lesions may resemble unicameral or multilocular bone cysts. [2] When fibrous dysplasia involves the proximal part of

femur, it results in weakening of the bone which may lead to progressive microfractures and is referred to as a "Shepherd's crook deformity". Patients with pathologic fractures or progressive skeletal deformity require curettage, bone grafting, and skeletal stabilization.

CASE HISTORY

A 47 years old female, housewife by occupation, came to the outdoor patient department of our hospital in rural set up with complaints of pain in her left hip and inability to walk since 2 weeks after she had a sudden jerk and twist to her left hip region. On clinical examination the patient had swelling in left hip region and external rotation of left lower limb with abnormal mobility in the subtrochanteric region. A radiograph of pelvis with both hips revealed

a well circumscribed area of cystic lesion in the trochanteric region of femur just below neck with a subtrochanteric fracture of femur. MRI evaluation of pelvis with both hips revealed well defined well circumscribed area of 6 cm × 3 cm × 2 cm of altered marrow signal in upper femoral diaphysis of left side just below neck mostly suggestive of benign neoplastic lesion like fibrous dysplasia. Patients did not have any similar lesions in other bones. Basic hematological investigations were within normal limits. During the surgery patient was in supine position under spinal anesthesia on a fracture table. The fracture site exposed with standard lateral approach. A guide wire was passed in the central part of neck and its position was confirmed on C-arm. Two small cortical windows were made just superior and inferior to guide wire entry point in the lateral cortex. With the help of curette, the fibrous tissue was curetted out completely. Cavity was packed with cortico-cancellous bone grafts from left iliac crest and G bone. A dynamic hip screw was inserted over the guide wire. A 135 short barrel plate with six holes was fixed to dynamic hip screw and appropriate size 4.5 mm cortical screws were drilled, tapped and inserted in the last five holes of plate. The incision was closed in layers with sutures over a suction drain. Intravenous antibiotics were given for three days and then oral antibiotics for one week. Patient was asked to do static quadriceps exercises as soon as anesthesia effect worn off. Suction drain was removed on post operative day two and sutures were removed after ten days. Patient was ambulated after removal of drain with help of walker without bearing weight on the operated limb for two weeks and then with toe touch weight bearing for another four weeks. Patient is doing very well on

follow up and is able to do her daily household activities bearing full weight on her left lower limb.

Microscopy: bony tissue shows curvilinear trabeculae of woven bone, giving fish hook appearance. Bone lacks osteoblastic rimming. Bony tissue is surrounded by moderately cellular fibroblastic proliferation.

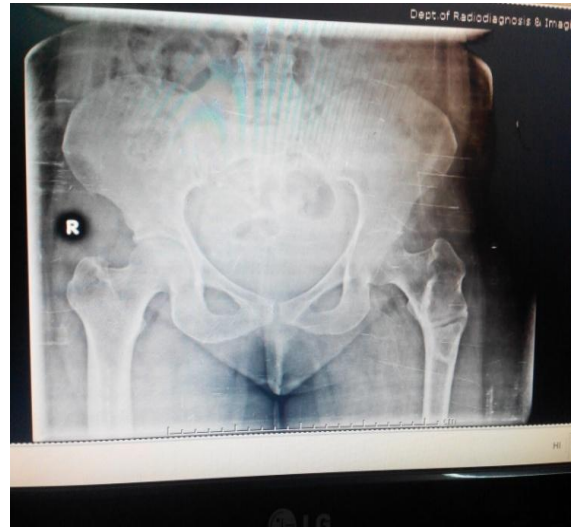


Fig 1: pre operative X-ray shows osteolytic lesion with pathological subtrochanteric fracture in left side



Fig 2: post operative X-ray shows DHS fixation with bone graft and G bone

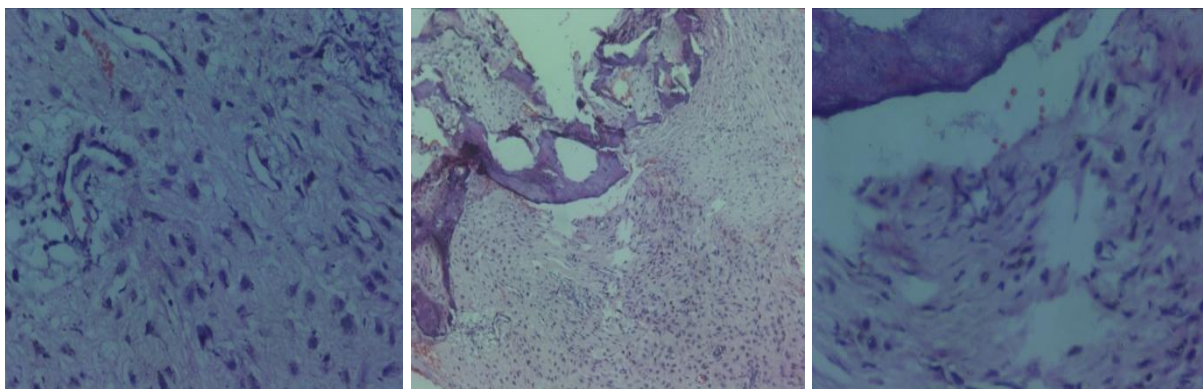


Fig 4: micro photograph showing slides of biopsy tissue and fibrous tissue

DISCUSSION

Fibrous dysplasia is a relatively rare condition with unknown etiology characterized by fibrous tissue replacement of the skeleton in which irregularly distributed spicules of bone lie in cellular fibrous stroma. The lesion is an abnormality of bone believed to occur because of development of hamartomatous tissue in the normal bone tissue. [4,5] The lesions are localized on the epiphysis, metaphysis or diaphysis of long bones. There is no sexual predilection. The monostotic variant has high prevalence. [2] It is usually diagnosed during the patient's growth phase and asymptomatic till pathological fracture occurs. It affects the femur, ribs, tibia, jawbone and Humerus. [1,6] When fibrous dysplasia involves the proximal femur, the resulting weakening of the bone may lead to progressive micro fractures and is referred to as a "Shepherd's crook deformity". [7]

The polyostotic form is observed in 30% of cases. It is usually diagnosed during the patients' infancy. It affects the cranium, face, pelvis, spine and shoulder. It is associated with the McCune-Albright syndrome in 2% of cases (fibrous dysplasia, skin pigmentation and precocious puberty) [4,8]

The monostotic variant is most of the time asymptomatic and is an incidental finding on radiographs. Pain and swelling are the other symptoms at the site of the lesion. Pregnancy can aggravate symptoms in females. This tumour can also present as a pathological fracture in a long bone that is mostly followed by a nonunion or malunion. [1,2,6]

Plain radiographs show circumscribed areas of decreased bone density having a ground glass or shower door glass appearance; a pattern resembling orange skin (peaud' orange), wool like appearance, or an amorphous, dense pattern. Early lesions may be more radiolucent than mature lesions and in rare cases may appear to have granular internal septa, giving the internal aspect a multilocular appearance. [9]

Endosteal scalloping of the adjacent cortex may be noted. Bone scans typically reveal increased uptake of radioisotope in the lesions. A cold bone scan does not exclude the diagnosis.

Bone scans are most useful for identifying the distribution of lesions in patients with polyostotic disease. On MRI, the bone lesions have decreased signal on T1-weighted images and variable signal intensity on T2-weighted images. [10]

Microscopically, lesions are composed of a fibrous stroma that is acellular and avascular. Fiber bone trabeculae are evenly spaced in a dense mature collagenous tissue. The bone develops by metaplasia from the collagenous tissue and there is absence of osteoblasts rimming the trabeculae. [11]

Asymptomatic bone lesions can be observed for any progression. The primary goal of treatment is to realign the deformed bone, particularly in weight bearing lower extremities. [12] If fractures occur in long bones, particularly proximal femur, surgical intervention is the preferred treatment in form of curettage, bone grafting and internal fixation by intramedullary rods or compression screws with side plates. [7,13,14]

Progressive deformity still might occur after adequate internal fixation and proper alignment of bone.

CONCLUSION

On a concluding note we suggest that an ideal treatment for a case similar to this is complete curettage of the lesion, adequate autologous bone grafting and stable internal fixation to be followed by close follow up, radiographic evaluation on a regular basis and non weight bearing mobilization of the patient till the consolidation of the lesion and uptake of graft is complete.

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