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Management of pathological fracture of the humerus secondary to osteofibrous dysplasia after curettage with elastic intramedullary nail: A case report

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Abstract
Introduction: Fibrous dysplasia is a rare non neoplastic bone disease of unknown aetiology which has a wide spectrum of symptoms such as pain, deformity, fracture depending on the area of involvement. Cases are diagnosed within the first three decades of life, though most commonly diagnosed in children and adolescents. Fibrous dysplasia presents in two forms monostotic or polyostotic. We present a case of pathological fracture of humerus secondary to fibrous dysplasia admitted in department with a background of posttraumatic pain in the right arm.
Conclusion: Pathological fracture of humerus in a child secondary to Fibrous dysplasia can be managed with curettage and bone grafting along with elastic intramedullary nail.

Keywords: Osteofibrous dysplasia, humerus, elastic intramedullary nail

1. Introduction
Fibrous dysplasia of bone is a rare disease often associated with severe clinical outcome, which ranges from bone pain to pathological fractures [1]. They represent 5-7% of all benign bone tumours [2]. Fibrous dysplasia presents in monostotic form which affects one bone or in polyostotic form which affects several bones. Fibrous dysplasia is usually found in the proximal femur, tibia, humerus, ribs, and craniofacial bones in decreasing order of incidence. Cases are diagnosed within the first three decades of life, most commonly in children and adolescents. Monostotic fibrous dysplasia may be completely asymptomatic and is often an incidental finding on x-ray. Pain and swelling at the site of the lesion can also be present. This tumor can also present as a pathological fracture that may go into a nonunion or malunion. It is the advised for intramedullary fixation with the strongest possible device is the best method for treatment FD lesions. We present a case of pathological fracture of humerus secondary to fibrous dysplasia, admitted in department with a background of posttraumatic pain in the right arm.

2. Case Report
13 year old male was admitted in our department with complaints of right humerus pain since last 10 days. He had history of trauma while playing. Physical examination reveals bony hard swelling over left forearm with tenderness and no associated scar, sinuses or dilated veins. Results of the lab analysis were Hb-11.5 gm/dl, with normal total and differential WBC counts, ESR 20 mm in the 1st hour, reticulocyte count 2%, and platelet count 2, 50, 000/cu mm which did not reveal any significant abnormality. Routine urinalysis was normal. Blood biochemistry including Blood sugar, urea and creatinine were within normal limit. X ray humerus AP and lateral revealed well circumscribed completely lucent lesions with ground-glass matrix without periosteal reaction. Fracture line was seen just above the area of lucency. MRI showed well defined intramedullary lesion measuring 3.2 × 2.7×9.6 cm with expansion and thinning of cortex. Lesion appeared heterogeneously hyperintense on T2 and iso to hypointense on T1 and enhancement on post contrast study. Findings were suggestive of benign neoplastic aetiology with pathological fracture. Patient underwent surgery for the same.

Humerus was exposed laterally followed by curettage and bone grafting. With 1 cm incision two intramedullary elastic nail were passed from medial and lateral supracondylar ridge respectively. Post operatively patient was immobilized with U slab for 6 weeks. Surgical biopsy on gross appearance showed multiple fragmented tissue, greyish white, soft to firm in consistency. Microscopically it revealed narrow, curved, irregular shaped trabeculae interposed in cellular fibroblastic stroma. No evidence of atypia/malignancy seen. Patient was followed up at six weeks, three months, five months after surgery (figure 7, 8). Patient improved symptomatically with significant reduction of pain. Serial x rays were done at 6 weeks and 3 months.

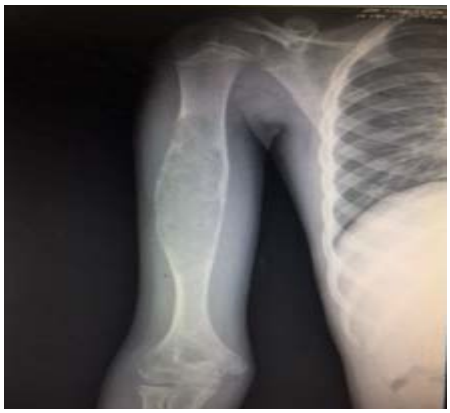

Fig 1: pre-operative AP view

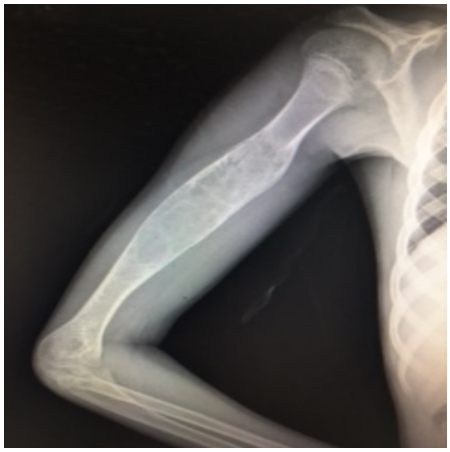

Fig 2: pre-operative lateral view

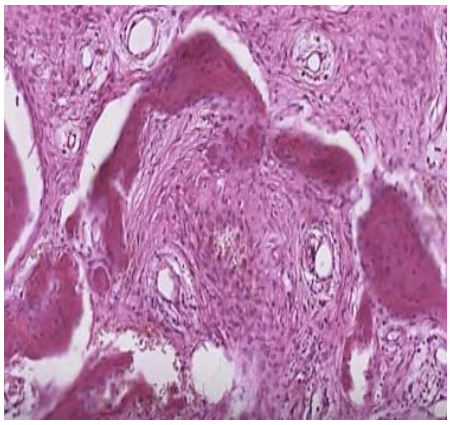

Fig 3: histopathology slide

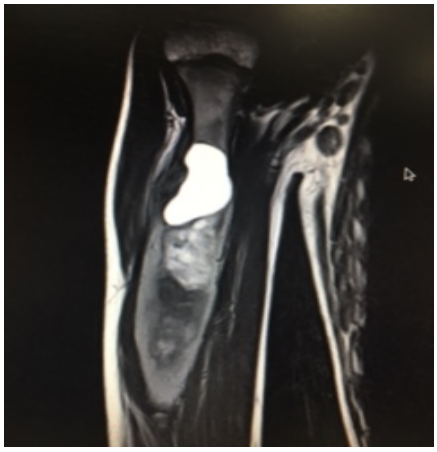

Fig 4: MRI coronal view

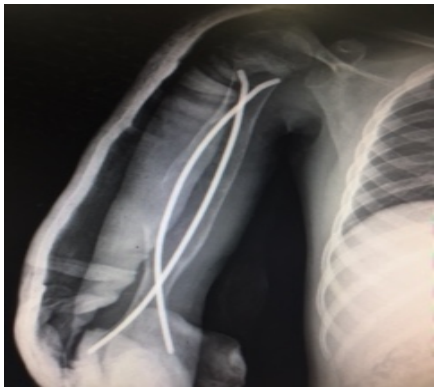

Fig 5: post-operative AP view

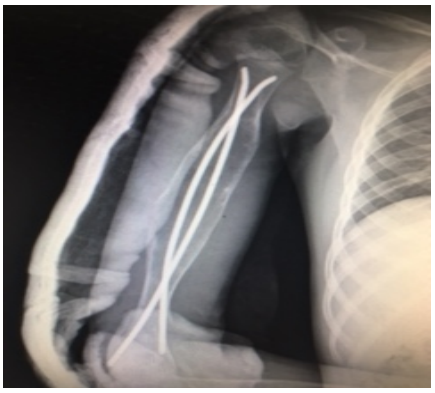

Fig 6: post-operative lateral view

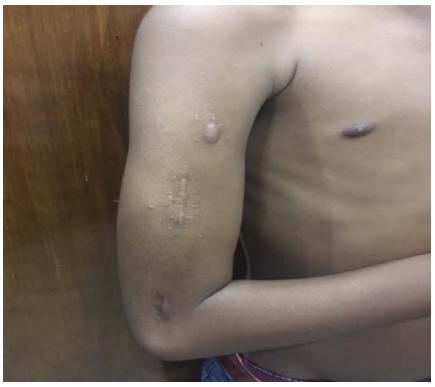

Fig 7: post-operative scar picture

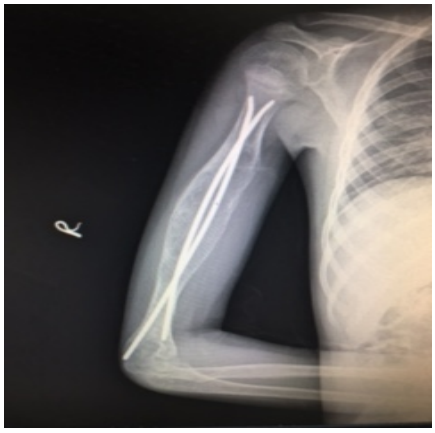

Fig 8: follow up x ray AP view

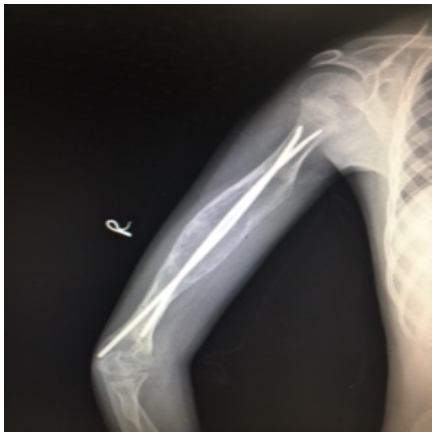

Fig 9: follow up x ray lateral view

3. Discussion

Osteofibrous dysplasia is a rare condition in which bone tissue is replaced by fibro-osseous lesions [3]. Most lesions of osteofibrous dysplasia affect the cortex of the bone, predominantly the middle third of the diaphysis [4]. The cortex often is expanded and thinned, with multiple radiolucencies mixed with intervening areas of sclerosis. The cause of fibrous dysplasia is unknown. Most cases of fibrous dysplasia display no particular pattern of inheritance [5]. Fibrous dysplasia can present as an autosomal dominant disorder affecting the mandible and maxilla bones in children in their teenage years. The tissue in the tumor is immature, woven bone that cannot differentiate in to mature, lamellar bone. This may be due to a mutation in a cell surface protein [6]. This is a somatic mutation, rather than in the germline. Fibrous dysplasia appears as irregular foci of woven bone arising from a cellular fibrous stroma. The stroma has a whorled appearance and is highly vascular. The short, irregular bone segments or trabeculae are not rimmed by osteoblasts. These irregular trabeculae have been described as "Chinese letters" or "alphabet soup" [7]. No lamellar bone is found within a fibrous dysplasia lesion. The abnormality is limited to the tissues within the lesions. The cells have an increased number of hormone receptors, which may explain why these lesions become more active during pregnancy [8]. Also, polyostotic fibrous dysplasia is known to have multiple associations with other disorders. The combination of polyostotic fibrous dysplasia, precocious puberty, and cafe au lait spots is called Albright's syndrome [9]. The association of fibrous dysplasia and soft tissue tumors has been given the name Mazabraud's syndrome [9]. Other endocrine abnormalities including hyperthyroidism, Cushing's disease, thyromegaly, hypophosphatemia, and hyperprolactinemia have been associated with fibrous dysplasia [9].

Radiographically, fibrous dysplasia appears as a well circumscribed lesion in a long bone with a ground glass or hazy appearance of the matrix. There is a narrow zone of transition and no periosteal reaction or soft tissue mass. The lesions are normally located in the metaphysis or diaphysis. There is sometimes focal thinning of the overlying cortex, called "scalloping from within" [10]. T-99 bone scan uptake may be normal or increased. Bone scans are not helpful in diagnosing these lesions but can be useful in identifying asymptomatic lesions. MRI scans or CT scans can be helpful in delineating the extent of the lesion and identifying possible pathological fractures. Sarcomatous change within the lesion can be identified by MRI or CT scans [10].

Diseases to be considered in the differential diagnosis of fibrous dysplasia include chondroma, simple bone cyst, non-ossifying fibromas, osteofibrous dysplasia, Paget's disease of bone, osteoblastoma, chondroblastoma, fibromyxoma of bone, adamantinoma and low-grade intramedullary osteosarcoma [11]. A biopsy may be needed to confirm the diagnosis, but surgery is necessary for a symptomatic lesion if there is a risk for pathological fracture.

Treatment for fibrous dysplasia generally consists of prophylactic surgery (curettage and grafting) and clinical observation. Recent studies have reported that bisphosphonate therapy may be effective in some patients with fibrous dysplasia [11]. Lesions whose behavior is latent and do not need any evaluation or treatment unless there is a risk of pathologic fracture. Painful long bone lesions can be managed with curettage and stabilized by cortical grafting or implant fixation [12]. Resection with curettage and bone grafting alone is best suited to lesions in non-weight bearing bones. These lesions should be evaluated carefully for risk of pathological fracture. Isolated Curettage and bone grafting of humerus lesions cannot be counted on to provide long-term healing, since the bone graft seems to be reabsorbed by the lesion in some cases. It is the advised for intramedullary fixation with the strongest possible device (a steel or titanium cephalomedullary nail) is the best method for treatment humerus lesions [13]. Persistent nonunion, malunion and refracture through lesions has been frequently observed [14]. It is advised to stabilize with strongest intramedullary fixation to prevent above complications.

4. Conclusion

This case report presents management of pathological fracture humerus secondary to Osteofibrous dysplasia with curettage and iliac crest bone graft along with intramedullary fixation. It also emphasizes on step wise management of Osteofibrous dysplasia including basic workup of patient along with rehabilitation. With proper counselling and compliance which can yield satisfactory functional outcome.

5. Abbreviations

OFD: Osteofibrous dysplasia, FD: fibrous dysplasia

6. Consent

For this case report to be published patient satisfactorily given informed consent for history, physical examination and publishing clinical photos and other relevant details.

7. Acknowledgements

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