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Case Report

MANAGEMENT OF OSTEOFIBROUS DYSPLASIA OF THE ULNAAFTER RESECTION WITH ELASTIC INTRAMEDULLARY NAIL AND NON VASCULAR FIBULAR GRAFT: A CASE REPORT

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ABSTRACT
Introduction: Osteofibrous dysplasia is a rare, nonneoplastic condition of unknown etiology that affects the long bones. It frequently is asymptomatic.Which usually affecting proximal femur, tibia, humerus, ribs, and craniofacial bones in decreasing order of incidence (McCaffrey *et al.,* 2003). Cases are diagnosed within the first three decades of life, though most commonly diagnosed in children and adolescents. Involvement at the ulna has also been reported in few cases. We are describing rare case of osteofibrous dysplasia of ulna managed after resection with fibular autograft and elastic intramedullary nail. *Case Presentation:* Present case is 27 year female housewife by occupation presented with swelling over left forearm since last six years (figure 1). Painless, insidious in onset and slowly progressive. Biopsy confirmed Osteofibrous dysplasia of ulna. We decided to manage with resection and non vascular fibular graft with elastic nail fixation. *Conclusion:* Osteofibrous dysplasia of ulna is very rare case which can be managed with resection and curettage with fibular bone autograft and which can be fixed with intramedullary nail.

Keywords: *Osteofibrous Dysplasia, Elastic Intramedullary Nail, Fibular Autograft*

INTRODUCTION
Osteofibrous dysplasia is a rare condition in which bone tissue is replaced by fibro-osseous lesions (Freeman *et al.,* 1987). These benign skeletal lesions may involve one bone (monostotic) or multiple bones (polyostotic). The lesions can occur throughout the skeleton but tend to occur in long bones, ribs and craniofacial bones.

They represent 5-7% of all benign bone tumours (DiCaprio and Enneking, 1987). 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, though most commonly diagnosed in children and adolescents.

Most lesions of osteofibrous dysplasia affect the cortex of the bone, predominantly the middle third of the diaphysis. The cortex often is expanded and thinned, with multiple radiolucency mixed with intervening areas of sclerosis (Campanacci and Laus, 1981). Polyostotic cases can affect multiple adjacent bones or multiple extremities.

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. Female patients may have increased symptoms during pregnancy. Unfortunately, this tumor can also present as a pathological fracture that is followed by a nonunion or malunion (Bullough, 1997).

The cause of fibrous dysplasia is unknown. Most cases of fibrous dysplasia display no particular pattern of inheritance. Fibrous dysplasia can present as an autosomal dominant disorder affecting the mandible and maxilla bones in children in their teenage years (Gray *et al.,* 2015).

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. This is a somatic mutation, rather than in the germline.

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. Also, polystotic 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. The association of fibrous dysplasia and soft tissue tumors has been given the name Mazabraud's syndrome. Other endocrine abnormalities including hyperthyroidism, Cushing's disease, thyromegaly, hypophosphatemia, and hyperprolactinemia have been associated with fibrous dysplasia.

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". The radiological appearance can also be cystic, pagetoid, or dense and sclerotic. Repeated fractures through lesions in the proximal femur can result in the formation of a so-called shepherd's crook deformity. 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 (Wootton-Gorges, 2009). Sarcomatous change within the lesion can be identified by MRI or CT scans.

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 (Lane *et al.,* 2001). Diseases to be considered in the differential diagnosis of fibrous dysplasia include chondroma, simple bone cyst**,** non-ossifying fibromas, osteofi- brous dysplasia, Paget's disease of bone, osteoblastoma, chondroblastoma, fibromyxoma of bone, adamantinoma and low-grade intramedullary osteosarcoma (Mathew and Joseph, 2007).

CASES

27 year female, Muslim by religion, housewife by occupation presented with swelling over left forearm since last six years. Painless, insidious in onset and slowly progressive. On physical examination reveals bony hard swelling over left forearm with no tenderness, scar and sinuses or dilated veins. Non adherent to skin without any discharging sinuses. X ray left forearm suggesting well circumscribed lesion in a ulna shaft with a ground glass appearance of the matrix (figure 2, 3). No periosteal reaction or soft tissue mass. Investigations revealed 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. Routine urinalysis was normal. Blood biochemistry including Blood sugar, urea and creatinine were normal. The serum calcium was 9.1 mg/dl and serum phosphate was 4.1 mg/dl. Liver function test revealed serum albumin 3.2 gm/dl, globulin 4.9 gm/dl, total serum bilirubin 0.5 mg/dl, SGOT 46 IU/ml, SGPT 44 IU/ml, and serum alkaline phosphatase 137 IU/l.

MRI was done suggesting of Osteofibrous dysplasia of left ulna. Which was later confirmed on Open excision biopsy. Histopathology shows on gross appearance, the tumor is a solid white or tan mass. The cut surface is gritty or sandy because of the fine bone spicules it contains. Microscopically, 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". No lamellar bone is found within a fibrous dysplasia lesion.

Patient underwent surgery for the same. Ulna was exposed and affected bone was resected and curettage was done. Created bone gap was filled up by non vascularized fibula graft (figure 6). And intramedullary elastic nail was passed from olecranon which was passed through fibula graft and distally engaged in distal ulna (figure 4, 5).

Patient was immobilized with above elbow slab for six weeks. Elbow range of motion was started after that. She was followed up at six weeks, three months, five months after surgery (figure 7, 8). At the end of two weeks she regained full range of movements of at elbow joint. Series of x rays were done at 6 weeks and three months.

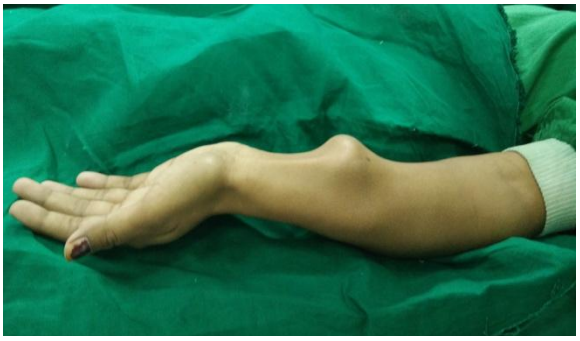

Figure 1: Clinical Picture

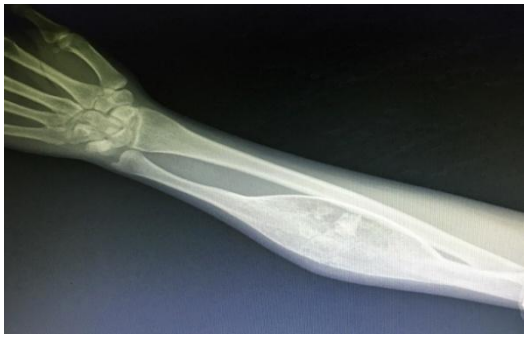

Figure 2: Pre-Operative AP View

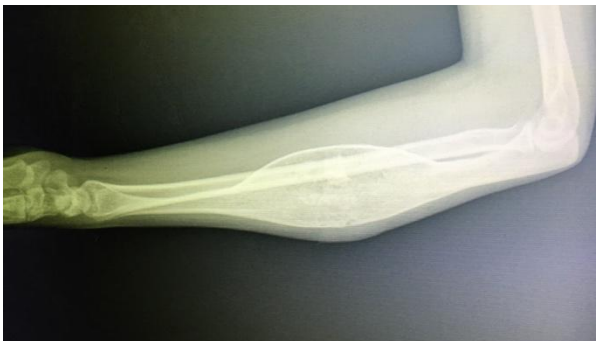

Figure 3: Pre-Operative Lateral View

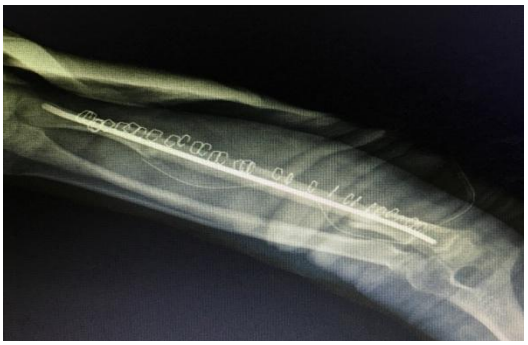

Figure 4: Post Operative AP View

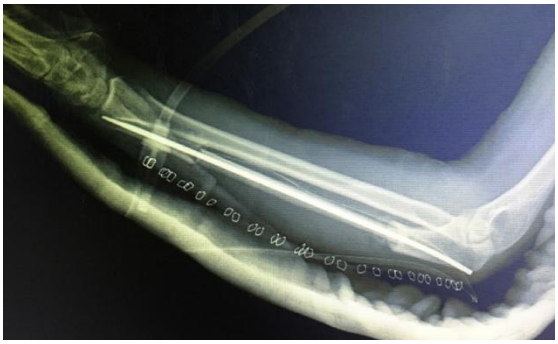

Figure 5: Post Operative Lateral View

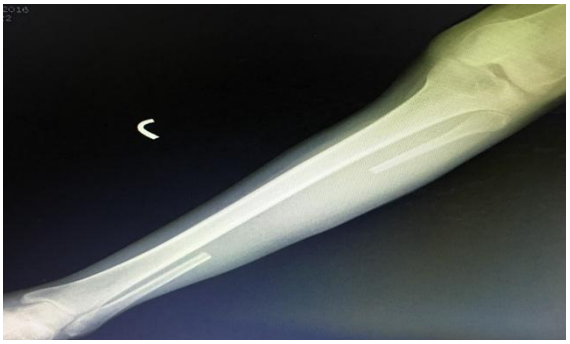

Figure 6: Post Operative Fibula Graft

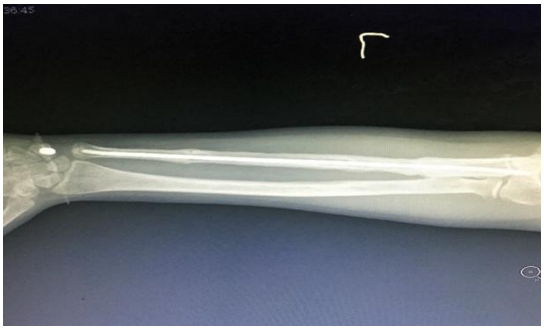

Figure 7: Post Op after 5 Month AP

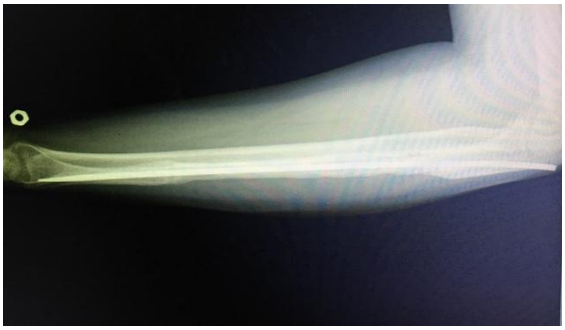

Figure 8: Post Op after 5 Month Lateral

DISCUSSION

Fibrous dysplasia is usually found in the proximal femur, tibia, humerus, ribs, and craniofacial bones. Its rare to find in ulna. 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. Lesions whose behavior is latent and do not need any evaluation or treatment unless there is a risk of pathologic fracture. Surgery with curettage of the lesion can be associated with high rates of local recurrence (Moretti *et al.,* 2012). Painful long bone lesions can be stabilized by cortical grafting or implant fixation. Cortical strut grafts are preferred to a morselized cortical cancellous grafts, which can become replaced with the same immature fibrous lamellar bone that comprised the lesion (Hahn *et al.,* 2007). Resection with curettage and bone grafting alone is best suited to lesions in non-weight bearing bones. Lesions within the ulna shaft are a particular challenge because they present in the young patients, and complications of treatment or from the tumor can lead to significant damage to the affected limb, joint and long-term disability. These lesions should be evaluated carefully for risk of pathological fracture. It is the advised for intramedullary fixation with the strongest possible device (a steel or titanium cephalomedullary nail) is the best method for treatment ulna lesions. Isolated Curettage and bone grafting of ulna lesions cannot be counted on to provide long-term healing, since the bone graft seems to be reabsorbed by the lesion in some cases (Huvos, 2015). Persistent nonunion, malunion and refracture through lesions has been frequently observed. It is advised to stabilize with strongest intramedullary fixation to prevent above complications. Our patient was managed with fibula graft and elastic nail for stabilization. Patient was kept on regular follow up. With proper rehabilitation.

Conclusion

This case report presents management of unusual osseofibrous dysplasia of ulna with fibula Cortical strut bone graft and intramedullary fixation. It also emphasizes on step wise management of osseofibrous dysplasia including basic workup of patient along with rehabilitation. With proper counselling and compliance which can yield satisfactory functional outcome.

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