Case Report

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Monostotic fibrous dysplasia of the proximal phalanx of left hand: a case report

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ABSTRACT

Fibrous dysplasia is a benign skeletal developmental anomaly where normal bone is replaced by fibrous tissue. There is a defect in osteoblastic differentiation and maturation of normal bone, which results in formation of structurally weak bone. Disease can affect one bone (monostotic) or multiple bones (polyostotic). Fibrous dysplasia usually involves femur, tibia, pelvis, ribs and skull. It rarely occurs in bones of hands and feet. We report a case of fibrous dysplasia of proximal phalanx of left middle finger in a 10 year old female.

KEY WORDS: Fibrous Dysplasia; Monostotic; Polyostotic; Phalanx; Enchondroma

INTRODUCTION

Fibrous dysplasia (FD) rarely occurs in hand. It is a disorder where normal bone is replaced with fibrous tissue. Majority of clinically significant lesions are detected in the first decade. The hallmark of the disease is an inability to produce mature lamellar bone and an arrest at the level of woven bone [1]. Monostotic fibrous dysplasia (MFD) is rare in upper extremity. Only a few cases have been reported in phalanges [2,3]. We report a case of FD involving proximal phalanx of left middle finger.

CASE PRESENTATION

A 10 year old female child presented to our department with pain and swelling of middle finger of left hand since 6 months. Initially, the patient noticed swelling in the middle finger which increased gradually. Subsequently she developed pain in the swelling since last one month. There was no history of trauma or fever prior to the onset of swelling. There were no other swelling elsewhere in the body. Clinically, the proximal phalanx of the left middle finger was swollen and tender. The movements of proximal interphalangeal joint and metacarpophalangeal were restricted and painful.

Radiographs (Fig 1) of the affected area showed a lytic, expansile lesion in the proximal phalanx with cortical thinning and ground glass appearance. The list of differential diagnosis included Aneurysmal bone cyst (ABC), Simple bone cyst (SBC), Non ossifying fibroma (NOF), Enchondroma, Spina ventosa and FD. Patient was under taken for surgery. Curettage of the lesion (Fig 2) along with autologous bone grafting was done. Appropriate splintage was given till suture removal at 2 weeks, following which hand mobilization was allowed. Radiographs showing bone graft incorporation at the lesion were taken at 6 weeks (Fig 3) and at the final follow up of 1 year (Fig 4).

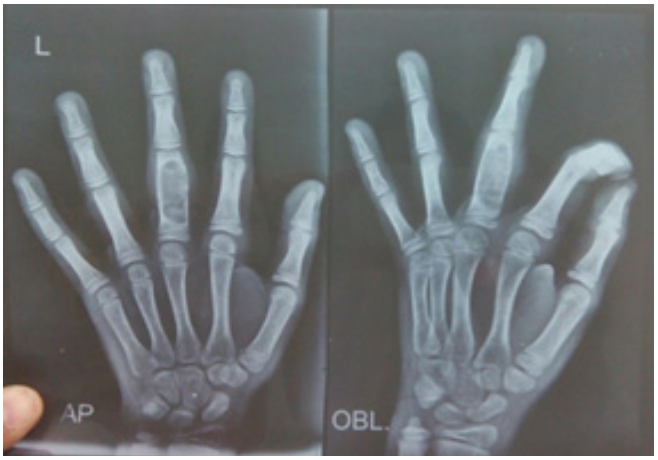

Fig 1. AP and Oblique view of left hand showing lesion in the proximal phalanx of middle finger

Fig 2. Window made in the cavity of proximal phalanx for curettage and bone grafting

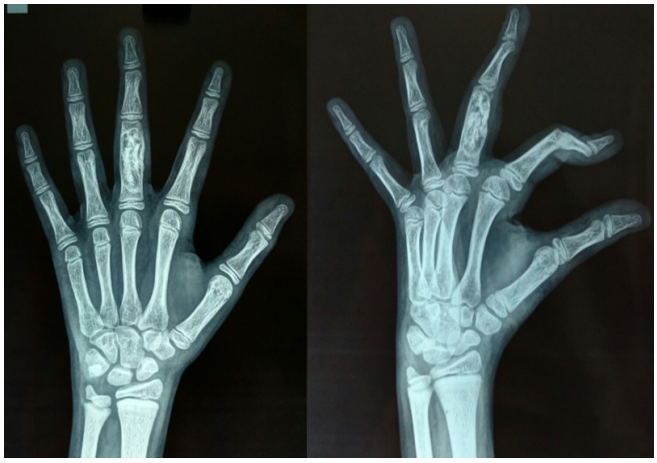

Fig 3. X rays at 6 weeks follow up

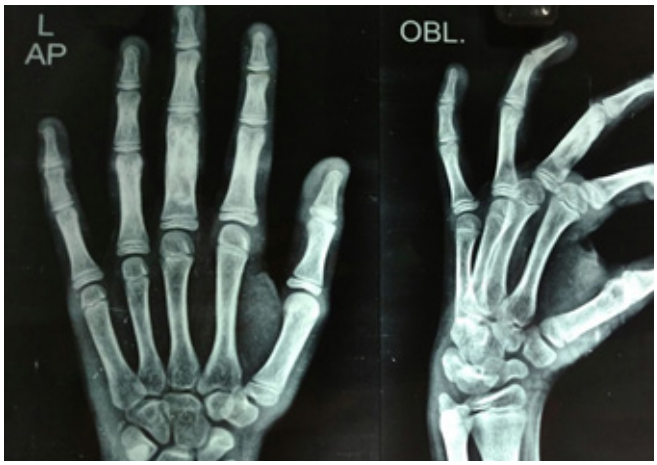

Fig 4. X rays at the last follow up of 1 year showing graft incorporation

DISCUSSION

FD can be considered as a dysplastic anomaly of bone forming mesenchymal tissue with an inability of bone forming tissue to produce mature lamellar bone [1]. FD can involve a single bone (monostotic) or multiple bones (polyostotic). FD accounts for 5 % of benign bone lesions [4]. MFD is present in as many as 75-80 % of the total cases. It is usually detected in the first decade when it becomes clinically significant. The lesion can cause pain, deformity and occasionally pathological fracture.

Most commonly, MFD involves proximal femur, tibia, skull and ribs [5]. Involvement of upper extremity especially the phalanx is rare with only few cases having been reported in the literature. FD has been known to occur with few syndromes. McCune –Albright syndrome is a rare multisystem disorder characterized by triad of polyostotic fibrous dysplasia, precocious puberty and café au lait spots [6]. Mazabraud syndrome is a rare syndrome comprising of polyostotic fibrous dysplasia and multiple soft tissue intramuscular myxomas [7]. On x-ray, phalangeal FD typically appear as lytic , expansile lesion with ground glass appearance with intact cortex as was in our case [8,9]. This x ray appearance can be seen in ABC, SBC, NOF, encondroma and spina ventosa [10]. MRI of the lesion show an isointense signal area and a high signal area on T2 weighted sequence, a homogenous low signal on T1 weighted images and a homogenous isointense signal on STIR images [9]. Thus MRI is very helpful in diagnosing the condition. However, we were not able to get MRI because of financial constraints.

Curettage of the lesion should be done and tissue should be send for histopathological examination to confirm the diagnosis. After the curettage, the cavity should be filled with autologous bone graft. Malignant transformation is present in only 0.4- 4% of the cases. Most common tumors are osteosarcoma, fibrosarcoma and chondrosarcoma [11]. The rate of malignant transformation is higher for polyostotic lesions than for monostotic lesions.

CONCLUSION

MFD of the hand is rare and the involvement of the phalanx is even rarer. Thorough clinico-radiological examination should be done to diagnose this condition and to look for involvement of other sites. It should always be included in the list of differential diagnosis of lytic lesion of phalanges.

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