Case Report

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Rare case of symptomatic (painful) fibrous dysplasia of 12th Rib

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ABSTRACT

Fibrous dysplasia (FD) is a common benign intra-medullary lesion of the skeleton characterized by proliferation of fibro-osseous tissue. It is viewed as a developmental abnormality that is typically seen in adolescents and young adults. It can present as monostotic (70% to 80%), polyostotic fibrous dysplasia (20% to 30%), McCune-Albright syndrome (2% to 3%), or Mazabraud's syndrome. Monostotic fibrous dysplasia accounts up to 30% in the ribs. It is often asymptomatic and incidentally detected on radiographs. Author report a case of a 35-year-old lady who presented with swelling on the right chest wall of 5-year duration which was increasing in size and painful for 8 months. Her chest radiograph showed a cystic mass of the right 12th rib. On computed tomography scan, mass turned out to be focal lytic lesion of the 12th rib. Surgical resection of 12th rib was done. Histopathology of the lesion confirmed the diagnosis of FD. Although it is fairly common in ribs, no previous literatures on FD of 12th rib was found.

Keywords: 12th Rib, Fibrous dysplasia (FD), Painful, Rare

INTRODUCTION

FD is a condition leading to abnormal differentiation of osteoblasts which leads to replacement of normal bone with fibrous stroma.1 The term FD was introduced by Lichtenstein in 1938 to describe the anomalous replacement of medullary bone by fibrous tissue. It is a benign lesion that may involve one (monostotic) or more bones (polyostotic) or be accompanied by other systemic alterations and endocrine disorders, such as in the McCune-Albright syndrome.² Polyostotic forms are observed in head, face, humerus, leg, upper limbs, iliac bone, ribs and vertebrae. On the other hand, monostotic forms are mainly observed in ribs.³ FD of ribs accounts for up to 30% of all benign chest wall tumours, and monostotic forms are about four times more common than polyostotic forms. It is typically present in the third or fourth decade of life as an asymptomatic mass.⁴ Author report a case of a 35-year-old lady who presented

with swelling on the right chest wall of 5-year duration and gradual increase in size and pain for 8 months. Radiologically, X-ray and CT scan showed a focal lytic expansion (2.5*1.9cm) with ground glass matrix of the 12th rib. The resected lesion was 12th rib with solid cystic lesion having focus of blackish discoloration. Histopathologically, benign fibrous spindle areas with disorganized irregular bony trabeculae were seen. The diagnosis was monostotic FD of the 12th rib. Although FD is seen in ribs but 12th rib involvement is rare and no previous reports about symptomatic FD of 12th rib was found.

CASE REPORT

A 35-year-old lady came to the orthopaedic clinic with gradual increase in size of the swelling of 5-year duration on the right side of chest for 8 months. Radiologically, X-

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ray and computed tomography (CT) showed a focal lytic expansion (2.5x1.9cm) of the 12th rib (Figures 1, 2 and 3).

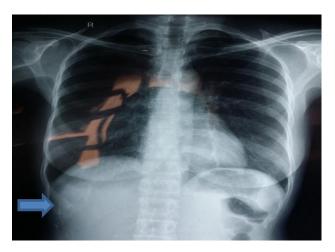


Figure 1: X-ray chest showing expansile lytic lesion of right 12th Rib.

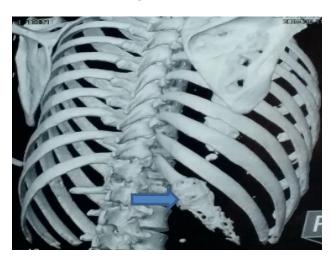


Figure 2: CT scan-with 3D construction showing focal lytic expansion (2.5*1.9cm) of the right 12th rib.



Figure 3: CT scan- on axial cut section showing focal lytic expansion of the right 12th rib.

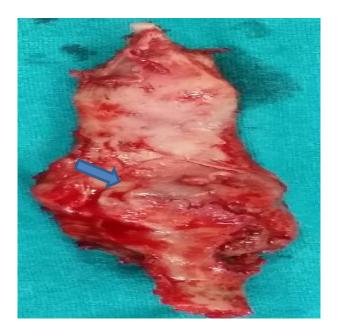


Figure 4: The gross appearance of bony hard tissue with soft tissue measuring 11x2.3cm. At a focus, there was presence of a solid cystic lesion measuring 2.3x1cm with focal blackish discoloration in the solid area (arrow).

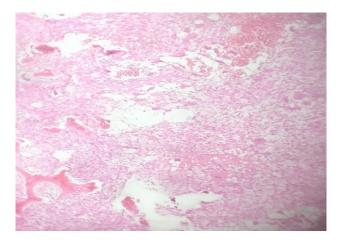


Figure 5: Microscopy showing Chinese alphabet spicule of woven bone with fibrous stroma (H and E).

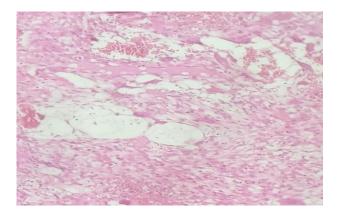


Figure 6: Microscopy showing no atypia of stromal cell (H and E).

The lesion with partial resection of the 12th rib was performed. The gross appearance was a bony hard tissue with soft tissue measuring 11x2.3 cm. At a focus, there was presence of a solid cystic lesion measuring 2.3x1cm with focal blackish discoloration in the solid area (Figure 4). Histologically, the lesion consists of osseous and fibrous components. The osseous component consists of disorganized irregular "Chinese alphabet" spicules of woven bone separated by abundant fibrous stroma (Figure 5). The fibrous component was composed of cytologically bland spindle cells with no atypia of stromal cells (Figure 6). The final diagnosis was monostotic fibrous dysplasia of 12th rib.

DISCUSSION

FD of a skeleton is an intraosseous neoformation of the fibrous tissue probably caused by an anomaly in the development of the bone.⁵ Fibrous dysplasia is usually diagnosed before the age of 30 years and, if considered with bone tumors, comprises nearly 1% of primary bone tumors.⁶ It is monostotic in 70-80% of cases and polyostotic in 20-30% of cases. The age of presentation ranges from 10-70 years with the majority of cases (75%) presenting before the age of 30 years.¹ It presents later in the ribs, probably because it is often asymptomatic in this site. The disease is equally distributed in both sexes. Reactivation may occasionally occur in later life and in pregnancy.⁷

The etiology has been linked with a mutation in the Gs α gene that occurs after fertilization in somatic cells and is located at chromosome 20q13.2-13.3. All cells that derive from the mutated cells manifest the dysplastic features. The clinical presentation varies depending on where in the cell mass the mutation is located and the size of the cell mass during embryogenesis when the mutation occurs. Severe disease may be associated with an earlier mutational event that leads to a larger number or a more widespread distribution of mutant cells. The sporadic occurrence of these diseases and the characteristic lateralized pattern of skin and bone involvement in the polyostotic forms of fibrous dysplasia suggest this mosaic distribution of abnormal cells. The Gs α mutation was first identified in patients with McCune-Albright syndrome, a rare disorder that combines polyostotic fibrous dysplasia, skin pigmentation, and one of several endocrinopathies.8

The three most common radiological features of FD are pagetoid, sclerotic, and cystic patterns. The tumors are homogeneous soft-tissue masses, showing low attenuation on CT. On MRI, they are hypointense to slightly hyperintense on T1-weighted images and hyperintense on T2-weighted images and show a complex pattern of enhancement after administration of gadolinium. Lesions of fibrous dysplasia are composed of fibrous tissue containing bone trabeculae. The fibrous stroma is a myxofibrous tissue of low vascularity, while the bone trabeculae are composed of woven bone. The

architecture of bone actually varies greatly in the degree of layering. Woven refers to bone with little layering, while lamellar means bone with well-formed layers. All gradations in between can occur, depending on the speed with which the osteoid is laid down. Fibrous dysplastic bone can have noticeable layering, but its architecture never approaches the lamellar structure of normal cortical bone. The outline of the bone trabeculae in fibrous dysplasia varies from solid, round islands to a wide variety of curved, serpiginous, or curlicue shapes that have been likened to Chinese characters or alphabet soup. The amount of bone is quite variable, but it is usually easily found. In some cases, many sections must be examined.¹¹

Malignant transformation with rapid expansion of the bone has been reported in about 0.5% of patients with monostotic FD but in nearly 4% of those with McCune-Albright syndrome. It may develop after irradiation of the involved bones. Malignant transformation is most common to osteosarcoma, although fibrosarcoma, chondrosarcoma, or malignant fibrous histiocytoma noted.¹² Fibrous dysplasia usually can be managed by observation. Large symptomatic lesions at risk of fracture may require a surgical procedure, such as curettage and bone grafting, with or without implants. Bisphosphonates are used for nonsurgical management when the disease cannot be managed surgically. The prognosis for fibrous dysplasia is generally good, although outcomes are poorer in young patients and those with the polyostotic forms.¹³ Surgery is indicated for chronic weight-bearing bone pain not relieved by medical management, for the correction of deformity that is interfering with useful ambulation, for the management of recurrent fractures, nonunion of fractures, and transformation of the lesions into either benign (i.e., aneurysmal bone cyst) or malignant tumors.¹⁴ Some surgeons prefer a surgical management than simple surveillance, especially in ribs location of FD, because it can raise the difficult problem of differential diagnosis with malignant tumours. Author believe that in ribs location of symptomatic monostotic FD, excision of involved bone must be indicated for both curative and diagnosis intention, to rule out malignancy and provide relief from symptoms. 15

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