

Polyostotic fibrous dysplasia of the ribs: An unusual cause of chest pain and dyspnea

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Abstract: Fibrous dysplasia is a benign bony abnormality that may be monostotic or polyostotic and is not included in the differential diagnosis of chest pain and dyspnea, since it is typically asymptomatic. A 36-year-old man presented with left-sided chest pain and dyspnea for three months. Chest X-ray and chest computed tomography scan revealed a large solid mass arising from the anterior parts of the left fourth, fifth, and sixth ribs and compressing the adjacent lung parenchyma. The tumor was completely removed surgically and histopathologic examination was consistent with fibrous dysplasia without malignant transformation. This report demonstrates that polyostotic dysplasia of the ribs may cause chest pain and dyspnea and present radiographically as a large mass.

Keywords: chest wall, imaging, computed tomography, thoracotomy

Introduction

Polyostotic fibrous dysplasia is a rare developmental disorder with focal areas of abnormal bony architecture. We present an exceptional case of a polyostotic fibrous dysplasia of the ribs.

Case report

A 36-year-old man was admitted to our hospital with complaints of left-sided chest pain and shortness of breath for the previous two weeks. Past history and family history were not significant. Physical examination was within normal limits except tenderness in the left side of the chest at the level of the fourth to sixth ribs. A chest X-ray (Figure 1) and computed tomography (CT) scan of the chest (Figure 2) showed a large thoracic mass arising from the lateral parts of the left fourth to sixth ribs.

Thoracotomy was performed. A frozen section biopsy at the time of surgery was consistent with fibrous dysplasia and thus, the tumor was completely removed with en-block resection with 2 cm margins. Also, three ribs were sectioned during invasion. The chest wall defect did not require reconstruction with prosthesis. The postoperative course was uneventful with resolution of the symptoms.

The surgical specimen was a hard, lobulated and partially calcified tumor arising from the lateral part of the left fourth to sixth ribs which measured 16 × 11 × 6 cm (Figure 3). The tumor had two components: 1) multiple cysts containing yellowish-serous fluid and 2) creamy solid areas. Cysts were multiple and the largest had a diameter of 9 cm. Microscopically, the tumor had a typical histologic appearance of fibrous dysplasia. Neither cellular atypia, mitotic figures, nor necrosis were observed (Figure 4).

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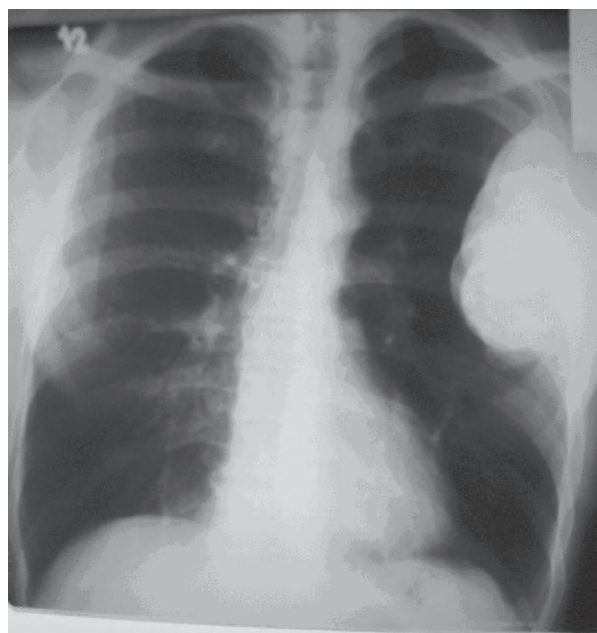


Figure 1 A chest X-ray demonstrates a large peripheral mass arising from the lateral parts of the left fourth to sixth ribs.

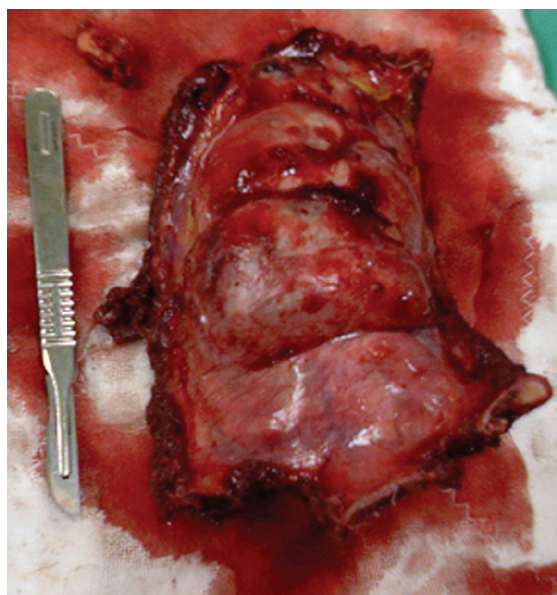


Figure 3 The surgical specimen was a hard, lobulated and partially calcified tumour arising from lateral part of the left fourth to sixth ribs which measured 16 × 11 × 6 cm. The tumor had two components: multiple cysts containing yellowish-serous fluid and solid areas.

Discussion

Fibrous dysplasia is a sporadic developmental condition affecting bones, which show poor mechanical strength leading to pathologic fractures and progressively enlarging deformities that impinge on adjacent structures. Monostotic involvement occurs more commonly than the polyostotic form. Monostotic disease does not usually progress to polyostotic disease, and the size and number of lesions generally remain the same over time as they were at initial radiologic evaluation.^{1,2} Bony involvement in polyostotic fibrous

dysplasia typically includes facial bones, skull base, long bones, and occasionally ribs.

Chest pain, an unusual symptom resulting from polyostotic fibrous dysplasia of the ribs, has been reported previously in only two papers.^{3,4} Rarely, multiple lesions are sufficient to result in progressive restrictive lung disease, pulmonary hypertension, and cor pulmonale.^{2,5,6}

Radiographs characteristically show unilateral fusiform enlargement and deformity with cortical thickening and increased trabeculation of one or more ribs. Amorphous or

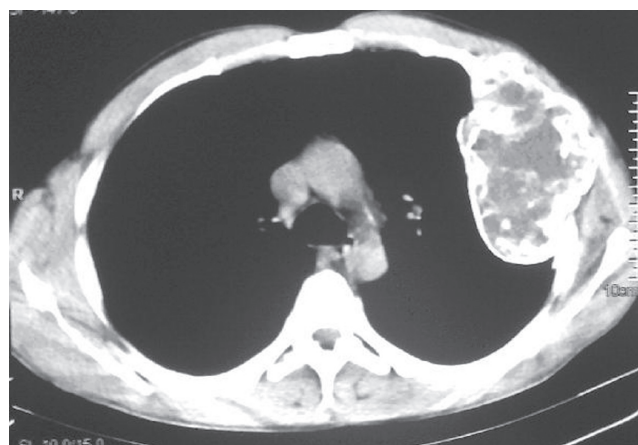


Figure 2 Axial computed tomography (CT) scan at the level of the aortic arch shows an expansile mass with areas of ground-glass attenuation and irregular calcification in left fourth to sixth ribs.

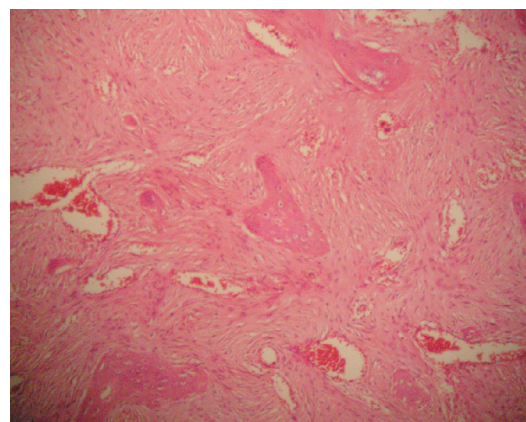


Figure 4 Histological appearance diagnostic of fibrous dysplasia. The mesenchymal stroma surrounding the dysplastic trabeculae is relatively hypocellular. There is a lack of osteoblastic rimming surrounding the dysplastic trabeculae (hematoxylin and eosin stain, × 200).

irregular calcification is often seen on CT sections of the lesion^{1,2} as in our case.

Costal lesions of fibrous dysplasia are also known to develop after the end of growth because of cystic degeneration, with occasional formation of an aneurysmal cyst.⁷ Development of a huge benign costal mass compressing the mediastinum is exceptional. Since this complication is rare, a prophylactic resection for small fibrous dysplasia is probably not justified. Even when the tumor is massive, as in our case, surgical resection is still possible since fibrous dysplasia of bone is a benign and noninfiltrative tumor. It must be kept in mind that malignancy occurs in less than 1% of cases of fibrous dysplasia.⁸ Thus, surgical approach is usually only performed for exact diagnosis.⁹

In conclusion, polyostotic fibrous dysplasia of the ribs, although it is rare, may be included in differential diagnosis of chest pain.

Disclosures

The authors report no conflicts of interest in this work.

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