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CASE REPORT

Sternal Polyostotic Fibrous Dysplasia: A Rare Tumor in an Unusual Location

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Abstract. Sternal tumors and masses are not uncommon and include a wide spectrum of diseases. The rarest and the strangest pathology is a patient who presented with a painless sternal mass. Histopathology showed proliferating fibroblastic tissues and irregular trabeculae of woven bone. This rarest sternal tumor is known pathologically as polyostotic.

Keywords: Sternum, Tumor, Chest wall, Polyostotic.

Introduction

Fibrous dysplasia is a benign lesion in which portions of the bone are replaced by fibrous connective tissue and poorly formed trabecular bone[1]. It may involve a single bone [monostotic fibrous dysplasia] or multiple bones [polyostotic fibrous dysplasia][2]. Polyostotic form is associated with endocrine anomalies in McCune-Albright syndrome (MAS)[3]. Polyostotic fibrous dysplasia involving the sternum or the ribs is rare with only a few reports of cases in association with thoracic spine involvement[4].

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Case report

A 36-years-old female was presented with a painless sternal mass that had a history of chest trauma during childhood. The mass was 3.2 x 4.5 cm nontender, hard, immobile arising from the body of the sternum. She also complained of tenderness along the left fourth ribs.

Method

Computerized tomography (CT) scans and magnetic resonance imaging (MRI) of the chest (Fig. 1) showed expansile, osteolytic and hyper translucent multilocular septated lesions of the sternum, the fourth rib and the fourth thoracic vertebral body. Bone scan revealed intense heterogeneous isotope uptake in the same sites. Hormonal assay to rule out the McCune-Albright syndrome were within normal limits. Open wedge resection of the tumor in the sternal body

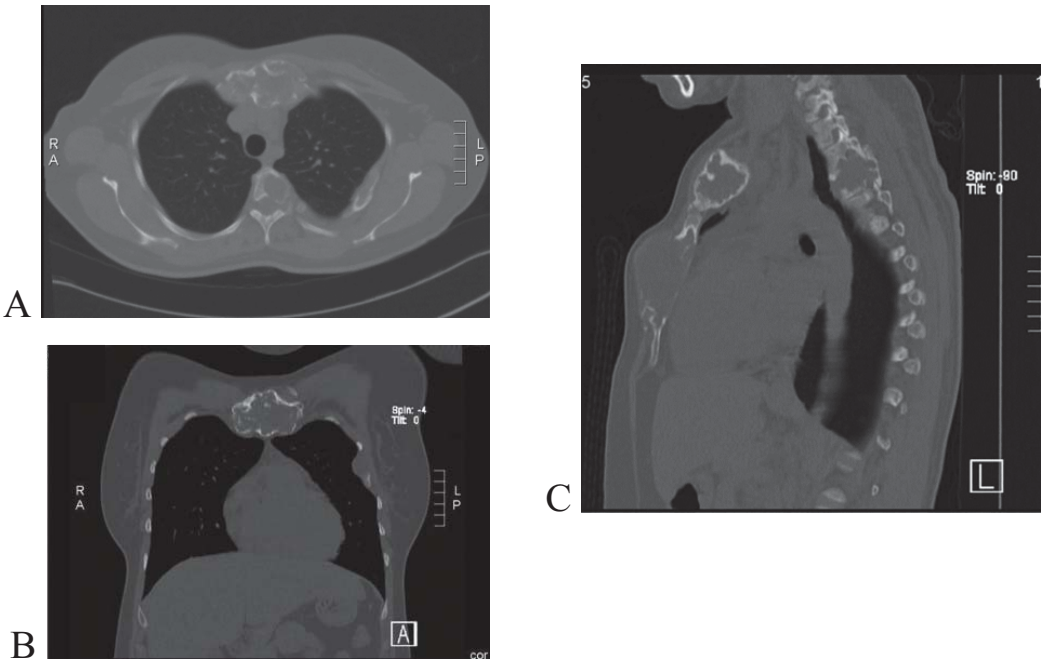

Fig. 1. **A) CT scan of the chest cross section showing involvement of the sternum and 4th vertebral body; B) MRI coronal view showing the multi septated hyper translucent sternum; C) MRI sagittal view showing the sternal and 4th vertebral body destruction.**

revealed fibrous tissue proliferation arranged in storiform pattern. Trabeculae of woven bone lacking the osteoblastic rimming are arranged like Chinese letters or alphabet soup, typical of fibrous dysplasia (Fig. 2). The sternal involvement was treated conservatively, and the patient was referred to neurosurgery for vertebral stabilization to avoid collapse fracture and spinal complications. Bone scan follow-up of such patients is recommended to detect new sites.

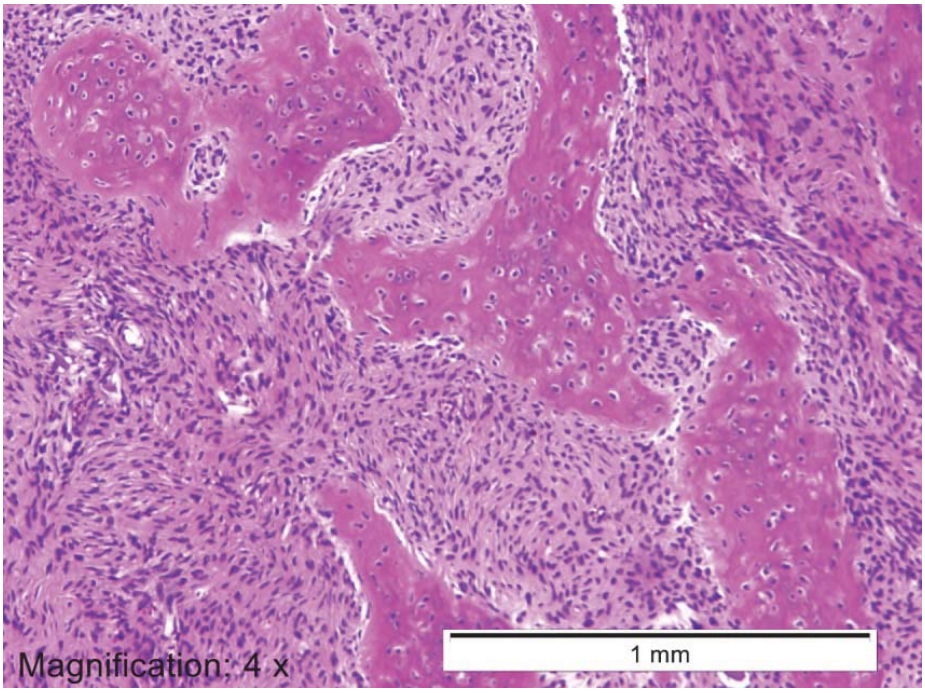

Fig. 2. **Photomicrograph using H&E staining and 4x-magnification showing typical features of fibrous dysplasia, with trabecular bone lacking an osteoblastic rim interspersed in proliferating fibroblastic tissue.**

Discussion

Fibrous dysplasia is a congenital, non-inherited connective tissue disorder in which medullary bone is replaced with fibro-osseous connective tissue[5]. The disorder can present in either a single skeletal site or multiple sites, respectively termed monostotic or polyostotic. The McCune-Albright syndrome is a severe form of fibrous dysplasia, which is associated with café-au-lait spots and

endocrine dysfunction. The primary etiology has been linked to a mutation in the alpha subunit of the guanine-nucleotide binding protein (Gs-α) leading to continuous activation of adenylate cyclase and increased cyclic adenosine monophosphate (cyclic AMP) formation, leading to abnormal bone development[6]. Long bones, lumbar spine, and craniofacial bones are the most common sites for fibrous dysplasia. The sternum is the rarest bone to be affected[7]. Extensive chest wall involvement can lead to restrictive lung disease and cor pulmonale requiring release by chest wall reconstruction[8,9]. Early cases like this require following up with periodical CT, MRI or bone scan to monitor progression in size or eruption of new sites. Surgery is indicated in symptomatic patients, compression of vital organs or fear of fracture.

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عجز النمو الليفى عديد الحويصلات لعظمة القص: ورم نادر فى مكان غير معتاد

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المستخلص. الأورام والكتل التى تصيب عظمة القص هى من الأمراض غير الشائعة ويعتبر الورم الذى أصاب مريضتنا من أندرها وأغربها على الإطلاق فى تصنيف علم الأمراض. الورم لم يصاحبه أى الم وتبين من الفحص المجهري وجود ولى متشعب يحوي حواجز غير منتظمة من النسيج العظمي. ويعرف هذا الورم النادر باسم بوليستوزس.