Giant Cell Tumour of the Sternum-Two Cases

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Abstract

Giant cell tumor (GCT) is a common, benign, locally aggressive bone tumour affecting long bones patients between the ages of 20 and 45 years. The incidence of this tumour is 5%. Multinucleate giant cells identify it, which consists of more than 100 nuclei and fibrous stroma. Amongst long bones the tibia and femur are affected. Other affected sites are radius, small bones of hand, sacrum, pelvis, vertebral bodies and ribs. The sternum is a flat bone. The commonest tumour affecting sternum is metastasis. Most common tumours affecting sternum are chondrosarcoma, osteosarcoma, myeloma, and lymphoma. Benign primary tumors of the sternum are even rarer than malignant primary tumors of the sternum. Giant Cell Tumour of the sternum is a rare condition and to the best of our knowledge only seven cases are documented in the literature.

We are presenting case reports of two patients who presented with sternal mass pathologically proven as giant cell tumour. One patient is a 38-year-old woman and another 28-yearold aged man. These patients presented with palpable chest wall masses. Plain radiographs showed an expansile osteolytic lesion of the sternum.

Computed tomography (CT) demonstrated focal cortical bone lytic lesion with destruction, soft tissue extension. Bone scan showed increase uptake. Fine needle aspiration cytology examination performed revealed a benign tumor composed of multi-nucleated giant cells compatible with a Giant Cell tumour. Histopathological analysis showed presence of multinucleated giant cells, and spindle cells with elongated nuclei without atypias although rare giant cell tumour of sternum must be considered as a differential in patients with lytic expansile lesion of sternum.

Keywords : Giant cell tumour of sternum, Osteoclastoma, Sternal tumours, CECT, Sternum, Osteoclasts

I. INTRODUCTION

Giant cell tumor (GCT), or Osteoclastoma, is a tumor of mesenchymal origin characterized by proliferation of multinucleated giant cells. Sir Astley Cooper first described this type of tumor in 1818.^{1,2} Giant cell tumour of the sternum is a rare tumour.¹ To date only seven cases have been described in literature to the best of our knowledge^{1,2,3,4,5,6,7}. We present case series two cases of giant cell tumours of sternum, which presented to our hospital.

The sternum is a flat bone, slightly convex anteriorly

and concave posteriorly. It consists of three parts: the manubrium, body, and xiphoid process. It tends to narrow at the junction between the body and manubrium and the junction between the body and the xiphoid process. Computed tomography (CT) is the modality of choice to evaluate anatomic detail as well as pathologic conditions of the sternum, sternoclavicular joints, and adjacent soft tissues. Most neoplasms of the sternum are metastases.^{1,2} Primary tumors are relatively uncommon in this site; however, primary tumors of the sternum are much more frequently malignant than benign. Benign primary tumors of the sternum are even rarer than malignant primary tumors of the sternum.^{7,4,5,6}

Case 1

CASE HISTORY:

38-year-old female patient was admitted to our hospital because of swelling and pain in the anterior chest region. The patient presented a one and half year history of mid sternal chest pain after heavy housework; the pain was constant localized and partially controlled with analgesics, and the pain and swelling developed gradually and breathlessness from 5 days. No history of recent respiratory infection or weight loss or fever. She was in good general health without any changes in the other systems. Physical examination revealed a 3.8×4 cm hard and fixed mass, which could be palpated in the mid-sternum. The surface was felt to be smooth, but the border was unclear. There were no other visible bony lesions.

She was referred for CECT for further characterization. CECT was done using six sliced Siemens Emotion, Pre and post contrast CT of Thorax was done. No solid component was identified. . No extension to steno costal joint. Chest CT revealed an expansile lytic lesion $(5.82 \times 6.2 \times 9.8 \text{ cm})$ affecting the cortical layer of the sternum, it showed contrast enhancement. Few areas of cortical destruction seen. On plain CT this lesion showed an attenuation of 30-40 HU; on post contrast studies it showed of about 70 to 80 HU. This lesion was epicentred in manubrium sternum with cortical erosion involving anterior and posterior aspect and no other chest wall bone abnormalities were seen .It abutted pericardium.

No invasion of surrounding structures or breast or cardia. On Bone scan uptake is shown in sternum. Figures 1,2 and 3).

Differentials considered were Plasmacytoma Giant cell tumour and Aneurysmal bone cyst. She was subjected to Pathological examination, which revealed showed numerous large multi-nucleated osteoclasttype giant cells with no atypical cells.

Case 2

28-year-old male patient was admitted to our hospital because of swelling and pain in the anterior chest region. The patient presented a history of midsternal swelling and pain from three years, No recent respiratory infection or weight loss or fever. He was in good general health without any changes in the other systems. Physical examination revealed a 10×8 cm hard and fixed mass, which could be palpated in the mid-sternum. The surface was felt to be smooth, but the border was unclear. There were no other visible bony lesions. Overlying skin was shiny. He was referred for CECT for further characterization.

CECT was done using six slice Siemens emotion .Pre and post contrast CT of Thorax was done. . No solid component was identified .No soft tissue component .No extension to sterno costal joint. Chest CT revealed an expansile lytic lesion (14.82 \times 6.2×9.8 cm) affecting the cortical layer of the sternum, it showed contrast enhancement. Few areas of cortical destruction seen. On plain CT this lesion showed an attenuation of 40 HU; on post contrast studies it showed of about 70 to 80 HU. On Bone scan uptake is shown in sternum. It extended anteriorly, it involved entire sternum. (Figure 4 and 5). Differentials considered were Plasmacytoma, Giant cell tumour and Aneurysmal bone cyst. He was subjected to Pathological examination, which revealed showed numerous large multi-nucleated osteoclasttype giant cells with no atypical cells.

Most neoplasms of the sternum are metastases. Primary tumors are relatively uncommon in this site; however, primary tumors of the sternum are much more frequently malignant than benign. Benign primary tumors of the sternum are even rarer than malignant primary tumors of the sternum According to the series of the Mayo Clinic; benign sternal tumors were found in only 3 out of 2,334 cases of all benign bone tumors. (0.1 %) Two of these three benign cases were sternal GCTs. 1,2,3,4,5,6,7 The spectrum of lesions includes enchondroma, osteochondroma, hemangioma, hemangiopericytoma, enostosis, osteoid osteoma, fibrous dysplasia, Paget disease, Langerhans cell histiocytosis, aneurysmal bone cyst, eosinophilic granuloma, giant cell tumor, brown tumor, nonossifying fibroma, and chondromyxoid fibroma. In what may be one of the largest studies of benign sternal neoplasms so far, Hoeffel et al reviewed 19 cases of benign tumors of the sternum and found 14 cases of cartilaginous tumor (16.66%), two cases of eosnophilic granuloma (2.38%), one case of fibrous dysplasia (1.19%), one case of giant cell tumor (1.19%), and one "miscellaneous" case.7

Conventional GCT of bone is a relatively common, locally aggressive neoplasm accounting for approximately 4% of all primary bone tumors. It

affects skeletally mature individuals, F > M, 20 to 50 years of age. It is extremely rare in children and patient older than 60 years. Most GCTs affect the long bones with the highest incidence (65%) in the distal femur, proximal tibia and distal radius. In the long bones, the tumor is invariably centered in the epiphysis. It can also be found in any other long bone, pelvis and sacrum, and spine (3%). GCTs of the sternum and hands and feet are very rare. In approximately 30% of cases, intravascular invasion is present. Common secondary changes in GCT are hemorrhage and necrosis. fibrohistiocytic(xanthomatous) change. and aneurysmal bone cyst formation. Complications include pathologic fractures and malignant transformation (dedifferentiation). Local recurrences are common (25% - 30%) and may involve bone and/or soft tissue. Secondary malignant transformation, a rare complication seen mostly in patients with prior irradiation, may be in the form of MFH, osteosarcoma or fibrosarcoma. Primary de-novo malignant GCT is extremely rare and is characterized by marked nuclear atypia of stromal cells and atypical mitoses. 1,2,3,4,5,6,7

The major clinical manifestation is intermittent, localized pain, with or without swelling in the affected region. The pain might be caused by the local penetration of the thin cortical layer of the sternum. Imaging studies suggest the expansile, lytic lesions that do not involve the cortical bone. With the growth of the tumor, it penetrates the cortical layer of the bone and causes pain.

There are only a few isolated reports of GCTs affecting the axial skeleton; so, sternum GCT is extremely rare, and only seven cases with detailed information have been reported to the best of our knowledge. Diagnostic criteria includes moderate pain, combines with the classical radiologic features such as eccentric lytic with the presence of cortical destruction and soft tissue swelling. The most common differential diagnosis includes:benign lesions such as chondroma, fibrous dysplasia, lipoma, fibroma, and aneurysmal bone cyst; malignant lesions such as chondrosarcoma, solitary plasmacytoma, metastatic carcinoma, lymphoma, Ewing's tumor, and desmoid tumor.^{7,6,5,4,1}

CECT is useful for the anatomical location and limits of the lesion. The histological morphology of multinucleated giant cells is useful for the diagnosis of GCT. Although, immunohistochemistry can help to eliminate some lesions from the differential diagnosis. It was not performed for our cases, since the diagnosis was based on the clinical symptoms, the CT examination, and pathology Campannacci defined radiographic patterns, but radiographic three appearance does not reflect histology or clinical behavior, Type I as quiescent lesions with welldefined margins, with defined sclerotic rim, rarely involvement, Type II being active lesions cortical with well-defined, but without sclerotic rim; cortex

thinned and expanded; 70–80% of GCTs are Type II and *Type III* are aggressive lesions and often with cortical destruction and soft tissue extension and Recurrence are seen as nodular, mass-like marrow replacement (not diffuse).^{3,4,5,6,7,}

On Bone scintigraphy most lesions show increased blood pool and bone activity To date, the recurrence rate of GCT ranges from 35% to 50%, the risk of metastasis being 2%. Surgical treatment is treatment of choice including curettage, wide radical resection, and reconstruction. The risk of local recurrence is 20-40%, and the risk of pulmonary metastasis is 2% following treatment by curettage and bone graft. ^{4,5,6,7}

Learning points

Giant cell tumour although rare should be considered as one of the differentials in lytic lesions of the sternum. Diagnostic criteria includes moderate pain, combines with the classical radiologic features such as eccentric lytic with the presence of cortical destruction and soft tissue swelling.

CECT/CEMRI is the investigation of choice.

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Figure 1- Axial CECT Image, Mediastinal Window, Showing Expansile Lytic Lesion of the Sternum.



Figure 2- Saggital CECT Image Showing Lytic Lesion of the Sternum with Cortical Breach. No Invasion of Surrounding Structures. no Evidence of Calcification



Figure 3- Bone Scan Showing Uptake.







Figure 5 - Sagittal CECT Bone Window Image Showing Lytic Lesion of the Sternum With Cortical Breach. No Invasion of Surrounding Structures.

