

Giant Cell Tumour on Anterior Arc of Tenth Rib - Report of a Rare Case

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Abstract: Giant cell tumour (GCT) of bone also known as osteoclastoma is a distinct clinical, roentgenographic and pathologic entity having specific characteristics. It is a benign but locally aggressive neoplasm. Classically it is seen as a purely lytic lesion of the epiphyseal or metaphyseal-epiphyseal region of long tubular bones. The most common locations include the distal femur, proximal tibia and distal radius. We report here a case of 25 years old lady presented with a swelling over right lower lateral part of chest wall with a provisional diagnosis of primitive neuro ectodermal tumour (PNET). Computed Tomography (CT) report showed destruction of right tenth rib antero-laterally with soft tissue density lesion. The biopsy revealed giant cell tumour of rib composed of regular distribution of large sized giant cells in a background of benign appearing stromal cells. This type of tumour on the anterior arc of the rib is a very rare occurrence. Even though occur in ribs, it usually occurs in the posterior aspect and quite often the diagnosis is delayed. The patient underwent complete primary resection of the tumour and bone grafting with amelioration of symptoms. Her post operative 2 years follow up is uneventful. So we want to highlight through this rare case that chest wall GCT should be included in the differential diagnosis of any chest wall masses.

Keywords: Bone neoplasm's, Chest wall tumour, Giant cell tumour, Ribs

INTRODUCTION

Giant cell tumour (GCT) or osteoclastoma is found most often in the epiphyseal portion of the mature long bones. It is a locally aggressive neoplasm, generally arising in adults, having metastatic potential [1]. The rib is a rare site with a reported incidence of less than one percent [2]. Even in the cases involving the rib, most are located in the posterior arc (the head and tubercle of ribs) [3, 4]. Involvement of the anterior arc of a rib is a very rare occurrence with only a few cases recorded previously in the literature. This article highlights such a rare location of GCT, which should be included in the differential diagnosis of a tumour originating from the anterior arc of the ribs.

CASE REPORT

This case was diagnosed at Nil Ratan Sir Car Medical College in Kolkata. We report here a case of 25 years old female, student by occupation presented to surgery outpatient department with complaints of a painful and gradually increasing swelling over right sided lower antero-lateral part of chest wall for last 5-6 months. There was no previous history of any trauma. Her family and medical history was insignificant. Only a vague soft tissue opacity was noticed at right costophrenic angle on chest X-Ray (CXR). Initially the provisional diagnosis was primitive neuro ectodermal tumour (PNET). Simultaneously a computed

tomography (CT) of thorax was done and report revealed destruction of right tenth rib antero-laterally with a soft tissue density lesion having central hypodensity along with scattered bone fragments. Post-contrast scan showed mild heterogenous enhancement with central non enhancing area. The lesion measured 5cm × 3.5cm × 3.0 cm. The lesion was causing compression of right lobe of liver. Lungs & mediastinal structures were within normal limits. No evidence of pleural effusion or thickening was seen. Pre operative report of fine needle aspiration cytology (FNAC) was suggestive of GCT. Serum acid phosphatase level was within normal limits. Following cytological diagnosis, a wide resection of the lesion was performed including rib and tumour followed by bone grafting in the department of cardio thoracic and vascular surgery. We received a part of rib with an attached brownish, irregular and ovoid mass at one end measured 5cm × 4cm × 3 cm. Cut section showed complete replacement of the medullary cavity of the rib by a friable, haemorrhagic and necrotic material with cystic degeneration at places. Histological examination revealed almost uniform and regular distribution of multi-nucleated giant cells along with the surrounding bland looking mononuclear stromal cells. The number of nuclei in giant cells varied from thirty to fifty. The nuclei of the giant cells were similar to the nuclei of the stromal cells. No necrosis, pleomorphism or mitotic

activity was identified. Chondroid or osseous elements were also absent. Both the surgical margins of excision were free of tumour. On strict follow up the patient remained well without evidence of recurrence after two years of surgery.



Fig. 1: Gross picture of part of tenth rib with an attached irregular and brownish mass at one end

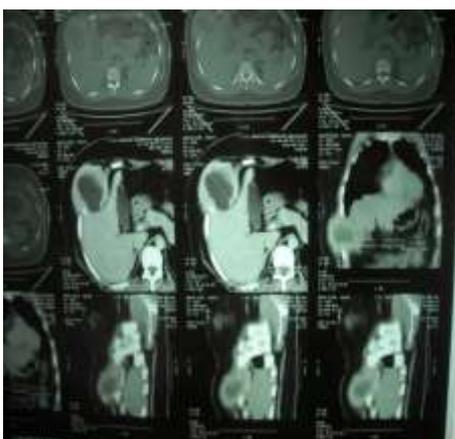


Fig. 2: CT scan of thorax shows a destructive and hypodense space occupying lesion involving the right tenth rib antero-laterally causing compression of the liver (central cross area)

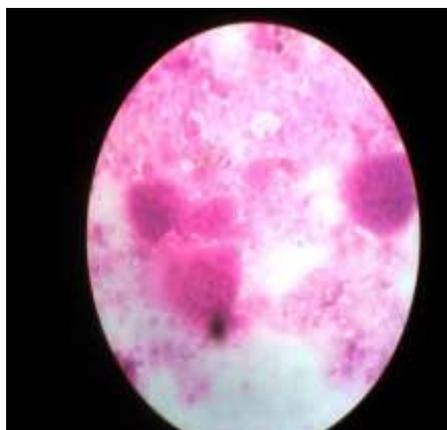


Fig. 3: Haematoxylin and Eosin stained FNAC smear showing diffuse distribution of giant cells in a

background of benign looking stromal cells (40X; High power)

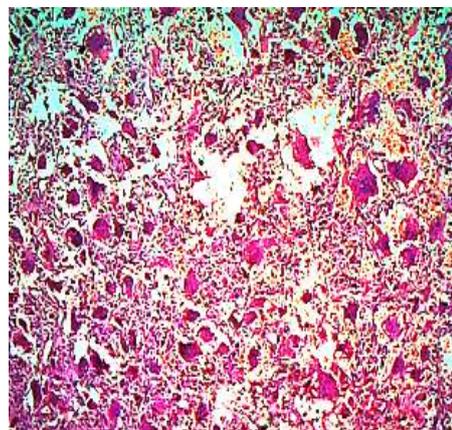


Fig. 4: Haematoxylin and Eosin stained section also shows uniform and diffuse distribution of giant cells in a background of benign looking stromal cells (10 X, Low power)

DISCUSSION

Giant cell tumour (GCT) is also known as osteoclastoma. It is a benign skeletal neoplasm arising most often from the epiphysis of the long bones. But it can involve the metaphysical area. Sometime it invades intramuscular septa by breaking through the cortex or even cross a joint space [1]. It generally arises in adults between the ages of 20 and 40 years that means after the skeletal maturation and is one of the rare bone tumour that more frequently affects women [1]. Our patient too was an adult female. It accounts for 5-9 percentages of all primary bone tumours [2]. The most common sites of involvement according to the order of frequency are lower end of the femur, upper end of the tibia and distal part of the radius [1]. The rib is a very rare site for a GCT with an incidence below one percent [2]. When it affects the ribs, it is usually at their posterior end [3, 4]. This type of tumour was first described in 1818 by Sir Astley Cooper. In 1860, Nelaton described the clinical and histological characteristics of GCTs, highlighting the fact that they are locally aggressive [5]. Gupta and Mittal did a review of the English literature and discovered 15 cases involving the rib; however, most of those involved the posterior aspect and grew inferiorly [6].

Radiographically, the typical GCT is entirely lytic and expansile without any periosteal reaction [1]. The clinical presentation of GCT is insidious onset of pain and swelling at the affected site. This non-specific symptom in many cases may be mismanaged as infection or as chronic sprain [7, 8]. A history of preceding minor trauma is infrequent. The clinical and histopathological characteristics of GCTs are similar to those of various tumours and pseudotumours such as aneurysmal bone cyst, solitary bone cyst, nonossifying

fibroma, chondromyxoid fibroma, osteoid osteoma, osteoblastoma, chondroblastoma, brown tumours seen in cases of hyperparathyroidism and telangiectatic osteosarcoma [1]. When differentiating GCTs of rib from simple bone cyst Oschner described that the latter are more likely to be formed in the anterior part of the ribs, whereas GCT are mostly located posteriorly in the epiphysis of bone (i.e., the head and tubercle of ribs) [9]. Only 3% of GCTs develops in the immature skeletons which distinguishes these patients from those with aneurysmal bone cysts, in whom the tumor maximally occurs prior to epiphyseal fusion [10].

Microscopically the tumour is composed of regular as well as uniform distribution of osteoclasts (giant and multinucleated) in a background of spindle-shaped mononuclear stromal cells. The giant cells are distributed irregularly in the above mentioned giant cell containing lesions in contrast to the true GCT [1]. The giant cells contain twenty or thirty nuclei, most of them arranged towards the center.

Microscopic grading of GCTs is not of great value except for the obviously sarcomatous lesions (grade 3). These lesions are characterised by the combination of pleomorphism, marked nuclear atypia and high mitotic activity in the neoplastic mononuclear component, often accompanied by necrosis. Giant cell-rich osteosarcoma (when osteoid production is present) and malignant fibrous histiocytoma (when osteoid production is absent) are the close mimickers of this grade 3 GCTs [1]. Our case was just a plain and simple case of GCT without any cytological atypia .

Several studies were done to know the pathogenesis of the tumour. All the evidences indicate that formation of giant cells occur by fusion of the recruited circulating monocytes inside the lesion and these are the non-neoplastic elements in comparison to the surrounding stromal cells which are rather neoplastic in nature [1].

The treatment of GCT is mainly surgical in the form of curettage with bone grafting or en-bloc excision with replacement with allograft or artificial material, depending on the location of the tumour [1]. Special care should be taken to prevent implantation of the tumour into the adjoining soft tissues.

The tumour is locally aggressive, but distant metastases are rare. The fate of metastases can be fatal. Pulmonary metastases may be solitary or multiple are seen in 2 % of patients with giant cell tumours, on average 3-4 years after primary diagnosis [11]. Some of these metastases may slowly grow (benign pulmonary implants) and some may regress spontaneously. Few factors are responsible for metastasis, such as local recurrence, surgical manipulation and location in distal

radius [12]. Hutter *et al.* reported that most recurrences (81%) appear in less than 2 years, and almost all have been manifested by 4 years. Thus at least 5 years of close follow up are recommended. However, it has been reported that the course of a benign giant cell tumor undergoing malignant transformation may take longer than 5 years [13]. However true malignant transformation is rare [14] and often follows radiotherapy. In our case the patient is under close follow up despite any evidence of recurrence even after 2 years of surgery.

Serum acid phosphatase level has been found to be increased in many cases of GCTs. It is therefore suggested that serum acid phosphatase can be used as a "tumour marker" for GCT and for monitoring treatment response and disease recurrence. It was also demonstrated that the level of this enzyme is proportional to the size of GCT [15]. In our case, serum acid phosphatase was normal preoperatively (probably due to the small size of the tumor) and remained at the same level postoperatively and during follow up.

CONCLUSION

Through this case the authors emphasize not only the rarity of a bone GCT but also its unusual costal localization. Rare sites of GCT may be diagnosed wrongly and mismanaged. Early and proper diagnosis of this type of tumour clinicoradiologically and histologically can lead to proper management. Due to the risk of local recurrence and pulmonary metastasis, the close follow up of patients is very much essential.

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