

Case Report

Giant Cell Tumor of the Thoracic Spine - A Case Report

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ABSTRACT

Giant cell tumor of the axial spine is a rare entity. The majority of cases affect sacrum. Lesions above sacrum are rare accounting for 1- 1.5% cases. They affect vertebral body primarily and may extend to neural arch and paraspinal soft tissue. Large paraspinal soft tissue component may simulate a posterior mediastinal mass. Our case was initially diagnosed as neurogenic tumor on imaging, but was subsequently proven to be a primary giant cell tumor of thoracic spine by histopathology.

Keywords: Giant cell tumor, Posterior mediastinal mass, Thoracic spine, En bloc excision.

INTRODUCTION

Giant cell tumor (GCT) of bone is a benign but aggressive lesion commonly affecting metaepiphyseal ends of long bones. It accounts for approximately 5% of all primary bone tumors in adults. However it is uncommon in axial skeleton, mostly occurring in sacrum. They are distinctly rare above the sacrum. Involvement of mobile spinal segment is seen in 1- 1.5% cases with roughly equal incidence seen in all three mobile spinal segments. They affect vertebral body primarily causing expansile lytic destruction and may extend to neural arch and paraspinal soft tissue. Large paraspinal soft tissue component may simulate a posterior mediastinal mass. [1-4]

Various modalities of treatment are available for spinal GCTs such as surgery, radiotherapy, embolization, cryosurgery, cementation, and chemical adjuvant like phenol or liquid nitrogen but optimal treatment has not been well established and the recurrence rate is high despite the best management. Whenever possible, en bloc excision should be pursued as the surgical

procedure of choice for spinal GCT as it has the least recurrence rate. They carry much worse prognosis than the lesions in long bones. [5]

CASE REPORT

A 38-year-old male patient presented with upper back pain and difficulty in lifting right upper limb. Chest radiograph (posteroanterior and lateral view) showed a oval shaped opacity in right postero-superior mediastinum (Figure 1).

MRI showed a highly enhancing soft tissue mass lesion of approximate size 52×46×54 mm in right paravertebral region with subtle lytic destruction of D2 vertebral body and there was intraspinal extension through D2, D3 neural foramen producing characteristic “dumbbell” shaped mass (Figure 2 & 3). There was homogenous enhancement on contrast MRI. Hence a provisional diagnosis of nerve sheath tumor was given. He was advised for computed tomography which well depicted bone destruction of D2 vertebral body, posterior elements and right second rib (Figure 4).

Post-op histopathological examination of the tumor was suggestive of GCT of bone (Figure 5).

He underwent right posterolateral thoracotomy and partial excision of soft tissue component of the mass. He is being planned for adjuvant radiotherapy.

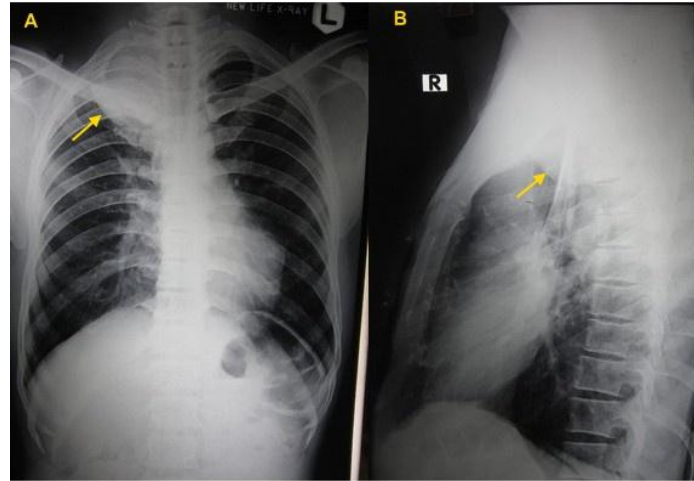


Figure 1: Posteroanterior (A) and lateral (B) chest radiograph showing an oval shaped mass in the right postero-superior mediastinum (arrow).

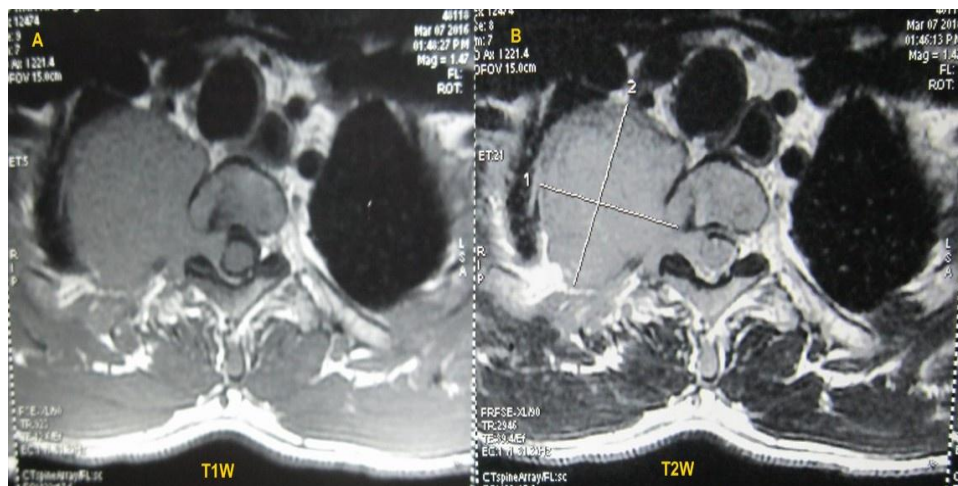


Figure 2: The mass is isointense to spinal cord on T1W imaging (A) and mild hyperintense to spinal cord on T2W imaging (B). Note the intraspinal extension of the mass through neural foramen causing “dumbbell” appearance.

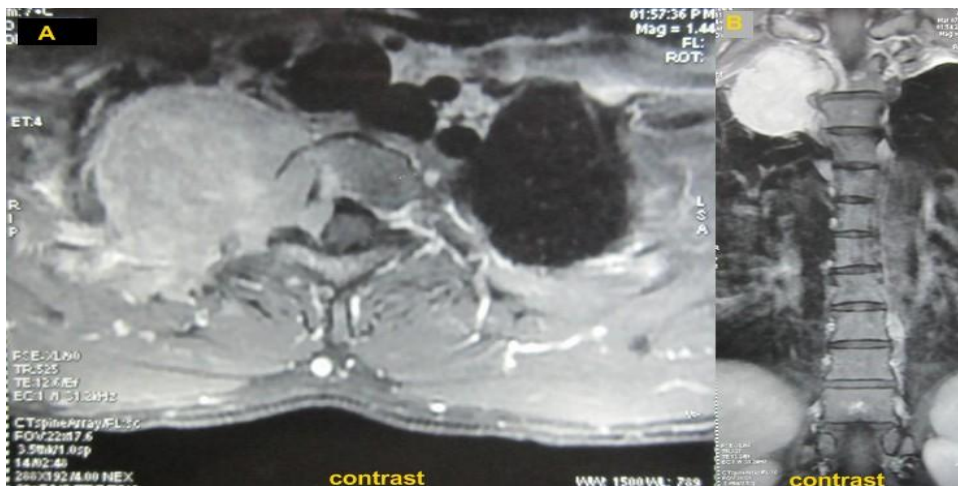


Figure 3: T1contrast MRI axial (A) and coronal (B) images showing homogenous enhancement of the mass. Also noted is the D2 vertebral body & posterior element involvement.

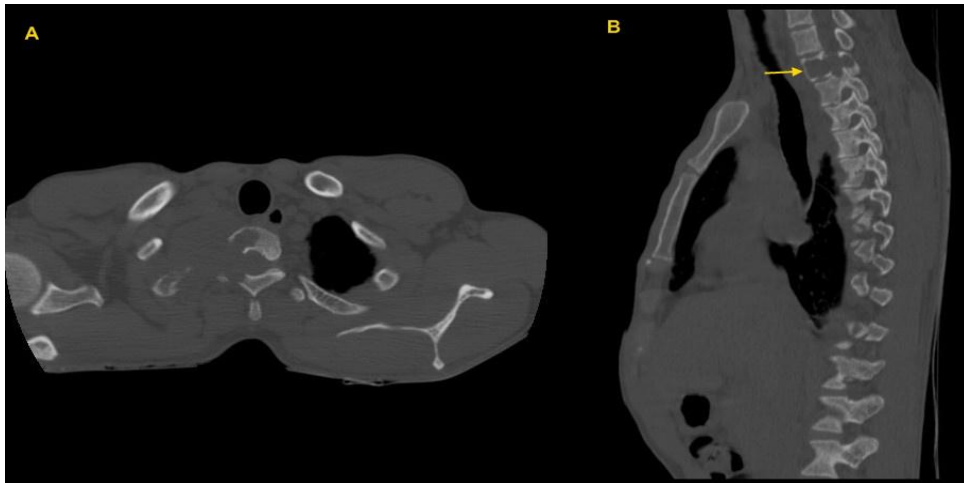


Figure 4: Computed Tomography (CT) axial (A) and sagittal (B) section nicely depicting lytic destruction of right half of D2 vertebral body, posterior element and adjacent second rib. Only a thin rim of anterior vertebral body is spared (arrow).

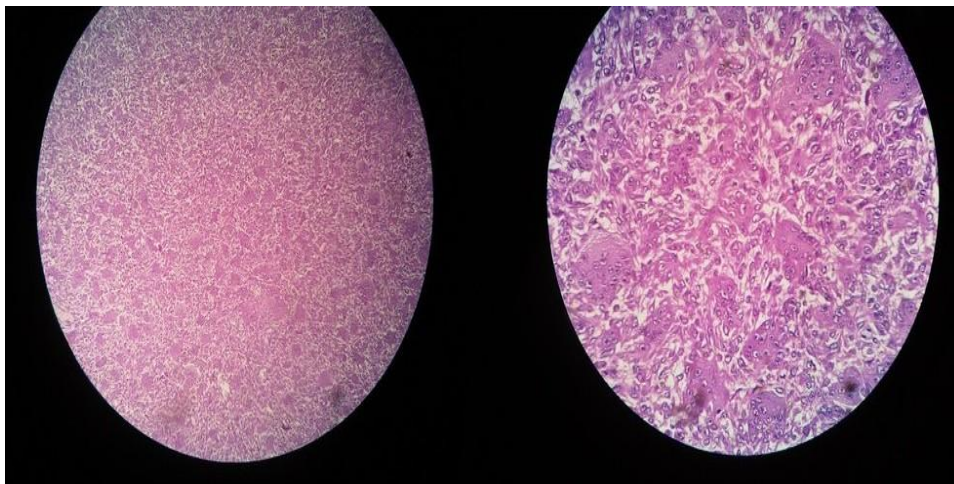


Figure 5: Photomicrograph showing uniform distribution of multinucleated osteoclastic giant cells against a background of round to spindle shaped mononuclear stroma cells.

DISCUSSION

GCT of thoracic vertebrae is quite infrequent. Sacrum is commonly affected in axial spine. They were previously termed as osteoclastoma because they were considered to arise from the osteoclasts. However, the exact cell of origin is unclear. [1-5]

It is common in age group of 20-45 with equal sex incidence. Most patients present with pain or neurologic deficit at the site of tumor involvement, and symptoms are usually present for many months prior to diagnosis. Other symptoms are arm or leg weakness, paresthesias, diaphragmatic paralysis, constipation and structural deformity of the spine. They even can be asymptomatic and diagnosed incidentally on imaging. [2,5,6]

The radiographic characteristics of spinal GCT are considered to be a round or oval extrapleural mass with shell-like calcification of the marginal lesion and the absence of a mineralized matrix. [2,4,5,7] Spinal GCT usually causes lytic destruction of vertebral bodies and there may be extension to posterior elements, spinal canal and paravertebral soft tissues. They can have intraspinal extradural extension through neural foramen and hence can cause compressive myelopathy. [7] Posterior elements are commonly involved in other spinal bone tumor like aneurysmal bone cyst, osteoid osteoma, and osteoblastoma. [4,5,7,8]

Like GCT of long bones, GCT of the spine and sacrum can develop benign lung metastasis which has been reported to occur

in up to 13.7% of the spinal lesions (vs less than 2% in long bones GCT). [5,6]

Spinal GCT has some characteristic findings on MRI which help in differentiating from other spinal bone lesion. [7,8] On MRI following parameter are considered while describing GCT of spine: the signal intensity on the T1- and T2-weighted images relative to spinal cord, enhancing pattern, presence or absence of curvilinear area of low signal intensity in the mass, tumor extent, presence or absence of spinal canal involvement, compression fracture of the involved vertebrae, cystic changes within the mass and fluid-fluid level within the mass. GCT usually has a low to intermediate signal on T1-weighted images. They have variable signal intensity on T2W imaging. They have low to similar signal intensity to the normal spinal cord on the T2-weighted MR images in 63-96% of cases. This appears to be caused by the relative collagen content of fibrous components and hemosiderin within the tumor. [7] This feature on T2W imaging is quite helpful in making a differential diagnosis as most of the spinal neoplasms (metastases, myeloma, lymphoma, and chordoma) show high signal intensity on T2W images. Cystic areas and fluid-fluid level can be seen due to secondary aneurysmal bone cysts in these lesions which are more common in sacral GCT. [4,7,8]

On computed tomography, these lesions are usually isodense (with respect to the paraspinal muscle) with a thin rim of sclerosis and show a homogeneous hypervascular appearance with contrast enhancement. Expansile lytic bone destruction with vertebral collapse is well seen. Involvement of the adjacent intervertebral disks and vertebrae is not unusual simulating an infectious process. Angiography of GCT usually reveals a hypervascular lesion. [7, 8]

At bone scintigraphy, GCT may demonstrate peripheral increased uptake of radionuclide (doughnut sign). [8] Definite diagnosis based solely on imaging is not

possible and biopsy of the lesion remains the mainstay of an ultimate diagnosis.

The histologic appearance of GCT is a uniform distribution of multinucleated giant cells against a background of round to spindle shaped mononuclear stroma cells as in long bone GCT. Multinucleated giant cells can be seen in other bone tumors like chondroblastoma, fibrous dysplasia, eosinophilic granuloma, chondromyxoid fibroma, telangiectatic or fibrogenic variants of osteosarcoma, and malignant fibrous histiocytoma and hyperparathyroidism and Paget disease of bone. [4,7-10]

Many treatment modalities are available for spinal GCTs such as surgery, radiotherapy, embolization, chemotherapy, cryosurgery, cementation with varying degrees of success. But definite treatment of GCT in the sacrum and spine is not well defined. Surgery is the treatment of choice and wherever possible en bloc surgical excision to completely remove tumor followed as it has the lowest recurrence rates. This is followed by stabilization and reconstruction of the spine with autografts and instrumentation. The disadvantage of en bloc resection is that it is associated with increased risk of permanent neurologic deficit. [5,6,11] For large tumors where en bloc resection is not attempted because of fear of neurologic deficit, pre-operative embolization followed by intralesional resection of tumor might be considered. The recurrence rate of spinal GCT following en bloc excision has been reported to range from 11-50% as compared to 0-71% in case of intralesional excision. [5] Use of adjuvant radiation therapy after surgery has been controversial as it is associated with relatively high risk of radiation-induced malignancy. In surgically inaccessible tumors, selective arterial embolisation is preferred to primary radiotherapy. [5,6,11] In case of recurrences, Serial selective arterial embolization until complete devascularization is achieved is the treatment of choice. Medical treatment included bisphosphonates and denosumab. [6]

Pulmonary metastasis is treated with chemotherapy and surgery. Since there is no specific treatment algorithm of this aggressive tumor, treatment option is tailored to individual case. Because of the possibility of local recurrence or the development of pulmonary metastases Close follow-up is required in all patients for 10 years. [11]

CONCLUSION

Though giant cell tumor of the spine is uncommon, it should be kept in the differential diagnosis of expansile lytic bone lesion affecting vertebral body. Large Soft tissue component simulating a posterior mediastinal mass is still rarer. Though MRI findings are somewhat characteristic, definitive diagnosis based solely on MR characteristics may not be always possible and histopathology remains the gold standard.

GCT of spine presents as a aggressive tumor with high local recurrence rates as compared to GCT of long bones. Among various treatment modalities available, en bloc surgical resection is the treatment of choice. Local recurrences are treated with selective arterial embolisation and radiotherapy. Despite this GCT of spine carry a much worse prognosis.

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