Case report

Giant cell tumor of the thoracic spine presenting as a huge posterior mediastinal mass: Case report and review of the literature

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There are a few case series of giant cell tumor (GCT) in the spine and sacrum. GCT in the thoracic vertebrae is even more exceptional in medical literature. In this article we report a GCT in a 25-year-old male who presented with progressive back pain and dyspnea. On contrast-enhanced multidetector computer tomography, a large heterogeneously enhancible posterior mediastinal mass was detected with adjacent bony erosion and right main bronchus narrowing. At surgery, resection of large posterior mediastinal mass and curettage of adjacent body vertebrae with stabilization were performed. The final histological examination revealed that these findings were compatible with GCT from thoracic body vertebra.

KEYWORDS: Giant Cell Tumor, Mediastinal Mass, Thoracic Vertebrae, Bone Neoplasm

BACKGROUND

Giant cell tumors (GCTs) are locally aggressive benign bony lesions with a high incidence of recurrence and have malignant potential in recurrence. They account for approximately 5% of bone tumors and over 20% of benign bone neoplasms.[1] GCT occurs most commonly at juxta-articular metaphysis and epiphysis of long bones.[2] In terms of frequency, the bones that can be affected include distal femur and proximal tibia (knee), distal radios, sacrum, distal tibia, proximal humerus, pelvis and proximal femur.[3]

The spinal axis is involved in 8-11% of GCTs and order of frequency in spine is sacral, thoracic, cervical and lumbar, so the majority of this tumor in spine is detected in sacrum.[1,4] In several large series, only 1-2% of GCTs occurred in the thoracic spine.[5]

GCT can be found with a slight predominance in female patients with a peak incidence in the third and fourth decade of life.[1] Although localized pain is the most familiar symptom of patients with GCT of the spine, but radicular and neurologic symptoms may appear in a subset of them.[6]

STAGING: Both CT and MRI accurately demonstrate the intraosseous and soft tissue extension of the tumor. CT is better as showing cortical destruction, whereas MRI is better at demonstrating intra-articular involvement.[7]

GCT is staged according to the Enneking's staging system for benign bone tumors: stage 1, latent lesion that is biologically static; stage 2, active, slow-growing lesion confined within bone; and stage 3, locally aggressive with soft tissue extension. GCTs may also be stratified according to the Campanacci system that is based on plain radiographs: grade 1, well marginated border with a slim rim of mature bone; grade 2, relatively well defined margins without a radiopaque rim; and grade 3, indistinct borders suggesting rapid permeative growth, soft tissue extension.[3]

The radiographic characteristics of spinal GCT are considered to be a round or oval extra pleural mass with rim–like calcifications of the peripheral aspect and the absence of a mineralized matrix. On non-contrast CT scans, a spinal GCT was reported to show a homogeneous expansile soft tissue density mass like region that has homogeneous hypervascular appearance with contrast enhancement.[1] Heterogeneous density or a fluid level due to hemorrhage or necrosis within the GCT[1,5] was not observed in previous cases.

The MR images provided more information on both tumor location and extension than did the CT images. GCT usually has a low to intermediate signal on T1-weighted images and a predominantly high signal with some low to intermediate signal intensity regions on T2-weighted images that caused by the high collagen content of fibrous components and the hemosiderin deposition within the tumor.[6-9] After administra-

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Received: 07.01.2012; **Revised:** 09.02.2012; **Accepted:** 26.02.2012

tion of gadolinium, a GCT shows several degrees of contrast enhancement on T1-weighted images.[6,7]

To the best of our knowledge, this article is the fourth reported case of thoracic spinal GCT that manifested with a huge mediastinal mass.[2,8,9] We report one case that had a large posterior mediastinal mass that is a rare manifestation.

CASE REPORT

A 25-year-old man was admitted to the hospital with 3 years history of diffused progressive back pain and dyspnea. His back pain was aggravated during the last 12 months that promoted radiographs and computed tomography (CT) scans.

Radiographs showed a huge radiolucent soft-tissue mass in the right mediastinum. Contrast enhanced multi-sectional CT of thorax in axial section revealed extrapleural hypervascular huge posterior mediastinal mass with contrast enhancement and heterogeneity that little adjacent bony erosion was present.

The tumor had spread to the mediastinum from local invasion and was in contact with vital structures involving the trachea, esophagus and aortic artery. Moreover, the mass had extended to the right mediastinum with pressure effects caused anterior shifting of main bronchus but other mediastinal structures were not displaced by the mass (Figure 1-4).

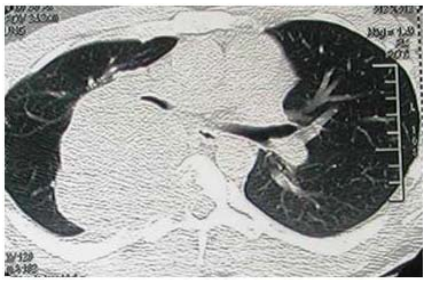

Figure1. A 22-year old male presenting with back pain and dyspnea that contrast enhanced CT scan (lung window) showed a large posterior mediastinal mass with adjacent bony erosion

The patient rejected additional imaging studies including magnetic resonance imaging (MRI) due to his claustrophobia. A fine-needle aspiration of the mass was performed and analysis of sections of the biopsy showed spindle-shaped cells with multinucleated giant cells which consisted the histological diagnose of GCT (Figure 5).

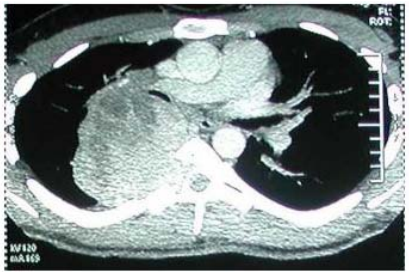

Figure 2. A 22-year old male presenting with back pain and dyspnea that contrast enhanced CT scan (mediastinal window) showed a heterogeneously enhancible large posterior mediastinal mass with bony erosion of adjacent thoracic vertebra

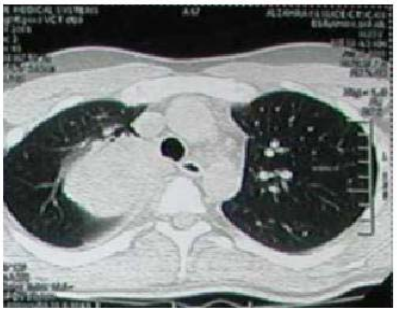

Figure 3. A 22-year old male presenting with back pain and dyspnea that contrast enhanced CT scan (lung window) showed a posterior mediastinal mass with RT paratracheal extension

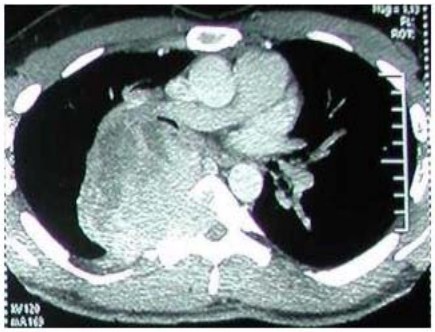

Figure 4. A 22-year old male presenting with back pain and dyspnea that contrast enhanced CT scan (mediastinal window) showed a heterogeneously large enhancible posterior mediastinal mass with adjacent bony erosion and narrowing and anterior shifting of RT–main bronchus

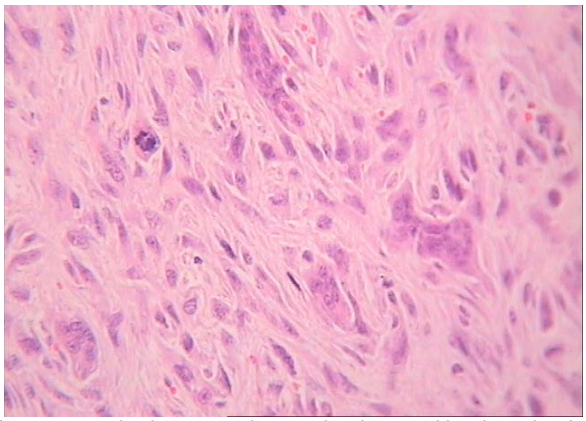

Figure 5. Histology specimen showing multinucleated giant cells composed of spindle shaped and mononuclear cells (magnification ×400)

After biopsy, the patient underwent resection of huge posterior mediastinal mass beside curettage of adjacent body vertebrae with stabilization. The final pathologic assessment of the incised tumor verified that these findings were compatible with GCT of bone arising in the thoracic vertebrae.

DISCUSSION

GCT documented by Cooper and Travers for the first time in 1818.[7] After near 2 century, several large cases of patients with GCT were observed. GCT of bone is a benign, but potentially aggressive tumor and may show local invasion; this tumor most commonly arises from the metaphysis and epiphysis of the long bones.[2] They are found most frequently in young adults, with a female predilection.[1]

Although the spine is the fourth location of GCT but occurs infrequently in the spine above the sacrum and only 5% of these tumors is seen there.[1] GCT presence in the thoracic vertebrae is even more exceptional that only involves 1% to 2% of GCTs.[5]

GCTs of the spine sometimes extend into the adjacent paravertebral soft tissue,[1] but a primary thoracic spinal GCT simulating a huge mediastinal neoplasm in medical literature is rare. Kim et al.[2] reported a case of 29-year-old man who presented with primary thoracic spine GCT manifesting as a huge posterior mediastinal tumor with benign pulmonary metastasis. Kathiresan et al.[8] described a 24-year-old woman with a large GCT tumor of the thoracic spine presenting with spinal cord compression during pregnancy which treated after delivery.

Gee et al.[9] reported a 44-year-old woman with an 8 cm mass arising from thoracic vertebral body GCT in which soft tissue extension to the spine complicated management. Sakurai et al.[5] reported a 34-year-old woman of GCT of the thoracic spine noticed by a well-defined oval mass in the right upper mediastinum, but in this case the mass was not as huge as cases cited above. We report a 25-year-old woman with GCT of the thoracic spine manifesting posterior mediastinal mass that was presented with back pain and dyspnea.

The primary tumor diagnostic approaches are plain radiography, computed tomography (CT) and magnetic resonance imaging (MRI). On plain film radiography, GCT of bone is commonly noticed as a destructive osteolytic lesion which makes a mouse-eaten appearance. Spine GCT most often involves vertebral bodies that having expansion and thinning.[10] Radiography can be more useful while GCT occurs in the epiphyseal and metaphyseal part of long bones; as a result, the radiographic changes are less diagnostic in the spine.

Chest radiography is suggested to diagnose huge expansile mediastinal tumor or pulmonary metastasis that may be presented in a very small subset of patients. However, it was mentioned in a recent study that benign lung metastasis occurs in up to 13.7% of the spinal GCT patients.[11]

On CT scans, usually a lytic expansile lesion with narrow zone of transition manifests; also thinning and erosion of the cortex is present but calcification within the tumor is absent.[3,12] Moreover, spinal GCT shows a homogenous hypervascular appearance with contrast enhancement.[13] Low grade of heterogeneity can be observed as a result of hemorrhage or necrosis in the mass.[13]

On MR imaging, giant cell tumors are well defined with a low signal intensity rim or halo surrounding the tumor.[3] On T1-weighterd images, the tumor usually has a low to intermediate signal intensity, on T2-weighted images; it has an intermediate to high signal intensity.[12]

The radiographic differential diagnosis include brown tumor of hyperparathyroidism, hematologic malignancies, metastatic disease, aneurysmal bone cyst, giant cell reparative granuloma, and chondroma as well as benign or malignant fibrous histiocytomas.[14] However, histological analysis of a biopsy specimen is required to satisfy the diagnose of GCT.

Other pathologic conditions that can manifest as a paraspinal soft tissue mass include neurogenic tumor, lymph adenopathy, infectious spondylitis, hematoma due to spinal trauma, spinal tumor and extramedullary hematopoiesis in anemia and myelodysplastic syndrome.

Spinal GCTs are usually expansile lesion with bone destruction that affects the vertebral body and can cross disk spaces and extend into the posterior elements, but involvement in the posterior elements was observed with other spinal bone tumors, as aneurysmal bone cyst and osteoid osteoma or osteoblastoma.[13]

In the spine, the initial surgery of GCT should be as aggressive as possible within neurological preservation and spinal stability. In case of partial removing of tumors, the radiation therapy is a useful added treatment

with less risk of recurrence even though it gives a chance of malignant transformation.[15]

CONCLUSIONS

We presented a rare case of giant cell tumor of the thoracic spine with posterior mediastinal mass that presented with back pain and dyspnea. There is no similar report for spinal GCT presenting with dyspnea. Our case is the first report of dyspnea as a presentation of spinal GCT.

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How to cite this article: Tayar N, Sirous M, Sirous AM. Giant cell tumor of the thoracic spine presenting as a huge posterior mediastinal mass: Case report and review of the literature. J Res Med Sci 2012; 17(Spec 2): S314-S317.

Source of Support: Nil, **Conflict of Interest:** None declared.