Giant-cell Tumor of Dorsal Vertebra Presenting as a Posterior Mediastinal Mass: A Rare Case Report

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ABSTRACT

Giant cell tumors (GCT) of bone also referred to as osteoclastoma or myeloid sarcoma are usually benign tumors but have the potential for local recurrence and metastasis. Giant cell tumors mostly involves the long bones with the proximal tibia and distal femur sites. Giant cell tumors of the spine presenting above the level of the sacrum is relatively less common. We report a patient with GCT arising from dorsal vertebra and presenting as a huge mediastinal mass.

A 25-year-old female presented with bilateral lower limb weakness, breathlessness, and difficulty in speaking. Chest X-ray showed bilateral upper lobe mass. Further evaluation with contrast-enhanced computed tomography (CECT) of the thorax revealed a well-defined heterogeneously enhancing posterior mediastinal mass with necrotic areas within and bony infiltration was noted. Sequential chest X-ray was suggestive of rapid progression within 15 days. Ultrasound-guided biopsy of mass lesion revealed features of giant cell tumor with immunohistochemical markers strongly positive for CD68, negative for S100, and β-HCG. The patient was subsequently started on denosumab given unresectable pulmonary metastasis.

Keywords: Case report, Dorsal vertebral lesion and denosumab, Giant cell tumor.

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ABBREVIATIONS USED IN THIS ARTICLE

CECT = Contrast-enhanced computed tomography; GCT = Giant cell tumor; MRI = Magnetic resonance imaging; RT = Radiation therapy.

INTRODUCTION

Giant cell tumor (GCT) commonly involves ends of long bone and usually occurs in the third or fourth decade with a female preponderance. Though predominantly a benign tumor, it is very aggressive with a high incidence of local recurrence. The most common sites are the distal femur, proximal tibia, and the distal radius. Vertebral giant cell tumor affects the spinal column in about 5% of cases. The most common site in the vertebral column is the sacrum and it is rarely reported above the sacrum with involvement of the thoracic spine in only 1–2%. The majority of GCT are benign with only 5% constituting malignancy.

CASE DESCRIPTION

A 25-year-old female presented to the Emergency services with complaints of weakness of both lower limbs, difficulty in breathing for 4 months, and difficulty in speaking for 10 days. She also had a history of back pain, and urinary and fecal incontinence. She had no history of altered sensorium, upper limb weakness, stridor, trauma, decreased neck movements, or fever. She also had dysphagia to solid foods. She had no cough with expectoration or chest pain. She had been treated elsewhere and was referred to our center. On examination, her vitals were stable. She was conscious and oriented and had no neck stiffness. Bilateral upper limb power, tone, and reflexes were found to be normal. But in bilateral lower limbs, power was 2/5 with spasticity and hyperreflexia noted. Respiratory system examination revealed bilateral decreased air entry. Heart sounds were heard normally. A routine chest X-ray was done in ED which was suggestive of bilateral upper lobe non homogenous opacity. On comparing with a previous X-ray done 15 days back, it was noticed that the size of the opacity has increased (Fig. 1). Given rapid progression, a CECT thorax and abdomen was done which revealed a large well defined heterogeneously enhanced mass lesion in the bilateral upper lobe with necrosis, with the lesion extending to the posterior mediastinum and infiltrating T2–T6 vertebral body levels with collapse of T2–T3 vertebral body with kyphosis.

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Anteriorly it was compressing the esophagus with close proximity to the great vessels. A filling defect was noted involving the right upper lobe pulmonary artery and its subsegmental branches and azygous vein suggestive of tumor thrombus. A loculated fluid attenuating collection with underlying lung collapse was noted in the apicoposterior segment of the left lung (Fig. 2). With the clinical and radiology images, a possible differential diagnosis of lung mass extending into the vertebra, neurogenic tumor, germ cell tumor, and lymphoma was considered. Abdomen cuts showed no organomegaly or free fluid. Tumor markers for possible mediastinal tumors were sent. Her alpha feto-protein level was 2.0 ng/mL (normal value 0–8.5 ng/mL). The β-HCG was 11.3 (normal values 0–5 IU/mL). Magnetic resonance imaging (MRI) was not done due to technical issues.

Ultrasound sonography (USG) guided biopsy taken from heterogenous mass in paraspinal region showed sheets of round to oval mononuclear cells with bland nuclear morphology. These cells were mixed with many osteoclast-type and giant cells. Immunohistochemical markers for these tumor cells were strongly positive for CD68, and negative for S100, and β-HCG which suggested the possibility of GCT of bone (Fig. 3). She was started on subcutaneous injection of denosumab 120 mg every 28 days, with two additional loading doses on days 8–15 of the first month given unresectable upper thoracic vertebral mass with pleural
effusion. She was also started on therapeutic anticoagulation given pulmonary artery thrombosis. After treatment for 1 month, patient defaulted for follow-up. Repeated attempts to contact her failed.

**Discussion**

Giant cell tumors of bone is a relatively uncommon, benign but locally aggressive osteolytic bone neoplasm occurring in young adults. Giant cell tumor accounts for 5% all primary bone tumors. Spinal GCT mostly occurs in the sacrum followed by the thoracic spine, lumbar and cervical vertebra. Giant cell tumors occurs after skeletal maturity, with a peak incidence in patients with age 30–40 years. Symptoms of spinal GCT include back pain, limb weakness due to spinal cord compression, bowel and bladder dysfunction, and structural deformity of the spine. Our patient was in her 20s and had dysphagia and dyspnea because of compression of the trachea and esophagus.

Radiologically GCT appears as a round or oval extra pleural mass with shell-like calcification of the marginal lesion with the absence of a mineralized matrix. Differential diagnoses of the mass can be neurogenic (nerve sheath tumor, parasympathetic ganglion tumor, sympathetic chain tumor), nonneurogenic (like chordoma, lymphoma, metastasis), infections (paraspinous abscess), foregut duplication cyst, vascular (Descending thoracic aortic aneurysm, varices) and lymphadenopathy. Giant cell tumors involve anterior elements of the vertebra like vertebral body and soft tissue. In contrast, other bony tumors like an aneurysmal bone cyst, osteoid osteoma, and osteoblastoma involve the posterior elements of the vertebra like articular processes, pedicle, lamina, and spinous process. This case presented with bilateral upper lobe mass with involvement of posterior mediastinum, dorsal vertebral (posterior elements) involvement, and loculated effusion.

Giant cell tumors of the spine occasionally extends to paraspinal soft tissue. In our patient, the lesion was arising from dorsal vertebra simulating a dumbbell tumor (bilateral upper lobe mass with communication through the posterior mediastinum) which is a rare presentation.

Pulmonary metastasis in GCT is seen in 1–6% and occurs in GCT of the spine compared to extremities. It is commonly diagnosed during the evaluation of a recurrent GCT. Several mechanisms have been suggested as to the reason for metastasis. These include tumor embolism, biologic predetermination of the tumor cells to invade the interstitium, and consequent spread to distant organs. It is also suggested that interventions such as curettage can sludge and deposit the tumor into the blood vessel with subsequent metastasis. The metastatic lesions are usually slow-growing and usually don’t require treatment and resolve spontaneously. Surgical excision of the metastasis is required in cases with tumor-related symptoms like hemoptysis, bronchial obstruction, and lung collapse. Surgical management includes metastasectomy, wedge resection, or lobectomy. In case of unresectable pulmonary metastasis, radiation therapy (RT) or denosumab may be offered.

In our case, as the lesion primary vertebral lesion was extending into the posterior mediastinum and resembling a posterior mediastinal mass, it was a diagnostic difficulty. For differentiation between primary vertebral tumor and posterior mediastinal mass histopathological diagnosis and immunohistochemical markers are necessary. The distinction between benign and malignant GCT may
be difficult as the transformed element is relatively difficult to detect in the presence of numerous reactive giant cells. The histological appearance of GCT is that of sheets of round to oval, polygonal, or elongated mononuclear cells that are interspersed with uniformly distributed, large osteoclast giant cells. In osteoclastoma, giant cells show positivity for CD45, CD68, and Cathepsin K. The p63 shall be positive in more than 95% of cases however it’s not specific.

The different recommended treatment protocols for spinal GCTs are surgery, radiotherapy, cryosurgery, cementation, and chemical adjuvant therapy. Surgery is the treatment of choice for long bone giant cell tumor, and surgical options include intralesional curettage (with or without bone cement), en bloc resection, or marginal excision. Surgery may be a difficult option for GCT of the spine and pelvis. Campanacci grading system (depending upon clinical and radiographic appearance) also influences the type and extent of surgery for GCT. Intralesional curettage have a high risk of local recurrence compared to extensive surgery but the addition of bone cement as an adjuvant to intralesional curettage can decrease the local recurrence rate. Local recurrence can be managed by curettage but extensive surgery may be needed for later recurrence. Chemotherapy (ifosfamide, cyclophosphamide, cisplatin doxorubicin) is not a standard approach in benign but can be used in malignant giant cell tumor. Lifelong posttreatment surveillance is important because of increased risk of locoregional recurrence and metastatic disease. The frequency of posttreatment imaging can be decided upon the clinical response. Recently, denosumab a monoclonal antibody that targets the RANK ligand is shown to be beneficial. Denosumab binds to receptor activator of nuclear factor Kb (RANKL) which stops the activation of RANK. This prevents further recruitment of osteoclast-like cells and bone resorption. Denosumab can be tried for patients with local recurrence and who are poor candidates for surgery.

**CONCLUSION**

Thoracic spine GCT though uncommon, should be considered in the differential diagnosis of lung mass with thoracic vertebral involvement. Clinicians need to suspect bone tumors when a patient presents with an aggressive spine lytic lesion.

**REFERENCES**