Giant Cell Tumor of the Sixth Thoracic Vertebra: Case Report

Altıncı Torasik Vertebranın Dev Hücreli Tüümörü: Olgu Sunumu

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ABSTRACT

Giant cell tumor is an uncommon but most aggressive benign tumour of the spine with unpredictable outcome and challenging treatment. Spinal giant cell tumors located above the sacrum are rare and treatment recommendations are still unclear. We report a rare case of this lesion in an adult and discuss the management and outcome of such uncommon tumors. A 31-year-old woman presented with progressive motor weakness of both lower limbs with back pain during the past month, associated with sphincter disturbances for the past two days. She was diagnosed with a lytic heterogeneously enhancing mass depending mainly on the T6 posterior arch with small vertebral body involvement. The tumor extent reached surrounding soft tissue and the spinal canal with marked spinal cord compression. A posterior approach was realized as an emergency. Histological examination showed evidence of a giant cell tumor and a complementary irradiation was used. The patient improved well post operatively. There was no recurrence or metastasis over 5 years of follow-up.

KEYWORDS: Giant cell tumor, Thoracic spine, Magnetic resonance imaging, Surgery

ÖZ


ANAHTAR SÖZÇÜKLER: Dev hücreli tümör, Torasik omurga, Manyetik rezonans görüntüleme, Cerrahi

INTRODUCTION

Giant cell tumors represent approximately 5% of bone tumors and over 20% of benign skeletal neoplasms. They typically arise in the metaepiphysseal ends of long bones (3) and occur with a slight predominance in female patients older than 19 years (10). Spinal giant cell tumors are rare and account for 2.7% to 6.4% of all giant cell tumors of bone (9). They represent the most aggressive benign tumor of the spine with unpredictable outcome (2). The management of giant cell tumors of the spinal column presents a challenge because of their anatomic location and it is agreed that radical resection achieves the best results but recommendations for rare locations like the spine are still unclear (2, 3, 4). This article describes a rare case of thoracic spine giant cell tumor and discusses the management and outcome of such uncommon lesions.

CASE REPORT

A 31-year-old woman presented to our hospital complaining of a three-week history of back pain and progressive motor weakness of both lower limbs, associated with sphincter disturbances for the past two days. Nothing special was noted in her medical history.

On admission, the patient’s neurological examination revealed paraparesis with motor strength of 3/5 in both lower limbs, a T8 sensory level, exaggerated knee and ankle reflexes along with extensor planter response on both sides. Chest X-ray was normal and magnetic resonance imaging (MRI) showed a lytic heterogeneously enhancing mass depending mainly on the T6 posterior arch with small vertebral body involvement. The tumor extent reached surrounding soft tissue and the spinal canal with marked spinal cord compression (Figures 1, 2).

A single-stage posterior approach was realized as an emergency. We found a hemorrhagic dark red-colored soft mass that expanded the bone of origin. The T6 posterior arch, tumor and intracanal portion were completely removed and no stabilization was required.
Histological examination of the tumor specimens revealed a giant cell tumor and a complementary irradiation was administered.

The patient’s immediate postoperative course was uneventful, and she recovered quickly. Her neurological condition improved continuously, and follow-up examinations showed that the paraparesis almost completely disappeared. At last follow-up, 5 years after surgery, she is able to walk alone with no radiological recurrence or metastasis (Figure 3).

**DISCUSSION**

Giant cell tumors were first described by Cooper & Travers in 1818. They are most frequently seen in young adults with slight female predominance (3) and account for approximately 5% of all primary bone lesions and 20% of benign skeletal tumors. Their osteolytic potential is mediated by RANK ligand (RANKL), a key factor for osteoclast differentiation and activation (6).

Spinal giant cell tumors are rare and account for 2.7% to 6.4% of all giant cell tumors of bone (9). Above the level of the sacrum, these tumors are equally distributed in the cervical, thoracic and lumbar spine (10). Spinal giant cell tumors are usually located in the vertebral body (55%) and less frequently within the body and arch (29%) and are more rarely limited to the vertebral arch (16%) (10).

In our case, the tumor developed mainly on the T6 vertebral arch with a small T6 vertebral body involvement that is not a commonly described feature in the literature.
Pain is the most common presenting symptom for spinal giant cell tumors, with an average duration of 6 months before diagnosis. This often gets compounded by nerve root symptoms (30%) or paraparesis (16%) due to local neurological involvement (10). An asymptomatic, incidental radiological occurrence is infrequent. In our case, back pain and motor weakness were reported for the past month only, suggesting a more rapid growth than previously noted.

Radiographically, giant cell tumors of the bone usually present as a destructive, osteolytic lesion on plain films. On computed tomography (CT), spinal giant cell tumors are often surrounded by a narrow zone of transition and show homogenous contrast enhancement. MRI is ideal for determining the extent of the disease, particularly for assessing tumor involvement of great vessels and spinal cord, as it provides high tissue contrast and multiplanar views. MRI of spinal giant cell tumors shows an intermediate to low signal mass on T1-weighted sequences, intermediate to hyper signal on T2-weighted sequences and variable enhancement with gadolinium. A surrounding rim of low T1 signal was described and may be due to haemosiderin deposition or reactive bone formation. In our case, the tumor had hypo signals on T1-weighted sequences and hyper signals on T2-weighted sequences, and showed heterogeneous enhancement with gadolinium. Its extent reached to the perivertebral soft tissue and the spinal canal.

Spinal giant cell tumors are radiologically classified according to the Enneking surgical staging system (8). In the present case, the tumor was considered as Stage III Enneking class due to the soft tissue mass accompanying the cortical breakthrough.

The radiographic differential diagnosis mainly includes brown tumor of hyperparathyroidism, metastatic disease, hematologic malignancies, chordoma and aneurysmal bone cyst. Nevertheless, definitive histological analysis before treatment should be performed and CT-guided needle biopsy is recommended.

Optimal management of giant cell tumors of the bone is complete tumor resection with wide margins if possible, (9) but this approach is significantly more challenging when tumors arise in the axial skeleton. These considerations oblige both surgeons and patients to choose between radical resection (total spondylectomy) with potentially devastating morbidity, and intralesional removal with higher rates of recurrence (10). In our situation, the patient presented with rapid neurological deterioration due to a posterior expansible lesion requiring immediate surgery.

Although Boriani et al. stated that Enneking stage III tumors were adequately controlled with total spondylectomy in their recent and largest literature series (5), we used posterior lesionectomy with T6 laminectomy, complete tumor and relevant soft tissue removal, and bone drilling in our case. There were no stability considerations in the present case.

Usually, some microscopic tumor tissue is expected to stay behind and our patient underwent post-operative adjunctive radiation therapy as giant cell tumors exhibit a propensity for aggressive local recurrence. The use of radiation therapy remains controversial because of the widely held sarcomatous transformation but this is no longer true with modern radiotherapy techniques, especially by keeping the total radiation dose under 50 Gy.

Recurrence is common in spinal giant cell tumors. In fact, Sanjay et al. (10) reported a recurrence rate of 41.7% in their series of 24 patients with a mean follow-up of 12.4 years. Boriani et al. reported a 22% recurrence rate in a series of 49 patients with mobile spine giant cell tumor with a mean follow-up of 145 months where 64% of their study recurrences occurred within 10 month and the latest one was after 5 years (5).

A 45% local recurrence rate for Enneking stage III tumors treated intralesionally and 35% recurrence rate in a sub group of patients treated with radiation therapy in an adjuvant manner after intralesional resection were also reported (5). In the present case, there was no recurrence after 05 years of close follow-up with intralesional resection followed by radiation therapy.

Although considered benign tumors, spinal giant cell tumors show a metastatic potential supposed to be higher than their long bone counterparts, (7) which outlines the importance of postoperative long-term radiological imaging. In our case there was no metastasis at the last clinical and radiological follow-up investigation.

Our great hope is that future research will give us more answers and more treatment options. In fact, the recent literature describes a positive effect of systemic bisphosphonate administration but their role in giant cell tumors is not fully understood (1). Due to the dismal prognosis of giant cell tumors of the spine and their relatively rare side effects, we nowadays recommend additional bisphosphonates in complicated cases and metastasis. A promising RANKL inhibitor (Denosumab) is increasingly used. In fact, a study guided by Branstetter et al. found that its use induces tumor reduction and bone formation in patients with giant-cell tumors of the bone (6). Such findings will certainly motivate further investigations for the role of Denosumab therapy in the treatment of spinal giant cell tumors, especially in complicated cases.

**CONCLUSION**

Although spinal giant cell tumors are challenging clinical entities with high local recurrence and metastatic potential, we consider that wide excision along with radiation therapy offers a good prognosis.

The future is promising and close follow-up is mandatory to spot recurrences early.
REFERENCES


