

## Primary Extradural Tumors of the Spine – Case Review with Evidence-guided Management

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Received: 26 April 14 Accepted: 21 June 14 Published: 28 August 14

### This article may be cited as:

Lam FC, Arle JE, Glazer PA, Kasper EM. Primary Extradural Tumors of the Spine - Case Review with Evidence-guided Management. Surg Neurol Int 2014;5:S373-5.

Available FREE in open access from: <http://www.surgicalneurologyint.com/text.asp?2014/5/8/373/139673>

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### Abstract

**Background:** Primary extradural tumors of the spine comprise only a small percentage of all spinal tumors. However, given their relative radioresistance and their typical malignant, invasive nature, surgery may be associated with fairly high morbidity and mortality rates. Furthermore, it may be especially difficult to achieve gross total resections with tumor-free margins.

**Case Descriptions:** We present two cases and review the literature regarding the presentation, diagnosis, and evidence-based guidance/treatment of primary extradural spinal tumors. The first patient with an L1 chordoma presented with cauda equina syndrome. Following surgery, the second patient, with a giant cell tumor of the cervicothoracic junction, responded well to the novel drug *denosumab*, a monoclonal antibody to the RANK ligand.

**Conclusion:** Primary extradural spine tumors pose significant challenges to surgeons as the aim is to achieve satisfactory surgical outcomes with clean tumor margins (e.g, thus avoid recurrence) while minimizing morbidity. Improvements in radiotherapy, chemotherapy, and novel molecular drugs may increase survival rates and improve overall outcomes.

**Key Words:** Chemotherapy, extradural, malignant, molecular drug treatments, primary, radiotherapy, spine, tumors

### Access this article online

**Website:**

[www.surgicalneurologyint.com](http://www.surgicalneurologyint.com)

**DOI:**

10.4103/2152-7806.139673

**Quick Response Code:**



## INTRODUCTION

Primary extradural tumors of the spine are rare and constitute approximately 4% of all spine tumors.<sup>[5]</sup> Despite their rarity, these tumors [Table 1] can pose significant surgical challenges as their aggressive nature makes achieving a clean tumor margin difficult; this may be particularly complicated by the critical surrounding anatomy (e.g, nerve roots/spinal cord). Here we reviewed the literature and focus on two cases of primary extradural tumors: An L1 chordoma and a giant cell tumor (GCT) at the cervicothoracic junction.

### Case 1 - L1 Chordoma

A 53-year-old male who presented with acute urinary incontinence and right leg numbness, exhibited saddle anesthesia, decreased rectal tone, and numbness in the right L2 distribution on physical examination. Sagittal T2-weighted magnetic resonance imaging (T2W MRI) of the lumbar spine showed a hypointense, nonenhancing intraosseous lesion at L1 [Figure 1a], while the axial T1WI demonstrated an exophytic component compressing the cauda equina [Figure 1b]. The patient underwent an emergency L1 vertebrectomy with insertion of an expandable carbon fiber cage,

**Table 1: Summary of primary extradural spine tumors**

Variables	Benign	Malignant
Tumor	Aneurysmal bone cyst (*)	Chordoma
Location	Mainly posterior elements of the thoracolumbar spine, with extension into the vertebral body in 40% of cases	Over 50% of cases occur in the sacrum followed by the clivus (35%). One-sixth arise in the cervical, thoracic, and lumbar spine
Frequency	15% of all primary spine tumors	1-4% of primary bony tumors
Tumor	Giant cell tumor (*)	Chondrosarcoma
Location	Sacrum most common, then thoracic, cervical, and lumbar	Cervical (20%); thoracic (20%); lumbar (20%); sacral (20%)
Frequency	2-4% of all primary spine tumors	10% of all bone tumors but rarely in the spine
Tumor	Hemangioma	Ewing's sarcoma
Location	Most common in the thoracic and lumbar spine	Most commonly occurs at the sacrum
Frequency	Most common tumor of the spine, 12% incidence	Very rare to have primary spinal disease
Tumor	Osteoblastoma (*)	Lymphoma
Location	Most common in the cervical spine, then in the sacrum	Mostly involved the thoracic and lumbar spines
Frequency	1-3% of all primary bone tumors	Exceedingly rare, 1.7% of all primary bone lymphomas
Tumor	Osteochondroma	Osteosarcoma
Location	About 60% located in the lumbar spine, cervical (27%), thoracic (12%), sacrum (2%)	Mostly in the sacrum
Frequency	Most common benign bone tumor (30-40%), 1-4% occur in the spine	<5% of cases arise in the spine; comprise 3.6-14.5% of primary spinal tumors
Tumor	Osteoid osteoma	Plasmacytoma/multiple myeloma
Location	50% of cases occur in the cervical spine, followed by thoracic region	Lower thoracic and lumbar spine
Frequency	3% of all primary bone tumors, 10% occur in the spine	Most common primary tumor of the spine

(\*): Aggressive benign tumors

followed by T11 to L3 pedicle screw instrumentation [Figure 1c and d]; postoperatively, this was followed by adjuvant radiotherapy. The patient has done well at one-year of follow-up at the time of manuscript preparation with no signs of disease recurrence.

**Case 2: Giant cell tumor of the cervicothoracic junction**

A 38-year-old female presented with several months of neck pain. On physical examination, she only demonstrated midline cervicothoracic tenderness; otherwise she was neurologically intact. Sagittal [Figure 2a] and axial [Figure 2b] T2W MRI of the spine demonstrated a large hyperintense lesion involving the T1 vertebral body with a hypointense rim associated with a large ventral soft tissue component. The patient underwent a staged circumferential surgical procedure first involving an anterior T1 corpectomy with strut graft placement and anterior plating, followed by a posterior instrumented fusion utilizing lateral mass screws from C5 to C7 and pedicle screws from T2 to T4 [Figure 2c]. Intraoperatively, hematoxylin and eosin staining showed a uniform distribution of osteoclast-like giant cells in a background of mononuclear cells consistent with the diagnosis of a GCT. Postoperative imaging confirmed a generous decompression of her spinal canal [Figure 2d]. Nevertheless, her residual disease involving the lateral masses warranted her enrollment in a clinical trial with *denosumab*, a monoclonal antibody to the RANK ligand. She tolerated the treatment well, and has not shown any signs of recurrence at 40 months follow-up at the time this manuscript was prepared.

**DISCUSSION**

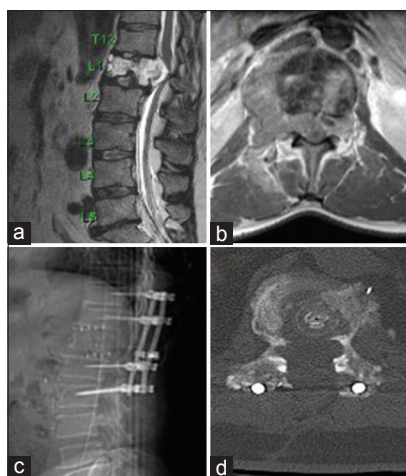
**Chordomas**

Chordomas are slow growing, locally aggressive lesions that can occur anywhere along the axial skeleton, but are most commonly found at the craniocervical junction and the sacrum.<sup>[1]</sup> Sacral chordomas are often associated with large, exophytic soft tissue masses that can contribute to a neurogenic bladder and obstipation. En bloc resection of chordomas decreases local recurrence rate to 22% while intralesional surgery leads to 78% recurrence.<sup>[2]</sup>

Conventional radiation therapy has little effect on chordomas, but may still be offered as palliation for subjective pain control and to delay the time to recurrence.<sup>[9]</sup> Recent advances in radiation therapy have allowed for higher conformal doses of radiation to be applied to a focal area with sparing of critical adjacent structures: (e.g. combined photon/proton beam therapy - approximately 70-80 Gy may increase response to treatment; inverse modulation protocols using LINAC; hypofractionation protocols; and radiosurgery).<sup>[7]</sup> The ability to achieve a wide en bloc resection offers the best prognosis and constitutes the only independent factor best avoiding tumor recurrence and tumor-related deaths.<sup>[2]</sup> Furthermore, patients with primary disease treated with surgery and radiotherapy have better local disease control than those with recurrent disease, regardless of surgical margins achieved.<sup>[8]</sup>

**Giant cell tumors**

GCTs comprise 5% of all adult primary bone tumors and occur most frequently at the epiphyseal–metaphyseal



**Figure 1:** a) 53-year-old male presenting with an L1 chordoma. b) Sagittal T2WI shows a hyperintense lesion of L1 with a large exophytic component compressing the conus on axial T1WI. c) Postoperative sagittal X-ray showing L1 vertebrectomy with T11 to L3 pedicle screw and rod fixation. d) Axial CT through L1 shows insertion of a carbon fiber cage packed with autologous bone graft

junction of long bones. They most commonly involve the sacrum, followed by the cervical and thoracic regions. Although they are aggressive benign tumors, their size and vascularity make resection difficult; therefore, patients may require preoperative embolization to decrease intraoperative blood loss.

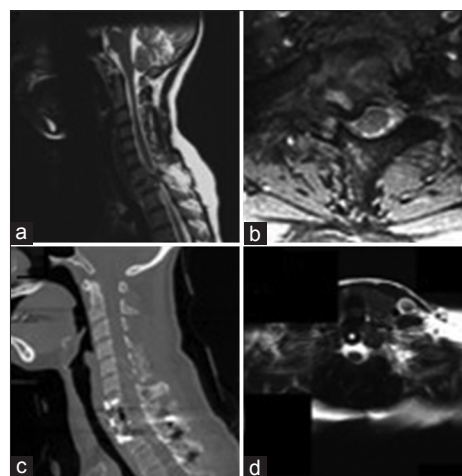
Surgical management of spine GCTs involving the thoracic and lumbar spine should include en bloc resection to decrease the rate of local recurrence. Intralesional resection can provide adequate control for Enneking stage II tumors. Cryotherapy, phenolization, and bone cement may be used as adjuvant treatments to wall off the resection cavity. Patients should be monitored for at least 5 years for disease recurrence.<sup>[3]</sup>

There is currently very low quality evidence supporting the role of radiation for treatment of recurrent GCTs.<sup>[4]</sup> Although radiotherapy may be used as an adjunct to surgery to treat local recurrences, there have been no documented benefits of adjuvant radiation following surgical debulking of sacral GCTs; furthermore, this may increase the risk for radiation-induced sarcomas.<sup>[6]</sup>

A recent clinical trial using *denosumab*, a monoclonal antibody to the RANK ligand to modulate osteoclast activation, showed a response rate of 86% and functional improvement with decreased pain in 84% of patients.<sup>[10]</sup> Our patient was enrolled in this trial following her surgery for treatment of residual disease; she tolerated the therapy well with good response at 40 months follow-up.

### Summary

Primary extradural spine tumors are rare, and there is little high quality evidence outlining the optimal treatment for these lesions. Trends in the literature support en bloc



**Figure 2:** 38-year-old female with a T1 giant cell tumor. a) Sagittal and b) axial T2WI of the cervical spine shows a hyperintense lesion at T1 eroding through the posterior cortex causing spinal cord compression. c) Postoperative sagittal CT scan of the spine showing T1 interbody fusion and plating with posterior C5 to T3 instrumentation and fusion. d) axial T2WI through T1 showing an anterior strut graft with a generous decompression of the spinal canal

spondylectomy for low grade malignant tumors, but this aggressive approach proves technically challenging and may be associated with relatively high morbidity and mortality rates. Improvements in radiotherapy technology, advances in chemotherapy, novel molecular drug targets, and other multimodality protocols may increase survival rates.

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