

# A rare case of extradural thoracic spinal chordoma with medullary compression and literature review

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## ABSTRACT

**Background.** Chordomas are low-grade, slow-growing, but locally invasive and aggressive malignant tumors, arising from the remnants of the embryonic notochord. Given its origin, the distribution of chordomas is along the axial skeleton: 50% of cases in the sacral region, 35% skull base and 15% occur in the vertebral bodies of the spine, which most commonly involve cervical (particularly C2), followed by lumbar, and then the thoracic spine. Without a prompt and aggressive treatment the chances of metastasis are low, but the local recurrence rate is high (about 50%) with a 40% chance of 10-year survival.

**Objective.** Our purpose is to present a rare case of an extradural thoracic chordoma, the importance of differential diagnosis and the management of this case for a better outcome.

**Case presentation.** A 61-year-old man presented with a 1-month history of thoracic back pain, followed by rapidly progressive paraparesis and paresthesia in both lower limbs, and 24 hour urinary retention. MRI of the thoracic spine revealed a contrast-enhancing lesion arising from T5 to T6 vertebral level, with pre and paravertebral extension and medullary compression. The diagnosis of chordoma was made by tissue biopsy and histopathological study. There are few reports of chordomas with this localization in English literature. After surgery, the patient showed slightly good clinical outcome in terms of improvement of muscle strength in lower limbs and back pain decrease in intensity of back pain.

**Conclusions.** Primary chordoma in the thoracic spine is extremely rare (less than 15% of the cases). This case highlights the importance of emergency surgery in the treatment of spinal tumors and the need to consider chordoma as a differential diagnosis for extradural spinal tumors. On the other hand the treatment is challenging because the current literature is lacking in evidence based guidelines due to low incidence.

**Keywords:** chordoma, extradural spinal tumors, intramedullary compression, rapidly progressive paraparesis

## INTRODUCTION

Chordomas are low-grade, slow-growing, but locally invasive and aggressive malignant tumors, arising from the remnants of the embryonic notochord (1). Given its origin, the distribution of chordomas is along the axial skeleton as follows: 50% of cases in the sacral region, 35% skull base and 15% occur in the vertebral bodies of the spine, which most commonly involve cervical (particu-

larly C2), followed by lumbar, and then the thoracic spine (2,3). They often extend across the intervertebral disc space, involving more than one vertebral segment and also into the epidural space compressing the spinal cord, or along the nerve roots enlarging the neural foramen.

Without a prompt and aggressive treatment, the chances of metastasis are low, but the local recurrence rate is high (about 50%) with a 40% chance of 10-year survival.

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Our purpose is to present a rare case of an extradural thoracic chordoma, the importance of differential diagnosis and quick surgical decision as main treatment option of this case, for a better outcome.

## CASE PRESENTATION

A 61-years-old male patient presented with insidious onset, non-traumatic back pain for 1-month followed by rapid progressive distal paresthesia and paraparesis for a week and 24h urinary retention. He has a history of hypertension and type II diabetes mellitus, but there was no history of trauma, anorexia, fever or contact TB patients, although his back pain was first considered to be due to a recent lung infection that was treated with antibiotics

On clinical examination, he had decreased motor strength with distal asymmetric paraparesis 3/5

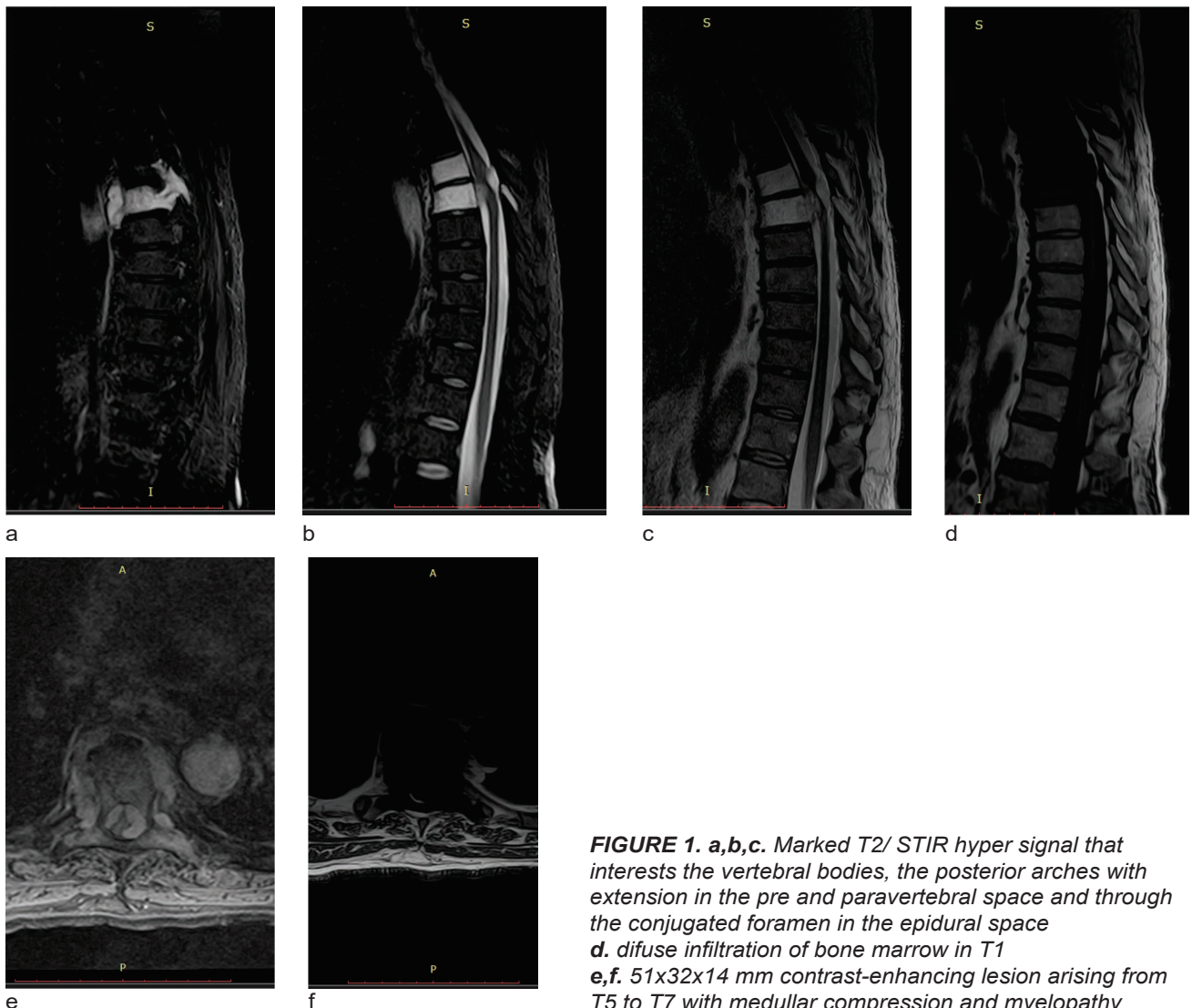
(Medical Research Council grade) on the left lower limb and 4/5 on the right lower limb, which rapidly progressed to paraplegia within 24 hours, hypoesthesia with a T5-T6 level of sensibility, with diminished deep tendon reflexes in both lower limbs, no Babinski sign and no signs of muscles atrophy, ileus paralyticus and acute urine retention. Sensory examination was abnormal with inguinal and lower limbs hypoesthesia, but without mioarthrokinetic loss of sensitivity.

## Investigations

Routine blood tests showed high levels of serum glucose due to his diabetes, leukocytosis, inflammatory syndrome.

The chest X ray revealed bilateral infrahilar alveolitis.

The abdominal ultrasonography showed abdominal bloating, and half-empty bladder with



**FIGURE 1.** a,b,c. Marked T2/ STIR hyper signal that interests the vertebral bodies, the posterior arches with extension in the pre and paravertebral space and through the conjugated foramen in the epidural space  
d. diffuse infiltration of bone marrow in T1  
e,f. 51x32x14 mm contrast-enhancing lesion arising from T5 to T7 with medullar compression and myelopathy



size and tumor necrosis are associated with poor prognosis (9).

Radiotherapy with target dose of greater than 60 Gy is the option in cases where resection is not suitable. Proton beam therapy is preferred over photon therapy, due to its straighter trajectory and so minimizing damage to surrounding critical structures (10).

Chemotherapy did not show to be effective and it is not recommended, hence the focus has shifted towards investigating molecular targeted therapy as a treatment modality (11). There have been done studies with promising results on Imatinib, a platelet-derived growth factor receptor (PDGFR) TKI,

Lapatinib in EGFR-positive chordomas and Sorafenib-kinase inhibitor.

## CONCLUSIONS

Chordomas must be kept in mind as differential diagnoses for extradural spinal tumors. Due to the rarity of chordomas, limited trials have been carried out and as a result, there is lack of standardized systemic and effective therapy. At present, unblock resection with possible radiotherapy remains the only effective treatment option, but still the recurrence rates are quite high and the long term prognosis is poor.

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