

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

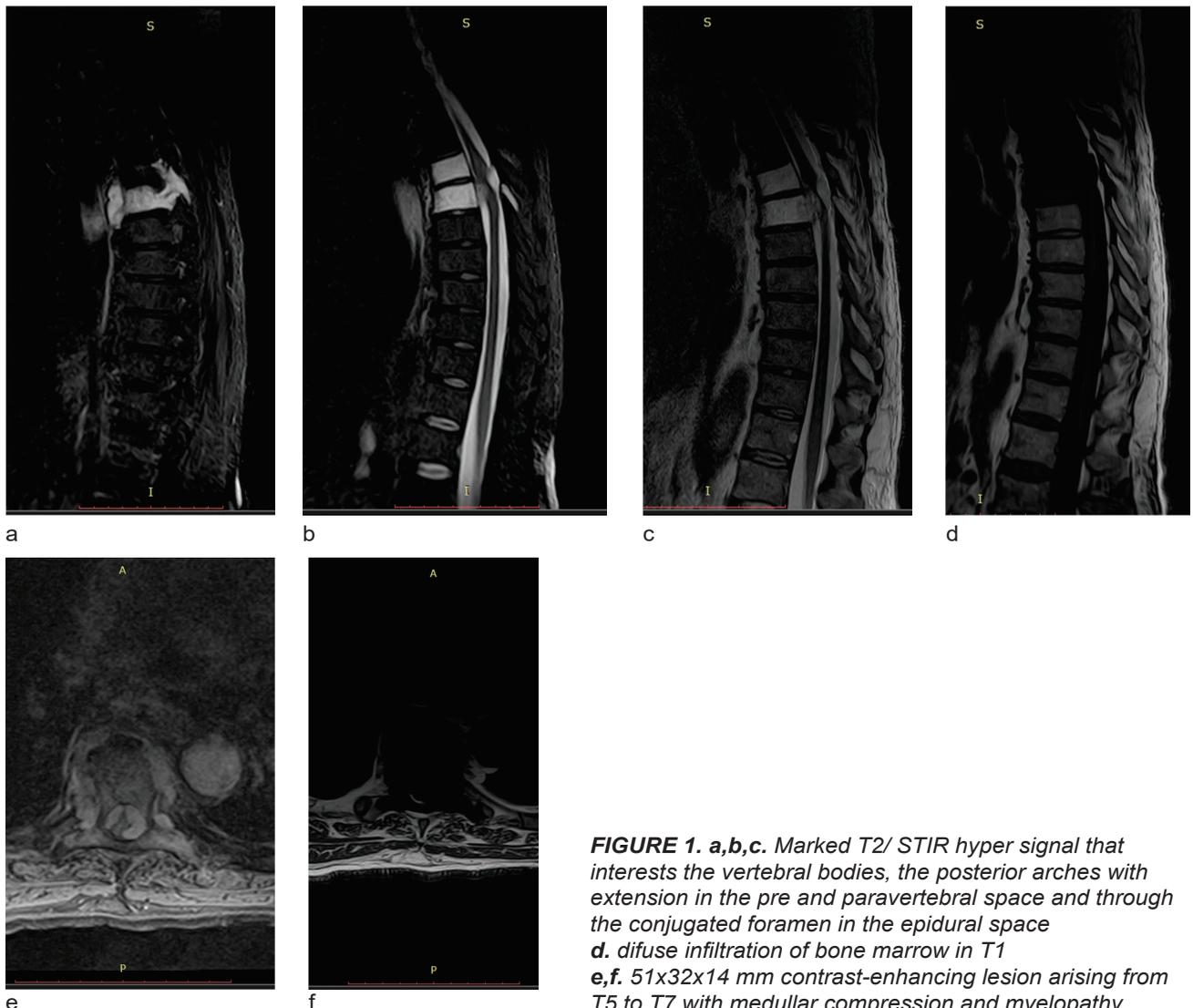


FIGURE 1. a,b,c. Marked T2/ STIR hyper signal that interests the vertebral bodies, the posterior arches with extension in the pre and paravertebral space and through the conjugated foramen in the epidural space
d. diffuse infiltration of bone marrow in T1
e,f. 51x32x14 mm contrast-enhancing lesion arising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

normal kidneys and prostate. We could not perform a urodynamic study. The head and thoracic spine CT were normal.

Due to the clinical examination that was suggestive for a lower motor neuron injury, a thoracic spine MRI was performed that revealed a 51x32x14 mm contrast-enhancing lesion arising from T5 to T6 vertebral level, with paravertebral extension and medullary compression (fig. 1).

After emergency surgery, the histopathological examination of a postoperative specimen indicated chordoma.

A contrast whole body CT was done to look for metastases, that came back negative.

Differential diagnosis

As differential diagnosis, we kept in mind metastatic vertebral involvement of lung cancer, breast carcinoma, thyroid carcinoma, renal cell carcinoma, osteosarcoma, chondrosarcoma, haemangioblastoma and a remote possibility of tuberculoma.

Treatment

He was transferred to the neurosurgery department, where he underwent urgent decompression laminectomy for spinal cord compression, following which his neurological symptoms slightly improved within 5 days.

Outcome and follow up

In the immediate postoperative period, there was a significant reduction in pain and muscle strength improved to 2/5 (MRC) in both lower limbs. However, there was no evidence of any improvement in urinary incontinence. Patient was then referred to oncology department for further investigations, treatment and follow up.

DISCUSSION

Chordomas are primary bone tumors, which arise from notochord remnants. They can affect any part of the axial skeleton, with 35% cases occurring intracranial, 50% in the sacrum, and 15% affecting the rest of the spine (1).

They are more frequent in male (2:1) and may occur at any age, but peak incidence is in fifth or sixth decades of life (1), with an exception: the skull base presentation affects teenagers and children.

Metastasis can appear in 40% of all cases, with lungs, liver, lymph nodes and cerebrospinal (CSF) as the commonly reported sites (1). Survival is modest ranging from 50% to 68% at 5 years and 28% to 40% at 10 years, due to the local aggressive course (2).

Histologically, chordomas consist of fibrous tissue separating chords of tumor from mucoid matrix. Microscopically they are characterized by foamy cells with vacuoles. There are 3 histological subtypes: classical, chondroid and de-differentiated, the last one being the most aggressive and affecting mostly children. Immuno-histological markers are needed to establish diagnosis, for molecular target therapy and to differentiate it from other tumors like chondrosarcoma. Chordoma stains positive for nuclear brachyury along with S-100, epithelial membrane antigen, vimentin, and cytokeratin (3). They are unusual primary vertebral malignancies that must be considered among the differential diagnoses for extradural spinal tumors (4).

Chordoma is a slow growing tumor, locally invasive and destructive, that can eventually lead to compression of surrounding structures including vessels and nerves. Symptoms depend on the site involved and size of the tumor: chordomas of the skull base may present cranial nerve paresis or brain stem compression syndrome, while the other types may be suggested by the clinical signs of spinal cord compression (5).

The diagnosis workup includes radiological imaging: X-ray that can show bone erosions with irregular calcium foci along with a lytic lesion, followed by Computed tomography scan used for evaluation of the lytic lesions and locally destructive pattern with hyper dense soft tissue mass with calcification and most important MRI which is considered superior to other imaging modalities and lesions appear hypointense on T1, hyperintense on T2 and Flair images and may have heterogeneous contrast enhancement (6). Pathological examination is the gold standard for diagnosis of disease.

Regarding treatment options, complete surgical resection with negative margins is the gold standard and may be combined with radiotherapy (7). This leads to better disease control and reduced local recurrence (8). An incomplete surgical resection, skull base location, advanced age, and large

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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