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Case Report

Surgical management of a thoracic chordoma: A case report and literature review

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ABSTRACT

Background: Chordomas are rare benign, but locally aggressive tumors that are regularly encountered in the clivus and sacrum. Although they are rarely found in the thoracic spine, they can contribute not only to local bone destruction and spinal instability, lead to cord compression, and major neurological deficits.

Case Description: A 56 year-old-male originally underwent a T12 laminectomy with debulking for a thoracic chordoma. Two years later, the lesion recurred contributing to a significant paraparesis. The new MR showed a T2 hyper intense lesion with huge epidural extension that warranted a 360° surgical decompression and fusion. The histopathology from both surgeries confirmed the diagnosis of a chordoma.

Conclusion: Combined 360° decompression and fusion was warranted to resect a T12 chordoma that recurred 2 years following an original laminectomy with debulking procedure.

Keywords: Combined anterior and posterior approach, Recurrence, Spinal surgery, Thoracic chordoma

INTRODUCTION

Chordomas are slow-growing but locally invasive/aggressive lesions that account for 2–4% of all primary malignant bone tumors.[9] They most commonly involve the sacrococcygeal (50%) and spheno-occipital (35%) spine; the remaining 15% occur in the spine (i.e., lumbar [57%], cervical [29%], and thoracic [13–17%]).[3,9] Despite initial radical *en bloc* resections, followed by radiotherapy and chemotherapy, these tumors still recur up to 66% of the time.[3] Here, we present a 56-year-old male whose T12 chordoma recurred 2 years following an initial debulking, warranting a secondary definitive 360° decompression/fusion procedure.

CASE STUDY

A 56-year-old male presented 2 years ago with T12-dorsal/intercostal pain. The MR documented an enhancing T12 vertebral lesion with ventral epidural extension contributing to marked cord compression [Figure 1]. He underwent a T12 laminectomy for partial tumor debulking. The pathology was diagnostic for a chordoma.

Recurrence of T12 chordoma 2 years later requiring 360° decompression/fusion

Two years later, the patient presented with recurrent dorsal pain, and a significant para paresis (i.e., motor exam 3–4/5 right leg/4–5/5 left leg, diffuse hyperreflexia, and T12 pin level, and anal incontinence). Thoracic and lumbar MRI/CT studies documented a lytic T12 vertebral body lesion with a large ventral canal extradural mass contributing to severe cord compression [Figure 2]. First, the patient underwent a thoracic laminectomy with a T10-L2 pedicle screw fusion. Two weeks later, he had a thoracotomy to perform a T12 anterior corpectomy/cage arthrodesis [Figures 3 and 4]. Notably, the second procedure was delayed by 2 weeks due to the patient's increased paraparesis after the first surgery. Following both operations and with rehabilitation therapy, the patient was discharged 10 days after the second surgery with no residual neurological deficits. One month post discharge, he was sent for follow-up conventional radiotherapy.

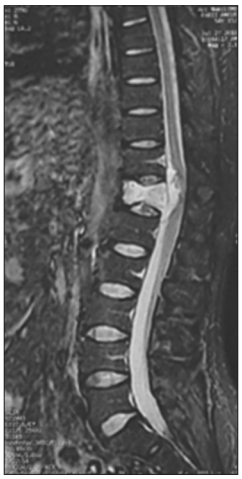

Figure 1: First preoperative MRI, T2 hyper intense D12 lesion with anterior epiduritis.

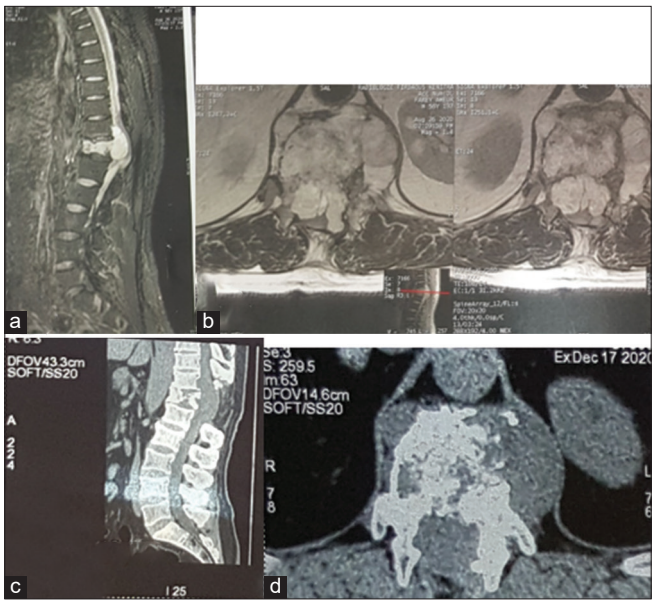

Figure 2: Pre operative images 2 years after the first surgery (a,b) sagittal and axial cut T2 MRI huge epiduritis with extra dural mass (c,d) CT scan sagittal and axial cut which show destruction of the vertebral body of D12.

DISCUSSION

Chordomas represent 2–4% of primary malignant bone tumors.[9] They have an incidence of 8.4 cases for 10 million persons.[2] Clinical manifestations, depending on their location, include; pain (i.e., about 12–24 months prior to diagnosis), motor, sensory, and/or sphincter deficits.[4] Plain X-rays/CT scans typically demonstrate lytic lesions, with CT and MR studies further document accompanying soft-tissue masses. Specifically on MRI, chordomas appear iso/slightly hypointense on T1, hyperintense on T2 studies, and markedly enhance with contrast. Treatment typically includes radical *en bloc* tumor resection with a focus on obtaining tumor-free margins.[9]

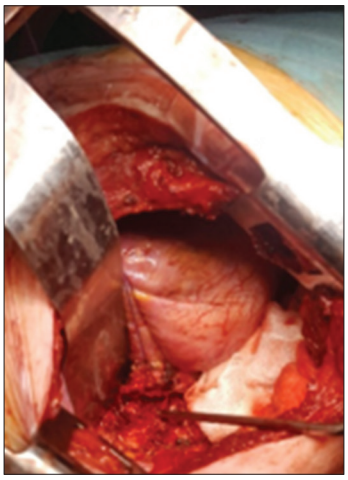

Figure 3: Intraoperative image of the anterior approach of D12.

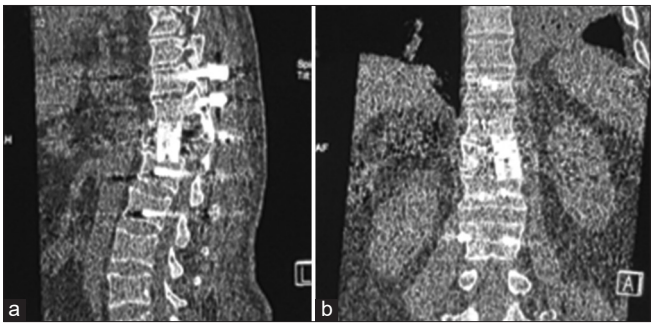

Figure 4: Post operative CT scan after circumferential approach (a,b) sagittal and coronal cut respectively.

Table 1: Case reports over last 10 years regarding thoracic chordoma.

Study	Chordoma Localization	Surgical Approach	Associated Therapy	Outcome	Reccurence Rate/ Follow-Up Time
Oppenlander *et al*. 2014[7]	T2T3	Both TH/POSTAP	No	Favorable	None - 6 months
Pu *et al*. 2016[8]	T10L2	ANTLAT	No	Favorable	None - 30 months
Liu *et al*. 2019[6]	T5T6	360°	No	Favorable	None - 6 months

RT: Radiotherapy, CT: Chemotherapy, TH: Thoracoscopic, POSTAP: Posterior Approach, ANTLAT: Anterolateral

Adjunctive radiation therapy (RT) and chemotherapy for chordomas

Chordomas are relatively radioresistant; although RT increases disease-free survival it does not prevent recurrence.[1] Unfortunately, their slow-growing nature makes these lesions resistant to most current chemotherapeutic agents.

Recurrence rate for chordomas

Chordomas recur in up to 25–89% of cases (i.e., dependent on their original location).[5] They typically recur by direct extension rather than systemic dissemination. Here, the patient had tumor recur in just 2 years following a debulking/laminectomy at the T12 level. Now, 6 months following a definitive 360 T12-decompression/fusion, the patient will warrant additional conventional RT (i.e., we have no access to proton therapy). Recent studies, dating back 10 years, showed that most chose to perform initial en bloc resections of these lesions (i.e. in 3 of 4 cases) resulting in no recurrences [Table 1].[6-8]

CONCLUSION

Despite gross total, *en bloc* resection, spinal chordomas still recur in up to 66% of cases. Here, 2 years following an original decompression/partial debulking of a T12 lesion, the patient underwent a definitive 360° *en bloc* resection (decompression/fusion) followed by appropriate adjunctive treatment.

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Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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