A C7 Chordoma Masquerading as Spinal Tuberculosis

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ABSTRACT
Tuberculosis of cervical spine is a fairly common pathology encountered by the spine surgeon in India. It is not uncommon to find that empirical treatment with anti-tubercular drugs is started based on clinical, hematological and radiological features. We present one such patient who presented to us with dysphagia and was on anti-tubercular treatment for 18 months. It was later diagnosed as a C7 chordoma after an intralingual excision. This emphasizes the fact that a tissue diagnosis is imperative for starting any antimicrobial therapy. The purpose of this report is to highlight the fact that a chordoma should be considered as an important differential diagnosis in the axial skeleton lesions anywhere from the craniovertebral junction to the sacrum; as an early diagnosis has significant impact on the tumor recurrence and long term survival. This report also emphasizes the importance of tissue diagnosis by fine needle aspiration cytology (FNAC) or biopsy before starting the definitive treatment.

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INTRODUCTION
Chordoma is a primary low grade malignant tumour arising from the notochordal remnants in the spine. The usual region of involvement is the craniovertebral junction or the sacrum, with about one third occurring in the mobile spine. Incidence in the cervical spine is about 8%, but it is extremely rare for a chordoma to occur in the lower cervical spine.1,2 The slow growing nature of the tumour and varied presentation often leads to delayed diagnosis or misdiagnosis. Tuberculosis of spine is a commonly seen pathology in India and there is often a tendency to start anti-tubercular treatment based on the clinical, radiological and haematological parameters without a confirmatory biopsy from the lesion. The purpose of this report is to highlight the fact that a chordoma should be considered as an important differential diagnosis in the axial skeleton lesions anywhere from the CV junction to the sacrum; as an early diagnosis and complete surgical excision has a significant impact on the tumour recurrence and long term survival. This report also emphasizes the importance of tissue diagnosis by fine needle aspiration cytology (FNAC) or biopsy before starting the definitive treatment.

CASE REPORT
A 57-year-old man presented to us with complaints of dysphagia of 6 weeks duration. He had a past history of neck pain and left arm pain, 18 months ago, when he was diagnosed to have a C7 destructive lesion and was started on anti-tubercular therapy based on magnetic resonance imaging (MRI) report and an elevated erythrocyte sedimentation rate (ESR), at another hospital (Fig. 1). He was also diagnosed to have idiopathic thrombocytopenia at the same time due to which a surgery or biopsy was deferred and empirical medical treatment was started with a working diagnosis of tuberculosis of C7. As his symptoms began to improve and ESR came down with the anti-tubercular therapy (ATT), the treatment was continued for 15 months. Clinical examination revealed cervical spine deep tenderness and painful flexion and extension of the neck. There was no neurological impairment and deep tendon and plantar reflexes were normal.

An X-ray of the cervical spine showed a destructive lesion of C7 with collapse of the body and a widened prevertebral soft tissue shadow (Fig. 2). MRI of the cervical spine showed a soft tissue lesion arising from C7 body and extending in to the prevertebral soft tissue indenting the esophagus; mostly hyperintense with interspersed areas of hypointensity on T2-weighted image, and was reported as suggestive of...
tuberculosis by the radiologist (Fig. 3). The size of the soft tissue lesion had increased as compared to the initial MRI scan. A frozen section analysis was done during the surgery, which revealed the possible diagnosis of chordoma. Complete intralesional surgical excision of the lesion was done through an antero-medial approach and C6 to T1 reconstruction was done with a titanium mesh cage and locking plate fixation (Fig. 4). The patient was relieved of the pain and dysphagia. He received radiotherapy postoperatively.

Histopathological evaluation revealed a neoplastic lesion composed of cells arranged in sheets and cords, separated by large amount of mucoid intercellular tissue within an abundant myxoid stroma. The neoplastic cells were polygonal in shape and had moderate to abundant pale eosinophilic vacuolated cytoplasm and round to ovoid nuclei with one or more prominent nucleoli (Fig. 5). Immunohistochemistry evaluation showed that the lesion was positive for S 100, vimentin, Pan CK and was negative for CD31, confirming the diagnosis of chordoma.

On follow-up at one and half years, the patient had a symptomatic recurrence of the tumor, extending into the adjacent vertebrae, epidural space, right paraspinol region, encircling the vertebral artery and the brachial plexus and a complete surgical excision of the lesion was not possible (Fig. 6).

**DISCUSSION**

Chordoma is a slow growing, locally aggressive primary malignant tumour arising from the notochordal remnants, mostly in the axial skeleton. Cervical spine chordomas form about 5 to 7% of all chordomas and 20 to 50% of spinal chordomas. The slow growing nature of the tumour allows it to reach a large size before detection, especially in the sacral region. The cervical spine chordomas are diagnosed relatively early, compared to the sacral chordomas, as they cause symptoms much earlier due to involvement of the vital structures in the neck. However, development of a chordoma in the cervical spine is often overlooked as a diagnostic possibility in patients with neck pain or mass. Cervical spine chordomas have been reported to masquerade as neurofibromas or schwannomas, or even a thyroid carcinoma. They have been reported to affect contiguous multiple levels, often looking like an infective pathology. Empirical treatment with anti-tubercular therapy in such cases can lead to a delayed or inadequate surgical treatment, significantly affecting the recurrence rate and survival rate. This report emphasizes the fact that a histopathological diagnosis must be obtained before initiating the definitive treatment in such lesions.

Surgery remains the mainstay of treatment of chordomas. Although newer radiotherapy techniques and medical agents like Imatinib mesylate have been shown to be effective, they are used mainly as adjuvant or palliative treatments and for recurrences. Chordomas are known to seed the biopsy tracks and recur in the soft tissues. So, it is imperative that the biopsy track is excised at the time of definitive surgery.

Tuberculosis of the spine is a very common pathology in India and it is not uncommon to find a patient started on
antitubercular treatment based on the clinical and hematological parameters without a histopathological proof of diagnosis. This decision in this reported patient resulted in a delay of 15 months before definitive surgery could be done. An intralesional excision done subsequently has led to a recurrence within 1 year despite postoperative radiotherapy.

CONCLUSION

Chordoma of lower cervical spine is extremely rare and can mimic other lesions including tuberculosis on the MRI. A tissue diagnosis and early enbloc excision gives the best possible result. Empirical treatment of spinal lesions as tuberculosis without tissue diagnosis can lead to delay in diagnosis and appropriate treatment. The lessons learnt from this case are as follows:

• Tissue diagnosis is a must be established for any lesion in the spine.
• Empirical treatment of spinal lesions as tuberculosis must be avoided.
• An enbloc excision must always be considered when possible in chordomas to achieve a long term recurrence free survival.

REFERENCES