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Case Report

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Upper Thoracic Chordoma: Case Report and Review of the Literature

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Abstract

Chordoma originates commonly in the sphenooccipital and sacrococcygeal regions has a general incidence of 0.08 per 100,000 people and constitutes 1–4% of all bone cancers. Thoracic chordoma is a rare but aggressive and sometimes life-threatening tumor due to local recurrence and distant metastases to the lungs. We report a case of a 65-year-old man admitted for nontraumatic spinal cord compression syndrome secondary to upper thoracic chondroma successfully removed "enbloc" with a positive outcome after eight months of physiotherapy.

Keywords: Thoracic Chordoma, Spinal Cord Compression, Management.

1. Introduction

A chordoma is a low-grade, slow-growing but locally invasive and locally aggressive tumor that is a type of sarcoma. Chordomas arise from remnants of the notochord and occur along the midline spinal axis between the clivus and the sacrum, anterior to the spinal cord [1,2]. Chordomas can present anywhere on the spine, the most common locations are at the sacral spine with 50%, the skull base with 35%, and the mobile spine with 15%. Chordomas located on the spine make up approximately 20% of reported chordomas, with the thoracic spine contributing less than 1% [1,3–5].

Despite the putative embryological derivation, middle-aged adults are the most commonly affected age group, with most series listing the average age at presentation in the sixth decade [6]. Up to a third of the reported patients were entirely asymptomatic and diagnosed incidentally on routine chest radiographs. Nevertheless, the most common complaints at presentation were chest or back pain, followed by anterior mediastinal compression syndromes (hoarseness, dysphagia, cough) with non-traumatic spinal cord compression syndrome when extended into the vertebral canal [7].

Innovative treatment approaches have been developed in the past 20 years, but the evidence generated by available studies is weak. Therefore, the degree of uncertainty in selecting the most appropriate treatment remains high, which results in suboptimum outcomes for many patients [3].

This case highlighted an underreported location, presentation, and management of the chordoma.

2. Case Presentation

2.1 Patient Information

A 65-year-old man presented with 4 months history of progressively worsening thoracic back pain. His past medical history was a medication for Arterial Hypertension. He reported progressive Dorsalgia becoming resistant to usual painkillers and followed by quick fatigue in the lower limbs, and intermittent claudication complicated by bladder and bowel disturbances at the admission. He denied any trauma or other inciting event.

2.2 Clinical Findings

Physical examination found T10 level hypoesthesia and normal muscle tone, with a Medical Research Council (MRC) scale for muscle strength of 3/5 in both lower limbs. The reflexes were diminished on the right lower limb with a Babinsky. Slight abdominal distension with a pollakiuria was found. The initial laboratory evaluation revealed a normal complete blood count with differential. The tests of inflammatory markers (C-reactive protein and procalcitonin), electrolytes, liver, renal, and thyroid function were normal. ECG was unremarkable.

3. Diagnostic Assessment

With this progressive nontraumatic spinal cord compression, complete spine magnetic resonance imaging (MRI) demonstrated a 37 mm mass of the T2 vertebral body with epidural extension. The lesion was hyperintense on T2-weighted imaging, intermediate on T1-weighted, and intensively enhanced by the Gadolinium. The lesion extends from T1 to T3, with a mass effect on the spinal cord (Figure 1).

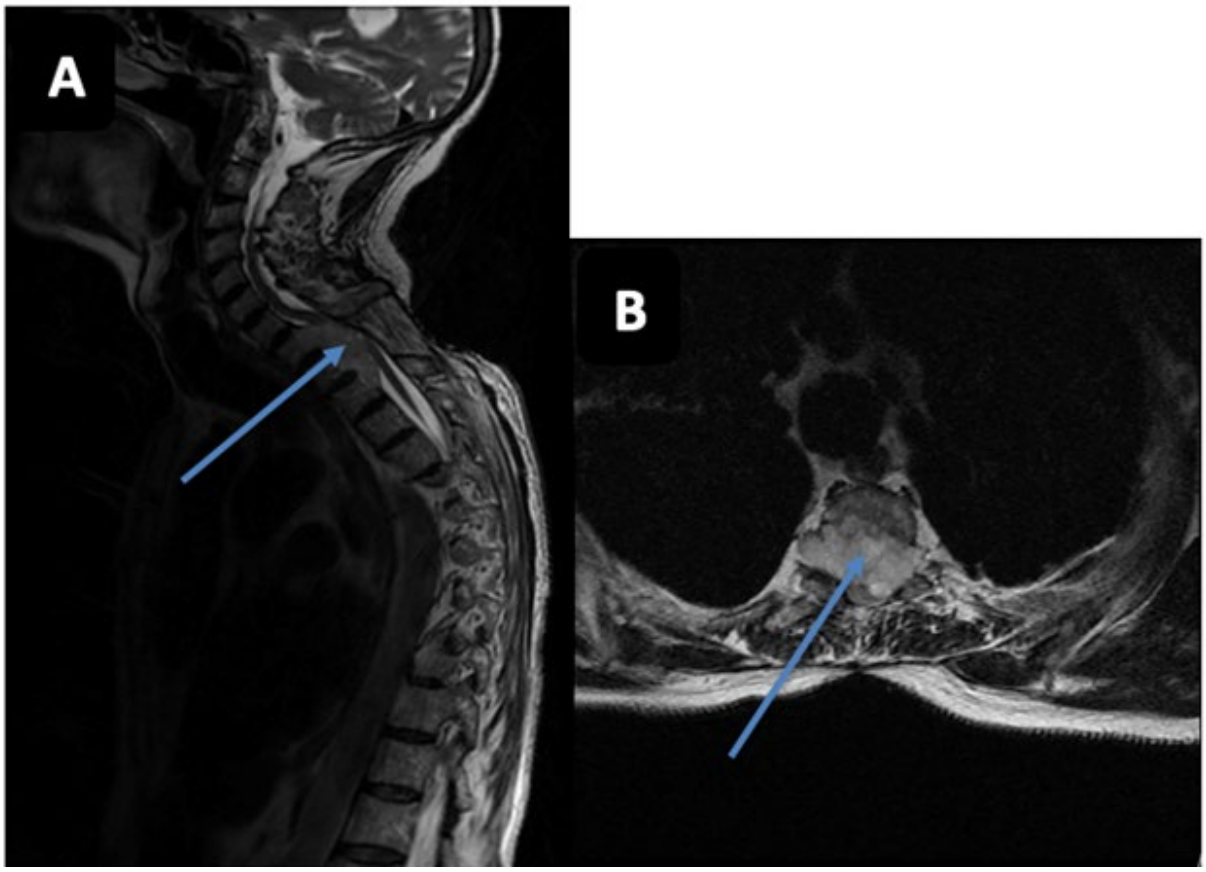

Figure 1: A: MRI Hyperintense Lesion on T2-Weighted Imaging. B: T1-Weighted Lesion Intensively Enhanced by The Gadolinium.

4. Therapeutic Intervention

The patient underwent surgery the next day. Laminectomy of T2 with en bloc resection of the lesion was done successfully. Histological examination of the lesion was consistence with a chordoma (Figure 2).

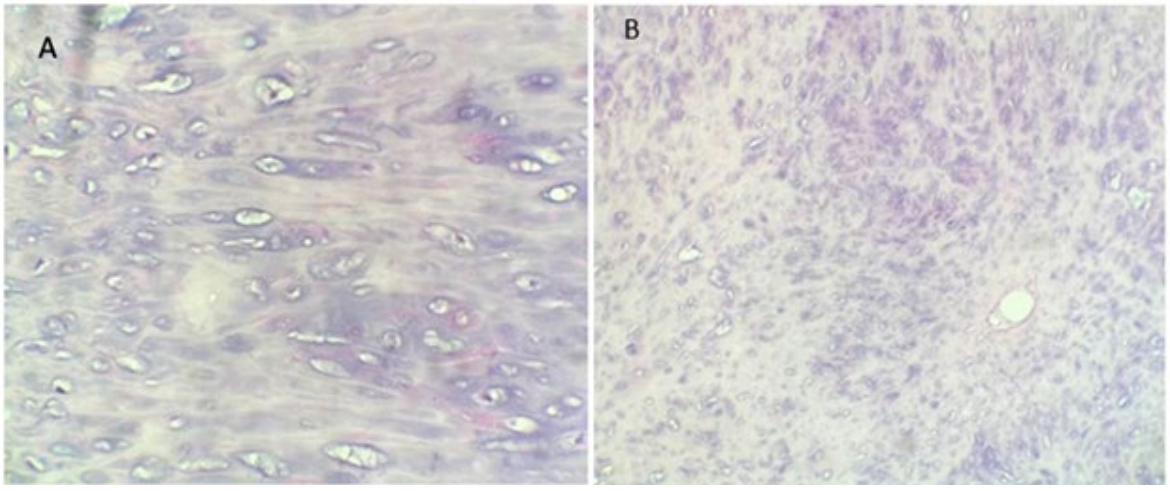

Figure 2: A: Histology Showing High-Density Cartilage Proliferation [HEx20]. B: Chondrocytes of Variable Size with Sometimes Binucleated Nuclei Surrounded by a Clear Halo (HEx40).

5. Follow-Up and Outcome

After eight weeks of follow-up with physiotherapy, the patient was doing good and able to walk with perfect control of his bladder and bowel movements. A thoracic spine MRI six months later showed a total removal of the chordoma with no evidence of local recurrence.

6. Discussion

This study provided details about the underreported location, presentation, imaging, and management of the spinal chordoma.

N°	Authors (year of publication)	Sex	Age (Years)	Spinal level involved	Symptoms	Treatment	Outcomes
01	Crowe et al (1951) [15]	M	30	Thoracic	Upper back pain, dysphagia	Resection of lesion and radiotherapy	Favorable
02	Topsakal et al. (2002) [16]	F	44	Thoracic (T4)	Upper back pain, paraparesis, sphincter dysfunction	Resection of lesion, corpectomy, bone fusion	Favorable
03	Fontes et al. [6] (2012)	M	89	Thoracic (T10)	Upper back pain, paraparesis, sphincter dysfunction	Resection of lesion and corporectomy	Favorable
04	Conzo et al. (2013) [17]	M	47	Thoracic (T1-2)	Asymptomatic	Resection of lesion	Favorable
05	Rena et al. [7] (2014)	F	69	Thoracic (T2-3)	Upper back pain, paraparesis	Resection of lesion, corporectomy and laminectomy.	Favorable
06	Oppenlander et al. (2014) [18]	M	51	Thoracic (T2-3)	Upper back pain	Resection of lesion, laminectomy, bone fusion	Favorable
07	Faheem et al. (2016) [19]	M	8	Thoracic (T11-L1)	Lower back pain, paraparesis, sphincter dysfunction	Resection of lesion and laminectomy	Favorable
08	Goomany et al. [10] (2016)	M	46	Thoracic (T11)	Upper back pain	Resection of lesion, laminectomy, bone fusion	Favorable
09	Liu et al. [9] (2019)	M	64	Thoracic (T5-6)	Upper back pain	Resection of lesions, laminectomy, bone fusion	Favorable
10	Yoon et al. (2020) [20]	F	52	Thoracic (T4-5)	Upper back pain, dysphagia	Resection of lesion and corporectomy	Favorable
11	Constanzo et al. (2021) [21]	M	62	Thoracic (T3-4)	Upper back pain, paraparesis, gait disturbance	Resection of lesion and laminectomy	Favorable
12	Kassels et al. [13] (2022)	F	36	Thoracic (T2-3)	Upper back pain	Resection of lesion, radiotherapy and proton therapy	Favorable

13	Moune et al. [14] (2022)	M	56	Thoracic (T12)	Upper back pain	Resection of lesions, laminectomy, bone fusion, radiotherapy	Favorable
	Our present case	M	65	Thoracic (T2)	Upper back pain	Resection of lesion and laminectomy	Favorable
	F indicates Female; M : Male						

Table 1: A Literature Review of Thoracic Chordoma Cases

Chordomas tend to occur at sites along the spinal axis: the sacrococcygeal and sphenooccipital junctions [6]. In the thoracic region, the occurrence of chordomas is less common. The literature review on this topic revealed only 13 reported cases with a male predominance of 69.2%. (Table 1). The mean age was 50.3±19.8. Twelve of the 13 presented with upper back pain while 3 had paraparesis, gait disturbance, and sphincter impairment. Lesion resection, laminectomy, and bonne fusion were the gold standard of surgical treatment with a favorable outcome in 92.3% of cases. Only 2 reported cases described radiotherapy as adjuvant treatment.

These tumors account for only 8 - 17% of mobile spine chordomas and an estimated 1% of all chordomas [6]. The symptoms and pattern of occurrence depend largely on the tumor location. Pain is the most common symptom of chordoma. In addition, spinal cord or nerve root compression can lead to neurological symptoms, while nerve root involvement in the sacrum leads to bowel and bladder dysfunction, including incontinence [8]. It is similar to our case.

Imaging studies, including MRI, bone scan, and PET/CT, are non-specific, making it difficult to distinguish thoracic chordoma from other common spinal lesions [9]. They may show consecutive spinal canal stenosis, spinal cord or nerve root compression, compression, or involvement of adjacent structures. However, imaging studies play a critical role in making surgical decisions. MRI of our patients showed an isointense lesion on T1WI and an isointense lesion on T2WI, consistent with previous case reports [9].

Surgery is the main treatment for the chordoma and, if possible, “en bloc” resection should be performed to achieve tumor-free margins and prevent spread [10-12]. Thoracic chordomas are difficult to completely resect. The planning of the operation follows a thorough preoperative evaluation and a careful assessment of the extent of the tumor in the spinal canal. Due to the rarity of thoracic chordomas and differences in tumor localization and presentation, surgical approaches vary. [13]. Although posterior debulking approaches have been used, they have recently been replaced by anterior or lateral approaches to ensure as complete a resection as possible and achieve modern spinal immobilization through interbody fusion [14].

In our case, we performed a post approach and a complete resection was possible. The tumor was easy to dissect. There was no adhesion with the tumor capsule on the dura mater. This condition allowed us to perform an “en bloc” resection in a single surgical procedure. Given the quality of tumor removal, adjuvant radiotherapy was not used in our case. Some authors recommend combining radiotherapy with surgery to improve local control [8]. In our case, no tumor recurrence was observed 6 months after the surgery.

7. Conclusion

Gross total resection of thoracic chordoma is doable and remains, so far, the main neurosurgical therapy necessary to save patients from its devastating complications [15-21].

Declarations

Ethics Approval and Consent to Participate: Informed consent was obtained from the patient prior to the submission of this article. Also, this article respects both the Consensus-based Clinical Case Reporting Guideline and the Recommendations for the Conducting, Reporting, Editing, and Publication of Scholarly Work in Medical Journals.

Consent for Publication: Informed consent was obtained from the patient to publish his case.

Availability of Data and Material: All data generated or analyzed during this study are included in this published article.

Competing Interests: The authors declare that they have no competing interests.

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References

1. Tenny S, Varacallo M. Chordoma [Internet]. StatPearls [Internet]. StatPearls Publishing; 2022 [cited 2023 Mar 25].
2. Tranesh, G., & Nassar, A. (2016). Metastatic Chordoma: A diagnostic challenge on fine needle aspiration. Case Reports in Pathology, 2016.
3. Stacchiotti, S., & Sommer, J. (2015). Building a global consensus approach to chordoma: a position paper from the medical and patient community. The lancet oncology, 16(2), e71-e83.
4. Zhou, Y., Hu, B., Wu, Z., Cheng, H., Dai, M., & Zhang, B. (2018). A giant lumbar chordoma: a case report. Medicine,

97(25).
5. Sobash, P. T., Vedala, K., Alfano, D., Pinckard-Dover, H., Muesse, J. L., & Desikan, R. (2021). A rare case of chordoma presenting as a Pancoast tumor. Rare Tumors, 13, 20363613211029493.
6. Fontes, R., & O'Toole, J. E. (2012). Chordoma of the thoracic spine in an 89-year-old. European Spine Journal, 21, 428-432.
7. Rena, O., Davoli, F., Allegra, G., Casadio, C., & Turello, D. (2014). Giant chordoma of the upper thoracic spine with mediastinal involvement: a surgical challenge. Asian Spine Journal, 8(3), 353.
8. Bai, R., Zhao, Z. Q., Wang, Y. X., Zhao, W., Wu, L. S., Cui, S. X., ... & Liang, C. L. (2021). Sacral and thoracic chordoma with pulmonary metastases: A case report and review of the literature. Molecular and Clinical Oncology, 14(1), 1-1.
9. Liu, S., Zhou, X., Song, A., Huo, Z., Yao, S., Wang, Y., & Liu, Y. (2019). Surgical treatment of giant chordoma in the thoracic spine combining thoracoscopic and posterior spinal surgery: a case report. Medicine, 98(35).
10. Goomany, A., Timothy, J., Robson, C., & Rao, A. (2016). En bloc resection of a thoracic chordoma is possible using minimally invasive anterior access: An 8-year follow-up. Journal of neurosciences in rural practice, 7(01), 138-140.
11. Leitner, Y., Shabat, S., Boriani, L., & Boriani, S. (2007). En bloc resection of a C4 chordoma: surgical technique. European Spine Journal, 16, 2238-2242.
12. Weil, A. G., Shehadeh, M., Ayad, T., Abboud, O., & Shedid, D. (2015). En bloc resection of a C2–C3 upper cervical chordoma. Surgical Neurology International, 6(Suppl 24), S615.
13. Kassels, A. C., Melamed, J., Rogers, A., & Johnstone, D. (2022). Thoracic chordoma in a 36-year-old female. Journal of Surgical Case Reports, 2022(12), rjac541.
14. Moune, M. Y., Sayore, C. M., Hemama, M., El Fatemi, N., & El Maaqili, M. R. (2022). Surgical management of a thoracic chordoma: A case report and literature review. Surgical Neurology International, 13.
15. Crowe, G. G., & Muldoon, P. L. (1951). Thoracic chordoma. Thorax, 6(4), 403.
16. Topsakal, C., Bulut, S., Erol, F. S., Ozercan, I., & Yildirim, H. (2002). Chordoma of the Thoracic Spine—Case Report—. Neurologia medico-chirurgica, 42(4), 175-180.
17. Conzo, G., Gambardella, C., Pasquali, D., Ciancia, G., Avenia, N., Della Pietra, C., ... & Santini, L. (2013). Multifocal thoracic chordoma mimicking a paraganglioma. Journal of Cancer Research and Therapeutics, 9(3), 497-499.
18. Oppenlander, M. E., Maulucci, C. M., Ghobrial, G. M., Evans III, N. R., Harrop, J. S., & Prasad, S. K. (2014). En bloc resection of upper thoracic chordoma via a combined simultaneous anterolateral thoracoscopic and posterior approach. Operative Neurosurgery, 10(3), 380-386.
19. Faheem, M., Zeeshan, Q., Ojha, B., & Agrawal, P. (2016). Thoracic intramedullary chordoma without bone involvement: a rare clinical entity. Case Reports, 2016, bcr2016216101.
20. Yoon, S. K., Moon, M. H., & Moon, S. W. (2021). Thoracic Chordoma Misdiagnosed as Primary Adenocarcinoma of the Mediastinum. Journal of Chest Surgery, 54(2), 158.
21. Costanzo, R., Scalia, G., Marrone, S., Umana, G. E., Graziano, F., Furnari, M., ... & Nicoletti, G. F. (2021). Thoracic dumbbell spinal chordoma mimicking a schwannoma. Surgical Neurology International, 12.