



Chordoma in the Thoracolumbar Spine: A Case Report

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Abstract

Purpose: Chordomas arise from embryonic notochordal remnants along the length of the neuraxis at developmentally active sites. They constitute less than 1% of CNS tumors and rarely occur in extra-axial locations. Chordoma in the thoracic spine is considered rare and comprises less than 15% of all chordomas.

Case study: A healthy, 42-year-old male presented with radicular low back pain, with a sudden onset four weeks prior to this examination. He complained of paresthesias, unstable walk patterns and difficulties in urination; however, no neurologic deficits were reported. On CT and MRI, an occupying soft tissue lesion was detected with pressure on the thecal sac at the level of D6-D9 vertebrae. The lesion was irregular and diagnosed initially as a dermoid tumour. After primary debulking and fixation, the pathological biopsy revealed a chordoma. The patient had a second surgery to change the primary fixators to carbon made rods prior to Proton Beam Therapy.

Discussion: According to the imaging and location of the lesion, chordoma was not the suggested diagnosis. Surprisingly, based on the histopathological results, a chordoma was diagnosed. Due to the multidisciplinary treatment approach, the patient was prepared for following radiotherapy treatment. Two years after treatment, there are no signs of a lesion or clinical signs of thecal sac involvement.

Conclusion: When suspicion of an irregular space-occupying lesion arises, always act as if a malignancy is present. We recommend a multidisciplinary approach in coordination with oncologists, pathologists, the surgical team and the radiation centre for management of such cases.

Keywords: Chordoma; Tumor; Thoraco-lumbar; Spine; Radiotherapy; Dermoid

Introduction

Chordomas are rare tumours that arise from embryonic notochordal remnants along the length of the neuraxis at developmentally active sites. They constitute less than 1% of CNS tumours and rarely occur in extra-axial locations. According to the literature, there are three major sites of occurrence in descending order of frequency: The sacrum, intracranially at the clivus, and along the spinal axis. When considering all locations, the male-to-female ratio is 2:1 [1]. The treatment of chordomas is optimally managed by aggressive surgery with care to preserve elemental structures, following postoperative radiation [1]. The overall survival of chordomas is enhanced by complete resection of the tumour [2]. Given the local invasiveness and large tumour burden, complete resection of these tumours is often difficult. Surgery should aim towards maximally safe cytoreductive surgery with wide en bloc resection along with preservation of neurological function and quality of life, even at the price of postoperative residual tumour. Due to the difficulty of obtaining a gross total resection and wide surgical margins, adjuvant post-operative radiotherapy is important or even essential for local tumour control even in these slowly growing tumours [3]. Chordoma is a radioresistant tumour which requires irradiation doses of at least 74 Gy using conventional fractionation (1.8-2 Gy per fraction) that are beyond the tolerance of several critical structures and administered with photon beams [3].

Advances in radiation technology and delivery with the introduction of hadrons (i.e., protons or charged particles, including carbon ions, helium, or neon) have led to higher doses being delivered to the target, with limited injury to the surrounding tissue and improved radiobiological effects. Unfortunately, the availability of hadron-based therapy is limited because of the associated construction and operational expenses [4-7]. Using technology with photons, such as IMRT or Stereotactic Radiation (SBRT), are also in use to enhance irradiation doses with maximum effect on the tumour and minimal effect on

critical organs [8-10]. Heavy ions, most frequently carbon ions, have been theoretically postulated to have a biological advantage in terms of Relative Biological Effectiveness (RBE) over photon and proton therapy (3-4-fold more), particularly in slow-growing, usually radio resistant, tumours [4-7]. The scarce number of heavy ion devices throughout the world and the limited number of treated patients are limiting factors for up-to-date results regarding irradiation of chordomas, although results are very promising [4-7].

Proton Beam Therapy (PBT) was shown to be superior to photons with RBE of 1.1-1.2. The energy profile with Bragg Peak allowed precision of high doses in target volumes with maximal protection of critical tissues. Particle therapy or proton treatment for patients with chordomas showed favourable local control and overall survival. Severe toxicities were successfully reduced by modifying the dose fractionation and treatment planning in the later treatment era. Thus, this therapeutic modality should be considered useful and safe [4-7,11]. In a recently published work, a retrospective analysis of the clinical outcomes of eligible patients with primary sacral chordoma who had undergone definitive PBT with 70.4 Gy (relative biological effectiveness) in 32 fractions demonstrated a 3-year estimated local progression-free survival, distant metastasis-free survival, disease-free survival, cause-specific survival, and overall survival rates of 89.6%, 88.2%, 81.9%, 95.7%, and 92.7%, respectively [10]. This study demonstrates the

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Received December 28, 2019; **Accepted** January 17, 2020; **Published** January 24, 2020

Citation: Batash R1, Simonovich A2, Lee E1, Schaffer M3, Debbi R, et al. (2020) Chordoma in the Thoracolumbar Spine: A Case Report. J Clin Case Rep 10: 1312

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Case Report

A 43-year-old healthy male patient was referred to an orthopaedic evaluation due to radicular low back pain with onset four weeks prior to his examination. The patient complained about paresthesias, unstable walk patterns, and difficulties in urination, but no neurologic deficits were reported. A history of back trauma was ruled out and no family history for malignancy was mentioned. On physical examination the patient demonstrated signs for myelopathy: Extremity paresthesias, ataxia, spastic gait, weakness and clumsiness, decreased sensation along L4 dermatome right leg, hyperreflexia, and a positive Babinsky sign. He was admitted for further evaluation and an MRI scan of the lumbar spine. On the primary MRI, no pathologic fractures, spinal deformity or stenosis was seen, nor was there any thecal sac involvement. The patient's symptoms continued to aggravate, and he was unable to bear weight in the Orthopaedic ward. After physical examination and re-evaluation, a cervical to sacral spine CT scan was performed. The dorsal spine CT displayed a lytic mass with smooth lobulated margins, involving posterior elements of the D7 and D8 vertebra, extending into the left pedicle of D8 (Figures 1a and 1b).

Consecutively, MRI of the spine was performed at 1.5 T (Ingenia, Philips, The Netherlands). MRI analysis was performed with T1 and T2-weighted Turbo Spin Echo sequences, with axial and sagittal planes, with and without fat saturation, completed with sequences after intravenous administration of 0.1 mg per kilo of paramagnetic contrast media (Gd-DTPA). Diffusion-Weighted Imaging (DWI) was performed also. The presence of an expansive lobulated mass, 5 × 3 × 1.5 cm, centred in the posterior epidural space, was confirmed (Figure 2). This mass showed heterogeneous hyper-intense signalling on the T1 weighted images, hyper-intense signal to cerebral spinal fluid in the long TR sequences. After intravenous administration of the contrast agent, no enhancement was observed within the lesion. The mass was compressing the Dural sac and partially occluding the left intervertebral foramina between D7 and D8 (Figure 3). The bulk also involved the left vertebral pedicle, the corresponding articular process of D8 and, to a lesser degree, the right articular process. The base of the spinal process of D7 was also involved. On DWI, the mass presented a high signal, suggesting restricted diffusion.

Based on the imaging characteristics on the MRI and the bone destruction pattern on the CT, the patient was prepared for a wide excision biopsy and posterior spinal fusion. The surgery was performed by two senior spinal surgeons. The pathologic biopsy revealed a chordoma in the D6-D9 vertebrae and soft tissue along the vertebral bodies (Figures 4 and 5). A follow-up MRI revealed residual tumour

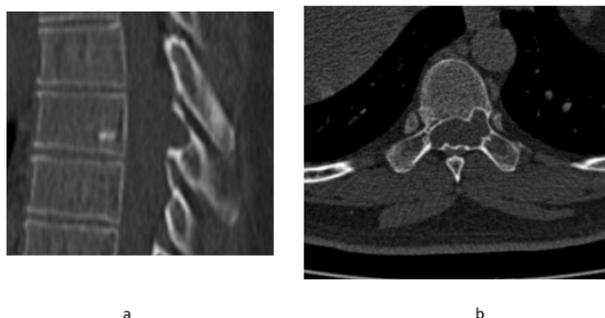


Figure 1: Sagittal (a) and axial (b) view. A lytic mass with smooth lobulated margins, involving posterior elements of the D7 and D8 vertebra, extending into the left pedicle of D8.

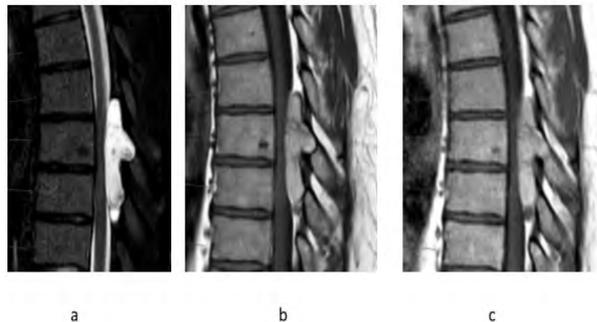


Figure 2: Sagittal STR (a), pre (b) and post contrast (c) T1 weighted images. Expansive lobulated mass, centered in posterior epidural space, showed heterogeneous hyper intense signal on the T2 (a) and T1 (b) weighted images. After intravenous administration of contrast agent, no enhancement was observed within the lesion (c).

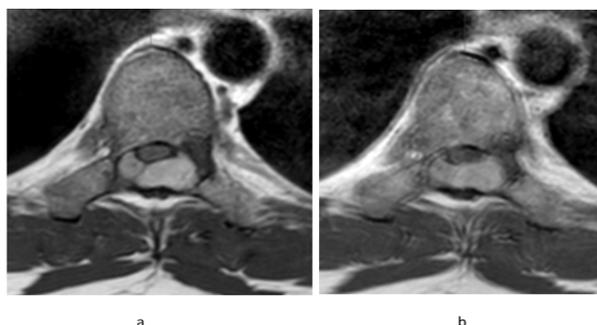


Figure 3: T1 pre (a) and post contrast (b) image. The mass was compressing the Dural Sac and partially occluding the left intervertebral foramina between D7 and D8 vertebra.

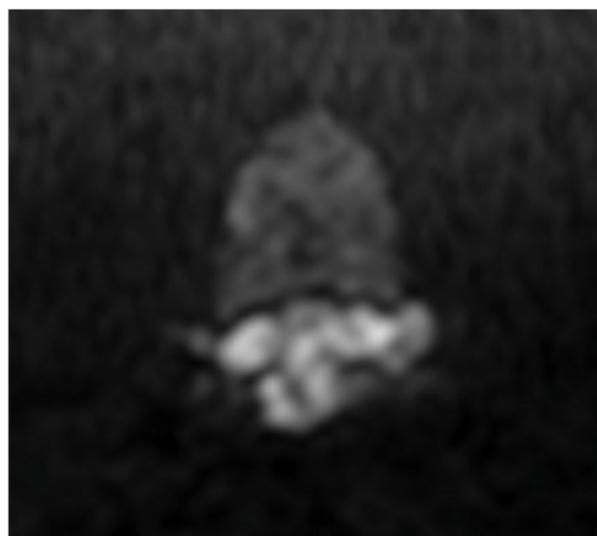


Figure 4: On DWI the mass presented high signal, suggested restricted diffusion.

at the left T8 pedicle. After consultation with an orthopaedic spinal oncologist and a tumour board, a second surgery was undertaken to remove as much residual tumour as possible and to replace the rod fixators by carbon implants to assist future irradiation therapies and

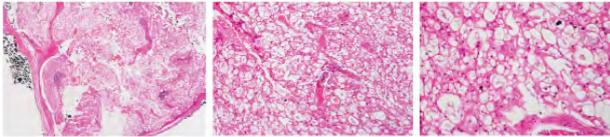


Figure 5: Pathologic biopsy revealed a chordoma in the D6-D9 vertebrae and soft tissue along the vertebral bodies.

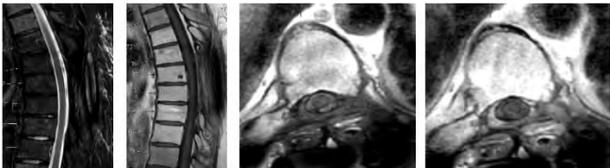


Figure 6: 2 years after treatment. MRI images sagittal and axial planes demonstrating tumour free vertebrae, pedicles and soft tissue surrounding D6-D9.

follow-up. Later, the patient was planned for 54 Gy RBE in 27 fractions and 20 Gy RBE in 10 fractions. Due to a possible excess to the spinal cord, the second series was reduced by one fraction, making the total dose received 72 Gy (RBE) in 36 fragments, utilizing the 250 MeV cyclotron at the Centre for Proton Radiation Therapy at the Paul Scherrer Institute in Switzerland without any concomitant chemotherapy. During therapy, the patient developed fatigue Grade 1 and radiation dermatitis Grade-1. His appetite was slightly reduced but no change in body weight was noticed. The patient reported a reduction in pain and an ability to walk almost with no aids. On physical examination two years after the radiation therapy, the patient is symptom free, walks with no assistance, and has no neurological deficits. On MRI, there are no signs of recurrence and the surrounding soft tissue is tumour free (Figure 6).

Discussion

Chordomas in the thoracolumbar spine comprise 15% of all chordomas. Hence, this location is rare. Due to the fact that chordoma is considered a local aggressive tumour with a high frequency of recurrence, multidisciplinary treatment is advised. In our case, we consulted oncologists and a radiation centre regarding treatment. Upon receiving the pathologic results confirming the diagnosis of chordoma, the rod fixators were changed to carbon rods to be compatible with radiation and following MRI series for follow-ups. The patient demonstrated vast recovery and immediately after the operation began walking in the department. At two years follow-up, the patient's MRI demonstrated a tumour free thoracolumbar spine with no signs of fixator rods loosening or vertebral instability. One might wonder whether the surgical team managing this case should have started with a biopsy of the lesion and continue to a major vertebral removal along with the surrounding soft tissue only after confirmation of the histopathology. According to the clinical symptoms of the patient demonstrating spinal cord compression and signs of cauda equina syndrome with fast deterioration, we decided to go for an emergency tumour debulking, decompression and posterior fusion surgery. Based on the MRI findings of an irregular space-occupying lesion, gross debulking of the involved soft tissue and involved vertebrae prior to the posterior spinal fixation seemed warranted.

Following the surgery, the patient was sent for radiation therapy. According to the latest literature, treatment with heavy ions achieved better results than photon radiation therapy, due to higher Relative Biological Effectiveness (RBE) and physical selectivity. Unfortunately,

this therapy is in phase II clinical trials and is not yet approved as a standard of care treatment modality. Our patient, therefore, underwent proton radiation therapy, as noted above. Despite all efforts, most chordomas will recur. According to the current literature, several studies are now focusing on future directions in hopes of revealing more tools in the management of the disease. Tyrosine kinases and transcriptional regulators, brachyury, were found in chordoma [12]. A Phase III study investigating the efficacy of carbon-ion therapy is undergoing [13]. Phase II trial with Everolimus and Imatinib demonstrated promising results (EUDRACT 2010-021755-34) [14].

Conclusion

In conclusion, chordomas are aggressive tumours and must be treated in a multidisciplinary approach; surgical treatment followed by radiation therapy is the standard of care. An MRI scan is the method of choice preoperatively to evaluate the tumour. Postoperative radiation treatment should be taken into consideration; hence, radiation centre involvement should be updated and should advice regarding the fixation material, e.g. carbon rods. In case of an emergency debulking operation, it is advisable to remove all suspected tissue as thoroughly as possible, even at the expense of stability which will indicate fixation later on.

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