Eur Spine J
DOI 10.1007/s00586-016-4877-6

CASE REPORT

Giant chordoma in the thoracolumbar spine: a case report and literature review

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Received: 27 May 2016 / Revised: 4 October 2016 / Accepted: 11 November 2016

Abstract
Study design Case report.
Purpose We present a rare case of a giant chordoma in the thoracolumbar spine and review the current literature. We describe its complicated clinical progression, hoping to shed light on the clinical management of this complex tumor.
Methods We present a previously healthy 41-year-old man who experienced progressive low back pain at T10-L2 for the past 2 years. A giant tumor was detected on magnetic resonance imaging, and aspiration biopsy was used to obtain a definite pathological diagnosis. The postoperative pathology confirmed that it was a chordoma. He underwent complete resection of the tumor and internal fixation of the vertebral bodies, which is a good way to control recurrence and preserve stability.
Results Histopathology confirmed the tumor was a chordoma via immunohistochemical study of both the biopsy sample and the surgically resected tissues. There has been no recurrence or metastasis at the 30-month postsurgery radiographic examination. The internal fixation has remained stable.
Conclusion Primary chordoma in the thoracolumbar spine is extremely rare. The treatment is difficult because the current literature is sparse and patients are rare. Complete resection and internal fixation are effective for reducing recurrences and metastasis.

Keywords Chordoma · Thoracolumbar · Spine

Introduction

Chordoma is a malignant bone tumor that accounts for 1–5 % of all bone tumors. Chordomas arise from notochordal remnants at sacrococcygeal and sphenooccipital junctions [1, 2]. It is unusual in the so-called "mobile" spine, with only 15 % of cases occurring there, and is especially rare in the thoracolumbar spine [3, 4]. In this study, we describe a rare case of chordoma in the thoracolumbar spine and review the literature on thoracolumbar spine chordomas.

Case report

A previously healthy 41-year-old man experienced progressive low back pain for the past 2 years. The neurological examination was normal. Spinal magnetic resonance imaging (MRI) showed a 7.8 × 5.6 × 12.1 cm mass at paraspinal T10-L2 on the left, and the left parts of

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Published online: 29 November 2016

T11-T12 demonstrated destruction (Fig. 1a). Aortic angiography showed that the tumor was close to the aorta and was pushing on it; fortunately, it was well-defined (Fig. 1b). Ultrasound showed an 8.0 × 10.0 cm acoustic area in the lower back, and there was no obvious blood flow signal within (Fig. 1c). No other primary tumors or metastases were detected via emission computed tomography (ECT) except active bone metabolism in T11-T12 (Fig. 1d). After this finding, the patient was proposed for diagnostic surgery.

Computed tomography (CT) was used to guide an aspiration biopsy (Fig. 2a). The pathological result of the aspiration biopsy was confirmed as chordoma via hematoxylin–eosin and immunohistochemical staining. The tumor was vimentin (+), S-100 (+), EMA (+), Ki67 (LI < 1 %), PCK (−), and CD34 (−) (Fig. 2b).

At surgery, we resected three ribs on the right to expose the tumor and complete resection was achieved. Histopathological examination of a postoperative specimen indicated chordoma based on the immunohistochemical profile (Fig. 3). After surgery, the patient did not receive chemotherapy or radiotherapy. At the 30-month post-surgery follow-up, radiography and CT showed spinal internal fixation stability (Fig. 4a, b), with no clinical or radiological evidence of local recurrence or metastases (Fig. 4c). He has achieved good stabilization of the spine and can ambulate with a walker; activities of daily living can be performed independently.

The study was approved by the Ethics Committee of Union Hospital Tongji Medical College of Huazhong University of Science and Technology, and the patient provided informed consent to publish this case report.

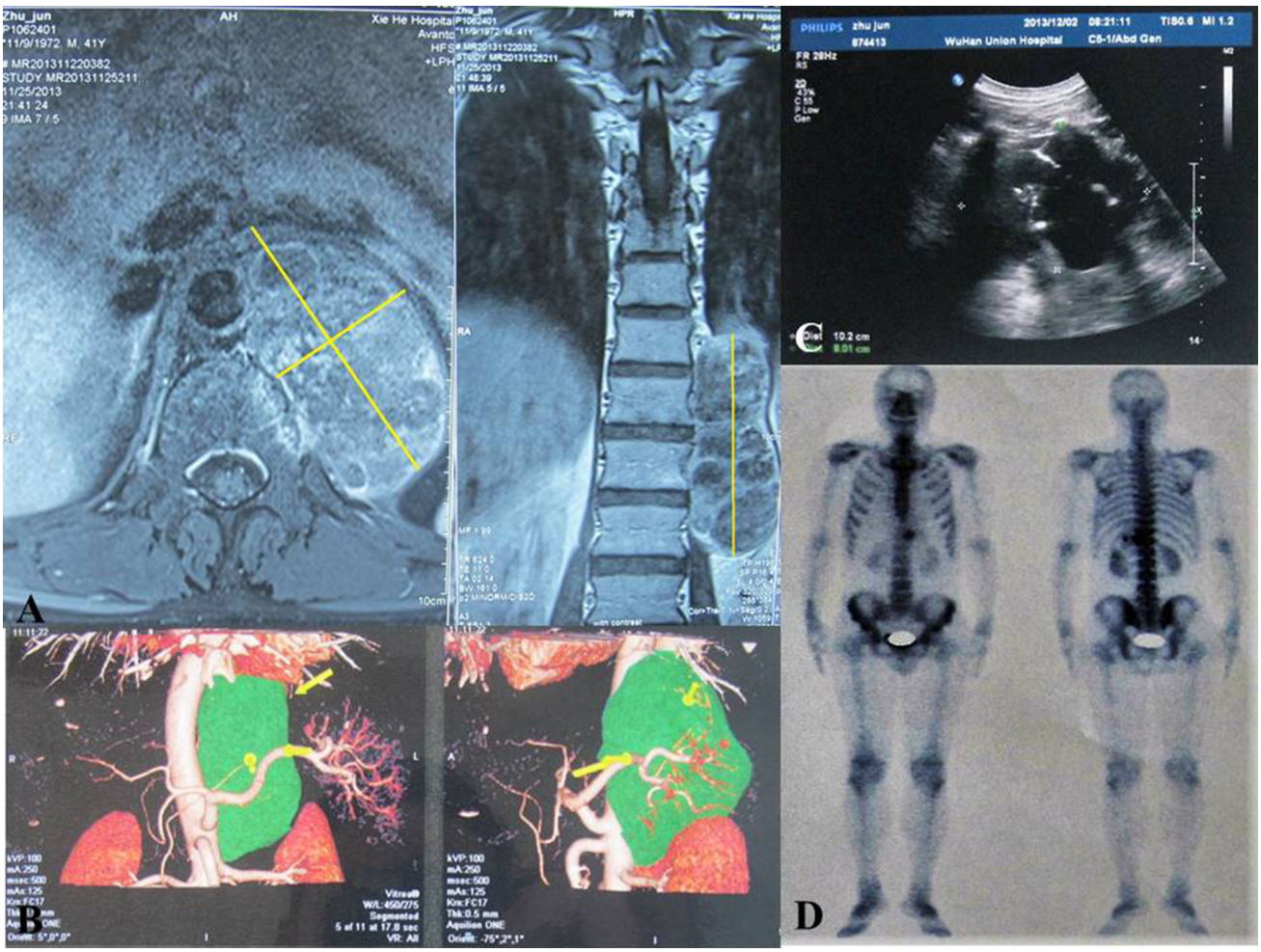

Fig. 1 MRI showing a 7.8 × 5.6 × 12.1 cm mass at paraspinal T10-L2 on the *left*, and destruction of the *left parts* of T11-T12 (**a**). Aortic angiography image showing the tumor is close to the aorta and pushing on it; fortunately, it is well-defined (**b**). Ultrasound image showing an 8.0 × 10.0 cm lower back acoustic area, and there is no obvious blood flow signal inside (**c**). ECT showed no other primary tumors or metastases except active bone metabolism in T11-T12 (**d**)

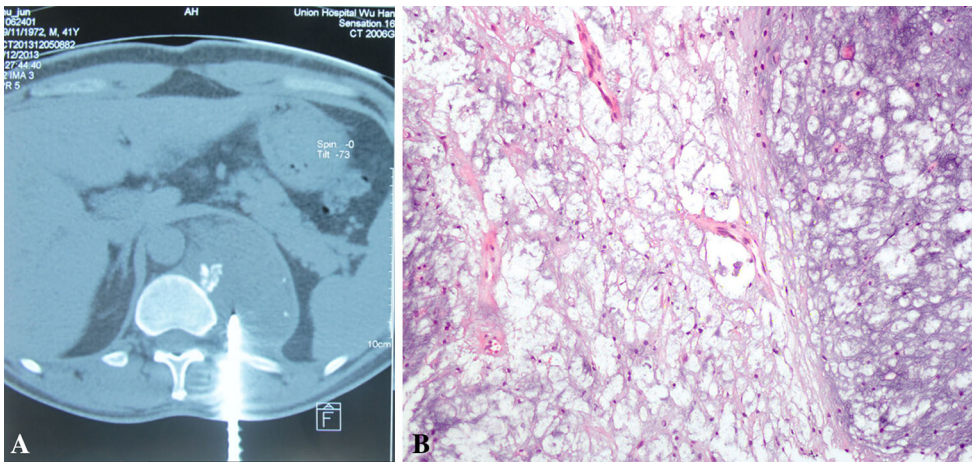

Fig. 2 CT-guided aspiration biopsy (**a**). H&E (×40) staining and immunohistochemistry. The tumor is vimentin (+), S-100 (+), EMA (+), Ki67 (LI < 1 %), PCK (−), CD34 (−) (**b**)

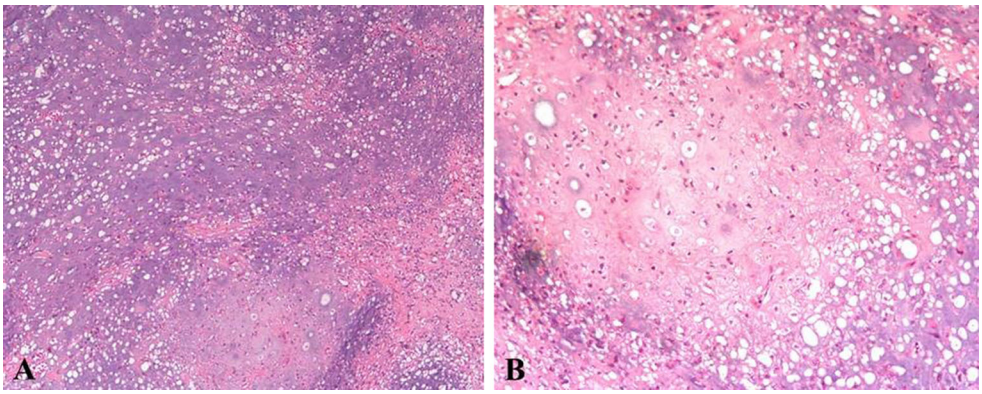

Fig. 3 Histopathological examination of a postoperative specimen indicated chordoma based on the immunohistochemical profile. H&E ×40 (**a**), H&E ×100 (**b**)

Discussion

Being remnants of the primitive notochord, chordomas may occur at the sacral (50 %), cervical (8–9 %), lumbar (5 %), and thoracic (1–2 %) levels, and also at the skull base (35 %) [5–8]. Chordomas are unusual primary vertebral malignancies that should be considered in patients with bony lesions in the mobile spine. We reviewed the published English literature by searching EMBASE, PubMed, Medline, and Scopus. Thus far, only two cases of chordoma in the thoracolumbar spine have been reported [5, 9].

Giant chordoma in the thoracolumbar spine is extremely rare. The early clinical manifestations of spine chordoma are usually subtle or atypical. As the mass increases in size, symptoms of nerve compression usually appear. In our case, the patient presented with progressive low back pain and the tumor was detected via MRI. He had no neurological symptoms, possibly owing to the absence of sympathetic nerve compression. CT and MRI can help doctors to determine the extent of the lesion, and are advantageous for surgical planning. MRI is more suitable for showing the tumor range and intrusion into surrounding organs, but CT is more suited to determine bone damage [10–12]. It is very difficult to identify the nature of a tumor via imaging, and laboratory examination is helpful for the differential diagnosis. In this case, laboratory results such as tumor markers, T-SPOT for tuberculosis, erythrocyte sedimentation rate, and C-reactive protein were all normal. Pathological examination is the gold standard for diagnosis of disease. Histologically, chordoma tumor cells are typical physaliphorous cells, and they possess vesicular nuclei and a soap bubble–like cytoplasm [13–15]. Tissue diagnosis is helpful for treatment planning, and in our opinion, CT guided aspiration biopsy is a safe and effective method for pathological diagnosis.

Surgical resection is the best treatment. Complete resection with a negative margin can reduce the risk of local recurrence and distant metastasis effectively [14].

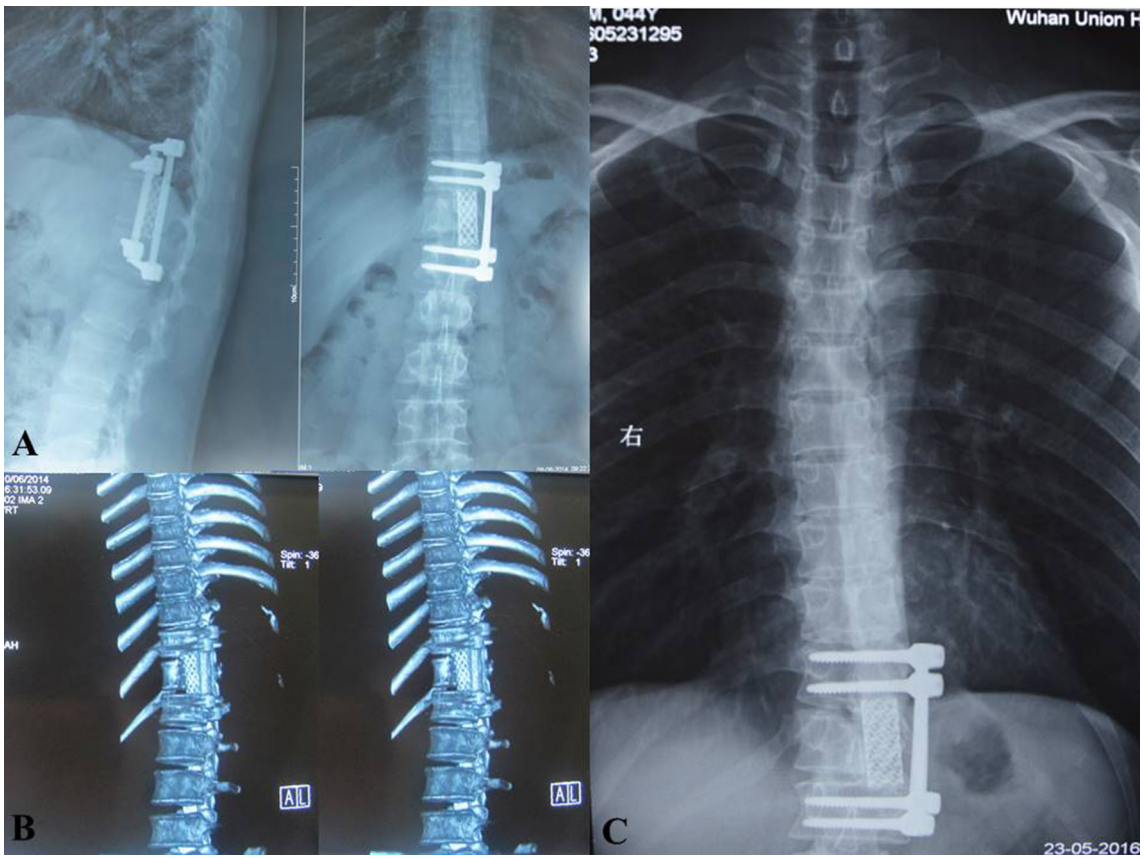

Fig. 4 Radiography and CT images obtained at the 30-month postsurgery follow-up showing the spinal internal fixation stability (**a**, **b**). There is no clinical or radiological evidence of local recurrence or metastases (**c**)

However, the paraspinal tumor was located at T10-L2, and the left parts of T11-T12 demonstrated destruction. Aortic angiography showed the tumor was close to the aorta and pushing on it, but fortunately, it was well-defined. Thus, we resected three ribs on the right to expose the tumor, and then, complete resection was achieved. An internal fixation of the vertebral bodies was performed to stabilize the spine.

Chordoma is not sensitive to radiotherapy or chemotherapy, but recent studies have found some possible methods of treatment. Some studies reported that, to a certain extent, high-dose proton or photon beam radiation can control local recurrence and distant metastasis [16–18]. A study showed that imatinib mesylate can inhibit the growth of chordomas, but we believe that additional research is needed to confirm this conclusion [19]. Our patient refused any postoperative adjuvant therapy. He has achieved good stabilization of the spine and can ambulate with a walker; activities of daily living can be performed independently. There has been no clinical or radiological evidence of local recurrence or metastases at 30 months of follow-up.

Conclusion

In summary, chordoma in the thoracolumbar spine is rare. It is difficult to manage because of its anatomical location and the high risk of local recurrence if the initial surgical margin is positive. Multimodality treatment via surgery and post-operative adjuvant therapy may improve patient outcomes.

Compliance with ethical standards

Conflict of interest The authors have no conflicts of interests.

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