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CASE REPORT

Huge myxoid chondrosarcoma expanded into the thoracic cavity with spinal involvement

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Abstract

Purpose En bloc resection is the treatment of choice of myxoid chondrosarcoma. These tumors can produce huge masses. Anatomical constraints limit the possibility to perform en bloc resection in the spine.

Methods A very huge myxoid chondrosarcoma (14.2 × 10.8 × 11.4 cm) arising from T2 to T5 and invading the whole higher left pleural cavity was observed. Surgical planning according to WBB staging system was performed.

Results The tumor was successfully submitted to en bloc resection achieving a tumor-free margin as demonstrated by the pathologist's report.

Conclusions A careful planning and a multidisciplinary collaboration make possible to perform en bloc resection even in apparently impossible cases.

Keywords En bloc spondylectomy · Spinal myxoid chondrosarcoma · WBB staging system · Multidisciplinary collaboration

Introduction

Myxoid variety of chondrosarcoma is known to produce huge masses [1–3]. This low-malignant tumor is radioresistent [4] and has a very high tendency to recurrence when submitted to intralesional excision [5–9, 13]. Chemotherapy has been proved ineffective and hence not recommended [10–12]. Its tendency to progress to higher level of malignancy at recurrence is also known. En bloc resection is therefore the treatment of choice [2, 3, 13–15]. Regional constraints and the dimension of some spine tumor can make impossible to perform en bloc resection [16–20].

This case is reported to demonstrate that careful planning, including multidisciplinary competences, makes possible to perform en bloc resection even in huge tumors.

Case report

A 34-year-old female was admitted to our department presenting with complains of increasing pain at the back, with a mass on the left back and complete paralysis of the lower extremities for 1 month. The patient initially presented to another hospital 3 months prior to this with a mass on the left back. An open incisional biopsy was performed at the left back in her primary hospital prior to our workup, and the diagnosis from histopathological analysis was of a low-grade myxoid chondrosarcoma.

A physical examination showed that there was complete paralysis of the lower extremities and decreased sensation below the trunk, with dysfunction of the bladder and bowel movement (ASIA impairment scale: B). Plain radiography, computed tomography and magnetic resonance imaging demonstrated an expansive mediastinal mass lesion with thoracic vertebral bodies and ribs involved (Fig. 1). Further

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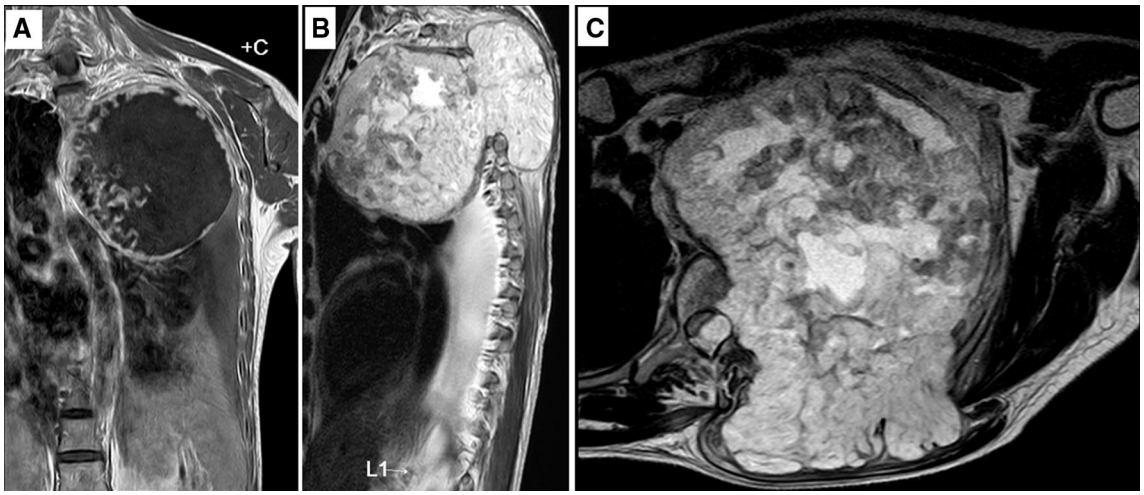

Fig. 1 **a** Coronal, **b** sagittal and **c** axial MRI images showing a huge expansive mediastinal mass lesion (14.2 × 10.8 × 11.4 cm) invading the thoracic vertebral bodies and ribs and spinal cord compression are demonstrated

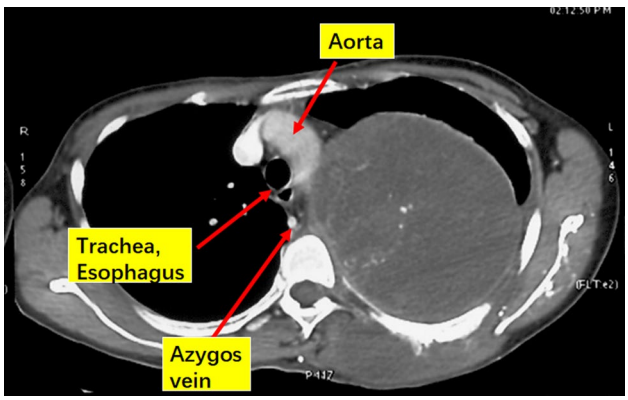

Fig. 2 Chest CT scan, showing the adjacent important structures of the mediastinum. There is a large soft tissue mass within the mottled calcification

investigation revealed no other tumor lesions existed. In the present case, the mass had become involved with the spinal canal and adhered to the aorta, trachea and esophagus (Figs. 2, 3) (Enneking stage IB and WBB stage 1-7/A-D). Selective arterial embolization was performed 1 day prior to the surgery. En bloc resection with a tumor-free margin by one-stage posterior approach was carefully designed by a multidisciplinary team.

Surgical planning by posterior approach (WBB-based en bloc resection type 2a, Fig. 4) [21] and detachment of the spinal muscles from the posterior elements from T2 to T5 were performed. The whole procedure was carried out in the prone position. First, the right pleural cavity was opened by section of the T2–T5 ribs, ligation of the azygos, release of the aortic arch and the esophagus. A dissection of the mass was performed by section of the T2–T6 left ribs and the left scapula. Lateral mass screws were implanted at C6, C7. Pedicle screws were implanted at the levels of T1, T6, T7 and T8. A wide right semi-laminectomy was performed by ultrasonic osteotome from T2 to T5, and the T2–T5 right nerve roots were sacrificed. Then, sectors 8-9-10 were piecemeal excised by high-speed burr to approach the spinal canal at levels T2–T5. A rod was connected on the right side, and osteotomy was performed by Tomita saw at T1/T2 and T5/T6 disk levels. At this time, the huge mass can be rotated on the longitudinal axis, allowing to separate the thecal sac from the mass in the spinal canal under direct vision. The T2–T5 left nerve roots are sectioned after ligation. Then, the whole mass is finally removed without any apparent surgical violation of the margins (Fig. 5a, c, d). The reconstruction was performed by four rods connection at bilateral C6, C7, T6 and T1, T7, T8, respectively. A titanium cage was inserted between the T1–T6 vertebral body, and two vertebral body screws were implanted at T1 and T6 to reinforce the fixation of the titanium cage. Reconstruction of the ribs defect was performed by using two rods connecting the screws and the defect ends in the left side (Figs. 5b, 6d). A chest tube was placed to re-expand the lung.

The operation took 15 h in total, with a total of 5500 mL of bleeding. Ventilator-assisted ventilation was used for 17 days after operation in intensive care unit. After respiratory function exercise for 1 month, she can breathe normally without hypoxic symptoms. Pulmonary infection occurred postoperatively, and etiological examination suggested Acinetobacter Bauman infection. Chest drainage sustained for 64 days. Anti-infective treatment regimen was based on the results of bacterial susceptibility. Imipenem combined with cefoperazone was used for the first month and then cefoperazone only until chest drainage was removed. A slowly skin sensation recovery was observed on the trunk postoperatively. No improvement in muscle

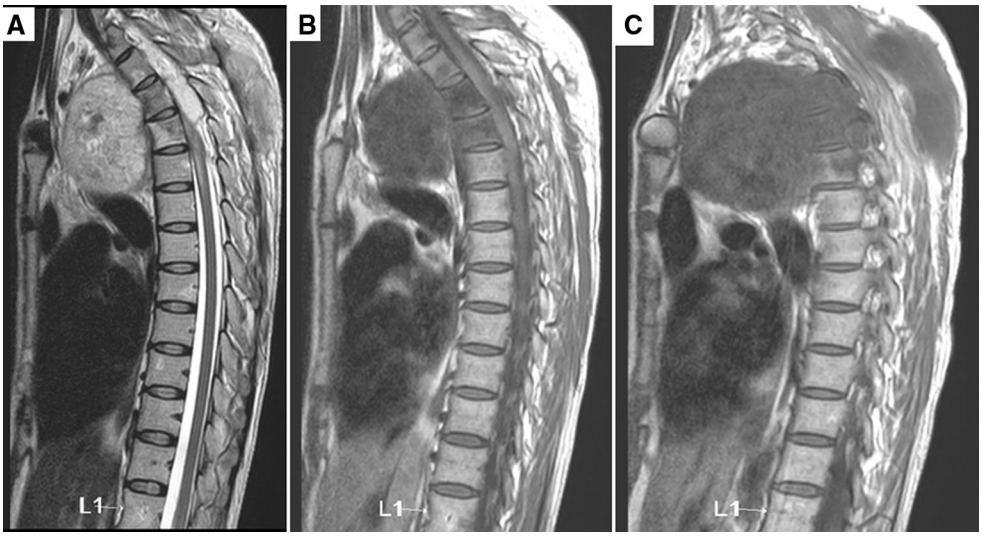

Fig. 3 **a** The sagittal T2-weighted MRI image demonstrates the epidural mass and spinal cord compression. **b**, **c** The sagittal T1-weighted MRI images show low signal intensity in the T2–T5 vertebral bodies and tumor mass

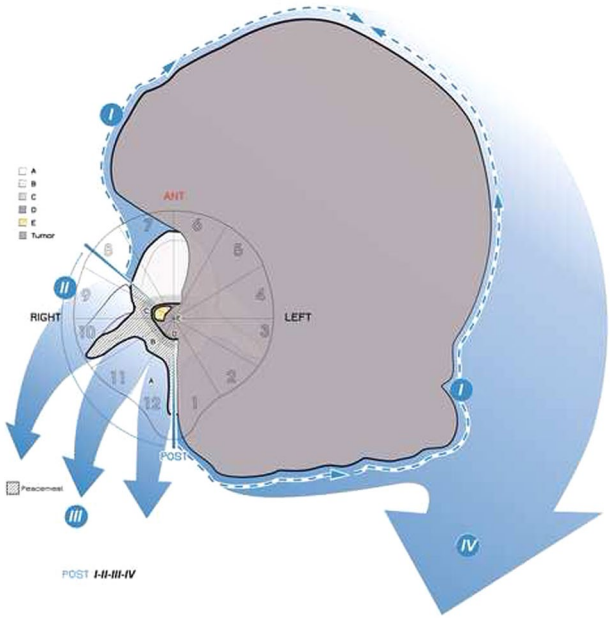

Fig. 4 WBB-based planning of the en bloc resection. Posterior approach only, patient in prone position (type 2b). Step I: completely release of the tumor mass from left and from right. Step II: definition of the approach to the canal: sectors 8–12. Step III piecemeal excision of sectors 8–12, release of the dural sac after section of the nerve roots. Step IV: En bloc tumor removal by careful clockwise rotation along the longitudinal axis

strength of the lower extremities and defecation functions was observed at 3 months postoperatively. Reconstructed postoperative CT scan at 3 months showed no signs of tumor recurrence and well-united bone at the reconstruction site (Fig. 6).

Discussion

From the earliest report by Lievre [22], Hamdi [23] and Stener [24, 25], en bloc resection was later on popularized by Roy Camille [26] and Tomita [27, 28] till more recent attempt to standardize the surgical planning according to the WBB staging system [21, 29]. En bloc resection with tumor-free margins remains the best oncological management of the spine, which aims to a better local control and longer survival rate.

Recurrence of chondrosarcoma usually occurs in 3–5 years postoperatively, and the relapse tends to be faster if an intralesional excision was performed [5, 30]. Thus, a worse prognosis could be achieved. These results were proved to be related to the inadvertent intraoperative contamination [5, 31]. Regional anatomical constraints and huge volumes remain, however, a severe limit to perform an oncological appropriate en bloc resection (it means resulting in tumor-free margins without any tumor contamination), and some reports in the literature of procedures including the separation of the tumor in two or more pieces with intralesional margins [16–20]. A long-term survival rate has only been observed in low-grade spinal chondrosarcoma patients after repeated intralesional excision surgeries, combined with radiotherapy [32].

Here, a case of en bloc resection of a huge myxoid chondrosarcoma is reported, resulting in a fully tumor-free margin resection. To our knowledge, this is one of the largest tumors ever resected by this technique. The peculiar steps were: the ligation of the azygos, the release of the aortic arch and of the esophagus, the opening of a window in the posterolateral vertebrae (T2–T5 in sectors

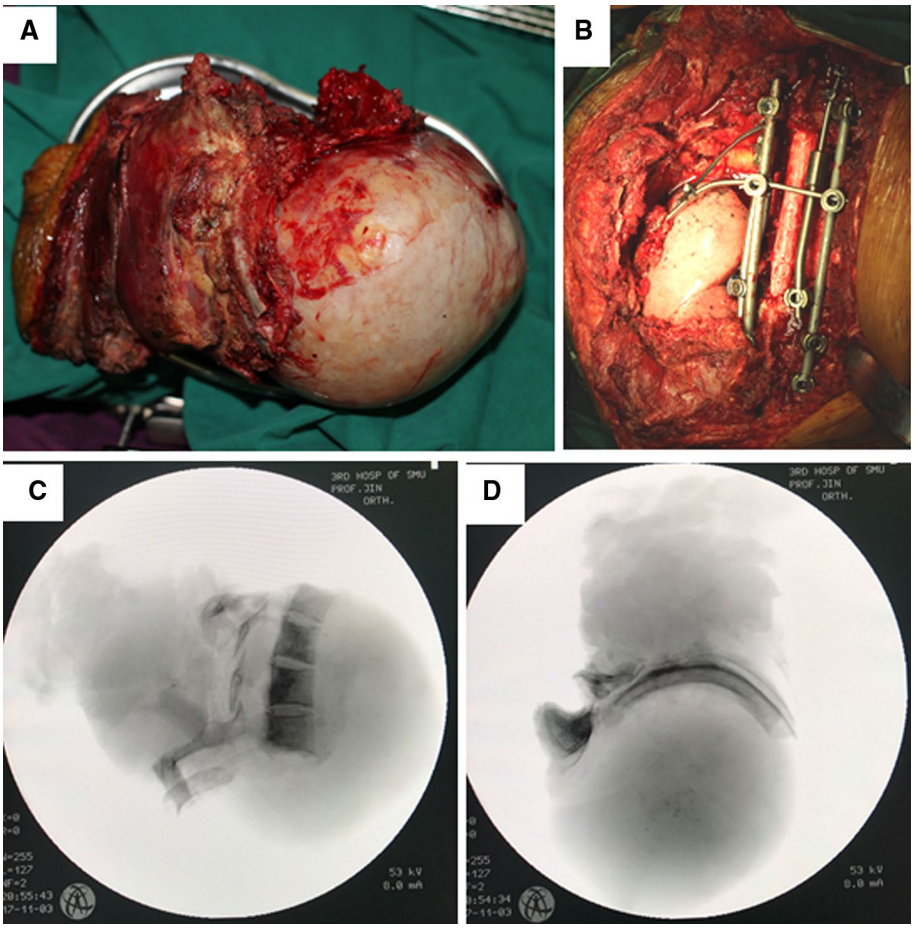

Fig. 5 **a** Photograph of the en bloc resected specimen. **b** Photograph demonstrates the reconstruction of the chest wall defect and spinal stability by screws and rods. **c**, **d** The X-ray images of the en bloc resected specimen

8-9-10) in order to release the dural sac. Once the osteotomies of the spine at the T1/T2 and T5/T6 disk levels were performed, the specimen could rotate, allowing to finalize the resection.

The goal of this surgical procedure was to remove the tumor en bloc with macroscopically healthy margins. Owing to its huge dimension and proximity to vital neurovascular structures, the surgical morbidity and functional impairment must be seriously considered. In the present case, multiple ribs and partial chest wall tissues were excised combined with the mass. The chest wall has both a protective structural around the vital organs of the body and functional role of respiratory movement. Large chest wall defects can cause devastating respiratory and circulatory functional consequences [33, 34]. Reconstruction of the chest wall defect was performed to recreate a stable chest wall with adequate functional capacity. Fadel et al. [35] reported an incidence of pneumonia of 35.3%: 6 patients suffered from pneumonia postoperatively in 17 patients following tumor resection involving the pleura and ribs. The use of sensitive antibiotics in the treatment of pulmonary infection and appropriate ventilator-assisted respiratory is critical for postoperative recovery.

Conclusions

The purpose of this article is to remark the role of preoperative planning in the surgical technique of en bloc resection. However, a careful follow-up will be mandatory for detecting local recurrence, whose risk—due to the histologically proven appropriate margin—is low but not negligible. The procedure

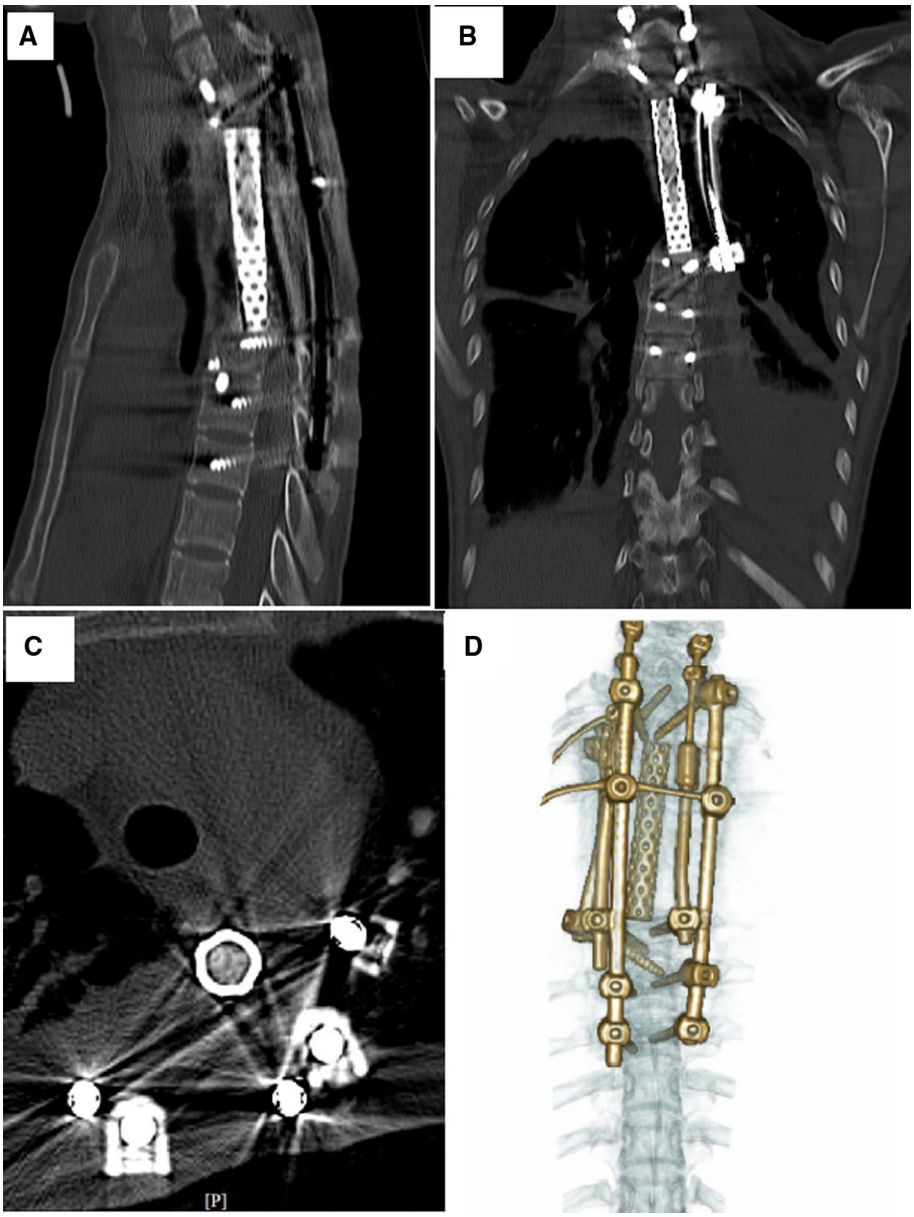

Fig. 6 Reconstructed **a** sagittal, **b** coronal, **c** axial and **d** three-dimensional computed tomography scan 3 months postoperatively

was successfully performed thanks to a very careful preoperative planning and to a multidisciplinary collaboration.

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Compliance with ethical standards

Conflict of interest statement None of the authors has any potential conflict of interest.

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