En bloc resection of a C4 chordoma: surgical technique

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Received: 8 March 2007 / Revised: 14 July 2007 / Accepted: 27 July 2007 / Published online: 23 August 2007
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Abstract The prognosis of aggressive benign and low-grade malignant tumors in the spine as in the limbs, seems to be mostly related to the feasibility of en bloc resection, while in the treatment of high-grade malignant tumors the protocols of treatment include the combination of chemotherapy, radiation and surgery. Indications, criteria of feasibility and surgical technique are extensively reported for the thoracic and lumbar spine. In the cervical spine few cases are reported of resection, due not only to anatomical constraint, but also to the rarity of finding a tumor accomplishing the criteria of feasibility. A case of double-approach vertebrectomy finalized to remove en bloc the body of C4 for a stage IA chordoma is reported. The first stage was posterior, aiming to remove the posterior healthy elements by piecemeal technique. The anterior approach consisted of contemporary right and left prevascular pre-sternocleidomastoid approaches. The specimen was submitted for the histological study of the margins, which resulted tumor-free. This technical note is finalized to confirm that en bloc resection of the vertebral body through total vertebrectomy is feasible in the midcervical spine by double approaches, provided the tumor involves only layers B and C, maximum extension sectors 5–8.

Keywords Chordoma · En bloc resection · Cervical spine

Introduction

A great amount of clinical data can be found in the literature supporting en bloc resection as the only treatment associated with long disease-free survival for chordomas [3, 4, 10, 18, 19, 20, 31]. Treatment of recurrences and intralesional excision cannot prevent local tumor recurrence even if survival can be quite long (with asymptomatic or, more frequently, symptomatic growing tumor). Recently, some evidence is reported that tumor growth can slow by new technique of radiation able to deliver high doses in the deeper areas [5, 12, 28] and by drugs like Imatinib mesylate [11].

In the cervical spine it is very rare to find a chordoma whose features (tumor extension, relationship with unresectable structures, and contamination of the epidural space) fulfill the criteria to perform tumor-free margins en bloc resection.

Only one case of en bloc resection for a chordoma is reported in the cervical spine [18], performed by single anterior approach using the T-saw. This is a very elegant but very demanding surgical technique. Another cervical spondylectomy is reported in a case of osteogenic sarcoma [13].

We report on a case of C4 chordoma, limited to layers B and C, sectors 5–8, therefore prone to be submitted to en bloc resection applying the same technique described for the thoracic and lumbar spine [7].

Case report

A 37-year-old white male complained of cervical pain that radiated to his right arm since 4 months. All possible forms of physical and medical treatments proved to be

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ineffective; the patient was submitted for C6–C7 disc excision and fusion, as the MRI showed major degenerative changes and prolapse (Figs. 1, 2). This surgery gave no substantial benefit to the patient, so more importance was given to signal alterations (hypolucency in T1 and hyperlucent in T2) observed on MRI and previously attributed to “Hemangioma.”

A biopsy was performed by trocar under a CT scan control and the reported diagnosis was chordoma. A specific question was submitted to the pathologist about the differential diagnosis of eccordosis phisaliphora but this option was discarded. The diagnosis of chordoma was confirmed on the basis of immunohistochemical studies. According to MRI the tumor involved the forth cervical vertebra from layers B and C sectors 5–8 (Fig. 3). Preoperative studies included an angiography to demonstrate the perviety of both vertebral arteries in case one of them had to be sacrificed.

Surgical technique included a posterior approach (step 1 in Fig. 4) to remove all the posterior elements (Fig. 5a), isolation of the nerve roots and of the vertebral arteries. Lateral mass screws were inserted from C3 to C7 (skipping of C4). A wide laminectomy of C4 was then performed revealing and isolation of the vertebral artery in this level. A posterior fusion of C3–C7 was applied through the lateral masses screws. At this stage the patient was changed to a supine position. A left pre-sternocleidomastoid incision was done. A formal anterior approach was used to reach the vertebra. The left vertebral artery was explored and

Fig. 1 a, b MRI: C4 changes of signal (low in T1 weighted, enhanced in T2 weighted), together with C6–C7 prolapsed disc

Fig. 2 Discectomy and fusion was performed without symptom remission

isolated removing peacemeal the bone surrounding (step 2 in Fig. 4). A right pre-sternocleidomastoid incision was done and the right vertebral artery was explored and isolated removing peacemeal the bone surrounding (step 3a in Fig. 4). the discs above and below were removed by combined left and right approaches. The vertebral body of

Fig. 3 MRI transverse slice on C4: tumor extends sectors 5–8, layer B and C

Fig. 4 WBB staging system of the reported case (sectors 5–8, layer B and C). En bloc resection of the tumor with appropriate margin is feasible after complete intralesional excision of all sectors 9–4 by posterior approach (step 1), isolating the nerve roots and the vertebral arteries. *Left* anterior approach (step 2) is required for a safe separation from the *left* vertebral artery and for *left* dissection of C4 vertebral body from the adjacent vertebrae. *Right* anterior approach is finalized first to isolate the *right* vertebral artery (step 3a), then to complete the disectomies and the section of the longitudinal ligament. This allow the en bloc resection of the vertebral body (step 3b)

C4 was carefully removed together with the transversely incised PLL (Fig. 5b). A Harms cage was inserted with iliac bone graft and an anterior Caspar plate was used for fixation (Fig. 6a, b).

Discussion

This is the report of a very rare case of cervical chordoma diagnosed while still enclosed inside the C4 vertebral body

Fig. 5 **a** Posterior arch resected. **b** Specimen of the resected vertebral body of C4 including the Chordoma. The margins were confirmed negative by the pathologist

(layers B and C of WBB staging system [8]), that means it could be included in Enneking stage [17] IA. Stage IA Chordoma amounted to 7 on 52 in a recently reported series [9], none of which in the cervical spine. This exceptional finding made possible to perform en bloc resection applying the same procedure proposed for thoracic and lumbar spine. In this peculiar case, due to the previous surgery performed for a prolapsed disk, the reconstruction had to cover three motion segments.

At least a 5-year follow-up is required for chordomas to confirm the effectiveness of the treatment [9], but we decided to submit earlier this case to demonstrate the possibility of early diagnosis of chordoma and to stress on the opportunity to perform—whenever possible—en bloc resection.

Chordoma is a slow-growing malignant tumor arising from the remnants of notochord (in most of cases Enneking stage [17] is IB-low grade malignant, extracompartmental). It shows a marked tendency to recur in case of intralesional excision or biopsy contamination [4]; seeding of the tumor along the wound [2] or even inside the dura is also reported [23]. Metastases are rare to the lung [26], or to the spine [15, 21] exceptional but possible in other sites of the musculoskeletal system or even in the skin, brain, and internal organs [32]. The death in most of the cases is related to the local aggressiveness of the tumor.

The surgical techniques of tumor margin-free en bloc resection have already proved their effectiveness in terms of local control and long-term prognosis for the chordomas occurring in the thoracic and lumbar spine [3, 4, 10, 19, 20, 31] and any effort is justified to perform those procedures even in the cervical spine [18], being accepted that the sensitivity of this tumor to different techniques of radiation is partial or still a matter of discussion on long term analysis [5, 12, 28].

The anatomy of the cervical vertebra is somewhat unique and therefore mandates the surgical technique.

Fig. 6 a, b Post-operative radiogram. The reconstruction was performed by a fixation C3–C7 due to the previous discectomy and fusion C6–C7

Cervical vertebrae have foramina in their transverse processes, which (except in the case of C7) transmit the vertebral vessels and nerves. Therefore before removing en bloc a cervical vertebra the arteries on both side should be released.

Prognosis is heavily affected by the appropriateness of the first treatment. Once recurred, further treatments are always unsuccessful at midterm.

The criteria for performing en bloc resection in the cervical spine are restricted by the surgical anatomy constraints (vertebral arteries inside the vertebra, close relationship with aerial and digestive viscera, bilateral carotid jugular vagus bundle, relative inextensibility of surrounding soft tissues), only under some circumstances (tumor confined inside the bone and not surrounding at least one of the vertebral artery foramen) this complex procedure is justified.

The size of these tumors at the diagnosis is sometimes so huge to prevent from oncologically adequate treatments [14] and allowed—especially in the past—only functional surgery aiming to neurologic decompression or to debulk the tumor [22, 25, 29]. The recent advances in imaging techniques [1, 16, 27, 30] and a more extensive use of MRI play an important role in improving the prognosis of Chordoma by earlier finding of these.

The combination of palliative or debulking surgery with high energy radiation [5] seems promising but not resolute at long term in recurrent tumors or in tumors not suitable for *en bloc* surgery [12, 28, 6, 24]

This case report wants to demonstrate that en bloc resection of the vertebral body is feasible in the cervical spine. The procedure is justified provided that early detection of the tumor is performed by more extensive use of MRI.

Key points

The en bloc resection of the vertebral body is feasible in the cervical spine only under restricted circumstances. Small tumors of the vertebral body (sectors 6 and 7), not expanding in the layer D are suitable to be resected with appropriate tumor-free margins. MRI is essential for early detection of tumors. The indication of such surgery is restricted to aggressive benign tumors like giant cell tumor or low-grade malignancies like chordoma.

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