

Case Report

360-surgery for a giant cervical chordoma with involvement of the right vertebral artery

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ABSTRACT

Background: Chordomas are malignant tumors that arise from the remnants of the notochord. Complete *en bloc* radical resection with postoperative radiation therapy is currently considered the gold standard. Here, we performed a 360-staged approach to manage a C3-C4 chordoma that involved the right vertebral artery.

Case Description: A 40-year-old woman presented with a C3-C4 chordoma that invaded the right vertebral artery. She responded well to a circumferential approach including resection and stabilization.

Conclusion: A 40-year-old woman with a C3-C4 spinal chordoma was optimally managed with a combined anterior/posterior surgical approach including decompression/fusion.

Keywords: Chordoma, Cervical spine, Vertebral artery, 360-degrees

INTRODUCTION

Chordomas are malignant tumors that arise from the remnants of the notochord, anywhere in the axial skeleton.^[1] Chordomas represent 3% of primary bone tumors with a worldwide incidence of 0.08 cases/100,000 person-years. The World Health Organization considers these to be malignant neoplasms.^[2] Pamir *et al.*, analyzed and reported data drawn from the National Cancer Institute's Surveillance, site results were 41% cranial, 30% spinal, and 28.5% sacral. The median survival rate is about 4.7 years across all sites.^[8] Our review of 13 patients over the past 10 years, revealed 38.46% cranial, 46.15% spinal, and 15.38% sacral lesions. Complete *en bloc* radical resection with postoperative radiation therapy is currently considered the gold standard for treating chordomas.^[5] However, this is rarely possible due to the tumor extension to surrounding vital organs and structures, vertebral artery (VA). Here we report a patient who underwent circumferential management of a cervical chordoma with preservation of the ipsilateral VA despite tumor invasion (i.e., due to the risks of major ischemia, and the high risk of tumor recurrence that may invade the contralateral VA).

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CASE REPORT

Presentation

A 40 YO woman presented with a 2-year history of the posterior neck and right shoulder pain; it worsened over the prior year.

Examination

On examination, she had 4+/5 mild weakness involving the right arm (C6, C7 distributions) along with hyperreflexia and decreased pinprick/hypoesthesia in the right C4-C6 distribution. Right Froment's sign positive.

MR and CT findings

T2-weighted MR presented an isointense, homogeneous, well-defined lesion localized in the right side of the vertebral body of C3 and C4 invading the ipsilateral VA, myelo-scan presented compression of the spinal cord. Bone window CT demonstrated lytic lesion of the body and right transverse foramen of C3 and C4 [Figure 1].

Staged surgery

Part I

Stage I was a minimally invasive surgery (MIS) cervical laminectomy on the right of C3 and C4 (using tubular retractors and neurophysiology monitoring). The tumor was soft, gray-looking, with surrounding small vessels. With the help of an ultrasonic aspirator, the lesion was partially resected. There was an additional dorsal and lateral

displacement of the right C5 root and 360° involvement of right VA. Only 50% of the lesion was resected and followed by routine closure. Pathology was consistent with a chordoma [Figure 2].

Part II

A preoperative angiogram was performed to evaluate the patency and dominance of both the compromised right VA and contralateral VA [Figure 3]. The second surgery included a right anterior cervical approach consisting of a microscopic discectomy and full corpectomy of C3 and C4. Tumor invasion was documented extending to the posterior longitudinal ligament (PLL) and the dura. Resection of the PLL was performed and a C2-C5 fusion was achieved using an expandable titanium cage and anterior cervical plate [Figure 4]. Thus, complete resection of the anterior portion of the lesion was successfully achieved.

Part III

The patient was readmitted 4 weeks later for a 3rd stage procedure. A balloon test occlusion of the VA circulation was performed; the test was not tolerated due to right side VA dominance. The patient then underwent a posterior procedure aimed at complete radical resection of remaining posterior arch of C3 and C4 and tumor surrounding the right VA. A gross total removal was achieved under the operating microscope. The patient was routinely discharged 8 days later without pain, and no signs of cerebrospinal fluid leakage and/or new neurological dysfunction. Artist representation (Figure 5).

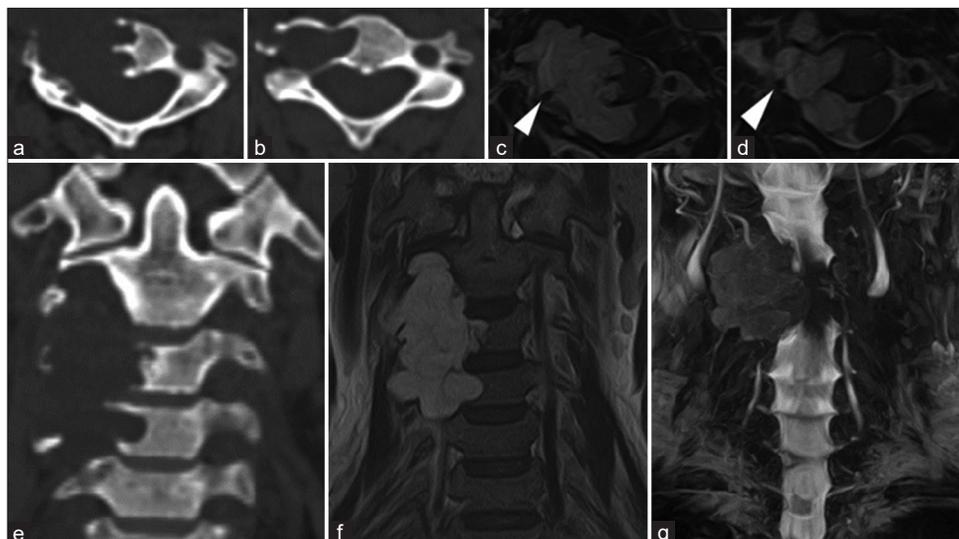


Figure 1: Pre-operative studies. (a) C3 CT bone window, (b) C4 CT bone window, (c) C3 T2-weighted MRI, right VA white arrow head, (d) C4 T2-weighted MRI, right VA white arrow head, (e) cervical spine CT bone window, (f) cervical spine T2-weighted MRI, (g) cervical myelo-scan. CT: Computed tomography, VA: Vertebral artery, MR: Magnetic resonance.

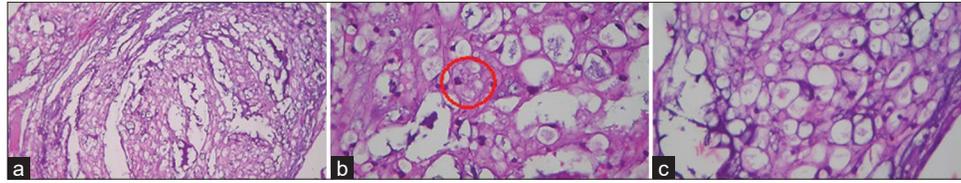


Figure 2: Microscopic images. Hematoxylin and eosin stain. (a) Low power view ($\times 5$) Chondroid chordoma. Neoplastic epithelioid cells with eosinophilic to clear, bubbly cytoplasm arranged in nests, sheets, and individual in a myxoid stroma. (b) Hematoxylin and eosin stain. High power view ($\times 40$) Red circle demonstrates cells with vesicular chromatin and bubbly to vacuolated cytoplasm called physaliphorous cells. (c) Hematoxylin and eosin stain. High power view ($\times 40$) Physaliphorous cells reside in abundant myxoid stroma.



Figure 3: Right vertebral artery angiogram, anterior view. White arrow showing filling defect, due to tumoral compression, at C3-C4 level.

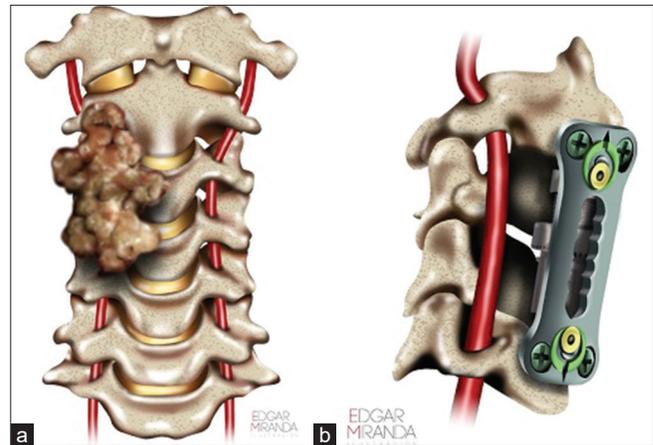


Figure 5: Artist representation. (a) pre-operative illustration. (b) post-operative illustration.

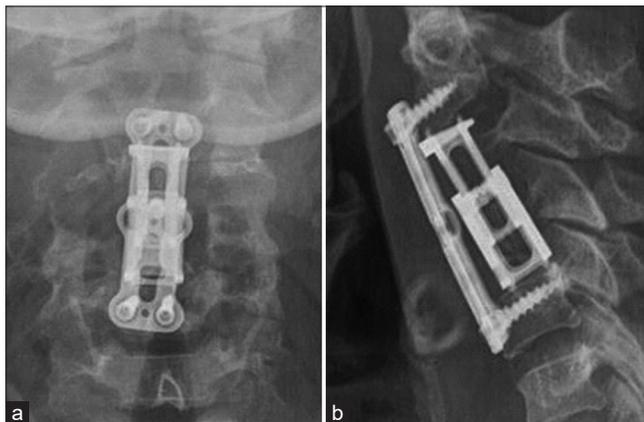


Figure 4: Cervical X-ray showing placement of expandable cage and anterior plate. (a) Anterior view, (b) lateral view.

DISCUSSION

In our institution, we treated 13 patients with histologically confirmed diagnoses of chordoma in the last 10 years. Six patients were diagnosed with spinal chordomas. They presented at a mean age of 57.50 years. The average time from symptom onset to surgery was 5.7 months.

Cervical chordomas are not a common disease, they are often overlooked as a diagnostic possibility in patients with neck pain or palpable mass.^[4,10] However there are few reports of imaging features in literature. In 1998 Wippold *et al.* published one of the most cited articles regarding this topic. They described radiological features of ten cervical chordomas. On gross inspection they form lobulated pseudoencapsulated masses, being the vertebral body the predominant site of affection, with a mean involvement of three levels, being C3-C4 the most common level affected. Lytic lesions were seen in 8 of 10 cases, two cases involved the VA. On MRI no tumor was hyperintense in T1-weighted and 5 out of 6 were hyperintense in T2.^[10] Cui *et al.* published 9 cases of CT and MRI features of cervical chordomas. 9 patients exhibited a multi-lobar soft tissue component that extended over the vertebrae, involved and revealed a collar button appearance in the sagittal plane. The VA was encased in seven patients. Six patients presented with lytic-sclerotic bone destruction. All cases were heterogeneous and hypodense compared with adjacent muscle.^[4] T1 and T2-weighted features were the same as those described by Wippold. These radiological features are the most common pattern as described by previous studies, being lobulated in gross appearance, T1-weighted hypointense, T2-weighted hyperintense, with multi-level involvement.

Table 1: Reports of cervical chordomas excisions with VA involvement.

Author	Patient characteristics	Surgery
Rhines <i>et al.</i> 2005 ^[9]	54 YO man, C2-C4 chordoma, involvement of right VA	2 stage procedure, sacrificed VA, achieved <i>en bloc</i> resection
Bailey <i>et al.</i> 2006 ^[1]	50 YO man, C1-C3 chordoma, involvement of right VA	3 stage procedure, sacrificed VA, achieved <i>en bloc</i> resection
Jiang <i>et al.</i> 2009 ^[7]	26 YO woman, C2-C3 chordoma, involvement of left VA	1 stage combined anterior and posterior approach, sacrifice of VA and C3 nerve root, achieved <i>en bloc</i> resection
Guppy <i>et al.</i> 2013 ^[6]	49 YO woman, C2-C3 chordoma, involvement of right VA	3 staged procedure, endovascular sacrifice of right VA, achieved <i>en bloc</i> resection

VA: Vertebral artery

The gold standard is *en bloc* resection, however, this was not initially considered in our patient due to the suspicion of right VA irruption, thus we had to perform a 360°, MIS, multiple staged approach to achieve maximal safe resection. For tumor extension, we used the Weinstein-Boriani-Biagini (WBB) classification.^[2] We classified the lesion for C3 as WBB 7-10 A-D, and C4 as 8-9 A-D. However, The Spine Oncology Study Group proposed a modification by adding a letter “F” to indicate VA involvement.^[3]

Currently, there are no guidelines regarding the management of cervical chordomas with invasion and encasement of the VA. In retrospect, a full angiography assessment of the patency of both VA prior to the first stage could have been made. After this case, we believe this step to be crucial in the management of this kind of pathology. Due to our own personal experience, we considered a MIS posterior approach to be the best option as the initial step, however an open full laminectomy of C3 and C4 is a suitable option as well, the decision should be made based on the surgeon expertise. Anterior tumor resection utilizing a C3, C4 corpectomy technique with an expandable cage and plating was effective for gross total anterior tumor resection. It additionally facilitated removal of tumor around the right VA.

There are limited reports of cervical chordomas managed with circumferential surgery where the VA was also involved, four cases are described [Table 1].

CONCLUSION

Cervical chordomas are slow-growing malignant tumors resulting in spinal cord compression and occasional invasion of the VA. With appropriate preoperative testing, these lesions may be resected *en bloc* in stages, and accompanied by selective VA sacrifice.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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