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# Novel reconstruction of the anterior craniocervical junction using an expandable cage with integrated fixation after total C2 spondylectomy for chordoma



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## ABSTRACT

Chordoma is a locally aggressive malignant tumor that generally occurs in the clivus, mobile spine and sacrum. While *en bloc* resection with wide margins has been advocated as the only cure for chordomas, tumor characteristics and violation of critical anatomical boundaries may preclude pursuing this treatment option in the cervical spine. We present a C2 chordoma in a 35-year-old man with epidural and prevertebral extension that was treated with a single stage anterior–posterior total C2 spondylectomy with novel reconstruction using an expandable cage with integrated fixation followed by stereotactic radiosurgery. Single stage intralaminar total C2 spondylectomy via anterior transoral and posterior approaches was performed. The anterior column was reconstructed using an expandable cage with integrated fixation from the clivus to C3. The patient maintained his intact neurological status at 6 month follow-up with full resumption of activities of daily living without any significant morbidity.

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## 1. Introduction

Chordomas originate from the notochord remnant and are the most common malignant primary spinal tumor after lymphoproliferative tumors. Cervical spine involvement accounts for only about 6% of all cases with a predilection to involve the upper cervical spine [1]. *En bloc* resection of chordoma is often recommended and has been shown to be associated with longer recurrence free survival, overall survival and potential cure in some cases [1–3]. Nevertheless, the necessity to preserve cervical nerve roots, protect the vertebral arteries, and the complex osseous anatomy makes this goal formidable [4]. We describe a patient with C2 chordoma who was treated with single-stage C2 spondylectomy via combined transoral–posterior approach with circumferential reconstruction of the anterior craniocervical junction using an expandable cage with integrated fixation.

## 2. Case illustration

A 35-year-old man with no significant past medical history presented with new onset of dysphagia, and neck pain for 2 months. His physical examination was otherwise unremarkable. MRI

demonstrated a destructive lesion involving the C2 vertebrae with extensions into retropharyngeal and ventral epidural spaces (Fig. 1) that was confirmed to be a chordoma on a CT scan guided biopsy. The tumor had violated numerous critical anatomic boundaries (anterior longitudinal ligament, longus colli, and fragmenting C2) making true *en bloc* resection impracticable. A decision was made to perform a total intralaminar C2 spondylectomy followed by stereotactic radiation.

After induction of general anesthesia and an elective tracheostomy, anterior access to the upper cervical spine was obtained via the transoral approach with pharyngotomy and soft-tissue palatotomy. As the tumor was encountered no clear border could be observed between tumor and normal tissue, the anterior longitudinal ligament was violated and a pathologic fracture was observed. The bony portion could not be mobilized as a single segment. This necessitated an intralaminar gross total resection. The anterior arch of C1 and dens were removed following separation of ligamentous attachments (Fig. 2). The C3 endplate and tip of the clivus were prepared for interbody arthrodesis and an expandable cage with integrated fixation (Fortify-I, Globus Medical, Audubon, PA, USA) packed with morcellized allograft was inserted into the space between the C3 endplate and the clivus. Following closure of the transoral incision, the patient was positioned prone and underwent removal of rest of the C2 vertebrae, and occipito-cervical fusion and instrumentation.

The patient developed a naso-palatal fistula that required repair and placement of a percutaneous gastric tube. He subsequently

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**Fig. 1.** T2-weighted sagittal (A) and axial (B) MRI of the cervical spine revealing a heterogeneous mass involving C2. T1-weighted sagittal (C) post-gadolinium MRI with diffuse enhancement of the anterior elements of C2.

received photon-based intensity-modulated radiotherapy followed by proton-beam boost. Post-operative radiographs revealed a

stable fusion construct and good anatomic alignment with no evidence of tumor recurrence on MRI (Fig. 3).

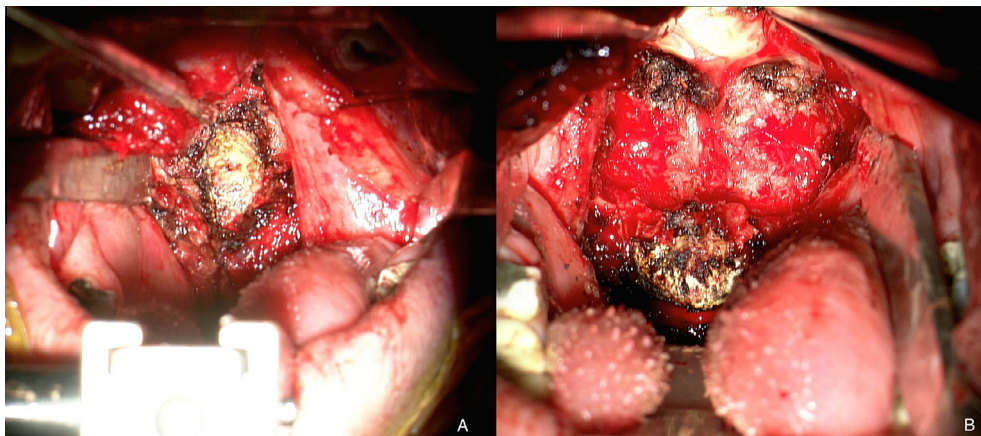
### 3. Discussion

Surgical treatment of chordoma involving the cervical spine remains one of the most technically challenging spinal operations because of the close proximity of vertebral arteries, nerve roots, spinal cord, and the unique spinal biomechanics in this region, making performance of an *en bloc* spondylectomy with reconstruction formidable. Studies have shown a rate of wide or marginal margin resection in patients with chordomas involving the mobile spine of only 21% [5]. Moreover, even in patients with wide margin resections, recurrence rates ranging from 17% to 60% have been reported in studies with longer follow up [5,6]. When the tumor extends into the epidural space, an intralesional gross total resection of the cervical spine chordoma with adjuvant post-operative radiation therapy remains a viable management strategy as *en bloc* resection is not feasible [7]. Though further studies with longer follow up are needed, sustained responses to high-dose stereotactic radiotherapy have been shown in recent studies.

Various strategies for spinal column reconstruction and stabilization after resection of a chordoma in the upper cervical spine have been reported [8,3,9]. The most common methods of C2 replacement include fibular graft, Harms cage, custom wire cages with anterior cervical plate, and even three-dimensional printed cages. Although these strategies are reasonably effective, they often require a larger anterior surgical corridor and suffer from the associated complications. An expandable cage such as illustrated here requires a smaller surgical corridor and may minimize approach-related morbidity. Additionally, strategies using the surgical techniques and adjuvant radiotherapy as described here may offer a valuable treatment option in patients who are not candidates for true *en bloc* excision [9].

### 4. Conclusion

Total spondylectomy of the C2 vertebrae is technically challenging due to the anatomical and biomechanical constraints inherent to the upper cervical region. The use of an expandable cage with integrated fixation for reconstruction of the anterior craniocervical junction may reduce the need for more extensive transoral exposures.



**Fig. 2.** Intra-operative view of a subperiosteal dissection of the odontoid process (A) and after odontoidectomy (B).

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