

En bloc resection of a C2–C3 upper cervical chordoma: Technical note

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Abstract

Background: Recently, *en bloc* spondylectomy for upper cervical chordomas has been reported. Most authors utilize the combined approaches (e.g., transoral tumor resection with anterior column reconstruction and primary pharyngeal closure without up-front flap repair). However, the 60% incidence of posterior pharyngeal wall dehiscence delays oral intake, typically requires an additional surgery (e.g. free-flap), and delays radiation therapy.

Methods: Here, we demonstrate the efficacy of *en bloc* C2–C3 spondylectomy for the treatment of upper cervical chordomas utilizing a combined transoral followed by posterior approach. We used a novel anterior de-epithelialized submental island flap (SIF) as an underlay graft between the pharyngeal wall and cage/hardware to prevent pharyngeal wound dehiscence.

Results: Despite a small pharyngeal fistula, the construct healed and the patient was disease-free 40 months later.

Conclusion: *En bloc* C2–C3 spondylectomy for the treatment of an upper cervical chordoma typically requires a combined transoral and posterior approaches. This required utilization of an anterior SIF to promote adequate wound healing. This maneuver avoided incurring the typical complications of combined approaches (e.g. transoral tumor resection with anterior column reconstruction and primary pharyngeal closure without up-front flap repair).

Key Words: Chordoma, *en bloc*, oncology, spine, submental island flap

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INTRODUCTION

Only 4–5% of chordomas arise in the cervical spine.^[7,8] *En bloc* resection is challenging due to the proximity of vital structures (e.g., the vertebral arteries [VAs], cervical nerve roots, and spinal cord). Traditional intralesional resection of upper cervical chordomas results in a high local recurrence rate, with up to a 70% long-term mortality rate.^[7,8] Over the last decade, 12 reports utilized a combined posterior and anterior transoral approach to achieve *en bloc* resection of upper cervical chordomas.^[1–5,10] Here, we performed a marginal-margin

en bloc resection of a C2–C3 chordoma utilizing a combined posterior-anterior (transoral) approach and

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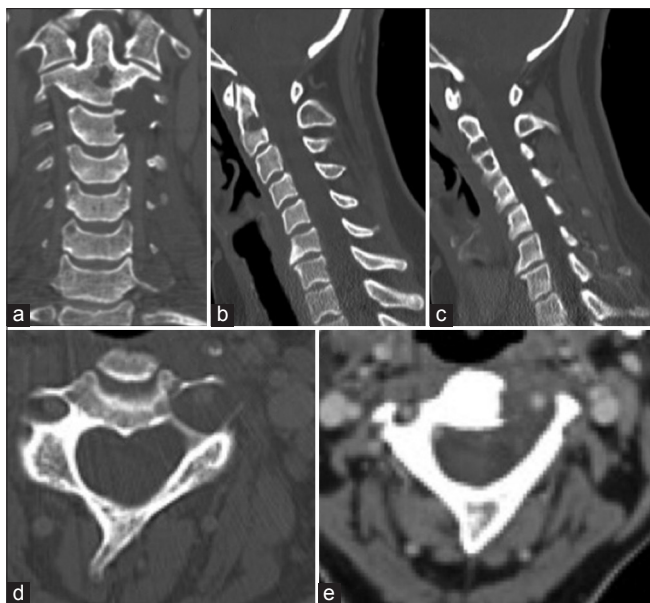


Figure 1: Preoperative coronal (a), sagittal (b and c), and axial (d) computed tomography scan showing the bony lesion involving the bodies of C2 and C3, as well as the left C2 and C3 transverse foramina, which are widened. Computed tomography-angiogram (e) confirms the left vertebral artery which is a patent at both C2 and C3 and pushed anteriorly by the tumor

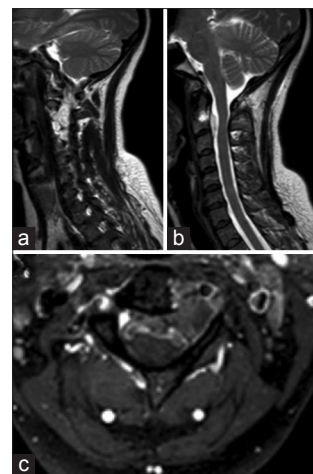


Figure 2: Preoperative sagittal T2-weighted image (a and b) showing the extension of the tumor from the left C2 and C3 vertebral body to the left epidural space. Axial T1-weighted magnetic resonance imaging with gadolinium (c) shows the left C2–C3 tumor involving the left C3 pedicle and transverse foramina

novel de-epithelialized submental island flap (SIF) as an underlay graft overlay between the pharyngeal wall and cage hardware to prevent pharyngeal wound dehiscence.

CASE REPORT

A 46-year-old female presented with neck and shoulder pain following a car accident. The cervical computed tomography (CT) scan and magnetic resonance imaging (MRI) studies revealed an osteolytic lesion involving the left C2 and C3 vertebral bodies. The lesion encased the left VA, and extended foraminally into the epidural space [Figures 1 and 2]. A CT-guided biopsy revealed a chordoma. The left VA was coiled/occluded preoperatively between the C1 and C5 levels (e.g., after placing a distal protective intra-arterial balloon, there were no postocclusion deficits). The initial posterior approach included: Excision of the posterior elements with facetectomies from C2 to C4, ligation of the left C2 and C3 nerve roots, isolation/clipping of the left VA between C1 and C2, and occipito-cervico-thoracic fusion.

The second stage performed 1-week later involved securing the airway with a tracheotomy, exposing the upper spine from C1 to C5 utilizing a transoral transmandibular, and an extended anterior cervical approach, followed by C2, and C3 corpectomy. This facilitated *en bloc* resection of the tumor, and anterior column reconstruction with a Harms cage filled with allograft. Protection of the instrumentation with a submental flap, and refixation of the split mandible was followed by closure of the posterior pharyngeal wall and the neck incision [Figures 3-5].

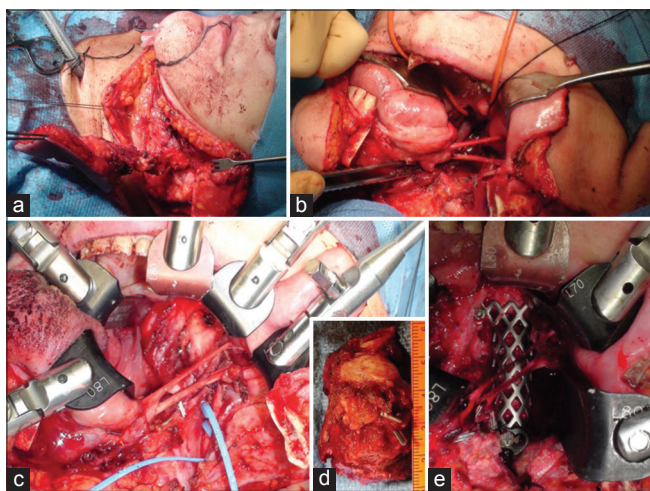


Figure 3: After preoperative embolization of left vertebral artery and posterior approach with C2–C3 laminectomy, left facetectomy and sectioning of the left C2 and C3 nerve roots and occipito-cervico-thoracic fusion, the patient underwent an anterior approach. Intraoperative photographs demonstrating the transoral/transmandibular/extended anterior cervical approach. A submental flap was included in the incision and later used to cover the hardware (a). The subaxial cervical spine was exposed following the mandibulotomy, submandibular gland resection, with the preservation of the lingual, hypoglossal and glossopharyngeal nerves (b). The upper cervical spine was exposed following the soft palate and posterior pharyngeal wall incision and retraction. With the syframe retractor in place, a wide anterior exposure extending from the mid-clivus to the level of C5 inferiorly was obtained (c). Following *en bloc* removal of the tumor (d), the anterior column was reconstructed with a titanium T-shaped Harms (e) which was fixed to the lateral masses of C1 with screws and buttressed in place on the body of C4 with an anterior plate extending from C4 to C5



Figure 4: Lateral (a) anteroposterior (b) and oblique (c) cervical radiographs demonstrating the posterior occipito-cervico-thoracic instrumentation and anterior reconstruction with titanium mesh cage and C4–C5 anterior plate. The embolized left vertebral artery has been removed with the tumor expert for coils above and below the points of ligation (d) Computed tomography scan showing the C1 lateral mass screw through the T-shaped titanium cage (e) and C1 lateral mass

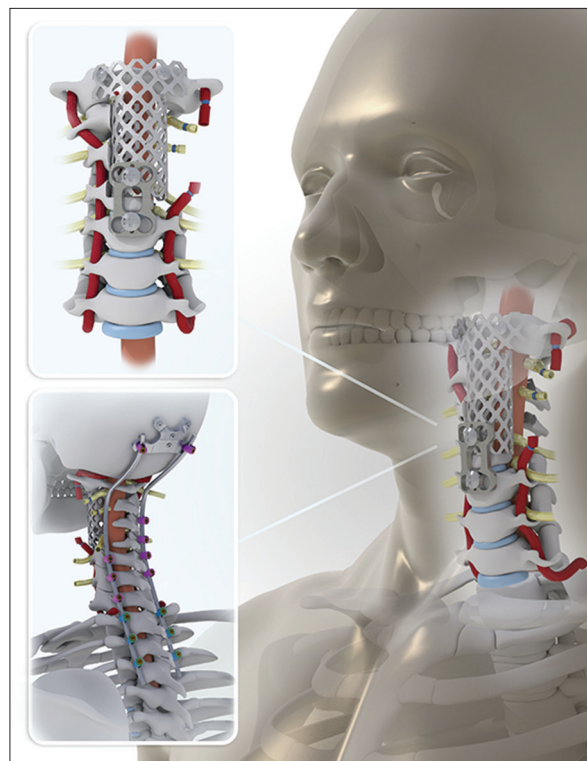


Figure 5: Medical illustration demonstrating the final anterior and posterior constructs. The nerve root and vertebral artery sacrifice are seen

Postoperative morbidity following the second surgery included: Transient facial/glossal edema, ventilation until the seventh postoperative day due to the diaphragmatic weakness from C2–C3 nerve root sacrifice (which progressively resolved), and partial left eleventh nerve palsy. Thirty-five days after the second surgery (anterior approach), the patient underwent a third surgery to realign the head by about 15° posteriorly at the occipito-cervical junction utilizing a posterior approach. Severe dysphagia initially required a nasogastric tube and later a gastrostomy; 60 days later, she was finally started on oral feeding. At the final 40 months follow-up, the patient was functioning normally, including no deficits in speech and swallowing. The repeat cervical CT and MRI revealed no tumor recurrence.

DISCUSSION

Controversial *en bloc* spondylectomy

Controversy still exists regarding optimal constructs for *en bloc* C2 spondylectomy. In clinical series involving spondylectomy and reconstruction of the upper cervical spine, high pseudarthrosis rates of 14–66% have been reported.^[7] Posterior occipito-cervical fusion alone is insufficient following complete C2 spondylectomy.^[9] Anterior reconstruction with a titanium mesh cage, such as the one used in our patient, fixed from C3 to the clivus, or C1 ring/odontoid has been shown to completely eliminate the biomechanical instability, even without posterior stabilization, and is advocated by most authors.^[9]

Intralesional resection

In upper cervical chordomas, intralesional resection with adjuvant radiation therapy is associated with a 40% recurrence and 67% mortality rate at 5 postoperative years.^[7] Only, nine prior cases and one multicenter study (involving three new cases) have previously described the successful *en bloc* resections of upper cervical spine chordomas.^[1–3,5,7,8,10] Although the survival benefits of *en bloc* spondylectomy over intralesional resection with adjuvant Proton Beam Radiation Therapy (PBRT) are well documented, as in our case with disease-free survival of 40 months, this technique is associated with a 70% morbidity (e.g., transient dysphagia, transient ventilator-dependence requiring prolonged Intensive Care Unit stay), the potential for construct pseudarthrosis, and pharyngeal wound dehiscence.^[5,7,8] Three of the five reported cases of transoral *en bloc* spondylectomy developed pharyngeal wall dehiscence.^[3,8] This was attributed to a thin pharyngeal wall whose sutures were too close to the anterior construct, leading to a delay of oral intake, need for a complex secondary surgery (e.g., requiring a free-flap pharyngeal wall repair), increased the risk of infection, and possible delay of adjuvant radiation therapy in cases

with residual tumor.^[4] Here, we demonstrated that a pedicled SIF can be readily harvested in combination with an anterior transoral approach, and that it can be readily mobilized to reach the prevertebral space. The de-epithelialized SIF, located in the prevertebral space behind the pharyngeal wall, protected the anterior construct from the saliva and permitted both the small postoperative pharyngeal fistula to heal spontaneously and decrease the risk of dehiscence infection.

CONCLUSION

Upper cervical C2–C3 spine chordomas may be managed with *en bloc* stand-alone resection with negative (marginal) margins, accompanied by the up-front use of a de-epithelialized SIF to prevent the posterior pharyngeal wall dehiscence. Although this approach is associated with a high frequency of pre- and post-operative complications, several technical pearls may reduce the morbidity and potential mortality.

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Conflicts of interest

There are no conflicts of interest.

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