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En bloc resection of a high cervical chordoma followed by reconstruction with a free vascularized fibular graft: illustrative case

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BACKGROUND Wide excision of chordoma provides better local control than intralesional resection or definitive radiotherapy. The en bloc excision of high cervical chordomas is a challenging endeavor because of the complex anatomy of this region and limited reconstructive options.

OBSERVATIONS This is the first case report to describe reconstruction with a free vascularized fibular graft following the en bloc excision of a chordoma involving C1–3.

LESSONS This report demonstrates the durability of this construct at 10-year follow-up and is the first case report demonstrating satisfactory long-term oncological outcomes after a true margin-negative resection of a high cervical chordoma.

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KEYWORDS high cervical chordoma; free vascularized fibular graft; FVFG; en bloc resection; chordoma

Chordomas are tumors that arise from vestigial notochordal remnants and are the most common primary spine tumor in adults.[1] Chordomas are invasive, can metastasize, and are lethal without appropriate treatment.[2] Surgical excision is typically indicated given the refractory nature of these tumors to chemotherapy and radiation therapy.[3] Chordomas occurring in the cervical spine, which account for 5%–7% of all chordomas, are challenging to resect due to the complex anatomy of this region.[4–7] Consequently, cervical chordomas have historically been treated with intralesional resection.[7–26] However, recent studies have shown that en bloc excision of a resectable cervical chordoma results in superior local control in comparison to intralesional excision or radiotherapy.[4,27–38]

The en bloc resection of high cervical chordomas (those involving the C1 to C3 vertebral levels) remains a particularly technically challenging endeavor due to the need for a complex multidisciplinary surgical approach, proximity of neurovascular structures, and limited reconstructive options in an area of significant strain.[30,32–35,39] Several case reports have reported negative margins following en bloc resection of a high cervical chordoma, but these cases have been fraught with postoperative hardware complications related to the anterior column reconstruction and only include midterm oncological follow-up.[30–35] As such, the long-term oncological outcomes and best method for obtaining anterior column arthrodesis remain unknown.

This is the first case report to describe anterior column reconstruction with a free vascularized fibular graft following the en bloc excision of a chordoma involving C1–3. This report demonstrates the durability of this construct at 10-year follow-up and is the first case report demonstrating satisfactory long-term oncological outcomes after a true margin-negative resection of a high cervical chordoma.

Illustrative Case

History and Examination

A 67-year-old male presented to his primary care physician because of a 3-month history of neck pain that began following a ground-level fall onto his right shoulder. The patient had no signs or symptoms of radiculopathy or myelopathy at the initial presentation. Physical examination demonstrated 5/5 strength on manual motor testing of the bilateral, deltoids, biceps, triceps, wrist extensors, finger flexors, and hand intrinsics per the American Spinal Cord Injury

ABBREVIATIONS CSF = cerebrospinal fluid; CT = computed tomography; FVFG = free vascularized fibular grafting; ICU = intensive care unit; O–C2 = occiput-to-C2; PEG = percutaneous endoscopic gastrostomy.

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Association/International Spinal Cord Society International Standards for Neurological Classification of Spinal Cord Injury. His sensory examination was remarkable for normal sensation to light touch at the occiput and decreased sensation to light touch involving the right ear, face, and neck. He also demonstrated normal upper and lower extremity reflexes, negative Hoffman's sign, plantar Babinski reflex, negative Romberg sign, and normal tandem gait.

Imaging

Radiographs of the cervical spine were obtained and demonstrated no evidence of pathology involving the cervical spine. Given the patient's persistent neck pain refractory to over-the-counter pain medications, advanced imaging was obtained to further evaluate the etiology of the patient's ongoing pain. Computed tomography (CT) scan of the cervical spine demonstrated a destructive lesion with peripheral sclerosis involving the C2 body and odontoid with destruction of the posterior wall of the vertebral body (Fig. 1). A subsequent magnetic resonance imaging scan of the cervical spine demonstrated a T2 hyperintense enhancing mass centered in the C2 body with extradural extension that displaced the spinal cord without evidence of myelomalacia (Fig. 2). Additionally, the mass extended into the odontoid, anterior arch and right lateral mass of C1, right C2–3 neural foramen, and right transverse process of C3, with complete encasement of the right vertebral artery. These imaging findings were deemed consistent with chordoma as the most likely diagnosis.

Staging and Preoperative Planning

A CT-guided needle biopsy was performed, and a pathology review of the tissue specimen was consistent with chordoma. A CT scan of the chest, abdomen, and pelvis demonstrated no evidence of metastatic disease. The patient was counseled extensively regarding the various treatment options, including isolated radiotherapy, gross-total resection with adjuvant radiotherapy, and en bloc resection with adjuvant proton beam therapy. After a thorough discussion of the risks and benefits of each approach, the patient elected to pursue en bloc tumor excision followed by circumferential fusion and reconstruction of the anterior column with free vascularized fibular grafting (FVFG).

Surgical Technique

Preoperatively, temporary balloon occlusion of the right vertebral artery was performed by our interventional radiology colleagues. This maneuver resulted in no changes in neurological potentials, and so the interventional radiologists proceeded with embolization of the right vertebral artery at the C6 level.

During the first stage of surgery, the patient was positioned prone with his head secured in the Mayfield head holder (Integra). Care was taken to position his neck in neutral alignment with an occiput-to-C2 (O–C2) angle of 33°—identical to the O–C2 angle measured on his preoperative standing radiographs. We began by harvesting 3 corticocancellous struts and additional cancellous autograft from the right iliac crest. The spine was then exposed from the occiput to C7 and laterally to the tips of the lateral masses, except at C2 and C3 to avoid violating the tumor at this location. A

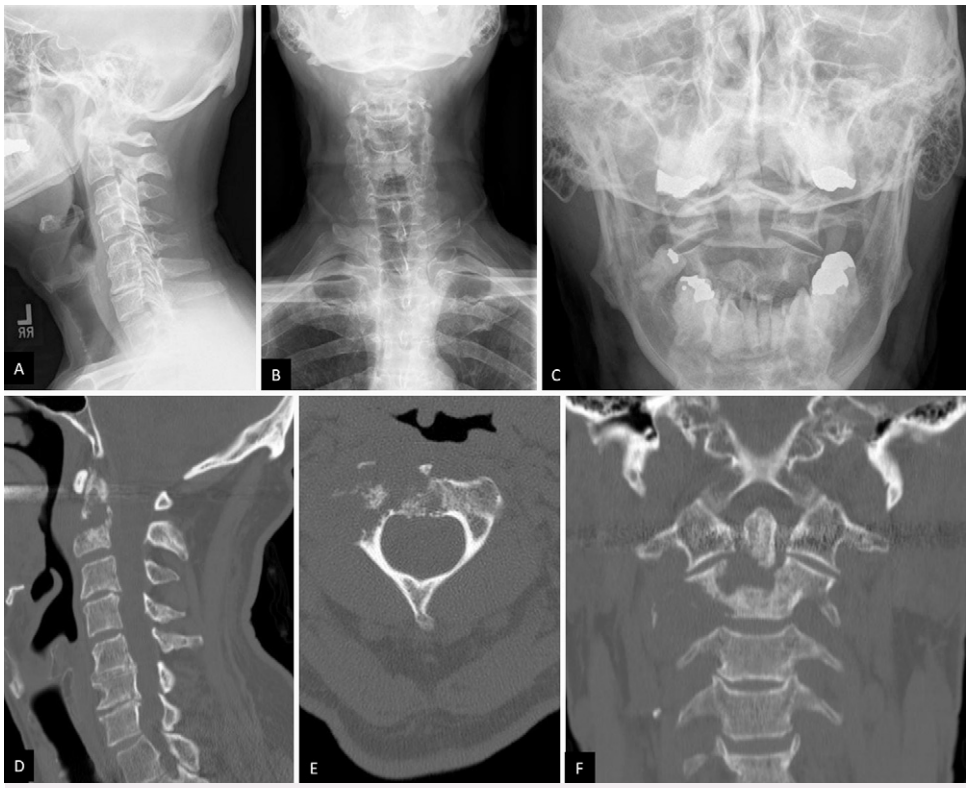

FIG. 1. Preoperative standing radiographs of the cervical spine, including lateral (**A**), anteroposterior (**B**), and open-mouth odontoid (**C**) views and preoperative CT scans of the cervical spine (**D–F**).

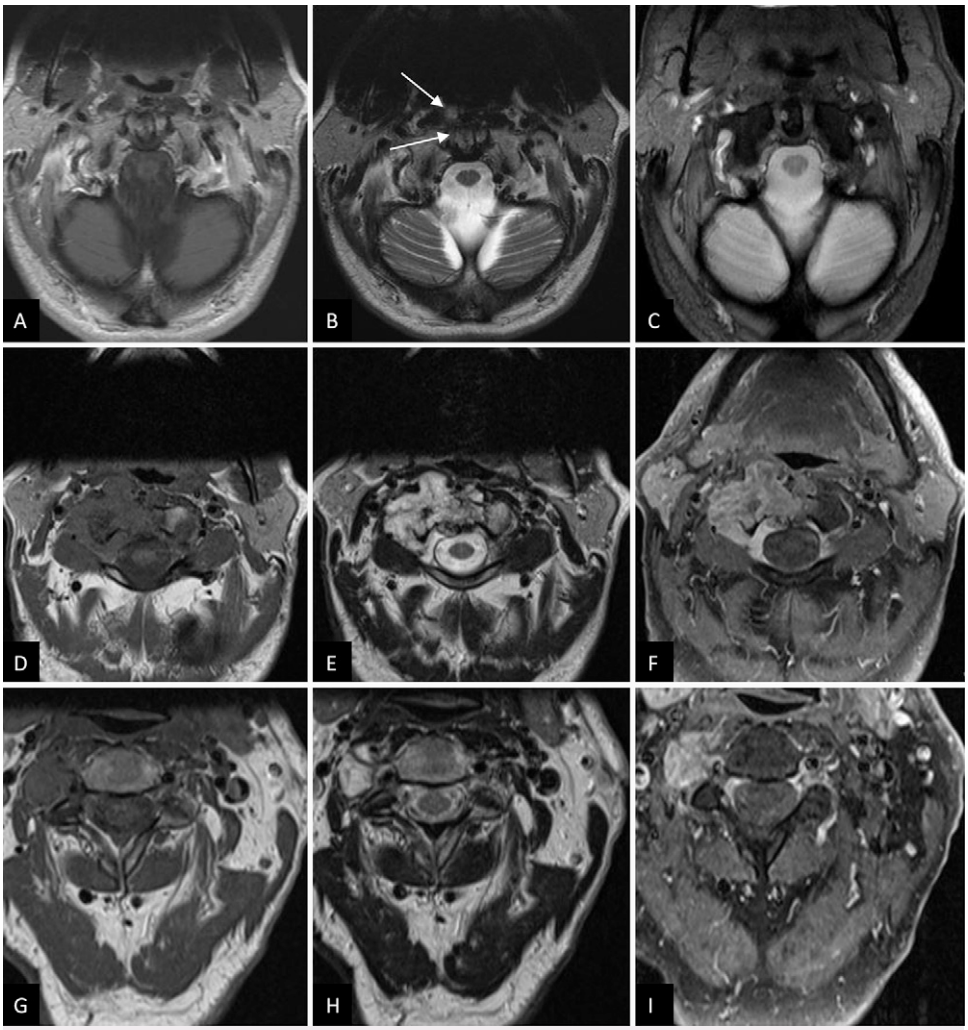

FIG. 2. Preoperative magnetic resonance imaging of the cervical spine including axial T1-weighted at C1 (**A**), axial T2-weighted at C1 (**B**) with *arrows* demonstrating tumor extension partially encasing the anterior arch, fat-suppressed post-gadolinium T1-weighted at C1 (**C**), axial T1-weighted at C2 (**D**), axial T2-weighted at C2 (**E**), fat-suppressed post-gadolinium T1-weighted at C2 (**F**), axial T1-weighted at C3 (**G**), axial T2-weighted at C3 (**H**), and fat-suppressed post-gadolinium T1-weighted at C3 (**I**).

plane was developed within the paraspinal muscles adjacent to the tumor, leaving a cuff of normal muscle attached to the mass. The right vertebral artery was then exposed and ligated at the superior aspect of the arch of C1. A complete laminectomy was performed at C1, and partial laminectomies were performed at C2 and C3 to avoid entering the tumor. Stealth navigation and the operating microscope were used to guide the completion of osteotomies through the lateral mass of C2 and the vertebral body of C3. The right C2 and C3 nerve roots were identified and transected. We then instrumented the spine posteriorly from occiput to C7, utilizing an occipital plate, 3.5-mm screws bilaterally at C4 through C7, and 3.5-mm cobalt chrome rods bilaterally. Rib allograft and fibular allograft struts were placed centrally between the 2 rods extending from the occiput to C4. Cancellous iliac crest autograft was then placed into each of the remaining facet joints, and an iliac crest strut graft was placed on the left side between the occiput and the C3–4 level just lateral to the rod. Postoperatively, the patient was transferred to the intensive care unit (ICU) in stable condition.

Two days later, the patient returned to the operating room for the second, anterior stage of his procedure. Following completion of a tracheostomy, our ear, nose, and throat (ENT) colleagues performed a mandibulotomy and transmandibular approach to the upper cervical spine. Dissection was carried through the floor of the mouth and along the lateral pharynx to expose the anterior cervical spine. The hypoglossal, vagus, and accessory nerves, as well as the internal carotid artery, were identified and retracted laterally. Despite this approach, significant difficulty was encountered in accessing the occipitocervical junction secondary to the posterior instrumentation that had been placed from the occiput to C7 during the first stage of surgery, which made it impossible to extend the neck to improve access. Despite the limited operating window, an osteotomy was performed at the left side of the anterior arch of C1, lateral to the attachment of the transverse ligament. The anterior soft tissues were dissected until they communicated with the previously performed osteotomies at C2 and C3. The anterior tubercle of the C4 transverse process was identified and removed to facilitate exposure of the right vertebral artery, which was then ligated

at the C4 level before it entered the tumor. A discectomy was performed at C3–4, and the posterior longitudinal ligament was excised at this level. A Caspar distraction screw was then placed into the vertebral body of C3 and utilized to lever the caudal portion of the specimen anteriorly. As the specimen was mobilized anteriorly and to the left side of the neck, the right occipitocervical joint opened. By distracting this joint via levering of the specimen, we were able to visualize the spinal cord posterior to the mass and safely transect the apical and alar ligaments. The specimen was then completely free of adherent tissues and able to be removed en bloc. Intraoperative pathologist review of the specimen demonstrated negative microscopic margins circumferentially.

An FVFG was harvested from the right leg and sized to span the gap in the anterior column from the clivus to C4. A notch was cut into the FVFG cranially to create a snug fit between the graft and the clivus. The FVFG was then docked caudally into the C4 vertebral body and secured in place with a plate and locking screws. A small dural tear was identified at the level of the clivus extending proximally, which was sealed with DuraGen (Integra) given that it was inaccessible for suture repair. The vasculature of the flap was anastomosed to the facial artery and vein of the right neck. The mandibulotomy was repaired with rigid fixation, and the mucosal defects were closed in a water-tight fashion. The patient was then placed into a halo vest and transferred to the ICU in stable condition.

Postoperative Hospital Course

Upon cessation of sedation on postoperative day 1, the patient was found to be disoriented, unable to follow commands, and moving all 4 extremities sluggishly. A CT scan of the head was performed and demonstrated new high-attenuation material within bilateral cerebellar hemispheres, consistent with remote cerebellar hemorrhage. Interval repeat CT scans of the head demonstrated no increase in the degree of hemorrhage, and the patient progressively recovered both gross and fine neurological function in his bilateral upper and lower extremities with normalization of his mentation over the first 72 hours postoperatively.

On postoperative day 5, the patient became febrile with a worsening leukocytosis and a global decline in his strength and mentation. His surgical incisions appeared benign. A CT scan of the neck with contrast was obtained, which demonstrated right-sided anterior and posterior fluid collections within the operative bed, concerning for possible seromas, abscesses, or cerebrospinal fluid (CSF) collections. These fluid collections were aspirated, and ultrasound-guided drains were placed within the fluid beds. Fluid from the drains returned positive for beta-2 transferrin and bacterial cultures grew multiple colonies of *Serratia marcescens*. Given the inaccessible nature of the dural tear that had been identified intraoperatively at the level of the clivus extending proximally, any attempt to address it surgically would have involved reopening of the patient's mandibulotomy and removal of the FVFG with the takedown of its vascular anastomosis. As such, the decision was made to remove the drains, oversew the drain sites in a water-tight fashion, and hope that the resultant pressure would be sufficient to seal the CSF leak. Additionally, the patient was placed on meropenem and vancomycin to control the deep *Serratia* infection. After initiation of antibiotics, the patient remained afebrile throughout the remainder of his postoperative course and experienced progressive improvement in his global strength and mentation back to his postoperative baseline over the subsequent 48–72 hours.

Throughout the patient's hospitalization, he demonstrated severe dysphagia, which was initially managed with nasogastric tube feeds until placement of a percutaneous endoscopic gastrostomy (PEG) tube on postoperative day 25. Following completion of that procedure, his tracheostomy was decannulated and allowed to close spontaneously. The patient was discharged home from the rehabilitation unit on postoperative day 32.

Post-Hospitalization Course

Following dismissal from the hospital, the patient's postoperative course proceeded uneventfully. A CT scan obtained at 3 months postoperatively demonstrated evidence of early osseous bridging between the FVFG and the clivus and C4 (Fig. 3). He continued to experience severe dysphagia, with a complete inability to tolerate food or liquid by mouth. After consultation with our infectious disease colleagues, all parties agreed that he would remain on lifelong suppressive antibiotics given the presence of *Serratia* in the postoperative bacterial cultures obtained from the fluid collections that

FIG. 3. Three-month postoperative CT scan of the cervical spine.

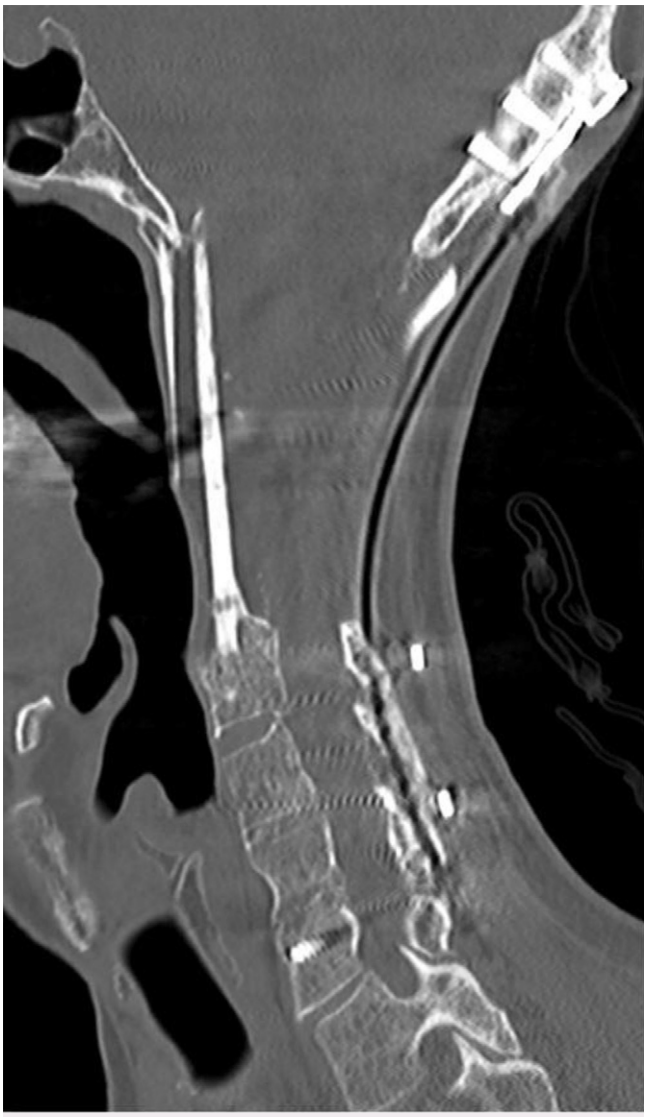

FIG. 4. Five-year postoperative CT scan of the cervical spine.

were in communication with the patient's spine hardware. Physical examination demonstrated intact strength in all key motor groups of bilateral upper and lower extremities.

At 5 months postoperatively, a CT scan of the cervical spine demonstrated complete fusion of the FVFG to the clivus and C4 with continued interval healing of the posterior strut grafts and no evidence of hardware failure. The patient's halo vest was removed, and he was instructed to begin isometric neck exercises. He was then treated with a course of proton beam radiation beginning at 6 months postoperatively.

Interval follow-up on an annual basis for the first 5 years and on a biannual basis thereafter demonstrated no evidence of tumor recurrence and solid circumferential arthrodesis on advanced imaging (Fig. 4). At 3.5 years postoperatively, follow-up endoscopic swallow evaluation demonstrated concern for thinning of the posterior pharyngeal wall overlying the plate at C4 that was utilized at the index surgery to affix the FVFG to the C4 vertebral body. He underwent an uneventful plate removal via a tracheostomy followed by a left transhyoid pharyngotomy. Repeat endoscopy performed 4 months after the removal of the plate demonstrated a significant improvement in the gross appearance of the posterior pharyngeal wall with no concern for impending rupture. His Neck Disability Index improved from 32% at 1 year postoperatively to 6% at 2 years postoperatively and 0% at 7 years postoperatively. The patient ultimately died 11 years postoperatively from an unrelated medical condition.

Discussion

Observations

Oncological Outcomes

The present case report is the first to report long-term oncological follow-up after en bloc excision of a high cervical chordoma, although multiple previous case reports and small case series have reported local tumor control at short- to midterm follow-up[30–35] (Table 1). Rhines et al.[35] performed en bloc resection of a multilevel cervical chordoma involving the C2–4 vertebrae and reported no evidence of local recurrence at 1 year postoperatively. Bailey et al.[30] found no evidence of local or distant tumor recurrence at 2 years postoperatively following en bloc spondylectomy of a chordoma involving C1–3. In their case series of 3 patients with high cervical chordomas, Chou et al.[31] reported no evidence of local recurrence in any of the patients at follow-up lengths of 12 months, 16 months, and 30 months. Similarly, Ozpinar et al.[34] found no evidence of local recurrence following en bloc resection of a high cervical chordoma at 26 months of follow-up. In a retrospective case series of 5 patients with cervical chordomas, Hsieh et al.[33] reported that both of the patients who underwent planned close-margin excisions due to the proximity of the tumor to the dura experienced local recurrence, whereas the 3 patients who underwent wide-margin excisions had no evidence of recurrence at final follow-up at a minimum of 4 years postoperatively. The 2 patients who experienced local recurrences did so at 25 months and 83 months postoperatively, which corresponds with previous studies identifying the propensity of chordomas to recur late.[40] Historically, high cervical chordomas were most commonly treated with gross-total resection with or without adjuvant radiotherapy; however, this approach is associated with higher rates of local recurrence than en bloc excision.[4,9–12,28,29,41] Surgeons should engage patients in a detailed preoperative discussion of the risks and benefits of each approach to determine how each approach aligns with patients' goals of care.

In the present case report, the patient underwent en bloc excision of a multilevel high cervical chordoma with negative margins, including at the location of the planned close margin where the tumor abutted the dura. At 10 years postoperatively, this patient remained disease-free, with no evidence of either local or distant tumor recurrence. This study is the first to report oncological outcomes at this extended interval and, as such, is the first report to demonstrate satisfactory long-term oncological outcomes following en bloc resection of a multilevel high cervical chordoma.

Additionally, this is only the second case report to describe the use of adjuvant proton beam radiotherapy following en bloc resection of a high cervical chordoma. Ozpinar et al.[34] reported no evidence of local disease at 26 months postoperatively following wide excision of a C1–2 chordoma followed by adjuvant radiotherapy. In the present study, the patient demonstrated no evidence of disease at 10 years postoperatively following en bloc resection of a C1–3 chordoma followed by proton beam radiotherapy. Although isolated

TABLE 1. Summary of reported cases of en bloc resection of multilevel high cervical chordomas

Authors and Year	Age (yrs), Sex	Involved Levels	Resection	Margin Status	Pst Instrumentation	Ant Reconstruction	Radiotherapy	FU (mos)*	Hardware Failures	Other Complications	Oncological Outcome (local recurrence, distant metastasis, cancer-specific mortality)
Rhines et al., 2005[35]	52, M	C2–4	Planned close margin (at dura)	–	Titanium rods coupled w/ occipital wiring, C5–T1 Wisconsin spinous process wiring, & T2–9 pedicle screw fixation.	Fibular allograft spanning from ant arch of C1 & remnant of dens (cranial) to superior endplate of C5 (caudal) & an ant cervical plate from C1 to C5.	None	12	Migration of fibular allograft & ant plate on POD 17 requiring revision to a titanium mesh cage.	Respiratory difficulty requiring prolonged ventilatory support. Pst pharyngeal wall dehiscence requiring reoperation for flap coverage.	No evidence of disease
Bailey et al., 2006[30]	50, M	C1–3	Planned close margin (at dura)	–	Occiput to C6 posterolat arthrodesis	2 Corticocancellous ilac strut autografts spanning from occipital condyle to C4 (rt) & from C1 lat mass to C3 (lt). The 2 struts were secured w/ screws at each end.	None	24	None	Immediately postoperative patient went into septic shock & ARDS, necessitating a 10-day ICU stay. Persistent dysphagia at 1 yr postoperatively; resolved at 2 yrs postoperatively.	No evidence of disease
Chou et al., 2009[31]	61, M	C2–4	Wide	NR	Occiput to T2 posterolat arthrodesis	Expandable cage spanning from C1 to C5 w/ ant cervical plate from C1 to C5.	None	12	None	Dysphagia Pneumonia	No evidence of disease
Hsieh et al., 2011[33]	32, M	C2–3	Wide	–	NR	NR	None	22	None	None reported	No evidence of disease
Guppy et al., 2013[32]	51, F	C2–3	Intralesional (planned close margin, but incidentally entered tumor)	+	Occiput to C6 posterolat arthrodesis w/ rib allograft.	Expandable cage spanning from C1 to C4 w/ ant plate.	Adjuvant proton beam therapy planned but deferred due to pharyngeal wall dehiscence. External beam radiation to sites of recurrence (6600 cGy using 200 cGy daily fractions).	20	None	Pst pharyngeal wall dehiscence that was not repaired due to identification of metastatic disease. Patient remained G-tube dependent at final FU w/ no plans to repair pharyngeal wall.	Local recurrence & supraclavicular lymph node metastases identified at 10 mos postoperatively.
Ozpinar et al., 2016[34]	48, F	C1–2	Wide	–	None	Expandable cage spanning from clivus to C3 & ant plate from clivus to C3. Sublaminar wires were used to secure cage to plate.	Adjuvant proton beam radiation	26	None	Pst pharyngeal wall full thickness defect due to erosion by the ant plate requiring reoperation 8 mos postoperatively for flap coverage. This defect recurred at 1 yr postoperatively, requiring radial forearm flap coverage.	No evidence of disease

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Authors and Year	Age (yrs), Sex	Involved Levels	Resection	Margin Status	Pst Instrumentation	Ant Reconstruction	Radiotherapy	FU (mos)*	Hardware Failures	Other Complications	Oncological Outcome (local recurrence, distant metastasis, cancer-specific mortality)
Present study	67, M	C1–3	Planned close margin (at dura)	–	Occiput to C7 posterolat arthrodesis w/ an occipital plate, lat mass screws at C4–7, & cobalt chrome rods. Rib, fibula, & iliac allograft struts.	Free vascularized fibular autograft spanning from clivus to C4 w/ ant plate securing caudal aspect of fibula to C4.	Adjuvant proton beam radiation	122	None	Remote cerebellar hemorrhage on POD 1. Deep serratia marcescens infection requiring lifelong antibiotic suppression. Inaccessible CSF leak that persisted postoperative managed by drain removal & watertight closure of drain sites. Severe dysphagia requiring lifelong PEG tube feeds. pst pharyngeal wall thinning requiring plate removal 3.5 yrs postoperatively.	No evidence of disease

ant = anterior; ARDS = acute respiratory distress syndrome; cGy = centigray; CSF = cerebrospinal fluid; FU = follow-up; G-tube = gastric tube; ICU = intensive care unit; lat = lateral; lt = left; NR = not reported; PEG = percutaneous endoscopic gastrostomy; POD = postoperative day; posterolat = posterolateral; pst = posterior; rt = right; + = positive; – = negative.
* Length of follow-up period.

proton beam radiotherapy is a viable option for inoperable mobile spine and sacral chordomas, en bloc resection with negative margins and adjuvant proton beam radiotherapy remains the gold standard to improve local and locoregional tumor control.[42,43]

Anterior Column Reconstruction

Previous reports of en bloc spondylectomies for high cervical chordomas have highlighted the inherent difficulty of and high complication rates associated with reconstruction of the anterior column following tumor resection.[32–35,44] Following resection of a high cervical chordoma involving C2–4, Rhines et al.[35] performed posterior instrumented fusion from the occiput to T9 and anterior column reconstruction with a fibular allograft spanning from the arch of C1 and the residual cranial-most portion of the dens to the superior endplate of C5, which was anchored in place by an anterior cervical plate spanning from C1 to C5. Unfortunately, on postoperative day 17, a CT scan demonstrated migration of the caudal end of the graft and anterior plate, and so the patient returned to the operating room for revision reconstruction with a titanium mesh cage. Final follow-up at 1 year postoperatively demonstrated no evidence of construct failure.

Bailey et al.[30] performed en bloc resection of a multilevel high cervical chordoma involving C1–3, followed by posterior instrumented fusion from occiput to C6 and anterior column reconstruction with 2 corticocancellous iliac crest autografts spanning the vertebrectomy site from the occipital condyle to C4 on the right and from the C1 lateral mass to the remaining portion of C3 on the left. The grafts were secured at each end with screws. The patient wore a halo vest for 3 months postoperatively. There was no evidence of hardware failure at final follow-up at 2 years postoperatively.

Hsieh et al.[33] reported pseudarthrosis and instrumentation failure in 2 of 3 patients who underwent instrumented anterior column reconstruction without adjunct autograft following multilevel high cervical chordoma resection, citing the inability of modern instrumentation to recapitulate the multidirectional forces present in the high cervical spine. In their case report of a patient who underwent C2–3 spondylectomies for resection of a high cervical chordoma, Guppy et al.[32] stabilized the cervical spine by performing posterior instrumented fusion from the occiput to C6 and reconstruction of the anterior column with an expandable cage from C1–4 with an anterior plate. At 10 months postoperatively, the patient was found to have a posterior pharyngeal wall defect that was not repaired due to concomitant identification of recurrence both locally and in the supraclavicular lymph nodes. At the final follow-up 20 months postoperatively, there was no evidence of hardware failure, but the patient remained feeding tube dependent due to the posterior pharyngeal wall defect. Ozpinar et al.[34] also reported hardware erosion through the posterior pharyngeal wall 8 months after anterior column reconstruction with an expandable cage spanning from the clivus to C3 and an anterior plate. Despite undergoing flap coverage of the pharyngeal wall defect, the patient developed a recurrent defect at 1 year postoperatively, ultimately requiring removal of the anterior plate. The authors reported radiographic evidence of solid arthrodesis at the final follow-up 26 months postoperatively.

The present case report is only the second to describe en bloc excision of a high cervical chordoma involving C1, and the first report to describe the utilization of an FVFG to span a multilevel upper cervical vertebrectomy defect and anchor proximally on the clivus. Utilization of a vascularized autograft allowed for relatively rapid healing in a high-strain, inhospitable environment for fusion, as evidenced by the presence of a solid arthrodesis on CT scans obtained 5 months postoperatively. Similar to other case reports in which anterior plates were utilized, the patient in the present study experienced erosion into the posterior pharyngeal wall of the plate used to secure the FVFG to C4 caudally. Fortunately, this was identified before the formation of a full-thickness pharyngeal wall defect, and the plate was removed uneventfully. At 10 years postoperatively, the patient's FVFG remained solidly fused without evidence of fracture or failure of his posterior instrumentation.

Complications

Of primary importance, the patient in the present case report demonstrated no evidence of tumor recurrence or failure of his complex cervical reconstruction at 10 years postoperatively. However, our patient did experience multiple complications that should inform future decision-making when performing en bloc resection of high cervical chordomas and the associated reconstruction.

Due to concerns that the posterior portion of the en bloc resection would result in potentially catastrophic upper cervical instability, we elected to perform an instrumented posterior cervical fusion during the first stage of surgery. Although a transmandibular approach was used to maximize exposure during the second, anterior stage of surgery, the inability to extend the patient's neck to access the occipitocervical junction made the anterior portion of the en bloc resection substantially more challenging, nearly to the point of rendering this dissection entirely impossible. After a painstakingly challenging dissection, the anterior portion of the resection was completed, and the tumor was removed en bloc. However, given the considerable difficulty encountered during the anterior portion of this case, consideration could be given to performing similar surgeries in the future in 3 stages, with the instrumented posterior fusion being reserved for the third stage of surgery after the tumor has been removed and the anterior reconstruction has been performed. A halo vest could be used in conjunction with strict spine precautions between stages. During each stage, the halo ring could be attached to an electro-mechanical head holder (Mizuho OSI) to control position with the greatest possible precision.

Postoperatively, our patient experienced severe and persistent dysphagia, rendering him PEG tube dependent for the remainder of his life. Although the etiology of his severe dysphagia was likely multifactorial, a primary driver of his dysphagia likely related to the final position in which his upper cervical spine was fused. In their study investigating the impact of O–C2 angle on the oropharyngeal cross-sectional area, Miyata et al.[45] determined that the patients with postoperative dysphagia following occipitocervical fusion all had a decrease in their O–C2 angle of greater than 10°, leading to a decrease in oropharyngeal cross-sectional area of 40%. Furthermore, no patients whose O–C2 angle increased from preoperative to postoperative radiographs developed dysphagia or dyspnea postoperatively. It is not possible to measure the O–C2 angle in our patient given the complete spondylectomy of C2. However, the O–C4 angle decreased from 33° preoperatively to 23° postoperatively, and the cross-sectional area of the oropharynx decreased from 361 mm^2 preoperatively to 268 mm^2 postoperatively (Supplemental Figure 1). Despite our best efforts to position the patient intraoperatively to mimic his preoperative O–C2 angle, these radiographic measurements suggest that perhaps he should have been fused in a more extended position to recreate his preoperative occipitocervical alignment. Other possible etiologies for his persistent dysphagia include extensive dissection, including mandibulotomy and C1

resection, and postoperative proton beam radiotherapy. Dysphagia was a reported complication in all but one of the currently available case reports on the resection of high cervical chordomas.[30–32,34,35]

Lessons

This case report describes the successful en bloc spondylectomy of a high cervical chordoma involving C1–3 followed by anterior column reconstruction with a free vascularized fibular graft. At 10 years postoperatively, this patient remained free of local or distant recurrence and had a solid arthrodesis without neurological deficit. En bloc resection in this anatomically complex region is fraught with complications, necessitating a thoughtful approach to preoperative planning and perioperative care of these patients.

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Author Contributions

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Supplemental Information

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