Single stage total endolesional C2 spondylectomy for chordoma

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Published online: 22 May 2013
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Keywords Chordoma · Total spondylectomy · Axis · C2

test. A novel approach to the reconstruction is also presented.

Learning targets

- Learn the surgical technique for a 360° C2 spondylectomy.
- Learn how to perform an upper cervical spine reconstruction.

Introduction

Chordomas are locally invasive, malignant bone tumors, which rarely occur in the cervical spine [5, 12]. En-bloc, or even marginal resection, offers improved survival and potential cure. Complete resection of tumors, involving the upper cervical vertebrae, requires combined anterior and posterior approaches and is complicated by the presence of vertebral arteries. A reconstruction of the post-resection defect is prone to failure.

We present a case of a chordoma, involving the axis that was treated by a single stage total endolesional C2 spondylectomy, with preservation of both vertebral arteries, as the patient did not tolerate preoperative occlusion

Case description

A 62-year-old man was seen on outpatient basis with a 3-month history of progressively worsening neck pain. He did not develop any neurological deficits. Plain radiographs pointed to an osteolytic lesion of the axis.

Subsequent CT and MRI scans confirmed a disease process, involving not only the body of the C2 vertebra, but also extending into the posterior elements via the left pedicle. Also a moderate epidural and extravertebral involvement, resulting in displacement of the left vertebral artery was confirmed.

There was evidence of a C2 vertebral compression deformity, but no spinal cord compression. The diagnosis of chordoma was confirmed via an open posterior microsurgical biopsy. The patient decided to proceed with the recommended C2 spondylectomy through a combined transoral and posterior approach.

The left vertebral artery displacement, found on diagnostic imaging, warranted a pre-operative occlusion test, as the artery would potentially need to be sacrificed. However, the occlusion test resulted in a transient confusion and hemiparesis, lasting for 48 h and the operation was delayed by 2 weeks. On the day of surgery, the patient was neurologically intact.

Electronic supplementary material The online version of this article (doi:10.1007/s00586-013-2813-6) contains supplementary material, which is available to authorized users.

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Surgical procedure

A combined transoral and posterior approach were planned. Both approaches consisted of two steps: (a) removal of vertebra with tumor and (b) reconstruction of the defect. The entire procedure was to be performed with continuous electrophysiological monitoring. A navigation system and fluoroscopy were also used.

The patient was positioned supine and tracheostomy was performed. The oral and nasal cavities, together with face and neck, were prepped in a standard fashion. The oral surgeon created a small right-sided submandibular channel through the floor of the mouth with an angle, suitable for the clival screws. The channel was kept open by a silicon tube for the duration of the procedure. The uvula was stitched to a vessel loop that was pulled out through the nose, and thus everting the uvula and soft palate cranially into the nasopharynx.

A self-retaining Crockard retractor system created adequate operative corridor from the inferior border of the C3 vertebral body to the clivus. This was verified with a lateral fluoroscopy. A midline incision in the posterior pharyngeal wall and subperiosteal dissection facilitated the access to the craniocervical junction.

The entire anterior arch of atlas was removed, the clivus was cleared of soft tissue and a C2/3 discectomy was performed. The dens was skeletized bilaterally and both C2 pedicles were exposed. The pedicles were then transected using an osteotome. Although an en-bloc resection is theoretically feasible and was indeed attempted, it proved to subject the spinal cord to unnecessary risk, due to the posterior and apical ligamentous attachments of the vertebra. The anterior portions of the vertebra and the tumor were therefore removed microsurgically step-by-step.

The defect was reconstructed using a 14-mm diameter Harms mesh cage filled with autologous, iliac crest bone graft, mixed with hydroxyapatite paste. The cage was fashioned in such a manner, that the cranial end was firmly anchored at the clival edge, and caudally on the C3 end-plate. The overhanging edges of the mesh were left anteriorly, to allow for screw fixation into the clivus and the C3 vertebral body (with 12 mm and 19 mm screws, respectively). The length of the clival screw was determined using the navigation system, and the appropriate angle was achieved through the prepared channel, created through the floor of the mouth. The pharyngeal wall was then closed in two layers and the nasogastric tube was inserted under direct vision.

The patient was then turned prone, taking extreme care to keep the head in a neutral position, and was fixed in a Mayfield head holder. A neutral head position is important for the planned occipitocervical fixation.

A standard midline posterior approach was performed for the posterior stage of the surgery. The exposure extended from the occiput to C6. A C2 laminectomy was performed and the adjacent facet joints identified. Both C2 nerve roots were tied off and cut. Attention was then turned to the left vertebral artery (VA), where the tumor extended outside the vertebral body. The artery was exposed at its entrance into the transverse foramen of the atlas, and just above the C3 superior articular process, part of which was to be removed. This facilitated both the proximal and the distal control of the VA. Unfortunately, the need to maintain tumor plane outside the vertebral body on the left, eventually required a complete removal of the superior articular process of C3. The dorsal aspect of the left C2 articular process was removed microsurgically to expose the VA, where it passed over the surface of the tumor. There were multiple arterial tumor feeders originating from the VA. These were coagulated and divided. The remainders of the bony elements were removed on both sides, thus completing a total C2 spondylectomy and a marginal resection of chordoma.

Reconstruction of the middle and posterior spinal columns was achieved by bilateral mesh cages (left diameter of 12 mm and right 10 mm), placed between the lateral mass of C1 and superior facet of C4 on the left, and C3 on the right. Cranially, both cages were fixed through prepared eyelets with C1 lateral mass screws, and formed part of the occipitocervical (OC) construct. On the right, a short lateral mass screw was placed in C3. Both cages were again filled with autograft and hydroxyapatite paste. The posterior construct was then finalized with OC fixation, extending down to C6 lateral masses. Autograft bone was then placed between the decorticated surfaces of the occiput and C3, and between the spinous processes below that level. A suction drain was placed subfascially, and the wound was closed in layers.

Electrophysiological monitoring remained stable throughout the procedure, all blood loss was replaced throughout and the whole operation took a total of 13 h. The patient's neck was stabilized in a Philadelphia collar postoperatively.

Postoperative information

The ventilatory support was removed on the second post-operative day and tracheostomy on day three. The patient was gradually mobilized and walked on post-operative day five. Initial parenteral nutrition was replaced by enteral feeds on day three and he gradually progressed to a regular diet by day 12. He was discharged home on day 17 with no neurological deficit, except for expected C2 distribution hypesthesia.

Discussion and conclusion

Recent advances in imaging, surgical and monitoring techniques have resulted in more radical spinal procedures being performed safely [2, 3, 8, 13, 14, 17, 18]. Lately, multiple reports exist on en-bloc resections of spinal tumors as well as on total vertebrectomies [7, 9, 19]. The goal of such radical procedures is not only to remove the tumor but also any potentially affected bone, and thus decrease the rate of recurrence [10]. However, the benefits of such radical procedures must outweigh its risks. An extensive radical resection should only be done, when longer chances of survival (more than 2 years) are expected [6, 15]. The en-bloc resection is indicated in cases where there is a concern for potential dissemination of the tumor cells in the surgical wound. In cases of malignant bone tumors, a total spondylectomy appears to be a more oncologic procedure with improved recurrence rates [1, 7, 16].

Upper cervical spondylectomies are complicated by a complex anatomy and the presence of vertebral arteries. Authors that had reported upper cervical spondylectomies, were usually forced to sacrifice the involved vertebral artery [4, 15] in order to achieve an en-bloc resection of the tumor and any potentially involved soft tissues.

This case demonstrates that vertebral artery sacrifice was only not feasible due to the failed occlusion test and the risk of stroke, but also not necessary as the VA was not directly involved in the tumor. The key to isolation of the VA is proximal and distal control. The distal dissection was straightforward, given a predictable vessel course over the atlas. The proximal control required removal of the superior articular process of the C3 facet. Given the natural history of chordomas, and the extent of C2 involvement in our patient, it was felt that a total C2 spondylectomy was indeed indicated. However, the tumor extended into the epidural space and also into the soft tissues, lateral to the C2 vertebra. Although the resection was radical, it can only be considered marginal (intralesional extracapsular) in oncologic terms.

Although the primary goal of the procedure is a radical resection of the chordoma, the reconstruction of the created defect poses its own challenges. Previous case reports demonstrate, that cervical constructs following spondylectomies are prone to failure [15]. A mesh cage secured to the C1 arch, or its remainder, anteriorly, probably does not provide an adequate biomechanical support of the construct. Furthermore, the preservation of the atlas arch anteriorly would make a safe removal of the odontoid tip impossible. The authors believe, that direct fixation to the clivus may represent a more robust alternative to previously described techniques [4, 15, 16].

Reconstruction of the middle column, using cages placed between the articular surfaces increases the chance

of bony fusion and also potentially offloads the strain on the posterior occipitocervical fixation. Multilocular support for the case of eventual tumor recurrence is the other reason for three-column reconstruction. All lateral mass screws were placed bicortically. In order to further increase the strength of the construct. A bony fusion was further reinforced by placing autograft bone between the occiput and each spinous process.

Although recent literature demonstrates that a wide, en-bloc resection of chordomas involving the spine, provides the patient with the best chance of disease-free survival [4, 15]. This and other previously reported cases [5, 6, 11] demonstrate, that this may not always be possible in the cervical spine. The authors believe, that as the experience with en-bloc resections for this rare condition widens, reconstructive techniques will be required, to match those improvements. This case report demonstrates another reconstructive option for the surgeon, who is challenged by cervical chordoma.

Conflict of interest None.

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