Aggressive, Multidisciplinary Staged Microsurgical Resection of a Giant Cervicomedullary Junction Chordoma

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Abstract
Chordomas of the cranial base are locally destructive tumors since they are surrounded by significant complex neurovascular structures. Thus, their surgical removal is challenging, recurrence rates are high, and their therapeutic strategies remain controversial. In this video, we present a 47-year-old man with a recent onset of swallowing difficulties, hoarseness, and weight loss for several weeks. In the neurological examination, he had complete paralysis of the 9th, 10th, 11th, and 12th cranial nerves. Magnetic resonance imaging (MRI) showed a heterogeneously enhancing expansile invasive mass lesion centered within the clivus and involving the C1, the occipitocervical junction, the retropharynx, and the hypoglossal canal. The decision was made to proceed with multiple staged surgeries. In the first surgical stage, we performed a mastoidectomy with the infralabyrinthine approach to perform a test clip ligation of the sigmoid sinus and to resect the tumor component that extended into the infralabyrinthine space. In the second stage, we performed a far-lateral transcondylar approach for tumor resection and occipitocervical fusion. In the third stage, we used a transoral approach with endoscopic assistance to complete the excision of the remaining tumor in the retropharyngeal space and anterior aspect of C1 and C2 bodies that were not accessible in the first two stages. The surgeries and postoperative course were uneventful. Postoperative MRI showed a gross total resection of the tumor. Histopathology indicated a chordoma. The patient subsequently received proton radiotherapy and has continued to do well without recurrence at 14 months’ follow-up.

Keywords
► cervicomedullary junction
► chordoma
► multidisciplinary
► skull base
► proton radiotherapy

The link to the video can be found at: https://youtu.be/uP9OSiKg_rE.

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Fig. 1 Preoperative MRI in (A) axial, (B) sagittal, and (C) coronal planes shows a heterogeneously enhancing expansile invasive mass lesion centered within the clivus and involving the C1, the occipitocervical junction, the retropharynx, and the hypoglossal canal and postoperative MRI in (A) axial, (B) sagittal, and (C) coronal planes shows a gross total resection of the tumor and the fat graft in the resection cavity without recurrence. MRI, magnetic resonance imaging.

Fig. 2 (A) The cadaveric dissections of each stage from our laboratory have demonstrated the extent of access and related anatomic and neurovascular structures via each stage. (B) The intraoperative picture shows the vertebral artery running in the sulcus arteriosus was exposed. (C) The intraoperative picture shows the vertebral artery was mobilized and transpositioned. (D) The intraoperative picture shows the dissection plane between the tumor and the vertebral artery. (E) The intraoperative picture shows the accessory nerve after opening of the dura. (F) The intraoperative picture shows the hypoglossal nerve which was encased by the tumor. (G) Intraoperative picture shows underneath the accessory nerve with more resecting of the tumor, we encountered posterior inferior cerebellar artery. JB, jugular bulb; PICA, posterior inferior cerebellar artery; SS, sigmoid sinus; VA, vertebral artery.