A Staged Strategy for Craniocervical Junction Chordoma with Combination of Endoscopic Endonasal Approach and Far Lateral Approach with Endoscopic Assistance: Case Report

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Abstract

Objective While the endoscopic endonasal approach (EEA) has gained widespread acceptance for the resection of clivus chordomas, conventional transcranial approaches still have a crucial role in craniocervical junction (CCJ) chordoma surgery. In repeat surgery, a carefully planned treatment strategy is needed. We present a surgical treatment plan combining an EEA and a far-lateral craniotomy with endoscopic assistance (EA) in the salvage surgery of a recurrent CCJ chordoma.

Case Presentation A 37-year-old woman who had undergone partial resection of a chordoma extending from the mid-clivus to the CCJ.

Technique A two-stage surgical intervention was planned. First, we opted for an EEA with the intention of removing only the extradural and medial compartments of the lesion. The rationale was to avoid intradural dissection of possibly adherent tissues from the previous procedures and to minimize the cerebrospinal fluid leak risk. One month after the first endonasal stage, a far lateral craniotomy was performed. After removal of the lateral mass and pedicle of C1, a large surgical corridor to the tumor was obtained. Tumor loculations disseminated in and around the CCJ and located in the areas blind to microscopic examination were then successfully resected with EA. An occipito-cervical fusion was then performed during the same procedure.

Conclusion In addition to the exact location and morphology of the tumor, history of previous surgery was an important factor in devising a treatment strategy in this case of clivus chordoma. EA was also found to be instrumental in improving the reach of the far lateral approach.

Keywords

- chordoma
- endoscopic endonasal approach
- far-lateral approach
- salvage surgery

Introduction

Chordomas, which arise from remnants of the notochord, have a higher tendency to express in two regions of the vertebral axis: the sacrococcygeal junction and the craniocervical junction (CCJ).^{1,2} Lesions at the CCJ often extend into the clivus, petrous bone, occipital condyle, and upper cervical vertebra (C1–C3)

with destruction of bony structures.^{1,2} In the case of CCJ tumors located in the midline, recent advances in endoscopic endonasal approaches (EEA) have greatly contributed to improve resection rates in this deep location. However, chordomas often extend laterally and involve the neurovascular structures, which may result in incomplete resection through a purely midline approach. Additionally, and even after “radical” resection, the

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risk of tumor relapse is high in this area.^{1,3,4} In recurrent cases, previously used surgical approaches have a significant impact on the following salvage treatment strategies.

In this report, we present a case treated with a staged treatment strategy with use of EEA for the extradural region, followed by a far-lateral craniotomy with endoscopic assistance (EA). A creative combination of both EEA and conventional craniotomy could provide more extensive and safer resection, even in salvage surgery.

Case Presentation

History and Examination

A 37-year-old woman was referred to our department with neck pain and lower cranial nerve dysfunction (hoarseness and dysphagia). She was previously diagnosed with a CCJ chordoma and two previous attempts at surgical resection had been performed in another hospital (one by retrosigmoid approach and one by an EEA). In the initial EEA, a mucosal flap with fascia lata was used to seal the wound and prevent cerebrospinal fluid (CSF) leakage. Histopathological examination of the surgical specimen led to a diagnosis of chordoma. Initial examination of the magnetic resonance imaging (MRI) at our institution revealed a tumor located in the lower clivus and extending to

the periodontoid region and the lateral masses of C1 and C2 on the right side involving the right vertebral artery. There was also tumor extension into the petrous apex and an intradural portion adherent to the brainstem (►Fig. 1). Computed tomography (CT) also showed extensive areas of bony infiltration by the tumor.

Treatment Strategy

Our treatment strategy was to resect the paramedian aspects of the tumor through an EEA in the first stage. There was a concern that the inferior pole of the tumor would be difficult to reach even with angled endoscopes and especially at the paraodontoid region (►Fig. 2). Based on the MRI, we also suspected that the chordoma infiltrated the pial planes and vessels of the posterior circulation. In addition, this patient had a history of surgical resection by a retrosigmoid approach, which was expected to have caused adhesions in the intradural space. Moreover, the right occipital condyle was completely replaced by tumor (►Fig. 1). Surgical stabilization and fusion of the CCJ was indicated. Taken together, these considerations suggested that the intradural part and lower aspect of tumor could be resected more safely during a second transcranial stage, more specifically through a far lateral approach (FLA) with EA (►Fig. 2).

Fig. 1 Preoperative imaging before endoscopic endonasal approach at our institute. This patient has had 2 previous transnasal resections and 1 craniotomy through a retrosigmoid approach at another institute. (A–D) MRI T2 WI showing a large chordoma extending in the prevertebral space and around the odontoid process. The tumor extended laterally to the hypoglossal canal and jugular foramen, and medially to the intradural space. The tumor extended from the middle clivus to the periodontoid region, at the level of C2.

Fig. 2 Treatment strategy. Paramedian part of the tumor could be resectable by an EEA. On the other hand, the periodontoid region (red circle in A and D), the intradural tumor extension with contacting the posterior circulation (blue circle in B and E), and adhesion from previous retrosigmoid approach (yellow circle in C and F), these parts seemed to be preferable for the second transcranial stage. (A–C) MRI T2 WI. (B–D) MRI T1 WI Gd enhanced.

First Stage: Endoscopic Endonasal Approach

Considering that the patient had a previous EEA and that a nasoseptal flap was used in the reconstruction, there was a concern that a clear dissection plane between the intradural and extradural spaces and through the adhesive scar would be difficult to obtain. A contralateral nasoseptal flap was thus harvested at the beginning of the case to ensure adequate closure (►Fig. 3). Intraoperatively, and as anticipated, the dura mater was found to be highly fibrotic. Tumor was infiltrating the dural planes and was resected along with the periosteal layer of the dura mater. A slight intraoperative CSF leak was observed. A multilayer closure was done with fat graft and the harvested nasoseptal flap (►Fig. 3).

After the first procedure, the patient did not present new neurological deficit, cranial nerve dysfunction, or CSF leakage. As expected, on the postoperative MRI, residual tumor was detected in the periodontoid region, lateral mass of C1, and intradural compartment, (►Fig. 4).

Second Stage: Transcranial Far Lateral Approach and Posterior Fixation

The second surgical stage was scheduled 1 month after the first EEA. This relatively long interval between the two surgical

stages was meant to let inflammatory healing of the fat graft and adequate intake of the nasoseptal flap occur. Otherwise, there would have been a risk of dislodging the fat graft and disrupting the nasoseptal flap during the following craniotomy, and cause inadvertent CSF leak. Regular endoscopic assessment confirmed adequate intake of the flap 3 weeks after the EEA. Therefore, 1 month after the first intervention, a right-sided retrosigmoid craniotomy with FLA was performed to approach the right occipital condyle, the ipsilateral vertebral artery, the periodontoid space and to gain access to the intradural extensions of the chordoma (►Fig. 5). After lateral subperiosteal dissection of the lamina of C1, the transverse process and lamina of C1 were removed with a high-speed drill. Next, the ipsilateral vertebral artery was translocated in a downward direction. The right occipital condyle was then completely drilled as it was found to be infiltrated by tumor. Intradurally, there were multiple sites where chordoma loculations were visible on the MRI. One of these was located in contact with the brainstem and was resected with a meticulous microsurgical technique. Other tumor compartments were removed with and without EA. For EA, a rigid 4-mm-diameter endoscope of 18 cm in length and a 30-degree lens (Karl Storz, Tuttlingen, Germany) was used. A rotative and malleable

Fig. 3 Intraoperative photographs with a 30-degree endoscope. (A and B) A left middle turbinectomy is followed by harvest of a nasoseptal flap. The flap was stored within the left maxillary sinus. (C) The right vidian nerve was sacrificed and the sphenopalatine ganglion was transposed to widen the surgical corridor. (D) The posterior nasopharyngeal wall was opened to approach the periodontoid region. (E) After drilling the clivus, the tumor seated in the epidural space was resected. (F) After exposure of the anterior and medial aspects of the ICA, the tumor located between the posterior aspect of the ICA and clivus was resected with angled instruments. (G) Only the extradural compartment of the tumor was resected to avoid the CSF leak and intradural dissection of fibrotic planes. The fascia lata used in the previous EEA was detected and was kept intact. (H) Closure was done with abdominal fat graft and a tissue-glue-coated collagen sponge (TachoSil, Nycomed Linz, Austria) supplemented with fibrin glue. (I) The nasoseptal flap was positioned atop the reconstruction and was held in place with a silicon sheet. ICA, internal carotid artery; MS, maxillary sinus; SPG, sphenopalatine ganglion; VN, vidian nerve.

suction was used to aspirate the lesion. EA was useful for the resection of tumor extensions located in microscopic “blind spots,” especially in the periodontoid region and the middle clivus region where we could obtain a wide surgical view through the corridor between the vertebral arteries inferiorly and the hypoglossal nerve superiorly (► Fig. 5).

After tumor resection, posterior fixation of C0–C1–C2 was performed. The pathological examinations in both stages confirmed the previous diagnosis of chordoma, with a Ki-67 index of 10%. The postoperative course was uneventful, and the neurological symptoms were stable. A few months after surgery, the patients complained of ear pain associated with a middle ear effusion that subsided after a right tympanostomy tube was implanted. On the postoperative MRI, subtotal resection of the tumor was demonstrated with only a small chordoma remnant in the contra-lateral C1 pedicle (► Fig. 6). Adjuvant photon radiation therapy (total of 70.2 Gy) was done in the following months. There has been no sign of tumor progression in the 2 years of follow-up since surgery.

Discussion

Anatomical Limitations of the Endonasal Approach

To achieve satisfactory resection of CCJ chordomas, several surgical approaches have been described in the literature, including EEA,^{4–6} transoral approach,⁷ and transcranial approaches.⁸ The precise localization of this slow growing tumor and all of its extensions has to be carefully considered when choosing the surgical approach to ensure effective and safe tumor resection. In the lower clivus, the anatomical landmarks defining the lateral limitations of the endonasal route are the hypoglossal canal, eustachian tube, and internal carotid artery.^{5,9–11} In addition, the hard palate also determines the reach of the EEA inferiorly.^{5,9–11} The more inferior the direction, the less surgical space there is, especially laterally. In fact, in subtotal resection of lesions centered on the CCJ through an EEA, tumor residue is often found in the lateral field, even with the use of angled endoscopes.^{5,9–12} As a practical rule of thumb, when entertaining

Fig. 4 Postoperative imaging after endoscopic endonasal tumor resection (first stage). (A–D) MRI T2 WI showing adequate decompression of the brainstem; residual tumor is identified intradurally and in the periodontoid region (green arrows).

either an EEA or a FLA for a CCJ tumor, it is often useful to determine if there is lateral extension of the tumor beyond the hypoglossal canal on preoperative imaging.¹⁰

Differences in the View between Far Lateral Approach and EEA

A recent cadaver study by the University of Pittsburgh group reported that the surgical view obtained through an EEA was significantly larger than the one from the transcondylar far lateral approach at the ventromedial region of the brainstem.¹⁰ On the other hand, they also reported that the surgical corridor in the far lateral approach was significantly larger than with an EEA.¹⁰ Obviously, each of these corridors has its own sets of advantages, limitations, and possible complications. FLA can require manipulation of the vertebral artery and more extensive condylar bone removal which may result in CCJ instability.⁸ In the literature, there were some reports of partial drilling of the condyle with the intention of preserving the stability of the condyle;^{13,14} however, for this aggressive tumor that often invades the surrounding bone, radical resection and extensive bony removal are preferable to reduce the risk of recurrence.¹⁵ Through an EEA, the main risk is the development of CSF leak and also, to a lesser degree, of injury to the internal carotid artery.

Advantages of Endoscope-Assisted Craniotomy

Although a nasal flap contributes in decreasing the rates of CSF leakage in repeat surgery,^{4,5,11} finding the anatomical landmarks and dissecting along the proper plane is more difficult than in virgin cases. In addition, in this case, the dura mater was opened during the previous endonasal procedure. The risk of postoperative CSF leak and finding dense arachnoid adhesions were felt to be high. Only the extradural and medial parts of the tumor were thus resected during the endoscopic endonasal stage and a FLA with EA was subsequently performed. Through the retrosigmoid and FLA corridors, the intradural chordoma implants were meticulously resected under the microscope with no neurological morbidity and a reduced risk of CSF leak. The advantages of the endoscope are to expand the surgical field and to allow for visualization in the microscopic blind spots as shown in our case (with use of endoscope, the tumor located anterior, and medial to the brain stem was successfully resected (–Fig. 5)).¹⁶

Other Combined Strategies for the Skull Base

There are several reports in the literature of combined approaches to different areas of the skull base using multiple surgical stages. Some groups have initially advocated the combination of endoscopic endonasal and transoral approaches.¹⁷

Fig. 5 Intraoperative photographs during the far-lateral approach with endoscopic assistance. (A and B) A right side retrosigmoid craniotomy is performed, with exposure of the C1 and C2 laminas. The right vertebral artery was dissected and transposed. (C) After drilling of the occipital condyle and lateral mass of C1 endoscopic assistance was used to deepen the surgical access. (D) Endoscopic view. (E) After inserting the endoscope through a corridor between the hypoglossal nerve and the vertebral artery (asterisk), a wide view of periodontoid region was obtained and tumor located in this area was resected. (F) Through a corridor crossing the hypoglossal nerve, abdominal fat that was placed in the previous EEA stage was found. G-I: MRI T2 WI showing the region where endoscopic assistance (EA) was performed. Tumors at Preodontoid region (red circle in G), cranio-cervical junction (red circle in H), and middle clivus (red circle in I) were successfully resected with help of EA. (J-L) After opening the dura matter of the posterior fossa, the jugular tubercle was drilled to obtain a larger surgical corridor. Tumor around the jugular foramen was resected through a corridor between the vertebral artery and lower cranial nerves. C1C, C1 condyle; C1LP, C1 lateral process; JF, jugular foramen; OC, occipital condyle; VA, vertebral artery.

The larger operating corridor obtained by combining both these approaches can in fact resolve the issue of the inferior limitation defined by the hard palate. However, one of the main advantages of the endonasal route is that it avoids the relatively high morbidity of transoral approaches. In fact, the mucosal incision is located higher in an EEA that in transoral approaches, which potentially limits the infectious risk, quickens extubation, and the reintroduction of oral feeding.¹⁸

In addition, there has been an interest for robotic surgery though transoral and transnasal routes, but these are still in the early development phase.¹⁹ While there are still points to improve in these strategies (intraoperative positioning, limited vision through the transoral route, steep learning curve, cost-effectiveness, etc.), these strategies may represent the future of the surgical treatment of lesions seated at the CCJ and nasopharynx.

Fig. 6 Postoperative imaging after the second-stage craniotomy by a far-lateral approach. (A–D) MRI T2 WI showing a gross total resection of chordoma throughout staged surgery. A slight remnant tumor was detected at the periodontoid region (red circle in A). The patient received subsequently a proton therapy. X-Ray (E) and CT (F) images showing the posterior fixation performed at the same procedure.

Conclusion

For chordomas of the lower clivus, EEA has been widely accepted as a treatment option. However, this tumor has the tendency to recur. In cases of lateral extension beyond the hypoglossal canal, where the endonasal route is limited, or in cases of recurrence in which safe margins of resection are difficult to obtain only through an EEA, a combined surgical strategy with staged EEA and FLA may be a valuable option. EA may also be used to improve the extent of resection through the FLA.

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