Combined Transnasal and Transcranial Removal of a Giant Clival Chordoma

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Abstract

Clival chordomas confront the surgeon with the task of resecting an aggressively invasive and destructive tumor in a critical surrounding. For many, mainly smaller, chordomas, the transnasal transclival approach is a feasible and safe surgical access. Larger tumors and especially those with extensive intradural, retrochiasmal, and/or deep cervical expansion are mostly approached by open craniotomy. Staged procedures are also commonly used in the case of expansive tumor growth. We present the first case of a single-session combined transnasal and transcranial approach to radically resect a large clival chordoma.

Keywords
- endoscopy
- chordoma
- skull base
- approach

Introduction

Arising from the embryonic rests of the notochordal, clival chordomas are slow-growing yet aggressively invasive and destructive tumors. They show a strong tendency for local recurrence even after combined surgical and radiosurgical treatment. The possibility of spreading to distant locations of the neuraxis may further complicate the treatment and causes additional morbidity.

The transnasal and transclival approach is for many chordomas a feasible and safe surgical access.[1–3] Larger tumors, especially those with extensive intradural retrochiasmal and/or deep cervical expansion, are most often resected by open craniotomy. A large number of transcranial approaches have been described in the last decade.[4–6]

We present a case of a single-session combined endoscope-assisted, transnasal, and transcranial retrosigmoidal approach for radical resection of a giant clival chordoma.

Case Report

A 30-year-old man presented with a history of recurrent clival chordoma. Upon diagnosis, a transnasal biopsy and later on a subtotal resection of the chordoma through a transnasosphenoidal route were performed. Due to a cerebrospinal fluid (CSF) leak, revision surgery was necessary. In the later course of the disease the patient required a ventriculoperitoneal shunt placement and 2 months later a revision of the shunt. All these procedures had been performed in an external clinic.

Three years later, the tumor showed marked growth with clinical and radiologic signs of progressive pontomedullary compression (►**Fig. 1**). As key symptoms the patient presented a progressive gait ataxia and difficulty swallowing.

Because the tumor extended perimedullary beyond the petrous apices into the posterior skull base on both sides, we planned a primary transnasal approach with the option of an additional transcranial access in a single session.

The patient was put in a supine position with the head held in a Mayfield clamp in a way that a transnasal as well as a transcranial approach was possible (►**Fig. 2**).

A preoperative computed tomography (CT) scan was performed, and neuronavigation was registered (BrainSuite, Brainlab, Feldkirchen, Germany; Siemens, Munich, Germany).

Neurophysiologic monitoring was used for complex brainstem and cranial nerve monitoring (facial nerve,

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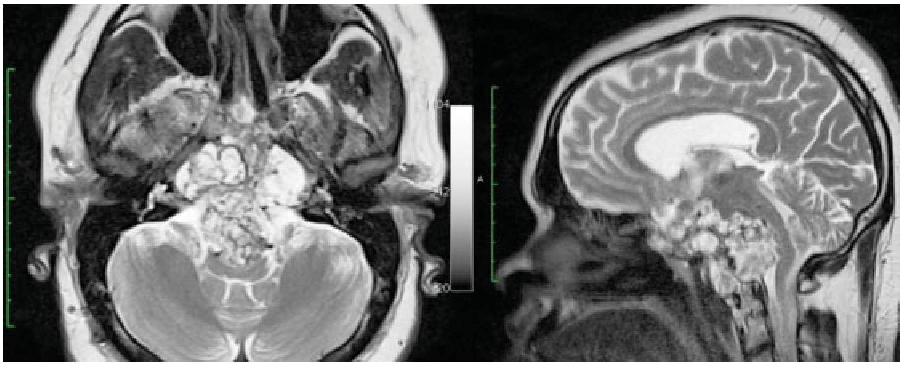

Fig. 1 Preoperative T2-weighted magnetic resonance imaging. Axial (left) and sagittal (right) sectioning.

glossopharyngeal nerve, hypoglossal nerve, somatosensory evoked potential, and motor evoked potential, all bilateral).

Part 1: Transnasal Endoscopic Approach

An extended transnasal endoscopic access was performed according to a bilateral sphenoethmoidectomy with harvesting of a vascularized septomucosal flap for later skull base reconstruction. In such a way a broad access to the central skull base was gained under endoscopic view, thus allowing removal of the tumor in a four-hand technique with one surgeon holding the endoscope and a sucker while the second surgeon was able to use two instruments. The clivus was reduced by the use of a high-speed drill and Kerrison punches. Tumor removal was initially mainly performed using curettes and an ultrasound aspirator. In the area of both petrous apices and the cavernous sinus, the tumor revealed a softer consistency, and both carotids in their petrous part were freed of the adhering tumor. However, toward the central skull base the tumor became firmer with adhesions to intracranial vessels. Further tumor removal through the transnasal access at that point was associated with unacceptable risks, and the decision was made to perform an additional transcranial exposure.

Part 2: Transcranial Retrosigmoid Approach

An intraoperative CT scan was performed to confirm the extent of the residual tumor volume and for the planning of further transcranial resection. The head was slightly rotated without opening the Mayfield fixation. After anatomical and neuronavigational orientation, a retrosigmoid craniotomy was performed on the left side. CSF was drained from the inferior cerebellomedullary cistern to relieve pressure within the posterior fossa. The tumor was entered between the vestibulocochlear and the glossopharyngeal nerve. Due to the firm texture of the tumor, enucleating was done mostly through sharp dissection and resection. The vertebral artery was followed toward the vertebrobasilar junction and freed of firmly adhering tumor tissue. Now the caudal aspects of the tumor were addressed and detached from the clival dura. First the left, then the right abducens nerve as well as the contralateral caudal cranial nerves, facial, and vestibulocochlear nerves were identified (►**Fig. 3**).

Part 3: Combined Transnasal and Transcranial Approach

Surgery was continued in a combined transnasal-transcranial way (►**Fig. 4**). Transnasal tumor resection was continued with direct visual control and guidance through the retrosigmoidal approach. This was also performed vice versa with resection through the transcranial approach and endoscopic transnasal control (►**Fig. 5**). After radical tumor resection, the large dural defect was covered transcranially with a flap of fascia lata. Abdominal fat was placed between the flap and the brainstem to improve the conditions of subsequent proton

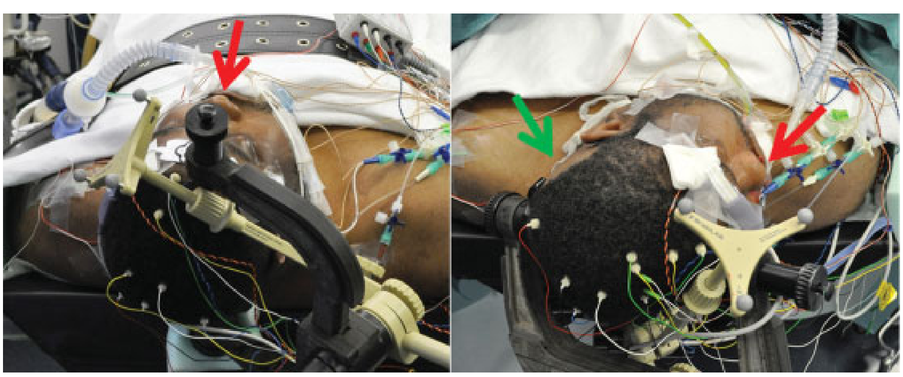

Fig. 2 Position of the patient using radiolucent head clamp for navigation-guided transnasal (red arrow) resection and after repositioning for combined transcranial (green arrow) and transnasal (red arrow) exposure.

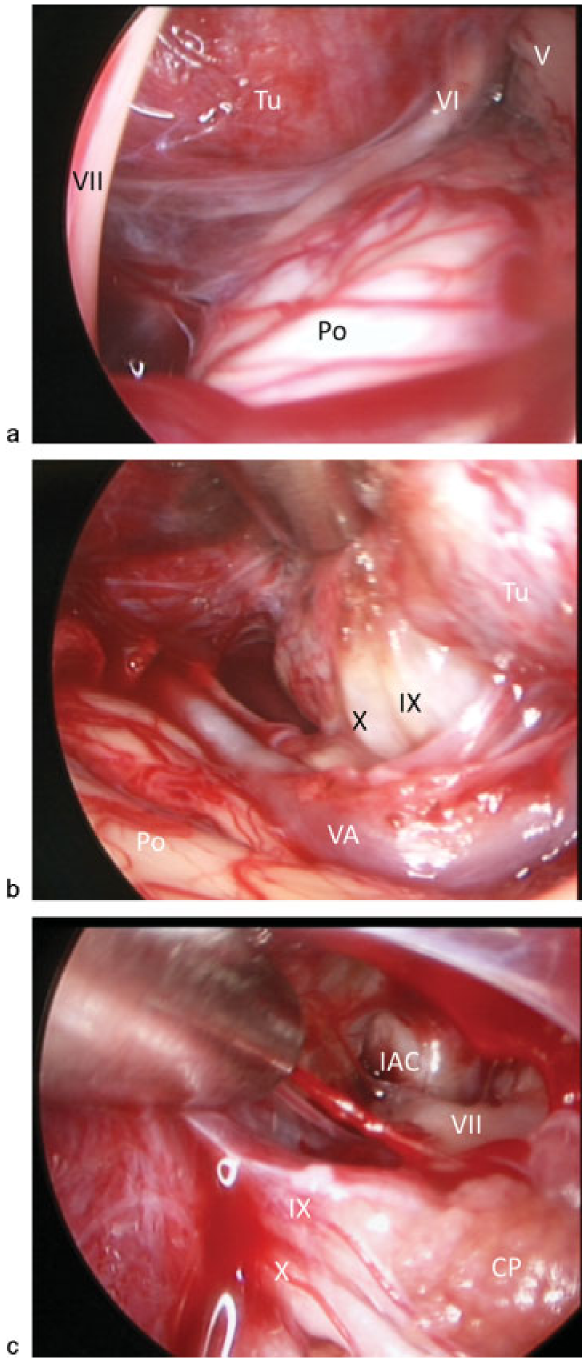

Fig. 3 (A) Endoscopic view of the giant chordoma via retrosigmoidal approach. Note the upper part of the tumor (Tu) with displacement of the trigeminal (V), abducent (VI), and facial (VII) nerves and compression of the pons (Po). (B) Lower part of the tumor (Tu) with marked adhesions to the vertebrobasilar junction and caudal cranial nerves (IX and X). VA, vertebral artery; Po, pons. (C) After further dissection, the contralateral cerebellopontine angle becomes visible. Note the opposite choroid plexus (CP), glossopharyngeal (IX), and vagal (X) nerves, and the facial nerve (VII) entering the internal auditory canal (IAC).

beam therapy. Now the covering of the defect was completed through the endonasal access in a multilayer technique by means of fat, fascia, and the free mucosal flap. Due to the extent of the defect, endonasal tamponades were applied. After the reconstruction of the dural defect, a second intraoperative computed tomography was performed to demonstrate the extent of resection and rule out major complications. Finally the retrosigmoidal approach was closed through direct sutures of the dura and osteosynthetic fixation of the bone fragment. Operative time was $\sim$ 17 hours; blood loss was 700 mL.

Postoperative Course

Postoperative magnetic resonance imaging revealed only small tumor remnants in the right cavernous sinus and a circumscribed pontine ischemia on the right side (►**Fig. 6**). Accordingly, the patient presented a transient right-sided hemiparesis with quick restitution to normal function. The ophthalmoparesis of the right eye remained unchanged. Otherwise the postoperative course was uneventful, and there was no rhinoliquorrhea. Later on, the patient underwent proton beam therapy and did not show tumor progression at follow-up after 24 months.

Discussion

The initial clinical presentation of clival chordomas is highly variable. Double vision due to partial ophthalmoparesis is the most common presenting symptom; decreased visual acuity, signs of endocrine dysfunction, hearing loss, facial numbness, difficulty swallowing, or even motor weakness may also be present.[5]

The mainstay of the current treatment is surgical gross total removal or tumor debulking before additional stereotactic radiotherapy or radiosurgery. In recent years, proton beam therapy has increasingly found an application in the adjuvant therapy of clival chordomas.[7,8]

Over the last decades there has been a shift toward the use of endonasal endoscopic approaches because they offer a direct access to these midline lesions. Angled endoscopes have broadened the operative region without the need for an extension of the surgical corridor.[9] Tools for neuronavigation have additionally facilitated the use of endoscopic techniques.

Tumor extension lateral to the internal carotid arteries (ICAs) is considered by many authors to be the most important limiting factor in achieving a higher degree of resection when endonasal endoscopic approaches are applied. Accordingly, a higher rate of radical resection may be achieved through open transcranial approaches in tumors $>$ 4 cm in diameter or 80 cm^3.[9]

Also, the extension of the tumor downward as far as the upper cervical levels may require the application of rigid instrumentation as Chau et al describe in their case of a combined endoscopic endonasal and posterior cervical approach to a clival chordoma in a staged procedure.[10] However, in their case the total operative time for both procedures is described as 29 hours. This, as well as the necessity of repositioning the patient into a prone position, renders the combined use of a transnasal endoscopic and an open posterior approach impractical. In our case, however, the tumor did not extend as caudally as the craniocervical junction, thus being approachable in its lower parts through a retrosigmoid craniotomy and not requiring additional spondylodesis.

Fig. 4 Combined transcranial and transnasal endoscopic surgery in a "real" rhinosurgical (RS) and neurosurgical (NS) cooperation.

a
b

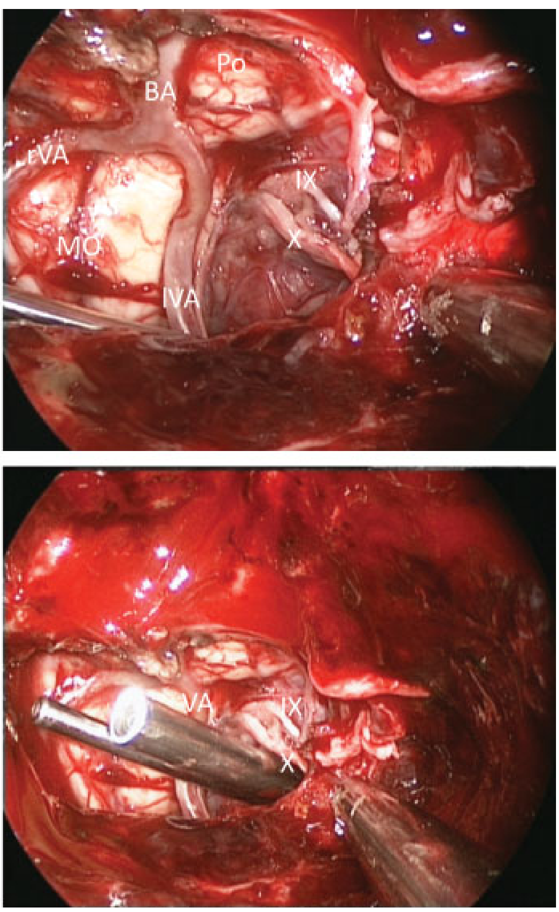

Fig. 5 (A) Transnasal endoscopic view after combined resection of the giant chordoma. Note left (lVA) and right (rVA) vertebral arteries, basilar artery (BA), medulla oblongata (MO), pons (Po), and left caudal cranial nerves (IX and X). (B) Endoscopic transnasal view of the cooperative resection: left vertebral artery (VA), glossopharyngeal (IX) and vagal nerves (X).

In regard to the long duration of this procedure with its inherent risk of surgeon fatigue and wound infection, it might also be considered to stage the operation, hence performing the endoscopic part in a first step and the transcranial part in a second step a few days later. The clear advantage of the biportal transnasal and transcranial exposure, however, is the possibility to safely dissect in hidden corners of the field under direct visual control from an additional angle. In cases, where a lesion shows such an extension that a removal might not be achieved through one approach, we therefore recommend preparing the patient in a way that a combined exposure is possible and to begin the procedure with the approach that is expected to enable most of the resection. At the point where no further tumor resection can safely be accomplished, the surgical team should then decide whether an additional exposure is to be performed right away or at a second stage later on.

One of the main challenges in transnasal endoscopic chordoma surgery remains the watertight closure of the dura.[1] In the largest series reported so far by Stippler et al, CSF leaks occurred in 25% of patients.[11] The extent of intradural tumor involvement is probably a major risk factor in developing a CSF leak. However, the possibility to address large defects following tumor removal through a combined transcranial and transnasal multilayered covering might improve the chances of achieving a watertight repair. In our hands a lumbar drain is not used routinely. A patient complaining of nasal CSF discharge postoperatively is closely monitored for 24 to 48 hours. If the CSF rhinorrhea does not resolve spontaneously, revision surgery is performed.

A cooperative rhino- neurosurgical effort is advocated by other exponents in the field of endoscopic transnasal skull base surgery.[9] Also, at our institution, all procedures involving a transnasal access to the central skull base are performed by a rhinosurgeon and a neurosurgeon in a cooperative manner.

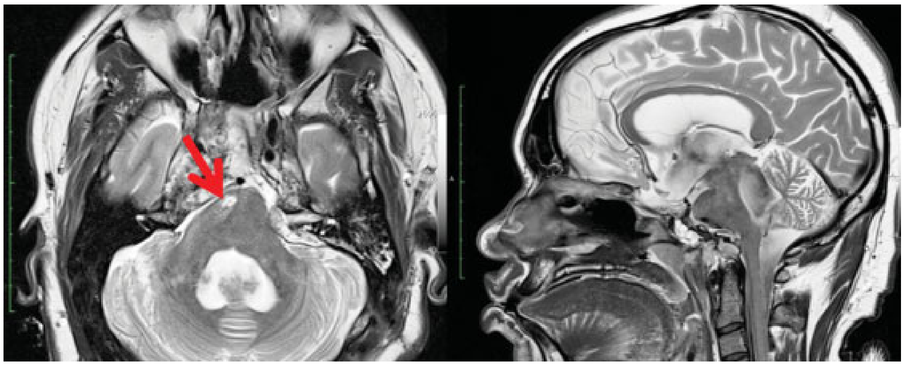

Fig. 6 Postoperative T2-weighted magnetic resonance imaging. Axial (left) and sagittal (right) sectioning; ischemic lesion (arrow).

Conclusion

In highly selected cases of clival chordoma and other lesions of the central skull base, the use of a combined open microsurgical and an endonasal endoscopic approach might increase the radicality of tumor resection while decreasing the operative and anesthesiological burden for the patient. In such complex cases, a cooperative rhino- and neurosurgical effort is recommended.

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