Endoscopic transsphenoidal resection of a large chordoma in the posterior fossa

Case report

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The authors report their encouraging experience using an endoscopic technique for transsphenoidal surgery in a patient with a large chordoma in the posterior fossa. The patient was a 40-year-old man with a 2-year history of progressive ataxia, a memory disorder, and emotional instability. A magnetic resonance (MR) image of the brain revealed a midline posterior fossa mass measuring 4 cm in diameter located between the clivus and the brainstem. The basilar artery and its bifurcation were encased by the tumor, which also distorted the brainstem. The patient had been treated at another hospital for obstructive hydrocephalus with a ventriculoperitoneal shunt and he received fractionated external-beam radiation treatment, although no histological diagnosis was ever made. The authors achieved a subtotal resection of the tumor through the patient's nostril using an endoscopic transsphenoidal technique. The portion of the tumor located behind the basilar artery was not resected to protect the brainstem perforating arteries. The patient showed dramatic improvement of his symptoms postoperatively. Residual tumor located behind the basilar artery was treated by stereotactic gamma knife surgery. This is the first reported case of a large posterior fossa chordoma treated by an endoscopic transsphenoidal technique.

Key Words * chordoma * endoscopy * pituitary neoplasm * skull base surgery * sphenoid sinus * transsphenoidal approach

Otolaryngologists have adopted endoscopic techniques to replace conventional sinus surgery for the surgical treatment of most inflammatory sinonasal disorders.[8] We have utilized an endoscopic technique for transsphenoidal resection of pituitary tumors in more than 50 patients.[3] We recently encountered a patient with a posterior fossa tumor based at the clivus; we were convinced that the endoscopic transsphenoidal technique was applicable. The following report involves a case of a large chordoma that was successfully treated using our endoscopic transsphenoidal technique.

CASE REPORT

This 40-year-old man was referred to The University of Pittsburgh Medical Center with a 2-year history of progressive ataxia, emotional instability, short-term memory disorder, and story fabrication.
History. The patient first noticed a balance disorder after drinking a single can of beer and then noticed gradual difficulty with his balance even without consuming alcohol. He had also suffered from uncontrollable emotional outbursts, ranging from pathological laughter to crying and was aware that his labile emotion was not normal. His family indicated that he fabricated stories and also displayed incomplete short-term memory. Eight months prior to his admission to the University of Pittsburgh Medical Center he developed right hemiplegia. A magnetic resonance (MR) image of the brain, obtained at another institution, showed a posterior fossa tumor located between the clivus and the brainstem with obstructive hydrocephalus. He was treated for hydrocephalus by placement of a right ventriculoperitoneal shunt and received fractionated external-beam radiation treatment for his tumor, although no histological diagnosis was obtained. His right hemiplegia had improved after placement of the right ventriculoperitoneal shunt. Despite the subsequent radiation treatment, he continued to deteriorate and his symptoms of ataxia, pathological emotional displays, and difficulty with his short-term memory increased.

Examination. On neurological examination, he had ataxia with a wide-based gait, hyperreflexia in his right upper and lower extremities with presence of a right Babinski sign, and emotional instability. While being examined, he burst out in laughter followed by instantaneous crying and stated that he could not control those emotional outbursts. Despite the short-term memory disorder and story fabrication that his family had noted, his memory examination was unremarkable.

An MR image of the brain revealed a tumor measuring 4 cm in diameter in the midline posterior fossa between the clivus and the brainstem (Fig. 1). The tumor was deeply indenting the midbrain and upper pons and had encased the basilar artery and its distal branches. The patient had an amply sized sphenoid sinus.

Surgical Decision Making. The patient was originally referred to us as a candidate for gamma knife surgery, but that treatment was determined to be inappropriate for his tumor. Radical resection of the tumor was recommended; however, the patient insisted that he wanted a minimally invasive method of treatment. Based on our previous experiences in endoscopic transsphenoidal pituitary surgery, we discussed the possibility of surgical resection of the tumor by means of an endoscopic transsphenoidal approach. The patient agreed to undergo endoscopic resection of the tumor followed by stereotactic gamma knife surgery for any residual tumor. The patient and his family were made well aware that this surgical technique is considered unconventional.
OPERATIVE TECHNIQUE

The details of our endoscopic transsphenoidal technique for pituitary tumors has been described elsewhere.[3] The patient was anesthetized by endotracheal intubation and maintained supine, with 10° elevation of the torso and the head tilted 10° to the left. The head was fixed in this position using a three-pin head fixation system. His face and nasal cavity were prepared with an antiseptic solution, and the periumbilical abdomen was prepared in a similar fashion to the donor site for a free fat graft. The patient, C-arm fluoroscope, and endoscope/video camera equipment were then draped following aseptic techniques. We used 4-mm rigid endoscopes with 0°, 30°, and 70° angled lenses. A lens cleansing irrigation-suction system was attached to the endoscope. The endoscope was also connected to a closed circuit video system that provided a continuously monitored view and allowed video filming of the surgery. An endoscope-holding device was mounted to the head fixation system.

We approached the patient’s tumor through the right nostril. The nasal mucosa was decongested using cottonoid patties moistened with a decongestant solution. Under 0° endoscopic visualization, the mucosa over the rostrum of the sphenoid, middle turbinate, and posterior septum was infiltrated with a solution of 1% lidocaine with epinephrine diluted at 1/100,000. The middle turbinate was gently pushed laterally to access the sphenoid sinus ostium and to identify the sphenoid sinus ostium. The mucosa at the anterior wall of the sphenoid sinus was then coagulated with a bipolar cautery. Using Kerrison rongeurs to perform an anterior sphenoidotomy, the sphenoid ostium was enlarged. The sphenoidal septum was removed, and the floor of the sella turcica, bilateral carotid protuberances, and clivus were exposed. Fluoroscopic imaging was used to corroborate the vertical dimension of the clivus and as guidance while extending the sphenoidotomy caudally to expose the lower clivus.

![Endoscopic view of the clivus showing a tumor (T) at the clivus. The sella turcica is exposed superiorly (S) and the carotid protuberances are located laterally (C).](image)

We encountered the tumor at the center of the exposed clivus (Fig. 2) with the area of a small bone and dural defect. The clival bone around the defect was removed, and the dura mater was exposed from the floor of the sella turcica to the lower clivus vertically and from one carotid protuberance to the other carotid protuberance horizontally. We extended the dural incision superiorly and inferiorly from the original dural defect and removed the tumor piecemeal with a micropituitary rongeur. The normal arachnoid membrane was identified at the caudal pole of the tumor.
We dissected the tumor further rostrally while keeping the arachnoid membrane intact as much as possible. The portion of tumor encasing the basilar artery was resected, which exposed the basilar trunk and its perforating arteries (Fig. 3 upper left). At this point we replaced the 0\(^\circ\) endoscope with a 30\(^\circ\) endoscope to explore the rostral portion of the posterior fossa (Fig. 3 upper right). The portion of tumor encasing the distal basilar artery, proximal superior cerebellar arteries, and proximal posterior cerebral arteries was removed with No. 5 and 7 suction cannulas and a micropituitary rongeur (Fig. 3 lower left). The tumor extending behind the basilar artery and its branches was deeply deforming the midbrain. So as not to damage the many small perforating arteries from the basilar artery and its branches, we did not attempt to remove the tumor that was located behind it. We used a 70\(^\circ\) endoscope to explore the basal hypothalamic region and to remove the segment of tumor located rostrally to the basilar tip (Fig. 3 lower right). We decided that the residual tumor located behind the basilar artery and its many small perforating arteries should be treated by stereotactic gamma knife surgery.
We obtained free fat graft material from the abdominal wall through a 2-cm periumbilical skin incision and placed one piece of the fat graft at the dural opening. A water tight seal was confirmed by repeated Valsalva maneuvers. We placed another piece of the fat graft in the sphenoid sinus and packed the sphenoid sinus with an absorbable gelatin sponge to provide further support to the graft. We laid absorbable gelatin film at the middle meatus allowing air flow at the level of the inferior meatus.

**Postoperative Course.** The patient was ready to be discharged from the hospital the following day but was kept in the hospital for an additional 2 days because of the possibility of cerebrospinal fluid (CSF) leakage. His postoperative course was benign, and his symptomatic improvement was dramatic, with improvement of ataxia, complete resolution of emotional instability, and subjective improvement of his memory. A postoperative MR image of the brain revealed fat graft material in the sphenoid sinus and posterior fossa and residual tumor indenting the midbrain (Fig. 4). His residual tumor was treated by stereotactic gamma knife surgery 10 days after the endoscopic resection.

![Fig. 4. Postoperative magnetic resonance images, axial and sagittal views, demonstrating a fat graft and residual tumor located between the basilar artery and the midbrain.](image)

**DISCUSSION**

Surgical treatment of chordomas has been challenging to neurosurgeons for many years.[9] Recently, radical resection of the tumor has been recommended for better outcomes.[2] When complete resection of the tumor is not achieved, radiation treatment for residual tumor is recommended.[1,4,7] Conventional microscopic transsphenoidal techniques have been used in the treatment of clival chordomas.[5]

While performing endoscopic transsphenoidal pituitary surgeries, we have observed the panoramic anatomical exposure provided by these techniques.[3] Endoscopic transsphenoidal techniques expose the planum sphenoidale, the optic protuberances covering the optic nerves, the carotid protuberances, and the clivus as well as the sella turcica. When we encountered a patient with a tumor based at the clivus, we were convinced that the tumor could be resected using an endoscopic technique. Other surgical techniques for skull base lesions were fully discussed with the patient and his family prior to his endoscopic operation.

Using an endoscopic technique for a clival lesion requires a hollow space large enough to expose the entire spectrum of the lesion. This case was unique in that the patient had an amply sized sphenoid sinus through which the entire vertical dimension of the tumor would be accessible. Although bone can be drilled under endoscopic exposure, thick clival bone may hinder exposure of the intradural lesion. The case reported here involved a patient who had a very thin clival bone that was easily removed with a
micro-Kerrison punch. Endoscopic techniques are best used on tumors that are relatively avascular; such avascular tumors in the posterior fossa include chordoma and epidermoid. The tumor must also be located in the midline posterior fossa so that it can be accessible through the 2-cm wide exposure at the clivus. The main concern preoperatively in our patient was the possibility of CSF leakage. To minimize the risk of a CSF leak, we kept the dural opening as small as possible. Although we did not measure the actual size of the dural opening, we estimated that it was approximately 2 cm in diameter. One piece of a large abdominal fat graft was placed at the dural opening. When the fat graft was placed at the dural opening, its shape resembled that of a dumbbell with the center portion at the dural opening and one end resting intradurally and the other extradurally. Fibrin glue may help in preventing CSF leakage but it was not used in our patient. We performed repeated Valsalva maneuvers to confirm water tight closure.

The case of the posterior fossa chordoma reported here involved unique elements that included a large sphenoid sinus, thin clival bone, avascular nature of the tumor, and the patient's desire in seeking a less invasive surgical technique. With the use of a 30° endoscope the residual tumor located behind the basilar artery was accessible through the small spaces among the numerous small perforating arteries arising from the basilar artery and its large branches. It might have been possible to remove the tumor located behind the basilar artery; however, so as not to damage the small perforating arteries, we made no attempt to remove this portion of tumor. To our knowledge, this is the first reported case of a posterior fossa chordoma resected by using an endoscopic transsphenoidal technique. A case of pathological laughter with a prepontine chordoma was previously reported.[6]

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References


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