Endoscopic endonasal transclival approach to a ventral pontine pediatric ependymoma

Case report

Prajwal RajaPPa, M.D., Konstantinos Margetis, M.D., Dimitri Sigounas, M.D., Vijay Anand, M.D., Theodore H. Schwartz, M.D., and Jeffrey P. Greenfield, M.D., Ph.D.

Departments of Neurosurgery and Otolaryngology-Head and Neck Surgery, Weill Cornell Medical College, New York-Presbyterian Hospital, New York, New York

The authors report a case of a recurrent pediatric ventral pontine ependymoma that they resected through an endonasal endoscopic transclival approach. Regarding the options for a surgical approach to ventral pontine tumors, traditional far-lateral approaches are associated with considerable morbidity due to the required muscle mobilization, brain retraction, and in-line obstruction of cranial nerves before reaching the target. The endoscopic endonasal transclival approach was made appealing by eliminating all of these concerns. The patient’s fully pneumatized sphenoid sinus, laterally displaced basilar artery, and the direct ventral location of the bulky disease all further supported this unconventional choice of surgical corridor to achieve a palliative brainstem decompression of an incurable recurrence.

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Key words • brainstem • ependymoma • neuroendoscopy • skull base • oncology

Ependymomas constitute the third most common pediatric brain tumor; they are of neuroectodermal origin and are usually midline posterior fossa tumors. Maximal survival rates are dependent on achieving gross-total resection (67%–93% 5-year overall survival) compared with subtotal resection (22%–52% 5-year overall survival). Chemotherapy and radiation therapy have proven ineffective for both primary and recurrent ependymomas. One-third of patients diagnosed with ependymoma will have local recurrence between 13 and 25 months. Many of these patients will undergo re-resection, highlighting the importance of aggressive surgical approaches both at diagnosis and recurrence. However, clearly the choice of approach, tumor location, and history of adjuvant therapies all play a role in determining the effectiveness of the resection. Conventionally, ventral pontine lesions have been accessed via the posterolateral suboccipital approach. Since this approach offers less of a surgical corridor for anterior brainstem lesions, variations have been described including the retrosigmoid, far-lateral, and extreme far-lateral craniotomies. Although these approaches provide access to ventral brainstem lesions, they result in significant morbidity due to cranial neuropathies and extensive bone removal incurred with open neurosurgical procedures.

The endoscopic endonasal approach has been more recently used for ventral skull base lesions. The advantages of this approach include the avoidance of neurovascular structures, elimination of the need for parenchymal retraction, and low overall morbidity. We report the feasibility and safety of the first reported purely endoscopic endonasal transclival approach for a purely intradural posterior fossa brain tumor and discuss the possible utility for biopsy or resection of posterior fossa and brainstem tumors in the pediatric population.

Case Report

History. A 16-year-old right-handed boy had undergone a subtotal resection of a posterior fossa mass at the age of 3 years. Gross-total resection was not achieved because of brainstem invasion. Pathology revealed a WHO Grade II ependymoma. The patient was then offered ad-
juvant therapy with stereotactic radiosurgery and 5 cycles of cyclophosphamide, cisplatin, etoposide, and vincristine. He subsequently underwent monitoring with serial MRI for residual disease.

At 16 years of age, the patient presented with headaches, dysphagia, and left hemiparesis. Magnetic resonance imaging demonstrated a 2.8 × 3.4-cm ventral intrinsic lesion at the pontomedullary junction with foci consistent with acute hemorrhage (Fig. 1A and B). The patient underwent decompressive surgery via an endoscopic endonasal transclival approach with near-total resection (Fig. 1C and D), and he experienced complete resolution of his focal neurological deficits.

**Initial Operation.** The patient was intubated, and triple antibiotics, glucocorticosteroids, and antihistamines were administered. A lumbar drain was placed, 10 ml of CSF was withdrawn, and 0.25 ml of 10% fluorescein (AK-Fluor, Akorn) was injected to help visualize any CSF leaks during the closure phase. The patient’s face was prepared with Betadine solution, and the nasal cavity was irrigated with a dilute solution of 80 mg gentamicin. A Mayfield head clamp and neurophysiological monitoring electrodes were applied (motor evoked potentials and cranial nerves VII–XII). Cottonoids soaked in 4% cocaine were inserted into the nasal cavity, and registration of the stereotactic image guidance system followed.

Under endoscopic visualization with the aid of a 0°, 18-cm-long, 4-mm-diameter rigid endoscope (Karl Storz), bilateral nasal septal flaps were developed based on the sphenopalatine artery and were tucked into the nasopharynx. Bilateral superior and partial inferior turbinectomies, partial sphenoidectomy, and partial ethmoidectomies were performed to widen the surgical corridor. Under stereotactic guidance the clivus was thinned out using a diamond bur. Curettes provided the initial bony opening to the dura, which was then widened using Kerrison rongeurs. The Doppler probe was used to identify the presumed location of the vertebral and basilar arteries, and the location was reconfirmed with intraoperative guidance. The dura mater was then opened in a cruciate fashion using a sickle knife and bipolar cautery (Video 1).

**VIDEO 1.** Video clip demonstrating the basic operative steps. Copyright Jeffrey P. Greenfield. Published with permission. Click here to view with Media Player. Click here to view with Quicktime.

The tumor was visible and samples were sent for pathological examination. The preliminary intraoperative report was recurrent ependymoma. Suction, ring curettes, and a tissue-shaving device (NICO Myriad, NICO Corp.) were used to remove tumor tissue until the ventral aspect of the brainstem was visualized. The brainstem was stained with hemosiderin and it was used as a margin for tumor debulking. The presurgical goal of brainstem decompression was achieved, and any further attempt to remove macroscopic tumor surrounding the basilar artery was deferred. Gelfoam and Floseal (Baxter) were necessary at various points during and after tumor resection to achieve hemostasis. Neurophysiological monitoring did not detect any signal changes during surgery. An autologous fascia lata graft was placed over the defect, and the gasket seal principle was applied by placing a Medpor plate (Porex Corp.). The nasal septal flaps were overlayed and were covered with DuraSeal (Covidien). Nasal packing with Telfa (Covidien) completed the procedure. The patient was extubated in stable condition.

**Initial Postoperative Course.** The tumor pathology was now consistent with a WHO Grade III anaplastic ependymoma. No adjuvant therapy was given because of family indecision regarding enrollment in a Phase I or II clinical trial.

**Second Operation.** Four months postoperatively, without having received any postoperative therapy, the patient presented again with identical speech and swallowing difficulties along with a left hemiparesis and was found to have a recurrent lesion on MRI (Fig. 2A and B). The patient underwent re-resection through the endoscopic, endonasal, transclival approach and once again demonstrated significant neurological improvement in cranial neuropathies and hemiparesis soon after the procedure (Fig. 2C and D). There were no significant technical limitations to the reoperation with respect to mobilizing the pericranial flap. Similar to his first endonasal procedure, an autologous fascia lata graft was placed over the defect and reinforced with Medpor for a gasket seal. DuraSeal was reapplied to the area for secondary repair of the dura. Because he did not meet the entry criteria for recurrent ependymoma trials, the patient was treated off of protocol with superselective intraarterial delivery of bevacizumab and cetuximab for 3 cycles totaling 6 months.

![Fig. 1. Ventral pontine ependymoma with significant brainstem invasion. Preoperative axial (A) and sagittal (B) T1-weighted, Gd-enhanced MR images. Postoperative axial (C) and sagittal (D) T1-weighted, Gd-enhanced MR images demonstrating near-total tumor resection via an endoscopic endonasal transclival approach.](Image)
Endoscopic approach to ventral brainstem lesions

Fig. 2. Early recurrence of the aggressive ventral pontine ependymoma. Axial (A) and sagittal (B) T1-weighted, Gd-enhanced MR images obtained prior to the reoperation. Axial (C) and sagittal (D) T1-weighted, Gd-enhanced MR images demonstrate the tumor debulking after the reoperation via an endoscopic endonasal transclival approach.

Second Postoperative Course. Eight months after the second endonasal procedure, the patient experienced a third recurrence that was far more asymmetrically located. We thought that a repeat endonasal approach would be fraught with added morbidity related to reaching most of the tumor, which was now situated lateral to the laterally displaced basilar artery and to closing the same skull base defect for the third time. We also discussed palliative care options for this incurable tumor, but did offer one last attempt at meaningful decompression via a retrosigmoid approach, contralateral to the initial surgical corridor. This surgery was complicated by a postoperative CSF fistula through the previously irradiated skin of the retrosigmoid incision, and the patient developed gram-negative meningitis. Despite external drain placement and intravenous antibiotics, the patient’s condition deteriorated neurologically, and comfort care measures were requested by the family. One month after the final procedure the patient died.

Discussion

This report demonstrates the feasibility and safety of an endoscopic endonasal transclival approach to ventrally located intrinsic brainstem masses in tumors encountered within the pediatric population. The endoscopic endonasal approach was first proposed for intra-, supra-, and parasellar lesions.5 It was soon expanded to extradural clival lesions.3,16 To our knowledge, the only other published cases of an endoscopic endonasal transclival intradural approach in a pediatric patient were for an invading extradural lesion (chordoma) and a pontine cavernoma.6,11,17 There are various options for an open, microsurgical approach to a lesion located anterior to the brainstem.14 The most commonly used approach for an extradural lesion is the transoral approach with its transpalatine/labiomandibular/labiomandibular modifications. Other options are the transcranial-transbasal, extended frontal, transsphenoidal, transmaxillary, and transcervical approaches. For intradural lesions, one or a combination of lateral approaches is usually selected.25 A pre- or retrosigmoid approach as well as the far-lateral approach and its modifications are also usually used. All of these approaches have significant drawbacks and limitations that may result in significant morbidity. In our patient, a viable alternative would have been a transoral approach. However, a transpalatine extension would have been necessary and possibly a labiomandibular extension to achieve a low angle and to approach the upper part of the tumor. Such an approach would be associated with significant morbidity in terms of dental malocclusion,14 proper functioning of the hard and soft palates, velopharyngeal incompetence, and CSF leakage.7 We thought that the added morbidity would result in an extended hospital stay and result in long-term impairment of the patient’s quality of life. In retrospect, our concerns over an inability for wound healing in the heavily radiated and poorly vascularized posterior fossa were realized when the patient developed a CSF fistula after his final operation done via a “traditional” approach to the posterior fossa.

The endoscopic endonasal approach in pediatric patients may be restricted to a few pathological entities and may be associated with more difficult anatomical constraints when compared with the pathology and anatomy encountered in adults. A recent anatomical study20 delineated the anatomical technique in pediatrics and highlighted, among other issues, the piriform aperture, the cavernous intercarotid distance, and the possible inability to raise an adequate nasoseptal flap as possible constraints.19 The differences in individual anatomy and in the lesion’s location and relationship with adjacent structures necessitate an individualized assessment and selection of the optimum method. We stress the importance of adequate preoperative imaging for the endoscopic endonasal transclival approach, usually in the form of MRI and CT angiography, the use of Doppler ultrasonography and a stereotactic image guidance system, and a combined otolaryngology/neurosurgical team.

The endoscopic endonasal transclival approach was attempted as a palliative option in this patient, and it provided exceedingly good visualization and facile tumor resection. It is unlikely to become a mainstay of approaches to the posterior fossa in children, but it demonstrates a viable and advantageous option for the treatment of purely intradural lesions in select pediatric patients.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Greenfield,
References


