Expanded endoscopic endonasal resection of an olfactory schwannoma

Case report

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Olfactory schwannomas are rare tumors of the anterior skull base that are possibly derived from ectopic Schwann cells, perivascular neural tissue, or sensory nerves of the meninges. The authors report the case of a 14-year-old boy with an olfactory schwannoma that extended inferiorly through the cranial base and superiorly into the frontal lobe. Because of the growth characteristics of the tumor and the significant overlying frontal lobe edema, the lesion was approached via an endonasal endoscopic route, as a strategy to minimize brain retraction. This tumor was characterized radiographically as contrast-enhancing with cystic areas and erosion into bone. The tumor showed immunoreactivity for S100 protein and leukocyte antigen 7 (CD57) but not epithelial membrane antigen, supporting the diagnosis of olfactory schwannoma. A gross-total resection was achieved. This approach represents a novel application of endoscopic endonasal surgery to the pediatric neurosurgical context, as well as a favorable outcome in an extremely unusual tumor type, that should be applicable to other appropriately selected pediatric brain tumors.

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Key Words • endonasal approach • endoscopic approach • olfactory schwannoma • pediatric brain tumor • skull base

Olfactory schwannomas are extremely rare tumors of the anterior skull base. The olfactory tract and bulb are part of the central nervous system and are unmyelinated, so the association with a tumor derived from a cell involved in peripheral myelination is perplexing. Additional sources of such skull-based schwannomas have been postulated, including hamartomatous displacement of neural crest cells, perivascular sympathetic neural plexus, the meningial branch of the trigeminal nerve, and the anterior ethmoidal nerve.10,13 To date ~ 23–26 olfactory schwannomas have been described in the literature, some under the title of subfrontal schwannoma or neurilemmoma.4,8,13,14 Compared with other intracranial schwannomas, olfactory tumors have a slightly higher tendency to be found in males and at a younger age (mean 4th decade of life).5 Patients usually present with headache, seizure, anosmia, frontal lobe dysfunction, and signs and symptoms related to elevated intracranial pressure.13 The following case highlights an effective surgical approach for olfactory schwannomas and reviews their histopathological features.

Case Report

History and Examination. In September 2007, this 14-year-old boy presented with a 2-year history of declining school performance, personality changes, difficulty concentrating, worsening daily headaches, and a 20-lb weight loss over the past 6 months attributed to a loss of taste. A CT scan obtained at an outside hospital revealed a large anterior fossa mass with significant associated frontal lobe edema, which prompted a transfer to our institution for further evaluation. On examination the child had normal findings during his neurological examination with the exception of diminished smell, bilateral papilledema, and a significantly flattened affect. Magnetic resonance images (Figs. 1 and 2) and a CT angiogram were obtained to further evaluate the lesion and its vascular supply. These studies disclosed erosion of the tumor through the cranial base into the left ethmoid sinus (Fig. 3).

Abbreviations used in this paper: EMA = epithelial membrane antigen; GTR = gross-total resection; OECT = olfactory ensheathing cell tumor.
Initial Operation. Because of the significant frontal lobe edema and the inferior extension of the tumor, an endoscopic endonasal trajectory was selected for tumor biopsy and initial debulking, as a way of minimizing brain retraction and also to establish a histological diagnosis. This was accomplished using an expanded endonasal transplanum and transcribriform approach as previously described by our group.5,7 Because of the unusual nature of the lesion and its uncertain histological characteristics, a 2-stage approach was planned. The first of these stages was designed to achieve exposure, biopsy, and debulking, with the thought that if this lesion proved to be a type for which aggressive resection was not warranted (for example, metastatic neuroblastoma and inflammatory process), we would have spared the patient the potential morbidity of dissecting the mass from the edematous frontal lobe and anterior cerebral blood supply.

Exposure of the skull base was carried out as described previously.5,7 Tumor was encountered within the ethmoid sinuses on the left (as suggested by preoperative imaging) and found to be intimately associated with the olfactory filaments. The anterior and posterior ethmoid arteries were identified bilaterally and cauterized at the junction of the orbit and skull base. The bone of the skull base was then thinned with a drill including the left superior/medial orbit and elevated from the underlying dura mater. The dura was exposed from the superior intercavernous sinus posteriorly to the most rostral extent of the exposure and cauterized. Anteriorly the dura was incised and the falx cauterized and transected to allow reflection of the dura from the skull base. The tumor was internally debulked and sharply dissected from the frontal lobes and left medial orbit (Fig. 4). There was clear subpial invasion requiring dissection along the left gyrus rectus. The tumor had a fibrous, tough consistency amenable to resection by a combination of sharp dissection and the use of a modified ultrasonic aspirator (Integra, Life Sciences). There were also friable portions of the tumor that were easily resected using a 2-suction technique. Approximately 70% of the tumor was debulked before the subpial dissection revealed tumor adherent to a frontopolar branch of the anterior cerebral artery. The pathological entity at this intraoperative point was indeterminant, and a decision was made to stage the resection of the tumor if the final pathological diagnosis suggested a GTR was necessary.

Adequate hemostasis was obtained and the defect was repaired with an in-lay fat and dural substitute graft fol-

Fig. 1. Axial (A), coronal (B), and sagittal (C) contrast-enhancing images showing erosion of the tumor through the cranial base into the left ethmoid sinus.

Fig. 2. Axial T2-weighted MR image showing tumor erosion.

Fig. 3. Coronal (left) and sagittal (right) CT reconstructions of the anterior skull base.
followed by an on-lay transposition of a vascularized septal mucosal flap. The child was extubated and awoke with no new neurological deficits, and he was transferred to the pediatric intensive care unit overnight for observation. Postoperative MR imaging (Fig. 5) and CT scanning confirmed a substantial internal debulking of the tumor with significant improvement in frontal lobe mass effect.

Pathological Findings. On microscopy, the tumor cells were somewhat spindle shaped and arranged in vague fascicles with indistinct cell borders. The cytoplasm was noted to be pale, eosinophilic, wavy, and vesicular. The nuclei were described as round-to-ovoid with pinpoint nucleoli. Mitotic figures were rare. Although classic Verocay bodies were not apparent, there were areas of alternating hypopericytosis.

Immunohistochemical studies showed that the tumor cells expressed S100 protein and stained negatively for EMA, glial fibrillary acidic protein, and synaptophysin. A tumor cell proliferative index of 2% was shown by Ki 67. The tumor was reticulin-rich, and the reticulin stain highlighted the fascicular architectural pattern. The tumor stained positively for leukocyte antigen 7 (Leu7/CD57).

Second Operation. Based on the histopathological diagnosis, it was believed that an attempt at achieving a GTR was warranted. Through an endonasal route, the previous reconstruction was visualized and the nasal-septal flap was mobilized with its attached vascular pedicle. The extent of the transplanum, transcribriform exposure was identified, and further tumor dissection and internal debulking was undertaken. The frontopolar artery was densely adherent to the tumor (Fig. 6), and the mass was carefully dissected from this vessel and the overlying frontal lobe. Gross-total resection of the tumor was achieved, and the resection cavity was closed by transposing the nasal-septal flap. Again
the child awoke with no new neurological deficits and was admitted to the pediatric intensive care unit for observation. The following day a cerebral angiogram was obtained, which confirmed patency of the frontopolar artery and the surrounding vasculature.

**Postoperative Course.** Postoperative imaging (Fig. 7) confirmed GTR of the tumor. The child was discharged home and on follow-up visits has demonstrated normal neurologic function (with the exception of anosmia) and a well-healed mucosal flap.

**Discussion**

In an adult, the first consideration in the differential diagnosis of large, partially cystic, enhancing subfrontal lesion is a meningioma, although such lesions are uncommon in childhood, and alternative possibilities are considered more likely. Olfactory schwannomas, which are exceedingly rare in children and adults, can have cystic areas like meningiomas but the key distinction is the relationship of the tumor with the adjacent bone: olfactory schwannomas tend to be erosive (see Fig. 3), whereas meningiomas tend to induce hyperostosis. Furthermore, on angiography meningiomas are characterized by a robust vascularity and a prominent tumor blush, whereas schwannomas of the skull base are relatively hypovascular. Other entities in the differential diagnosis include esthesioneuroblastoma and carcinoma of the paranasal sinuses, which both tend to have a more aggressive, destructive appearance and clinical presentation, and metastatic abdominal neuroblastomas, which occur more commonly in younger children and characteristically manifest with a “raccoon eye” appearance.

To date this is the first description of the use of the endonasal approach for resection of an olfactory schwannoma, and one of the first descriptions of an expanded en-

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**Fig. 6.** Intraoperative image of tumor adherent to frontopolar artery. C = cottonoid; D = dissector.

**Fig. 7.** Axial (A), coronal (B), and sagittal (C) MR images showing GTR of the olfactory schwannoma.
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donasal approach for the management of a lesion with these growth characteristics in a child. The patient tolerated the procedure well with no sequela and subjective improvement in neurocognitive function. Alternative surgical approaches include interhemispheric or subfrontal dissections with or without orbital osteotomies. The advantages of the endonasal approach include the ability to devascularize the tumor blood supply from the anterior and posterior ethmoidal arteries during the initial approach. Additionally, there is minimal retraction on the frontal lobes during the resection while also avoiding potential cosmetic issues associated with other techniques. Generally speaking, disadvantages include a higher risk of anosmia and cerebrospinal fluid leakage with the endonasal approach, although specifically in this case the child presented with anosmia and the nasoseptal flap has proven to be an effective means of closure in our experience.

Lack of EMA expression and strong S100 reactivity supported a diagnosis of schwannoma over meningoima. The possibility of a recently introduced tumor of the olfactory groove, ÖECT, was entertained. A single case of ÖECT was described by Yasuda et al. as being histologically similar to olfactory schwannoma but distinguishable based on immunohistochemical findings. In their study, they used lack of Leu7/CD57 expression to differentiate ÖECT from olfactory schwannoma, which are usually Leu7/CD57 positive, as in our case. Olfactory ensheathing cells do not myelinate olfactory nerve fibers but do surround them and act in a supporting manner that includes creating an environment conducive to axon regeneration and repair. Their phenotype is very similar to both astroglia and Schwann cells, and ultimately they function in both roles as the cells that accompany the olfactory system through its transition from intracranial to extracranial central nervous system tissue. In this case, the histological findings after H & E staining supported a diagnosis of schwannoma, and immunohistochemistry was used to confirm the diagnosis.

Conclusions

The endonasal endoscopic approach for tumor resection provides advantages in the resection of large subfrontal tumors. In this case GTR with no morbidity was achieved in a 14-year-old boy by using these techniques. Immunohistochemical studies for S100, EMA, and Leu7/CD57 are useful in distinguishing schwannoma from meningoima and olfactory ensheathing cell tumor.

Disclosure

Drs. Kassam and Prevedello are consultants for Karl Storz Endoscopy. Dr. Kassam is also a consultant for Stryker Navigation.

References
