Endoscopic management of pediatric sinonasal schwannoma: case report

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Abstract. Endoscopic management of pediatric sinonasal schwannoma: case report. Objectives: To describe an extremely rare pediatric sinonasal schwannoma, and to review management strategies and relevant literature. Methods: Case report of pediatric sinonasal schwannoma, that was imaged with computed tomography and magnetic resonance imaging and managed endoscopically. Immunohistochemical analysis was performed to determine pathology. Results: A 12-year-old girl presented with a 2-month history of progressive left exophthalmos. Imaging studies showed a large heterogeneous tumour arising from the ethmoid sinus and extending to the base of the skull and to the orbital cavity. The lesion was removed with an endonasal radical excision. The final pathological diagnosis was schwannoma. There was no tumour recurrence or any major complication during the 2-year follow up. Conclusion: Schwannoma should be considered in the differential diagnosis for pediatric patients with intranasal masses. Depending on the location and extent of the tumour, endonasal endoscopic excision could be a suitable management strategy.

Introduction

Schwannomas comprise Schwann cells of the nerve sheath. These rare tumours can occur in any part of the body. Schwannomas that occur in the head and neck region account for 25-45% of all schwannomas, and they develop most frequently in the eighth cranial nerve, followed by the spinal nerves.¹,² These sporadic head and neck schwannomas, unlike those that occur in other body sites, are not associated with BRAF or KRAS mutations.³ Only 4% of these head and neck lesions involve the nasal and paranasal sinuses.¹ The most frequent sites, in descending order, are the nasoethmoidal complex, maxillary sinus, nasal cavity, and sphenoid sinus.¹ Schwannomas are relatively benign, encapsulated tumours, and they are not generally locally aggressive. Most schwannomas are radioresistant; thus, surgery remains the best treatment option.⁴ An endonasal endoscopic approach eliminates an external incision and allows a shorter hospital stay than traditional approaches.⁵ Most cases of sinonasal schwannomas reported in the current literature have been found in adults without any sexual predominance. However, they have been reported rarely in the pediatric population. Due to the scarcity of these cases and the limited number of reports that address treatment experiences, we herein present a case of sinonasal schwannoma in a 12-year-old girl who was treated with endonasal endoscopic excision.

Case report

A 12-year-old girl presented with a 2-month history of progressive left exophthalmos and persistent nasal obstruction. No diplopia, blurred vision, cerebrospinal fluid rhinorrhea, or epistaxis was reported. An ophthalmological survey disclosed normal light reflex, visual acuity, and intraorbital pressure. Orbital computed tomography (CT) revealed a large heterogeneous tumour arising from the ethmoid sinus and extending to the base of the skull superiorly and to the orbital cavity laterally (Figure 1A, arrows). A T2-weighted magnetic resonance imaging (MRI) scan revealed a well-circumscribed tumour that appeared to be growing in an expansile fashion. There was no evidence of brain parenchyma invasion or extensive surrounding edema, which suggested a benign lesion (Figure 1B). The patient initially underwent endoscopic biopsy. An examination of the intraoperative frozen pathology specimen revealed that the lesion was a benign neurogenic tumour,
narial wound was packed with a bioresorbable nasal dressing (Nasopore; Polyganics, The Netherlands). Histopathological analysis revealed a spindle cell tumour with compact Antoni type A tissue and loosely structured Antoni type B tissue. In Antoni type A areas, we identified Verocay bodies, which formed parallel rows of tumour nuclei separated by an eosinophilic, anuclear zone (Figure 3A & B). Immunohistochemical analysis showed that the tumour cells were labeled with S100 and vimentin antibodies (Figure 3C & D), but not with EMA, GFAP, CD34, synaptophysin, or chromogranin-A antibodies. The morphology and immunohistochemical staining results were consistent with a pathologic diagnosis of benign schwannoma.

During the 2-year follow up, no tumour recurrence was noted under repeated CT scans (Figure 4), and there were no major complications, such as visual loss, diplopia, or focal neurologic defect.

Discussion

Sinonasal schwannomas develop mostly in autonomic nerves of the paranasal sinus or in maxillary or ophthalmic branches of the trigeminal nerve.\textsuperscript{5} A recent study reported that the average age of patients with sinonasal schwannomas was 46 years old (range 17-68 years).\textsuperscript{5} Our patient was 12 years old; this type of case is very rare in the literature. Sinonasal schwannomas present with various signs and symptoms that relate to the...
Exophthalmos, facial swelling, and epiphora are less frequently observed. In this case, our patient initially presented with exophthalmos. Because this symptom is not frequently observed at first visit, it could have easily led to misdiagnosis. Sinonasal schwannomas are typically benign and well-encapsulated. However, some reports have suggested that these tumors can also lack encapsulation and become locally aggressive, or even worse, enter malignant transformation.6,7 Regardless of whether a sinonasal schwannoma is benign or malignant, they frequently cause bony erosion and extend intracranially.8,9

Surgery is the best treatment for both benign and malignant schwannomas. Calceterra et al. described an excision of a benign neurilemmoma of the sphenoid sinus with a transantral approach. They suggested that a combination of lateral rhinotomy, a transeptal approach, and even skull base surgery may be necessary for complete excision of the tumor extensions.10 The choice of a surgical
endoscopic excision is associated with lower morbidity, shorter hospital stays, and better cosmetic outcomes compared to surgery. On the basis of those reports and our patient’s characteristics - a 12-year-old girl with a well-encapsulated tumour - we chose the endonasal endoscopic approach as the operative method. We removed the lesion successfully, and the patient showed good preservation of aesthetic and visual function in the 2-year follow up. A further, long-term follow up should be conducted.

There are multiple methods for reconstructing skull base defects. In 2013, a review article concluded that multilayered free grafts were adequate for small defects (<1 cm), but that pedicle vascular flaps with low postoperative CSF leak rates (5%) were the method of choice for large defects (>3 cm). In managing pediatric skull base defects with a pedicle flap, a special concern is the growth pattern of nasoseptal mucosa. Shah and associates (2009) reported that the septum achieves adult length at about age 13, and thus, it is not recommended to reconstruct anterior skull base defects in children less than 10 years old. Children between the ages of 10 and 13 years require careful clinical consideration on a case-by-case basis. Hence, we used a nasoseptal flap, a type of pedicled flap placed on the posterior nasal septal artery, to reconstruct the skull base defect in our patient. To date, the prognosis is good without any ascending intracranial infections.

**Conclusion**

The endonasal endoscopic approach provided complete, excellent visualization for resection of an intranasal tumour that extended into the base of the skull. We found that endonasal endoscopic surgery was a safe, useful method for the complete resection of a benign schwannoma. This approach, although relatively conservative, showed equivalent effectiveness compared to external approaches. When diagnosing intranasal masses in children with an initial presentation of exophthalmos, schwannoma should be considered in the differential diagnosis.

**References**


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