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Endoscopic Excision of a Rare Sinonasal Schwannoma: A Case Report and Review of Literature

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Abstract

Schwannomas are rare slow-growing benign peripheral nerve sheath tumors arising from Schwann cells. Among all cases,approximately 25-45% are found in the head and neck area with 4% incidence of sinonasal schwannomas. Diagnosis is mainly done by histology and immunohistochemistry. Complete excision of tumor is standard modality of treatment.Endoscopic excision is widely practised in present time.

In our study, we present a case of 44-year-old female with a 3-year history of left-sided nasal obstruction and rhinorrhoea. Nasal endoscopy revealed a smooth, well-circumscribed mass arising from the lateral wall of the nasal cavity. Following detailed radiological evaluation, the patient underwent complete endoscopic excision of the mass with histopathological diagnosis of Schwannoma. During post-operative period, patient noticed complete relief of symptoms with no recurrence of disease in 1 year follow up. These rare tumors present diagnostic challenges due to their nonspecific clinical presentation and imaging features. The purpose of this study is to contribute to the limited literature on sinonasal schwannomas and highlights the importance of considering this diagnosis in patients presenting with unilateral nasal masses.

Keywords: Schwannoma, nasal cavity, sinonasal tumor, endoscopic surgery, S100 protein

Introduction

Schwannomas, also termed as neurilemmoma.These are benign slow-growing tumors originating from Schwann cells that form the myelin sheath around peripheral nerve fibers.[1] These tumors can occur anywhere in the body where peripheral nerves are present, but they show a predilection for the head and neck region, accounting for 25-45% of all schwannomas.[2]

Sinonasal schwannomas represent a rare subset of head and neck schwannomas, comprising less than 4% of all head and neck schwannomas.[3] with over 100 cases reported in the literature, these tumors remain uncommon entities that present diagnostic challenges due to their nonspecific clinical symptoms and imaging findings. The rarity of sinonasal schwannomas often leads to delayed diagnosis and may be initially misinterpreted as more common sinonasal pathologies such as inflammatory polyps, inverted papilloma or other benign tumors.[4]

The most commonly affected anatomical locations within the sinonasal tract include the ethmoid sinus, maxillary sinus, nasal cavity, sphenoid sinus, and nasal septum. These tumors typically present in middle-aged adults without gender or racial predilection, most commonly in the 5th to 6th decades of life. Clinical symptoms are generally nonspecific and include nasal obstruction, epistaxis, rhinorrhea, anosmia, headache, and facial swelling. [4-5]

Case Presentation

A 44-year-old female presented to the Department of Otorhinolaryngology with a 3-year history of left-sided nasal obstruction and rhinorrhoea. The patient denied any history of trauma, epistaxis, nasal discharge, diplopia or facial pain. There was no

significant past medical history or family history of neurofibromatosis.

Physical examination revealed a smooth, well-circumscribed, yellowish mass in the left nasal cavity abutting both the nasal septum and lateral wall was

noted along with deviated nasal septum with spur to left. The mass was insensitive to touch, and it did not bleed on touching. A Probe could be passed all around the mass. Diagnostic nasal endoscopy confirmed the presence of a well-defined mass obscuring the entire nasal cavity. The contralateral nasal cavity appeared normal, and head and neck examination was unremarkable.

High-resolution computed tomography (HRCT) of the paranasal sinuses demonstrated a single ovoid soft tissue mass measuring about 5.7 * 1.7* 3.5 cm in size in the left nasal cavity, extending posteriorly through the left posterior choana into the nasopharynx with associated secondary sinusitis. The left middle and inferior turbinates were obscured by the mass.

The mass caused lateral bowing of the medial wall of the left maxillary sinus and thinning of the bony medial wall structures.

Based on the clinical presentation and imaging findings suggestive of a nasal cavity tumor, the patient was planned for endoscopic excision of mass. Complete endoscopic excision of the mass was performed under general anesthesia along with septoplasty. Intraoperatively, the tumor was found to have an origin to lateral nasal wall anterior to the middle turbinate. The mass was completely excised and sent for histopathology.

Gross examination revealed a yellowish, well-encapsulated mass measuring 5 x 2.0 x 3 cm. Microscopic examination demonstrated a spindle cell lesion with alternating hypercellular and hypocellular areas characteristic of schwannoma. The hypercellular areas (Antoni A pattern) showed spindle cells with peripheral palisading of nuclei, while hypocellular areas (Antoni B pattern) displayed loose myxoid stroma. Nuclear atypia was minimal, and no mitotic figures were identified. Hemangiopericytomatous vasculature was observed throughout the lesion.

Immunohistochemical analysis revealed strong diffuse positivity for S100 protein, confirming the neural origin of the tumor. The lesion was negative for smooth muscle actin (SMA) and desmin, helping to exclude other spindle cell tumors. The MIB-1 labeling index was low at 1-2% in the highest proliferating areas, consistent with the benign nature of the lesion.

The patient had an uneventful postoperative recovery with prompt resolution of nasal obstruction symptoms. Follow-up nasal endoscopy at 3 and 6 months postoperatively showed complete healing. After 1 year of surgery, the patient is asymptomatic and does not have any recurrence of disease.

Discussion

Sinonasal schwannomas are exceptionally rare benign peripheral nerve sheath tumours. These tumours account for less than 4% of all head and neck schwannomas.[6]

The clinical symptoms of sinonasal schwannomas include nasal obstruction, epistaxis, rhinorrhea, anosmia, headache, facial swelling.[6-7]

Schwannomas arise from Schwann cells, which are responsible for forming the myelin sheath around peripheral nerve fibers in the peripheral nervous system.[8]

The pathogenesis of schwannomas is primarily driven by genetic mutations affecting the NF2 gene, which encodes merlin (schwannomin), a tumor suppressor protein. Loss of merlin function leads to dysregulation of multiple signaling pathways including mTORC1, EGFR-RAS-ERK, and PI3K-Akt pathways, resulting in uncontrolled Schwann cell proliferation and tumor formation.[9]

In the sinonasal tract, schwannomas are believed to arise from branches of the trigeminal nerve (ophthalmic and maxillary divisions), the sphenopalatine ganglion, or autonomic nerves innervating the nasal mucosa. The lateral wall of the nasal cavity, as seen in our case, is supplied by the anterior ethmoidal nerve and lateral posterior superior and inferior nasal nerves, which could serve as the origin of these tumors. However, identifying the specific nerve of origin can be challenging, and further research is needed to definitively determine the neural origins of sinonasal schwannomas.[9-10]

Radiological evaluation plays a crucial role in the assessment of sinonasal schwannomas, though imaging findings are generally nonspecific. High-resolution computed tomography (CT) provides

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valuable information about tumor size, location, and extent, as well as any associated bony changes.[7] Schwannomas typically appear as well-circumscribed masses with low to intermediate attenuation and mild to moderate contrast enhancement. Small tumors usually demonstrate homogeneous enhancement, while larger tumors may show heterogeneous enhancement due to cystic degeneration or hemorrhage.[7,11]

Magnetic resonance imaging (MRI) is superior to CT in differentiating tumors from inflammatory changes and evaluating extranasal extensions. On MRI, schwannomas characteristically show intermediate signal intensity on T1-weighted images and heterogeneously hyperintense signal on T2-weighted images. The T2 signal characteristics depend on the cellular composition, with hypercellular Antoni A areas appearing relatively hypointense and hypocellular Antoni B areas showing high signal intensity.[12]

Histopathological examination remains the gold standard for diagnosing sinonasal schwannomas due to their nonspecific clinical and radiological features. Microscopically, schwannomas demonstrate characteristic alternating patterns of hypercellular and hypocellular areas, known as Antoni A and Antoni B patterns, respectively.[7]

Immunohistochemical staining is essential for confirming the diagnosis and differentiating schwannomas from other spindle cell tumors. Schwannomas show strong and diffuse positivity for S100 protein, a marker of neural crest origin. Other supportive markers include calretinin and CD56, which are highly specific for schwannomas. Negative staining for CD34, smooth muscle actin, and desmin helps exclude other spindle cell tumors such as solitary fibrous tumors and smooth muscle tumors. [7, 13]

Complete surgical excision is the treatment of choice for sinonasal schwannomas, with the surgical approach determined by tumor size, location, and extent.[14]

The goals of surgery include complete tumor removal, preservation of function, and prevention of recurrence.[8] The endoscopic endonasal approach has become the preferred method for most sinonasal schwannomas due to its minimally invasive nature, excellent visualization, and reduced morbidity compared to external approaches.[15]

Endoscopic resection offers several advantages including shorter hospital stays, better cosmetic outcomes, reduced postoperative pain, and faster recovery. The technique allows for precise tumor removal with preservation of surrounding normal structures.[3] In cases where tumors extend into the orbit or skull base, endoscopic approaches can still be utilized, often in combination with image guidance systems to ensure complete resection while minimizing complications.[17]

Complications of endoscopic sinonasal schwannoma resection are generally rare and include cerebrospinal fluid leak (in cases with skull base involvement), epistaxis, dental numbness (with pterygopalatine fossa involvement), and epiphora.[18] The overall complication rate for endoscopic approaches ranges from 3% to 26%, which is significantly lower than open surgical approaches.[19]

Long-term outcomes following complete endoscopic excision are excellent, with recurrence rates being very low. In a review of endoscopic resections of sinonasal schwannomas, no recurrences were reported during mean follow-up periods ranging from 2 to 5 years. This excellent prognosis emphasizes the importance of achieving complete tumor removal during the initial surgical intervention.[18]

Postoperative surveillance typically includes regular nasal endoscopy and, in some cases, follow-up imaging to monitor for recurrence. The frequency and duration of follow-up depend on factors such as completeness of resection, tumor characteristics, and patient-specific factors. Given the benign nature of these tumors and low recurrence rates following complete excision, long-term surveillance beyond 2-3 years may not be necessary in most cases.[16-19]

Conclusion

Nasal schwannomas are exceptionally rare benign peripheral nerve sheath tumors that present significant diagnostic challenges due to their nonspecific clinical and radiological features. This case report contributes to the limited literature on sinonasal schwannomas and highlights the importance of maintaining a high index of suspicion for these rare tumors when evaluating patients with unilateral nasal masses.

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The diagnosis of nasal schwannoma relies primarily on histopathological examination with characteristic Antoni A and B patterns and strong S100 immunoreactivity.

Complete surgical excision via endoscopic approach represents the treatment of choice and is associated with excellent outcomes and minimal recurrence rates

This case emphasizes the importance of considering schwannoma in the differential diagnosis of sinonasal masses and demonstrates the effectiveness of endoscopic surgical management for these rare tumors.

Future research should focus on better understanding the molecular mechanisms underlying sinonasal schwannoma development and optimizing surgical techniques to ensure complete tumor removal while preserving nasal function.

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