International Journal of Otorhinolaryngology and Head and Neck Surgery
Azman AB et al. Int J Otorhinolaryngol Head Neck Surg. 2025 Feb;11(1):75-78
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20250122

Case Report

Unveiling the unseen: anterior nasal septal schwannoma

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Received: 28 August 2024
Accepted: 05 December 2024

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ABSTRACT

Schwannomas also known as neurilemmomas are slow-growing, encapsulated benign peripheral nerve sheath tumours that arise from Schwann cells. It has no gender or racial predilection and commonly occur between the 3rd to 6th decades. Approximately 45% of schwannomas arise from the head and neck region. Sinonasal and paranasal schwannomas however are relatively rare tumours and not commonly documented, accounting less than 4% of all head and neck schwannomas. Schwannomas arising from the nasal septum are rarely encountered, with only few cases reported. Here we report a case of a 50-year-old lady presented with unilateral nasal congestion and intermittent left sided epistaxis. Physical examination revealed a soft tissue mass occupying the left nasal cavity. A complete tumour excision via endoscopic approach was performed and the histopathologic examination identified the tumour as a schwannoma. A prompt resolution of the symptoms ensued following complete excision of the tumour.

Keywords: Sinonasal schwannoma, Schwannoma, Peripheral nerve sheath tumours, Anterior nasal septal schwannoma

INTRODUCTION

Peripheral nerve tumours are classified based on their specific cellular differentiation. The commonest benign peripheral nerve tumours are schwannomas and neurofibromas. They may occur sporadically or may be part of genetic syndromes such as neurofibromatosis type 1 or neurofibromatosis type 2.[1] Neurofibromas are benign peripheral nerve sheath tumours that can include components from several elements of peripheral nerves, including axons, Schwann cells, endoneuria fibroblasts, and perineurial cells. Schwannomas on the other hand originate from Schwann cells of the myelin sheath.[2] They can develop at multiple anatomical sites but most commonly affect the vestibular branch of cranial nerve VIII where it is known as an acoustic neuroma or vestibular schwannoma. Less common sites for schwannomas are scalp, face, oral cavity, and the respiratory tract.[3,4] The head and neck accounts for about 45% of schwannomas but only 4% arises from nasal and paranasal sinuses.[4] Of the benign peripheral nerve sheath tumours involving the sinonasal tract, only 32 reported cases of nasal septal schwannomas have been documented in the literature.[5]

CASE REPORT

A 50-year-old Malay lady with underlying hypertension presented with left sided nasal congestion, intermittent left epistaxis, and clear mucinous nasal discharge for 6

months prior to initial presentation. The bleeding was painless, spontaneous and mild in severity. There was no history of cacosmia, purulent nasal discharge, facial pain, hyposmia or facial distortion. She denied any constitutional symptoms or neck swelling. Rigid endoscopy of the nose revealed a large mass occupying the entire left nasal cavity. It had a smooth surface with prominent blood vessels and minimal bleeding upon manipulation (Figure 1). The origin of the tumour could not be identified with probing due to patient discomfort. The right nasal cavity was normal. A biopsy of the mass was undertaken under local anaesthesia, which was initially suggestive of a neurofibroma. Examination of the rest of the head, neck ear, nose and throat were unremarkable and there were no palpable neck nodes.

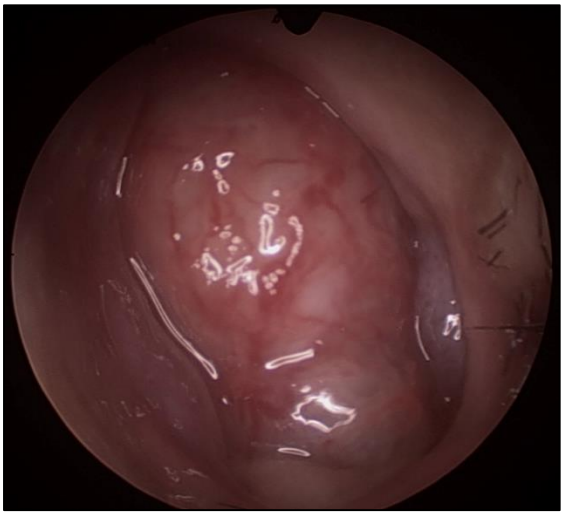

Figure 1: A large smooth surfaced mass occupying the left nasal cavity with prominent blood vessels.

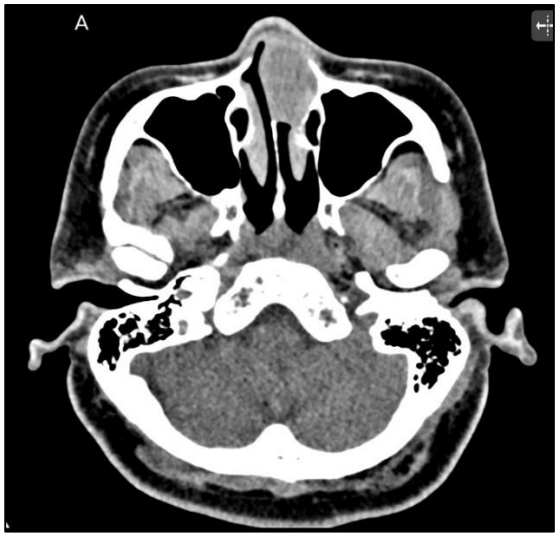

Figure 2: Computerized tomography (axial view) of the paranasal sinuses showed a well circumscribed soft tissue mass occupying the anterior part of the left nasal cavity with significant obliteration of the left nasal cavity, causing septal deviation to the right.

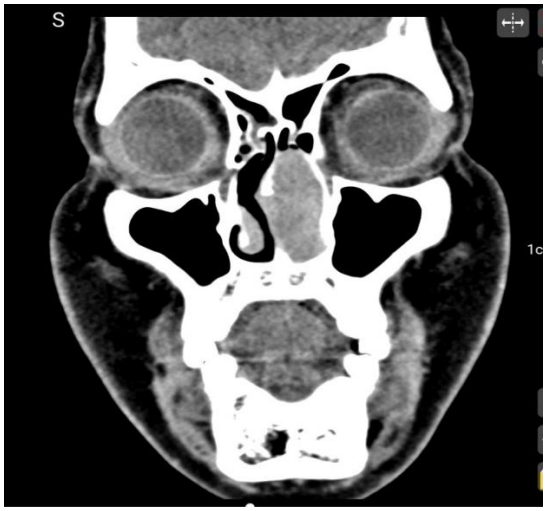

Figure 3: Computerized tomography (coronal view) of the paranasal sinuses showed a well circumscribed soft tissue mass occupying the left nasal cavity with obliteration of left osteomeatal complexes causing septal deviation to the right.

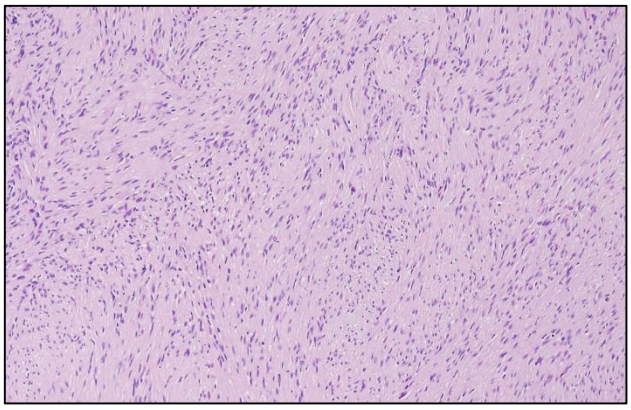

Figure 4: Spindle cells in fascicular arrangement with wavy nuclei and pale eosinophilic cytoplasmic processes (haematoxylin-eosin, original magnificationX200).

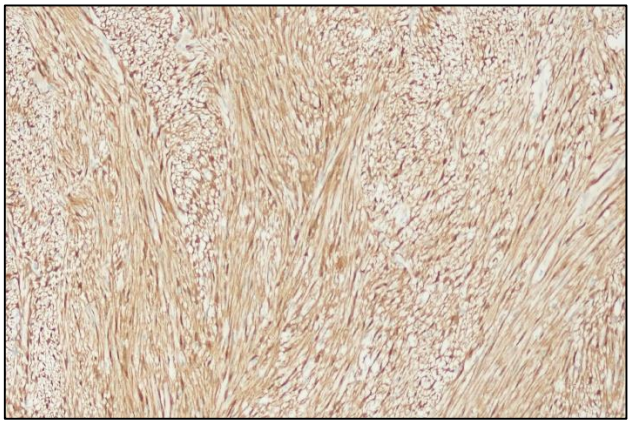

Figure 5: The spindle cells show diffuse positivity for S100 (Immunohistochemistry, original magnification X200).

Computerized tomography (CT) scan of the paranasal sinuses showed a well circumscribed soft tissue mass occupying the anterior part of the left nasal cavity measuring approximately 2.2×3.3×3.4 cm with significant obliteration of the left nasal cavity, causing nasal deviation to the right with no adjacent bone erosion (Figure 2 and 3).

The patient underwent a left endoscopic excision of the mass under general anaesthesia. Intraoperatively, there was a smooth surfaced mass occupying the whole wall of the left nasal cavity taking origin from the left anterior aspect of nasal septum.

Postoperative histopathological examination found features of a benign peripheral nerve sheath tumour favoring a schwannoma. The tumour composed of neoplastic spindle cells with wavy nuclei and pale eosinophilic cytoplasmic processes arranged in intermediate fascicles with areas showing vague nuclear palisading (Figure 4). Immunohistochemical staining of the tumour cells showed they were diffusely positive for S100 (Figure 5) with CD34 highlighting interspersed fibroblasts only at the edges of tumour which is indicative of schwannoma. The tumour cells were negative for CKAE1/AE3, EMA, SMA and Desmin.

One month postoperatively, the patient was well and reported marked improvement in nasal congestion. There were no signs of recurrence seen after 3 months. Endoscopic examination of left nasal cavity appeared to be clear. The patient was later discharged well and remains asymptomatic.

DISCUSSION

Schwannomas are benign peripheral nerve sheath tumours that arise from the Schwann cells of the myelin sheath. These tumours are slow-growing, and rarely undergo malignant transformation.[6,7] The presentation varies and depends on the anatomical location and specific nerve involved.[1] There is no racial or gender predilection and these tumours can occur at any age, but they most commonly occur in the 30s to 60s.[6] Only 32 reported cases of nasal septal schwannomas have been documented in the literature.[5] This tumour most frequently involves the posterior third of the septum which comprises 60% of nasal septal schwannomas, followed by the middle and anterior parts.[8]

The most common symptoms at presentation are nasal obstruction (92.4%), epistaxis (30.3%), and rhinorrhea (18.1%) 9. Other symptoms include headache, deformity of the nasal pyramid, loss of smell, hyposmia, facial swelling and pain.[4,10] Of the sinonasal masses arising in the nasal septum, 87.5% are benign neoplasms while another 12.5% are malignant. The differential diagnosis for lesions arising from the nasal septum include pyogenic granuloma, inverted papilloma, schwannoma, and mucosal melanoma.[11] Other causes that may mimic the tumour include nasal polyps, neuroblastomas, angiofibromas, neurofibromas, and meningiomas.[1]

The diagnosis of schwannoma is confirmed by histopathological examination due to the lack of specific radiological features. Computerized tomography (CT) scans are fundamental to determine the origin and extent of the tumour, but it cannot differentiate tumour from infections and retained secretions.[10,12] Magnetic resonance imaging (MRI) scans on the other hand provide better soft tissue resolution than computerized tomography allowing for the differentiation of tumours from inflammatory disorders and normal tissue. MRIs also provide better visualization of any intracranial extension of the tumour.[13]

On MRI, sinonasal schwannomas appear to have intermediate intensity on T1 weighted images and intermediate to high intensity on T2 weighted images contingent on the characteristic of the tumour whereas in the computerized tomography (CT) scan of the sinonasal schwannoma typically shows homogeneous heterogeneous enhancement with contrast, homogenous soft tissue density, or bony erosion.[13,14]

The gold standard for the diagnosis of schwannomas is by histopathological examination.[5] Macroscopically, Schwannomas are usually encapsulated tumour with various shapes including oval, round and fusiform. They appear fleshy, and can be glistening grey to yellow in color.

Microscopically, they typically have two distinct morphologic areas, namely Antoni A and Antoni B. Antoni A refers to more cellular areas with spindle cells forming fascicles and often palisades around eosinophilic cytoplasmic processes known as Verocay bodies, while Antoni B are hypocellular myxomatous areas.[12,15] The tumour cells have spindled to ovoid nuclei which appear wavy with tapered ends and intranuclear inclusions may be observed. Immunohistochemical studies of Schwannomas typically diffusely express S100 compared to Neurofibromas which show more heterogenous expression.[16,17] The tumour described in this case showed almost exclusively cellular Antoni A areas with typical nuclear features but absence of well-formed Verocay bodies. Diffuse expression of S100 further supported the diagnosis.

Treatment of this tumour depends on the location, size and the invasion of the surrounding structures. The gold standard treatment for sinonasal schwannoma is complete surgical excision which can be achieved via endoscopic or open approach.[9,10] Open surgical approaches include lateral rhinotomy, Denker's operation, trans palatal incision, gingivobuccal incision, or midfacial degloving are used for more extensive tumours.[9] In current era of endoscope, excision of the tumours is generally undertaken endoscopically, and is more preferred than open surgery.[10] It is safe, effective and confers the

advantage of providing a shorter duration of hospital stay and it is also a minimally invasive procedure without any external incision needed . This treatment provides a good prognosis with complete resection of the tumour and postoperative recurrence is highly unlikely.[10,17]

CONCLUSION

In conclusion, sinonasal schwannomas are rare benign peripheral sheath tumours which can manifest as unilateral nasal congestion, epistaxis or nasal discharge. Despite being rare, this tumour should be considered as a differential diagnosis in patients presenting with a unilateral nasal mass. Diagnosis is established by histopathological examination of the tumour morphology and supported by immunohistochemistry. The treatment is achieved by complete resection of the tumour with excellent prognosis and low rate of recurrence is reported following excision.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Azman AB, Gill RS, Marimuthu SG, Kodiappan R, Nair SB. Unveiling the unseen: anterior nasal septal schwannoma. Int J Otorhinolaryngol Head Neck Surg 2025;11:75-8.