Schwannoma of nasal septum: A rare case report with literature review

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Abstract Schwannomas are benign tumors of nerve sheath and quite uncommon in the nasal septum. In contrast to the earlier reports in the literature, a confounding case of a nasal septal schwannoma presenting as a symptomatic growth on the left side of nasal septum in a 24-year-old man is discussed here.

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1. Introduction

Schwannomas (neurilemmomas or neurinomas or perineural fibroblastoma) are benign encapsulated nerve sheath neoplasms composed of Schwann cells first described by Verocay in 1908.1 Stout (1935) coined the term neurilemmoma believing that this tumor arose from cells of sheath of Schwann (neurilemmoma) which may also develop in any part of the body. This tumour most frequently originating from the acoustic nerve in the head and neck region, 45% of all schwannomas occur in body and 25% as extracranial schwannomas.2 It has also been observed in the neck, pharynx, larynx, scalp, face, oral cavity, middle ear, and internal auditory canal. However, involvement of the nasal cavity, paranasal sinuses and especially the nasal septum is rare. Incidence is 1 in 3000 schwannomas.3 Tumors arising from nasal septum are extremely rare with only 16 cases having been reported in the literature.4–11 A case of neurilemmoma of nasal septum was first described by Bogdasanian and Stout.5

2. Case details

A 24-year-old man was referred to our department with a 6-month history of progressive left-sided nasal obstruction, rhinorrhea, epistaxis. There was no history of anosmia, facial pain, headache and recent nasal trauma. Patient was neither suffering from any comorbid diseases nor reported any such family history. Anterior rhinoscopy revealed a large polypoid mass almost completely filling the left nasal cavity. The polyp was firm in consistency and appeared to be covered by normal
Figure 1  CT scan image of the nasal septal mass.

Figure 2  Photomicrograph showing low power schwannoma lying just beneath the septal cartilage[A–D], with the presence of Verwucay Bodies[C] and surrounding mucosal glands[D]. H&E 10x.
nasal mucosa. It bleeds on touch. The attachment of the polyp was difficult to determine. The nasal septum was slightly deviated to opposite side and right nasal cavity was clear. A CT scan of the paranasal sinuses was advised to determine the nature and extent of the polyp. This showed a homogeneous mass measuring $30 \times 16 \times 15$ mm, lying within the midportion of the left nasal cavity, arising from the left side of nasal septum (Fig. 1). The lesion was well defined with smooth margins and without calcification (Fig. 2). The paranasal sinuses were clear. A contrast study was not performed due to the absence of bony destruction. The patient agreed and consented to undergo removal of the polyp under general anesthesia. The polypoid mass was attached to middle part of the left side of the nasal septum, opposite the anterior end of the middle turbinate. The mass was excised completely along with a cuff of septal mucoperichondrium. Histopathological examination shows both spindle shaped schwann cell rich area with nuclear palisading (Antoni A) and schwann cell poor loose myxoid areas (Antoni B). Verocay Bodies are present without any evidence of ancient or malignant changes. The histopathological diagnosis was benign schwannoma (Figs. 2 and 3). Immunohistochemistry shows tumor cells are strongly positive for S-100 protein (Fig. 4) confirming the diagnosis and distinguishes neurilemmoma from neurofibroma. The patient was under follow up for six months and there was no recurrence at six months.

3. Discussion

The neural origin of the schwannomas is considered to be from peripheral motor, sensory, sympathetic and cranial nerve sheath. The optic and olfactory cranial nerves are not potential sites of the origin, since they lack sheaths that contain schwann cells and since nasal schwannomas are sometimes removed without loss of original nerve functions, it is usually difficult to determine the neural origin. Nasal schwannomas are presumed to be arising from the sheath of the ophthalmic and maxillary branches of the trigeminal nerve and autonomic ganglia. Schwannomas of the sinonasal tract are very infrequent, representing less than 4% of the schwannomas of the head and neck. In this location they have been reported in patients between the ages of 6 years and 78 years. There is no sex or racial predilection. The ethmoidal sinus is most commonly involved, followed by the maxillary sinus, nasal fossa and sphenoid sinus. Localization to the nasal septum is exceedingly rare. Septal schwannomas arise from the autonomic or sensory nerves within the nasal septum. There is no apparent site predilection on the septum.

Symptoms are non-specific and are the result of the mass effect or tumor necrosis. Patients may present with nasal obstruction, rhinorrhoea, or recurrent epistaxis, as for this patient. Nasal obstruction is the most common clinical symptom followed by epistaxis. Facial swelling and pain are associated with paranasal sinus involvement. These lesions rarely undergo malignant transformation.

Computerized tomography (CT) delineates an image of the soft tissue tumor and simultaneously outlines the skeletal margins well enough to rule out invasion and demonstrated central lucency and peripheral enhancement after contrast administration in case of schwannomas because peripheral neovascular areas of the tumor are enhanced in contrast with nonenhancing necrotic or cystic regions. Although magnetic resonance imaging (MRI) is superior in defining soft tissue tumors, CT offers better resolution of bony invasion. However, as benign schwannoma can erode bone by pressure, bony erosion is not a criterion for malignancy. With either approach, final diagnosis rests solely on the histologic examination. Micro-
scopically, schwannomas can exhibit two architectural patterns, Antoni type A and Antoni type B, in different proportions. Antoni type A area is composed of an organized compact cellular stroma with elongated spindle cells. Parallel rows of palisading nuclei can be seen in this highly differentiated tissue. Antoni type B area is composed of disorganized loose myxoid stroma with few spindle cells. Additionally, schwannomas usually show intense immunostaining for S100 protein (particularly Antoni A areas), which may help to distinguish peripheral nerve sheath neoplasms from other tumors.

The nasal septum appears to be the most common location for this lesion. The main differential diagnoses of nasal masses with similar MRI/CT findings as schwanna include nasal polyp, malignant peripheral nerve sheath tumors (MPNST), myxoma & fibromyxoma, Lobular Capillary hemangioma (LCH) and sarcoma. Nasal polyps represent hyperplasia of the mucosa in response to chronic inflammation, usually from chronic sinusitis. Nasal polyps usually have high signal intensity on T2-weighted images, which helps to distinguish them from tumors. Myxoma & fibromyxoma are benign neoplasms of uncertain histogenesis with characteristic histologic appearance. When relatively a greater amount of stromal collagen is present, the term fibromyxoma (or myxofibroma) is used. Histologically these tumor show scanty, loose cellular proliferation containing spindle-shaped or stellate-appearing cells embedded in abundant mucinous stroma. This tumor can manifest as a well-margined mass but may also show local infiltration and local invasion.17 In patients with a known history of NF, a nasal mass with such MRI/CT findings provides a clue for this diagnosis of schwanna,18 which requires confirmation by histology. MPNS tumors show features of malignancy, high mitotic count and with or without heterologous elements which will be lacking in schwannomas. Other sarcomas similarly show higher pleomorphism with absence of neural differentiation. LCH is most often found in the anterior portion of the nasal septum.19,20 Histologically LCH is characterized by submucous vascular proliferation arranged in lobules or clusters composed of central capillaries and smaller ramifying tributaries.

The only treatment for schwanna is wide local excision through an approach allowing adequate exposure as schwanna are generally radioresistant.4,5 But in treating benign schwanna, functional and cosmetic considerations should be taken into account. Recently, the technique of endoscopic nasal surgery has rapidly developed and transnasal endoscopic excision of benign tumors of the nose, paranasal sinuses and nasal septum has been successful.14,17 The tumor mass in our case was limited to the septal mucosa and successfully excised by transnasal endoscopic approach. A single schwanna does not recur when completely excised, but intracranial extension of a nasal schwanna has been reported.21,22

4. Conclusion

We report a 24-year-old male patient with nasal septal schwanna which was successfully treated by transnasal endoscopic excision. Although schwanna of the nasal septum is extremely rare, the possibility of their existence should be realized and included in the differential diagnosis of any nasal mass.

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