Case Report
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Nasoethmoid Ancient Schwannoma with Bilateral Intracranial Extension: A Case Report with Review of the Literature

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Abstract

Introduction:

Nasoethmoid schwannomas are rare benign tumors of the nasal cavity and paranasal sinuses that arise from local peripheral nerve sheaths. Up to 45% of all schwannomas develop in the head and neck region; however, fewer than 50 cases have been described in the nasoethmoid area in the medical literature. This case underscores the importance of including schwannoma in the differential diagnosis of nasoethmoidal masses with skull-base involvement.

Case Report:

A 60-year-old female presented with diminished sensitivity to smell for 12 months, swelling near left medial canthus and headache for the past 10 months. Nasal endoscopy revealed a smooth encapsulated mass in the anterior roof of nasal cavity between middle turbinate and septum bilaterally. CT and MRI revealed a smooth mass in the roof of nasal cavity having intracranial extra-axial extension. Biopsy displayed features of schwannoma. Patient underwent excision of tumor via combined approach of endoscopy and bifrontal craniotomy. Post-operative histopathology was consistent with features of schwannoma.

Conclusion:

Nasoethmoidal schwannoma is a benign neoplasm that infrequently exhibits intracranial extension. To date, there have been 17 documented cases in the literature, and this report presents the 18th reported case overall and 6th bilateral case. Because radiographic results are ambiguous, a histological investigation is required for a precise diagnosis. Bifrontal craniotomy combined with endoscopic transnasal excision is the preferred surgical treatment for tumors that extend into the anterior cranial fossa. Long-term clinical follow-up is critical for detecting future recurrences.

Keywords: Ancient schwannoma, Bifrontal craniotomy, Frontoethmoid schwannoma, Nasoethmoid schwannoma with intracranial extension

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Introduction

Nasoethmoid schwannomas are rare benign tumors of the nasal cavity and paranasal sinuses arising from local peripheral nerve sheaths. Ancient schwannomas are long-standing variants of these tumors that demonstrate degenerative changes, including cystic degeneration, hyalinization, calcification, and nuclear atypia. Although up to 45% of all schwannomas develop in the head and neck region, fewer than 50 cases have been described in the nasoethmoid area in the medical literature (1). Among these 50 cases, only a small subset of cases extend intracranially to involve the orbit or anterior cranial fossa. The precise origin of nasal schwannomas is difficult to determine due to the sparse and intricate neural supply of the nasal cavity, making intraoperative identification of the nerve of origin challenging. Sympathetic, parasympathetic, or sensory nerves are considered as their possible origin (2,3).

The majority of these lesions are benign, encapsulated, and spread locally by eroding the surrounding bony structures. To the best of our knowledge, only 17 cases of nasal schwannoma with intracranial extension have been reported in the medical literature to date. We hereby report the 18th case of nasal schwannoma with intracranial extension with relevant clinical, surgical and histological findings. Additionally, we conducted a comprehensive review of all previously documented cases of nasoethmoid schwannoma with intracranial extension.

Case Report

A 60-year-old female presented to the ENT out-pat of a tertiary care centre with complaints of diminished sensitivity to smell for 12 months, swelling near left medial canthus and headache for the past 10 months. The swelling was 2cmx1cm in size, insidious in onset, gradually progressive, not associated with pain or fever. It was not associated with any nasal blockade, mouth breathing, sneezing, nasal discharge or bleeding. Patient denied symptoms of excessive lacrimation, visual disturbances, earache, loss of sensation over cheek, difficulty in speech, loss of nasal twang or history of trauma. There was no history of any surgical procedure in past. Patient had no known comorbidities or similar complaints in past. General examination revealed no abnormality. On local examination, there was a 2cmx1cm diffuse swelling between the dorsum of nose and left medial canthus which was associated with flattening of nasal bridge (Figure. 1).

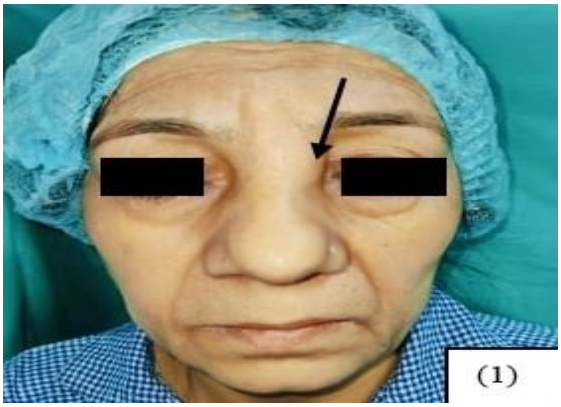

Fig. 1. swelling on the left nasal bridge and flattening of nasal bridge.

Swelling was smooth, soft, non-pulsatile, immobile, non-tender, non-fluctuant, non-compressible, non-reducible with diffuse margins and no overlying skin changes. There was no increase in size of the swelling with cough impulse. On performing diagnostic nasal endoscopy (DNE), a smooth well encapsulated mass between the nasal septum and left middle turbinate was noted. Similar mass was seen on the roof of right nasal cavity extending between the right middle turbinate and septum (Figure. 2 (a,b)).

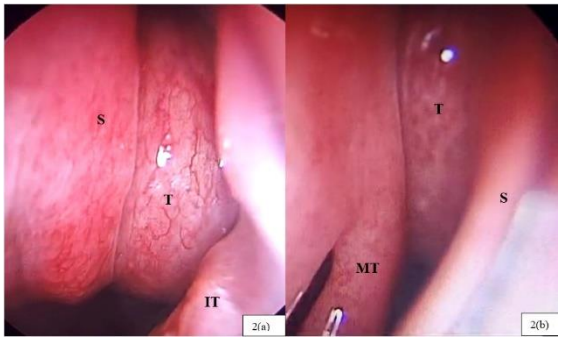

Fig. 2. (a) Left DNE displaying relation of tumor (T) to septum (S) and inferior turbinate (IT); **(b)**: Right DNE depicting tumor mass(T) between septum(S) and middle turbinate (MT)

Probe test revealed that the mass was firm in consistency, insensitive and did not bleed to touch. The site of origin of this nasal mass was speculated to be from the roof of nasal cavity on both sides. Bilateral nasal patency was present. Paranasal sinus tenderness was absent

on both sides. Eye, ear and oral cavity examination were unremarkable. No focal neurological deficit was present. On cranial nerve examination, all nerves except olfactory nerve were intact. Contrast enhanced computed tomography (CECT) of nose, paranasal sinus, orbit and head revealed a minimally enhancing soft tissue lesion involving superior nasal cavity and left frontal sinus with bony remodeling and focal areas of osteolysis involving bilateral ethmoidal lamella, lamina papyracea, anterior skull base, left middle turbinate, cribriform plate and bony nasal septum. Contrast enhanced magnetic resonance imaging (CE-MRI) revealed a well-defined, irregularly shaped, multi-lobulated soft tissue measuring 2.8x3.8x4.3cm epicentered in the region of superior nasal cavity causing expansile remodeling of bilateral anterior ethmoidal air cells (Figure.3(a,b,c)).

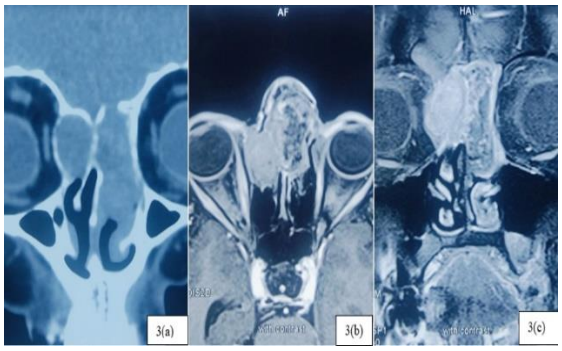

Fig. 3. (a) CT coronal image showing tumor in superior nasal cavity causing a bulging effect on right lamina papyracea; **(b,c)** MRI axial and coronal images respectively depicting a well-defined, multilobulated soft tissue in the region of superior nasal cavity and bilateral anterior ethmoids with extra-axial intracranial extension without intraorbital extension;

The lesion was hypo-intense on T1W images and heterogeneously hyperintense on T2/STIR images. Laterally, the lesion was causing bulge of the right lamina papyracea with no obvious intra-orbital extension and bulbous protrusion was seen at the level of left medial canthus. Posteriorly, the lesion was in close relation with the olfactory recesses. Superiorly, the lesion was extending into the extra-axial space of anterior cranial fossa in the basi-frontal region and causing disruption of CSF sleeve with smooth dural enhancement. Biopsy was taken from the nasal mass to establish tissue diagnosis which revealed it to be an ancient schwannoma with features of hypercellular Antoni A cells exhibiting nuclear palisading with presence of verocay bodies, hypocellular Antoni B cells, scattered hemosiderin laden macrophages and intense S-100 staining on immunohisto-chemistry (IHC) (Figure. 4 (a,b,c,d)).

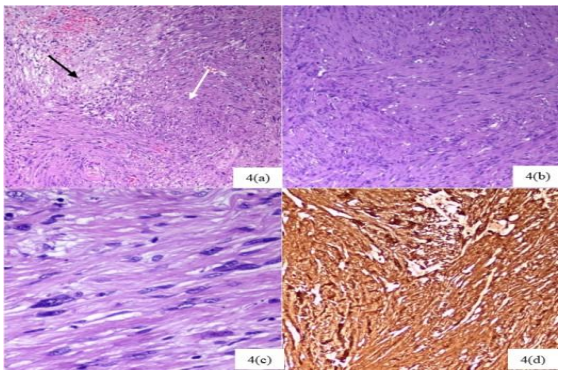

Fig. 4. (a) histopathological slide with Antoni A (white arrow) and Antoni B (black arrow) areas, **(b)** verocay bodies on 10x power, **(c)** atypia on 40x power, **(d)** intense cytoplasmic and nuclear staining for S100 marker

Patient was planned for excision of the nasal schwannoma under general anesthesia via a combined approach of endoscopic transnasal and bifrontal craniotomy with anterior skull base reconstruction by multi-disciplinary team (ENT and Neurosurgery). Following a bifrontal craniotomy, the tumor was exposed and noted to be an encapsulated pink-grey mass with solid consistency and moderate vascularity. The tumor was adherent to left frontal lobe dura and was removed by meticulous dissection (Fig. 5 (a,b)).

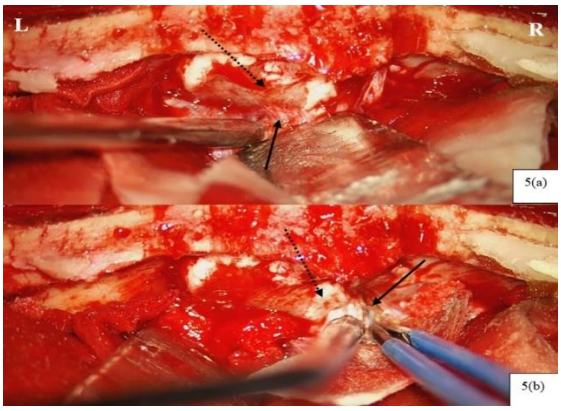

Fig. 5. (a) intraoperative image showing tumor (black dotted arrow) in the anterior skull base region tightly adhered to dura (black arrow) on right side, **(b)** tightly adhered dura (black arrow) on left side with tumor mass (black dotted arrow) with L and R depicting left and right respectively.

Major part of the tumor was excised by transcranial approach, but the tumor in the anterior nasal cavity was inaccessible by this route. Hence, nasal part of the tumor was excised after middle turbinectomy with endoscopic transnasal approach to achieve a gross total resection. Post excision of the tumor by this combined approach, anterior skull base was reconstructed transcranially by carpeting layers of fat, fascia, pedicled pericranial flap and tissue glue. Post-operatively, patient was kept on broad spectrum antibiotics for 2 weeks. No CSF leak was noted in the post-operative period. Hence, following an uneventful post-operative stay, patient was discharged and is in follow-up since then without any complication or recurrence. The patient provided informed consent for the publication of this report.

Discussion

Schwannoma, also known as neurilemmoma, is a benign tumor that develops from the cells forming the nerve sheath. It can originate from any nerve that possesses a Schwann cell covering, including most cranial nerves (with the exception of the optic and olfactory nerves), as well as nerves of the spinal and autonomic nervous systems. The earliest documented description of a schwannoma was published by Verocay in 1910 (3). Ackerman and Taylor in 1951 found that the schwannoma presented with clear areas of hypocellular tissues and attributed the changes to the prolonged degenerative changes. They coined the term “ancient” schwannoma for such type of benign neurogenic tumor. These degenerative features are attributed to the long-standing growth and "aging" of the tumor, hence the term “Ancient schwannoma”(4). Preoperative identification of the nerve of origin is difficult. There are three possibilities; the autonomic nerve and branches of the ophthalmic and maxillary nerve. In general, as nerves divide into smaller branches toward their peripheral ends, the density of Schwann cells rises, making the nasal cavity and paranasal sinuses more frequent sites for neurinoma development (2). The olfactory nerves are ensheathed by glial cells and lack schwann cells, therefore, cannot give rise to schwannoma (5). Localization allows sinonasal schwannomas to be categorized into four forms: subfrontal (involving the olfactory groove), nasoethmoidal (within the nasal and ethmoid regions), frontoethmoidal (chiefly intracranial with downward extension), and ethmofrontal (arising in the sinonasal tract with superior intracranial spread) (6).

The origin of schwannoma in our case is obscure. Its origin was probably an intranasal nerve, from which it gradually expanded, causing erosion of the cribriform plate and subsequent involvement of the anterior cranial fossa. Its highest prevalence is observed among people in their fourth to sixth decades of life, with no associated sex or racial predilection (3). The clinical presentation of nasal schwannomas often mimics that of inflammatory conditions of the sinonasal tract. Our review showed that headache, nasal obstruction, anosmia, epistaxis, and rhinorrhea were the most frequent symptoms, while nasal swelling, pain, exophthalmos, diplopia, and facial distortion occurred less often. In this case, anosmia pointed toward an origin in the olfactory cleft or ethmoid region, where the tumor’s presence likely obscured the olfactory apparatus and led to hyposmia. The differential diagnosis includes esthesioneuroblastoma, meningioma, papilloma, sarcoma, nasopharyngeal carcinoma, and lymphoma. Notably, nasoethmoid schwannomas do not display specific CT features and may be radiologically indistinguishable from esthesioneuroblastoma, fungal granuloma, or nasoethmoid carcinoma (7). CT imaging demonstrates central lucency with peripheral contrast enhancement in schwannomas. This pattern reflects the enhancement of peripheral neovascular regions, in contrast to the non-enhancing necrotic or cystic areas of the tumor. MRI surpasses CT in its ability to differentiate neoplastic tissue from normal anatomy due to superior contrast resolution, though CT remains the preferred modality for detecting bone destruction. MRI is particularly helpful in distinguishing tumors from inflammatory changes and assessing their extension beyond the nasal cavity. On T1-weighted sequences, the lesion usually displays intermediate signal intensity, while T2-weighted images may reveal intermediate to markedly hyperintense areas, reflecting differences in cellularity and cystic degeneration. These imaging findings assist in both diagnosis and in tailoring the surgical approach (3) Although histological confirmation is necessary, the biopsy should be carried out meticulously under

endoscopic visualization due to the risk of complications. A CSF leak has previously been reported after biopsy of an intranasal schwannoma (2). Because clinical signs and imaging appearances are often ambiguous, histopathology serves as the diagnostic gold standard. Under the microscope, schwannomas show two hallmark patterns: Antoni A tissue, made up of densely packed spindle cells exhibiting nuclear palisading around Verocay bodies, and Antoni B tissue, consisting of loosely dispersed, variably shaped cells within a myxoid matrix. Given the potential overlap with other lesions, immunohistochemistry especially strong reactivity for S-100 protein is useful in confirming the diagnosis (3). Although both schwannomas and neurofibromas are immunoreactive with the S-100 protein, Min and Kim reported that S-100 protein immunostaining was more intense on schwannomas than on neurofibromas (8). Moreover, Calretinin and CD-56 are also highly specific for schwannomas, while CD34 and factor XIII are more sensitive for neurofibromas. These markers further assist the differential diagnosis (3). Our case had an intense immunostaining for S-100 protein and negative for CD34, leading to the diagnosis of schwannoma.

The type of surgical procedure is determined by the tumor size, its position, and whether it has invaded nearby structures. For schwannomas that are confined to the nasal cavity, the typical approach is transnasal excision, which may or may not include the use of endoscopy. More extensive cases typically require open surgery. According to the literature, these tumors can be resected gross totally by bifrontal craniotomy. Among the 17 cases mentioned in literature, only one case was managed with subtotal resection and concurrent radiotherapy was given as it was malignant schwannoma on histopathology (9). Below is the summary of all 18 cases (including present case) of nasoethmoid schwannoma with intracranial extension (Table 1).

Table 1. Summary of all published cases of nasoethmoid schwannoma with intracranial extension.

Sr no.	Author	Year	Age (years)	Sex	Laterality	Clinical presentation	Surgical approach
1.	Zovickian et al.[5]	1986	40	M	Unilateral (Left)	Headache, nasal congestion	Frontal craniotomy with transnasal approach
2.	Enion et al.[10]	1991	28	M	Unilateral (Left)	Headache	Bifrontal craniotomy
3.	Bavetta et al.[11]	1993	41	M	Unilateral (Left)	Blurred vision, nasal blockage	Frontal craniotomy with transnasal approach
4.	Fujiyoshi et al.[12]	1997	38	M	Not specified	Epistaxis, nasal congestion	Not specified
5.	Gatscher et al.[13]	1998	50	F	Bilateral	Anosmia, headache, visual deterioration	Bifrontal craniotomy
6.	Sharma et al.[7]	1998	35	M	Bilateral	Anosmia, epistaxis, nasal obstruction, seizures	Bifrontal craniotomy
7.	Siqueira et al.[1]	2001	40	F	Bilateral	Anosmia, frontal deformity, headache	Bifrontal craniotomy with lateral rhinotomy
8.	Ogunleye et al.[9]	2006	31	F	Bilateral	Anosmia, midfacial swelling, unilateral blindness	Not specified
9.	George et al.[14]	2009	27	F	Unilateral (Right)	Blurred vision, headache	Frontal craniotomy
10.	Hong et al.[15]	2016	24	M	Bilateral	Incidental finding after road-traffic accident	Bifrontal craniotomy
11.	Eichberg et al.[16]	2017	41	M	Unilateral (Left)	Anosmia, headache	Bifrontal craniotomy
12.	Narang et al.[17]	2019	33	F	Unilateral (Left)	Left eye proptosis, epistaxis	Frontal craniotomy with transnasal approach
13.	Brahmbhatt et al.[18]	2003-2022	59	F	Not specified	Headache, Fullness	Endoscopic resection
14.			58	M	Not specified	Headache, Sinusitis	Endoscopic resection
15.			33	F	Not specified	Nasal Obstruction	Endoscopic resection
16.			49	F	Not specified	Dizziness, Nonspecific Neurologic	Craniotomy
17.			43	M	Not specified	Headaches, Incidental finding	Endoscopic resection
18.	Present case	2025	60	F	Bilateral	Headache, hyposmia, bulge at left medial canthus	Bifrontal craniotomy with transnasal endoscopic approach

While schwannomas located in the nasal cavity, particularly those that extend intracranially, are uncommon, they should be considered in the differential diagnosis of lesions in the anterior cranial fossa. Typically benign, these lesions have a propensity to recur significantly if any small remnants remain, frequently damaging parts of the skull base. However, complete surgical excision is associated with an excellent prognosis. Long-term follow-up with periodic endoscopic examination and imaging, such as MRI or CT, is recommended to monitor for residual or recurrent disease, particularly in cases with intracranial extension or complex anatomical involvement.

Conclusion

Nasoethmoidal schwannoma is an uncommon benign neoplasm that may rarely extend intracranially. As imaging findings are often nonspecific, definitive diagnosis relies on histopathological and immunohistochemical evaluation. Extensive tumors are best managed using a combined bifrontal craniotomy and endoscopic transnasal approach, with ongoing long-term monitoring to identify any recurrence.

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