Nur Syakirah Raja Ariffin[1], Muhammad Nasri Abu Bakar[1], Aifaa Abdul Manan[1], Norsyamira Aida Mohamad Umbaik[2*], Norasnieda Md Shukri[2]

[1] Department of Otorhinolaryngology, Hospital Sultan Ismail Petra, 18000 Kuala Krai, Kelantan, Malaysia.
[2] Department of Otorhinolaryngology-Head and Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia.

Sinonasal Ancient Schwannoma Presenting as Unilateral Blindness

Abstract— Schwannomas, also known as neurilemmomas, are benign nerve sheath tumors originating from Schwann cells of the peripheral nervous system. Sinonasal schwannomas are rare, comprising approximately 4% of all schwannomas. These tumors are typically slow growing, often leading to delayed clinical presentation. Despite their benign nature, their indolent progression can result in significant morbidity due to mass effect on adjacent vital structures. We report a case of a large, late-diagnosed right sinonasal ancient schwannoma causing extensive compression of surrounding tissues, including the right frontal and temporal lobes, ultimately resulting in vision loss due to compressive optic neuropathy. Ocular symptoms may be the initial manifestation of sinonasal tumors and should raise concern for potentially aggressive lesions. Prompt recognition and early referral are critical to minimize complications and improve patient outcomes.

Received 20 Sept 2024
Revised 23 May 2025
Accepted 22 June 2025
Published Online 01 Aug 2025

Keyword – **Sinonasal, Schawannoma, Neurilemmoma, Ischemic optic neuropathy**
.

*Corresponding Author
Norsyamira Aida Mohamad Umbaik
Email: syamira@usm.my

1. INTRODUCTION

Schwannomas are benign neoplasms derived from Schwann cells, which are responsible for the myelination of peripheral nerves [1]. While approximately 25–45% of all schwannomas occur in the head and neck region, only about 4% are in the sinonasal tract, making sinonasal schwannomas an uncommon clinical entity [2,3]. The most frequently encountered schwannomas in the head and neck region are vestibular schwannomas—formerly known as acoustic neuromas— which arise from the vestibulocochlear nerve (cranial nerve VIII) [4]. In contrast, schwannomas originating from the nasal cavity and paranasal sinuses are rare, often presenting with nonspecific symptoms such as nasal obstruction, rhinorrhea, or facial pressure. These symptoms may mimic those of chronic rhinosinusitis, potentially delaying accurate diagnosis and timely management.

A histological subtype known as “ancient schwannoma” refers to tumors that have undergone degenerative changes due to long-standing growth [5]. Although benign, these tumors may reach considerable sizes over time, resulting in pressure necrosis of adjacent bony structures, as well as compression of surrounding soft tissues, leading to functional impairments [6]. Surgical excision remains the mainstay of treatment, offering an excellent prognosis when complete removal is achieved. However, the surgical approach must be carefully selected based on the tumor’s size, location, and extent of involvement with adjacent anatomical structures [7].

2. CASE PRESENTATION

A 61-year-old woman with a history of hyperlipidemia presented with progressive right eye proptosis and vision loss over the past year. She also reported persistent right-sided nasal obstruction, hyposmia and yellowish nasal discharge of the same duration. As the right proptosis progressed, the patient began experiencing epiphora in the right eye. However, she denied systemic symptoms such as fever, headache, or limb weakness. Due to the COVID-19 pandemic, she was not able to seek treatment

early. After the movement restriction was over, she sought medical advice from nearest health clinic. Initially referred to the Ophthalmology team for suspected cataract by the primary care doctor, assessment revealed no light perception in the right eye, likely secondary to ischemic optic neuropathy. She was subsequently referred to our team to evaluate the right nasal obstruction, which was suspected to be related to her ocular symptoms.

On examination, there was evident proptosis of the right eye, resulting in incomplete eyelid closure. Ocular movements were restricted in all directions, the corneal reflex was absent, and the relative afferent pupillary defect (RAPD) test was positive. Nasoendoscopy revealed a medialized medial wall of the right maxillary sinus with a reddish nasal mass located between the septum and the medialized wall (Fig. 1). The surface of the mass appeared ulcerative and bled upon contact during probing. Minimal mucoid discharge was also noted over the mass. Further assessment beyond the mass was not possible due to total blockage of the posterior nasal cavity by the mass. Left nasal cavity and visualized nasopharynx were clear.

Computed tomography (CT) of the brain, orbit, and paranasal sinuses demonstrated a large lesion occupying the right sinonasal region, extending posteriorly into the infratemporal fossa and superiorly into the middle cranial fossa, with extensive bony erosion seen at the pterygoid plate and skull base areas (Fig. 2 and 3). Antero-superiorly, the mass involved both the intraconal and extraconal spaces of the right orbit, displacing the right globe, extraocular muscles, and optic nerve. Antero-inferiorly, there was bowing of the anterior wall of the right maxillary sinus. Medially, the mass filled most of the nasal cavity, as well as the ethmoid and sphenoid sinuses, and extended to the posterior choana. Magnetic resonance imaging (MRI) of the brain and paranasal sinuses revealed an isointense mass on both T1- and T2-weighted sequences.

Post-contrast images demonstrated strong, heterogeneous enhancement of the lesion. The mass was seen compressing the right frontal and temporal lobes; however, there was no evidence of intradural extension.

An endoscopic biopsy of the right sinonasal mass was performed under general anaesthesia due to the lesion's friability and tendency to bleed upon contact during clinic examination. Histopathological analysis revealed characteristic Antoni Type A and B, with no evidence of malignancy. Immunohistochemical staining demonstrated strong positivity for S100 protein, confirming the diagnosis of sinonasal ancient schwannoma. Given the complexity of the case, a multidisciplinary team discussion was conducted involving the Otorhinolaryngology, Ophthalmology, Neurosurgery, and Oromaxillofacial Surgery teams.

The consensus was to proceed with debulking surgery via combined endoscopic and open approach. However, the patient refused surgical intervention and subsequently defaulted on follow-up.

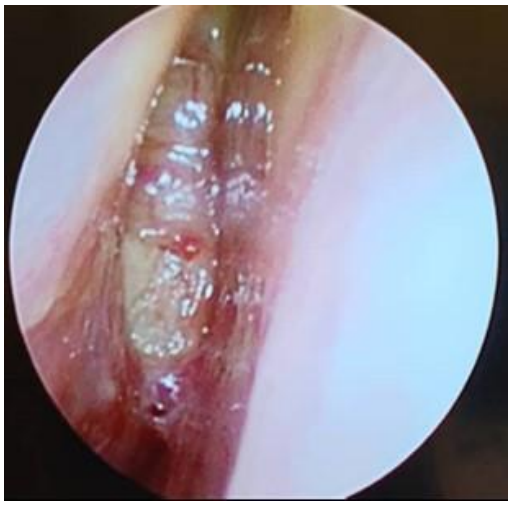

Figure 1: Endoscopic view of the right nasal mass with medialized medial wall of maxillary sinus.

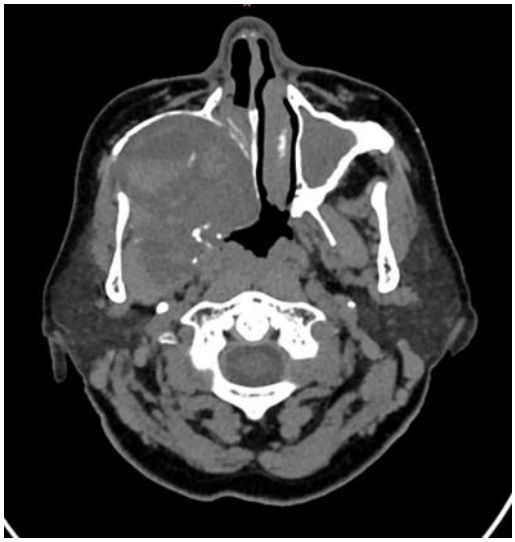

Figure 2: Axial view of CT brain showing large sinonasal mass arising from the right maxillary sinus extending superiorly to ethmoid sinus, medially obliterating ipsilateral nasal cavity, posteriorly to infratemporal fossa and anteriorly causing bowing of maxillary sinus wall.

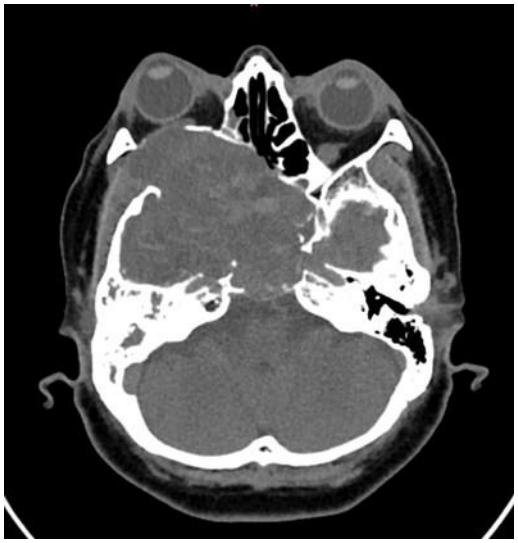

Figure 3: Superior cut on the CT brain showing the sinonasal mass extending to the orbital apex compression on the optic nerve region, eroding skull based and extends intracranially.

3. DISCUSSION

Sinonasal schwannoma (SS) is postulated to originate from the ophthalmic and maxillary divisions of the trigeminal nerve, or the neural supply to the septal vessels and mucosa. The nasal septum is the most frequently reported site of origin [8]. In the present case, the tumor is most likely derived from the maxillary branch of the trigeminal nerve.

The clinical presentation of sinonasal schwannoma varies based on the tumour's location. Most patients present with rhinological symptoms such as unilateral nasal obstruction, epistaxis, olfactory disturbances, and facial pain [8,9]. In this case, the patient had prolonged nasal symptoms that were initially disregarded until visual disturbances emerged, indicating the orbital involvement. The gradual loss of vision was initially misdiagnosed as cataract, while the nasal complaints were mistakenly treated as chronic rhinosinusitis at the primary care level. In its early stages, SS may resemble chronic rhinosinusitis. As the tumor enlarges, it may exert compressive and destructive effects on surrounding structures, including the orbit and intracranial compartments [8].

Complete surgical excision is the treatment of choice for SS, offering favorable outcomes and an overall good prognosis. Surgery is crucial not only to alleviate symptoms but also to address the potential risk of malignant transformation. The selection of the surgical approach is guided by the tumor's size, location, and extent of involvement. Recent advances in endoscopic technology and refinements in endonasal techniques have enabled the successful endoscopic resection of many sinonasal tumors. Endoscopic transnasal complement with transorbital and/or transcranial approaches is selected based on tumor extension [9]. In this case, however, a debulking procedure with combined approach was considered more suitable due to the extensive nature of the tumor, with involvement near the vital structures such as the internal carotid artery and invasion of the orbital compartment.

CyberKnife radiosurgery has emerged as a viable alternative to surgical intervention, particularly for patients who are unfit for or decline surgery. Its effectiveness has been demonstrated in both extracranial facial schwannomas and intracranial schwannomas, including vestibular, trigeminal, and facial nerve schwannomas [10,11]. In a study by Sasaki et al., an extracranial facial schwannoma showed a 23% reduction in size over 15 months following a three-day course of fractionated CyberKnife

radiosurgery [10]. A meta-analysis conducted by Mahboubi et al. reported a tumor control rate of 96.3% for vestibular schwannomas treated with CyberKnife radiosurgery [11]. In other hand, a meta-analysis by Yang et al. reported a tumor regression rate of 88.75% following CyberKnife radiosurgery for trigeminal schwannomas [12]. Thus, for appropriately selected patients, radiosurgery offers a non-invasive treatment option that can effectively control tumor growth, minimize the risk of progressive mass effect, and potentially achieve tumor regression or cure in some cases.

4. CONCLUSION

Sinonasal schwannoma, although benign, can grow extensively and lead to significant complications such as visual impairment. Early recognition is crucial, and clinicians should maintain a high index of suspicion by obtaining a detailed history, particularly of nasal symptoms, and performing a thorough physical examination. This case highlights the need for early referral to an Otorhinolaryngologist in patients with persistent unilateral nasal obstruction to exclude the possibility of sinonasal polyps or neoplastic lesions.

REFERENCES

[1] Anshit Goyal, Allan J. Belzberg, H. Richard Winn (Ed.), Benign and Malignant Tumors of the Peripheral Nerve, Neurological Surgery (7th ed.), Elsevier (2016).

[2] Dosemane D, Kabekkodu S, Jaipuria B, Sreedharan S, Shenoy V. Extracranial non-vestibular head and neck schwannomas: a case series with the review of literature. Braz J Otorhinolaryngol. 2022 Nov-Dec;88 Suppl4(Suppl4):S9-7.doi:10.1016/j.bjorl.2021.05.013. Epub 2021 Jun 9. PMID: 34154970; PMCID: PMC9756070.

[3] Umbaik NA, Ramli R, Abdullah B. Endoscopic excision of nasal septal Schwannoma: Case report and review of literature. Bangladesh Journal of Medical Science. 2019;18(3):660.

[4] Serban O, Poanta LI, Mihaileanu F, Fodor D. Schwannoma of the Lower Leg with Malignant Transformation. A Case Report. European Journal of Case Reports in Internal Medicine. 2015 Jul 31;2(5).

[5] Kumar S, Sayoo C. Sinonasal schwannoma: a rare sinonasal neoplasm. Indian Journal of Otolaryngology and Head & Neck Surgery. 2017 Sep;69:425-7.

[6] Dhawle MS, Rathod SG, Bhatkule MA, Bindu RS. Sinonasal schwannoma–A case report. Journal of clinical and diagnostic research: JCDR. 2017 May;11(5):ED22.

[7] Lee CW, Grammatopolou V, Bagwan I, Sunkaraneni V. Schwannoma of the sinonasal tract: case report with review of the literature. The Annals of The Royal College of Surgeons of England. 202 Jul;103(7):e216-22.

[8] Riveros AC, Proietti V, Serrano C, Ruggeri CS. Sinonasal Schwannomas. K Otolaryngol Rhinol. 2021;7:108. doi.org/10.23937/2572-4193.1510108.

[9] Corvino S, de Divitiis O, Corazzelli G, Berardinelli J, Iuliano A, Di Domenico C, Lanni V, Altieri R, Strianese D, Elefante A, Mariniello G. Naso-Ethmoidal Schwannoma: From Pathology to Surgical Strategies. Cancers. 2025 Mar 22;17(7):1068.

[10] Sasaki A, Miyazaki S, Hori T. Extracranial facial nerve schwannoma treated by hypo-fractionated cyberknife radiosurgery. Cureus. 2016 Sep 21;8(9) doi:10.7759/cureus.797.

[11] Mahboubi H, Sahyouni R, Moshtaghi O, Tadokoro K, Ghavami Y, Ziai K, Lin HW, Djalilian HR. CyberKnife for treatment of vestibular schwannoma: a meta-analysis. Otolaryngology–Head and Neck Surgery. 2017 Jul;157(1):7-15.

[12] Yang S, Wang J, Li X, Li H, Zhong Y, Zhou X, Chen G. Gamma knife radiosurgery for trigeminal schwannomas: A systematic review and meta-analysis. Journal of Clinical Neuroscience. 2022 Jul 1;101:67-79.