

## CNS Neurinoma Presenting as Unusual Skull Base Tumors

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### 1. Abstract

Schwannomas are benign peripheral nerve sheath tumors found throughout the body with 4% possibility to arise in sinonasal region compared to other head and neck schwannomas. Due to their unpredictable trend to affect multiple sites of human organ systems, no specific signs or symptoms are recognized for these lesions and is based on affected location. In this study, authors described 2 cases of neurinoma that occurred in unusual parts of the skull base. Case 1 is a sinonasal schwannomas in a 53-year-old patient originating from sphenoid region to nasal cavity that treated by endonasal endoscopic trans-sphenoid resection approach with no post-operative complications and recurrence in 18-month post-operative follow-up period. Case 2 was a glossopharyngeal neurinoma that was presented with dysphagia and neck pain and located in the jugular foramen. The patient was operated on through infra labyrinthe approach and the tumor was totally resected with no complication and no recurrence after 2 year follow up.

### 2. Introduction

Schwannomas (Neurilemmas) are benign neoplasms that arise from Schwann cells that surround the peripheral, cranial, or autonomic nerve sheaths. These slow-growing encapsulated tumors arise when proliferating Schwann cells form a tumor that encompasses the nerve sheath [1]. These tumors may occur in any organ or nerve trunk with cranial nerves I and II exception due to their lack of Schwann cells. Also, distinguished syndromes may be associated with Schwannomas including Von Recklinghausen's disease [2].

Approximately, 25–48% of Schwannomas are reported in head and neck regions with a trend to affect patients in their second to fifth decade [3]. Also, no racial or gender preference have been found regarding these benign tumors. [4] Due to various presentation locations, no specific presentations are defined. However, tenderness to palpation and secondary neurologic symptoms are common presentations. Although schwannomas represent a relative common finding in head and neck, 4% occur in sinonasal cavity and are usually misdiagnosed with other lesions due to lack of specific clinical signs or symptoms [5]. In this report, authors will describe 2 case of CNS neurinoma that occurred in unexpected sites of the skull base. Case 1 is an intra-sphenoid schwannoma as a rare subclass of sinonasal neurinoma and case 2 is a glossopharyngeal neurinoma that was placed in the jugular foramen.

### 3. Case Presentation 1

A 53-year-old female presented to our outpatient clinic of department of neurosurgery complained of progressive headache, blurred vision and sense of nasal congestion in the past 6 months.

Patient's previous nasal endoscopic evaluation revealed a left-sided nasal septum deviation and a protruding boggy and hyperemic mass from the right posterior nasal wall. Patient's neurological examination revealed right sided hyposmia, decreased visual acuity (OD=20/120, OS=20/80) and decreased right visual field (Figure 1). Also, neurological survey did not reveal papilledema and facial asymmetry, along with normal ocular movement and gag reflex.

The patient underwent pre-operative Computed Tomography (CT) and Magnetic Resonance Imaging (MRI). Axial CT scan demon-

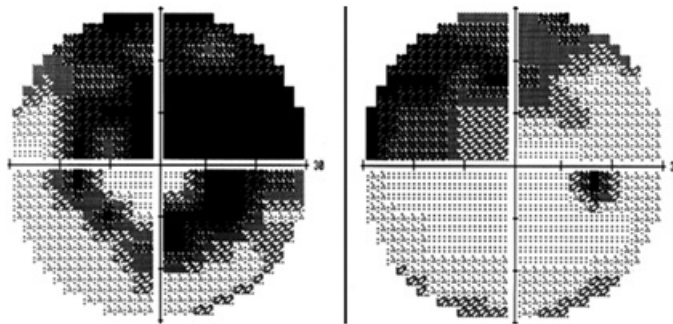
strated an irregular mass extending from sphenoid region to bilateral nasal cavities and nasal septal deviation (Figure 2).

Pre-operative MRI revealed a hypo intense lesion in T1 hypo intense irregular well defined lesion with mixed consistency protruding anteriorly from sphenoid region to nasal cavities (Figure 3). The lesion enhances vividly after contrast injection (Figure 4).

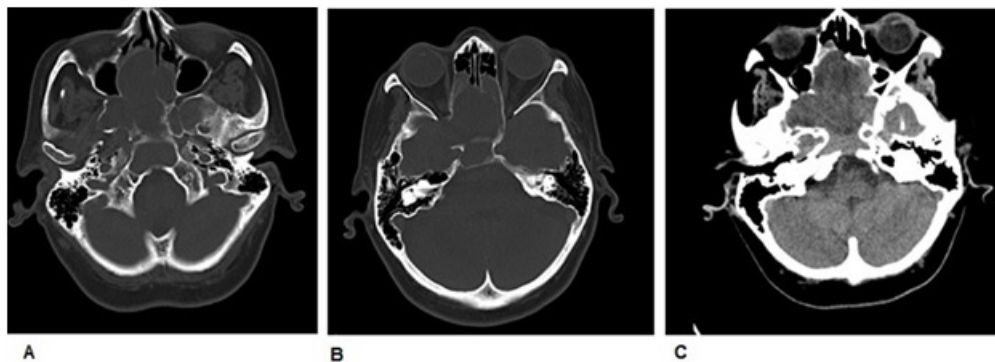
A trans-sphenoidal endonasal endoscopic surgery was performed and excised sample were sent for histopathological examination. The tumor was located within the sphenoid sinus and was multilobulated and destroyed the bony skull base and expanded the ethmoidal air cells and sphenoid sinus and had pressure effect over the optic recesses bilaterally. The tumor was adherent to the bony skull base and had fibrous consistency and rather vascular (Figure 5). The tumor was totally resected through endoscopic endonasal approach with the guide of neuronavigation. Microscopic sections showed sinus tissue with respiratory epithelium infiltrated by a

neoplasm composed of uniform spindle cells with elongated nuclei, scant eosinophilic cytoplasm arranged in diffuse growth pattern with rather whirling appearance. In addition, some hyalinized blood vessels, hemosiderin deposition and foamy histiocytes are also present. Also, no evidence of mitotic activity or necrosis was seen. Immuno Histo Chemistry (IHC) study revealed S100 strong positivity in conjunct with GFAP, Vimentin, CD68 (in background histiocytes) and CD99. Moreover, nuclear positivity for Ki67 was found in 2-4% of cells and final histopathological conclusion of Schwannoma.

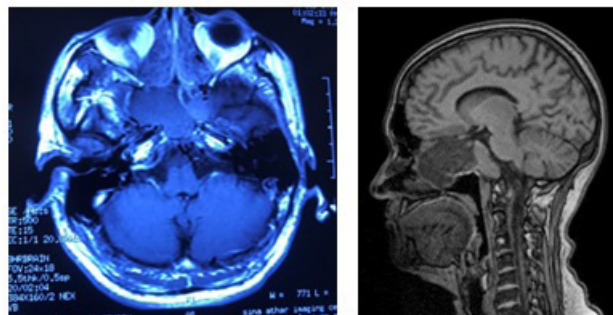
Post-operative MRI demonstrated decompressed sinonasal cavity and sphenoid region with no tumor remnant. (Figure 6) Also, post-operative neurological examination revealed no surgery related complications and during 18 months' follow-up period, no evidence of recurrence was observed. Written informed consent form was obtained from patient under ethical declaration of Helsinki.



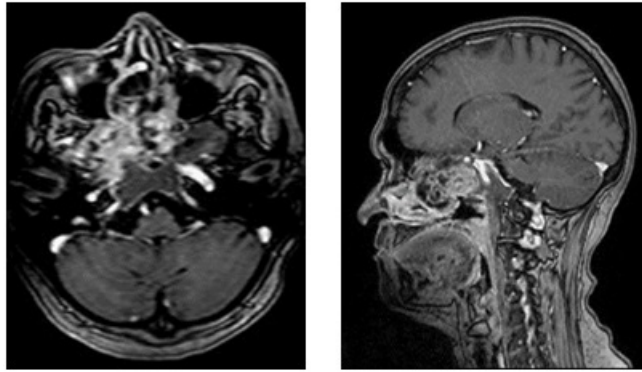
**Figure 1:** Visual field impairment of the patient 1



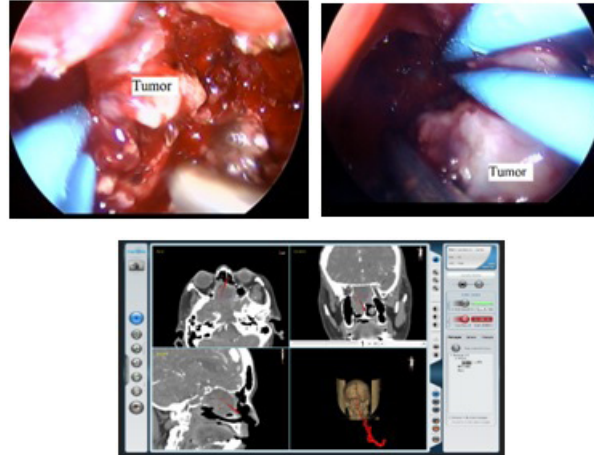
**Figure 2:** Pre-operative axial CT scan of the patient 1 that showed erosion and expanding of the ethmoidal air cells and sphenoid sinus.



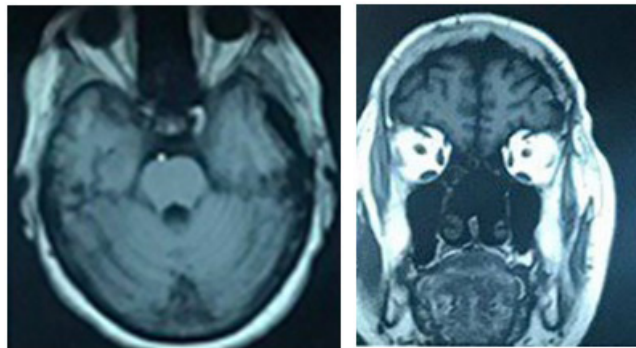
**Figure 3:** Pre-operative MRI without contrast, shows the skull base hypo to isosignal mass in the anterior skull base. Left: axial and Right: sagittal view.



**Figure 4:** Pre op MRI post contrast that shows the vividly enhanced tumor within the anterior cranial fossa. Left: axial and Right: sagittal view.



**Figure 5:** Left and middle: Intraoperative endoscopic view of the tumor within the skull base and Right: the navigation snap shot on the right which showed localization and the extent of the tumor.

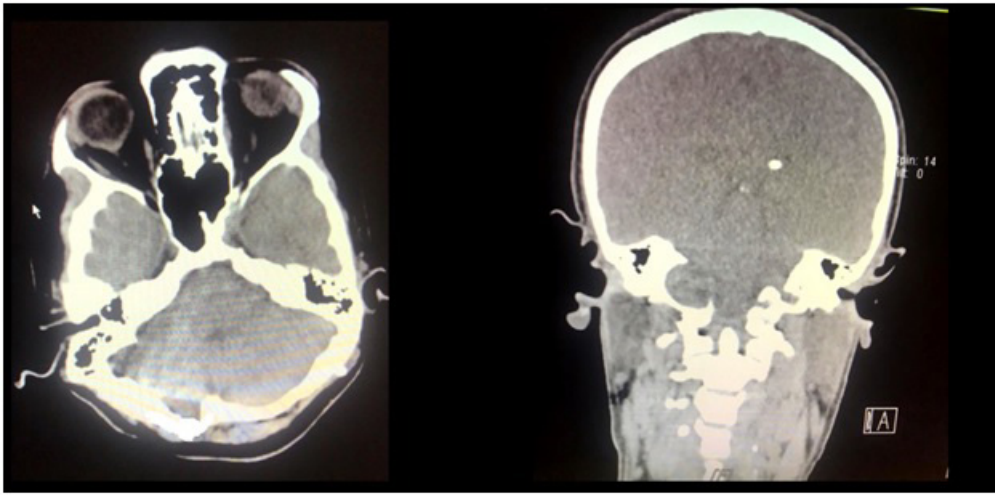


**Figure 6:** Post-operative MRI with Gad that shows total tumor resection. Left: axial and Right: Coronal View.

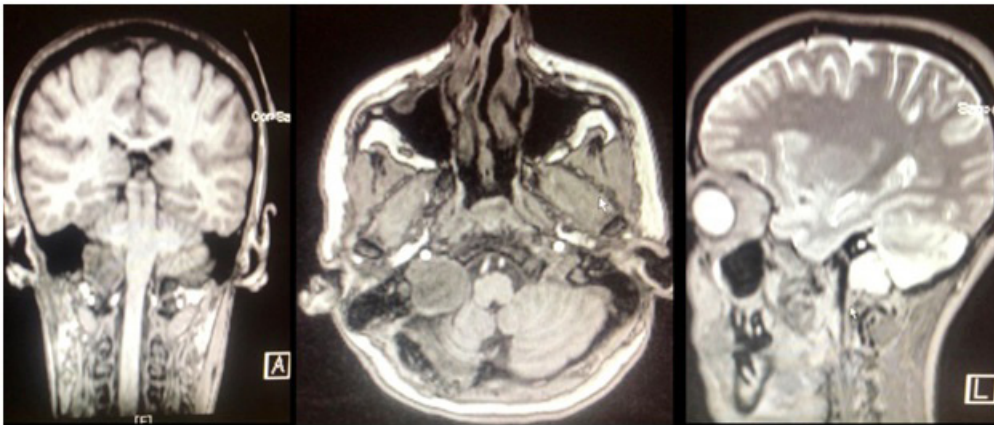
#### 4. Case Presentation 2

The patient was a 49 y old man, presented with headache, dysphagia and nuchal pain. Neuro exam showed impaired right sided gag reflex. Brain CT Scan showed dilated right sided jugular foramen (Figure 7). MRI study showed the jugular foramen mass which was hypo intense on T1 and hyper intense on T2 (Figure 8) and enhanced with contrast (Figure 9). The patient was operated through microscopic infra labyrinthine approach under the guide of navigation. Surgery: The incision was retro auricular. The mastoid was drilled and the mastoid antrum was opened. The drilling was con-

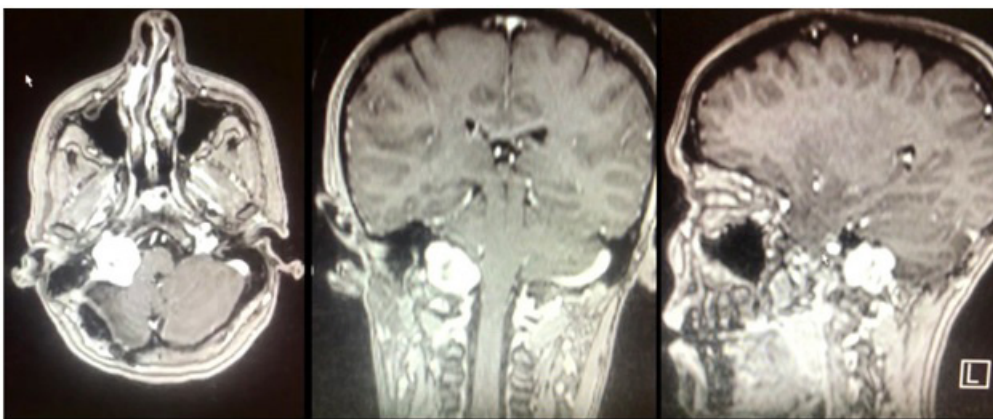
tinued under the guide of navigation in the sub cochlear region, above the jugular bulb and behind the internal carotid artery. The entry to the post fossa was done through Trautman triangle. The dura was opened and the tumor was exposed. It was grayish pink in color and had firm elastic consistency with relative vascularity. The tumor was totally removed and the 9th nerve was seen and preserved. The dura was repaired using duragen and fibrin glue and all mastoid air cells were waxed completely (Figure 10). Post Op CT scan showed complete tumor resection and the procedure was event free (Figure 11). No recurrence occurred during the 2 year follow up.



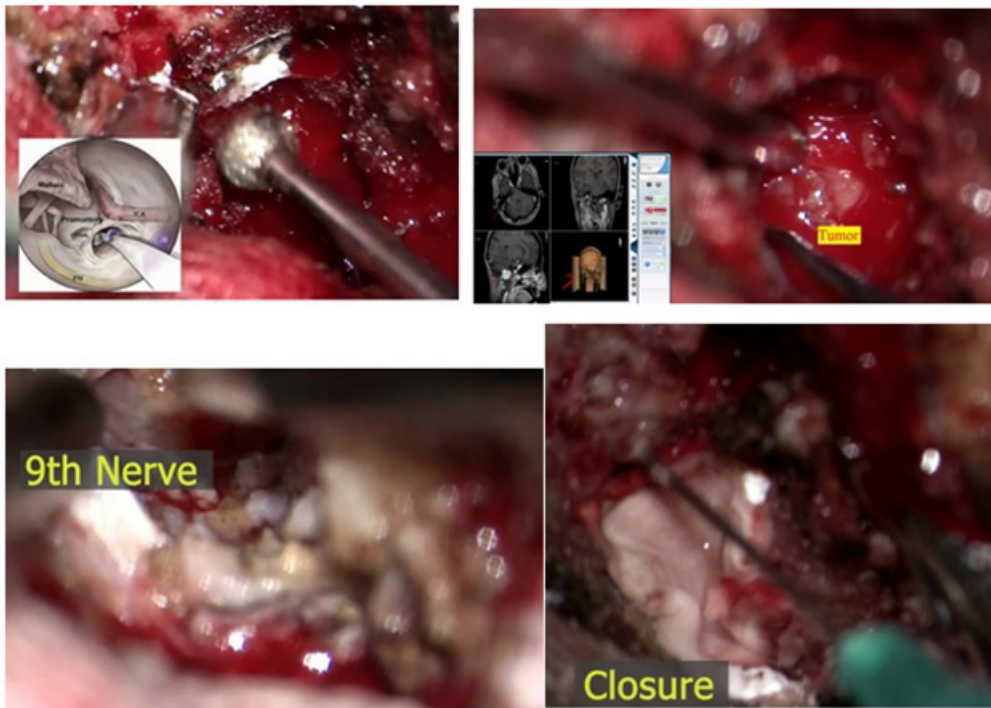
**Figure 7:** Brain CT Scan of the case 2, which showed enlargement of the jugular foramen and an iso dense mass Left: axial and Right: coronal.



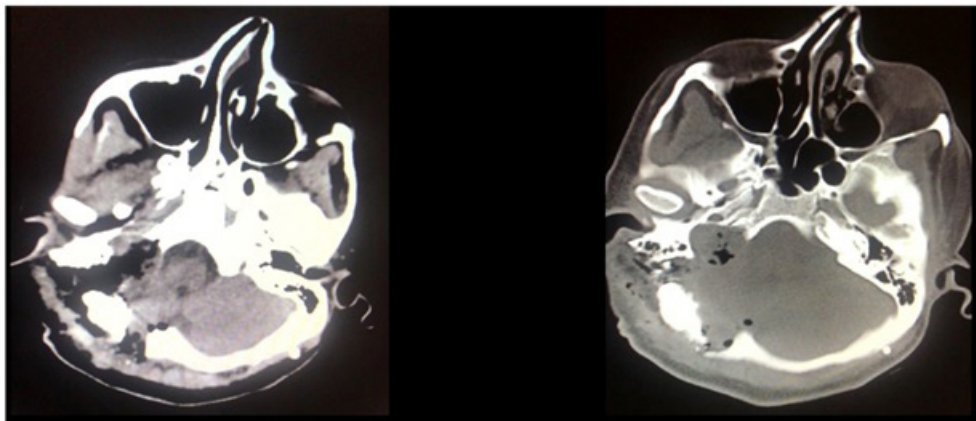
**Figure 8:** Brain MRI of the second case that showed an iso intense mass in T1 and hyper intense in T2. In the right jugular foramen. Left: coronal, Middle: axial and Right: sagittal view.



**Figure 9:** Brain MRI of the second case with contrast injection that showed vivid enhancement of the mass of the jugular foramen. Left: axial, Middle: coronal and Right: sagittal view



**Figure 10:** Intraoperative view of the second case. Upper left: Subcochlear drilling. Upper right: Tumor exposure. Lower left: Tumor originated from the 9th N. Lower right: Dural repair with duragen and fibrin glue.



**Figure 11:** Post op CT scan of the second patient which showed total tumor removal. Left: parenchymal view. Right: Bone view.

## 5. Discussion

Schwannomas essentially manifest themselves with a combination of cosmetic deformity, a palpable mass and compressive neuropathy manifestation. Late presentation of neurological symptoms and their vague nature frequently results in average 5-year delay prior to diagnosis [6]. Intracranial schwannomas are reported to be found in Cerebello Pontine Angle (CPA), para-cavernous sinuses and sinonasal cavity. Based on existing evidence, schwannomas originate from ophthalmic and maxillary branches of the trigeminal nerve or from autonomic nerves to the septal vessels and mucosa in the sinonasal cavity. Also, nasal cavity and ethmoid sinuses are more frequently affected with schwannomas compared to other sinuses [7]. However, another study revealed sinonasal schwannomas distribution in maxillary sinus, ethmoid sinus and frontal sinus in decreasing order [8]. Evidence suggests close proximity

with the pathway of cranial nerves and their division is a major contributor behind the statistics. For instance, lack of schwann cells of olfactory nerve may account for diminished likelihood of frontal sinus schwannoma [9].

Confronting a sinonasal lesion bring up polyps, papilloma, hemangioma, fibrous-osseous tumors, intrasellar adenoma and neurilemmoma as benign and Squamous Cell Carcinoma (SCC), adenocarcinoma, lymphoma, esthesioblastoma, angiosarcoma, chondrosarcoma, chordoma and plasmacytoma as malignant differential diagnosis. Schwannomas usually enhance variably and often demonstrate bone remodeling rather than cortex destruction on computed tomography.

Clival chordoma frequently presents as a midline sphenoid mass, but it is associated with cortex rarefaction, destruction of the sphenoid

noid or with mass protruding into the pre-pontine cistern. Chondrosarcoma is usually situated more eccentrically and contains matrix of calcifications [10, 11].

Schwannoma of the jugular foramen, is a very rare entity. It is originated from the ninth and 10th cranial nerves and constitutes only 2.9% of all intracranial schwannomas [12].

Jugular foramen schwannomas are sometimes difficult to differentiate from acoustic tumors. The other tumors located in the jugular foramen are glomus jugulare tumor, meningioma, choroid plexus papillomas, chordomas, exophytic pontine gliomas, chondromas, cerebellar hemangioblastomas, myxofibrosarcomas, epidermoid cysts, and carcinomas of the tympanic cavity [13].

Glossopharyngeal neuroma can be somehow differentiated from the above mentioned tumors based on MRI study. In T1-weighted images, neurinomas isointense or hypo intense, becoming hyper intense on T2-weighted images. It shows significant enhancement after contrast injection. Absence of tumor tail within the acoustic meatus usually differentiates them from acoustic schwannomas. Enhancement is relatively greater in meningiomas, whereas signal-void areas, are more prominent in glomus jugular tumors [14, 15, 16].

From histopathological point of view, concurrent arrangement of hypercellular or Antoni A areas composed of spindle cells in interlacing fascicles and hypocellular or Antoni B areas in loose myxoid stroma are suggestive of schwannomas. Also, parallel rows of palisading nuclei known as Verocay body, can be seen in highly differentiated tissue. Strong S100 positivity in IHC is another indicator suggesting toward schwannomas [12]. In conclusion, diagnosis of sinonasal schwannoma could sometimes impose difficulties to clinicians and radiologist due to its infrequency, varied symptoms and nonspecific imaging features. However, as this report provided evidence of schwannoma in a relatively rare location, mentioned diagnosis should always be kept in our differential diagnosis.

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