

Schwannoma Base of Tongue: Report of a Rare Case and Review of Literature

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ABSTRACT

Schwannomas or neurilemmomas are benign, slow growing, usually solitary and encapsulated tumor, originating from Schwann cells of the nerve sheath. Extracranially, 25% of all schwannomas are located in the head and neck region. The intraoral lesions show a predilection for the tongue, followed by the palate, buccal mucosa, lips and gingiva respectively. Among these, base of tongue schwannoma are extremely rare. Schwannomas involving the tongue base remain asymptomatic unless they attain appreciable size. Diagnostic investigations include ultrasound scanning, computed tomography, magnetic resonance imaging (MRI), and fine-needle aspiration cytology. MRI is superior to other imaging modalities for the examination of the base of the tongue. The definitive diagnosis is based on histopathology. Surgical excision or enucleation with preserve nerve function is the treatment of choice for this rare tumor. The most common approach for complete excision is transoral route for base of tongue schwannoma. Authors report a case of base of tongue schwannoma which was treated with surgery by transoral route.

Keywords: Schwannoma, Base of tongue schwannoma, Extracranial schwannoma.

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INTRODUCTION

Schwannomas or neurilemmomas are benign, slow growing, usually solitary and encapsulated tumor, originating from Schwann cells of the nerve sheath.¹ They originate more frequently from sensory nerves and can affect all cranial nerves, except the olfactory and optic, which are extensions of white matter from the brain.² When the nerve of origin is small, its association with a given tumor may be difficult to demonstrate. On the other hand, if a larger nerve is the site of origin, the nerve fibers are found to be splayed out over the outer aspect of the capsule rather than incorporated within the mass of the tumor.³ Extracranially, 25% of all schwannomas are located in the head and neck region, but only 1% show an intraoral region. The intraoral lesions show a predilection for the tongue, followed by the palate, buccal mucosa, lip and gingiva. They remain asymptomatic unless they attain appreciable size. In the tongue, the distinction between nerve of origin whether hypoglossal, glosso-pharyngeal or lingual nerve is difficult, given their proximity.⁴

CASE REPORT

A 55-year-old female patient presented to outdoor of our department with 1.5 month's history of difficulty in swallowing which was progressive in nature, more with solid food in comparison to liquid. She also complained of mass at the posterior part of the tongue on the right side for the same duration. The patient described that the mass was very small at onset and gradually became larger with time. The swelling was associated with minimal pain and mild difficulty in swallowing. Inspection of oral cavity and oropharynx revealed a multilobulated, dark red colored and well-demarcated mass of approximately 22 mm in diameter on the base of the tongue right side (Fig. 1). On palpation, swelling was tender and extending to right tonsillar pillar with partial obliteration of oropharyngeal airway. The swelling was firm, multilobulated and did not bleed to touch. No lymph nodes were enlarged and the remainder of the head and neck examination revealed no other lesions. The patient's medical history was unremarkable. Patient was ordered CT scan of neck with contrast enhancement which showed well defined, heterogeneously enhancing, soft tissue attenuation lesion, measuring approx $2.2 \times 1.2 \times 2.3$ cm, involving base of tongue on right side and right tonsillar pillar with obvious medial bulge in the oropharynx with partial obliteration of oropharyngeal airway (Fig. 2).

Routine investigations were within normal limits. After obtaining proper informed consent and all routine investigations, biopsy from the lesion was advised. HPE of

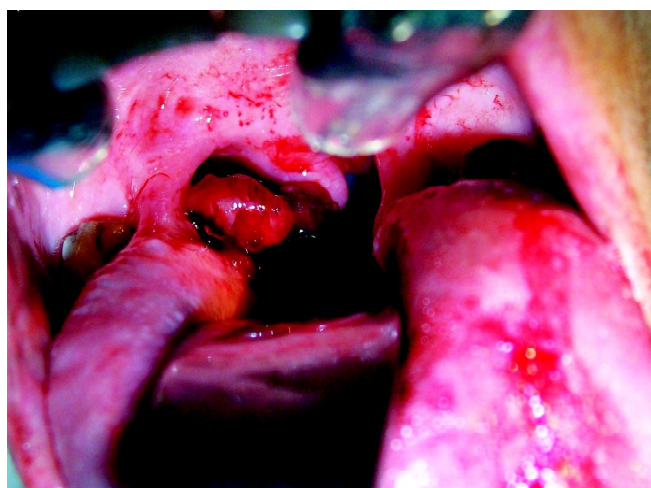


Fig. 1: Intraoral photograph of the patient showing tumor at base of tongue

tissue sent for biopsy revealed schwannoma (Fig. 3). Patient was taken up for surgery under general anesthesia. Wide local excision, i.e. excision with a cuff of clinically uninvolved surrounding tissue was performed through intraoral route. The postoperative course was uneventful. The mobility of the tongue was good. Postoperatively, there were no complaints related to swallowing. Patient was discharged home after 6 days of surgery. Histopathological examination of surgical specimen confirmed the diagnosis of schwannoma (Fig. 4). Patient was followed up till 6 months on monthly basis during which she remained symptom free.

REVIEW OF LITERATURE AND DISCUSSION

Schwannomas are benign encapsulated nerve sheath neoplasm composed of Schwann cells. They were first described by Virchow in 1908.^{5,6} Shortly thereafter, Stout recognized their schwannarian derivation.⁵ Embryologically, Schwann cells arise during the fourth week of development from a specialized population of ectomesenchymal cells of

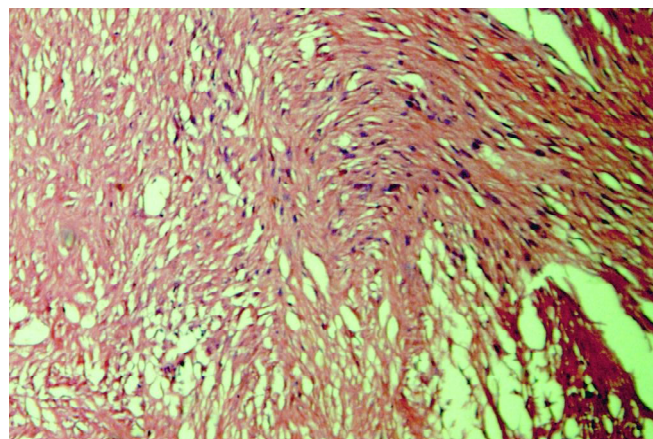


Fig. 4: Photomicrograph showing Antoni A and B type cells in sheets. Among the sheets, there are acellular eosinophilic bodies called Verocay bodies, formed by thin cytoplasmic fibers (stain—H and E magnification 5x)

neural crest.⁶ These cells form a thin barrier around each extracranial nerve fiber of motor and sensory nerves and wrap larger fibers with myelin sheath to enhance nerve conductance.

Schwannomas commonly arise from spinal nerve roots, intracranial nerves of the face, neck, extremities, mediastinum and pelvis. Most commonly affected nerve is the VIII cranial nerve (acoustic neuroma).⁷ Schwannomas in the head and neck regions constitute 25% of all extracranial schwannomas, but only 1% show intraoral origin.⁸ The intraoral lesions show a predilection for the tongue, followed by palate, buccal mucosa, lip and gingiva. Within the tongue schwannomas may arise from the hypoglossal, glossopharyngeal or lingual nerve. Typically, they remain asymptomatic and are slow growing neoplasm present several years before diagnosis. They produce symptoms by virtue of their large size and impingement on the affected nerve.^{7,5}

Diagnostic investigations include ultrasound scanning, computed tomography, MRI, and fine-needle aspiration cytology. MRI is superior to other imaging modalities for the examination of the base of the tongue.⁹ The definitive diagnosis is based on histopathology. The tumor tissue consists of the so called Antoni A and B type cells. Type A tissue shows densely packed, elongated spindle cells, in the form of parallelly formed thin reticulin fibers, fusiform shaped cells and curled nuclei while type B tissue has a more myxoid consistency. Among the sheets, there are acellular eosinophilic bodies called Verocay bodies, formed by thin cytoplasmic fibers. In addition, hemorrhage from adjacent tissue, necrosis, hyalinization and cystic degeneration may also occur in the tumor tissue.¹⁰

Malignant lesions, such as squamous cell carcinomas and sarcomas and benign lesions as granular cell tumors, salivary gland tumors, schwannomas of the oral cavity, leiomyomas, rhabdomyomas, lymphangiomas, hemangiomas, dermoid

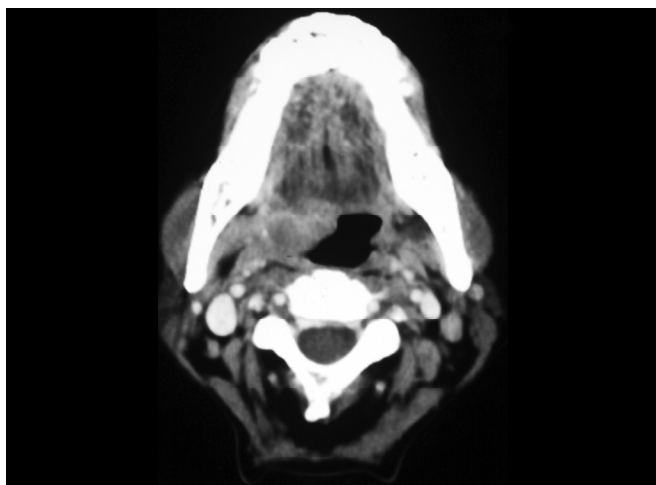


Fig. 2: Computed tomographic scan showing tumor at base of tongue schwannoma



Fig. 3: Excised surgical specimen

cysts, lipomas, inflammatory lesions and lingual thyroid are the differential diagnoses of this entity.¹¹

Surgical excision or enucleation with preserve nerve function is the treatment of choice for this rare tumor. Excision is usually easy to perform and the prognosis is excellent as malignant transformation is rare.¹²

The most common approach was the transoral route as most of these tumors are easily accessible via this route. Several other approaches have also been reported to have success including submandibular, suprahyoid pharyngotomy and transhyoid approach.^{12,13} All of these approaches were used for base of tongue schwannomas which were deemed difficult to approach by transoral route. More recently, the use of CO₂ laser for excision of a base of tongue schwannoma has also been reported.¹⁴

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