

Case Report

Tongue base schwannoma: a case report

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ABSTRACT

Schwannoma is a benign, slow growing tumor which is most commonly seen in the VIII cranial nerve. Schwannoma was first described by Verocay in 1910. We have reported a case of a 52 year old female who presented to the hospital with complaints of difficulty swallowing, change of voice and a growth on the posterior one third of the tongue. Biopsy was done and sent for histopathological evaluation which confirmed the diagnosis of Schwannoma. She underwent Lip Split with Mandibulotomy surgery which was done with a awake fiberoptic nasal intubation followed by general anesthesia. The tumor was excised in toto. Her postoperative period was uneventful. Mandibulotomy with lip split provides more than adequate visualization of the masses on the posterior region of the tongue, allowing better maneuvering, ease and precise results for a skilled surgeon. Transoral approach may be the most widely used technique but the decision should be based on the clinical evaluation such as the size, shape and the anatomy of the patient. Narrowing down to the technique of choice should be done after a comprehensive evaluation of that patient. A team of various specialities like Otolaryngologist, Anaesthetist and radiologist is required for optimal execution for this surgery. To enhance our understanding a literature review between 2006 and 2023 was done which included the reports on tongue base schwannoma. A large number of them underwent transoral excision of the mass with a minority choosing mandibulotomy with lip split. A limited array of literature is available for tongue base schwannoma cases.

Keywords: Schwannoma, Tongue base, Neurilemmoma

INTRODUCTION

Schwannoma is a nervous tissue tumor also known as neurilemmoma, neurinoma and perineural fibroblastoma.¹ They are benign, solitary, slow growing encapsulated painless nerve sheath tumors, which can be found in any nerve of the body - autonomic, peripheral or cranial nerve, except the optic and olfactory cranial nerve. Most commonly seen in the eight cranial nerves.²⁻⁵

Around 25-45 % of these tumors are found in the head and neck region, 1% is found on the tongue.^{1-3,5} Parapharyngeal space is the most common location in the head and neck region.^{6,7} Intra orally they are mostly found in the anterior two thirds of the tongue followed by the palate, floor of the mouth, cheek mucosa and gums.² It is very rarely seen at the tongue base for which only sporadic cases are noted.

They show no gender predisposition and are mostly seen in the 20-55 age group.^{4,5,7,8,10}

After extensive research very few articles were found on tongue base schwannoma articles. A PubMed and Ovid search engine was used to gather information on the review of literature of this case report. Only these cases with reports claiming to be schwannoma of the base of tongue with English literature were included. All of the cases had reached the final diagnosis with the histological report. Most of these cases used a transoral approach for the excision of the tumor.

Articles of English Literature from the year 2006 to 2023 were studied.

CASE REPORT

After a complete evaluation a 52 year old female presented to us with complaints of gradually increasing mass on the back of her tongue. She complained of dysphagia, 'hot potato voice', difficulty with articulation and restricted tongue movement from the past 1 year. No discoloration of the surrounding tissue, no difficulty breathing, no sensory or taste abnormalities, no history of bleeding or hematemesis was noted. The mass moved upwards on gagging and no cervical lymph node enlargements or neurological deficits were noted.

On examination a large easily palpable rubbery, non tender, non ulcerated slough covered mass was noted which was causing significant difficulty while swallowing food substances. MRI (neck) revealed a large enhancing lobulated submucosal lesion (hyperintense on T2 and hypointense on T1) involving the base of the tongue and completely obscuring the vallecula causing oropharyngeal airway obstruction. The well circumscribed mass measured 3.0* 3.2* 3.5 cm (TR*AP*SI). It superiorly reached the soft palate and the inferior margin lay 1.6 cm above the hyoid bone and 2.6 cm above the glottis.

The slough covered grayish tan tissue was sent for biopsy which revealed a benign peripheral nerve sheath tumor consistent with Schwannoma which described the composition of spindle shaped cells with Verocay bodies.

The patient was scheduled for the operation after 3 weeks of the checkup. Awake Fiberoptic nasal intubation was performed for the insertion of the endotracheal tube. The procedure performed was Lip Split + Mandibulotomy + Tracheostomy under General Anaesthesia. Midline tongue split along with medial mandibulotomy was performed to approach the entirety of the tumor which was located at the base of the tongue. A notched incision designed within the midline of the lower lip was taken - the incision was extended inferiorly till it met the underlying neck dissection incision near the hyoid bone and the inferior labial artery was cauterized. The periosteum was incised 2 cm on both the sides of the osteotomy site till the inferior border of the mandible was well exposed. After exposing the mandible the reconstruction plates were used to map the position for

proper alignment before medial mandibulotomy. Followed by an oscillating saw to perform the mandibulotomy anterior to the mental foramen and a surgical blade was used to incise the gums. After the osteotomy was completed the floor of the mouth was incised and the mandible was retracted laterally. The mylohyoid muscle was divided and the submandibular ducts were retracted laterally. The base of the tongue tumor was visible and blunt dissection of the mass was done.



Figure 1: Excision of the mass through lip split and mandibulotomy technique.



Figure 2: Location of the tumor on the radiological image.

After the removal of the smooth encapsulated tumor the mucosa of the floor of the mouth was sutured from the posterior to the anterior. The skin was approximated and the wound was closed in layers. Tracheostomy was done after the excision of the tongue base Schwannoma as a

preventive measure for airway protection. She was started on Ryles tube feeding on post operative day 2 following the operation she recovered quickly and was taken on a soft diet. The patient was discharged on postoperative day 5 with normal functional status. She was successfully decannulated on post operative day 10. The recovery was uneventful without any postoperative complications.

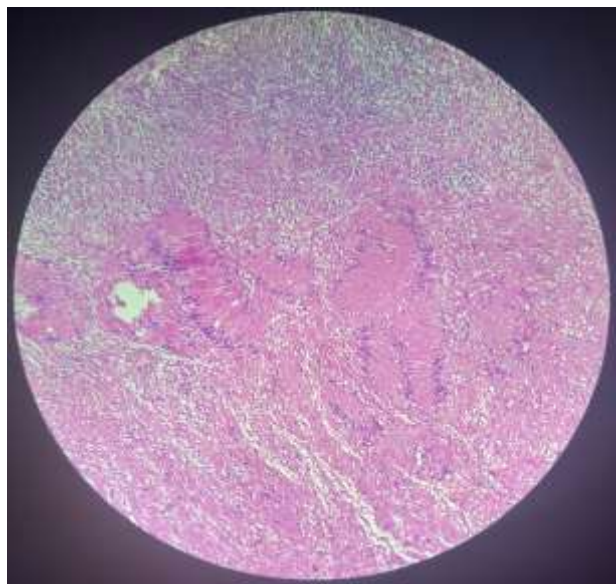


Figure 3: Palisading pattern and verocay bodies.



Figure 4: POD- 10; postoperative surgical site with good wound healing.

DISCUSSION

Schwann cells are actually neural crest cells derived from glial cells that provide insulation to the nerves. The optic and the Olfactory nerves do not have a Schwann cell covering and hence schwannomas are not seen to have an origin from them.⁵ Schwann cells arise in the fourth week of the embryological development, and schwannomas are noted when there is an unchecked proliferation

surrounding the nerves. Most of the Schwannomas are slow growing, non-tender, with a minimum predilection to malignant transformation. It could take almost 13.3 months to present.⁶ Etiology is unknown. Some of the factors are spontaneous growth, external injury, radiation exposure or irritation from a long-standing component.⁶ Schwannoma commonly arises from the spinal nerve roots from the face, neck, extremities, mediastinum and pelvis.

Schwannoma was first described by Verocay. If the schwannoma arises from a small nerve it is easy to prove its connection but if the nerve of origin is big, the nerves are splayed all over the tumor making it difficult to pinpoint the exact location.^{1,12} Only in half of the cases originating nerve can be identified.^{3,7}

The schwannomas may be indistinguishable from the other tumors so histological biopsy may be needed for a definitive diagnosis.¹ A systemic approach is required to rule out other more common diagnoses. The common differential diagnosis would be lipoma, hemangioma, eosinophilic granuloma, epidermoid and dermoid cysts, epithelial hyperplasia, granular cell tumor, benign salivary gland tumors, rhabdomyoma, leiomyoma, lingual thyroid and lymphangioma.³

The most common presentations of schwannomas were dysphagia and lump sensation followed by breathing difficulty, and change in voice - mostly the "hot potato voice".^{3,4} Schwannoma can be presented with pain, swelling fasciculations and loss of tongue control.⁵

Oral bleeding episodes are not usually seen.^{1,2} These tumors are usually solitary lesions but in Von Recklinghausen's neurofibromatosis they can be multiple in nature with a tendency of 15% malignant transformation.^{3,5,12}

Schwannomas are well circumscribed with homogeneous soft tissue on CT which are seen as hyperintense on T1 and hypointense on T2 and take up contrast sufficiently well.

MRI is used as the gold standard for the imaging techniques and signs could include split fat sign, target sign and fascicular sign.² Radiotherapy is not done as schwannomas offer a high degree of resistance to radiotherapy.^{10,11} Histologically the growth has two different appearances which can be found in combinations - Antoni A (type A) and Antoni B (type B). Type A shows tissues of well developed cylindrical bands of Schwann cells and connective tissue fibers the tendency of palisading of the nuclei and the central mass of cytoplasm (hyper-cellular areas), were as type B tissue are loosely arranged stroma in which fibers and cells have no distinctive pattern (hypo-cellular areas). Characteristic signs include palisading of the spindle shaped Schwann cells and central acellular area. Immunohistochemical testing shows positivity for S-100 and is usually the diagnostic for this condition.

Surgical intervention is the procedure of choice as the recurrence rates are very low.²⁻⁵ A number of approaches are available for the excision of tongue base schwannoma, each having its own benefits and postoperative morbidity. Namely two types: conventional methods and transoral approach. Out of which transoral surgeries have many advantages but the limitation of site exposure and less visual optimization rules out these techniques for a large tumor.³ Schwannomas pose a significant airway risk hence an early intervention is required.³ If the patient experiences dysphagia and Apea tracheostomy should be considered.

Many surgical techniques can be used- CO2 LASER excision, transoral robotic surgery, intraoral excision using diathermy, suprahyoid pharyngotomy, lateral pharyngotomy, submandibular approach and midline mandibulotomy.³ In benign lesions, excision must be carried out smoothly without any functional disability and with the least comorbidity possible which depends on the size of the area available for the approach. Transoral surgeries are usually preferred for tumors small in size. The base of the tongue surgery is an inherently difficult area to operate on due to its neurovascular anatomy, inadequate exposure, skills could lead to dysarthria, dysphagia, aspiration and breathlessness.^{6,7} Complete preservation of the nerve function is the primary objective during surgical resection. Even though the presentation may be asymptomatic, the patients have a high chance of airway distortion during or after anesthesia induction.⁶

In our scenario we preferred lip split with mandibulotomy as the intra oral tumor was big, sizing to more than 3cm. Earlier attempts to excise the mass using a transoral approach was attempted which caused bleeding as the area of access was not adequate enough for complete removal and hence the procedure was aborted. Our surgical technique of choice was selected for better exposure of the tumor and complete removal without damaging the other neurovascular structures.

CONCLUSION

A schwannoma of the base of the tongue has to grow to a particular size to produce symptoms. This tumor should be considered among the differential diagnosis among the neoplastic, infectious, congenital causes to avoid delay in identification and treatment. The choice of Mandibulotomy and tongue split was because transoral approaches did not provide a good exposure to ensure clear margins as the area had many critical structures. This approach provided adequate area to work on due to its anatomical limitations.

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