Clinical Features and Surgical Treatment of Schwannoma Affecting the Base of the Tongue: A Systematic Review

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Int Arch Otorhinolaryngol 2017;21:408–413.

Introduction

Schwannomas, or neurilemmomas, are benign neoplasms that arise from Schwann cells that surround the peripheral, cranial, or autonomic nerve sheaths.1 These slow-growing, benign, encapsulated tumors arise when proliferating Schwann cells form a tumor that encompasses the nerve sheath.1,2 Although the etiology is unknown, some causative factors such as spontaneous growth, external injury, chronic irritation, or exposure to radiation have been hypothesized.2 Typically these growths are solitary lesions with no genetic or gender predisposition, although they may rarely occur in multiples when associated with neurofibromatosis.3

Objectives

Here, a systematic review was conducted to include 15 cases of patients with schwannoma of the base of the tongue.

Data Synthesis

Most patients presented with a single, painless, well-encapsulated nodule at the base of the tongue. These nodules were slow-growing, with an average of 13.3 months from onset to presentation. Most cases were accompanied by airway obstruction, indicated by symptoms of dysphagia, dysarthria, snoring, and sleep apnea. Overall, the histological studies were consistent with a benign schwannoma with a palisading Antoni A and Antoni B pattern without malignant changes in cell morphology. These tumors were treated via complete surgical excision, and all cases achieved full remission by final follow-up.

Conclusion

Surgical removal is the primary mode of treatment with excellent postoperative prognosis and rare instances of recurrence. Given the rarity of this tumor, this review of available case studies serves to comprehensively describe clinical presentation and surgical treatment approaches to tongue base schwannoma.

Abstract

Introduction

Schwannomas of the head and neck account for 25–40% of all cases, with presentation at the base of the tongue as the most frequent site for intraoral tumors.

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Keywords

► base of tongue
► schwannoma
► neurilemmoma
► peripheral nerve sheath tumor
► systematic review

Approximately 25–48% of reported schwannomas occur in the head and neck region of patients between 20–50 years of age.2,4 However, only 1% originate in the intraoral cavity, mostly involving the tongue.4 Schwannomas of the tongue present as a painless mass with conventional histology demonstrating Antoni A patterns of nuclear palisades surrounding Verocay bodies with alternating Antoni B patterns.5 The presence of S-100 protein in immune-histochemical staining is a classic marker for diagnostic confirmation and magnetic resonance imaging (MRI) is the gold standard for preoperative imaging.6 As schwannomas are typically benign, well circumscribed, and minimally invasive tumors, complete surgical excision is the standard of care for ensuring no recurrence.7

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Intraoral schwannomas commonly produce symptoms of throat discomfort, dysphagia, dysarthria, snoring, and impaired breathing. Complete surgical excision leads to favorable outcomes with no recurrence.\(^2\,^8\) About 130 cases of tongue schwannoma have been reported in the English literature, and of those, only 19 cases have been documented at the base of the tongue.\(^5\) Surgical resection of schwannomas located at the base of the tongue are inherently difficult due to limited operative exposure amidst intricate neurovascular anatomy, which may cause significant morbidity when damaged, such as impaired speech, aspiration, dysarthria, and dysphagia.\(^5\) Due to the rarity of this lesion and the difficulties inherent in complete excision, surgical complications may occur, including airway management, nerve preservation, and anesthetic complications.\(^1\,\,^1^0\)

The present article will review and comprehensively analyze relevant literature and patient data to more definitely explain the treatment options and optimal surgical approaches for base of tongue schwannomas.

### Review of Literature

#### Literature Search

We systematically searched the National Library of Medicine’s PubMed database up to June 2016. The following search terms were used: “tongue” and “lingual” combined with “schwannoma,” “neurilemmoma,” “nerve sheath tumor,” and “Schwann cell tumor.” We screened titles and abstracts for possible inclusion, and subsequently, retrieved the full text of potentially relevant articles for review. We chose articles based on the following criteria: it must be written in English, report cases using primary human subjects, and discuss malignancies of the base of the tongue only, not of the palate, cheek, and other tongue regions. All other articles were effectively excluded.

#### Results

Initial search of PubMed returned 293 articles (see Fig. 1). After screening the titles and abstracts for relevance, the full text of 14 articles were retrieved for review. Upon completion of full text review, we excluded one article because it was a literature review. Ultimately, 13 articles were included in this review, consisting of 15 cases total. All included articles were case reports (\(N = 11\)) or case series (\(N = 2\)). The highest number of cases was reported in India (\(N = 4\)), followed by the United States of America and Taiwan (both \(N = 3\)). Summary of the study characteristics are reported in Table 1.

#### Clinical Presentation

The mean patient age was 28.6 years (range 9–49 years), and gender distribution nearly equal with 8 female patients (53.3%) and 7 male patients (46.7%). The clinical presentation of included cases is summarized in Table 2. All cases presented as a single nodule on the base of the tongue and all were symptomatic (\(N = 15, 100\%\)). Patients presented with either non-tender masses (\(N = 9, 60\%)\) or painful lesions (\(N = 6, 40\%)\) with reported discomfort at the back of the throat. Physical examination of the lesions revealed that the majority were smooth (\(N = 9, 60\%)\), non-ulcerated (\(N = 13, 86.7\%)\), round (\(N = 3, 20\%)\), well-defined and/or encapsulated (\(N = 13, 86.7\%)\) tumors.

The most common symptomatic complaint was noticeable swelling (\(N = 13, 86.7\%)\) frequently causing

### Table 1  Study characteristics and patient demographics

| Country     | Studies (Cases) |
|-------------|-----------------|
| India       | 4 (4)           |
| Brazil      | 1 (1)           |
| USA         | 3 (3)           |
| Spain       | 1 (1)           |
| England     | 1 (1)           |
| Netherlands | 1 (1)           |
| Sweden      | 1 (1)           |
| Taiwan      | 1 (3)           |

#### Study Design

- **Case report**: 11 (11)
- **Case series**: 2 (4)

#### Demographics

| N (range) |
|-----------|
| Total patients: 15 |
| Mean Age: 28.6 (9–49) |
| Gender |
| Male: 7 |
| Female: 8 |
Table 2: Clinical presentation of tongue base schwannoma

| Number of nodules       | N (%)   |
|-------------------------|---------|
| Single                  | 15 (100%) |
| Well Defined            | 13 (86.7%) |
| Non tender/painful      | 9 (60%) |
| Pain/discomfort         | 6 (40%) |
| Smooth                  | 9 (60%) |
| Submucosal              | 4 (26.7%) |
| Firm                    | 4 (26.7%) |

| Symptoms                          | N (%)   |
|-----------------------------------|---------|
| Swelling                          | 13 (86.7%) |
| Dysphagia                         | 7 (46.7%) |
| Dysarthria/change in voice        | 5 (33.3%) |
| Snoring                           | 5 (33.3%) |
| Compromised Airway                | 3 (20%) |
| Impaired tongue mobility          | 2 (13.3%) |
| Hemorrhage                        | 2 (13.3%) |
| Paresthesia                       | 1 (6.7%) |
| Otalgia                           | 1 (6.7%) |

| Mean duration, months (N = 12)    |
|-----------------------------------|
| 13.3 (range, 0.03–36)             |

 (...continued text)
hypointense ($N = 3, 23\%$), and isointense to muscle ($N = 3, 23\%$), all of which are enhancement patterns characteristic of schwannomas. Less common image findings included necrosis and blooming, indicating hemorrhage. All cases had well-defined margins, were well-encapsulated, and revealed no indication of invasion. Computed tomography was reported in 4 cases, and was primarily used to rule out metastases ($N = 4$).

**Outcome**

In a majority of the cases, preservation of nerve function was a primary concern during complete surgical resection. Despite a wide variety of surgical techniques utilized in the 15 cases, all postoperative recoveries were uneventful with no postsurgical complications such as loss of tongue function, sensation, or nerve damage. Of the papers that reported follow-up for their patients ($N = 9$), the mean follow-up period was 13 months (range 1.5 months - 60 months) with no evidence of recurrence in any of the cases.

The most common route used in resecting tongue base schwannomas was trans-orally ($N = 7$), which resulted in successful, short postoperative recovery times with no visible scarring, nerve damage, or loss of tongue function.

Another surgical approach was transcervical incision with blunt dissection ($N = 1$), resulting in a simple, quick procedure that protected surrounding nerves. Advantages to this approach included minimal morbidity, adequate exposure, no bony cuts, and minimal postoperative care.

One case utilized a suprathyroid pharyngotomy procedure ($N = 1$), during which a tracheotomy was first performed followed by an incision made at the hyoid bone, lacerating the platysma muscle, and enabling the removal of the suprathyroid muscles from the hyoid bone. This surgical technique led to a long postoperative period of 8 days and a more extensive and arduous recovery.

Other surgical approaches that were reported included mandibulotomy with lip splitting and mandibular swing ($N = 1$), which is unfavorable as it leaves aesthetically deforming scars around the mouth. Alternatively, the submandibular approach ($N = 1$) leaves a less noticeable scar below the mandible. Despite a resulting visible scar, the transhyoid approach ($N = 1$) allowed adequate resection and access to the base of the tongue with no morbidity and acceptable functional results. Excision via transoral laser microsurgery ($N = 2$) was used without any complication and the patient was discharged that same day post-surgery. Fiber optic bronchoscope (FOB)-guided awake nasotracheal intubation followed by general anesthesia was utilized in one case. The patient in this case was

| Histology (N varies by info reported) | N (%) |
|--------------------------------------|-------|
| Spindle/elongated cells              | 13/13 (100%) |
| Antoni A pattern or Verocay bodies  | 12/12 (100%) |
| Palisading                           | 10/12 (83.3%) |
| Antoni B                             | 11/12 (91.7%) |
| Well delineated and/or encapsulated | 15/15 (100%) |
| Necrosis                             | 1/15 (6.7%) |
| Hemorrhagic                          | 2/15 (13.3%) |
| Dilated, congested vessels           | 4/8 (50%) |
| Immunostaining ($N = 15$)            |       |
| Positive S100; S100 diffuse staining | 15/15 (100%) |
| Radiology ($N = 13$)                 |       |
| T2 Hyperintense                      | 8/13 (61.5%) |
| Hypointense T1                       | 3/13 (23%) |
| Isointense to muscle                 | 3/13 (23%) |

Fig. 4 Sagittal STIR MRI image of the head and neck (A) reveals a well-defined hyperintense mass (red arrow) within the base of the tongue. A coronal postcontrast fat-saturated image (B) shows heterogeneous enhancement of the lesion abutting the right submandibular gland. Reproduced under CC-NC-ND from Badar et al (2016).
successfully intubated with no other complications following the procedure.

Discussion

The overall clinical presentation of tongue base schwannoma is fairly consistent, generally presenting as an encapsulated, well-defined mass at the back of the tongue. Oftentimes patients will delay seeking medical attention until the airway is compromised, presenting with symptoms of snoring, sleep apnea, difficulty breathing, dysphagia, and dysarthria.\(^8\)–\(^11\) Generally, blood and urine chemistries are within normal limits.\(^9\) Biopsies and further histological analyses show characteristic histology of a schwannoma, revealing spindle shaped cells that are often palisading in a mixture of Antoni A and Antoni B patterns.\(^5\),\(^12\) MRI investigation usually demonstrates a well-delineated, encapsulated lesion with enhancement patterns characteristic of schwannomas such as T2 hyperintensity, T1 hypointensity, and isointensity to muscle.\(^11\) All cases stain positively for S100.\(^13\) Complete surgical excision is the gold standard of treatment for schwannomas, leading to very low rates of recurrence.\(^9\),\(^13\)

Surgical treatment of tongue base schwannomas is difficult due to limited operative exposure.\(^9\),\(^10\),\(^14\) Several surgical techniques are employed to remove schwannomas at the base of the tongue due to varying levels of surgical difficulty based on tumor size and location.\(^14\) Traditional surgical techniques for treatment of lesions at the base of the tongue include transoral excision, suprathyroid pharyngotomy, submandibular approach, and mandibulotomy with lip splitting.\(^1\),\(^5\)–\(^8\),\(^13\)–\(^14\) The transoral surgical approach is the most common and preferred method for small, well-encapsulated tumors, which do not cause any distortion of the airway.\(^13\) While transoral operations eliminate cosmetic disfigurement from scars, this technique is more likely to cause postoperative bleeding and swelling, especially in cases of larger, deeper lesions.\(^14\) Of the traditional surgical methods used to treat tongue schwannomas, the transoral approach is recommended in favor of suprathyroid pharyngotomy and the submandibular approach, which should be reserved for schwannomas that are malignant or deemed too difficult to approach transorally.\(^10\),\(^11\) Although mandibulotomy with lip splitting is frequently used for treatment of larger schwannomas due to better exposure, this approach has a higher risk of damaging the intricate neurovasculature within this region and can cause significant morbidity.\(^9\)

The utilization of laser treatment in tumor excision is an emerging alternative to traditional surgical methods. A CO\(_2\) laser surgery has high organ and nerve function preservation, minimal postoperative care and complications, and bypasses difficulties posed by excessive bleeding and obstruction of the lesion site.\(^7\) Laser surgery is very precise and readily seals any blood vessels, providing a dry field during dissection; however, higher cost and inadequate training of professionals limit this technique.\(^7\)

Another surgical challenge inherent in tongue base schwannoma treatment is difficult intubation. Fibre optic bronchoscopic-guided awake nasotracheal intubation followed by general anesthesia proved advantageous for schwannomas that profoundly obstructed the airway (\(N = 2\)).\(^1\),\(^2\) This technique was performed to preserve the airway anatomy and permit intact muscle tone to keep airway structures open, leading to easier visualization of the upper airway.\(^1\),\(^5\) Postoperative bleeding and swelling in the pharyngeal cavity and mouth floor is a common complication that can lead to dangerous obstruction of the airway after excision.\(^1\),\(^5\) Thus, nasal intubation and careful postoperative monitoring is recommended to maintain a viable airway even after a successful, complete excision.\(^1\) Proper anesthetic management for surgical excision of schwannoma of the tongue depends on the location, size, and surgical approach.\(^1\) Swellings located at the base of the tongue, though asymptomatic, may cause a distorted airway upon operative proceedings, causing life threatening airway obstruction after the induction of anesthesia.\(^1\)

The methodology of this review was designed a priori with explicit procedures to reduce bias in article selection, appraisal, and data extraction and analysis. However, there exist limitations to the present review. Use of a single search database is one limiting factor. Additionally, the study is limited by the consistency of the literature in reporting data, even with thorough extraction of available data.

Final Comments

Tongue base schwannoma is a rare neoplasm with an excellent prognosis. The clinicopathological characteristics are consistent with that of benign peripheral nerve sheath tumors. Surgical removal is the primary mode of treatment with excellent postoperative prognosis and rare instances of recurrence. Preservation of lingual function and minimal postoperative complications, however, still remain a challenge. Traditional surgical techniques, though routinely performed as the primary treatment, frequently result in poor potential outcomes such as swelling, scarring, bleeding, and longer recovery period. CO\(_2\) laser surgery is a new, upcoming treatment option that has been performed in several cases with excellent results. Investigation into the efficacy of this modality on primary lesions should be further explored as a minimally invasive alternative to traditional surgical treatment is highly desirable.

Acknowledgements

This research received no specific grant from any funding agency. This study did not involve human or animal subjects. The authors declare no conflicts of interest.

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