Schwannoma base tongue: Case report and review of literature

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Abstract

Schwannomas are benign peripheral nerve sheath tumors. These are rare in the oral cavity (1%). The most common site of involvement in oral cavity is the tongue. Posterior third of tongue is not frequently involved. The aim of this paper is to present a case report of base tongue schwannoma and review literature of this rare tumor. Data from literature were analyzed for age, gender, presenting symptom, size at presentation, and surgical approach.

We report a case of 26 year-old male who presented with swelling posterior 1/3rd tongue and change in quality of voice. He was evaluated for the same with MR and incision biopsy and was planned for surgery. Surgery was abandoned at a district hospital due to difficulty in intubation. At our center he underwent fibro optic bronchoscopy guided intubation followed by general anesthesia. He underwent excision of mass using left paramedian lip spitting approach with mandibulotomy and mandibular swing. Tumor was excised in toto. His post-operative recovery was uneventful.

Literature review between 2001 and 2012 was done. 15 cases of base tongue schwannoma were identified. The most common age group involved was between 30-40 years. There was a slightly higher incidence in females. All patients were symptomatic at presentation. Most common complaints were related to swallowing and throat pain. Most patients underwent transoral excision of the tumor.

Keywords: schwannoma, base of tongue, tongue schwannoma, lingual schwannoma, tongue neurilemmoma

Introduction

Schwannoma also known as neurilemmoma, is a benign, usually solitary encapsulated nerve sheath tumor arising from Schwann cells of the nerve sheath. It can virtually arise from any peripheral, autonomic or cranial nerve. It was first identified by Virchow in 1908. 25-40% of extra cranial schwannomas are located in the head and neck region and from these, only 1-12 % are intra-oral. Tongue is the most common site of involvement in the oral cavity.

A comprehensive literature search of intraoral schwannomas was done by Hatzlotis et al in 1967. He had reported 5 cases of schwannomas arising from the base of tongue over a period of 21 years (1945-1964). Later Marc Cohen et al. reported 6 cases of base of tongue schwannoma over a period of 57 years between 1955-2006, based on results of PubMed search. We further carried out literature search between 2001-2012 and we were able to identify 15 cases. We contribute to the list by presenting a case of base of tongue schwannoma recently operated in our institution.

Materials and Methods

An internet search of the terms “tongue schwannoma,” “lingual schwannoma,” “tongue neurilemmoma”, and “lingual neurilemmoma” was performed with the date range of 2001–2012. The search was limited to case reports in English. Schwannomas of the base of tongue were included. All of the case reports have histologically confirmed the identity of the masses as schwannomas. The elements looked into for data analysis included age, gender,
presenting complaints, size of the mass at presentation, difficulties at intubation, treatment modality and surgical approach.

**Case report**

A 26 year old male had presented with complaints of a painless swelling over posterior part of tongue and change in the quality of voice for 4 months. He was evaluated at a District hospital for the same. He underwent MRI of the head and neck region which revealed a diffuse heterogeneous lesion arising from posterior 1/3rd tongue of 4 x 3.4cm x 3.5 cm size. The lesion was hypo-intense on T1W1 images (Figure1) with internal hyper-intense areas. It was heterogeneously hyper-intense in T2 W1 (Figure 2) images and on STIR images. Blooming was noted, possibly due to hemorrhage. The lesion had a large exophytic component, protruding into oropharynx and compromising airway. It

![Figure 1: T1 W images](image1)

![Figure 2: T2 W images](image2)
was in close proximity with the left tonsillar fossa laterally and soft palate superiorly. Epiglottis was pushed posteriorly. MRI findings were consistent with cystic schwannoma. Incision biopsy was taken from the lesion, which was confirmed the diagnosis of Schwannoma. Immunohistochemistry showed strong and diffuse S100 positivity. He was taken up for surgery, but the procedure was abandoned in view of difficult intubation. He was further referred to us.

He was re-evaluated at the Regional Cancer Centre, Trivandrum. On examination, he had a swelling of 4 cm size involving posterior 1/3rd of tongue with overlying slough covered ulceration of 1.5 x 1 cm size. The swelling was more towards the left side and abutting left tonsil. The posterior limit of the swelling could not be palpated. His tongue movements were normal. There were no palpable neck nodes. IDL was not possible because of the size of the swelling. CT scan, MRI films and pathology slides were reviewed.

After proper assessment and evaluation, he was planned for surgery. Fibro optic bronchoscopy guided awake nasotracheal intubation was done, following which the patient was intubated. Examination under anesthesia was done. The findings included a 4 x 3.5 x 3 cm sub mucosal tumor with an overlying ulcerated area of 1.5 x 1 cm, involving posterior 1/3rd tongue, more towards left side, abutting the left tonsil and pushing the epiglottis posteriorly.

We used a left paramedian lip split approach with left paramedian mandibulotomy and mandibular swing. The schwannoma (Figure 3, 4, 5) was excised in toto. Tongue defect was repaired primarily. Mandibulotomy was fixed with two 2mm, 4-hole mini plates and 8 mm screws.

His post-operative course was satisfactory. Oral sips were started on post-operative day 2. He was taught jaw physiotherapy. He was on pureed diet by post-operative day 5 and was graduated to normal diet by post-operative day 14.

Microscopic examination findings (Figure 6) were that of a fairly circumscribed neoplasm composed of closely packed spindly cells with eosinophilic cytoplasm elongate nuclei and inconspicuous nucleoli arranged in fascicles. Nuclear palisading was seen. The pattern was consistent with Antony type A areas. Free bands of amorphous substance between rows of nuclei constituted the Verocay bodies. The second pattern was consistent with Antony type B and was composed of very loosely arranged Schwann cells set in a meshwork of reticulum.
fibers. Based on these findings, a diagnosis of schwannoma was made. Excision was complete on pathological examination.

Results

An internet search from 2001 to 2012 identified 15 cases of schwannoma of base of the tongue adding our report to this list makes the total number of cases 16. The most common age of presentation was between 20-40 years (Figure 7). Base tongue schwannoma were also reported in the pediatric age group. Out of the 16 cases, the gender of patients was mentioned only in 14 cases where there were 8 females (57.14%) and 6 males (42.86%)

Out of the 16 cases, 13 cases reported the presenting symptom (Table 1). The most common presenting symptom was difficulty in swallowing or globus seen in 9 out of 13 cases (69.23%). 3 (23.07%) patients complained of snoring and 3 (23.07%) patients complained of change in quality of their voice. 3 (23.07%) patients have reported a swelling. 2 out of 13 (15.38%) patients reported difficulty in breathing. One (7.69%) patient presented with sore throat.

Size of the tumor was mentioned in 12 out of 16 case reports. The average size at removal was 35.08 mm (range: 18mm-85mm).13 out of the 16 cases reported surgical approach. 9 (69.23%) out of the 13 patients underwent transoral excision. Out of these 9 patients, 2 patients (22.22%) underwent transoral laser microsurgery. In these two patients, size of the tumor was not available for one patient and the other one had a 22mm tumor. Surgical approach to the tumor (Figure 8) was varied from either transoral, transhyoid, suprahyoid, transcervical or mandibulotomy approach. 1 (7.69%) patient out of 13 underwent transhyoid excision and 1 (7.69%) underwent suprahyoid excision. In each of these cases the tumor size was 40mm. 1 (7.69%) patient out of these 13 patients underwent excision of schwannoma using transcervical approach. The size of tumor was not identified in this case report.

The case in the current study (1 patient) underwent excision of schwannoma using left paramedian lip split and left mandibulotomy and mandibular swing.

Discussion

Schwannoma is a slow-growing benign tumor of the nerve sheath. It was first reported by Virchow in 1908. Neurilemmoma, neurilemmoma, neurinoma, perineural fibroblastoma, peripheral glioma and peripheral nerve sheath tumor are the synonyms of schwannoma. It originates from the Schwann cell of the peripheral, autonomic, and cranial nerve. It is usually a single, circumscribed, firm, painless lesion of variable size. The etiology of schwannomas is unknown. Approximately 25–45% of all schwannomas occur in the head and neck with the parapharyngeal space being the most common site. Less commonly, schwannomas present in the oral cavity, and of this subset the tongue is the most frequently involved organ.

Table 1: Patient symptoms

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Number of patients (n=13)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dysphagia</td>
<td>9</td>
</tr>
<tr>
<td>Change in quality of voice</td>
<td>3</td>
</tr>
<tr>
<td>Snoring</td>
<td>3</td>
</tr>
<tr>
<td>Difficulty in breathing</td>
<td>2</td>
</tr>
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</table>

Figure 7: Age distribution at presentation of base tongue schwannoma

Figure 8: Surgical approaches to the tumor
<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age (years)</th>
<th>Gender</th>
<th>Presenting Symptom</th>
<th>Size (mm) [Greatest Dimension]</th>
<th>Surgical approach</th>
</tr>
</thead>
<tbody>
<tr>
<td>Goto Takashi et al</td>
<td>2001</td>
<td>6</td>
<td>M</td>
<td>NR</td>
<td>NR</td>
<td>Transoral</td>
</tr>
<tr>
<td>Bassichis et al</td>
<td>2004</td>
<td>9</td>
<td>M</td>
<td>Snoring, Breathing difficulty</td>
<td>23</td>
<td>Transoral</td>
</tr>
<tr>
<td>T Nakasato et al</td>
<td>2005</td>
<td>9</td>
<td>F</td>
<td>Sore throat</td>
<td>20 Multilobar neurilemmoma</td>
<td>Transoral</td>
</tr>
<tr>
<td>Hsu et al</td>
<td>2005</td>
<td>39</td>
<td>F</td>
<td>Dysphagia</td>
<td>40</td>
<td>Transhyoid</td>
</tr>
<tr>
<td></td>
<td></td>
<td>32</td>
<td>F</td>
<td>Globus</td>
<td>18</td>
<td>Transoral</td>
</tr>
<tr>
<td></td>
<td></td>
<td>20</td>
<td>M</td>
<td>Globus</td>
<td>50</td>
<td>Transoral</td>
</tr>
<tr>
<td>Mehrzad et al</td>
<td>2006</td>
<td>49</td>
<td>M</td>
<td>Throat discomfort</td>
<td>22</td>
<td>Transoral Co2 laser</td>
</tr>
<tr>
<td>Ying et al</td>
<td>2006</td>
<td>26</td>
<td>F</td>
<td>Dysphagia, Snoring, Change of voice</td>
<td>40</td>
<td>Suprahyoid</td>
</tr>
<tr>
<td>Ballestros et al</td>
<td>2007</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>Transoral laser microsurgery</td>
</tr>
<tr>
<td>Philip et al</td>
<td>2007</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Sawhney et al</td>
<td>2008</td>
<td>37</td>
<td>F</td>
<td>Dysphagia, Dysarthria</td>
<td>NR</td>
<td>Transcervical incision (blunt dissection)</td>
</tr>
<tr>
<td>Cohen &amp; Wang</td>
<td>2009</td>
<td>18</td>
<td>F</td>
<td>Painless mass</td>
<td>18</td>
<td>Transoral</td>
</tr>
<tr>
<td>Kayhoscrow Mordanpour et al</td>
<td>2009</td>
<td>18</td>
<td>M</td>
<td>Swelling, Difficulty in swallowing, Speech difficulty</td>
<td>20</td>
<td>NR</td>
</tr>
<tr>
<td>Upma B Batra et al</td>
<td>2011</td>
<td>38</td>
<td>F</td>
<td>Difficulty in swallowing, Change in quality of voice</td>
<td>40</td>
<td>NR Difficult airway [FOB guided awae intubation]</td>
</tr>
<tr>
<td>L Nisa</td>
<td>2011</td>
<td>38</td>
<td>F</td>
<td>Swelling, Progressive dysphonia, Sever dysphagia</td>
<td>85</td>
<td>Transoral</td>
</tr>
<tr>
<td>Current report</td>
<td>Current study</td>
<td>26</td>
<td>M</td>
<td>Swelling, Change in quality of voice</td>
<td>45</td>
<td>Left paramedian lip split with left mandibulotomy &amp; mandibular swing</td>
</tr>
</tbody>
</table>

Table 2: Patient and tumor characteristics and surgical approach
Schwannomas of the tongue occur at all ages with a higher predilection towards second to fourth decades of life. They demonstrate an equal gender predilection. The majority of them present as a painless mass. However, as they approach 3 cm in greatest dimension, they are more likely to produce symptoms such as throat discomfort, dysphagia, and voice changes. Moreover, if the schwannomas arises in the posterior two-thirds of the tongue, they are more likely to produce debilitating symptoms (6).

Schwannomas usually present as a solitary lesion. When multiple, however, they can be associated with neurofibromatosis. The differentiation between schwannoma and neurofibroma is essential because an apparently “solitary” neurofibroma may be a manifestation of neurofibromatosis (9). Approximately 15% of patients with neurofibromatosis will have malignant transformation in one or more lesion, which is in marked contrast to the typical behavior of a schwannoma (10). Malignant transformation is described in 8-10% of the cases of schwannoma (1).

To support a diagnosis of malignant transformation in benign schwannoma, the following features should be confirmed: (a) the tumor demonstrates, to some extent, benign schwannoma; (b) the tumor contains unequivocal malignant foci as manifested by the presence of increased cellularity, numerous mitoses, anaplastic cells, and invasiveness; (c) transitional features between malignant and benign areas can be seen; and (d) the patient has no evidence of von Recklinghausen’s disease (12).

In most intraoral lesions, identification of the nerve of origin is difficult. Direct relation with a nerve can be demonstrated in approximately 10-50% of the cases only (11, 13, 14).

The imaging modality of choice for schwannomas of the tongue is an MRI. MR is superior to computed tomography (CT) in several aspects. The MR image is not degraded by dental amalgam or the beam-hardening artifacts that plague CT scanning of the oral cavity. In addition, MR allows an accurate measurement of tumor size and precise localization in relation to other structures. On MRI, tongue schwannomas appear iso-intense to muscle on T1-weighted images and homogenously hyper-intense on T2-weighted images. Moreover, these tumors usually appear smooth, well demarcated, and do not invade the surrounding musculature (15).

Histologically, schwannomas display several classical features. Virtually, all of these tumors are encapsulated. Two main patterns are seen intermingled but sharply defined from each other. The first pattern is referred to as Antoni type A which consists of closely packed Schwann cells that form bundles or are arranged in rows with palisading, elongated nuclei. Free bands of amorphous substance between rows of nuclei constitute the Verocay bodies. The second pattern is known as Antoni type B and is composed of very loosely arranged Schwann cells set in a meshwork of reticulum fibers and microcysts (6). In addition to these characteristic patterns, diagnosis is aided by immunohistochemical markers, S-100 and Leu 7, which support the Schwann cell nature of these tumors.

Anesthetic management for surgical excision of schwannoma of the tongue depends upon the location, size, and the surgical approach to provide proper exposure of the tumor and distortion of the airway caused by the tumor. Swellings located at the base of the tongue though asymptomatic, may subsequently be found to have a distorted airway and may cause airway obstruction after the induction of anesthesia. A high index of suspicion should be kept in mind while managing any tumor at the base of tongue. Symptoms such as dysphagia, loss or change in voice, breathlessness, or wheeze should warn the anesthetist of potential airway problem (16, 17).

Surgical removal of a schwannoma arising at the base of tongue is technically difficult because of limited operative exposure and intricate neurovascular anatomic relationships that when damaged may result in significant morbidity, such as impaired speech, compromised deglutition, and aspiration. Schwannomas of base of tongue are treated by surgical excision. A number of surgical approaches have been described. The preferred approach depends upon
the size and location of tumor. These include trans-oral, mandibulotomy with lip splitting, mandibulectomy with lip splitting, submandibular (visor), suprahypophyseal pharyngotomy and transhyoid approaches\(^6,7,18,19,21\). More recently, the use of CO2 laser for excision of a base of tongue schwannoma has also been reported\(^{20}\). In our case, size of lesion would have made trans-oral excision impossible. The goal of surgical therapy is complete resection. If this is accomplished, recurrence is rare.

**References**


