

Schwannoma of tongue with review of literature

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Abstract

Schwannoma is relatively uncommon, slow-growing benign tumors. It is a benign nerve sheath tumor composed of Schwann cells, which normally produce the insulating myelin sheath covering peripheral nerves. Schwannoma of the head and neck account for 25–40% of all cases. However, intra-oral schwannoma account for only 1% of all head and neck tumors. Here we report a rare tumour of lingual Schwannoma in a 28 year old male patient which presented as a slow growing asymptomatic mass at the tip of the tongue. The mass was excised and the histopathology study of the lesion showed Schwannoma of tongue.

Introduction

Schwannoma also known as neurilemmoma, neurolemoma and Schwann cell tumors are defined as a benign, encapsulated, slow growing neoplasm that arises in the nerve fiber. This neoplasm is composed primarily of Schwann cells in a poorly collagenized stroma.^{1,2}

The etiology is unknown and there is no sex predilection.³ They originate from any peripheral, cranial (except the optic and the olfactory nerves), spinal and autonomic nerves that have nerve supporting Schwann cells.¹ The neoplasm can occur alone or is a part of genetically inherited diseases such as neurofibromatosis type 1 (NF1), NF type 2 (NF2) and schwannomatosis.⁴

Approximately 25–48% of reported schwannoma occur in the head and neck region of patients between 20–50 years of age. However, they are quite rare in the oral cavity, accounting just over 1% of benign tumors.⁴ In the oral cavity, tongue is the most common location followed by palate, floor of mouth, buccal mucosa, and mandible.^{5,6}

We present here a rare unsuspected case of Schwannoma presenting with a lesion on the tip of the tongue.

Case Study

A 28 year old male patient presented to the outpatient department with a six months history of slowly enlarging lesion on the tip of tongue associated with pain. He also had difficulty in swallowing and chewing. His personal and past medical history was non contributory. He had no symptoms of loss of weight or appetite.

Intraoral examination showed a well circumscribed mass of 2X2 cms on the tip of the tongue with normal overlying mucosa. On palpation, the lump was non tender, smooth, noncompressible and firm with well defined borders. There was no evidence of ulceration or paraesthesia. Adjacent oral mucosa showed no abnormality and there was no cervical lymph node enlargement. The clinical diagnosis was given as benign tumor of the tongue.

Routine haematological investigations were within normal limits. No radiological investigations were done as the lesion was easily visible and palpable.

Following this, the patient underwent wide local excision and the tissue was sent for histopathological examination. The post operative course was uneventful and the patient was discharged. On follow up, no record of recurrence or any other complaints were recorded.

On gross examination, the excised mass was well encapsulated, grey white soft tissue measuring 2x1.3x1 cms. Cut surface was grey yellow. Microscopic examination revealed a well encapsulated neoplasm composed of two growth patterns Antoni A and Antoni B type (Fig. a). Antoni A was hypercellular with presence of Verocay bodies (Fig. b) while Antoni B (Fig. c) were fewer and had loosely arranged cells. There was no pleomorphism or mitosis. These features led to the conclusive diagnosis of Schwannoma.

Discussion with Review of Literature

Schwannoma is a benign tumor, apparently derived from the Schwann cells, which may arise from any myelinated nerve fiber. The tumor is composed primarily of Schwann cells in

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a poorly collagenized stroma.² Embryologically, Schwann cells arise from ectomesenchymal cells of the neural crest during the fourth week of development. They enhance nerve

conductance by either forming a thin barrier around peripheral nerve fibers or a thick myelin sheath.¹

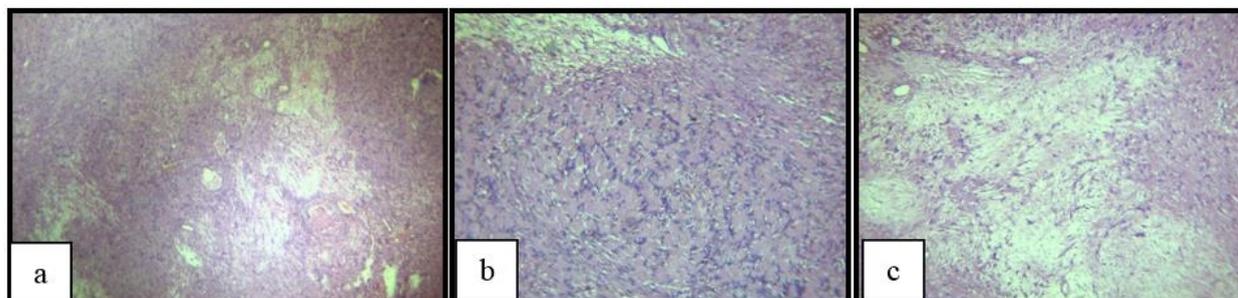


Fig. (a): Schwannoma of tongue showing Antoni A& Antoni B areas (H&E, 10X); **(b):** Antoni A showing verocay bodies and palisading nuclei, **(c):** Antoni B showing loose hypocellular.

Table 1: Patient and tumor characteristics of tongue schwannoma reported in past 10 year's literature.

| S. No | Author | Year | Age/Gender | Location of tumor | Size (greatest dimension) mm | Presenting symptoms | Surgical approach |
|-------|--|------|----------------|-------------------------|------------------------------|--|--------------------------------|
| 1 | Cohen <i>et al</i> ¹¹ | 2009 | 77/M 19 / F | Lateral / Posterior | 18 / 7 | Painless mass | Trans-oral |
| 2 | Jeffcoat <i>et al</i> ¹² | 2010 | 68/M | Lateral | | Painless mass | Trans-oral |
| 3 | Naidu <i>et al</i> ¹³ | 2010 | 12/M | Posterior | 20 | Painful mass with dysguesia | Trans-oral |
| 4 | Nisa <i>et al</i> ¹⁴ | 2011 | 38/F | Anterior | 85 | Dyspnoea, dysphonia, Severe dysphagia. | End-oral |
| 5 | Batra <i>et al</i> ¹⁵ | 2011 | 38/F | Anterior | 40 | Dysphonia & dysphagia | Trans-oral |
| 6 | Hussain <i>et al</i> ¹⁶ | 2011 | 10/F | Anterior | 50 | Dysphonia & dysphagia | Trans-oral |
| 7 | Kurupet <i>et al</i> ¹⁷ | 2012 | 56/M 31/ F | Posterior / Anterior | 40 / 30 | Painless mass / Dysphagia | Trans-oral |
| 8 | Manna <i>et al</i> ¹⁸ | 2012 | 15/M | Posterior | 30 | Dysphagia | Trans-oral |
| 9 | Lira <i>et al</i> ¹⁹ | 2013 | 26/F | Anterior | 25 | Parasthesia | Trans-oral |
| 10 | Moreno- Garcia <i>et al</i> ²⁰ | 2014 | 13/F | Anterior | 20 | Painless mass | Trans-oral |
| 11 | Bhola <i>et al</i> ²¹ | 2014 | 15/F | Anterior | 15 | Painful mass | Trans-oral |
| 12 | George <i>et al</i> ²² | 2014 | 26/M | Posterior | | Dysphonia | Paramedian mandibulotomy |
| 13 | Sharma <i>et al</i> ³ | 2016 | 20/F | Anterior | 40 | Dysarthria & dysphagia | Trans-oral |
| 14 | Medhiet <i>et al</i> ²³ | 2016 | 22/M | Posterior | 50 | Dysphagia & dysphonia | Paramedian mandibulotomy |
| 15 | Kavci <i>et al</i> ⁴ | 2016 | 20/F | Anterior | 13 | Painless mass | Trans-oral |
| 16 | Badaret <i>et al</i> ²⁴ | 2016 | 24/F | Posterior | | Pressure sensation | Trans-oral (robotic) |
| 17 | Abreu <i>et al</i> ²⁵ | 2017 | 20/M | Lateral | 15 | Painful mass | Trans-oral |
| 18 | Lee <i>et al</i> ⁷ | 2017 | 71/M | Posterior | 35 | Painless mass | Trans-oral |
| 19 | Nair <i>et al</i> ²⁶ | 2018 | 19/M | Posterior | 15 | Painless mass | Trans-oral |
| 20 | Diplanet <i>et al</i> ²⁷ | 2018 | 40/F | Anterior, Posterior | 60 | Dysarthria & dysphagia | Anterior midline glossotomy |
| 21 | Ameret <i>et al</i> ²⁸ | 2018 | 13/F | Anterior | 17 | Difficult to chew | Trans-oral |
| 22 | Present Study | 2019 | 28/M | Anterior | 20 | Dysphagia | Trans-oral |

On exploring PubMed we identified 23 cases of Schwannoma of tongue reported in literature in the past decade (Table 1) and elucidated some of their characteristics.

Schwannoma of the tongue demonstrated an equal gender predilection. The mean age at diagnosis was 29 years. The age groups most affected were the 2nd (47.82%) and 3rd, 4th (17.39%) decades of life. Majority of them presented as painless mass. The most common location was anterior part (47.82%) of tongue. The most common treatment was transoral excision, performed in (86.95%) cases. In two cases, paramedian mandibulectomy was done. One of the case was associated with Neurofibromatosis-2(NF-2) as reported by Amer *et al.*²⁸

Head and neck schwannoma account for 25–40% of such tumors in the body, with more than 90% being schwannoma of the vestibulocochlear nerve. Intraoral schwannoma make up only 1% of all head and neck tumors, with the base of the tongue being the most frequent site.³ It can occur at any age although when present in the oral cavity it tends to occur more often in adults than in children.⁵ The usual clinical presentation is a gradually growing painless mass. It is encapsulated and almost always solitary. The surface is mostly smooth and rarely exophytic or fungating. Ulceration of the overlying mucosa is uncommon and is generally the result of trauma.^{1,8} Oftentimes patients will delay seeking medical attention until the airway is compromised, presenting with symptoms of snoring, sleep apnea, difficulty breathing, dysphagia and dysarthria.⁶

Computed tomography (CT) usually shows well defined homologous lesions. However, MRI is superior to CT at depicting lingual schwannoma, as it is not degraded by dental artifacts that plague CT in the intraoral area.⁷ On MRI, schwannoma appear as a well-circumscribed mass with no infiltration into the surrounding structures. On T1-weighted imaging the lesions appear isointense relative to muscle, while on T2-weighted imaging they present as hyperintense lesions.^{3,7} However, radiology too has its own limitations i.e. sometimes it cannot differentiate schwannoma from other encapsulated benign tumors. Therefore, the definite diagnosis of schwannoma tongue is based only on histopathological examination.¹

Histologically, 2 patterns have been described, both of which make up the tumor mass. The first pattern (Antoni type A) shows tightly packed Schwann cells that form bundles or rows with elongated nuclei that appear in a palisading arrangement. An amorphous substance known as Verocay bodies are interspersed between the nuclei. The second pattern (Antoni type B) is composed of loose, hypocellular material arranged in myxoid matrix. Occasional mast cells may be identified in Antoni B area. Thrombosis and hyaline thickening of adventitia are common.^{1,8,9} Immunohistochemically, the protein S-100 is found on the supporting cells of both central and peripheral nervous systems, therefore allows the use of immunohistochemical S-100 in identifying the tumors arising from nerve sheath Schwann cells.³ Electron

microscopy shows basement membrane deposits encasing single cells and collagen fibers.¹

Schwannoma are usually treated by surgical excision with involved originating nerve. In the literature, transoral excision is the most common approach used. Recently, the use of CO2 laser for excision of a base of tongue schwannoma has also been reported.

Conclusion

Schwannoma of the tongue is a rare tumor of the oral cavity and generally remains unrecognized by treating physicians. It should therefore be considered among the differential diagnoses of tongue lesions. Clinical and radiological examination aids in diagnosis but histopathology is confirmatory. Complete excision of the tumor is the treatment of choice, usually by trans-oral approach. Following complete resection, recurrence is rare. The chance of malignant transformation of schwannoma is exceptionally rare.

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