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Schwannoma (Neurilemmoma) on the Base of the Tongue: A Rare Clinical Case

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Female, 20
Final Diagnosis: Schwannoma of the tongue
Symptoms: Dysarthria • dysphagia
Medication: —
Clinical Procedure: Excision of the mass via trans-oral approach
Specialty: Surgery

Objective: Rare disease

Background: Schwannomas are slow-growing benign tumors. They can arise from any peripheral nerve, including the cranial nerves (except the olfactory and optic nerves), spinal nerves, and autonomic nerves. Schwannomas of the head and neck account for 25–40% of all cases. However, intra-oral schwannomas account for only 1% of all head and neck tumors. Complete surgical excision is the treatment of choice. Malignant transformation and recurrence following this treatment are rare.

Case Report: A 20-year-old woman presented with a slow-growing mass over the back of her tongue first noticed 8 months before. Examination of the oral cavity exposed a 4×4 cm mass over the posterior aspect of the tongue. The remaining oral cavity examination was normal, with no cervical lymph node enlargement. The patient underwent excisional biopsy by the trans-oral approach under general anesthesia. Histopathological reports discovered features of schwannoma. The patient was followed up for 1 year; she had an uneventful recovery and no evidence of recurrence. We report a case of schwannoma over the base of the tongue, a rare location for this type of tumor.

Conclusions: In this article we report a case of schwannoma over the base of the tongue. Despite the rarity of this condition, physicians should consider schwannoma as a differential diagnosis for a mass over the tongue, as there can be a favorable outcome and prognosis for the patient when this condition is correctly identified.

MeSH Keywords: Neurilemmoma • Peripheral Nervous System Neoplasms • Tongue Diseases

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/897063>



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Background

Schwannomas are slow-growing, benign neoplasms arising from Schwann cells in the nerve sheath [1–4]. The etiology is unknown and there is no sex predilection [5,6]. Detecting the nerve of origin is often challenging. When the schwannoma arises from a small nerve, the association between the 2 can be difficult to establish [7]. Dreher et al. reported that in more than 50% of the intraoral tumors, it is difficult to differentiate between tumors of the lingual, hypoglossal, and glossopharyngeal nerves [8]. Interestingly, if these lesions arise from a large nerve, the nerve fibers are spread out over the capsule surface and not integrated within the mass itself [7].

Surgical excision remains the treatment of choice [5,9,10]. Malignant transformation and recurrence following complete excision are rare [11].

Case Report

A 20-year-old Indian woman presented with a slow-growing swelling over the back of her tongue that was first noticed 8 months before. She complained of difficulty swallowing and speaking during this period. Oral cavity examination showed a mass of 4×4 cm on the posterior aspect of the tongue, with normal-appearing overlying mucosa. On palpation, the lump was found to be non-tender, sub-mucosal, smooth, and firm, with well-defined round borders. The remaining oral examination was unremarkable and there were no cervical lymph node enlargements. She had no co-existing diseases or any exposure to relevant health hazards.

The patient's routine hematological and urine examination were normal.

The patient underwent magnetic resonance imaging pre-operatively, which showed a circumscribed mass, left of midline in the posterior base of the tongue. The mass was homogeneously T2 hyperintense and had a smooth, well-defined border, with no invasion into adjacent muscle. Additional MRI sequences demonstrated the mass to be isointense to muscle on T1 and to enhance modestly post-contrast (Figure 1).

The patient underwent excisional biopsy by the trans-oral approach under general anesthesia (Figure 2). The lesion was sub-mucosal, well encapsulated, and had a good cleavage plane. The excised mass measured 4×4 cm, had a smooth surface, well-defined round borders, and was tan-white in color. The postoperative course was uneventful. She was followed up regularly for 1 year and showed no evidence of recurrence.

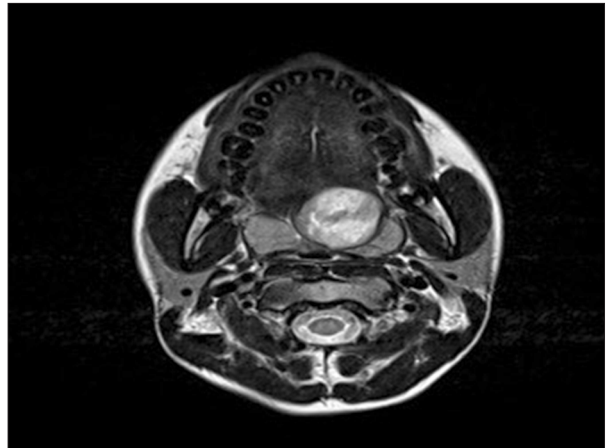


Figure 1. Axial T2 TSE through the base of the tongue shows a circumscribed mass centered left of midline in the posterior base of the tongue. The tumor has a smooth, well-defined border, with no invasion into adjacent muscle.

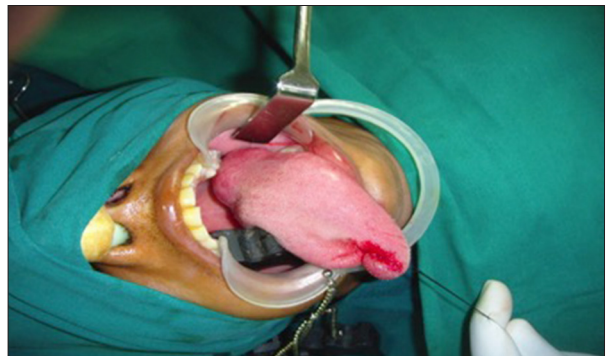


Figure 2. Intra-operative image of schwannoma over the posterior part of the tongue.

Histopathology reports of the excised surgical specimen on low-power imaging revealed Antoni A areas with ill-defined fascicles of spindle-shaped cells (Figure 3), while high-power imaging found fewer cellular Antoni B areas, with loosely arranged cells (Figure 4).

Immunohistochemistry was positive for S-100 protein, confirming schwannoma as the diagnosis.

Discussion

When evaluating a patient presenting with a tumor of the tongue, schwannoma should be considered along with the other differential diagnoses like lipoma, neurofibroma, hemangioma, lymphangioma, lingual thyroid, leiomyoma, and benign salivary gland tumors [10].

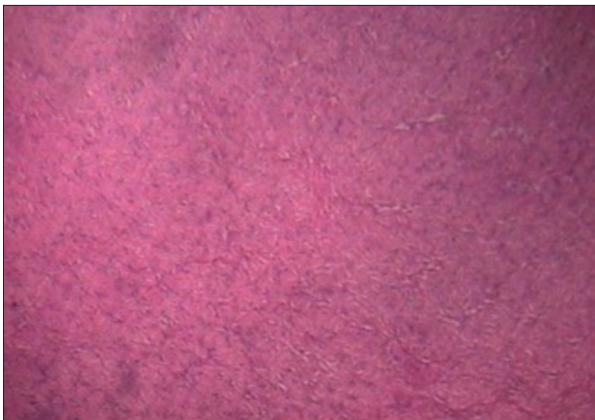


Figure 3. Schwannoma of the tongue under low power (H&E) showing Antoni A area with ill-defined fascicles of spindle-shaped cells.

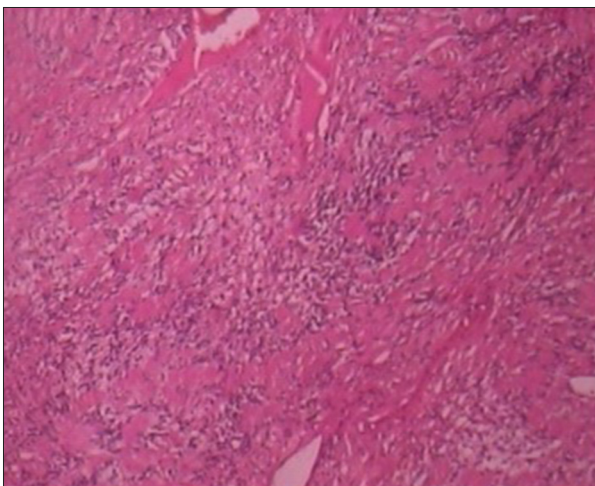


Figure 4. Schwannoma of the tongue under high power (H&E) showing palisading nuclei and Verocay bodies.

Isolated tumors are the most common presentation of schwannomas; however, if there are multiple tumors there may be an association with neurofibromatosis. Only neurofibromas have the potential for malignant transformation; therefore, an accurate diagnosis is essential to differentiate between an isolated neurofibroma and a schwannoma [12].

Head and neck schwannomas account for 25–40% of such tumors in the body, with more than 90% being schwannoma of the vestibulocochlear nerve [6, 10]. Intraoral schwannomas make up only 1% of all head and neck tumors, with the base of the tongue being the most frequent site [13].

Intra-oral schwannomas most often produce symptoms of throat discomfort, dysphagia, and voice changes, and snoring or ulceration if located over the tongue [10]. The presentation varies according to the location of the tumor. Our patient

presented with difficulty swallowing and speaking, in addition to her slow-growing mass.

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 not organized in any particular arrangement [9].

Immunohistochemical markers such as S-100 and Leu 7 further aid the diagnosis of schwannoma and allow Schwann cells to be identified as the precursor of the tumor [14].

Magnetic resonance imaging (MRI) is the modality of choice for evaluating schwannoma of the tongue and is preferred over computed tomography, which degrades the image by dental amalgam [9]. On MRI, schwannomas appear as a well-circumscribed mass, with no infiltration into the surrounding structures [5,9]. On T1-weighted imaging the lesions appear isointense relative to muscle, while on T2-weighted imaging they present as hyperintense lesions [9,10].

A complete surgical excision with an attempt to preserve the nerve is the treatment of choice for a schwannoma. The most common route used in resecting the mass is via the trans-oral approach, which we used for our patient [5,9,10]. Following complete resection, recurrence is rare [9,10].

The chances for malignant transformation of a schwannoma are rare. Head and neck schwannomas have a malignancy risk of only 8–10% [11].

Conclusions

Tongue schwannomas are rare tumors of the oral cavity. Nevertheless, schwannomas should be considered among the differential diagnoses when evaluating a patient presenting with a tongue mass, especially since favorable outcome and prognosis are likely. Complete excision of the tumor is the treatment of choice, usually by trans-oral approach. The chance of malignant transformation is extremely rare. Following complete resection, recurrence is rare.

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