International Journal of Otorhinolaryngology and Head and Neck Surgery
Viswanatha SC et al. Int J Otorhinolaryngol Head Neck Surg. 2023 Aug;9(8):682-684
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

Case Report

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20232230

Schwannoma of oral tongue

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Received: 12 June 2023
Accepted: 21 July 2023

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ABSTRACT

Schwannomas also known as Neurilemmoma, are benign encapsulated slow growing tumors, which arise from Schwann cells of the peripheral nerve sheath. They usually occur in the head and neck region, but rarely in the oral cavity. We report a case of Schwannoma of the oral tongue in a 49-year-old male patient, who presented with a slow growing, painless lesion involving left side of the dorsum of the oral tongue. Patient underwent Magnetic Resonance Imaging with contrast followed by complete surgical excision. Histopathological and Immunohistochemical examination confirmed the diagnosis.

Keywords: Schwannoma, Neurilemmoma, Tongue

INTRODUCTION

Schwannomas (Neurilemmoma or Neurinomas) are benign,commonly well encapsulated, slow-growing and generally solitary tumors that arise from Schwann cells of the peripheral nerve sheath.[1] These tumors can arise from any nerve covered with a Schwann cell sheath, which include cranial nerves (except for optic nerve and olfactory nerve), spinal nerves, and the autonomic nervous system.[2] When the nerve of origin is large, the nerve fibers are found to be splayed out over the outer aspect of the capsule rather than being incorporated within the mass of the tumor. Otherwise, if the site of origin is a smaller nerve, its association with the tumor maybe difficult to demonstrate.[3] Around 25-45% of all schwannomas occur in the head and neck region. Out Of these, approximately 1-12% occur intraorally with the tongue being the most common site.[4,5] Schwannomas usually remain asymptomatic, until they attain appreciable size. They almost never undergo malignant transformation and do not show recurrence if completely excised.[6] We report a case of schwannoma of oral tongue in a 49-year-old male patient.

CASE REPORT

A 49-year-old male patient presented to the otorhinolaryngology outpatient department, with complaints of painless lesion over the tongue noticed since 7 years, initially small in size, gradually progressive. Patient did not have symptoms of pain, dysphagia, bleeding from the lesion, paresthesia or altered taste sensation. He was a teetotaller without any co morbidities. Intraoral examination revealed an exophytic firm, nodular mass over the left side of dorsum of the tongue measuring about 1.5x1.5 cm, about 2.5 cm away from the tip, just touching the midline but not crossing it. Magnetic Resonance Imaging (MRI) with contrast showed a T2 weighted hyperintense lesion with homogenous post contrast enhancement, along the dorsum of the left side of the tongue (Figure 1), measuring 1.8x1.3x0.9 cm, suggestive of a neoplastic lesion. The lesion was almost reaching the midline, approximately 2.5 cm from the tip of the tongue. There was no significant cervical lymphadenopathy.

Excision biopsy of the lesion was done under local anaesthesia. Microscopic examination showed stratified

squamous epithelium with ulceration and granulation tissue.

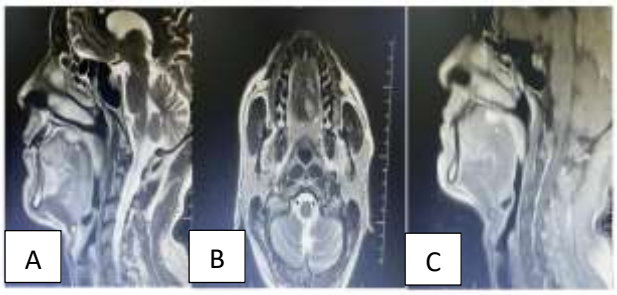

Figure 1: MRI of the tongue showing A) T2 weighted hyperintense lesion, Sagittal section (white arrow). B) T2 weighted hyperintense lesion, Axial section (white arrow), C) Homogenous post contrast enhancement, Sagittal section (white arrow).

The subepithelial stroma showed lobular well circumscribed neoplasm, composed of spindle cells arranged in diffuse sheets and fascicles (Antoni A), with hypocellular Antoni B area. Few areas resembled Verocay bodies. No nuclear atypia or pleomorphism or increased mitosis or necrosis were seen. Further Immunohistochemistry examination revealed strong positivity for S100 and negative for Pan CK (Figure 2). The morpho-immunophenotype was consistent with Schwannoma of the tongue. Post excision, patient did not develop any motor or sensory deficits and no recurrence was noted in 4 months of follow up.

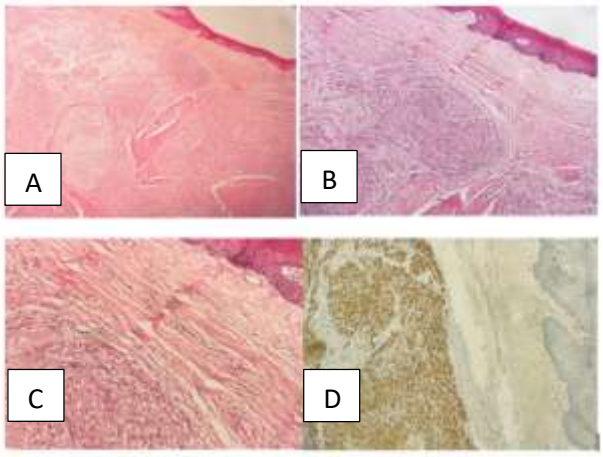

Figure 2: A) Histopathological examination with Haemotoylin and Eosin staining showing typical biphasic appearance of Schwannoma with compact celular Antoni A area adjacent to hypocellular Antoni B area (magnification x 100) B, C) Antoni A area characterized by spindle cells arranged in sheets and fascicles (magnification x 100, x 200) D) Immunohistochemistry examination showing diffuse and strong S-100 positivity(magnification x 200).

DISCUSSION

Schwannomas are benign encapsulated nerve sheath neoplasms composed of Schwann cells.[1,6] They were first described by Verocay in 1908.[7] Around 25-45% of schwannomas arise in the head and neck region, out of which 1-12% occur intraorally.[1] The intraoral lesions have a predilection for the tongue, followed by the palate, floor of mouth, buccal mucosa, gingiva, lip and vestibule.[4,5] The etiology of Schwannoma is unknown. However, factors such as extrinsic injury, chronic stimulation, and exposure to radiation have been suggested as possible etiologies.[8] High prevalence of schwannomas in the tongue may be due to the higher incidence of trauma to the tongue and the role of Schwann cells in repair of injured neurons.[9]

Schwannoma of the tongue commonly affects the age group between the first and the fourth decade and has no gender predisposition. Two-thirds of cases involve the oral portion of the tongue, and only about one-third involve the base of the tongue.[10] They are slow growing and are mostly asymptomatic, submucosal nodules as in our case, although larger tumors in the base of the tongue may cause pain, dysphagia and dysphonia.[6]

Schwannomas are usually solitary, but in patients with multifocal lesions, Multiple localized neurilemmomas, Neurofibroma in von Recklinghausen's disease, and Schwannomatosis (a non-hereditary disease characterized by multiple subcutaneous and intradermal schwannomas along with variety of intracranial tumors) should be considered.[5,11]

MRI is the imaging modality of choice for lingual schwannomas. The better tissue contrast of the MRI, compared to computerized tomography (CT), allows a precise localization with good spatial resolution, as well as a more accurate measurement of tumor size. MRI can also overcome artifacts caused by dental fillings or dense bone, which are frequently seen on CT scans of the oral cavity region.[1] Characteristically, Schwannoma has a homogeneous well-circumscribed border and does not infiltrate surrounding tissues.[12] MRI shows the tumor as a smooth and well-demarcated lesion which is isointense to muscle on T1-weighted images, homogeneously hyperintense on T2-weighted images and homogeneous enhancement after contrast administration.[1,13]

Malignant transformation of schwannoma is rare and when it does occur, it is usually in the form of Malignant Peripheral Nerve Sheath Tumor (MPNST) or more rarely angiosarcoma. The lifetime risk of MPNST in the setting of Neurofibromatosis (NF) 1 is about 2%.[14] Due to the low incidence and nonspecific clinical presentation, diagnosis of schwannoma is usually confirmed by histopathologic and immunohistochemical evaluation.[15] Histologically, most of the schwannomas have a capsule. The tumor is composed of alternating hypercelullar spindle cell areas (Antoni A) and hypocelullar round cell areas (Antoni B). Nuclear palisading is usually present in Antoni A areas

which may be prominent and form nuclear palisades around a collagenous hyalinised core (Verocay bodies). Antony B areas are paucicellular with small round cells, within a myxoid stroma. Schwannomas are characterized by strong and diffuse immunoreactivity for S-100 protein, which gives clue for the diagnosis, as in our case.[16,17]

The treatment of choice for Lingual Schwannoma is complete surgical excision. Schwannomas of oral tongue can be managed with transoral surgical excision. On the other hand, Schwannomas at the base of the tongue are usually approached by cervical access (transhyoid or submandibular) or rarely via transoral CO2 laser excision.[18] It is difficult to identify the nerve of origin of oral Schwanonomas, because most of the lesions originate from smaller nerves.[21] Recurrence of Schwannoma is rare, but when it does, it is usually due to incomplete excision.[19]

CONCLUSION

Schwannoma of Tongue is a rare benign neoplasm. It can be considered as one of the differential diagnosis for a smooth painless lesion of the tongue. Diagnosis is confirmed by histopathological and immunohistochemical examination. Treatment is complete surgical removal, transoral excision being the standard approach for majority of the lesions. Recurrence is very low if completely excised. Malignant transformation is extremely rare.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Viswanatha SC, Raghavan N. Schwannoma of oral tongue. Int J Otorhinolaryngol Head Neck Surg 2023;9:682-4.