Case Report

Schwannoma of the base of tongue in a 26-year old male: A rare case report with a short review of literature

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Abstract

Schwannoma is a benign nerve sheath tumor that arises from Schwann cells of the peripheral nerve sheath with uncertain etiology. It is well-encapsulated and a slow-growing tumor. Approximately 25–48% of cases are seen in the head and neck region. Schwannoma of the oral cavity has an approximate incidence of 1%. Tongue base Schwannoma is a rare entity. It can affect all age groups and typically presents as a painless lump. However, when it grows larger than 3 cm, it may produce dysphagia, pain, or discomfort and change in the quality of voice. Hence, Schwannoma should be considered as one of the differential diagnoses of exophytic mass of the tongue. We report a rare case of Schwannoma of the base of the tongue in a 26-year-old male who presented with a complaint of lump, along with a review of the literature published in the last 64 years.

Keywords: Base of tongue, nerve sheath tumor, rare, Schwannoma, tumors of oral cavity

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INTRODUCTION

Schwannomas are benign nerve sheath tumors, arising from Schwann cells, an integral component of the myelin sheath, ensheathing the peripheral nerves.[1] Only 1% of the Schwannomas are located intraoral and are rare at the tongue base.[1,2] It is normally solitary and slow-growing. Most Schwannomas are asymptomatic.[2] We present a case of Schwannoma in the base of tongue and review the literature available from the last 64 years (1959–2022).

CASE REPORT

A 26-year-old male presented to the ENT department with a mass on the tongue, which he noticed one month back. During examination, a pedunculated mass was seen on the base of the tongue. It was smooth, well demarcated, firm, and non-tender. Contrast-enhanced MRI of the face and neck revealed a well-circumscribed heterogeneously enhancing exophytic lobulated lesion with no definite sign of any deeper invasion. A clinical diagnosis of tongue papilloma was made and swelling was planned for surgical excision. Histopathological examination revealed a grayish brown nodule measuring 1.7 × 1.5 × 0.6 cm. The cut surface was solid and grayish-yellow. Microscopy revealed a capsulated tumor composed of alternating hypercellular Antoni A and hypocellular Antoni B areas. Verocay bodies were also noted. The histopathological features were suggestive of Schwannoma. To confirm the diagnosis, immunohistochemistry was performed. The neoplastic cells showed intense cytoplasmic and nuclear immunopositivity to S100.

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DISCUSSION

Schwannoma was first discovered by Verocay in the year of 1908.[2] It is a benign nerve sheath tumor arising from differentiated Schwann cells. They can arise from any cranial, peripheral, or autonomic nerve.[2,3] They account for 25–40% of occurrences in the head and neck region, while others involve the flexor surfaces of the upper and lower extremities, posterior mediastinum, and retroperitoneum.[3] 1–2% of Schwannomas occur intraorally, the most common site being tongue, followed by palate, floor of the mouth, and buccal mucosa.[4] Tongue Schwannomas arise from the hypoglossal nerve.[4] As they are rare, Schwannomas are usually not considered in the differential diagnosis of oral cavity lesions. Peak incidence is between the fourth and fifth decades.[4,5] There is no race or gender predilection.[5] Most Schwannomas are solitary. Multiple lesions occur in: (1) Neurofibromatosis type 1 or type 2; (2) Schwannomatosis.[6]

Merlin protein, which is produced by the NF2 gene and is positioned at 22q12.2, is crucial in the pathogenesis of Schwannomas. Loss of merlin function in the nucleus leads to an increased expression of membrane proteins, including integrins and growth factor receptors.[6] The absence of merlin favors their activation, stimulating mitogenic and survival pathways, and favoring a lack of cell polarization. Due to these changes, Schwannoma cells are unable to attach to an axon, which results in tumor development. The genes LATS1, LATS2, ARID1A, ARID1B, and DDR1 also frequently exhibit alterations. In 10% of cases, an in-frame SH3PXD2A-HTRA1 fusion is discovered. The majority of Schwannomas are caused by a 3-hit or 4-hit pathway involving two genes.[6,7]

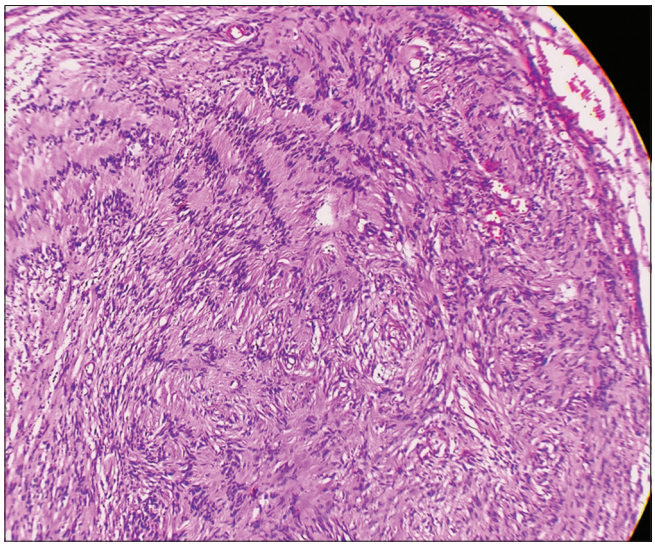

Figure 1: Photomicrograph showing biphasic tumor with hypercellular component (Antoni A) and Verocay bodies (H & E stain, 100×)

Schwannomas present as non-tender, slow-growing, soft to firm, globular, expansile submucosal masses, sessile or pedunculated, varying in size from 1 to 4 cm in diameter.[6] They are often asymptomatic or are discovered as incidental findings in imaging studies. Rarely, trauma cause ulceration of the epithelium.[7] Causative factors are unknown but chronic irritation, injury, or radiation exposure might be considered.[8]

Macroscopically, Schwannomas are mainly globoid and encapsulated with a smooth surface. Sectioned tumors reveal firm, light tan, glistening tissue, interrupted by white/yellow areas and/or patches of hemorrhage. On microscopy, these tumors are biphasic with compact areas exhibiting fascicles of spindle-shaped Schwann cells [Figure 1] AQ4 showing occasional nuclear palisading (Verocay bodies), alternating with loosely arranged hypocellular, eosinophilic areas [Figure 2]. Tumor cells are strongly and diffusely immunopositive for S100.

On review of literature published from 1959 to 2022, we found 75 cases of Schwannoma of the tongue. 54% of cases were male patients, and the rest were female. Present case is a male. 56% of cases were in the posterior tongue. Present case was in the base of tongue, which is a rare presentation. According to this review, the patients had a feeling of lump mostly, as was in our patient. Mean age of diagnosis was 25 years. Our patient was 26 years old. It was found that the average size of tongue Schwannomas at presentation was 2.4 cm. However, when the mass exceeds 3.0 cm, dysphagia, pain (or discomfort), dysphonia, and voice changes are usually present. The present case measured 1.7 cm in size and the patient was asymptomatic.

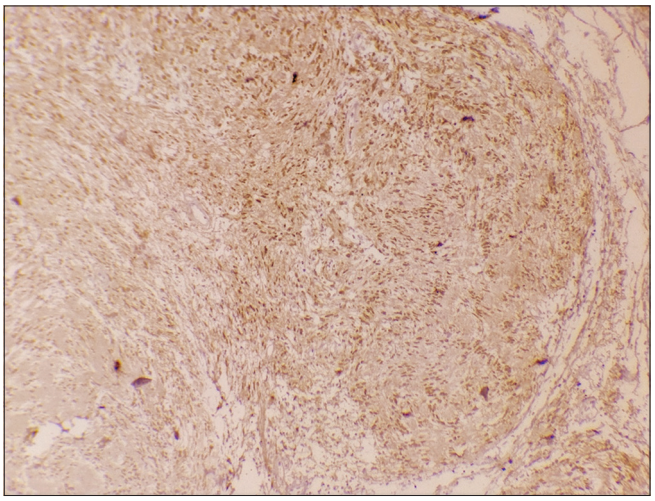

Figure 2: Photomicrograph showing immunostaining of S-100 protein-strong cytoplasmic and nuclear positivity seen in the tumor cells (100×)

Complete excision of Schwannomas has an excellent prognosis.[8] Conventional Schwannoma that develops into cancer is quite rare. In the case that is being discussed, excision has been complete, and years after surgery, there have been no signs of recurrence. Transoral resection with care to preserve the nerve remains the standard protocol.[9] There have been reports of the base of tongue Schwannomas being treated with carbon dioxide laser excision.[9,10] Radiation therapy is not effective against Schwannomas.[10] Because masses are encapsulated, it is simple to remove them completely. Recurrence could happen if the surgical excision is incomplete. In this instance, the tumor was completely removed without causing any mucosal injury, preventing tongue dysfunction and recurrence.

CONCLUSION

Schwannoma in the base of the tongue is extremely rare and has a non-specific clinical presentation. Clinically, it is a challenge to distinguish Schwannoma from other encapsulated benign tumors, so biopsy and histopathological examination are mandatory to provide a confirmatory diagnosis. Hence, during the evaluation of a smooth, painless, firm swelling in the tongue, the possibility of Schwannoma should be kept in mind.

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Conflicts of interest

There are no conflicts of interest.

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Patients and tumor characteristics of tongue Schwannoma

Author	Year	Gender	Age (years)	Size (cm)	Site	Presentation
Mercantini and Mopper	1959	M	22	1	Anterior	Intermittent pain
Cameron	1959	M	25	1.5	Anterior	Lump
Chadwick	1964	F	20	2.2	Posterior	Lump
Craig	1964	F	8	3	Posterior	Lump
Pantazopoulos	1965	F	45	4.5	Posterior	Dysphasia/change in voice
Chamber	1965	M	29	5	Posterior	Throat discomfort
Fifer *et al*.	1966	F	28	3	Anterior	Lump
Hatziotis and Aspride	1967	M	25	Hazelnut	Posterior	Lump
Oles and Werthemier	1967	M	52	1	Anterior	Lump
Paliwal *et al*.	1967	M	32	2.5	Anterior	Lump
Das Gupta *et al*.	1969	F	21	5	Posterior	Pain
Bitici	1969	M	40	2.5	Anterior	Slight discomfort
Sinha and Samuel	1971	M	23	1.5	Posterior	Dysphagia
Mosadomi	1975	M	19	3	Anterior	Painful mass
Swangsilpa *et al*.	1976	M	26	3	Anterior	Lump
Sharan and Akhtar	1978	F	30	1.5	Anterior	Change in voice
Akimoto *et al*.	1987	M	15	1	Anterior	Lump
Sira *et al*.	1988	F	18	3	Posterior	Lump
Flickinger *et al*.	1989	F	28	3	Anterior	Lump
Talmi *et al*.	1991	F	75	1	Posterior	Lump
Gallesio and Berrone	1992	F	21	1.9	Anterior	Dysphonia/paresthesia/chewing difficulty
Lopez and Ballistin	1993	M	24	0.6	Anterior	Lump
Haring	1994	F	49	2	Anterior	Lump
Nakayama *et al*.	1996	F	40	5.5	Anterior	Lump
Dreher *et al*.	1997	F	31	3	Base	Dysphagia
Spandow *et al*.	1999	M	37	7.9	Posterior	Throat discomfort
de Bree *et al*.	2000	F	24	5	Posterolateral/base	Lump
Pfeifle *et al*.	2001	F	30	0.3	Anterior	Lump
Cinar *et al*.	2004	M	7	1	Anterior	Lump
Bassichis and McMlay	2004	M	9	2.3	Posterior	Snoring
Nakasato *et al*.	2005	F	9	2	Posterolateral	Bleeding/ulceration
Hwang *et al*.	2005	M	23	2.8	Anterior	Lump
Lopez-Jornet and Bermejo-Fenoll	2005	M	39	0.8	Posterolateral	Lump
Vafiadis *et al*.	2005	M	18	3.1	Anterior	Lump
Bansal *et al*.	2005	M	26	4	Posterolateral/ventral	Paresthesia/dysphonia
Hsu *et al*.	2006	M	20	5	Posterior	Bleeding
Ying *et al*.	2006	F	26	4	Posterior	Dysphagia/otalgia
Enoz *et al*.	2006	M	7	2.5	Anterior	Dysphagia/pain
Mehrzad *et al*.	2006	M	49	2.2	Posterior/ventral	Pain
Batra *et al*.	2007	M	30	3	Posterolateral	Dysphagia, dyspnea, abscess
Ballesteros *et al*.	2007	F	31	2	Base	Pain
Sawhney *et al*.	2008	F	37	4.6	Posterolateral	Dysphagia/snoring
Sethi *et al*.	2008	F	28	1	Anterolateral/ventral	Lump
Pereira *et al*.	2008	M	12	1.5	Posterolateral/ventral	Lump
Cohen and Wang	2009	M	77	0.7	Posterolateral/ventral	Lump
Gupta *et al*.	2009	F	18	1	Anterior/ventral	Lump
Mardanpour and Rahbar	2009	M	18	2	Posterior	Dysphagia/change of voice
Karaca *et al*.	2010	F	13	2	Posterolateral/ventral	Dysphagia
Cigdem *et al*.	2010	M	13	2	Anterior/ventral	Lump
Jeffcoat *et al*.	2010	M	68	1.5	Lateral	Lump
Naidu and Sinha	2010	M	12	2	Anterolateral/base	Paresthesia/bleeding/ulceration
Lukšić *et al*.	2011	M	10	1.5	Posterolateral/ventral	Lump
Batra *et al*.	2011	F	38	4.2	Posterior/ventral	Dysphagia/change of voice
Nisa *et al*.	2011	F	38	8.5	Posterolateral/ventral	Dysphagia/dysphonia/dyspnea
Monga *et al*.	2013	M	20	2	Posterolateral	Lump
Lira *et al*.	2013	F	26	2.5	Posterior/ventral	Cervical pain
Erkul *et al*.	2013	M	21	3	Posterolateral/ventral	Chewing difficulty
Jayaraman *et al*.	2013	F	25	3	Anterolateral	Lump
George *et al*.	2014	M	26	4	Posterolateral	Dysphagia/dysphonia
Bhola *et al*.	2014	F	14	1.5	Anterolateral/ventral	Lump
Moreno-García *et al*.	2014	F	13	2	Anterior/ventral	Lump
Nibhoria *et al*.	2015	F	18	1.5	Posterolateral/ventral	Lump
Gopalakrishnan *et al*.	2016	M	32	3	Posterolateral/ventral	Dysphagia
Sharma and Rai	2016	F	20	4	Posterolateral/ventral	Dysphagia/dysphonia

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Author	Year	Gender	Age (years)	Size (cm)	Site	Presentation
Kavčič and Božič	2016	F	20	1.3	Anterolateral/ventral/tip	Lump
Lee *et al*.	2016	M	28	4	Posterolateral/ventral	Lump
Zain *et al*.	2016	F	24	Not clear	Posterior	Lump
Steffi Sharma *et al*.	2018	F	20	4	Posterior	Lump
Gayen *et al*.	2020	Male	48		Left lateral border	Growth
Keshwar *et al*.	2020	Male	20	3	Posterior	Lump
Ahmed *et al*.	2020	Female	14	2	Dorsal surface of tongue	Growth
Shibata *et al*.	2020	Female	28	2.5	Right Lateral	Nodule
Fayez A Alrohaimi *et al*.	2021	Male	12	2.3	Ventral	Cystic mass
Jagtap *et al*.	2021	Male	32	2.8	Right lateral	Lump
Rana SS, Ohri N	2022	Male	17	1.2	Base	Lump
Present case	2022	Male	26	1.7	Base	Lump/Throat discomfort