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Case Report

Ancient schwannoma (Neurilemmoma) of the tongue: A rare case report

Deepthi TR[1,*], Prem Sasikumar[2], Adarsh VJ[3], Vani MH[4], Anu Vijayan[5], Kavya Maheesan[6]

[1]*Dept. of Oral Medicine and Radiology, Malabar Cancer Care Society, Kannur, Kerala, India*
[2]*Dept. of Oral and Maxillofacial Surgery, Annoor Dental College, Muvattupuzha, Kerala, India*
[3]*Dept. of Conservative Dentistry and Endodontics, Mahe Institute of Dental Sciences, Mahé, Kerala, India*
[4]*Oral Physician and Maxillofacial Radiologist, Karela, India*
[5]*Dept. of Oral Medicine and Radiology, Mar Baselios Dental College, Ernakulam, Kothamangalam, Kerala, India*
[6]*Dept. of Conservative Dentistry and Endodontics, Kannur Dental College, Kerala, India*

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ABSTRACT

Schwannoma or neurilemmoma is an uncommon, benign peripheral nerve tumor derived from Schwann cells. It is usually a slow growing, asymptomatic, well encapsulated tumor that arises in association with a nerve trunk.Here we present a rare case of Ancient Schwannoma in a 20 year old male. The patient presented with a small, slow growing asymptomatic mass on the dorsal surface of the tongue, present since past 1year. Lesion was completely excised. Diagnosis was confirmed by histopathology and immunohistochemistry.

1. Introduction

Schwannoma or neurilemmoma is an uncommon, benign peripheral nerve tumor derived from Schwann cells. It is usually a slow growing, asymptomatic, well encapsulated tumor that arises in association with a nerve trunk.[1,2] Schwannomas are rare in the head and neck region and among the cases reported, only 1% show intraoral origin. The most common intraoral site is tongue, but can occur anywhere in the oral cavity.[2,3]

Schwannoma is of neurogenic origin, arising from the schwann cells of the nerve sheath. The lesion is believed to arise from the proliferation of schwann cells within the perineurium, causing displacement and compression of the surrounding normal nerve.[4,5] In this article the clinical presentation, histopathologic and immunohistochemical findings and management of a case of schwannoma of the tongue are discussed.

2. Case Report

A 20 year old male patient reported with complaints of a growth on the dorsal surface of the tongue present since past 1year. The mass was reported to be very small in size (size of a peanut) initially and a gradual increase in size was noted. No associated pain or tenderness was reported. On intraoral examination, a well circumscribed lesion of about 1.5x2cm diameter was noted on the dorsal surface of the posterior tongue.

A complete surgical excision of the lesion was done. Tissue specimen was sent for histopathological examination. On gross examination, the specimen was well encapsulated, firm, yellowish white in colour measuring 1.5cm in diameter. H&E stained section

* *Corresponding author*.
E-mail address: deepuraj.tr@gmail.com (Deepthi TR).

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showed stratified squamous epithelium lining and was focally ulcerated. Submucosa showed a partially encapsulated lesion composed of spindle cells with vague hypo and hypercellular area, hypercellular areas being prominent, admixed with many hyalinised blood vessels with hemorrhage in the background of a myxoid stroma. Subtle palisading of nuclei was noted at places. The lesional cells were spindle to oval with predominantly wavy nuclei and ill defined cytoplasm, seen interspersed with collagen fibres. Many cells with moderate atypia in the form of enlarged cells, hyperchromatic nuclei, nuclear vacoulation and occasional prominent nucleoli were also seen. Mitotic figures were seen (<4/10hpf). Also seen were admixed chronic inflammatory cells composed of lympocytes and occasional foamy macrophages.

The features were suggestive of Spindle cell lesion-Low to intermediate grade with a possible diagnosis of Ancient Schwannoma. Pathologist suggested IHC for cell typing and definitive diagnosis. Immunohistochemistry revealed S100 diffusely positive spindle cells and the Ki67 proliferation index was around 10% which was suggestive of a definitive diagnosis of Schwannoma (S100 diffusely positive, Ki67 ~10%).

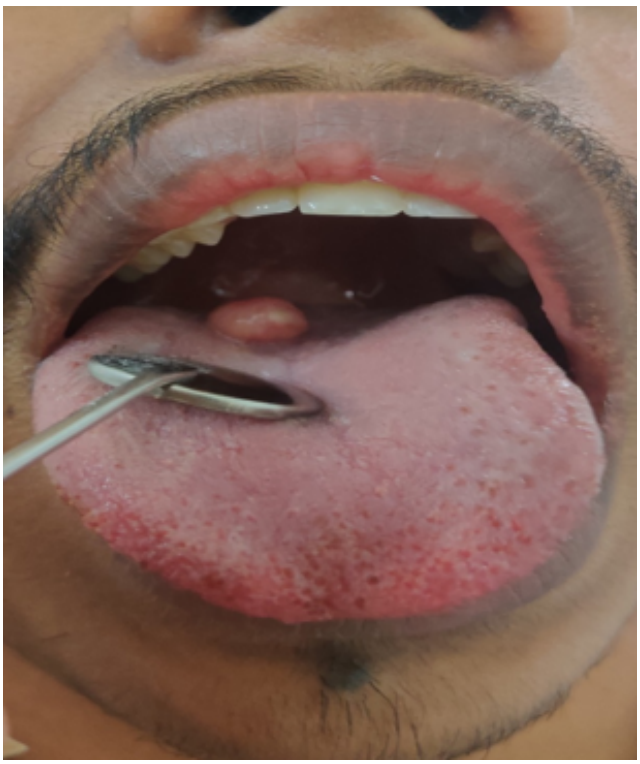

Fig. 1: Asymptomatic growth on the dorsal surface of posterior to tongue

Fig. 2: Gross specimen surgically excised from the dorsal surface of the tongue

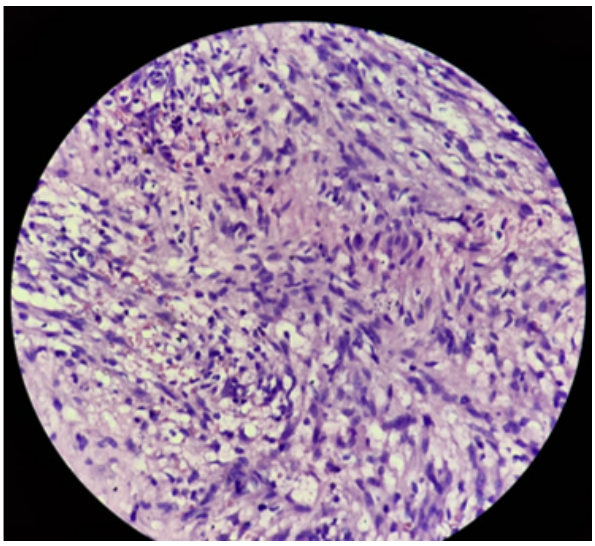

Fig. 3: Photomicrograph of the lesion showing the characteristic findings of ancient schwannoma

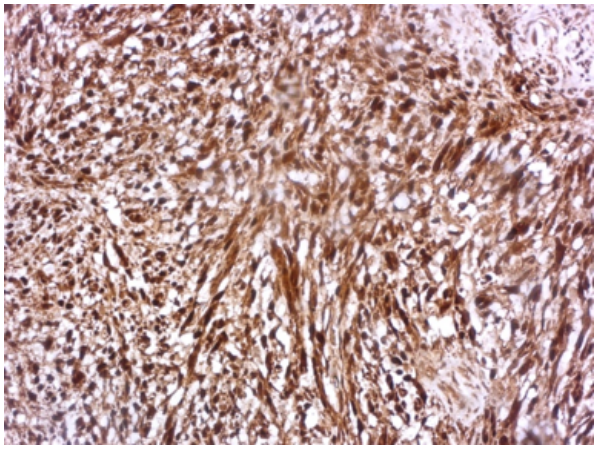

Fig. 4: Immunohistochemistry showing spindle cells diffusely positive for S100 and Ki67

3. Discussion

Schwannoma, also known as Neurilemmoma or neurinoma of 'Verocay', is derived from the schwann cells of any peripheral autonomic or cranial nerves except olfactory and optic.[6,7] Mostly they appear as solitary lesions, while multiple lesions may be associated with syndromes or schwannomatosis.[5] About 25 to 48% of the cases occur in the head and neck region with only 1% showing an intraoral origin.[2,3] Tongue is the most common intraoral site followed by palate, floor of the mouth, buccal mucosa, lip and gingiva.[3]

Schwannomas of the tongue arise from the hypoglossal nerve, but it is difficult to distinguish between tumors of the lingual, hypoglossal and glossopharyngeal nerve origin. The age of onset is usually between 20 to 50 years of age, but recent reports show a predilection for children between 10-13 years of age. Lesion does not show any gender predilection. Typically they are asymptomatic and slow growing benign neoplasms. They may present with mild pain when they become large in size and impinges on the affected nerve.[1,2] Being uncommon, schwannomas are not generally included in the differential diagnosis of intraoral soft tissue tumors. Even though tongue is the most common intraoral tumour location of schwannoma, a Pubmed search revealed only less than 50 cases of lingual schwannomas being reported in the literature in the past 20 years.[8] This may point out towards the cases being overlooked or under reported.

Schwannomas may pose a diagnostic challenge as they are clinically indistinguishable from other benign soft tissue tumors. Hence biopsy and histological examination are essential for a confirmative diagnosis. Complete surgical excision is the treatment of choice with solitary lesion and the chances of recurrence/ malignant transformation are very rare (8-10%).[9,10] In the present case, an excisional biopsy was done and the patient did not present with any symptoms of nerve injury post operatively as the lesion was small and well defined.

The H & E stained tissue section usually shows a thin fibrous capsule and proliferation of schwann cells in two microscopic patterns: Antoni type A and type B. The Antoni type A cells are closely packed, well organised bundles with elongated, palisaded nuclei. The palisaded arrangement is noted around central acellular, eosinophilic areas known as Verocay bodies. Antoni type B tissue meanwhile is less cellular and the spindle cells are randomly arranged within a loose, myxomatous stroma. The tumor cells show a S100 diffuse, positive immunohistochemical reaction. Degenerative changes such as hemorrhage, hemosiderin deposits, inflammation, fibrosis, and nuclear atypia can be seen in Ancient schwannomas.[2,7] In the present case, histopathologic findings were pointing out towards an Ancient schwannoma as degenerative changes were also noted. S100 diffuse positivity and Ki67 proliferation index around 10% were suggestive of a definitive diagnosis of Schwannoma.

4. Conclusion

The schwannoma of the tongue is uncommon, especially in younger age group, and there are only a very few cases reported in the literature. This may point out towards the cases being overlooked or under reported. This case report emphasizes the inclusion of uncommon tumors of connective tissue origin in the differential diagnosis of a growth on the tongue. Schwannoma of the tongue is often not even considered as a possible diagnosis during clinical practice. The rarity of this lesion warrants careful consideration as this may be clinically indistinguishable from other soft tissue tumors of the oral cavity such as fibroma, neurofibroma, benign tumors of salivary glands, leiomyoma, rhabdomyoma, lymphangioma, and hemangioma. The definitive diagnosis requires a histopathologic evaluation. Treatment is complete surgical excision of the lesion. Chances of recurrence or malignant transformation of the lesion are highly unlikely.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Author biography

Deepthi TR, Medical Officer https://orcid.org/0000-0002-5744-3916

Prem Sasikumar, Associate Professor

Adarsh VJ, Associate Professor

Vani MH, Private Practitioner

Anu Vijayan, Associate Professor

Kavya Maheesan, Assistant Professor https://orcid.org/0000-0003-1653-7665

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