Available online at https://www.mmpubl.com/

International Journal of Head and Neck Pathology

Case Report

Neurilemmoma at the Junction of Hard and Soft Palate: A Case Report

Kaaviya Tharsini P S [1], V Veeravarmal [2], Madhavan Nirmal[3], Mohamed Nassar[4]
[1]Post Graduate student, [2]Professor & Head,[3]Professor, [4]Associate Professor, Department of Oral and Maxillofacial Pathology and Microbiology & Oral Microbiology, Government Dental College & Hospitals, Cuddalore, Chidambaram

How to cite*: *Kaaviya T P S, Neurilemmoma at the Junction of Hard and Soft Palate: A Case Report. Int J Head And Neck 2024; 7(1):15-19.

DOI: *https://doi.org/10.56501/intjheadneckpathol.v7i1.1123*

Received:20/03/2024 *Accepted:28/03/2024* *Web Published:26/04/2024*

Abstract

Neurilemmoma is a benign peripheral nerve sheath tumour arising from schwann cells. It is relatively uncommon; around 25% to 48% of all cases occur in the Head and Neck region. Only 1% occur in the oral cavity and tongue being the most common site. Here, we report a case of a 40-year-old female patient with swelling in the junction of the hard and soft palate on the left side for 15 years. Due to its duration, site, and nature, it mimicked a glandular neoplasm. Clinical, histopathological, and immunohistochemical findings confirmed it as a neural tumour -Neurilemmoma.

Keywords: neurilemmoma, palate, S100 proteins

Address for Correspondence:
Dr. Kaaviya T P S
Post Graduate
Department Of Oral & Maxillofacial Pathology & Oral Microbiology
Government Dental College & Hospital,
Cuddalore, Annamalai nagar, Chidambaram – 608002
Email: varmal.opath@yahoo.com
Ph: 9443659412

INTRODUCTION

Benign soft tissue lesions that occur in the oral cavity comprised of salivary gland neoplasms, neural tumours, muscle tumours, as well as non-neoplastic lesions like fibroepithelial polyps, and mucoceles.[1] Neurilemmoma is a benign neural tumour arising from schwann cells present on the nerve sheath of peripheral, cranial (except for the optic and olfactory), or autonomic nerves.[2] It mostly occurs as an encapsulated, slow-growing, painless, solitary tumour. Here, we report a case of a neural tumour in a 40-year-old female which presented as a swelling in the junction of the hard and soft palate on the left side.

Case report

A 40-year-old woman presented with a swelling in the left throat region that has persisted for approximately 15 years which was accompanied with mild pain and dysphagia over the last year. She gave the medical history of iron deficiency anemia and under medications for the past 5 months. She was moderately built and nourished. No other abnormalities were detected on general examination. On intra-oral examination, a well-defined, solitary swelling measuring approximately 2x2 cm was observed at the junction of the hard and soft palate on the left side. It appeared smooth and had a reddish-pink hue, likely due to dilated capillaries (Figure [i]). Palpation confirmed the inspectory findings, and it was firm, mobile, non-tender, and blanched on pressure. It was provisionally diagnosed as pleomorphic adenoma based on location, duration, and clinical presentation by the clinicians. The lesion was excised in four bits under local anesthesia (Figure [ii]) and two bits were processed for microscopic evaluation. On microscopic examination, varying numbers of spindle-shaped cells with elongated fusiform nuclei and capillaries were seen. This excluded the differential diagnosis of pleomorphic adenoma, lipoma, and fibroma. The spindle cells in the tumour area made us suspect myoepithelioma, leiomyoma, and benign neural tumours too. Immunohistochemistry was carried out using smooth muscle actin, for which it was negative. This excluded the differential diagnoses of myoepithelioma and leiomyoma. In the meantime, other bits were processed which on microscopic evaluation revealed areas of Antoni A and Antoni B region (Figure [iii], [iv], [v] and [vi]). Therefore, the present case was diagnosed as Neurilemmoma and further confirmed by immunohistochemistry using S100 protein (Figure [vii]).

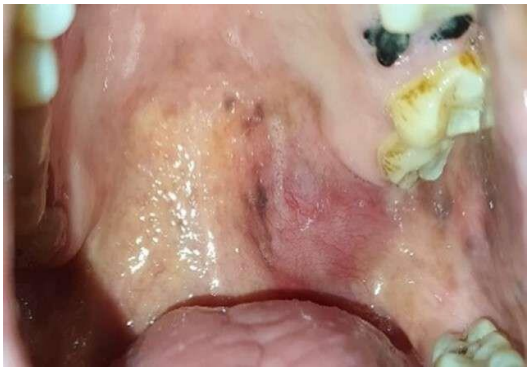

Figure [i] A swelling of size 2x2 cm seen in the left junction of soft and hard palate.

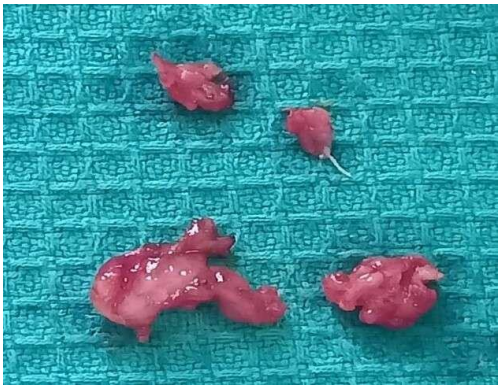

Figure [ii] - Post-operative specimen received as multiple bits.

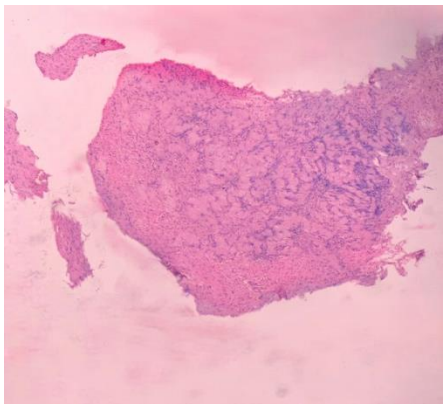

Figure [iii] - H&E stained lesional tissue showed well-localized spindle cell tumour. (x40)

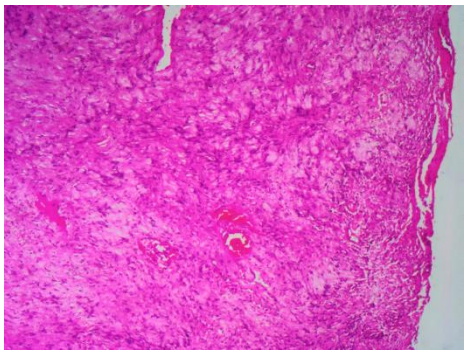

Figure [iv] - An encapsulated mass with antoni A areas and verocay bodies are seen in a fibrous background under H&E (x100)

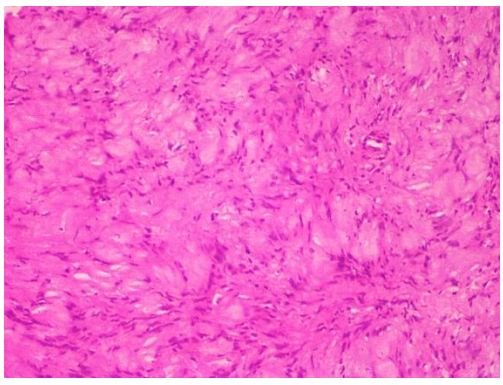

Figure [v] Verocay bodies with palisaded nuclei and acellular eosinophilic areas. (x400)

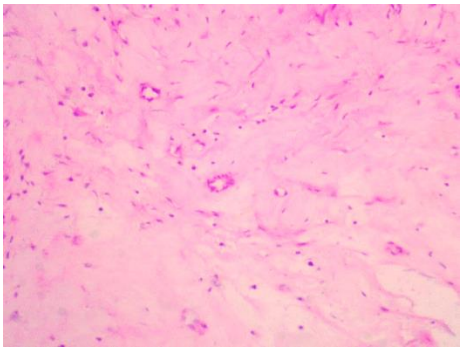

Figure [vi] Antoni B area showing myxoid stroma. (x400)

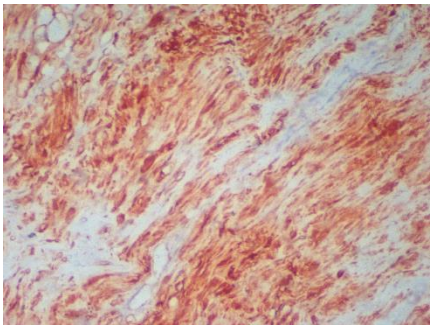

Figure [vii] Intense positivity for S-100 in spindle cells observed in the tumour area. (x400)

Discussion

Neurilemmoma is a benign nerve sheath tumour arising from schwann cells. It was first reported as 'neurinomas' by Jose Juan Verocay (1910), later as 'neurilemmomas' by Arthur Purdy Stout (1935). In 1968, Harkin and Reed used the term 'schwannoma' since ultrastructural studies demonstrated that these are almost entirely composed of schwann cells.[3]

It usually arises 90% sporadically, 5% in association with multiple meningiomas in patients with or without Neurofibromatosis 2 (NF2), 3% in patients with NF2, 2% in those with schwannomatosis, and also in few cases of Carney's complex.[4],[5] The exact etiology is unknown, however, chronic irritation, external injury, oxidative stress, exposure to radiation, and genetic susceptibility have been presumed as risk factors in the literatures.[6] The possible pathogenesis for most cases points to a mutation in the NF2 gene that results in loss of merlin in both sporadic and other NF2-related neurilemmomas. No other consistent genetic alterations have been identified. [4],[5],[7]

Neurilemmoma has a slight female predominance and occurs commonly in the third to fourth decades as in our case, however, it can occur at any age. Around 25% to 48% of cases occur in the head and neck region, with the parapharyngeal space being the most common site.[8] Only 1% occurs in the oral cavity where tongue is the most common site, followed by buccal mucosa, floor of the mouth, lips, palate, and gingiva. Rarely, it also has been reported as a central lesion within the mandible, arising from the mandibular nerve. Typically, the lesion grows within a capsule and remain peripherally attached to the parent nerve. The location of the tumour, in the present case, made us believe that it is from the greater palatine nerve. Intraorally, these present as slowly enlarging, painless submucosal nodules which are slightly mobile.[9] The presenting symptom depends on the size and location of the tumour. Schwannomas of the base of the tongue may present with dysphagia or a feeling of a lump in the throat.[10] Size of the tumour ranges between 0.5 to 3 cm in size, rarely exceeding 5 cm.[6] Due to slow growing nature it may go unnoticed by the patient for years, which was the exact presentation of the present case.

Microscopically, it is encapsulated and has two cellular patterns namely Antony A and Antony B. Antony A areas are highly cellular comprising of spindle cells. Nuclear palisading, whorling of the cells, and verocay bodies are seen in highly differentiated Antoni A areas.[5] Verocay bodies are observed as a stacked arrangement of two rows of spindle cells with palisading nuclei, separated by an acellular zone. This acellular zone is made up of reduplicated basement membrane material rich in laminin and collagen IV [3],[4] which was first described by Jose Juan Verocay, a Uruguayan physicist.[3] Antoni B areas are less cellular and loosely arranged in a myxoid matrix accompanied by strands of collagen fibers. These might exhibit hyalinization around vessels, hemorrhage, microcysts, and inflammatory infiltrate. [3],[4] The present case also showed both Antoni A with verocay bodies and Antoni B areas.

Histological variants include ancient/degenerative, cellular, plexiform, and epithelioid type. The ancient/degenerative type shows marked nuclear atypia and is usually infiltrated by large numbers of siderophages and histiocytes. This should not be confused with malignant change due to marked nuclear atypia.[4] Cellular type is predominantly or exclusively composed of Antoni A areas that lack verocay bodies. The cells are arranged in intersecting fascicles and whorls of Schwann cells. The Antoni A areas may display long, sweeping fascicles of schwann cells, sometimes arranged in a *herringbone fashion*. Since, it had more cellularity, mitotic activity, and occasional presence of bone destruction, it was diagnosed as malignant in more than one-fourth of cases. [4],[5] Epithelioid neurilemmoma has predominantly epithelioid schwann cells. These are small, superficial, well-encapsulated lesions with a high level of architectural organization and cytologic differentiation like conventional neurilemmoma. It is composed of small, round

schwann cells arranged singly, in small aggregates, or in cords within a collagenous or partially myxoid stroma. The cells may be associated with dense collagen cores forming irregular collagen rosettes like those seen in the neuroblastoma-like schwannoma. Plexiform or multinodular type may or may not be apparent macroscopically. These are encapsulated and usually occur in the skin. [5] Rarely, hybrid tumours can also arise which show areas of both neurofibroma and schwannoma within the same tumours, and these appear to be more common in schwannomatosis and NF1 and 2. [11]

Immunohistochemically, the tumour cells of neurilemmoma show strong expression for S100 protein and Sox 10. It also shows positivity for Calretinin (Antoni B areas), Podoplanin (Antoni A areas), TLE1 [Transducer like enhancer of split 1], CD 34, CD 56, and PGP 9.5[Protein Gene Product 9.5].[12] The present case showed strong immunopositivity for S100 protein confirming the diagnosis of Neurilemmoma.

Conclusion

Neurilemmoma is a slow-growing, benign neural tumour that presents rarely in the oral cavity. This should be differentiated from other lesions like salivary gland neoplasms especially when it occurs in the posterolateral aspect of the palate. It is a radioresistant tumor treated by conservative surgical excision.[10] The prognosis is good, chances of recurrence and malignant transformation are rare.[5]

Financial support and sponsorship
Nil

Conflicts of interest
There are no conflicts of interest

REFERENCES

[1] Amir R, Altman KW, Zaheer S. Neurilemmoma of the hard palate. Journal of Oral and Maxillofacial Surgery. 2002;60:1069–1071.
[2] Cohen M, Wang MB. Schwannoma of the tongue: two case reports and review of the literature. Eur Arch Otorhinolaryngol. 2009;266:1823–1829.
[3] Joshi R. Learning from eponyms: Jose Verocay and Verocay bodies, Antoni A and B areas, Nils Antoni and Schwannomas. Indian Dermatol Online J. 2012;3:215.
[4] Hilton DA, Hanemann CO. Schwannomas and Their Pathogenesis. Brain Pathology. 2014;24:205–220.
[5] Goldblum JR, Folpe AL, Weiss SW. Enzinger & weiss's soft tissue tumors. Seventh edition. Philadelphia, PA: Elsevier; 2020.
[6] Bhola N, Jadhav A, Borle R, et al. Schwannoma of the Tongue in a Paediatric Patient: A Case Report and 20-Year Review. Case Reports in Dentistry. 2014;2014:1–5.
[7] Helbing D-L, Schulz A, Morrison H. Pathomechanisms in schwannoma development and progression. Oncogene. 2020;39:5421–5429.
[8] Franzen A, Koegel K. Neurinome im Halsbereich. Laryngo-Rhino-Otol. 1996;75:250–253.
[9] Baliga M, Uppal N, Ramanathan A. Schwannomas of the head and neck: a case series. J Maxillofac Oral Surg. 2009;8:283–286.
[10] Hsu Y-C, Hwang C-F, Hsu R-F, et al. Schwannoma (neurilemmoma) of the tongue. Acta Oto-Laryngologica. 2006;126:861–865.
[11] Harder A, Wesemann M, Hagel C, et al. Hybrid Neurofibroma/Schwannoma is Overrepresented Among Schwannomatosis and Neurofibromatosis Patients. American Journal of Surgical Pathology. 2012;36:702–709.
[12] Guedes-Corrêa J, Cardoso R. Immunohistochemical Markers for Schwannomas, Neurofibromas and Malignant Peripheral Nerve Sheath Tumors—What Can the Recent Literature Tell Us? Arq Bras Neurocir. 2018;37:105–112.

Published by MM Publishers
https://www.mmpubl.com/ijhnp