Neurilemmoma of the Hard Palate: Report of a Case and Review of Literature

Nitin P Chikhale, Aradhana Mishra, Richa D Patel, Uma Pankaj Chaturvedi, V Jayalakshmi, Susan Cherian

ABSTRACT

Neurilemmoma, also known as schwannoma, is a benign tumor arising from the neural sheath Schwann cells. This tumor has a predilection for the head and neck region but it is rarely found in the oral cavity (1%) where tongue is reported to be the favored site. We report a rare case of neurilemmoma of the hard palate along with a review of literature of this entity.

Keywords: Neurilemmoma, Schwannoma, Schwann cells, Hard palate, Antoni A, Antoni B, Oral cavity, Head and Neck.

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INTRODUCTION

Neurilemmoma (schwannoma) is a benign, encapsulated neoplasm that arises from Schwann cells of the neural sheath of the peripheral, cranial or autonomic nerves. It can be found at almost any site. Approximately, 25 to 45% of the neurilemmomas are seen in the head and neck region; however, intraoral lesions are rare. Most of the intraoral schwannomas are located in the tongue. Other less frequent locations are the buccal mucosa, palate, base of mouth, gingiva and lips. The typical tumor is solitary, slow growing occurring in a middle aged adult.

Clinically these tumors could be mistaken with other entities such as fibroma, lipoma or salivary gland tumors as well as mucous retention cyst, palatal abscess, lymphoma and squamous cell carcinoma on account of their site of occurrence.⁴⁻⁷

We report a case of schwannoma presenting as a slow growing hard palate mass.

CASE REPORT

A 42-year-old female presented to the ENT department of our hospital for some unrelated complaint. A mass on the hard palate was incidentally found during the course of examination. Patient was unaware of the mass. On examination a solitary firm mass measuring 2×2 cm was seen on the left side of the hard palate. Overlying mucosa was similar in color and texture to the surrounding mucosa.

Computed tomographic (CT) scan showed an abnormal soft tissue mass along the oral surface of the hard palate

(Fig. 1). The mass measured $0.8 \times 1.8 \times 2.5$ cm. Erosion of the cortex was noted on the left side of the hard palate starting from the level of the second premolar extending up to the level of the second molar. There was no extension of the soft tissue in to nasal cavity.

The mass was excised intraorally and sent for histopathological examination. On gross examination, there were three irregular brownish tissue bits. Cut surface of all the bits was firm, grayish white.

Histopathological examination showed an encapsulated lesion composed of spindle cells arranged in two patterns: Antoni A and B. The Antoni A areas showed closely packed spindle cells arranged in rows with elongated, palisaded nuclei with eosinophilic Verocay bodies between them. The Antoni B areas showed hypocellularity with spindle cells arranged in a loose myxoid matrix (Figs 2 and 3).

Based on these findings, a diagnosis of neurilemmoma was made.

DISCUSSION

Neurilemmoma is a benign tumor arising from the neural sheath Schwann cells of the peripheral cranial or autonomic nerves. It is solitary slow growing, encapsulated and usually asymptomatic. Neurilemmoma was first reported by Verocay in 1910. It is almost always solitary. It can be found at almost any site but it is more common on the head and neck. Approximately one-third of all neurilemmomas





Fig. 1: CT scan image in coronal section showing abnormal soft tissue mass along the oral surface of the hard palate causing erosion of the cortex on the left side of the hard palate without extension into the nasal cavity

occur in the head and neck region but they are rarely found in the oral cavity (1%).²⁻⁵

Within the oral cavity, the neurilemmomas occur most frequently in the tongue. ¹² Other less frequent locations are the buccal mucosa, palate, base of mouth, gingiva and lips. ³⁻⁵

Leu and Chang reviewed a series of 52 cases of schwannomas of the head and neck region. Seven of these cases were located in the oral cavity. Out of these seven cases, one was located in the tongue, two in the submasseteric region, one in the soft palate, one in the lower lip and one in the hard palate.¹³

Wright and Jackson reported 146 cases of neurilemmoma of the oral cavity soft tissue. Of these, 52% involved the tongue, 19.86% buccal or vestibular mucosa, 8.9% the soft palate, and the remainder 19.24% were in the gingiva and lip.¹

Gupta et al studied 303 cases of benign neurilemmomas of which 136 were located in head and neck region. Out of these, 18 cases were located in oral cavity, eight were located in the tongue, three each in soft palate, hard palate, lip and one in the floor of mouth.¹⁴

Wakoh et al reported 22 cases of neurilemmomas of the head and neck of which seven were located on the palate, four on the tongue, three in the floor of mouth, two in the buccal mucosa, two on the mental skin, two on the lip, one on the gingiva and one in the temporal region. ¹⁵

Clinically, these benign tumors are easily mistaken for other entities, such as lipoma and pleomorphic adenoma on account of their slow growth and absence of neural symptoms and also to mucous retention cyst, minor salivary gland tumors, palatal abscess, fibroma, lymphoma and squamous cell carcinoma on account of their site of occurrence.⁴⁻⁷

Histologically, the characteristic features of a neurilemmomas are encapsulation and dimorphic growth pattern composed of Antoni A and B areas (Fig. 2). The Antoni A areas are cellular and composed of spindle cells with wavy nuclei. Highly differentiated areas show 'nuclear palisading' and Verocay bodies (eosinophilic cell bodies encircled by rows of nuclei) (Fig. 3). The Antoni B areas are hypocellular and the cells lack orientation. They are loosely arranged in a myxoid matrix accompanied by thin strands of collagen. Occasional mast cells may be identified in the Antoni B areas. In addition thick walled blood vessels are unusually prominent. Degeneration with cyst formation should not be mistaken for necrosis, and rare atypical nuclei (so called ancient change) should not be taken as a sign of malignancy. Cellular neurilemmoma differs from the classic neurilemmoma on account of its increased cellularity, nuclear pleomorphism and several mitotic figures (up to 4 per 10 hpf on an average). 16

Immunohistochemically neurilemmomas show consistent positivity for S100 protein. Tumoral cells within Antoni A show greater intensity scores compared to Antoni B. Capsular EMA and CD34 staining is observed in neurilemmoma.¹⁷

Treatment of benign neurilemmomas consists of local excision of the tumor. According to Asaumi et al ultrasonography, CT and magnetic resonance imaging (MRI) may be helpful diagnostics tools for estimation of tumor margins, the lesion composition and infiltration in to the surrounding structures. Recurrences as well as malignant transformation are rare events. ¹⁸

CONCLUSION

Neurilemmoma must be considered in the differential diagnosis of lesions of the oral cavity as it is clinically indistinguishable from other benign lesions in the same region. Final diagnosis requires a histopathological evaluation. Local excision of the tumor is the treatment of choice. Prognosis is good as recurrence is unknown.

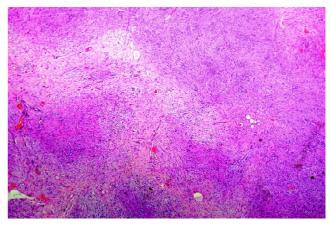


Fig. 2: Photomicrograph showing highly ordered cellular component (Antoni A) area with Verocay bodies and a loose myxoid component (Antoni B) area (H and E: 100x)

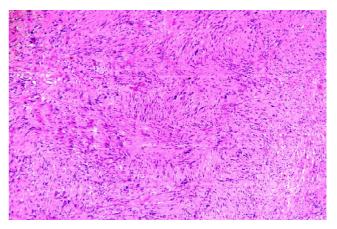


Fig. 3: Photomicrograph from hypercellular (Antoni A) area showing closely packed spindle cells with wavy nuclei and Verocay bodies (eosinophilic cell bodies encircled by rows of nuclei) between them (H and E: 400x)



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ABOUT THE AUTHORS

Nitin P Chikhale

Senior Resident, Department of Pathology, Bhabha Atomic Research Centre, Mumbai, Maharashtra, India

Aradhana Mishra

Senior Resident, Department of Pathology, Bhabha Atomic Research Centre, Mumbai, Maharashtra, India

Richa D Patel

Postgraduate Resident, Department of Pathology, Bhabha Atomic Research Centre, Mumbai, Maharashtra, India

Uma Pankaj Chaturvedi (Corresponding Author)

Consultant Pathologist, Department of Pathology, Bhabha Atomic Research Centre, Mumbai, Maharashtra, India, Phone: 02225598355 e-mail: umapc@rediffmail.com

V Jayalakshmi

Postgraduate Resident, Department of Pathology, Bhabha Atomic Research Centre, Mumbai, Maharashtra, India

Susan Cherian

Consultant Pathologist, Department of Pathology, Bhabha Atomic Research Centre, Mumbai, Maharashtra, India