

CASE REPORT

Neurofibroma of Soft Palate: A Rare Case Report

Anil Hanakere Thimmaiah, Lakshmi Shantharam, Smitha Soubhagya Gangaraj, Suguna Vijay

ABSTRACT

Neurofibroma of soft palate is a rare tumor with this report being the fourth case reported in English literature. We are reporting this case of isolated neurofibroma of soft palate for its rarity. The mass was excised by intraoral approach without prior tracheostomy. Histopathological examination showed features of neurofibroma with secondary changes.

Keywords: Neurofibroma, Soft palate, Excision, Schwannoma.

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INTRODUCTION

Neurofibroma is a tumor arising from the nerve sheath cells of Schwann. The lesions are usually discrete firm masses often showing cystic degeneration. Neurofibromas are fibrous lesions with nerve fibers traversing the tumor. They are usually part of generalized neurofibromatosis. Solitary neurofibromas are very rare.

CASE REPORT

A 75 years old man presented in ENT OPD with a swelling in the oral cavity, noticed by him 3 months ago. Associated symptoms were altered speech and snoring. There were no symptoms of dysphagia, dyspnea, weight loss or loss of appetite.

Examination of the oral cavity revealed a huge mass occupying the left side of the soft palate, extending anteriorly to the hard palate and crossing the midline, with shift of uvula to right side. It was soft swelling with cystic consistency and nontender.

The routine laboratory tests and urine analysis were normal. CT scan showed a large hypovascular mass in the left side of the soft palate. FNAC showed benign cystic lesion of soft palate.

Under general anesthesia, with tracheostomy arrangement standby, a blind nasotracheal intubation was done. A tonsillar Boyle Davis mouth gag was applied and tumor exposed well. A vertical incision was made over it (Fig. 1). The mass was dissected and excised *in toto*. The wound closed in layers with absorbable suture materials. Postoperative period was uneventful. Patient was kept in propped up position and observed for development of respiratory difficulty.

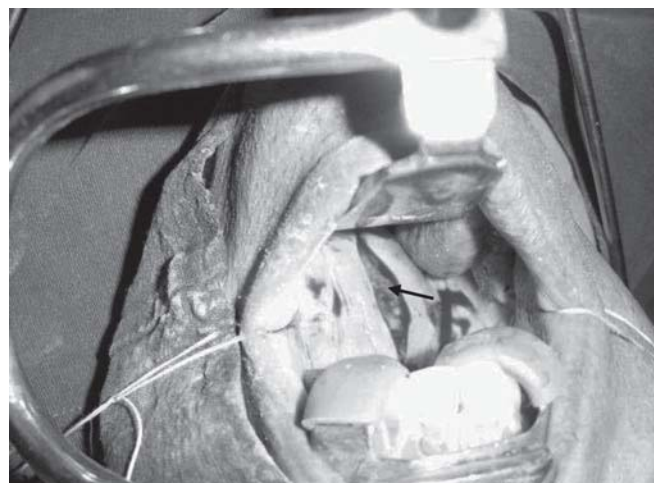


Fig. 1: Intraoperative picture showing a vertical incision over palatal swelling

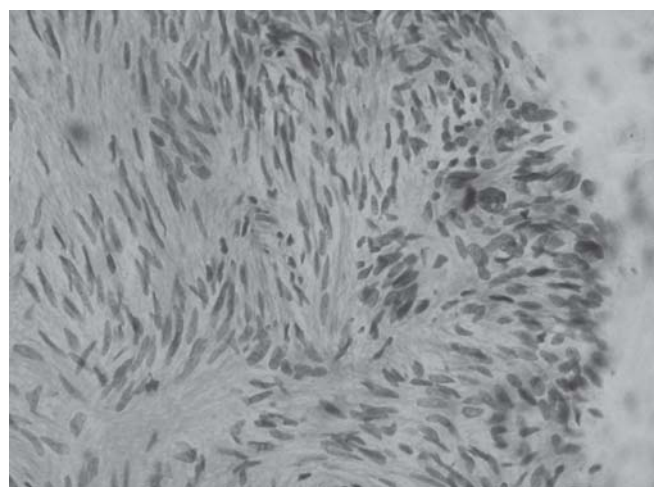


Fig. 2: Photomicrograph showing elongated cells, bend or wavy nuclei, separated by abundant fine and equally wavy collagen fibers (H&E, 40x)

Histopathological examination of the tumor showed a cellular spindle cell lesion with cells arranged in sheets and fascicles with focal areas showing palisade. Extensive secondary changes with hyaline degeneration, cystic changes and hemorrhage were noted. There was no evidence of malignancy (Fig. 2). A diagnosis of neurofibroma with secondary changes was made. Patient was followed up for 3 years and there was no recurrence.

DISCUSSION

Neurofibromas contain a mixture of neural axonal or dendritic fibers and Schwann cell elements. They are

frequently associated with Von Recklinghausen's disease as part of a generalized neurofibromatosis.¹ The incidence of neurofibromas reported in the head and neck is 37%.² Neurofibromas of oropharynx are extremely rare.³

The most common location of neural tumors in the oral cavity is the tongue and, thereafter, the sites in order of frequency are the buccal mucosa, the floor of the mouth, palate, lips and gingiva.⁴

There are three cases of solitary neurofibroma of soft palate reported in literature to our knowledge. This is the fourth case of solitary neurofibroma not associated with neurofibromatosis.

Sinha et al reported the first case of isolated neurofibroma of soft palate not associated with Von Recklinghausen's disease in 2002. They faced difficulty in intubation and removed the tumor after tracheostomy.⁵ Tracheostomy arrangement should be kept standby in case of difficult intubation that is common in those cases. This can be avoided by blind nasotracheal intubation or fiberoptic-assisted nasotracheal intubation and careful postoperative monitoring.

The second case was reported by Mazzoleni et al in 2009 in which they had to excise the mass along with extraction of upper left-hand second molar, which was attached to the mass through its palatal root.⁶ The third case was reported by Choi et al in 2011.⁷

A solitary neurofibroma has to be differentiated from the schwannoma. A schwannoma is encapsulated, eccentric to the nerve and is composed of Schwann cells. A neurofibroma merges with the neighboring tissues and incorporates the nerve. So, it is not possible to separate the lesion from the nerve in case of a neurofibroma and axons can be demonstrated within the tumor.⁸

Microscopically, neurofibromas are formed by combined proliferation of all the elements of a peripheral nerve, axons, Schwann cells, fibroblasts and perineural cells. Schwann cells are the predominant cellular element. Most have markedly elongated nuclei with a wavy, serpentine configuration and pointed ends. They are immunoreactive to S-100 protein.⁸

The cause of solitary neurofibroma is unknown. However, neurofibromatosis is inherited as an autosomal dominant trait with a high degree of penetrance but variable expressivity. Management of neurofibroma is by surgical excision.²

Few neurofibromas show small cystic changes due to myxoid degeneration. Cystic degeneration is more commonly seen in schwannomas than neurofibromas.⁹

Malignant transformation is also noted in neurofibromas.

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ABOUT THE AUTHORS

Anil Hanakere Thimmaiah (Corresponding Author)

Associate Professor, Department of ENT, Kempegowda Institute of Medical Sciences, KR Road, VV Puram, Bengaluru-560004, Karnataka India, e-mail: smitha_praveen@hotmail.com

Lakshmi Shantharam

Senior Resident, Department of ENT, Kempegowda Institute of Medical Sciences, Bengaluru, Karnataka, India

Smitha Soubhagya Gangaraj

Assistant Professor, Department of ENT, Kempegowda Institute of Medical Sciences, Bengaluru, Karnataka, India

Suguna Vijay

Professor, Department of Pathology, Kempegowda Institute of Medical Sciences, Bengaluru, Karnataka, India