

# Intraoral neurilemmoma of mental nerve mimicking as traumatic fibroma: An unusual presentation - A case report and literature review

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## Abstract

Neurilemmoma, neurinoma or schwannoma is a benign tumor of neurogenic origin, arising from the myelin sheath of a neuron. Head and neck region accounts for 25%–48% of these tumors reported till date. On the contrary, intraoral sites form only 1% of the cases accounted. Although a few cases of intraoral neurilemmoma of mental nerve has been reported in English literature, none has been reported to arise from the terminal fibers of the mental nerve. In this particular article, we present a case where the tumor was noted in the lower labial mucosa.

**Keywords:** Neurilemmoma, neurinoma, schwannoma

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## INTRODUCTION

Neurilemmoma also known as schwannoma, neurinoma, perineural fibroblastoma and peripheral glioma is a benign tumor of nerve sheath or myelin sheath which are formed by Schwann cells.<sup>[1-5]</sup>

Although head and neck region accounts for about 25%–48%,<sup>[1-5]</sup> this tumor is not very common in the oral cavity. Among the intraoral sites, tongue is the most common site of origin.<sup>[6]</sup> Rare sites of occurrence include intrabony lesions usually seen in the posterior mandible.<sup>[4]</sup>

Unlike neurofibromas, neurilemmoma presents as a solitary growth. Although pain may be present at certain instances, the tumor is usually painless. Usually, a slow growing tumor but also associated with rapid growth in some cases.<sup>[7]</sup>

Neurilemmomas, though can occur in cranial, spinal and peripheral nerves, have a strong predilection for sensory nerves as they have a thick layer of myelin sheath.<sup>[3]</sup> These tumors do not show any sex predilection and are usually found in young- and middle-aged persons.<sup>[1-7]</sup>

Histopathologically, neurilemmomas are encapsulated tumors composed solely of Schwann cells. There are two different patterns, in which the tumor cells are arranged. The more organized and distinctive pattern is the Antoni A pattern, where the Schwann cells are arranged in palisaded clusters. These palisaded clusters occur around an eosinophilic mass to form a Verocay body. The Antoni B pattern comprises an irregular arrangement of Schwann cells in a loose stroma with small vacuoles.<sup>[6,8]</sup>

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Neurilemmomas might undergo degenerative changes with cyst formation, calcification and nuclear atypism. Such tumors are termed as ancient schwannomas and are seldom associated with small intraoral tumors.<sup>[9]</sup> Although controversial neurilemmomas have been shown to undergo malignant transformation.<sup>[10]</sup>

Excision is the treatment of choice and is associated with nearly 100% cure rate, and the tumor does not recur.<sup>[1]</sup>

In our case, unlike the usual presentation, the tumor showed a rapid growth in a relatively less common site, the lower labial mucosa.

### CASE REPORT

A 16-year-old girl reported to the Department of Oral and Maxillofacial Surgery, Sri Ramachandra Medical Centre with complaints of a swelling associated with pain in the left lower lip region for a duration of 1 month.

Detailed history taking revealed that the patient has the habit of lip biting since childhood. The patient was apparently normal 1 month back after which she developed a small swelling on her lower lip which rapidly progressed over a period of 2 weeks. The swelling interfered with the patient's daily activities. Pain was present only when there was accidental irritation to the swelling.

On inspection, the swelling was present as a pedunculated growth on the left lower labial mucosa in relation to 33, 34 and 35. The swelling was oval-shaped measuring about 2 cm × 1.5 cm. The surface of the swelling was smooth and pale pink [Figure 1].

On palpation, the swelling was firm in consistency associated with tenderness, freely mobile and attached to the underlying tissue through a peduncle.

Based on this clinical examination and history taking, a

provisional diagnosis of traumatic fibroma was made and was planned for an excisional biopsy of the lesion.

The surgical procedure was performed under local anesthesia after obtaining the informed consent of the patient. Two percent lignocaine with 1:200,000 adrenaline was administered through local infiltration. The peduncle was ligated with the help of silk ties to avoid any hemorrhage [Figure 2]. The lesion in-toto was excised from the underlying tissue [Figure 3]. There was no mucosal defect at the excised site [Figure 4]. The surgical site was closed using black silk sutures.

The excised lesion was fixed in 10% buffered formaldehyde solution. Grossing of the specimen was done [Figure 5]. The specimen was sectioned and stained using hematoxylin and eosin. The obtained sections were observed under a light microscope in ×4, ×20 and ×40 magnifications.

At ×4 magnification, the mounted specimen showed spindle cells dispersed in connective tissue stroma [Figure 6]. At ×20 magnification, the specimen showed streaming fascicles of spindle-shaped Schwann cells with wavy nuclei [Figure 7]. Antoni A pattern of cellular arrangement was noted at ×40 magnification. The Verocay bodies, which are the palisaded arrangement of nuclei around a central acellular eosinophilic area, were also noted at ×40 magnification [Figure 8].

Based on the above said histopathological findings, a final diagnosis of conventional schwannoma was given. Excision of the lesion had already been done, and the patient was followed up for 6 months without any evidence of recurrence.

### DISCUSSION

Initially termed as neurinoma in the year 1910 by Verocay, later, the term neurilemmoma was coined in the year 1935 by Stout. Now, commonly referred to as schwannomas after



Figure 1: Intraoral view showing the tumor in the left lower lip



Figure 2: Ligation of the peduncle



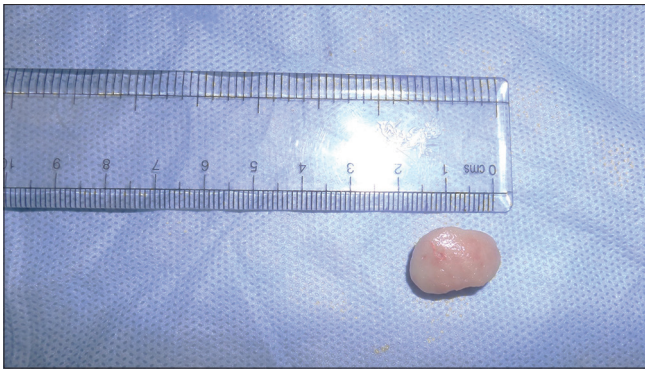


Figure 3: Excised tumor

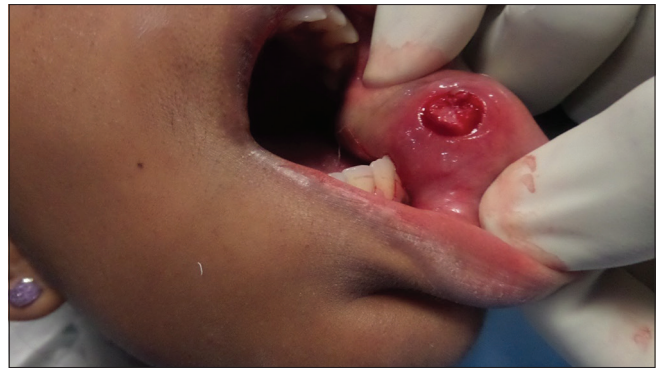


Figure 4: Surgical site postexcision showing the defect



Figure 5: Grossing

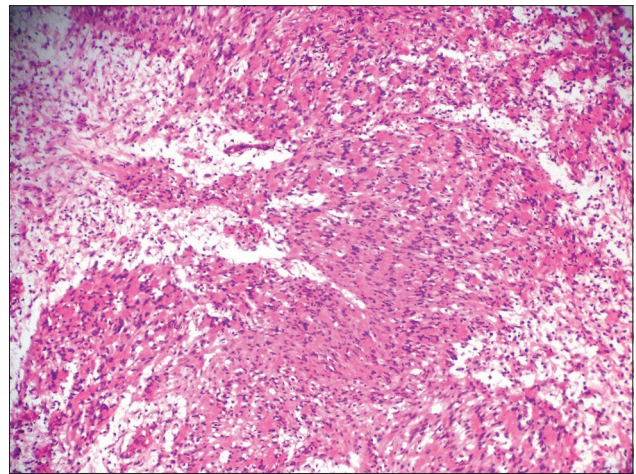


Figure 6: Spindle cells dispersed in connective stroma (H&E, x4)

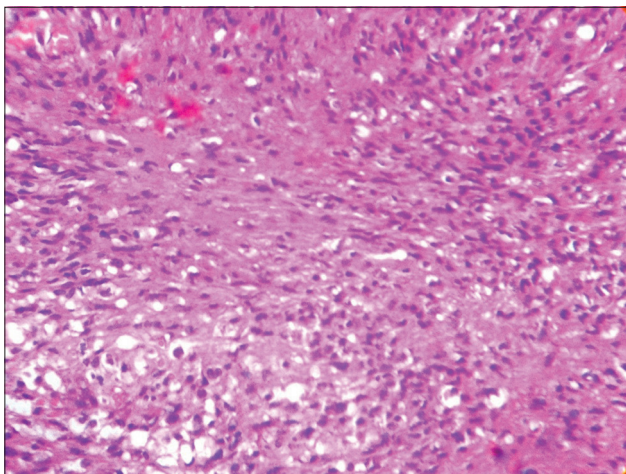


Figure 7: Streaming fascicles of spindle-shaped Schwann cells having wavy nuclei (H&E, x20)

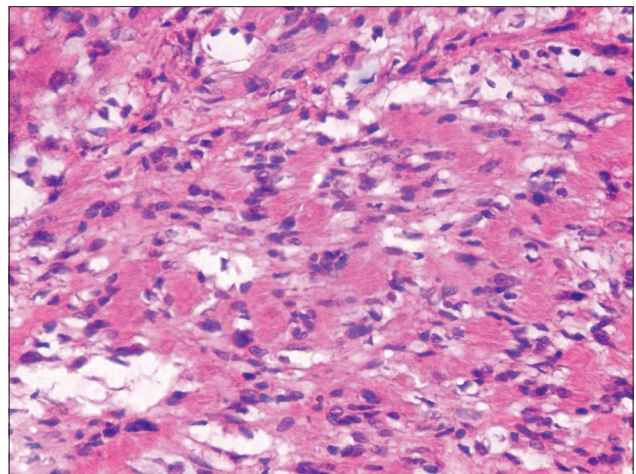


Figure 8: Antoni A: Cellular region - Nuclei palisaded arrangement around central acellular eosinophilic areas - Verocay bodies (H&E, x40)

the term was coined by Masson in 1932,<sup>[4]</sup> these tumors of neurogenic origin are usually benign and seldom show malignant transformation.

Head and neck area accounts for about 25%–48% of these tumors. Neurilemmomas most frequently occur along the

course of the vestibulocochlear nerve, followed by scalp, face, pharynx, parotid gland, middle ear and external acoustic canal.<sup>[1-7]</sup> Oral cavity accounts for only about 1% of all these neurogenic tumors, with the mobile tongue being the most common site of origin.<sup>[1-7]</sup> The other intraoral sites



include floor of the mouth, palate, mental nerve trunk, gingiva, buccal mucosa and lip.<sup>[1-7]</sup>

The sites of occurrence of neurilemmomas according to various authors were reviewed. Wright and Jackson analyzed 146 cases of intraoral neurilemmomas and found tongue accounts for 52% of the cases, followed by buccal mucosa and mucosa of gingiva and lip accounting for 19.8% and 19.24% of the cases, respectively, and the least being soft palate with 8.9%.<sup>[11]</sup> Gallo *et al.* studied 157 cases and found a result similar to that of Wright and Jackson, with 45.2% of their cases involving the tongue and 13.3% involving the buccal mucosa.<sup>[12]</sup> Gupta *et al.* in their study on 136 cases of neurilemmoma of the head and neck region found the neck to be the most common site with sixty cases, followed by parotid gland with ten cases. In the intraoral region, buccal mucosa accounted for nine cases and eight cases each in the tongue and pharynx.<sup>[13]</sup> Kun *et al.* analyzed 49 cases of neurilemmoma found 18 of those tumors were in the neck followed by 11 cases in the tongue.<sup>[14]</sup>

Clinically, neurilemmoma usually presents as an asymptomatic, slow-growing tumor. However, in case of intraosseous tumors, where mandible is the most common site of origin, pain, paresthesia and bony expansion are not uncommon.<sup>[4]</sup> Neurilemmoma can affect individuals of all age groups but most commonly found in the 2<sup>nd</sup> and 3<sup>rd</sup> decades of life.<sup>[1-7]</sup> Various authors have given different rates of sex predilection. William *et al.* in his study found a strong predilection for males,<sup>[15]</sup> while Lucas found a greater predilection for females.<sup>[16]</sup> Hatziotia and Asprides and Enzinger and Weiss in their studies found an equal sex predilection.<sup>[17,18]</sup>

The common differential diagnosis of intraoral neurilemmomas according to Wright and Jackson include cysts such as epidermoid and dermoid cysts or tumors such as lipomas, hemangiomas, granular cell tumor, leiomyoma and lymphangioma.<sup>[11]</sup> Histopathology is the only available modality for differentiating these clinically similar entities.<sup>[11]</sup>

The histological variants of neurilemmoma according to Kurtkaya-Yapicier *et al.*<sup>[19]</sup> include conventional, cellular, plexiform and melanotic variants. Conventional schwannomas are the most common variants and large old tumors with nuclear atypia are termed as ancient schwannomas and are a subtype of conventional schwannoma. According to Woodruff, although rare, only the conventional type undergoes malignant transformation. Cellular schwannoma first described by Woodruff in 1981, is a more recently described variant which shows

histological similarity to malignant peripheral nerve sheath tumors because of the disorderly pattern of arrangement of spindle cells. Plexiform schwannomas are relatively rare and vary in size. The small cutaneous tumors might not show the exact architecture of the lesion, whereas larger lesions are firm, multinodular and consist of “worm-like” components. The melanotic variant is the rarest among the four and usually involves the spinal nerves and the paraspinal ganglia.

Unlike the normal clinical presentation of a slow growing tumor, our case showed a rapid progression over a period of 1 month misleading us to give the first provisional diagnosis of traumatic fibroma as it was associated with a parafunctional habit.

Dentists should be aware of this rare benign neoplasm which might mimic as other common benign neoplasms. The treatment modality is being the same for most of such lesions. Still, this particular benign lesion has very minimal chance of recurrence and cases of malignant transformation have also been reported. Hence, patients with neurilemmoma have to be followed up on a regular basis.

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#### Conflicts of interest

There are no conflicts of interest.

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