

## **Case Report**

# **A RARE CASE OF SUBLINGUAL SCHWANNOMA: A CASE REPORT**

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## **ABSTRACT**

Schwannomas are benign, solitary, encapsulated and usually gradually increasing tumor origination from Schwann cells of the peripheral, cranial and autonomous nerve sheath. Most of the cases are seen in the head and neck region being uncommon in the oral cavity. Schwannomas of the sublingual space are exceedingly rare. This is a case report of schwannoma in a 46 year old male patient who had a 10 year history of asymptomatic swelling on the right side floor of the mouth. The clinical presentation and FNAC were inconclusive. Computerize Tomography imaging with contrast and histopathological findings were found to be consistent with Schwannoma. Complete surgical sublingual excision of tumor was done under general anaesthesia and the tumor enucleated with primary closure.

**Keywords:** Schwannoma, Floor of Mouth, Contrast CT Scan, Histopathology

## **INTRODUCTION**

Schwannoma is a benign neural tumor derived from spindle- shaped Schwann cells or nerve fibre sheath cells. The term Schwannoma has numerous synonyms such as neurilemmoma, neurinoma, neurolemoma, peripheral glioma, perineural fibroblastoma and peripheral nerve sheath tumor, but among these neurinoma, neurilemmoma and schwannoma are presently used (Baranovic *et al.*, 2006; Martins *et al.*, 2009).

Schwannoma is solitary, slow growing, benign encapsulated neural tumour arising from the nerve sheath of Schwann cells of the peripheral, cranial or autonomic nerves and it has a predilection for sensory nerves (Martins *et al.*, 2009; Kawakami *et al.*, 2004).

Schwannoma may arise anywhere in the body, but they have an affinity for the head and neck region and extrimities. Approximately 25 – 48 % of schwannomas occur in the head and neck area- the intracranial region being the most common site, but schwannoma in the oral floor is extremely rare. Schwannoma does not recur and the malignant transformation is rare (Husain *et al.*, 2011). Here, we report a case of Schwannoma of sublingual origin wherein the nerve of origin was not identified.

## **CASES**

A 46 year old male presented with complaints of painless swelling in the right floor of mouth since 10 years. It was increasing in size gradually without causing discomfort in speech or deglutition with normal sensory and taste sensations.

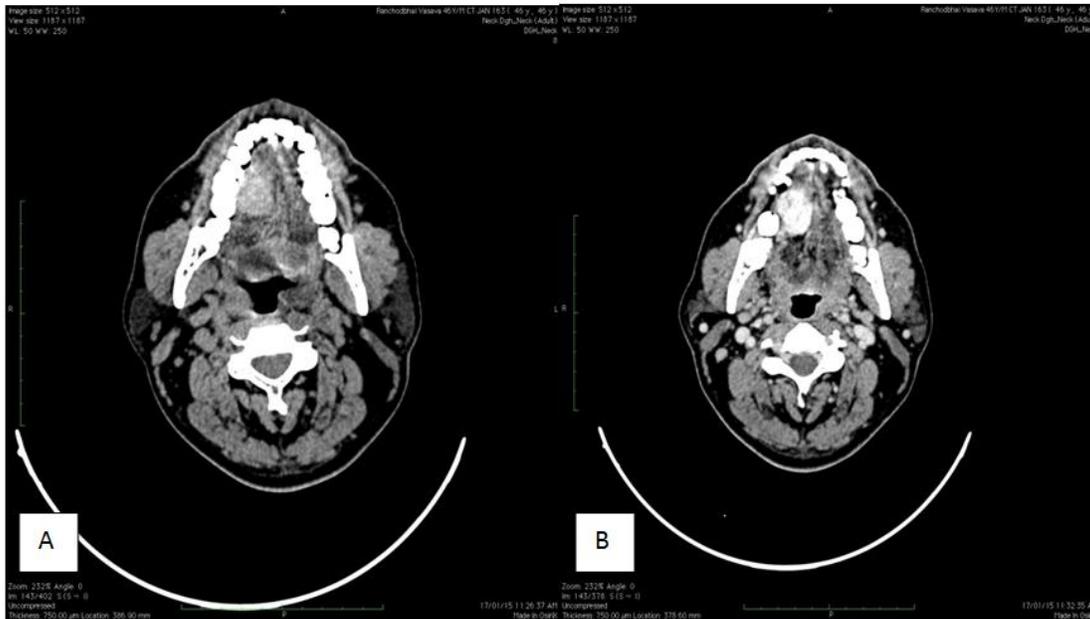
There was no history of trauma, local infection or systemic illness. An intraoral examination revealed well defined, large, circumscribed yellowish- white mass in the right anterior sublingual region measuring 3cm x 2cm in size, with a smooth surface. The swelling was firm in consistency, non- tender and mobile. He underwent routine investigations and Fine Needle Aspiration Cytology. The FNAC was inconclusive and differential diagnosis of benign masses such as neurofibroma, mesenchymal tumour and minor salivary gland tumor were considered. Contrast computerized tomography and excision biopsy was planned.

CT Scan revealed a very well defined, sharply margined, centrally hyperdense lobulated lesion measuring about 2.7 cm x 2.4 cm x 2.0 cm in the right side floor of mouth in the sublingual region (Figure- 1A). Post contrast CT Scan shows homogenous marked enhancement of the lesion (Figure- 1B). The tumor is compressing and displacing the tongue to left side of midline and posteriorly (Figure 2A and 2B). A complete surgical sublingual tumor excision and biopsy was done under general anaesthesia and the tumor enucleated with primary closure. Care was taken not to damage lingual nerve and Wharton's duct during surgical excision.

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Macroscopic appearance of specimen revealed a solitary, pinkish, well encapsulated mass measuring 3.0cm x 2.5 cm x 2.0 cm. External surfaces is smooth and glistening. On microscopic examination, study shows fascicles of spindle cells with serpentine nuclei having pointed ends (Figure- 3A). At places whorled cart- wheel appearance is seen; but majority areas show fascicles and bundles (Figure 3B). Obvious verocay bodies are not seen.

Based on Contrast CT Scan and Histopathological findings, the tumor was diagnosed as a Schwannoma of sublingual region.

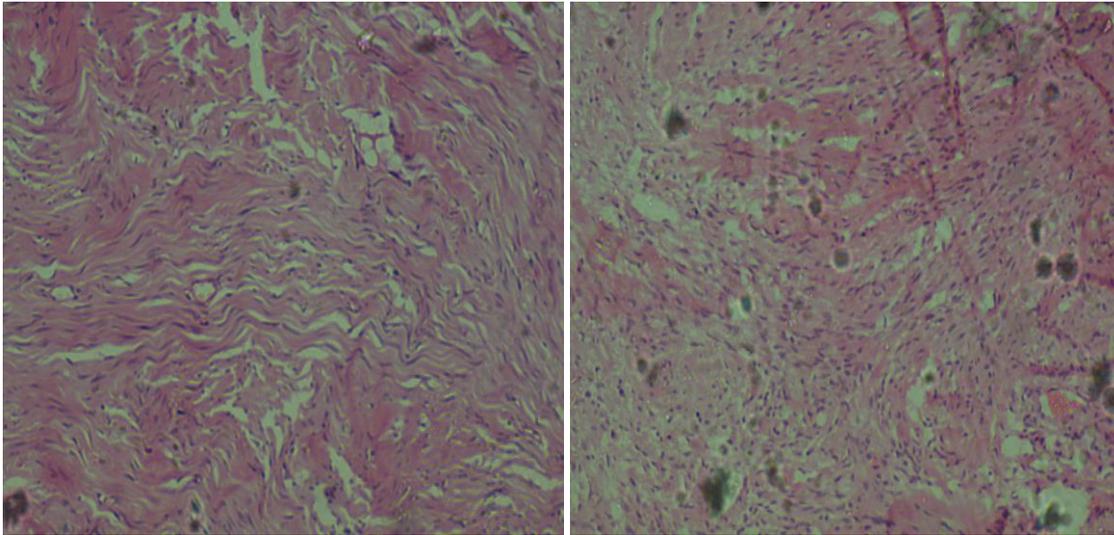


**Figure 1: A. Non-enhanced B. Enhanced axial CT cuts show a very well-defined sharply margined centrally hyperdense lobulated lesion in the right side floor of mouth, in the sublingual region**



**Figure 2: A. Coronal B. Saggital contrast enhanced CT cuts show homogenous marked enhancement of the lesion. The tumor is compressing and displacing tongue to left side of midline and posteriorly**

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**Figure 3: A. show fascicles of spindle cells with serpentine nuclei having pointed ends B. whorled cart-wheel appearance with alternating hypocellular and hypercellular areas**

### **DISCUSSION**

Embryologically, Schwann cells arise during 4<sup>th</sup> week of development from neuroectoderm (Ducic, 2003). Schwannoma is a rare, benign, neurogenic neoplasm composed of Schwann cells. (Lira *et al.*, 2013) It was first described by Verocay in 1910 and named it neurinoma.

These benign tumors occur regardless of age or sex but most reports suggest that the majority of tumors are present between the ages of 20 and 50 years and are equally distributed between the two sexes (Lira, 2013). It is usually solitary, encapsulated, painless, insidious and slow growing. So, they are of long duration at the time of presentation and rarely show a rapid course.

In the head and neck region, the tongue is the most common site, followed by the scalp, face, palate, floor of mouth, buccal mucosa, lips, jaws, pharynx, larynx, trachea, parotid gland, middle ear and external auditory meatus (Husain *et al.*, 2011). Other common sites include the flexor surface of upper and lower extremities and less often the mediastinum and peritoneum. Occasionally the tumor can arise centrally within bone and may produce the bone expansion (Lish *et al.*, 2006). Soft tissue or bone may be the site of origin of intraoral schwannoma. Those in soft tissue appear as a smooth submucosal swelling, thus resembling other lesions like mucocele, fibroepithelial polyp, fibroma, lipoma and benign salivary gland tumors (Ackerman *et al.*, 1951). So, many conditions come in the differential diagnosis of swelling of floor of mouth such as fibroma, lipoma, mucocele, epithelial hyperplasia, benign salivary gland tumors, hemangioma, granular cell tumor, neurofibroma, neuroma, nerve sheath myxoma, leiomyoma and rhabdomyoma. Although schwannoma in the oral floor is rarely observed, it should be taken into consideration while making a differential diagnosis (Baranovic *et al.*, 2006; Martins *et al.*, 2009).

The etiology is not known. They can arise from nerves covered with a schwann cell sheath, which include the all cranial nerves excluding the optic and olfactory nerve, and also involved the spinal nerves in the autonomic nervous system. More commonly it develops from the sensory nerves and rarely from the motor nerves (Husain *et al.*, 2011). If the nerve of origin is small, its association with a given tumor may be difficult to demonstrate. Whereas, if it originates from a larger nerve, it appears to be splayed out over the outer aspect of the capsule rather than incorporated within the tumor (Martins *et al.*, 2009; Lopez-Carriches *et al.*, 2009). Schwannoma has two clinical forms, the most frequent being the encapsulated one in which the tumor is surrounded by dense fibrous connective tissue; the other is pediculate, resembling a fibroma (Lopez-Carriches *et al.*, 2009).

Diagnostic investigations include an ultrasound scan, computed tomography (CT), magnetic resonance imaging (MRI) and fine needle aspiration cytology (FNAC). MRI is the best choice in detecting the

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extent of the tumor and correlates well with operative findings (Martins *et al.*, 2009; Lopez-Carriches *et al.*, 2009). MRI was not advised in the present case as patient already underwent CT scan.

In more than 50% of intra-oral lesions, it is not possible to differentiate between tumors arising from the lingual, hypoglossal and glossopharyngeal nerves (Mirza *et al.*, 2012) Also, there are reported cases of schwannoma arising from sublingual gland, (Okada *et al.*, 2012) mylohyoid nerve (Pattani *et al.*, 2010) and hypoglossal nerve (Fakhri *et al.*, 2009).

Ideally two histological patterns are identified, Antoni A and Antoni B. Antoni type A consists of schwann cells arranged in compact, twisted bundles, associated with delicate reticulin fibers and spindle-shaped nuclei aligned in parallel rows forming a typical palisading pattern. Between the rows there are fine cytoplasmic fibrils with acellular, eosinophilic masses called Verocay bodies. Antoni type B tissue is formed by irregularly arranged masses of elongated cells and fibres similar in appearance to neurofibroma, with areas of cystic degeneration and edema. Immune-staining analysis critical in the diagnosis of these neoplasms. Immune-histochemical tests can reveal a high affinity of the schwann cells to S-100 (Lira *et al.*, 2013; Lish *et al.*, 2006). The histopathological examination is the definitive diagnosis in the present case.

Surgical excision is the treatment of choice and relapse is uncommon in the well encapsulated variety. The encapsulated form is enucleated easily whereas the non- encapsulated requires normal tissue margins to avoid relapse. If the nerve of origin is visualized, an attempt should be made to separate carefully to preserve function, although this is sometimes not possible. The prognosis of schwannoma is quite favourable. Malignant transformation of benign schwannoma is controversial, with a few isolated cases documented (Baranovic *et al.*, 2006; Lira *et al.*, 2013; Lish *et al.*, 2006). Malignant transformation was not likely in our patient because examination of the excised mass revealed benign microscopic features and complete removal was confirmed.

### **Conclusion**

The schwannoma represents a lump not often encountered in clinical practice. The submucosal form of this lesion is, usually, indistinguishable from other benign neoplasm that also, usually, seen in the same region. Therefore, schwannoma should be included in the differential diagnosis of well circumscribed mucosal masses. Malignant transformation not seen in any of the intraoral schwannomas but the definite pre-operative diagnosis is necessary to avoid wide excision when the tumors can be easily enucleated without recurrence. The final diagnosis should be done after appropriate radiological investigations, histopathological examination and in some cases after immune-histochemical analysis.

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