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Case Report

# Cellular Schwannoma of Oral Cavity: A Case Report

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## Abstract

Neurilemmoma or Schwannoma are typically solitary, slow growing, encapsulated neoplasm that originate in a nerve and are composed of Schwann cells in a collagenous matrix. The frequency of multiplicity is apparently 2 - 6% in the body which has been given the term Schwannomatosis. Unlike classical schwannoma, cellular schwannoma discloses a markedly increase in cellularity, comprising fascicles of spindle cells which can occasionally be associated with herringbone or storiform pattern. We report here a case of intraoral multiple schwannoma of oral cavity showing histopathological findings of cellular schwannoma, in a 38 year old female patient.

**Keywords**: Neurilemmoma;Schwannoma;Nerve Sheath;Neoplasms;Neuroma;Neuroendocrine Tumors;Germ Cell and Embryonal.

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## Introduction

Schwannoma is a benign, encapsulated perineural tumor, that originates from the Schwann cells of the nerve sheath derived from the neuroectoderm.[1] It was first described by Verocay in 1910.[2,3] The tumor is solitary, with a smooth surface, slow asymptomatic growth; and the clinical symptomatology depends on the nerve of origin.[1,4] The reports of multiple intraoral schwannoma have been very few. This multiplicity can or cannot be associated with Von Recklinghausen's Syndrome.[5] We report here a case of multiple schwannoma which is not associated with any syndrome in a 38 year old female patient.

## Case Report

A 38 year old female patient presented with multiple swelling in the right quadrant of both jaws since six months. According to the patient, the lesion was always been asymptomatic, with no associated pain or paresthesia. The patient's medical history was noncontributory and has not undergone any surgery before. The clinical examination revealed multiple swellings; the largest being measured approximately 3 x 3 cm in dimension extending from distal of tooth #14 to tuberosity region (Figure 1a). On palpation it was firm in consistency and nontender. Swelling was also noted in the tooth #13, #21, #41, #42, #43 region involving gingiva on the labial side. A provisional diagnosis of pyogenic granuloma was given. Since the lesions were multiple, small and appeared to be benign, the surgeons chose to excise it for therapeutic purpose. The patient's postoperative course was uneventful and follow up for one year has shown no recurrence.

Microscopic examination revealed marked cellularity, comprising fascicles of spindle cells which in some areas are associated with herringbone pattern. Compact and hypercellular fascicles recapitulating Antoni-A areas are identified but not showing typical Verocay bodies (Figure 1b). In some areas it displayed short intersecting fascicles and whorls of Schwann cells (Figure 1c). Cells were also arranged in Antoni B type pattern with haphazard arrangement showing xanthomatous areas (Figure 1d). Areas of dense inflammatory cell infiltrate were also seen (Figure 1e). Because of the hypercellularity and lack of Verocay bodies we called it as "Cellular Variant of Schwannoma". Cellular schwannoma are known to be diffusely positive for S 100 and markedly positive for vimentin. Immunohistochemical staining with vimentin was performed which showed marked positivity (Figure 1f). On the basis of routine hematoxylin and eosin staining along with immunohistochemical evaluation, the above said diagnosis of Cellular Variant of Schwannoma was confirmed. Lack of necrosis, hyperchromatism and atypical features helped in differentiating from other spindle cell tumors like fibrosarcoma and malignant peripheral nerve sheath tumor. The patient healed uneventfully with no

recurrence further suggesting a benign nature of the tumor.

## Discussion

Schwannoma is an ectodermal benign nerve sheath tumor arising from Schwann cells.[6] Twenty five to 45% of Schwannomas occur in the head and neck[4] and 1% only demonstrate an intraoral origin[7]; in which case they occur, from greater to lesser frequency, in the mobile portion of the tongue, floor of the mouth, palate, gingiva, vestibule, lips, salivary glands and mental nerve region.[1,8] It is believed to originate from a proliferation of Schwann cells in the perineurium causing displacement and compression of the adjacent nerve.[1]

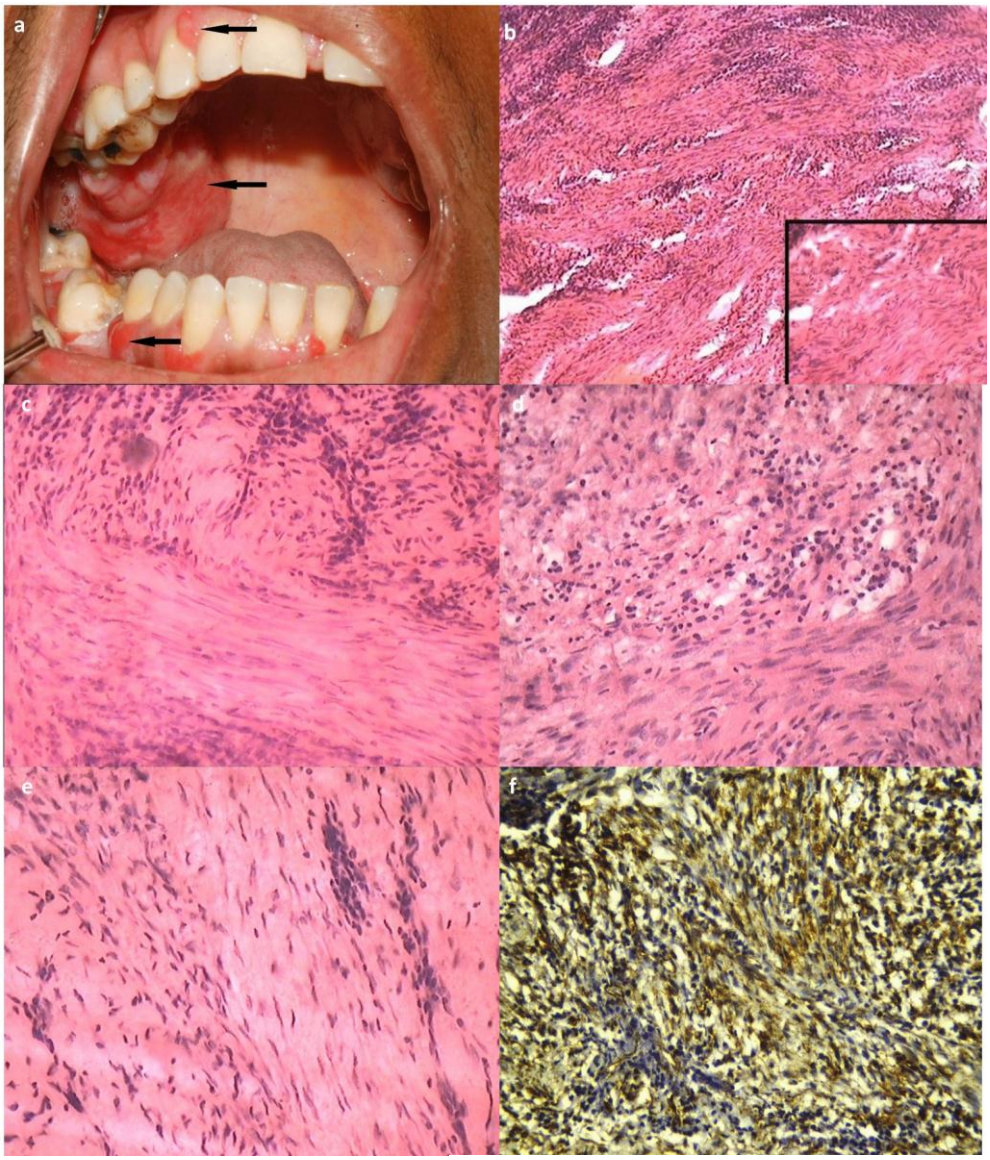


Figure 1: The intraoral photograph showing lesion on the right palate and labial side of maxillary and mandibular gingiva (a). The photomicrograph shows markedly increase in cellularity, comprising fascicles of spindle cells (b), short intersecting fascicles and whorls of Schwann cells (c), Antoni B type pattern with xanthomatous areas (d), inflammatory cell infiltrate (e) (H and E stained section 10X and 40X) and cells positive with vimentin (f).

Two types are distinguished: central or peripheral schwannoma, located in bone or in soft tissues respectively. Schwannoma can occur at any age, although when present in the oral cavity it tends to appear more often in adults than in children. There is a definite female predilection with female to male ratio 1.6:1. These are typically slow-growing, solitary tumors.[1] If multiple lesions are seen, it is called as “Schwannomatosis” which may either involve cranial, spinal or peripheral nerves[9] and is associated either with or without neurofibromatosis or Von Recklinghausen’s disease.[5]

Radiographically, intraosseous schwannoma of either jaw are well-demarcated, unilocular radiolucency's with a thin sclerotic border. Additional features such as external root resorption, cortical thinning, cortical expansion, and peripheral scalloping can be evident.[10] As diagnostic tools, ultrasonography, computed tomography and magnetic resonance imaging may be helpful for estimation of tumor margins as well as infiltration of surrounding structures[8]. But the present case was extra osseous variety.

The preoperative diagnosis was quite difficult in the present case, because of occurrence in less common areas like palate and gingiva. Multiple lesions of neurofibromas have been reported but multiple neurilemmoma of oral cavity is been rarely reported. Therefore histopathological examination provided a definitive diagnosis for the case.

There are many histopathological variants of schwannoma. Classically two histological patterns are defined, Antoni-A (with hypercellularity) and Antoni-B (with hypo cellularity). The ancient schwannoma includes various degenerative changes like cyst formation, haemorrhage and hyalinization along with nuclear atypia. Pseudo glandular schwannoma shows cystic spaces lined by schwann cells that assume a round or epitheloid structures resembling glands or dilated lymphatics. Plexiform schwannoma exhibits conventional or mixed appearance with increased cellularity and mitosis. Epitheloid schwannoma shows schwann cells which are round with sharp cytoplasmic border with nuclear inclusions resembling epitheloid cells but immunohistochemistry helps in the diagnosis. Melanotic schwannoma has polygonal to spindle shaped cells arranged in a syncytial pattern along with the melanin pigment that stains positively with Fontanna stain.[11] Unlike classical schwannoma, cellular schwannoma discloses a marked increase in cellularity, comprising fascicles of spindle cells which can occasionally be associated with herringbone or storiform pattern. Compact and hypercellular fascicles recapitulating Antoni-A areas can be identified in the form of long sweeping, short intersecting fascicles and whorls of Schwann cells. Although classical Verocay bodies are seldom identified, there may be occasional suggestion of palisades. Antoni-B areas are, however, not prominently featured. The spindle cells may exhibit mild nuclear atypia. Mitotic activity usually does not exceed four per ten high power fields.[11,12] The immunohistochemical tests reveal that the cells of cellular schwannoma show diffuse positivity for the protein S-100 and marked positivity for vimentin.[4,8] Therefore, all these features prompted us to call it as cellular variant of schwannoma.

The treatment of choice is excision. 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.[1,13] The prognosis is very good since it does not usually recur, and malignant transformation is rare. Since it usually develops in the extremities, malignant neurilemmoma is very rare in the oral cavity, although Hamakawa et al. described a case in the mandible with parotid and lung metastasis.[14] Kun et al. described six cases in the maxillofacial region, two of these had malignant transformation.[15] In our case the postsurgical healing was normal despite of the multiplicity of the lesion.

## Conclusion

Schwannoma should not be discarded when observing a tumor in the oral cavity. Any painless nodule in the head and neck region must include schwannoma. The definitive preoperative diagnosis could only be carried out with a biopsy and histopathological study. The treatment consists of surgical excision.

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