CASE REPORT

ANCIENT SCHWANNOMA-A RARE CASE

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ABSTRACT

Schwannoma is a relatively uncommon, slow-growing benign tumor that is derived apparently from the Schwann cells lining the nerve sheath. The schwannoma is usually a solitary lesion, and can be multiple when associated with neurofibromatosis. A case of 40-year-old women with a swelling in her mid-portion of the lower lip is presented. The diagnosis was established based on clinical, histopathological, and immunohistochemical aspects.

Keywords: Benign tumor, neural tumors, oral lesions, Schwann cells, schwannoma, soft tissue tumors.

INTRODUCTION

Schwannoma (Neurilemmoma) is a benign tumor of nerve sheath origin. These tumors can arise from any nerve sheath origin. These tumors can arise from any nerve covered with a Schwann cell sheath, which include the cranial nerve (except for optic, olfactory), spinal and autonomic nervous system (1).

Intra oral schwannoma can arise both in soft tissue or bone. Those in soft tissue appear as a smooth submucosal swelling, thus resembling other lesions like mucocele, fibro epithelial polyp, fibroma, lipoma and benign salivary gland tumors. However, the histological differential diagnosis is made with other neural origin lesions, which could be neurofibroma and neuroma or muscular or fibroblastic origin tumor. This is a report of a case of benign schwannoma in mid portion of the lower lip extending from vermillion border on to the skin, whose diagnosis was established upon clinical, histological and immunohistochemical findings and a review of literature is presented (2).

CASE REPORT

A 40 year old female patient reported to the department of oral medicine and radiology, AECS Maaruthi College of dental sciences, Bangalore with a chief complaint of a painless mass in the lower lip since 10 years. On eliciting presenting illness patient developed a small swelling on her lower lip which was peanut

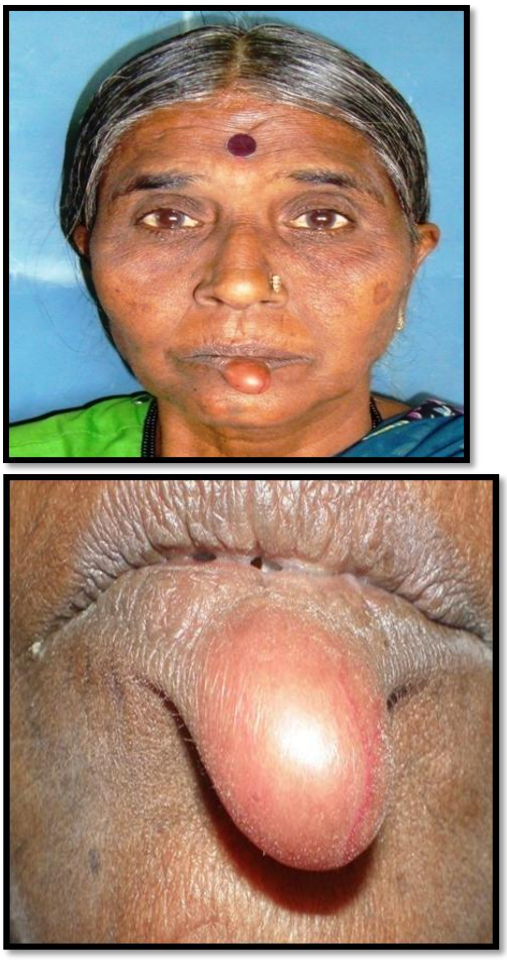

Figure 1: The first, extraoral photograph showing lower lip swelling; and the second, lower lip swelling with skin over swelling stretched and shiny with enlarged capillaries

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size 10 years back, it gradually started increasing in size since 3 years to present size. The patient's medical and family history was noncontributory.

Extra oral examination revealed a solitary spherical swelling in mid portion of the lower lip extending from vermillion border onto the skin. Approximately measuring about 2cmX1.5cm and skin over the swelling was stretched and shiny with engorged capillaries (fig 1).

On palpation there was no local rise of temperature, firm inconsistency, freely movable, nontender and not fixed to the underlying skin. On clinical basis we arrived at provisional diagnosis of benign soft tissue tumor.

All the laboratory investigations were under normal. As the size of tumor was small excision biopsy was planned under general anesthesia and complete excision of the mass was done. Grossly the specimen revealed a well encapsulated mass measuring 2cmX1.5cm with soft grayish surface (fig 2a and b). The specimen was subjected for histopathological examination. Patient was put on antibiotics and analgesics for a period of 5days. The post-operative period was uneventful.

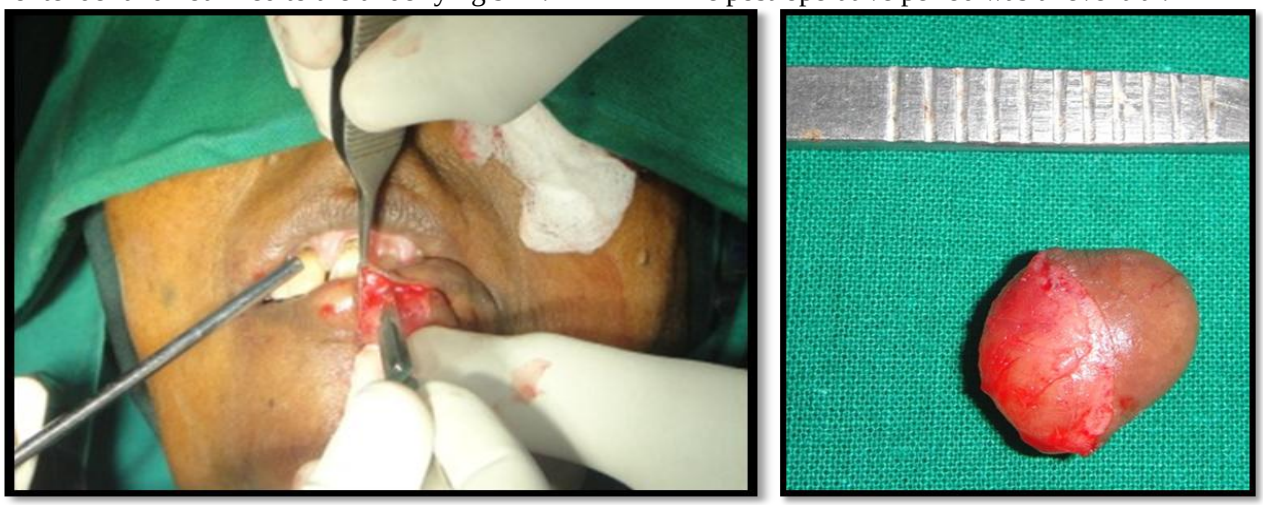

Figure 2: The first one, eexcision of the swelling under local anesthesia; and the second one, excised specimen grossly measuring about 3cmX 2cm.

Histopathological findings: Histopathological report revealed an encapsulated connective lesion in few areas suggestive of Antoni A cells. Randomly arranged spindle cells suggestive of Antoni B cells are also observed. Abundant Myxoid stroma, nuclear hyperchromatism & with fascicles of spindle shaped cells with wavy nuclei arranged in palisaded appearance pleomorphism are seen suggestive of degenerative changes.

Immunohistochemical staining was positive for S-100 fig 5 & negative for SMA ruling out tumor of muscle origin (fig 3a and b).

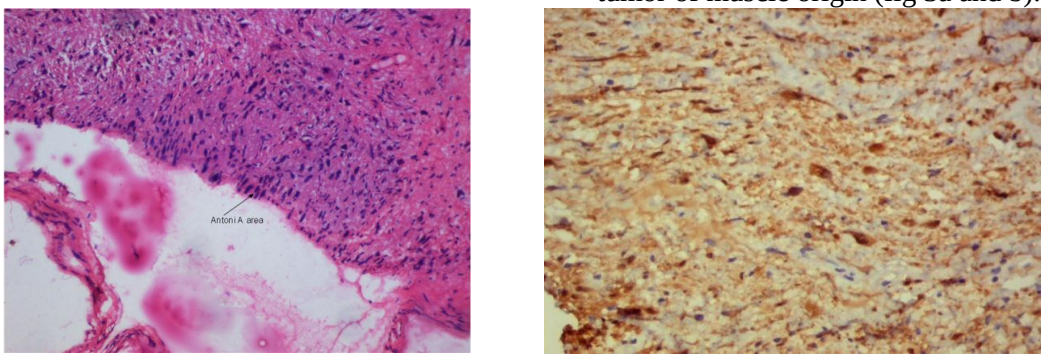

Figure 3: H and E staining of the specimen showing tumor cells composed of proliferating groups of Schwann cell nuclei in Antoni A tissue forming Verocay bodies; and the second; Immunohistochemical staining was positive for S-100 & negative for SMA ruling out tumor of muscle origin.

All these histological features and Immunohistochemical results were suggestive of Ancient Schwannoma.

DISCUSSION

The schwannoma, also known as Neurilemmoma, Neurinoma or perineural fibroblastoma. It is a rare benign neural tumor, arising from the neural sheath Schwann cells of the peripheral, cranial, or autonomic nerves[3]. The etiology is unknown. Oral schwannoma is a rare solitary, slow growing, generally asymptomatic neural tumor that can present itself at any age. However it is more common between the second and third decades of life. William *et al* findings showed that in 83% of the cases studied by them the schwannomas presented in males, while for Lucas there was a greater predilection for females, and for Hatziotis and Asprides; Enzinger and Weiss there was an equal distribution between both sexes. In most of the cases, between 25% -45%, extra cranial schwannoma occurs in the head and neck region. In the oral cavity the lesion in usually presented in soft tissue, more commonly the tongue, followed by the palate, buccal mucosa, and may have clinical aspects similar to other benign lesions like mucocele**,** fibroma, lipoma, and benign salivary gland tumors. In some cases the tumor could be intraosseous, being more frequent in the mandible where it may cause bone expansion, pain, and paresthesia. In these cases differential clinical diagnosis of cysts and odontogenic tumors are commonly formulated. Gallo *et al*. reported on 157 cases, where 45.2% of the cases involved the tongue and 13.3% involved the cheek. Gupta *et al.* on 136 cases of schwannoma in the head and neck that consisted of 60 cases in the neck, ten cases in the parotid gland, nine cases in the cheek, eight cases in the tongue, and eight cases in the pharynx. Kun *et al*. reported in their study that 18 out of 49 cases were in the neck and 11 in the tongue.

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

The clinical differential diagnosis is fibroma, lipoma, neurofibroma, or salivary glands tumor. However, the histological differential diagnosis is made with other neural origin lesions, which could be neurofibroma and neuroma.

Table 1: Review of the literature

Authors	Year	Age yrs	Sex	Location
Eversole & Howell[4]	1971	58	F	Floor of the mouth and ventral tongue
Marks et al[5]	1976	65	F	Floor of the mouth(right)
McCoy et al[6]	1983	36	F	Maxillary vestibule (left)
Dayan et al[3]	1989	52	F	Maxillary vestibule (left)
Nakayama et al[7]	1996	40	F	Floor of mouth and ventral tongue
Ledesma et al[8]	1999	21	F	Floor of mouth and ventral tongue
Chen et al[9]	2006	34	M	Floor of the mouth (left)
Krishna raj, Subhash raj et al[10]	2008	18	M	posterior vestibule (left)
Present case	2009	40	F	Lower lip

Ancient schwannoma: Ackerman and Taylor in 1951 found that the schwannoma presented with clear areas of hypocellular tissues and attributed the changes to the long standing degenerative changes. They coined the term "ancient" schwannoma for such type of benign neurogenic tumor.

These degenerative features are attributed to the growth and "aging" of the tumor, hence the term "Ancient schwannoma". Despite these degenerative changes, ancient schwannomas

behave similar to schwannoma. They are benign, slow-growing tumors with rare malignant transformation.

Reviewing the literature, only eight cases of ancient schwannoma had been reported[3-10] (Table 1).

Among the published cases of ancient schwannoma 6 cases were seen in females. The mean age being 44 years. The common coincidences for this tumor among the cases seen in the literature were the location at floor of the mouth, mean age of 44 years and female sex.

Recurrent and malignant transformation were not reported in any of the reported intraoral ancient schwannoma. The histological picture is dominated by an encapsulated lesion arising from a nerve end composed of an intimate mixture of spindle cells forming highly cellular Antoni A and less cellular, myoxid Antoni B areas. Vero cay bodies, which are cellular eosinophilic zones, are frequently seen.

In summary, this could be the second reported case of Ancient Schwannoma arising from the mental nerve found in a 40 years old female patient.

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