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CASE REPORT

NEURILEMMOMA OF UPPER LIP: A RARE CASE REPORT

Divya Pandya[1], Anil Kumar Nagarajappa[2], Sonalika Ghate[3], Sanyog Pathak[4]

[1]PG student, Department of Oral Medicine and Radiology, Hitkarini Dental College and Hospital, Jabalpur, Madhya Pradesh, India.

[2]Professor and Head, Department of Oral Medicine and Radiology, Hitkarini Dental College and Hospital, Jabalpur, Madhya Pradesh, India.

[3]Reader, Department of Oral Pathology, Hitkarini Dental College and Hospital, Jabalpur, Madhya Pradesh, India.

[4]Reader, Department of Oral and maxillofacial Surgery, Hitkarini Dental College and Hospital, Jabalpur, Madhya Pradesh, India.

Corresponding Author: Dr. Divya Pandya, F-2 Atul Vihar II, SBI Colony Near, Hathital Railway Crossing, Gorakhpur, Jabalpur (M.P), 482001

ABSTRACT:
Schwannomas are a rare type of benign slow growing nerve sheath tumors predominant in soft tissues of head and neck. Less than 1% schwannomas are intraosseous with affliction of mandible over maxilla. This submucosal lesion must be differentiated from other benign lesions that also appear in same regions. This article documents a rare case of peripheral soft tissue schwannoma of upper lip in a 30 year old male patient presenting as asymptomatic swelling in mucosal aspect of upper lip with no radiographic finding and a confirmatory diagnosis by histopathological examination. It also highlights the various imaging modalities and differential diagnosis that can be considered.
Key words: Antoni cells, Cranial nerves, Nerve sheath, Neurilemmoma, Schwann cells

INTRODUCTION

Neurilemmomas are uncommon, solitary, slowly growing benign neoplasm derived from the sheath cells that cover myelinated nerve fibres.[1,2,3] They are well encapsulated soft tissue or intrabony lesions deriving from neural crest cells.[2] Although the etiology is unknown, it is believed that the lesion arises from proliferation of Schwann cells at a point inside the perineurium, which causes displacement and compression of surrounding normal nerve.[4] Its origin is most commonly associated with a nerve trunk and most of the time it affects the whole nerve throughout its course in the peripheral nervous system.[5] There is a large variety of terms for neurilemmoma but only three are still in current use: neurinoma, schwannoma and neurilemmoma.[3] Schwannomas can arise from any cranial (except for optic and olfactory), peripheral or autonomic nerves that contain Schwann cells, the sheath that cover myelinated nerve fibres. Schwannoma was first reported by Jose Verocay in 1910 and called this benign neurogenic tumor as neurinoma.[2] In 1935, the term “neurilemmoma” was coined by Arthur Purdy Stout.[6,7] This tumor is often associated with nerve sheath and adjacent to

parentral nerve extrinsic to the nerve fascicles. Approximately 25% to 45% of schwannomas are seen in head and neck region which include scalp, face, pharynx, parotid gland, middle ear and external auditory canal[5] and are found rarely in oral cavity (only 1%). Most of the intraoral schwannomas are located in tongue with the tip being least affected part.[2,4,6] Other less frequent locations are buccal mucosa, palate, floor of the mouth, gingiva and lips.[2] Other common sites include the flexor surface of upper and lower extremities[3] including sacrum, verterbra, clavicle, ribs, humerus, radius, ulna etc[6] and less often mediastinum and peritoneum.[3]

Neurilemmoma are said to occur more frequently in the 25-55 year age group, but can occur at any age, with 1.6:1 female to male predilection, however there is no definite gender predilection.[2] The most common site of occurrence of central neurilemmoma is mandible where the posterior segments of body and ramus are most frequent sites of occurrence because of the protracted intraosseous path of inferior alveolar nerve[8,9] with intraosseous schwannoma of mandibular symphysis is exceptionally rare[8] however maxillary schwannomas have equal predilection for anterior and posterior segments of jaw.[7] In the current medical literature, there are 44 acceptable cases of intraosseous neurilemmomas of the jaw, 39 of the mandible and five in the maxilla, representing less than 1% of primary tumors of the bones.[6]

Most neurilemmomas are asymptomatic and reveal no radiographic findings that characteristically differentiate them from odontogenic cysts and tumors.[1]The clinical symptoms depends on nerve of origin.[5] We present here a rare case of neurilemmoma of upper lip a relatively rare site of occurrence and unusual presentation.

CASE REPORT

A 30 year old male presented to our Department of Oral Medicine and Radiology with a painless swelling in upper lip since 2 months. Swelling occurred gradually and initially it was small and slowly progressed to present size. Patient did not reveal any history of pain, paresthesia, any discharge or bleeding from the swelling. There was no history of trauma. Patient's medical and family histories were noncontributory.

Thorough physical examination revealed no abnormality. All vital signs were within normal physiological limits. Extraoral examination did not reveal any gross facial asymmetry due to swelling. On intraoral examination, an ill defined, solitary, non-ulcerated smooth surfaced swelling of approximately 3x4 cms was observed in relation to maxillary central incisors and right lateral incisor, roughly oval in shape involving labial vestibular depth. Swelling was sessile and originated from mucosal surface of upper lip and involved labial frenum (Figure 1). Swelling did not reveal bleeding or cortical plate perforation and was covered with normal mucosa with indentation of opposing arch teeth on inferior aspect of swelling. The permanent maxillary incisors were malpositioned, which was hindering occlusion due to anterior teeth cross bite. There was no evidence of any carious lesions with associated teeth, however all teeth were periodontally compromised with generalized class II gingival recession and inflamed marginal and papillary gingiva (Figure 1). On palpation, this protruding mass was non tender, firm in consistency with an intact mucosa which revealed no blanching on stretching the upper lip. There was no evidence of lymphadenopathy.

Maxillary anterior teeth were normal and responded positively to electric and thermal pulp vitality tests. This eliminated the possibility of periapical cyst or an odontogenic cyst to be the cause of swelling. Aspiration biopsy revealed no evidence of any cystic fluid or blood thus excluding cysts and hemangioma. Intraoral periapical radiograph of maxillary central

incisor region was made which revealed no pathology indicating no involvement of periapical or maxillary alveolar bone (Figure 2). All hematological and laboratory findings were normal. A provisional diagnosis of Mucous retention cyst was made owing to continuous trauma from opposing arch teeth on upper lip. A list of differential diagnosis was framed which included minor salivary gland tumor, traumatic fibroma, lymphoma, lipoma, neurofibroma and neurilemmoma. Incisional biopsy of tumor was performed and specimen was sent for histopathological examination.

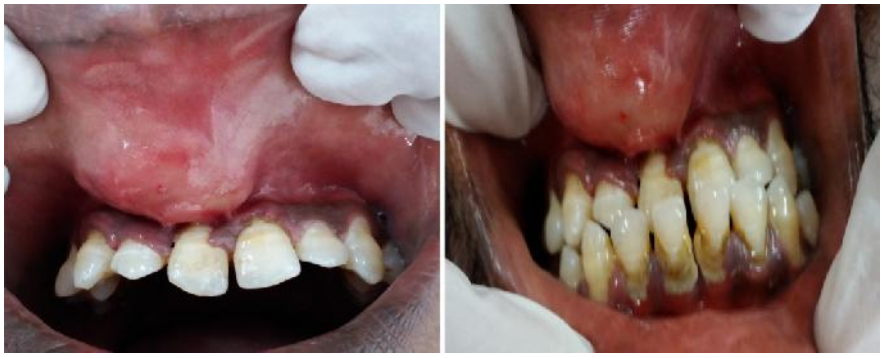

Figure 1: Intraoral clinical picture of solitary ill defined swelling on mucosal aspect of upper lip in relation to maxillary central incisors and right lateral incisor with indentations of opposing arch teeth on lower aspect of swelling due to anterior teeth crossbite

Figure 2: Intraoral peripaical radiograph of maxillary incisor region revealing no pathology

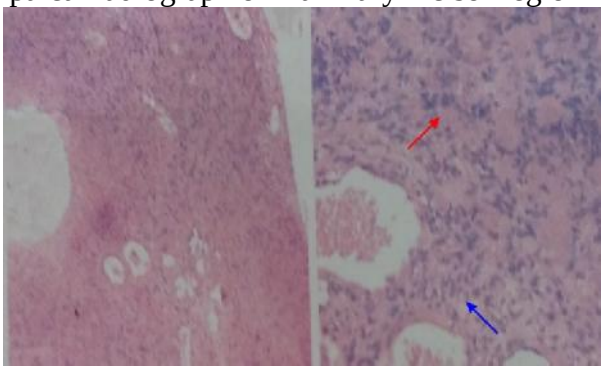

Figure 3: H & E stained histological section showing Antoni type A (marked by red arrow) and B (marked by blue arrow) cells with Verocay bodies

Histopathological report revealed hematoxylin and eosin stained section of tumor mass consisting of spindle cells arranged in two patterns. Antoni A and Antoni B. Antoni A represented area where spindle cells were arranged in palisaded pattern with parallel intercellular fibres. Homogeneous eosinophilic areas were seen in these areas, termed as Verocay bodies. Whereas Antoni B type cells showed haphazard arrangement of cells (Figure 3). Impression in hisotopathology report was schwannoma (neurilemmoma)

Hence, on the basis of history, clinical examination and investigations including biopsy a final diagnosis of Peripheral neurilemmoma of upper lip was made.

DISCUSSION

Schannoma is a solitary, slow growing, usually encapsulated, generally asymptomatic neural tumor. It can present at any age, however it is more common between second and third decade of life as with our case. The tumor is derived from the Schwann cell sheath, which enlarges, expands and causes displacement and compression of nerve of origin.[2] The present case occurred in male patient though literature showed female predilection. Neurilemmoma may occur in soft tissues (peripheral) commonly involving tongue, followed by palate, buccal mucosa and lip and central within jaw bones.[2] In the head and neck region, they arise medially from the last four cranial nerves or the sympathetic chain and laterally from cervical or brachial plexus. Schwannomas have a predilection for the sensory nerves, especially for eighth cranial nerve, but a motor nerve like facial nerve can also be affected.[3] Schannomas of hypoglossal nerve are very rare and seldom found in sublingual space.[3,10] Although schwannomas originate from nerve tissue, a direct relation with a nerve can be demonstrated only in 50% of cases.

Etiology remains unknown and tumor is generally asymptomatic and can remain quiescent for a long period. The usual presenting symptom is a gradually enlarging mass with pain and neurological symptoms are uncommon, unless tumor becomes large to invade submucosal areas leading to pain and discomfort. If it involves tongue, it may cause dyspnea or dysphagia.[3] Our patient was asymptomatic and presented for growing mass on mucosal aspect of upper lip which was increasing in size without any other symptoms like pain, paresthesia or hypersalivation. The tumor presents with smooth, glistening, pink, grey white or yellowish surface as seen with our case. Cystic areas and foci of hemorrhage and calcification can also be found with variable size ranging from few millimeters to several centimeters usually less than 5 cm in size. There are variants of benign schwanomas such as plexiform or multinodular, cellular, Wagner-Meissner, granular cell schwannomas,[3] but in our present case a conventional type schwannoma with mixed pattern of Antoni A and Antoni B areas and well formed verocay bodies is shown.

Clinical differential diagnosis includes, benign neural tumors like fibroma, lipoma, neurofibroma, minor salivary gland tumor, large retention mucocele, lipoma, pleomorphic adenoma on account of their slow growth and absence of neural symptoms,[2,5] leiomyoma, rhabdomyoma, neurosarcoma, ganglion cyst, giant cell tumor of tendon sheath.[3] Histopathology and imaging modalities are helpful in confirming diagnosis. Ultrasonography, computed tomography and magnetic resonance imaging (MRI) are modalities in diagnosing and treatment for estimation of tumor margins and determination of infilteration to surrounding structures. MRI is particularly helpful in showing the internal characteristic of encapsulated mass. Although soft tissue schwannomas have no useful radiographic findings as with our case but intrabony tumors shows varying radiographic presentations like unilocular/multilocular radiolucency with thin sclerotic border and its extension can be

determined by plain film radiography to differentiate it from other odontogenic and non-odontogenic cysts and tumors. However, use of advanced imaging modalities is not recommended for routine use.[2,7,8,11]

Microscopically, schwannomas can present two patterns: Antoni type A and Antoni type B areas. Antoni type A areas consists of spindle-shaped Schwann cells fascicles that often form a palisade arrangement around central acellular, eosinophilic areas known as Verocay bodies with parallel formed thin reticulin fibres, fusiform shaped cells and curled nuclei. Antoni type B region is less cellular and less organized. The spindle cells are randomly arranged within a myxomatous stroma.[2,3,8,9] In our case, the inspection of morphological stained slides revealed all the aspects needed to diagnose a conventional type schwannoma. Immunohistochemmically, schwannomas show an intense and relatively uniform staining for S-100 protein, a neural-crest marker antigen, present in supporting cells of nervous system.[3,9]

Local excision of tumor is the treatment of choice. The non encapsulated form requires a normal tissue margin and careful separation to preserve normal function.[3] Radiation therapy is not indicated because schwannomas exhibit high degree of radio resistance.[4] Recurrence is rare with complete resection. Malignant transformation of benign schwannoma is rare being mentioned in 8-10% of cases. Prognosis is usually good.[2,3,4]

The clinical course and the radiographic appearance of neurilemmoma is not characteristic, therefore clinical diagnosis is not possible and can only be established on histologic examination.[1]

CONCLUSION

We presented a rare case of schwannoma of oral cavity which presents as slow growing painless benign swelling in oral cavity rarely encountered in clinical practice and requires a systematic work up for an accurate diagnosis and classification, which must integrate histological analysis and with clinical data. This submucosal lesion must be differentiated from other benign lesions that also appear in same regions. The final diagnosis can only be given after histopathological examination. Prognosis is good with unknown recurrence.

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