INTRAORAL SCWANNOMA- A CASE REPORT

Abstract

Benign nerve cell tumors include schwannoma, which rarely has an intraoral presentation. Schwannoma is a relatively uncommon, slow-growing benign tumor that is derived apparently from the schwann cells. It’s an asymptomatic tumor which may sometimes present with pain. The tongue is the most common site, followed by palate, floor of mouth, buccal mucosa, lips, and jaws. Usually it is a solitary lesion but it can present as multiple lesions when associated with neurofibromatosis. Schwannoma resembles with many other benign tumours like salivary gland tumors, lipoma, fibroma, etc. Sofinal diagnosis is established after complete histologic and immunohistochemical analysis. Presence of Anti s-100 antibody is confirmative of schwannoma. Here, we report a rare case of intraoral schwannoma in right submental region of mandible, in a 16-year old male patient with emphasis on it’s differentiation from neurofibroma.

Keywords: Neurilemmoma, schwannoma, schwann cells

INTRODUCTION

Schwannoma also known as neurilemmoma is a rare, benign neural tumour, arising from neural sheath and schwanncells of the peripheral, cranial, or autonomic nerves. The etiology is unknown, but it’s postulated that the lesion arises by proliferation of schwann cells at one point inside the perineurium causing displacement and compression of surrounding normal nerve tissue. This tumor has predilection for the head and neck region where one-third of the cases are reported, however intraoral lesions being rare. When it is found in oral structures, the tongue is reported to be the favoured site. It usually occurs as an asymptomatic, solitary, smooth-surfaced growth, emerging at any age, with as such, no gender predilection.

It is known that schwannoma is a slowly growing lesion and is usually of long duration at the time of presentation by the patient, however occasionally it may exhibit a relatively rapid course. Although these tumors are neurogenic in origin, but still they are painless, exhibiting pain only in case if they are causing pressure on adjacent nerve rather than on nerve of origin. Schwannomas can be divided into central/intraosseous and peripheral lesions, also in addition to these a variant known
as ancient schwannoma has been diagnosed which presents with degenerative phenomena such as cystic cavities, hemorrhage, hyalinization or calcifications. A variant characterized by a nodular growth pattern (plexiform intraosseous schwannoma) is also present.

Radiological examinations such as a computed tomography (CT) scan with contrast and magnetic resonance imaging (MRI) may be performed to show the extension of the tumor. 

This tumour typically shows closely packed spindle cells, often with palisaded nuclei and verocay bodies (Antoni A areas) and less cellular areas with a loose reticular pattern and microcystic degeneration sometimes containing numerous xanthoma cells (Antoni B). The degree of cellularity of the neoplasm can be high or low. The spindle cells frequently are moderately pleomorphic, but mitotic figures are rare. The presence of pleomorphism does not necessarily denote a malignant tendency, but in rare cases undoubted malignant changes can appear associated with an increased growth rate. Thrombosis and necrosis may be present focally.

The clinical differential diagnosis could be with any other benign tumoral lesions such as 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, or muscular or fibroblastic origin tumor.

Treatment of schwannomas includes complete surgical excision of the lesion with no reported cases of recurrence. The prognosis is good and malignant transformation is very rare although a few isolated cases have been reported.

The purpose of the following article is to discuss a case of benign schwannoma present on the inferior border of mandible in right submental region. The diagnosis was confirmed on clinical and histological examination.

**CASE REPORT**

A 16 year old male patient presented with a growth in the right side of mandible that had been present for 6 months. Although the patient was well aware of the swelling but he reported after sometime as the swelling was painless and asymptomatic. On intraoral examination a well defined firm and non tender soft tissue swelling measuring 1.5 cm x 1cm was seen involving the inferior border of mandible in right submental region. The swelling was covered by normal appearing mucosa.

The swelling was ovoid in shape, reflecting a reddish white colour, was firm on palpation and not fixed to the underlying and surrounding tissues. From the above obtained data, a provisional diagnosis of fibroma was established. Radiographic examination was non relevant. To establish a confirmative diagnosis excisional biopsy was done under local anaesthesia.

Microscopic examination revealed a connective tissue stroma covered by stratified squamous keratinized epithelium. There were seen streaming fascicles of spindle shaped cells which at places formed a palisaded arrangement around acellular, eosinophilic areas (verocay bodies) indicative of Antoni type A pattern (Figure 1). The spindle cells in other areas were randomly arranged within a loose myxomatous stroma, indicative of Antoni type B pattern (Figure 2). In other areas small hyaline structures were present.

Based on the clinical and histological evidence obtained, a confirmatory diagnosis of schwannoma was given. As of now, the healing following biopsy has been uneventful with no evidence of recurrence.
DISCUSSION

Tumors arising from peripheral nerves in the oral and paroral tissues are uncommon. Oral peripheral nerve sheath tumors are rare and include neurofibroma, schwannoma, palisaded encapsulated neuroma (PEN), nerve sheath myxoma, mucosal neuroma associated with multiple endocrine neoplasia type 2B, traumatic neuroma and granular cell tumor. \(^\text{11}\) Schwannoma was first established as a pathological entity by Verocay in 1908 who later called it neurinoma in 1910. Later the term neurilemmoma was coined by Stout in 1935. \(^\text{12}\)

About 25-45% of all the extracranial schwannomas have been reported in the head and neck region. The most common site of the extracranial schwannomas in the head and neck region is the parapharyngeal space. \(^\text{12}\) The first case of neurilemmoma within the parapharyngeal space was reported in 1933 by Figi. Other sites in the head and neck like submandibular space, para-nasal sinuses, cheek, oral cavity etc. are rare. \(^\text{13}\)

In a review of 303 solitary neural tumors, 136 lesions (45%) were located in the head and neck region and among these, only 30 patients presented lesions in oral cavity. Gallo et al. \(^\text{14}\) reported on 157 cases of schwannoma, where 45.2% of the cases involved the tongue and 13.3% involved the cheek. Kun et al. \(^\text{15}\) reported in their study that 18 out of 49 cases were in the neck and 11 in the tongue. In a review made by Leu and Chang, \(^\text{16}\) out of 52 cases of schwannomas which originated in the head and neck region, seven cases showed lesions which were located in the oral cavity, which included one in the hard palate, one in the soft palate, two in submasseteric area, one in the tongue, and one in lower lip. \(^\text{4}\)

Schwannoma presents as an asymptomatic and slow growing lesion. Although schwannoma can be found to occur in any age; but it is more common between the second and third decades of life. William et al. \(^\text{17}\) revealed that in 83% of the cases studied by them, predilection for male patients was seen; while findings by Lucas \(^\text{18}\) presented with a greater predilection for females. On the other hand, Hatziotis and Asprides \(^\text{19}\); Enzinger and Weiss agreed over an equal prevalence of schwannoma in both sexes. Our case also presented with similar demographic and clinical findings in being asymptomatic, slow growing and occurring in a male patient who is 16 years of age.

Macroscopic features reveal a well-delineated but non-encapsulated globular, firm to rubbery yellow-tan mass. The cut surfaces show tan-grey, yellowish, solid to myxoid and cystic tissue, commonly with haemorrhagic areas.

Since schwannoma and neurofibroma clinically resemble each other, so to overcome the dilemma, histologic as well as immunohistochemical findings of both need to be understood. Microscopically, both schwannoma and neurofibroma contain elongated cells with irregular nuclei lying between bundles of collagen fibers; however schwannoma is derived from schwann cells and neurofibroma on other hand is derived from fibroblasts of the perineurium. Schwannoma shows degenerative changes such as cystic alterations and haemorrhagic necrosis whereas such changes are not seen in neurofibroma. In schwannomas, complete perineural encapsulation is seen with a total lack of axons unlike neurofibroma where there is lack of encapsulation and presence of fewer axons with myelin sheath. Also, true schwannoma rarely occurs in dermis. \(^\text{11}\) In the present case, histological features were of a classical schwannoma revealing Antoni A & Antoni B pattern.

Immunohistochemical staining of tumoral cells in neurilemmoma within the parapharyngeal space was performed in 1933 by Figi. Other sites in the head and neck like submandibular space, para-nasal sinuses, cheek, oral cavity etc. are rare. Due to rarity, about 25-45% of these tumors were reported in the head and neck region. \(^\text{11}\)

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Immunohistochemical staining of tumoral cells in schwannoma shows greater positivity for s-100 protein when compared with neurofibroma. Since positivity for s-100 protein is seen in both so to further delineate the two additional immunohistochemical stains are done.

Cd34 is a useful stain for differentiating between the two, since neurofibromas typically demonstrate a significant subpopulation of CD34-positive stromal cells, unlike most schwannomas wherein only slight positivity for CD34 cells can be found and that too in antoni B areas. Other markers that may be of some utility include Factor XIIIa (reportedly positive in neurofibroma but negative in schwannoma), and CD56 (reportedly negative in neurofibroma and positive in schwannoma. Schwannoma shows high positivity for Calretinin (a calcium-binding protein) when compared to neurofibroma. The pericapsular region of a schwannoma may contain EMA (epithelial membrane antigen) positivity. A very small minority of schwannomas may show rather extensive immunoreactivity with cytokeratin antibodies, which is thought to represent cross-reactivity with glial fibrillary acidic protein (GFAP) rather than true expression of cytokeratin proteins.

The treatment is complete surgical excision of the benign tumour. Recurrence after successful en bloc removal of the tumour is very rare.

CONCLUSION

Being a very rare neurogenic tumor, the diagnosis of schwannoma is usually based on exclusion criteria from other similar lesions. Definitive diagnosis of schwannoma is established only after complete histopathologic examination and mostly immunohistochemical analysis. Only definitive treatment is complete surgical excision of schwannoma, and while performing such procedure the potential risk of nerve damage should always be kept in mind. Recurrence after surgical removal is a rare phenomenon.
REFERENCES