Schwannoma of the hard palate: A case report and review of literature

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Abstract:
Schwannoma is a benign tumour that originates from the Schwann cells of the peripheral nerves. It presents as slow growing painless swelling in the oral cavity or head and neck region not often encountered in clinical practice. This sub mucosal lesion must be differentiated from other benign lesions that also appear in the same regions. Approximately 25 – 45% Schwannomas are seen head and neck region and found rarely in oral cavity, about 1%. Most of the intraoral schwannomas appear in the tongue. Other less frequent locations include buccal mucosa, palate, and base of the mouth, gingiva and lips. In this article, we report a case of schwannoma of the hard palate, which was excised intraorally.

Keywords: Schwannoma, neurilemmoma, neurinoma, hard palate.

Introduction:
Schwannomas are usually a solitary, slow growing benign neoplasm and well encapsulated soft tissue or intrabony lesions deriving from neural crest cells.1-3 Schwannomas can arise from any cranial, peripheral, or autonomic nerves that contain Schwann cells, the sheath cells that cover myelinated nerve fibers.4-6 Schwannoma was first reported by Verocay in 1910, called this benign neurogenic tumor as neurinoma.5 This tumour is often associated with the nerve sheath and adjacent to the parental nerve but extrinsic to the nerve fascicles. Approximately 25% to 45% of the schwannomas are seen in the head and neck region and are found rarely in the oral cavity (only 1%).6-9 Schwannomas of the head and neck region, including the oral and maxillofacial region are rare. Most of the intraoral schwannomas are located in the tongue with the tip being the least affected part.7,8 Other less frequent locations are the buccal mucosa, palate, base of the mouth, gingiva and lips.7-9 Schwannomas are said to occur more frequently in the 25-55 year age group with 1.6-1 female to male predilection, but can occur at any age.7-11 Although said to occur more frequently in women than in men, there is no definite gender predilection.2,9,12 Clinically, these benign tumors are easily mistaken for other entities such as lipoma and pleomorphic adenoma on account of their slow growth and absence of neural symptom, and also to mucous retention cyst, minor salivary gland tumour, palatal abscess, fibroma/papilloma, lymphoma, squamous cell carcinoma on account of their site of occurrence.1-2,8-9

Case Report:
An 18 year old male patient was referred by a general physician regarding swelling in the palate, thought to be result of impacted maxillary third molar. History of present illness revealed asymptomatic, single, slowly growing swelling in the left hard palate. The medical history was non-contributory. Mucosa over the swelling is similar in colour and texture with surrounding mucosa, measuring 3x2.5 cm. Clinical examination revealed a sessile, rubbery, non tender, non ulcerated mass on the left posterolateral part of hard palate (Figure 1). Computed tomography scan was made which showed soft tissue lesion in relation to left posterolateral part of hard plate and also eliminated the possibility of impacted maxillary third molar causing the swelling (Figure 2). From these data, the provisional diagnosis was established as some benign soft tissue neoplasm process of minor salivary gland tumour. The tumour was excised under local anesthesia and the specimen was sent to Histopathologic examination (Figure 3, 4).
Histopathologic examination of haematoxylin and eosin stained slides showed a well defined, neural lesion surrounded by a fibrous capsule. The tumour mass consists of neural tissue arranged in predominantly Antoni A pattern (Figure 5) with prominent nuclear palisading and formation of characteristic verocay bodies(Figure 5). Myxoid areas with Antoni B pattern are also seen. Large blood vessels with thrombus formation can be seen interspersed throughout the tumour mass. The diagnosis was schwannoma of the hard palate.

Fig 1: Preoperative view of the lesion

Fig 2: CT showing the tumor

Fig 3: Operative view of the lesion

Fig 4: Excised tumour mass

Fig 5: High power magnification demonstrating neural tissue arranged in Antoni A pattern with prominent nuclear palisading and formation of verocay bodies (highlighted in the inset).(hematoxylin and eosin, 100× [inset, 400×]).
Discussion:
The schwannoma is also called as neurilemmoma, neurinoma, perineural fibroblastoma and is a solitary, slow growing, usually encapsulated, generally asymptomatic neural tumour.\textsuperscript{[10-12]} It can present at any age, however it is more common between the second and third decade of life. The tumour is derived from the Schwann cell sheath, which enlarges, expands and causes displacement and compression of the nerve of origin.\textsuperscript{[1-13]} The present case occurred in the male patient, though literature showed female predilection.

The extracranial schwannoma occurs in the head and neck region. The oral schwannomas usually present in the soft tissue, more commonly the tongue, followed by palate, buccal mucosa and may have similar clinical features to other benign lesions like mucocele, fibromas, lipomas and benign salivary gland tumours.\textsuperscript{[1,9]} The present case was entirely soft tissue lesion, well encapsulated without showing any signs bony erosion.

On occasion, the tumour arises centrally with in bone and may produce bony expansion. Intraosseous schwannomas are most common in the posterior mandible and usually appear as either unilocular or multilocular radiolucencies on radiographs. Pain and paresthesia are not unusual for intrabony tumors. In these cases differential clinical diagnosis of cysts and odontogenic tumors are commonly formulated.\textsuperscript{[1,7,10-12]}

The clinical differential diagnosis could be with any other benign neural tumoral lesion such as fibroma, lipoma, neurofibroma, salivary gland tumor. However the histological differential diagnosis is other neural origin lesions, which could be neurofibrom and neurona, or muscular or fibroblastic origin tumour.\textsuperscript{[1,13,14]}

While arriving to the differential diagnosis of benign tumours of the hard palate, includes traumatic neuroma, schwannoma, neurofibroma and mucosal neuromas representing multiple endocrine neoplasia type 3. Among these, schwanna is unique in that it is composed primarily of a distinct pattern of neoplastic Schwann cells.\textsuperscript{[15]}

Gallo et al\textsuperscript{[16]} reported on 157 cases where 45.2\% of the cases involved in the tongue and 13.3\% involved the cheek. Wright and Jackson\textsuperscript{[17]} 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\% soft palate, and the remainder 19.24\% were in gingival and lip. Das Gupta et al\textsuperscript{[13]} reported on 136 cases of schwannoma in the head and neck that consisted of 60 cases in the neck, 10 cases in the parotid gland, 9 cases in the cheek, 8 cases in the tongue, and 8 cases in the pharynx. Kun et al\textsuperscript{[18]} reported in their study 49 cases, 18 cases in the neck, 11 cases were in the tongue. Wakoh M et al\textsuperscript{[19]} reported 22 cases of schwannomas among these, tumours located in palate 7 cases, tongue 4 cases, submandibular region or oral floor 3, buccal mucosa 2, mental skin 2, lip 2, gingiva 1, temporal region 1.

The microscopic picture of schwannoma is characteristic and can seldom be confused with that of other lesions. Schwannomas are unilobular masses surrounded by a capsule of epineurium and residual nerve fibers often with the edge of the neoplasm attached to the peripheral nerve. The substance of the tumor is composed of a mixture of two cellular patterns Antoni A and Antoni B.\textsuperscript{[1-21]} Antoni A areas are composed of compact spindle cells with twisted nuclei arranged in bundles or fascicles. In highly differentiated areas there may be nuclear palisading and formation of Verocay bodies, which are formed by alignment of two rows of nuclei and cell processes which assume oval shape. Antoni B variant is less cellular and less organized, representing degenerated Antoni A areas composed of haphazardly arranged spindle or oval cells within myxoid, loosely textured, hypocellular matrix punctuated by microcyst, inflammatory cells and delicate collagen fibers.\textsuperscript{[1-21]}

An immunohistochemical examination of the tumor may show positive results with S100 antigen.\textsuperscript{[1,10,14,20,21]} In the present case, the histologic analysis revealed a majority of Antoni A pattern with prominent nuclear palisading and formation of Verocay bodies.

Ancient neurilemmoma exhibits benign degenerative changes in a classic neurilemmoma occurring over time. These changes include cystic, myxoid, edematous and fibrotic areas, vascular abnormalities and atypical cells with pleomorphic nuclei. Ancient neurilemmoma behaves much like a benign neural neoplasm.\textsuperscript{[2,21]}

Cellular schwannoma is classified based on microscopic examination. However, support for a microscopic diagnosis of cellular schwannoma can be obtained by immunostaining. It differs from classic schwannoma by its increased cellularity, nuclear pleomorphism and hyperchromatism, lack of Verocay bodies, and frequently higher mitotic activity.\textsuperscript{[22]}

Microscopic features of both schwannoma and the neurofibroma contain elongated cells with irregular nuclei lying between bundles of collagen fibers. They differ histologically and histogenitically, the schwanna is derived from the schwan cells where as the neurofibroma from the fibroblasts of the perineurium. Neurofibroma is unencapsulated, consisting of a mixture of Schwann cells, perineural cells and endoneurial fibroblasts.\textsuperscript{[1,2,14]}
Asaumi et al,[23] in their study described ultrasonography, computed tomography and magnetic resonance imaging may be helpful in diagnostic and treatment tools, for the estimation of tumour margins and the determination of infiltration to surrounding structures. Magnetic Resonance Imaging was particularly helpful in showing the internal characteristic of the encapsulated mass. However, to rule out resorption due to a malignant tumor, computed tomography scans are useful. Although soft tissue schwannomas have no useful radiographic findings, in the rare case of intrabony (central) schwannoma the role of plain film radiography in verifying locations and determining extent should be appreciated.[9,24] However most intra oral tumours present as relatively small lesions, establishing the differential diagnosis using ultrasonography, computed tomography and magnetic resonance imaging should not be considered as routine or necessary.[23]

Local excision is the treatment of choice. The non encapsulated form requires a margin of normal tissue and careful separation from the involved nerve is also necessary to preserve normal function.[2,9,11,25] Recurrence is rare.[11-21] Malignant transformation of a benign schwannoma is rare.[26,27] In the present case connection with the nerve could not be seen, the mass was well encapsulated and could be totally excised. The prognosis is good.

Conclusion:

The schwannoma present as slow growing painless swelling in the oral cavity or head and neck region not often encountered in clinical practice. This submucosal lesion must be differentiated from other benign lesions that also appear in the same regions. The final diagnosis can only be done after histopathological examination of the lesion. Prognosis is good and recurrence is unknown.

References:
