

INTRAORAL SCHWANNOMAS- DIAGNOSTIC CHALLENGES WITH CASE DESCRIPTION

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ABSTRACT

AIM-To emphasize on a comprehensive approach for correct diagnosis and treatment of intraoral schwannomas.

BACKGROUND—Intraoral schwannomas are benign, encapsulated neurogenic tumours accounting for less than 1% of all head and neck tumours.

CASE DESCRIPTION—We report a case of a 32 yr old female with a painless, progressive cheek swelling of right side for last 4 yrs. It was misdiagnosed earlier which led to its incorrect management. This resulted in a considerable increase in size of the lesion at presentation, thereby requiring a complex surgery for excision.

CONCLUSION—Due to the rare presentations of the intraoral schwannomas, a compartmentalized approach by the clinician may lead to misdiagnosis and incorrect treatment.

CLINICAL SIGNIFICANCE—The rare intraoral presentations have led to frequent delay in diagnosis as well as inadequate treatment of such tumors. Correct diagnosis at primary presentation should be advocated for such rare entities.

KEYWORDS

schwannomas, neurilemmoma, oral cavity, buccal mucosa

INTRODUCTION-

Schwannomas or Neurilemmomas are benign, slow-growing, encapsulated tumours arising from the nerve sheath cells. About 25-48% of these tumours occur in the head and neck region with intraoral schwannomas accounting for less than 1% of all head and neck tumours[1].

Schwannomas can present as intraosseous or peripheral lesions. Some variants with degenerative changes like cystic cavities, hemorrhages, hyalization or nodular growth pattern have also been described. Intraoral peripheral schwannomas are most commonly located in the tongue followed by palate, floor of mouth, cheek mucosa and gums

The rare occurrence of intraoral tumours can lead to misdiagnosis and should be differentiated from clinically similar entities such as neuromas, neurofibromas, granular cell myoblastoma, neuroepitheliomas, fibromas or adenomas.

Computed tomography(CT) or magnetic resonance imaging(MRI) are useful for lesion description and extent but a histopathological examination(HPE) is required for definitive diagnosis. Characteristic tissue patterns on HPE referred as Antoni A and Antoni B and immunohistochemically positive protein S-100, vimentin, neuron-specific enolase(NSE) etc. confirm the diagnosis of schwannomas.

Surgical excision of the encapsulated lesion is the treatment of choice. The causal nerve is rarely identified intraoperatively but relapses or neurosensory defects are infrequent.

CASE DESCRIPTION

A 32 year old female presented to the ENT OPD with complaints of a painless, progressive swelling of the right cheek for last 4 yrs.

On examination, a firm, smooth, well-circumscribed, non-tender mass with well-defined border of size around 4 cm × 3 cm was present over the right cheek. Intraorally, a bulge was seen in the right buccal region with intact mucosa. It was non-compressible and non-fluctuant with no skin fixation.

CT Scan revealed a well-marginated soft tissue mass measuring about 4.1 cm × 3.2 cm in the right cheek anterior to the ramus of mandible and posterior to the maxillary antrum causing elevation of the masseter muscle laterally [Figure 1]. There was no obvious bone erosion.

Fine Needle Aspiration Cytology showed a few small clusters of benign spindle cells with uniform, slender and curved nuclei and

scanty cytoplasm against a background of few freely scattered benign spindle cells and collagenous matrix. There was no evidence of malignancy or salivary gland lesion. A provisional diagnosis of benign spindle cell tumour was made but excision biopsy was advocated for confirmation.

Surgical excision under general anaesthesia was planned and the encapsulated mass was removed in toto [Figure 2,3]. Histopathology showed solid hypercellular areas (Antony A) and hypocellular areas (Antony B) with Verocay bodies thereby confirming the diagnosis of intraoral schwannoma [Figure 4(A) and 4(B)].

DISCUSSION

Schwannoma is a benign, encapsulated perineural tumour of neuroectodermal origin. Neurilemmoma was first described by Verocay in 1910 which he called "Neurinoma". Later in 1935, the term "Neurilemmoma" was coined by Stout[2]. Chi CA et al in their study stated that the age of presentation of schwannomas ranges between 8-72 yrs averaging at about 34 yrs with a female predilection[3]. However, sex predilection cannot be ascertained due to different outcomes of studies by William et al who found male preponderance (83%) while Lucas reported vice versa. Hatziotis and Weirs stated no gender preference [4,5].

Due to its rarity and overlapping clinical symptoms, a patient with an intraoral schwannoma may present to a general physician, dental surgeon, otolaryngologist, head and neck surgeon and even to a general pediatrician or surgeon.

Being asymptomatic and slow growing in nature, the tumour is often diagnosed late. Due to its rarity, the diagnosis at its primary presentation is difficult. The line of treatment often results in inadvertent use of a range of antibiotics or unnecessary investigations, eventually causing a delay in proper management.

In our case also, the patient was a 32 yr old female presenting with a painless slow growing swelling in the cheek and depicted a similar picture.

Intraoral presentation of schwannomas are infrequent with tongue being the most common site followed by palate, floor of mouth, buccal mucosa, gingiva, lips and vestibular mucosa as reported by Hatziotis and Aspidides[5]. Leu and Chang studied 52 cases of schwannomas arising in the head and neck region and found only 7 intraoral lesions amongst them (one in hard palate, one in soft palate, two in submasseteric area, one in the tongue and one in lower lip)[6].

Wright and Jackson studied 146 cases of schwannoma of which 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 [7]. In our case, the lesion was in the submasseteric region which is a rare entity often leading to misdiagnosis and incorrect treatment.

The Schwannomas exhibit characteristic histological findings which is crucial to its correct diagnosis. It consists of a thin fibrous capsule with two types of distinct tissue arrangements of the tumoral proliferation: Antony type A and Antony type B. According to Marx and Stern, the term "Verocay Body" refers to a structure that results due to occurrence of two clusters of palisaded nuclei around an eosinophilic mass, and the eosinophilic area consists of cytoplasmic material and replicated basal membrane [8].

Immunohistochemical examination forms the cornerstone of definite diagnosis of neural tumors. All neural origin tumours show S-100 protein positivity. Passador-Santos et al studied A-100, laminin, fibronectin, Epithelial Membrane Antigen (EMA) and collagens I and II in neural benign neoplasms. They observed an intense positive reaction for S-100 in Schwannomas [9].

Total surgical excision of the tumour is the treatment of choice. Recurrence or malignant transformation is extremely rare.

CONCLUSION

The rare intraoral presentations of schwannoma makes it prone to misdiagnosis and delay in treatment. The condition can be easily overlooked if the clinician adopts a compartmentalized approach and fails to see the full clinical picture. Surgical dexterity should be available to excise the lesion completely to avoid recurrence.

CONFLICTS OF INTERESTS

The authors declare that there is no conflict of interests.

Figure 1-CT Scan showing a well marginated mass anterior to ramus of right mandible and posterior to right maxillary antrum



Figure 2-Intraoperative picture of encapsulated mass being excised intraorally



Figure 3-Specimen of excised intraoral encapsulated mass



Figure 4 (A)-Photomicrograph showing Antony A tissue pattern on HPE

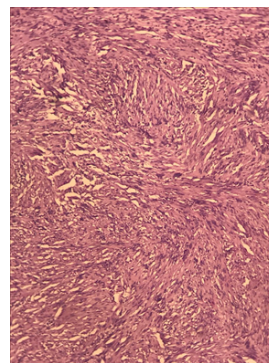
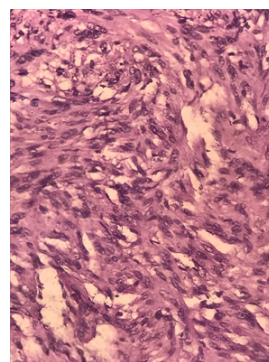


Figure 4 (B)-Photomicrograph showing Antony B tissue pattern on HPE



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