Rare disease

Intraoral schwannoma – a report of two cases

Seema Kurup,¹ Krishnakumar Thankappan,² Nitin Krishnan,¹ Preeti P Nair³¹Oral Medicine & Radiology Department, Amrita School of Dentistry, Kochi, Kerala, India;²Head & Neck Institute, Amrita Institute of Medical Sciences, Kochi, India;³Oral Medicine & Radiology Department, People's College of Dental Sciences & Research Centre, Bhopal, India**Correspondence to** Professor Preeti P Nair, shajihoss@gmail.com**Summary**

Schwannomas, also known as neurilemmomas, are uncommon neoplasms, derived from schwann cells. These neoplasms, although rare, should be considered in the differential diagnosis of slow growing tumour masses of the oral cavity. The growth of these tumours sometimes causes displacement and compression of the nerve of origin, giving rise to clinical signs and symptoms. It can develop at any age and there is no sex predilection. Intraorally, the tongue is the most common site followed by the palate, floor of the mouth, buccal mucosa, lips and jaws. Surgical excision is the treatment of choice for schwannomas, with few reports of recurrence or malignant transformation. In this paper, the authors report two cases of patients with intraoral schwannoma, where the preoperative clinical diagnosis was inconclusive and final diagnosis was established based on radiographic and histopathological examination.

BACKGROUND

Schwannomas originate from schwann cells of the neural sheath and are solitary, well-encapsulated slow growing tumours. It is also named as neurilemoma, neurinoma and schwann cell tumour.¹ Approximately 25%–45% of all schwannomas are seen in the head and neck region and are rarely found in the oral cavity.² Also it is the most common neural tumour found in the parapharyngeal space and vagus nerve is reported to be the origin for 50% of schwannomas in parapharyngeal space.³

Intra oral schwannomas are usually seen in the tongue, palate, buccal mucosa, gingiva and lips. It can present itself at any age and without any gender predilection. However, it is most commonly seen in young and middle aged. The clinical presentation varies with the anatomical area involved.^{3,4}

The clinical differential diagnosis could be with any other benign tumoural lesions such as fibroma, lipoma, neurofibroma or salivary glands tumour. Radiological examinations such as a CT scan with contrast and MRI reveal the extension of the tumour.⁴ Final diagnosis of the neoplasm is achieved with radiological, histopathological or immunohistochemical evaluations. Surgical excision is the treatment of choice and recurrence is very rare.³

This paper describes two cases of schwannoma in head and neck region at different sites involving the floor of the mouth and posterior third of the tongue.

CASE PRESENTATION

Case 1: a 56-year-old male patient, presented with swelling in the floor of the mouth of 2 months duration. There was no history of trauma, bleeding, discomfort from the swelling or pain during eating. He denied dysphagia and dyspnoea. Apart from being a known diabetic, his medical, family, dental history and review of systems were not significant.

There was no cervical lymphadenopathy. Intraoral examination revealed a 4×3 cm firm, non-tender and mobile

swelling in the floor of the mouth. Overlying mucosa was intact (figure 1).

Case 2: a 31-year-old female patient, otherwise healthy person, presented with difficulty in swallowing food since 1 month. There was no history of pain or cough. Medical, family, dental history and general examination were not significant. Hopkins rod endoscopy revealed a large sub-mucosal swelling arising from the base of tongue on the right side, of size about 3×3 cm hanging onto the right vallecule (figure 2). Larynx appeared normal. Neck examination was unremarkable.

INVESTIGATIONS

Case 1: laboratory investigations, carried out as routine preoperative procedure did not yield anything remarkable. The swelling in the floor of the mouth was close to the mandible. Hence a CT scan which is a superior

Figure 1 Photograph revealing a swelling of the floor of the mouth.

imaging modality for delineating any bone involvement was chosen for radiological evaluation. Contrast enhanced multi-detector CT revealed well-defined heterogeneously enhancing lesion in the left floor of the mouth (figure 3).

Case 2: contrast enhanced MRI of the neck revealed a well-defined enhancing mass in posterior third of tongue on the right side (figure 4)

Histopathologic findings of tissue sections in both cases revealed a fairly circumscribed neoplasm composed of spindle shaped cells arranged in fascicles and palisading sheets. Both cellular and hypocellular areas (Antoni A and Antoni B) with the formation of verocay bodies in the cellular areas were seen. There were no mitotic and necrotic

Figure 2 Endoscopic examination revealing large swelling in the base of tongue with extension to the vallecula.

Figure 3 Contrast enhanced multi-detector CT revealing well-defined heterogeneously enhancing lesion in the left floor of the mouth.

changes observed (figures 5 and 6). Similar histological features were evident in case 2 (figures 7 and 8)

Thus a final diagnosis of schwannoma was put forth in both the cases.

DIFFERENTIAL DIAGNOSIS

Case 1: ranula, salivary gland tumour, rhabdomyoma, fibroma, leiomyoma.

Case 2: lymphangioma, leiomyoma, neurofibroma.

TREATMENT

Case 1: the patient was posted for surgery and wide excision under general anaesthesia was carried out.

Case 2: the patient underwent surgery-excision by lateral pharyngectomy under general anaesthesia.

Figure 4 MRI showing well-defined enhancing mass in the posterior third of the tongue.

Figure 5 Low power view showing the alternating hypercellular-Antoni A (thick arrow) and hypocellular-Antoni B (thin arrow) areas (case 1) (H&E, 10x).

Figure 6 High power magnification showing the palisading spindle cells surrounding central stromal cells -Verocay bodies (arrow) (case 1) (H&E, 40x).

Figure 7 Low power photomicrograph (case 2) (H&E, 10x).

Figure 8 High power photomicrograph (case 2) (H&E, 40x).

OUTCOME AND FOLLOW-UP

Case 1: postoperative period was uneventful. The patient was stable and afebrile. Ryle's tube removed at the time of discharge. Examination on day of follow-up, 10 days later revealed no abnormality.

Case 2: postoperative period was uneventful and on sixth postoperative day, started on oral feed which was well tolerated and Ryle's tube was removed at the time of discharge. Suture removal was carried out on day of follow-up.

Both patients were recalled after 1 month for routine check up and were found to be normal.

DISCUSSION

Benign nerve sheath tumours, including schwannoma (neurilemmoma) and neurofibroma, are encountered in oral and perioral locations.⁵ Schwannomas are infrequent, benign neoplasms that can arise from any cranial, peripheral or autonomic nerve that contain schwann cells. In the head and neck, the optic and olfactory nerves lack schwann cell encasement.⁶ Approximately 1%–12% of schwannomas occur intraorally, the tongue being the most common site.^{7–10} Leu and Chang¹¹ reviewed a series of 52 cases of schwannomas originating in the head and neck region area over an 8-year period. In seven of these cases, the lesions were located in the oral cavity, one in the hard palate, one in the soft palate, two in submasseteric and one each in the tongue and lower lip. Wright and Jackson¹² 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.

A total of 126 cases of schwannoma of the tongue have been reported in the English literature over the past 51 years. Schwannomas of the tongue typically present in the third decade of life, display no gender predilection and often present as a painless mass. Schwannomas are likely to elicit distressing symptoms when they occur in the posterior one-third of the tongue. The location of the schwannoma was categorised as either anterior two-thirds or posterior one-third of the tongue. This classification was possible in 56 of the 126 cases with 37 cases occurring anteriorly and 19 cases identified posteriorly. Of the 19 patients with posterior one-third tumours, 12 patients reported symptoms.¹³

A review of literature revealed very few cases of intraoral schwannoma in the floor of the mouth. Enoz and Souglu¹⁴ in 2006 reported a case of lingual schwannoma. Okada *et al*¹⁵ in 2011 reported a rare case of schwannoma in the floor of the mouth, arising from the sublingual gland. Liesa¹⁶ in 1993 reported a schwannoma of the floor of the mouth which had the biggest tumour diameter of 7.5 cms.

Parapharyngeal space tumours are very rare and account for only 0.5% of all head and neck tumours. Approximately 50% of the tumours have a salivary origin, 20% are neurogenic and the remaining 30% are represented by tumours such as benign and malignant lymphoreticular lesions, metastatic lesions and carotid body tumours. Schwannoma (neurilemmoma) is the most common neural tumour next to salivary gland tumour found in the parapharyngeal space. Vagus nerve is reported to be the origin for 50%

of parapharyngeal schwannomas and cervical sympathetic chain is the next common source. Generally schwannomas are characterised by slow and asymptomatic growth; however, its progressive growth in parapharyngeal region may result in pressure effect manifestations like dysphasia and hoarseness of voice.³

Histologically, schwannomas display several features. Virtually, all of these tumours are encapsulated. Beneath this capsule, two main patterns are seen intermingled but sharply defined from each other. The first pattern is referred to as Antoni type A which consists of closely packed schwann cells that form bundles or are arranged in rows with palisading, elongated nuclei. Free bands of amorphous substance between rows of nuclei constitute the Verocay bodies. The second pattern is known as Antoni type B and is composed of very loosely arranged schwann cells set in a meshwork of reticulum fibers and microcysts.¹⁷ In addition to these characteristic patterns, diagnosis is aided by immunohistochemical markers, S-100 and Leu 7 antigen, which support the schwann cell nature of these tumours. According to Chrysomali *et al*,¹⁸ the tumoural cells with Antoni A show greater intensity scores compared with Antoni B tumour pattern. In our cases, final diagnosis of schwannoma was established by histopathology.

Rarely, long-standing schwannomas can undergo degenerative changes, dominated by large cystic myxomatous areas with variable bizarre spindle cells and exhibit a thick fibrous connective tissue capsule, a relative absence of necrosis, and distinct areas of increased hypercellularity, with nuclear pleomorphism and hyperchromatism. The histologically misleading nature of these lesions, which could be suggestive of a malignancy is termed as 'ancient schwannomas'.⁵

The Antoni-B structure of a schwannoma is sometimes difficult to distinguish from a neurofibroma as tumours of the nerve sheath with hybrid features of neurofibromas and schwannomas have been identified. Immunohistochemically, schwannomas stain for S-100 protein and CD34, but not for Factor XIIIa. However, neurofibromas stain for S-100 protein, CD34 and Factor XIIIa. The staining intensities for Factor XIIIa therefore clearly differ between schwannomas and neurofibromas.¹⁹

According to Asaumi *et al*,²⁰ ultrasonography, CT and MRI may be helpful diagnostic and treatment tools, for the estimation of tumour margins, the lesion composition and the determination of whether there is infiltration to surrounding structures or not. Lack of surrounding tissue infiltration and well-circumscribed borders are common radiographic features in schwannoma.² These imaging modalities also help in determining the nerve of origin pre-operatively and may help to reduce postoperative neural deficits.³ The imaging modality of choice for schwannomas of the tongue is MRI. MRI is superior to CT in several aspects. The MR image is not degraded by dental amalgam or the beam-hardening artifacts that plague CT scanning of the oral cavity. In addition, MR allows an accurate measurement of tumour size and precise localisation in relation to other structures. On MRI, tongue schwannomas appear isointense to muscle on T1-weighted images and homogeneously hyperintense on T2-weighted images. Moreover, these tumours usually appear smooth, well demarcated, and do not invade the surrounding musculature.²¹

Differential diagnosis must be made in relation to numerous benign neoplasms based on epithelial and connective tissues (lipoma, fibroma, leiomyoma and adenoma) and in relation to malignant tumours depending on speed of growth and clinical appearance of the neoplasia. However, the histological differential diagnosis is made with other neural origin lesions, which could be neurofibroma, neuroroma, malignant schwannoma, muscular or fibroblastic origin tumour.^{22 23}

Treatment is always surgical. The schwannoma should be excised in its entirety to avoid tumour recurrence, even if the nerve of origin cannot be preserved, because of the possibility of malignant transformation, which although rare has been described in the literature.²⁴

Learning points

- Schwannomas can present intraorally and should be considered in the differential diagnosis of any solitary, slowly enlarging oral cavity lesion.
- Depending on location of tumour, patients may report symptoms such as pain, hoarseness, dysphagia and cranial nerve neuropathies.
- Diagnosis of schwannoma is confirmed with histopathologic evaluation.
- Early treatment which includes surgical excision can prevent chances of malignant transformation.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. Artzi Z, Taicher S, Nass D. Neurilemmoma of the mental nerve. *J Oral Maxillofac Surg* 1991;**49**:196–200.
2. Colreavy MP, Lacy PD, Hughes J, *et al*. Head and Neck Schwannomas - a 10 year review. *J Laryngol Otol* 2000;**114**:119–24.
3. Giraddi G, Vanaki SS, Puranik RS. Schwannoma of parapharyngeal space. *J Maxillofac Oral Surg* 2010;**9**:182–5.
4. Anil HT, Gowda BV, Lakshmi S, *et al*. Schwannoma of the palatine tonsil. *J Laryngol Otol* 2005;**119**:570–2.
5. Humber CC, Copete MA, Hohn FI. Ancient schwannoma of upper lip: case report with distinct histologic features and review of the literature. *J Oral Maxillofac Surg* 2011;**69**:e118–22.
6. Lollar KW, Pollak N, Liess BD, *et al*. Schwannoma of the hard palate. *Am J Otolaryngol* 2010;**31**:139–40.
7. López JI, Ballester C. Intraoral schwannoma. A clinicopathologic and immunohistochemical study of nine cases. *Arch Anat Cytol Pathol* 1993;**41**:18–23.
8. Das Gupta TK, Brasfield RD, Strong EW, *et al*. Benign solitary Schwannomas (neurilemmomas). *Cancer* 1969;**24**:355–66.
9. Hatziotis JC, Asprides H. Neurilemmoma (schwannoma) of the oral cavity. *Oral Surg Oral Med Oral Pathol* 1967;**24**:510–26.
10. Jones JA, McWilliam LJ. Intraoral neurilemmoma (schwannoma): an unusual palatal swelling. *Oral Surg Oral Med Oral Pathol* 1987;**63**:351–3.
11. Leu YS, Chang KC. Extracranial head and neck schwannomas: a review of 8 years experience. *Acta Otolaryngol* 2002;**122**:435–7.
12. Wright BA, Jackson D. Neural tumors of the oral cavity. A review of the spectrum of benign and malignant oral tumors of the oral cavity and jaws. *Oral Surg Oral Med Oral Pathol* 1980;**49**:509–22.
13. Cohen M, Wang MB. Schwannoma of the tongue: two case reports and review of the literature. *Eur Arch Otorhinolaryngol* 2009;**266**:1823–9.
14. Enoz M, Suoglu Y, Ilhan R. Lingual schwannoma. *J Cancer Res Ther* 2006;**2**:76–8.
15. Okada H, Tanaka S, Tajima H, *et al*. Schwannoma arising from the sublingual gland. *Ann Diagn Pathol* 2011 (In Press).
16. Liesa FR, Gili BA, Arevalo BA, *et al*. Tumor of the mouth floor: submucosal schwannoma. *Acta Otorrinolaryngol Esp* 1993;**44**:147–5.

17. **Cherrick HM**, Eversole LR. Benign neural sheath neoplasm of the oral cavity. Report of thirty-seven cases. *Oral Surg Oral Med Oral Pathol* 1971;**32**:900–9.
18. **Chrysomali E**, Papanicolaou SI, Dekker NP, et al. Benign neural tumors of the oral cavity: a comparative immunohistochemical study. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1997;**84**:381–90.
19. **Kubota Y**, Yanai Y, Kumamaru W, et al. Multiple schwannomas in the oral floor: case report. *Br J Oral Maxillofac Surg* 2011;**49**:e33–5.
20. **Asaumi J**, Konouchi H, Kishi K. Schwannoma of the upper lip: ultrasound, CT, and MRI findings. *J Oral Maxillofac Surg* 2000;**58**:1173–5.
21. **Flickinger FW**, Lozano RL, Yuh WT, et al. Neurilemoma of the tongue: MR findings. *J Comput Assist Tomogr* 1989;**13**:886–8.
22. **Requena L**, Sanguenza OP. Benign neoplasms with neural differentiation: A review. *Am J Dermatopathol* 1995;**17**:75–96.
23. **López-Carriches C**, Baca-Pérez-Bryan R, Montalvo-Montero S. Schwannoma located in the palate: clinical case and literature review. *Med Oral Patol Oral Cir Bucal* 2009;**14**:e465–8.
24. **Kun Z**, Qi DY, Zhang KH. A comparison between the clinical behavior of neurilemmomas in the neck and oral and maxillofacial region. *J Oral Maxillofac Surg* 1993;**51**:769–71.

This pdf has been created automatically from the final edited text and images.

Copyright 2012 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Kurup S, Thankappan K, Krishnan N, Nair PP. Intraoral schwannoma – a report of two cases. *BMJ Case Reports* 2012;10.1136/bcr.12.2011.5389, Published XXX

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow

Keep up to date with all published cases by signing up for an alert (all we need is your email address) <http://casereports.bmj.com/cgi/alerts/etoc>