

A case report on lingual schwannoma

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Summary

Schwannomas are benign tumours of peripheral nerves originating in the nerve sheaths. Only 1% are reported in the oral cavity. We report a 24-year-old female who presented with a 10-year history of a mass at the base of the tongue with associated odynodysphagia, referred otalgia and loss of weight. Oral examination revealed a large left sided base of tongue mass extending into the vallecula. An open transcervical suprahyoid resection was performed with a good outcome.

Keywords: benign oral tumour, lingual schwannoma, transcervical suprahyoid resection, Antoni A cells, Antoni B cells.

Case report

A 24-year-old female presented with a 10-year history of a mass at the base of the tongue. There was associated odynodysphagia, left side referred otalgia and loss of weight. She had no associated dysphonia, gustatory

disturbance or evidence of upper airway obstruction. Oral cavity examination revealed a large mass on the left side of the base of the tongue extending into the vallecula, with no impairment of lingual mobility, and no associated cervical

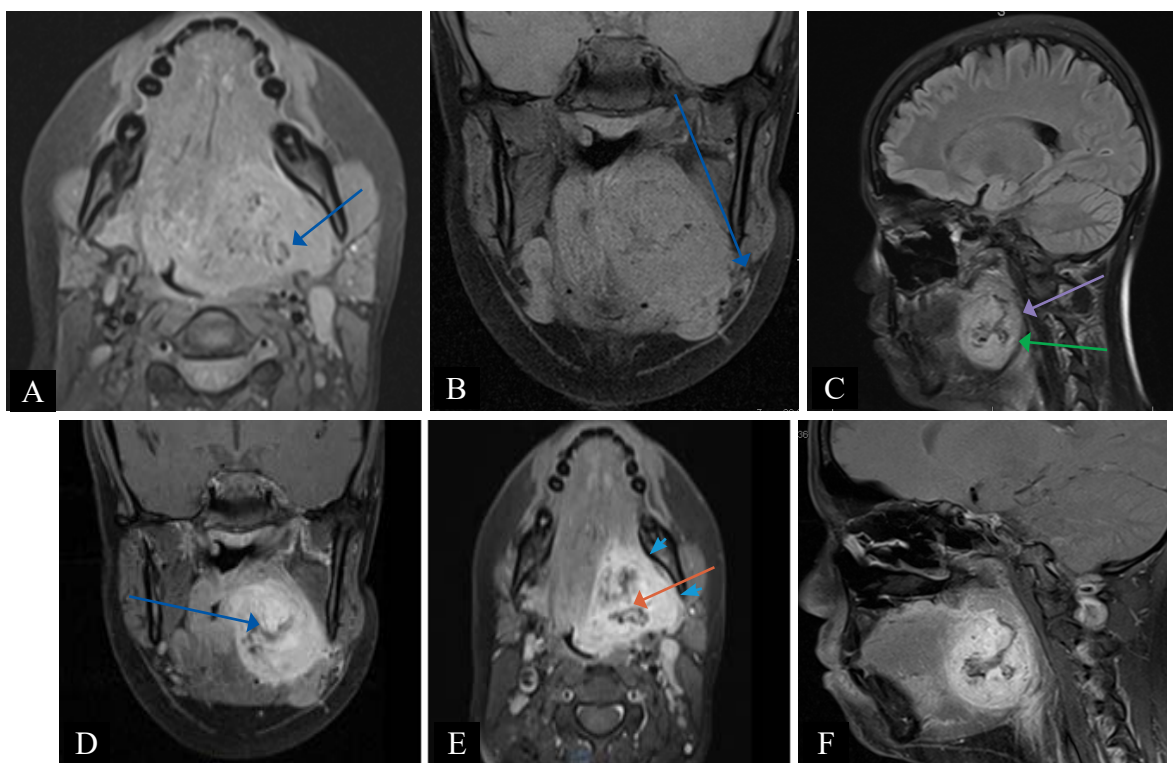


Figure 1: A – Axial pre-contrast fat saturated T1 weighted image MRI scan of the neck showing a well-defined centred mass (blue arrow) within left tongue base that is isointense to muscle. B – Coronal pre-contrast fat saturated T1 weighted image MRI showing mass abutting the deep lobe of the left parotid gland. C – Sagittal FLAIR T2 weighted image MRI showing a hyperintense mass with no plane of cleavage between mass and left posterior intrinsic muscles of the tongue. Multiple ring-like lesions are seen in the mass – fascicular sign (purple arrow). Target sign is a central area of low intensity surrounded by a T2 hyperintense rim (green arrow). D-F – T1 weighted MRI neck images post-gadolinium contrast in three planes: coronal (D), axial (E) and sagittal (F). Lingual schwannoma of the base of tongue (blue arrows) is largely solid and vascular with brisk enhancement and central necrosis (orange arrow). The fat split sign is demonstrated by the blue arrowhead. Sagittal post-contrast T1 image of the neck (F) shows no involvement of the prevertebral space.



Figure 2: Steps of transcervical suprahyoid resection of lingual schwannoma
A – A linear incision was placed just above the hyoid bone. B – Base of tongue mass accessed by dissection through the platysma, mylohyoid and entering the vallecular space via a suprahyoid pharyngotomy. C – Mass excised en-bloc.

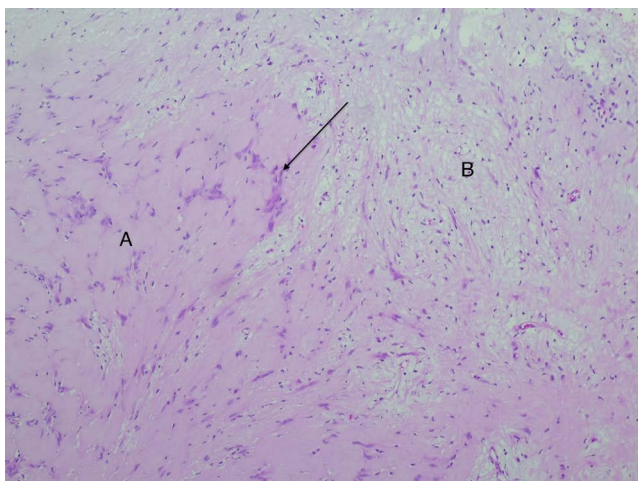


Figure 3: Low magnification reveals cellular Antoni A areas (A) with Verocay body formation (arrow), interspersed with paucicellular Antoni B areas (B) (haematoxylin & eosin, 100x).

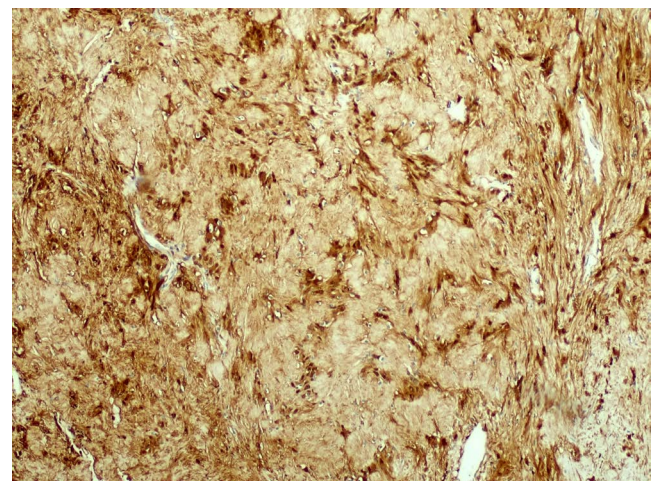


Figure 4: Immunohistochemistry for S100 protein showed strong diffuse nuclear and cytoplasmic staining.

lymphadenopathy. The initial clinical impression was of a large submucosal tumour.

Contrast-enhanced computed tomography (CT) scan revealed a heterogeneously enhancing mass at the base of the tongue on the left side, measuring 40 mm x 40 mm x 20 mm. There was extension anteriorly to the posterior third of the true tongue abutting the left lateral pharyngeal wall and prestyloid parapharyngeal space.

A magnetic resonance imaging (MRI) was ordered for better soft tissue delineation, increased tissue contrast and spatial resolution. The mass was hypointense on T1 weighted image (Figure 1), and hyperintense on T2 weighted image with post-contrast enhancement. Additional features suggestive of lingual schwannoma demonstrated on MRI included a split fat sign (thin peripheral rim of fat on T1WI), a target sign (central low signal within the lesion), and a fascicular sign (multiple internal small ring-like structures). There was no associated cervical lymphadenopathy or evidence of distant metastases.

The patient was scheduled for an examination under anaesthesia. Deep biopsies were taken from the mass via rigid laryngoscopy. The histology was consistent with a lingual schwannoma.

Following discussion with the patient, a temporary tracheostomy and a transcervical suprahyoid resection of the mass was performed. A linear incision was placed just above the hyoid bone (Figure 2A). The base of tongue mass was

accessed by dissecting through the platysma, mylohyoid and entering the vallecular space via a suprahyoid pharyngotomy (Figure 2B). The lingual artery, lingual nerve and hypoglossal nerve was isolated. The mass was excised macroscopically en-bloc and sent for histopathology analysis (Figure 2C). The defect was closed primarily over a suction drain.

The postoperative course was uncomplicated. The tracheostomy was decannulated after 24 hours and she was discharged home tolerating a normal diet. Specifically, she had no postoperative lingual, glossopharyngeal or hypoglossal nerve fall-out. On three-month postoperative follow-up she had no concerns and complete symptomatic resolution.

Histological sections showed a well-circumscribed neoplasm composed of bland spindle cells within a collagenous to fibrillary stroma, demonstrating bi-phasic architecture. Cellular areas with nuclear palisading and formation of Verocay bodies (compatible with Antoni A areas) were seen interspersed with paucicellular areas, comprising loose microcystic tissue with myxoid stroma (compatible with Antoni B areas) (Figure 3). Mitotic figures were sparse, and no features of malignancy were seen. Immunohistochemistry for S100 protein showed strong diffuse nuclear and cytoplasmic staining (Figure 4).

Discussion

Schwannoma was first reported by Virchow in 1908 and is a benign tumour of the peripheral nerves originating from the nerve sheaths.¹ Schwannomas of the oral cavity are rare and

limited to a few case reports in the literature. They account for 1% of benign tumours reported in the oral cavity and oropharynx, with the most common sub-site being the base of tongue.²

In more than 50% of oral schwannomas, identifying the nerve of origin, specifically the lingual, hypoglossal, or glossopharyngeal, can be challenging.³ These tumours usually appear between the second and fourth decade of life, with no predilection for gender or ethnicity. In a systematic review of 15 cases, Sitenga et al. found that the most common presenting symptoms included lingual swelling (87%), dysphagia (46.7%), dysarthria (33.3%).⁴

Large schwannomas, exceeding 3 cm in greatest dimension, are more likely to produce debilitating symptoms including throat discomfort, dysarthria, snoring, dyspnoea, and impaired tongue movement.⁵ Interestingly, pain is generally identified in subjects with neurofibromatosis type 2 (schwannomatosis) rather than in sporadic cases.⁶ Our patient, despite being a relatively large, sporadic case of lingual schwannoma, presented with mild odynodysphagia and referred otalgia only.

MRI is the optimal form of imaging due to its superior soft tissue resolution and lower risk radiation exposure. As described in the case report, schwannomas have typical pathognomonic features on MRI scanning.

The diagnosis is confirmed by cytological or histological sampling via a transoral or transcervical approach. Histologically, conventional schwannomas are bland spindle cell tumours that demonstrate characteristic biphasic zonation into compact areas (Antoni A areas) showing occasional nuclear palisading into parallel rows (known as Verocay bodies), alternating with less cellular, loosely arranged, often myxoid-appearing areas (Antoni B areas). The lesional cells possess poorly discernible cell borders, modest amounts of eosinophilic cytoplasm, and normochromatic elongated tapered nuclei. Microcystic change and the presence of thick-walled hyalinised blood vessels can additionally be seen in Antoni B areas. Degenerative changes may occur in schwannomas (known as ancient change), seen as scattered atypical to bizarre-appearing nuclei. However, mitotic figures are rarely seen, and malignant transformation to malignant peripheral nerve sheath tumour (MPNST) is exceedingly rare and reported at between 8 – 13.5%. The immunohistochemical profile of schwannomas confirm their peripheral nerve sheath origin, with diffuse nuclear and cytoplasmic staining for S100 protein. Extensive nuclear immunoreactivity for SOX10, and variable expression of GFAP can also be demonstrated.⁷

Literature supports surgical resection as the mainstay of treatment. When complete excision is performed, recurrence has not been reported.⁸ A transoral approach is the most widely reported in the literature. Multiple surgical approaches are available to access the tongue base, including transoral, lip split with mandibulotomy and mandibular swing, lip split with mandibulectomy, transcervical lingual pull through and suprahyoid pharyngotomy.

Tongue base tumours can pose challenges in perioperative airway management. In our centre, we routinely perform an elective tracheostomy for base of tongue tumours to secure the airway in view of potential postoperative swelling and haemorrhage.

The above case report is amongst the largest lingual schwannoma described in the medical literature to

date measuring 40 x 30 x 20 mm, and the second case reportedly excised via a suprahyoid pharyngotomy.⁹ The largest documented lingual schwannoma of tongue base measured 65 × 50 × 30 mm on gross examination and required a submental flap for the functional reconstruction of the defect.¹⁰ Other potential reconstructive options would include skin grafts, local tongue flaps, myocutaneous pedicle flaps, and microvascular free tissue transfer.¹¹

Base of tongue schwannomas are rare but should still be considered as part of the differential for lingual mass lesions. MRI is the diagnostic imaging modality of choice due to characteristic features, lower risk of radiation and superior soft tissue resolution. Complete excision, when possible, is the treatment of choice. The suprahyoid pharyngotomy illustrated is an elegant, low-risk approach to the base of tongue often forgotten in the era of expanding transoral approaches. This approach offers low morbidity, preservation of normal speech and swallowing as well as excellent cosmesis. Although not essential, a perioperative tracheostomy is recommended.

Funding source

None.


Ethical approval

Ethical approval was obtained from the University of Cape Town Faculty of Health Sciences Human Research Ethics committee (Ref: 954/2023).

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