Introduction

Schwannomas (neurilemmomas) are benign, slowly growing, single, painless and encapsulated neurogenic tumors. The etiology is unknown, but it is neuroectodermal in origin arising from the proliferation of Schwann cells of the peripheral, autonomic and cranial nerves. Only 50% have identified the associated peripheral nerve. This lesion is not usually considered in the differential diagnosis of numerous benign epithelial and connective tissues growth affecting the tongue. The cornerstone for its diagnosis is definitive histopathological examination.

Case report

A 20-year-old female presented with slowly progressive F.B. sensation, muffled voice, dysphagia and snoring for 11 years duration. There was neither paresthesia, nor choking, aspiration, nasal regurgitation or bleeding from the mouth. The patient was healthy, non-smoker with unremarkable past medical and family history.

On examination, a non-ulcerating mass was seen in the left side of the tongue base and its lower limit cannot be assessed. Mobility of the tongue and soft palate were normal. On palpation, it was firm, non-tender, with no bleeding on touch, not attached to surrounding tissue. NPL confirmed the findings and the lesion was reaching vallecula pushing epiglottis forwards with intact mucosal covering. CT scan of neck with contrast enhancement has showed superficial, well defined soft tissue lesion, involving the base of tongue on the left side and left tonsillar pillar with no infiltration of adjacent tissue and partial obliteration of oropharyngeal airway.

MRI has demonstrated a well-circumscribed mass, uniform isointense to the surrounding muscles in T1, heterogeneously enhancing hyperintense with a fascicular sign on T2, and markedly enhanced after intravenous administration of gadolinium DTPA (Fig.2 A–D). No significant intra-operative bleeding was encountered. The lesion was submucosal, well encapsulated and with a good cleavage plane. No significant intra-operative bleeding was encountered. The bed of the tumor was sutured primarily with Vicryl 3/0 to maintain haemostasis.

Postoperative the tongue mobility was good. Patient was followed up regularly for 18 months on monthly basis during which she remained symptom free and no evidence of recurrence.

Discussion

Head and neck schwannomas represent 25% of extracranial schwannomas. However, intrar oral locations are very rare (only 1%) and commonly seen at the tongue (60% in anterior third of the tongue vs 34% in the posterior two thirds) followed by palate, floor of mouth, buccal mucosa, gingiva, lips and the vestibular mucosa in descending manner.

Clinically, schwannomas are indistinguishable from other intraoral slow-growing encapsulated smooth submucosal swellings like neurofibromas, granular cell tumor, salivary gland tumors, traumatic fibroma, leiomyomas, rhadomyomas, lymphangiomata, hemangiomas, epidermoid cysts, lipomas, inflammatory lesions and ectopic lingual thyroid.

Malignant schwannomas are an exceptionally rare. They account for 5% of all soft tissue sarcomas. Of these, only 6–14% are in the head and neck. MRI is superior to CT for schwannomas, because of its increased tissue contrast and spatial resolution. Most schwannomas appear isointense or isointense relative to muscle on T1-weighted images, hyperintense on T2-weighted images, and show strong enhancement after contrast administration.

CT scan of neck with contrast enhancement has showed superficial, well defined soft tissue lesion, involving the base of tongue on the left side and left tonsillar pillar with no infiltration of adjacent tissue and partial obliteration of oropharyngeal airway.

The goal of surgical therapy is complete pericapsular excision. Different surgical approaches have been described for tongue base schwannomas. They include trans-oral (robotic surgery, CO2 Laser excision, or using diathermy, submandibular pull-through, suprahypoglossal (trans-hyoid) lateral pharyngotomy approaches and midline submandibulotomy and tongue split.

Most lesions could be safely excised via transoral access except those with deep extension towards the floor of the mouth or vallecula; an open approach would be required. The advantages of transoral approach could be summarized as: (1) avoidance of external scar, (2) improved preservation of sensation and function, (3) earlier swallowing rehabilitation and (4) decreased frequency of fistula formation. However, it has the disadvantage of limited exposure and less optimal visualization of deeper structures.

Conclusion

Despite their rarity, tongue base schwannomas should be included whenever a benign submucosal swelling is suspected in this area.

Transoral approach is still the standard approach with the least morbidities and best functional outcomes.

References