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Case Report

Tongue base schwannoma: differential diagnosis and imaging features with a case presentation

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ABSTRACT

Schwannomas are slow growing, encapsulated neoplasms that arise from the nerve sheath. A vast majority of these benign neoplasms occur in the head and neck region, most commonly involving the 8th cranial nerve. Schwannomas arising from the base of tongue are very rare and, thus, can easily escape the list of differential diagnosis for a posterior tongue mass. A systematic approach is recommended for diagnosis of a posterior tongue mass, with neoplastic, infectious, and congenital categories. We report a case of a 24-year-old female, who presented with pressure sensation in the throat. On imaging, she was found to have a mass in the right posterior tongue with follow-up biopsy results yielding a schwannoma. Characteristic imaging features for various possible etiologies of a posterior tongue mass are also discussed.

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Introduction

Schwannomas are benign tumors of nerve sheath origin, most commonly arising from 8th cranial nerve [1]. About 1% of these neoplasms can be intra-oral in origin, most commonly occurring in the tongue. A vast majority of schwannomas arising from the tongue occur in the anterior two thirds, whereas those arising from the base of tongue are exceedingly rare [2–4]. We present a case of a young female with a tongue-based schwannoma. A differential diagnosis for a tongue-based mass is also presented.

Case report

A 24-year-old female presented with pressure sensation in the throat, without any dysphagia or dyspnea. Physical examination revealed tenderness in anterior/medial aspect of the neck. Initial evaluation was done with a neck computed tomography (CT), which displayed an enhancing mass arising from the right posterior aspect of the tongue (Fig. 1). This warranted further evaluation with magnetic resonance imaging (MRI; Figs. 2 and 3) that showed a well circumscribed, heterogeneously enhancing, T2 hyperintense, and T1

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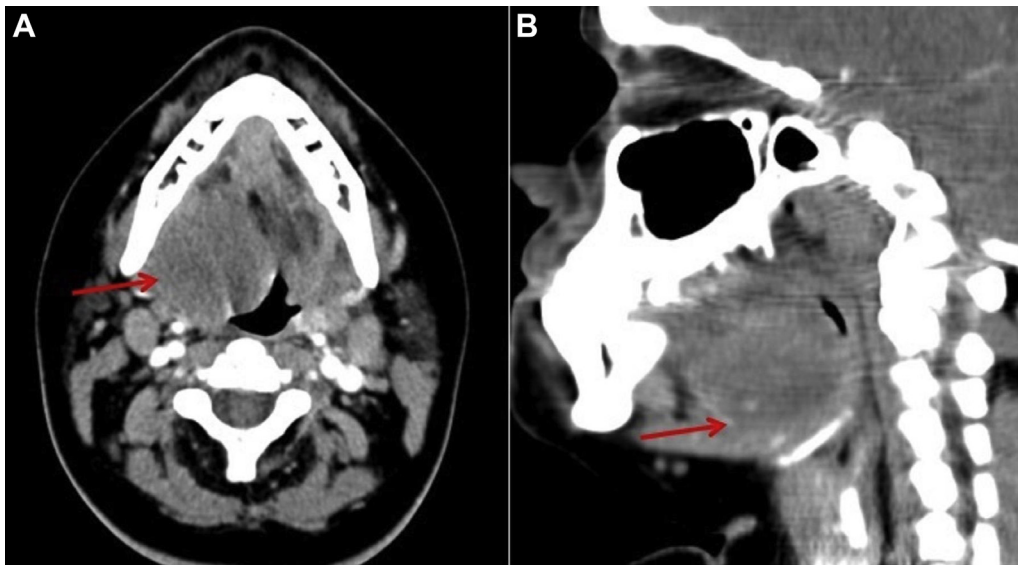


Fig. 1 – Contrast-enhanced axial (A) and sagittal reformatted (B) CT images of the neck show a globular lesion (red arrow) positioned in the right tongue base with demonstration of mild homogeneous enhancement resulting in mild narrowing of oropharyngeal airway on the right.

isointense mass with multiple intrinsic small ring-like heterogeneous lesions (fascicular sign).

Subsequently, the patient underwent direct laryngoscopy and biopsy. Pathology demonstrated lymphocytic infiltrate with well-defined hypocellular and hypercellular areas and Verocay bodies (Figs. 4 and 5). Confirmation was also obtained by a positive S100 and negative CD 68 stain. Thus, a diagnosis of schwannoma was established.

Patient underwent a transoral robotic excision of the mass.

Discussion

Schwannomas can arise from all cranial nerves (except CN I and CN II), with CN VIII being the most common. Other locations include spinal nerve roots, flexor surfaces of upper and lower extremities, and tongue [5,6]. Although most schwannomas are known to be solitary, there is an association with neurofibromatosis type 2 [7].



Fig. 2 – Axial STIR image (A), axial precontrast T1WI image (B), and axial postcontrast T1WI image (C). Axial image of the neck at the level of C3 vertebra shows a well-defined centered mass within right tongue base that is isointense to muscle on T1WI (red arrow in B) and heterogeneously STIR hyperintense (A); demonstrating heterogeneous enhancement on axial postcontrast fat-saturated T1WI (red arrow (C)). The lesion demonstrates the “fascicular sign” on STIR image as shown by yellow arrow (A). This lesion is noted to abut the right submandibular gland laterally as well as bulges posteromedially with mild compromise of the oropharyngeal airway at this level.

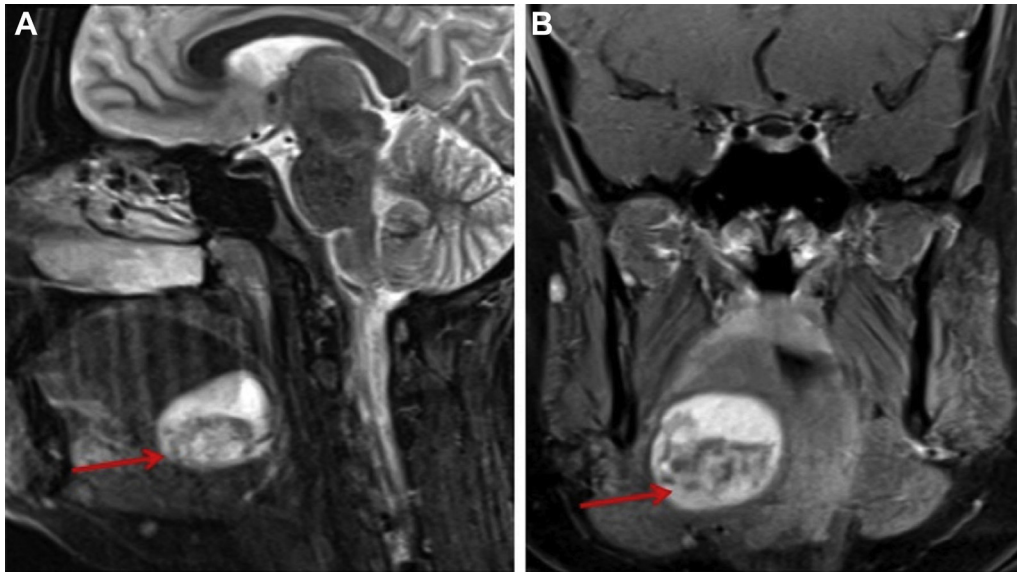


Fig. 3 – Sagittal STIR image of the neck (A) and coronal postcontrast fat-saturated image (B). Sagittal STIR image of the neck shows a well-defined STIR hyperintense mass (red arrow in A) within the right tongue base, which demonstrates heterogeneous enhancement on coronal postcontrast fat-saturated T1WI (red arrow in B). This lesion is again noted to abut the right submandibular gland laterally.

Imaging has become an integral part of evaluation for tongue base lesions, and thus, a systematic imaging approach should be considered. As demonstrated in Fig. 6, lesions of the tongue can be divided into infectious, neoplastic, and congenital categories. An infectious process, such as an abscess, appears hyperintense on T2WI with a thick rim-enhancing margin [8].

Benign neoplastic lesions like leiomyoma, neurofibroma, and schwannoma can have similar imaging appearance. On contrast-enhanced CT, a schwannoma demonstrates heterogeneous enhancement [9,10]. Additionally, it can demonstrate cystic and fatty degeneration. On MRI, schwannoma is typically a well-defined lesion which demonstrates hypointensity on T1WI, hyperintensity on T2WI, and intense postcontrast enhancement [11]. Furthermore, helpful radiological MRI signs include split fat sign (thin peripheral rim of fat on T1WI), target sign (central low signal within the lesion), and a

fascicular sign (multiple internal small ring-like structures), as visualized in our presented case [12]. Plexiform neurofibromas demonstrate enhancing infiltrating masses with a T2 hyperintense swirled appearance on MRI [13]. Although rare, malignant aggressive neoplasms such as rhabdomyosarcoma and alveolar soft part sarcoma are enhancing tumors with indistinct margins and lymph node involvement. Conclusive diagnosis is obtained via biopsy [14–16].

Congenital lesions can be subcategorized into vascular and nonvascular categories (Fig. 6). Lymphatic malformations are generally nonenhancing lesions with high T2 signal that can be either unilocular or multilocular with fluid-fluid levels [17]. Venous malformations usually enhance and are visualized as a lobulated soft tissue lesion with phleboliths. Venolymphatic malformations share characteristics of both

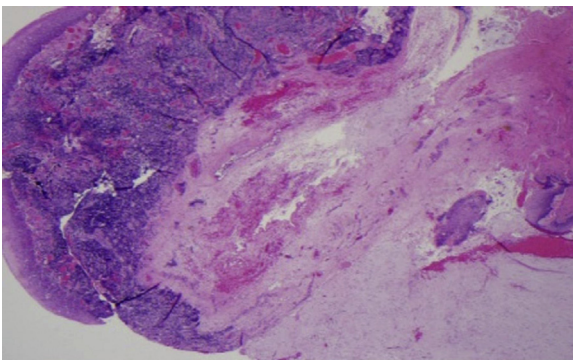


Fig. 4 – Low power microscopic image showing squamous epithelium of base of tongue, with underlying lymphocytic infiltrate and somewhat well-defined hypocellular lesion.

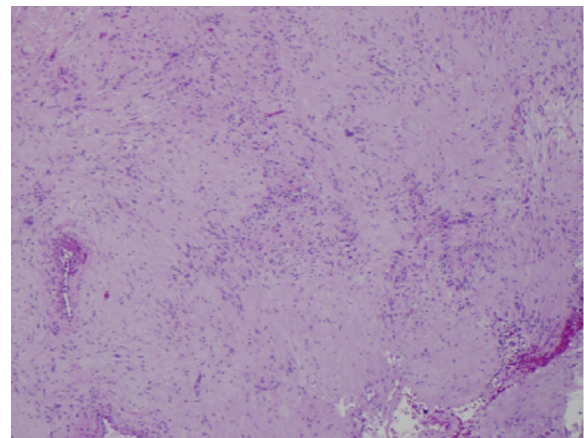


Fig. 5 – The lesion is composed of spindled cells with hypocellular and hypercellular areas. There are focal nuclear-palisading areas (verocay bodies).

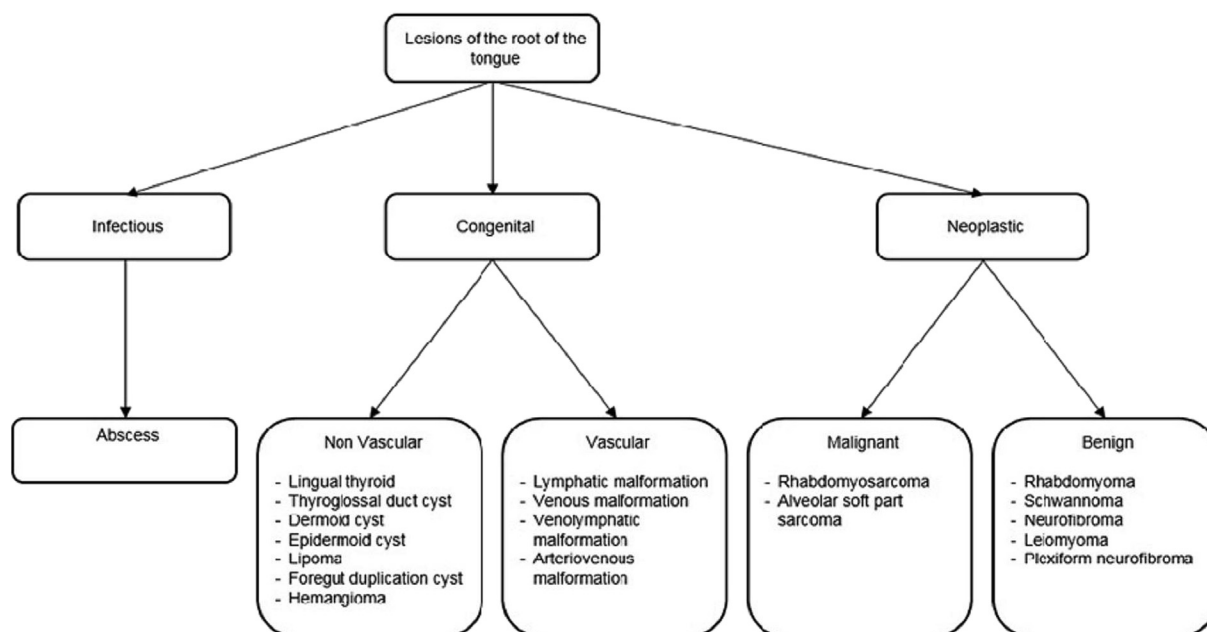


Fig. 6 – Systematic imaging approach for a tongue base lesion.

lymphatic and venous malformations. Arteriovenous malformations are high-flow vascular lesions, which are typically characterized by an arterial feeding vessel and a nidus [17].

Congenital nonvascular lesions are usually cystic in appearance with a few exceptions. A lingual thyroid is hyperdense on nonenhanced CT due to the high iodine content within the thyroid tissue. A thyroglossal duct cyst is described as a midline suprahyoid cystic neck lesion on CT and MRI [17]. Additional nonvascular cystic structures include dermoid and epidermoid cysts. Both these lesions have high T2 signal; however, epidermoid cysts will show restricted diffusion on diffusion weighted imaging, which is pathognomic. Dermoid cysts do not show restricted diffusion and are usually comprised of fat that coalesces into globules, creating a typical “sack of marbles” appearance. According to Kong et al, only 21 duplication cysts of the oral cavity have been reported [18]. A foregut duplication cyst is only diagnosed after resection with the cystic lesion showing intestinal epithelium. A lipoma is a well-encapsulated lesion with internal fat signal. Finally, lingual hemangiomas show intense enhancement on CT and MRI modalities, with prominent vasculature.

Surgical excision is the primary treatment of tongue schwannomas, with transoral route being the most commonly used approach [19].

Conclusion

A systematic approach toward a posterior tongue mass, that divides it into neoplastic, congenital, or infectious categories, can be helpful. Neoplasms arising from the base of tongue,

such as schwannoma, are rare and should be considered in an appropriate clinical context.

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