

Case Report

Novel surgical technique for management of tongue schwannoma: case report

Neha Jain*, Shama Shishodia, Ruchima Dham, Suparna Roy, Sachin Goel

Department of Otorhinolaryngology, Chacha Nehru Bal Chikitsalaya, Geeta Colony, New Delhi, India

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*Correspondence:

Dr. Neha Jain,

E-mail: nehaajmera84@gmail.com

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ABSTRACT

Schwannomas are rare, solitary, slow growing, smooth surfaced and well encapsulated tumors. Schwannomas of head and neck region account for 25-40% of all the cases. Approximately 1%–12% of schwannomas occur intraorally, the tongue being the most common site. Complete surgical excision is the treatment of choice. In this article, we describe a case of tongue schwannoma in a child, along with diagnostic and treatment options of tongue lesions. The tongue mass was completely excised via trans-oral approach using coblation method. The patient followed up for 1 year; he had an uneventful recovery and no recurrence.

Keywords: Schwannoma, Coblation, Tongue

INTRODUCTION

Schwannomas or neurilemmomas are benign tumors that arise from Schwann cells of peripheral nerve sheath.¹ It is a rare, solitary, slow growing, smooth surfaced and well encapsulated tumor.¹ Most common age groups affected are 30-60 years and is rare in pediatric population with no predilection for race or gender.^{2,3} Extracranially, about 25–40% of all schwannomas are seen in the soft tissues of the head and neck.² Approximately 1%–12% of schwannomas occur intraorally, the tongue being the most common site followed by palate, floor of mouth, buccal mucosa and mandible.^{2,3} Etiology of schwannoma remains unknown however; spontaneous growth, external injury, chronic irritation and radiation exposure are some of the factors that have been attributed to the etiopathogenesis of schwannoma.⁴

Schwannomas are generally asymptomatic but sometimes they may present with pain, paresthesia, dysphagia and neurological deficits, depending on their size and location.² Differential diagnosis of intraoral mass

includes neuromas, hemangiomas, neurofibromas, fibromas or adenomas, granular cell myoblastoma, neuroepitheliomas.² Malignant transformation and recurrence following this treatment are rare.⁵ Complete excision of schwannomas lowers the risk of tumor recurrence.⁶

CASE REPORT

We present a rare case of an 11-year-old boy with a solitary painless swelling on the right side of the tongue from seven months. There was no associated history of pain, discomfort, altered taste sensation, sensory abnormalities, dysphonia & dyspnea. There was no previous history of trauma, radiation exposure and chronic irritation. Clinical examination revealed a single, smooth, pinkish colored spherical swelling on right antero-lateral aspect of tongue, approximately 2 cm × 2 cm in size, which was non-tender, non-fluctuant and firm in consistency. Dentition, oropharynx and rest of the oral cavity were normal on clinical examination. There was no lymph node enlargement in the neck. Remaining

physical examination was not significant. Initial diagnosis of lympho-vascular swelling of tongue was made on clinical examination. To formulate a provisional diagnosis MRI with contrast (Figure 1) was done which showed the evidence of a well-defined, space occupying lesion of 20×17×17 mm in right side of anterior tongue appearing hypointense on T1 and hyperintense on IR sequence reaching up to midline suggestive of benign soft tissue lesion (likely of vascular origin).

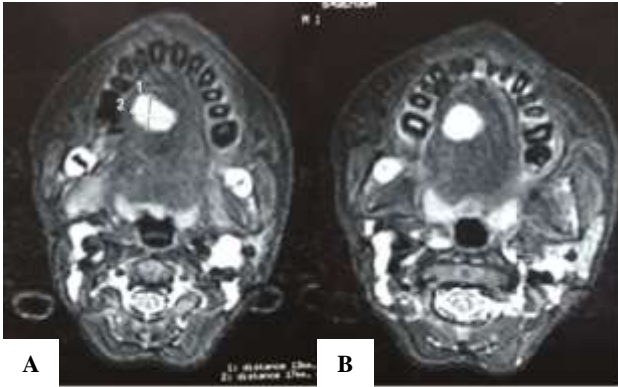


Figure 1: MRI of tongue schwannoma. (A) well defined, space occupying lesion of 20x17x17 mm in right side of anterior tongue; (B) mass is hyperintense on IR sequence reaching up to midline.



Figure 2: Intra-operative images of tongue schwannoma. (A) mass over right anterolateral side of tongue; (B) submucosal, encapsulated mass; (C): excised tumour 2x1 cm in size, well defined smooth margins

Following imaging, provisional diagnosis of benign vascular mass was made and intralesional bleomycin (0.25 mg/kg body weight) was administered to the patient. The size of the swelling initially increased but later on it decreased in size, became firm and pale in color (Figure 2A).

The patient underwent excisional biopsy by the trans-oral approach under general anesthesia. The lesion was submucosal, well-capsulated and had a good cleavage plane (Figure 2B). With the help of Coblator wand, swelling was excised in toto and sent for histo-pathological evaluation. The excised mass measured 2 cm×1 cm, was capsulated and had smooth surface (Figure 2C). The postoperative course was uneventful. Child was followed up regularly for 1 year and showed no evidence of recurrence.

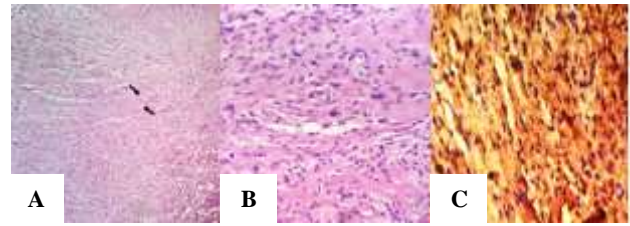


Figure 3: Photomicrograph of tongue schwannoma. (A) Schwannoma of the tongue under low power showing spindle shaped cells and verocay bodies (arrow) (haematoxylin-eosin stain, original magnification x20); (B) schwannoma of the tongue under high power (haematoxylin-eosin stain, original magnification x40); (C) tumor cells are immunopositive for S-100 (x 40, IHC S-100).

Pathology (Figure 3) demonstrated lymphocytic infiltrate with well-defined hypocellular and hypercellular areas with nuclear palisading around fibrillary process (Verocay bodies). Blood vessels with thick hyalinized walls and focal thrombus formation were seen. Immunohistochemistry showed strong S-100 positivity in the spindle tumor cells. Thus, a diagnosis of schwannoma was established. The patient is under regular follow up and has not shown any recurrence in the period of 1 year.

Although the tongue is one of the most common tumor locations in the mouth, very few cases of lingual schwannoma have been reported in pediatric group in the English literature so far.

DISCUSSION

Nerve sheath tumors are of two types: neurofibroma and schwannoma. Neurofibromas are benign neoplasm derived from fibroblasts of the perineurium.⁷ Schwannomas originate from Schwann cells of nerve sheath and are also known as neurilemmomas, neurinoma and Schwann cell tumor.^{7,8} Schwannomas are solitary, slow growing and encapsulated benign tumors.¹ It can originate in any peripheral, autonomic, or cranial nerve, except the olfactory and optic nerves, as these nerves lack Schwann cells.⁸

Twenty-five to forty five percent of schwannomas have been reported to occur in the head and neck region, of which 90% are located at the level of cranial nerve VIII and only 1% in the oral cavity and oropharynx.^{3,8} Tongue is the most common site in intra oral lesions followed by buccal mucosa, floor of mouth, palate, lips and gingiva.^{3,7}

Etiopathogenesis

Etiology of schwannoma remains unknown.⁶ The possible etiologies of the tongue schwannoma are spontaneous growth, external injury, chronic irritation, or exposure to radiation; but the certain cause of the tongue schwannoma is unknown.⁴

Diagnosis

Clinical presentation: Lingual schwannomas often present as a painless lump¹ and they are usually noticed due to discomfort caused to the patient such as difficulty in swallowing, chewing, and phonation.² Schwannomas of the tongue typically present in the third decade of life, display no gender predilection.^{2,3}

Lump in the oral cavity could be due to reactive or neoplastic lesions like lipoma, neurofibroma, hemangioma, retention cyst, lymphangioma, lingual thyroid, leiomyoma and benign salivary gland tumors.^{2,6} Therefore, a detailed history, local and systemic examination, histopathological examination and radiological assessment are essential tools for making a definitive diagnosis.

Imaging

Imaging tools like ultrasonography, CT and MRI are helpful for the estimation of tumor margins, site, extent, lesion composition and its relation to the adjacent surrounding structures.⁹ The imaging modality of choice for tongue schwannoma is MRI.⁹

On MRI, tongue schwannoma appears as a well circumscribed nodule which is homogeneously isointense to muscle on T1- weighted images and homogeneously hyperintense on T2- weighted images. Contrast enhanced MRI and CT shows intense heterogenous enhancement. Tongue schwannomas usually appear smooth, well demarcated and do not invade the surrounding structures.⁹ Schwannomas are very commonly misdiagnosed as hemangiomas due to similar imaging picture. Radiological findings of various lingual lesions are discussed in Table 1.

Table 1: Radiological pattern of various lingual benign lesions.

Lingual benign lesions	Radiological patterns on MRI	Radiological patterns on CT
Tongue Schwannoma	1.T1W1images-homogeneously isointense to muscles 2.T2W1-homogenous hyperintense. 3Contrast MRI- intense heterogenous enhancement	Heterogenous enhancement
Hemangioma	T1-hypointense T2-hyperintense	Intense enhancement
Lymphangiomas	Fluid fluid levels seen. T1: variable depending on protein content T2: high signal	Non enhancing lesions
Neurofibromas	Enhancing infiltrating mass T2 hyperintense swirled appearance	
Venous malformations	Enhancing lesions-lobulated soft tissue mass with phleboliths	
Lingual thyroid	T1; iso to hyperintense to muscle T2; variable (iso/hypo/hyper) Homogenous enhancement with Gd contrast	Hyperdense on CT due to high iodine content
Lipoma	Well encapsulated lesion with internal fat signal	

Cytology

The preoperative FNAC rarely helps in the diagnosis of neural tumor and might be inconclusive as blood usually contaminates the slide but cytology may help in differentiating benign and malignant lesions of soft tissue. Hence definitive diagnosis of schwannoma is often made postoperatively and requires histopathological examination of the excised mass.

Histologically, schwannomas are well circumscribed and encapsulated mass which may show cystic degeneration. Microscopically it consists of two types of pattern: Antoni A and Antoni B. Antoni A pattern consist of highly cellular and elongated Schwann cells with palisading nuclei whereas Antoni B consist of disorganized, elongated Schwann cells with less dense myxoid morphology. Verocay bodies are the regions

found in between the palisading cells which are devoid of nuclei.⁷ Schwannomas usually showed immunostaining for S-100, which may help to distinguish peripheral nerve sheath neoplasm from other soft tissue tumors.^{1,3}

In our case we did S-100 immunohistochemical staining to confirm the diagnosis.

Treatment

Schwannomas are not responsive to radiotherapy.¹⁰ These benign masses are well capsulated¹ and hence complete surgical excision is the treatment of choice.²

Transoral approach is most commonly used for tongue schwannomas. Besides conventional method, other tools like CO2 laser, coblation, bipolar cautery may be used for intraoral excision.

However trans-cervical approach (trans-hyoid or submandibular) is preferred for oral masses which are inaccessible via mouth, due to large size or posterior location.¹¹ Recurrence has been not reported if complete excision is performed.²

Follow up

Post-operative prognosis is good. The length of follow up remains debatable, long term follow up is not necessary if clearance margins are tumor free, but in doubtful cases, the patient should be annually reviewed.⁸

Schwannomas are usually solitary lesions however multiple lesions are seen in Von Recklinghausen's disease and schwannomatosis.¹ Therefore, after confirming the histological diagnosis, a thorough examination of the patient should be done to rule out von Recklinghausen's syndrome and Neurofibromatosis -2.¹² Malignant transformation is rare with incidence of 8% to 13.9%, and mostly seen in patients of von Recklinghausen's syndrome.^{1,5,6,12}

In the present case, tongue schwannoma presented as a slow growing, well circumscribed mass without any particular features to distinguish it from other benign soft tissue lesions and on imaging it was provisionally diagnosed as a vascular lesion. Among approaches for removal of lingual schwannoma, transoral approach was suitable for our patient. No case has been reported in literature in which tongue schwannoma has been excised with the help of coblation after injection of bleomycin. Therefore, we are adding our experience of treating schwannoma with a different technique and yielding satisfactory results.

CONCLUSION

Lingual schwannoma is a rare entity and is generally misdiagnosed as it is indistinguishable from other benign tongue lesions. Hence, a proper approach to diagnose it correctly should be adopted. We are reporting an alternative technique (coblation) for a rare tumor like schwannoma at a rare site (intraoral) with excellent postoperative result, minimal pain, minimal bleeding and no recurrence in the follow up period of 1 year.

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