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Recurrent schwannoma of the tongue in a pediatric patient—report of a rare case with an updated review of literature

Anup Singh*, Vaisakh Kuzhikkali and Arvind Kumar Kairo

Abstract

Background: Head and neck is a relatively common site of occurrence for the peripheral nerve sheath tumors, and majority of these tumors are seen involving neck, involving vagus nerve, and cervical sympathetic chain. Schwannomas involving mobile tongue are rarely encountered, especially in the pediatric population

Case presentation: We present a case of recurrent tongue schwannoma in a 13-year-old female successfully managed with transoral excision. At a follow-up of 3 years, no recurrence is observed.

Conclusion: Surgical excision is the recommended modality of treatment for lingual schwannomas, and when excised adequately, recurrences are not expected. A clear margin of surrounding normal tissue should be aimed for to avoid possible recurrence.

Keywords: Tongue, Child, Schwann cell, Schwannoma, Soft tissue neoplasms, Recurrence

Background

Schwannomas are benign tumors arising from the nerve sheath cells (Schwann cells), cranial nerves/autonomic nerves, and peripheral motor-sensory nerves. Head and neck involvement is common, constituting ~ 25–45% of all the cases and the commonest site of involvement in head and neck is cerebello-pontine angle/8th nerve complex [1]. Of the extracranial/non-vestibular cases, the commonest location is parapharyngeal space (vagus nerve and cervical sympathetic trunk). Oral cavity schwannomas constitute only ~ 1% of head and neck schwannomas and as such are quite low down in the list of differential diagnosis of oral tumors. The age group usually affected with pathology is 3rd to 6th decade [2]. We discuss a pediatric patient with recurrent schwannoma of tongue managed with surgical excision and review pertinent literature regarding the schwannomas of tongue in the pediatric population.

Case presentation

A 13-year-old female patient presented to us with a 6-month history of gradually enlarging mass lesion involving tongue. The patient did not report pain, bleeding, chewing difficulty, dysphagia, breathing difficulty, snoring, neck swelling, or any systemic symptoms. The patient had a history of excisional biopsy under general anesthesia for swelling at the same site 3 years back at another center and the post-operative histopathology was suggestive of benign nerve sheath tumor (schwannoma). On examination, the lesion was ovoid, smooth surfaced, mucosa covered, yellowish-pink, ~ 2–1.5 cm in size, non-tender, firm with a broad base located at the junction of anterior two-third and posterior one-third of the left side of the tongue (Fig. 1a). Tongue mobility was normal. The rest of the systemic examination did not reveal any abnormality. With a possibility of a recurrent lesion, a contrast-enhanced MRI was obtained, which showed a 1.6 mm × 1.4 mm × 1.4 mm well-circumscribed lesion, hypointense on T1, homogenously hyperintense on T2, and with avid homogenous enhancement on contrast administration (Fig. 2). A wedge

* Correspondence: anoop.aiims1@gmail.com
Department of Otolaryngology and Head & Neck Surgery, All India Institute of Medical Sciences, New Delhi 110029, India

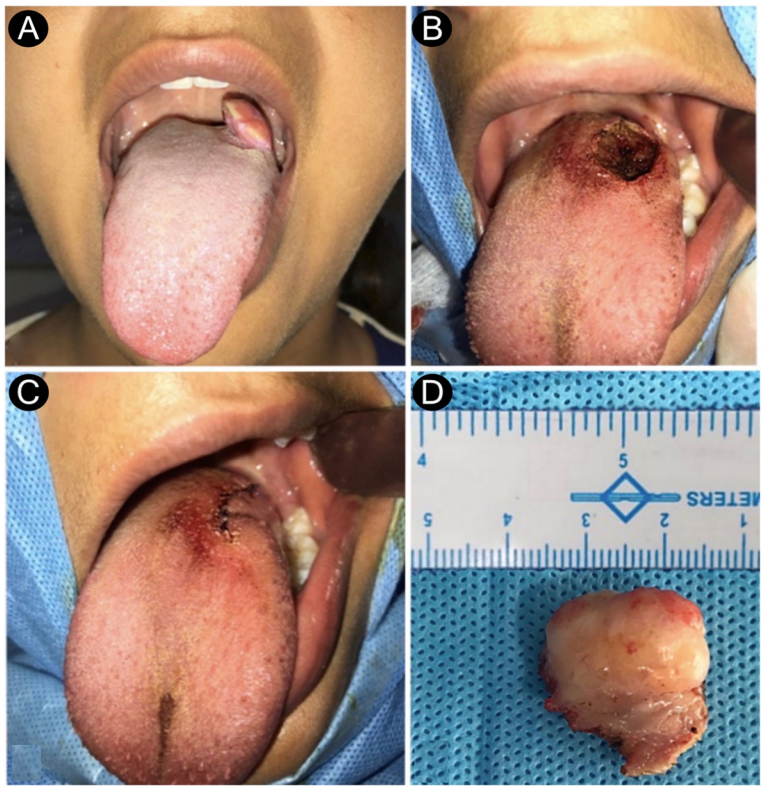

Fig. 1 **a** Showing the tumor involving the left posterior tongue with a wide base. **b** Intraoperative picture of the wound bed after complete excision. **c** Wound closed primarily, and **d** The specimen after complete excision with adequate margins

biopsy taken under local anesthesia from the lesion showed features suggestive of benign nerve sheath tumor. The patient and the relatives were explained about the need for surgical intervention and opted for the excision of the tumor under sedation (ketamine 1 mg/kg along with intravenous ondansetron 4 mg stat dose). With a good mouth opening and mobile tongue, the tumor could be adequately exposed transorally and wide excision with a cuff of surrounding soft tissue (~ 0.5–1 cm) was carried out (Fig. 1b, d). The nerve of origin was not discernible intraoperatively. The defect was primarily closed (Fig. 1c) and the post-operative period was uneventful. Post-op histopathology confirmed the diagnosis of schwannoma (Fig. 3) with clear margins. The post-operative period was uneventful. During a follow-up of 3 years, the surgical site is well healed without any evidence of recurrence.

Discussion

Intraoral schwannomas tend to involve the tongue followed by the floor of mouth, palate, buccal mucosa, gingiva, lip, and vestibule [3]. Involvement of tongue in the pediatric age group is a rare occurrence. A PubMed English language literature review of lingual schwannomas in pediatric patients, dated 28th February 2021, including the case reports and series with clearly defined individual patient characteristics revealed a total of 34 cases of patients in age group ≤ 18 years (Table 1) [3–16]. The first documented case of pediatric lingual schwannoma was by Craig [4] in 1964 in an 8-year-old female patient. The youngest patients (7 years old) were reported by Cinar [9] in 2004 and Enoz [10] in 2006 and recently by Thompson et al. [16]. There were 16 male and 18 female patients. The most common symptom, present in nearly three-fourth of the patients, was a painless lump. Other symptoms at the time of presentation included paresthesia, ulceration, bleed, and mechanical obstructive symptoms in the form of snoring, difficulty in mastication, swallowing, breathing, and phonation in varying combinations. The average size at the time of presentation was 19.6 mm (standard deviation: 10.5; range 5 to 50 mm) and the tumor was present in anterior/ anterolateral part of the tongue in 58.8% of the cases. An uneventful and complete excision of the tumor could be carried out in all the patients transorally without resorting to more invasive and

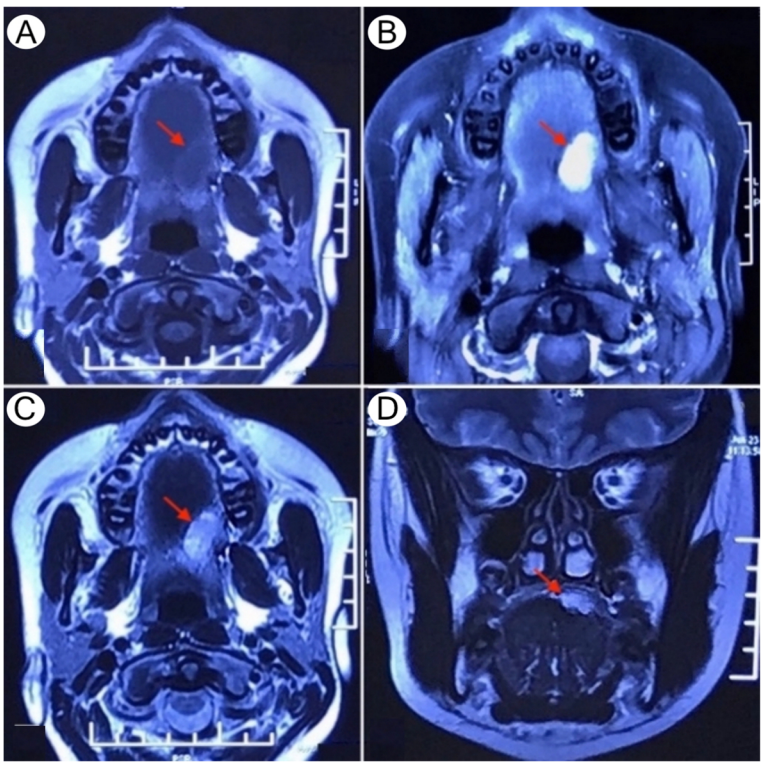

Fig. 2 CE-MRI face (coronal cuts **a**–**c** axial cuts **d**) showing the tumor involving left side posterior tongue (red arrow). **a** The tumor is isointense on T1, **b** brightly and uniformly enhancing on contrast administration, and **c** heterogeneously hyperintense on T2

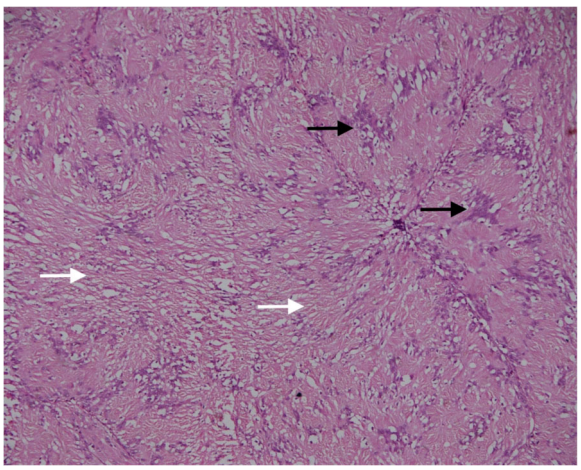

Fig. 3 Post-operative histopathology showing 'Antony A' (black arrows) and 'Antony B' (white arrows) areas characteristic of Schwannoma. (hematoxylin and eosin stain; original magnification × 10)

morbid approaches. Follow-up was available in 70.6% of the cases, with a median duration of follow-up being 13.5 months (range 1 to 136 months), without any reported recurrences.

It is interesting to note that despite having nerve sheath origin, these tumors are seldom painful. In tongue, these tumors may arise from hypoglossal, lingual, or glossopharyngeal nerve but the nerve of involvement may be difficult to discern. It is easy to misinterpret these tumors with other soft tissue tumors like granular cell tumors, neurofibromas, mucoceles, salivary gland tumors, and leiomyomas on the basis of clinical appearance.

The gold standard of diagnosis remains histopathology. The light microscopic findings are distinctive enough to confer a diagnosis of schwannoma. If needed, the diagnosis can be reinforced with immunohistochemical staining with S-100 protein, which is seen to be present diffusely and uniformly in the tumor tissue [12].

Surgical excision remains the treatment of choice given the radioresistant nature of these tumors and relatively straightforward transoral access in cases of tumors involving the mobile tongue. In the current review, all the tongue tumors could be accessed

Table 1 Pediatric lingual schwannomas—review of literature

SN [ref]	Author	Year	Age (years)	Sex	Symptom	Size[a] (mm)	Site	Surgical approach	FU (months)
1 [4]	Craig	1964	8	F	Painless mass	30	Posterior	Transoral	NA
2 [5]	Uj	1967	13	F	Painless mass	NA	NA	Transoral	NA
3 [6]	Barbosa et al.	1984	12	M	Painless mass	5	NA	Transoral	NA
4 [7][b]	Piatelli et al.	1984	18	F	Painless mass	NA	Anterior	Transoral	NA
5 [8]	Akimoto et al.	1987	14	M	Painless mass	10	Anterior	Transoral	NA
6 [8]	Siar et al.	1988	17	F	Painless mass	30	Posterior	Transoral	12
7 [8]	Siar et al.	1988	13	F	Painless mass	44	NA	Transoral	8
8 [3]	Pfeifle et al.	2001	18	M	Painless mass	20	Anterior	Transoral	NA
9 [8]	Bassichis et al.	2004	9	M	Snoring, breathing difficulty	23	Posterior	Transoral	60
10 [9]	Cinar et al.	2004	7	M	Painless mass	10	Anterior	Transoral	NA
11 [8]	Nakasato et al.	2005	9	F	Bleeding/ulceration	20	Postero-lateral	Transoral	17
12 [8]	Vafiadis et al.	2005	18	M	Painless mass	31	Anterior	Transoral	36
13 [10]	Enoz et al.	2006	7	M	Dysphagia/pain	25	Anterior	Transoral	60
14 [8]	Hsu et al.	2006	9	M	Painless mass	12	Anterior	Transoral	56
15 [8]	Hsu et al.	2006	15	F	Painless mass	12	Anterior	Transoral	136
16 [8]	Hsu et al.	2006	12	F	Painless mass	16	Anterior	Transoral	13
17 [8]	Pereira et al.	2008	12	M	Painless mass	15	Postero-lateral	Transoral	12
18 [8]	Gupta et al.	2009	18	F	Painless mass	10	Anterior	Transoral	NA
19 [8]	Karaca et al.	2010	13	F	Dysphagia	20	Postero-lateral	Transoral	12
20 [8]	Naidu et al.	2010	12	M	Paresthesia/bleed/ulceration	20	Antero-lateral	Transoral	3
21 [11]	Husain et al.	2011	10	F	Difficulty in mastication and phonation	50	Antero-lateral	Transoral	12
22 [8]	Lukšić et al.	2011	10	M	Painless mass	15	Postero-lateral	Transoral	60
23 [11]	Manna et al.	2012	15	M	Difficulty in mastication/ swallowing	17	Posterior	Transoral	6
24 [11]	Bhola et al.	2014	14	F	Painless mass	15	Antero-lateral	Transoral	12
25 [8]	Moreno-Garcia et al.	2014	13	F	Painless mass	20	Anterior	Transoral	12
26 [12]	Franco et al.	2017	14	M	NA	15	NA	NA	NA
27 [13]	Fan et al.	2017	16	M	Painless mass	25	Posterior(tongue base)	Transoral	22
28 [14][c]	Amer et al.	2018	13	F	Painless mass with chewing difficulty	20	Anterior	Transoral	NA
29 [15]	Ahmad et al.	2020	14	F	Dysphagia	30	Posterior	Transoral	24
30 [16]	Thompson et al.	2020	12	F	Lingual mass	7	Anterior	Transoral	48
			12	F	Lingual mass	15	Anterior	Transoral	9
			18	M	Lingual mass	7	Anterior	Transoral	70
			17	F	Lingual mass	31	Anterior	Transoral	14
			17	M	Lingual mass	7	Anterior	Transoral	1
31.	Present case		13	F	Recurrent painless mass	20	Posterior	Transoral	36

SN serial number, *M* male, *F* female, *mm* millimeter, *NA* not available, *FU* follow-up
[a]Largest dimension of the tumor
[b]Case of malignant schwannoma)
[c]The patient with neurofibromatosis-2

transorally. However, in cases with large tumors especially involving the posterior tongue/base of tongue or in cases with restricted mouth opening, an external approach (submandibular, lip split, mandibulotomy, or anterior midline glossotomy approach) may need to be resorted to for adequate surgical exposure. A complete surgical excision leaves no room for recurrence.

Conclusion

Schwannomas involving tongue are rare neurogenic tumors, especially in the pediatric age group. A complete excision with sufficient margin leaves no room for recurrence. However, enucleation of the lesion or excisional biopsy in the tongue can result in recurrence of the lesion predisposing the patient to the morbidity of repeat surgical procedure.

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Authors' contributions
Study concept and design: AS, AKK, and VK. Acquisition, analysis, or interpretation of data: AS, AKK, and VK. Drafting of the manuscript: AS and AKK. Critical revision of the manuscript for important intellectual content: AS, AKK, and VK. Administrative, technical, or material support: AS, AKK, and VK. All authors have read and approved the manuscript.

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Declarations

Ethics approval and consent to participate
Ethics approval is not applicable. Consent to participate was obtained from the parents of the patient.

Consent for publication
Written and informed consent to publish this information was obtained from the parent of the study participant.

Competing interests
The authors declare that they have no competing interests.

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