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Pediatric Lingual Schwannoma: A Rare Case with a Clear Diagnostic and Therapeutic Approach

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Abstract

Lingual schwannomas are rare benign peripheral nerve sheath tumors and are particularly uncommon in pediatric patients, which may delay diagnosis. We report the case of a 14-year-old girl presenting with a long-standing, bilobulated, firm submucosal nodule on the right lateral tongue, with recent accelerated growth and functional impairment. Contrast-enhanced cervicofacial computed tomography revealed a well-circumscribed lesion measuring 18 × 8 mm, without deep extension or suspicious cervical lymphadenopathy, consistent with localized nodular disease. Complete transoral excision was performed. Histopathological examination confirmed schwannoma, and immunohistochemistry demonstrated diffuse S100 and CD34 positivity, EMA negativity, and preserved nuclear INI1 expression, supporting the diagnosis and excluding perineurioma or epithelioid malignant peripheral nerve sheath tumor. Postoperative recovery was uneventful, with no evidence of recurrence at short-term 1 month follow-up. In adolescents presenting with persistent tongue masses, schwannoma should be considered in the differential diagnosis. Imaging is essential for surgical planning, while definitive diagnosis is histopathological; complete excision is typically curative with excellent functional outcomes.

Highlights

- Persistent tongue nodule in an adolescent—consider schwannoma early.
- CT delineated a well-circumscribed, enhancing, localized lesion aiding transoral planning.
- S100+, CD34+, EMA, INI1 preserved supports schwannoma over mimics.
- Complete transoral excision was curative with functional preservation.

1. Introduction

A schwannoma is a benign, encapsulated tumor arising exclusively from Schwann cells. Although head and neck involvement are relatively common, intraoral localization is uncommon (1%), most frequently affecting the tongue. Lingual schwannomas themselves remain infrequent, and pediatric presentations are particularly rare, which may delay timely diagnosis [1, 2].

Symptoms and signs of peripheral nerve tumors are caused by direct nerve invasion, involvement of surrounding tissues, or mass effect. Typical presentation is slow-growing and painless [3, 4].

When a mass can be clearly related to a nerve on ultrasound, CT, or MRI, schwannoma is suggested. In addition, a heterogeneous internal architecture with cystic degeneration is more frequently associated with schwannomas. On CT, schwannomas usually appear hypodense relative to muscle and demonstrate contrast enhancement [5].

Schwannomas show a biphasic architecture of Antoni A (dense) and B (loose) histologic patterns, as well as nuclear palisading (Verocay bodies) and a fibrous capsule containing the parent nerve. Neoplastic Schwann cells typically show spindle-shaped nuclei. In addition to these characteristic patterns, diagnosis is aided by immunohistochemical markers, such as S-100, which support the Schwann cell nature of these tumors [6–8].

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The standard management of lingual schwannoma is complete surgical removal, most often achieved through a transoral approach. For lesions arising at the tongue base, alternative access routes such as transhyoid or submandibular approaches, as well as CO laser excision, may be considered. Recurrence has not been documented following total excision [9].

2. Case Presentation

2.1. Clinical Presentation

A previously healthy 14-year-old girl presented with a bilobulated, hard and elastic submucosal nodule (2 cm) on the right lateral tongue, mildly tender, with functional impact on feeding, speaking and faster recent growth; no adenopathy.

2.2. Diagnostic Assessment

Contrast-enhanced computed tomography (CT) showed a well-defined, oblong enhancing lesion (18 × 8 mm), without deep extension to the tongue root or floor of mouth; no suspicious lymph nodes (Figure 1).

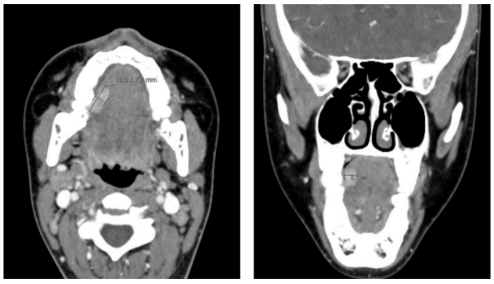

Figure 1. Axial contrast-enhanced CT showing a well-circumscribed, enhancing, oblong lesion (18 × 8 mm) in the right lateral tongue, without deep extension or lymphadenopathy.

2.3. Surgical Procedure

A complete transoral excision under general anesthesia was performed. The mass was submucosal and once a mucosal flap was raised, the tumor was readily shelled out using blunt dissection and a yellowish, encapsulated, firm mass corresponding to imaging dimensions was enucleated (Figure 2).

2.4. Histopathological Examination

Histopathological examination revealed a mesenchymal neoplasm compatible with schwannoma. Most schwannomas are encapsulated by a thick or thin, fibrous capsule and show a variable admixture of high and low cellularity zones – Cellular Antoni A zones and loose edematous or myxoid Antoni B zones. Another often conspicuous and characteristic finding in schwannoma is the presence of vascular channels with hyalinized walls and the presence of Verocay bodies.

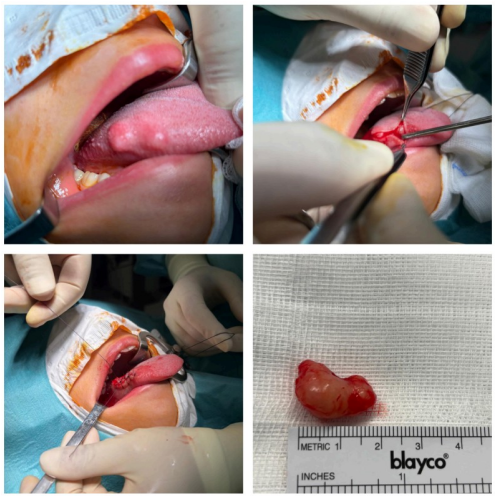

Figure 2. Intraoperative view of the encapsulated, yellowish mass before and after enucleation.

Immunohistochemistry showed diffuse S100 and CD34 positivity, EMA negativity, and preserved nuclear INI1 expression—findings that support the diagnosis of schwannoma and help to exclude perineurioma or epithelioid malignant peripheral nerve sheath tumor (MPNST). Surgical margins were free of tumor (Figure 3).

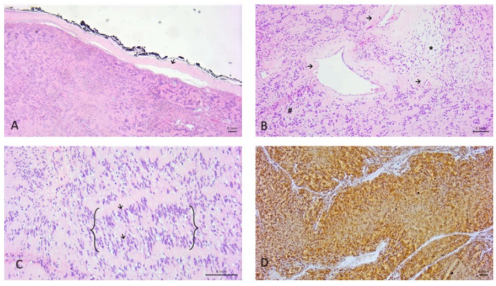

Figure 3. Histopathological features of the schwannoma. A - Fibrous capsule surrounding the tumor; B - Antoni A (dense) and B (loose) zones; C - Vascular channels with hyalinized walls and Verocay bodies; D - Diffuse S100 positivity.

2.5. Post-operative Course and Follow-Up

At early follow-up, 1 month later, the patient showed an uneventful recovery with no clinical recurrence.

3. Discussion

Lingual schwannoma is rare in adolescents yet well documented; tongue predilection and slow growth are typical [1–3]. The differential diagnosis includes fibroma, lipoma, minor salivary gland tumours, vascular lesions, and other neurogenic masses [2, 3]. In the present case, the recent accelerated growth and functional impairment prompted further investigation.

CT/MRI help define limits and plan a conservative

transoral approach and in our patient, CT revealed a well-circumscribed, enhancing lesion of the lateral tongue, consistent with these typical imaging features and supporting the diagnosis of schwannoma [5]. Therefore, imaging plays a crucial role in narrowing the differential diagnosis and guiding surgical planning.

Despite suggestive clinical and imaging features, definitive diagnosis is histopathological and immunohistochemical. Classic schwannomas demonstrate Antoni A and Antoni B areas, Verocay bodies, and encapsulation. Immunohistochemistry is essential in distinguishing schwannoma from histologic mimics. Diffuse S100 positivity confirms Schwann cell differentiation, while EMA negativity helps exclude perineurioma. Preservation of nuclear INI1 expression is particularly important in ruling out epithelioid malignant peripheral nerve sheath tumor, which typically shows INI1 loss [6–8]. CD34 positivity, as observed in this case, has been reported in cellular schwannomas and further supports the diagnosis [8].

Complete surgical excision remains the treatment of choice. For lesions of the mobile tongue, transoral excision is the most commonly reported and preferred approach, offering excellent exposure, functional preservation, and minimal morbidity [9]. In this case, the encapsulated nature of the tumor allowed straightforward enucleation with clear margins.

As described in the literature, recurrence following complete excision is exceedingly rare, and malignant transformation has not been documented in lingual schwannomas [9].

4. Conclusions

Lingual schwannoma, although rare in pediatric patients, should be included in the differential diagnosis of persistent tongue masses in adolescents. Imaging is essential for lesion characterization and surgical planning, but definitive diagnosis relies on histopathological and immunohistochemical confirmation. Complete transoral excision is curative in the vast majority of cases and is associated with excellent functional outcomes and minimal risk of recurrence. Early recognition allows timely management and avoids unnecessary morbidity.

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