



Case Report

Schwannoma of Tongue in A Child

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ABSTRACT

Schwannoma is a benign nerve sheath tumor composed of Schwann cells, which normally produce the insulating myelin sheath covering peripheral nerves. Here, we report a rare unsuspected case of 6 year old female child with schwannoma on the right side of the middle third of the dorsum of the tongue. This case highlights the clinicopathological features of schwannoma tongue and the importance of including schwannoma as one of the differential diagnosis among the lesions of tongue especially in children.

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Introduction

Schwannomas also known as neurilemmomas, neuromas, neurolemomas and Schwann cell tumors are one of the benign, slow growing, usually solitary and encapsulated neurogenic tumors. They originate from any peripheral, cranial (except the optic and the olfactory nerves), spinal and autonomic nerves that have nerve supporting Schwann cells.^[1] This entity was first described by Verocay in 1910^[2], later the term neurilemmoma was coined by Stout in 1935.^[3] About 25-45% of all the extracranial schwannomas have been reported in the head and neck region^[4] and the most common site is the lateral part of the neck (parapharyngeal space). Occasionally, they can occur in submandibular space, para-nasal sinuses, cheek and oral cavity.^[5] Intraoral schwannoma accounts for 1% of all the head and neck region tumors and its usual location is tongue, followed by buccal mucosa, floor of mouth, palate, lips, gingiva and mandible.^[6] The exact etiology of this tumor is still unknown. However, some etiological factors have been postulated like spontaneous growth, external injury, chronic irritation, or exposure to radiation.^[7] It usually presents between the third and sixth decades, with no predilection for gender or race.^[8]

We present here a rare unsuspected case of schwannoma in a child presenting with a lesion on the dorsum of the tongue.

Case Report

A 6 year old female reported to the outpatient department with a six month history of slowly enlarging lesion on the right side of the middle third of the dorsum of the tongue. She now had difficulty in speech, chewing and swallowing. Her past personal, birth and medical history were non-contributory. There were no constitutional symptoms such as loss of appetite or weight loss. Intraoral examination revealed 2 cms x 1.5 cms, firm, non-tender, well-defined, smooth, solitary lesion on the right side of the middle third of the dorsum of the tongue (Figure 1a). It did not bleed on touch. Oral hygiene was fair but there was mild halitosis. There was no neurological deficit and no cervical lymph nodes were palpable. The clinical impression was of a benign tumor of the tongue. Routine laboratory investigations were within normal limits. No radiological investigations or initial biopsy was done as the lesion was easily visible and palpable. Following this, fine needle aspiration cytology (FNAC) of the tongue swelling was performed using a 23 gauge needle. Both May-Grunwald-Giemsa (MGG) and Papanicolaou stained smears were examined. Cytology showed cellular smears with cohesive clusters and aggregates of benign spindle cells with wavy vesicular nuclei and ill-defined eosinophilic cytoplasm

embedded in the fibrillary myxoid matrix material. At places, nuclear palisading was also noted. No mitosis or pleomorphism was observed (Figure 1b). Based on these findings, a presumptive diagnosis of a benign spindle cell tumor, possibly schwannoma was made. The lesion was completely excised by intraoral approach and the defect was closed with vicryl. The patient was discharged the same day after an uneventful recovery.

On gross examination, the excised mass was well encapsulated measuring 2x1.5x1cm in size. The cut surface was grayish white with focal myxoid areas (Figure 1c). Histopathology revealed keratinized stratified squamous epithelium and underlying connective tissue showing a well demarcated lesion comprising of two growth patterns, namely Antoni A and Antoni B types (Figure 2a). The former was highly cellular and composed of elongated spindle like cells with abundant fibrillar cytoplasm, which in areas, formed nuclear palisades, namely Verocay bodies. Antoni B areas also showed these spindle cells which were more disorganized and were arranged in a less dense myxoid morphology (Figure 2b). There was no pleomorphism and mitosis. Immunohistochemistry (IHC) with S-100 protein revealed strong positivity, confirming the neural origin of this tumor. These features lead to a conclusive diagnosis of schwannoma of the tongue (Figure 2c). The patient was reviewed after a week, there was complete healing of the tongue. On monthly follow up for a period of one year, no recurrence or any other complains were recorded.

Discussion

Embryologically, Schwann cells arise from ectomesenchymal cells of the neural crest during the fourth week of development. They enhance nerve conductance by either forming a thin barrier around peripheral nerve fibers or a thick myelin sheath.^[9] Peripheral nerve tumors of the oral cavity include schwannoma, neurofibroma, traumatic neuroma, the palisaded encapsulated neuroma and neurothekeoma. Intraoral schwannomas are quite rare and may occur in all ages, with predominance in the third and sixth decade of life and very rarely below 10 years of age.^[10] Its most common site is the base of the tongue and tip of the tongue is the least affected part.^[11] Thus, the present case is rare to occur in a 6 year old child with lesion on the middle third of the dorsum of the tongue.

The usual clinical presentation of schwannoma tongue is a gradually growing painless mass. The surface is mostly smooth and rarely exophytic or fungating. Ulceration of the overlying mucosa is uncommon and is generally the result of trauma.^[12] It usually manifests as a solitary lesion. However, when multiple, it can be associated with neurofibromatosis. The differentiation between

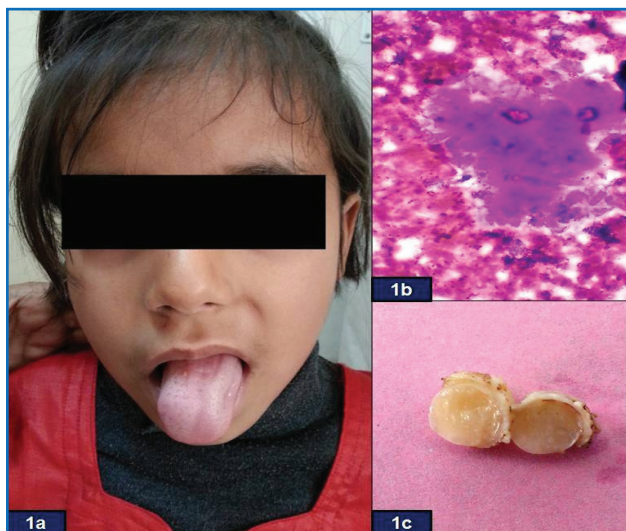


Fig. 1: (a) - Tongue swelling on clinical inspection., (b) - Cytological smears showing benign spindle cells with peripheral palisading and fibrillary background (MGG, x 40)., (c) - Gross specimen (cut section) of a well circumscribed tongue schwannoma.

schwannoma and neurofibroma is important because an apparently “solitary” neurofibroma may be a manifestation of neurofibromatosis.^[13] Approximately 15% of patients with neurofibromatosis will have malignant transformation in one or more lesion, which is in marked contrast to the typical behavior of a schwannoma.^[14]

Tongue schwannomas are generally not suspected clinically with considerable accuracy, therefore radiographic, cytological and histopathological investigations play an integral role in its diagnosis. The role of FNAC in the intraoral lesions is still debatable and has variable outcomes.^[15] Cytomorphological features of FNAC smears of schwannomas have been described in literature. These include the presence of spindle cells with fibrillary cytoplasm, fishhook nuclei, Verocay bodies, and pseudoinclusions in the nuclei. However, there are several pitfalls on cytology, firstly, the inadequate cell yields/paucicellularity and secondly, the other varied cytomorphological appearances (nuclear pleomorphism, stromal edema, fibrosis), leading to misdiagnosis and confusion with other types of malignant soft tissue tumors.^[16] Though in our case, FNAC was quite helpful. Magnetic Resonant Imaging (MRI) is the best choice in detailing the tumor, its capsule, origin and its extent.^[17] However, radiology too has its own limitations i.e. sometimes it cannot differentiate schwannomas from other encapsulated benign tumors. Therefore, the definite diagnosis of schwannoma tongue is based only on histopathological examination.

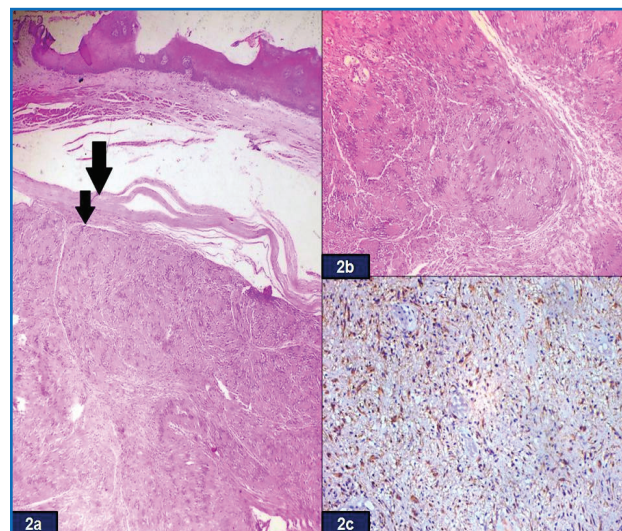


Fig. 2: (a) - Histopathology of tongue schwannoma showing keratinized stratified squamous epithelium and an underlying well defined encapsulated tumor (H and E, x10)., (b) - Tumor exhibiting Antoni A (Verocay bodies) and Antoni B areas (H and E, x20)., (c) - S-100 positivity of the tumor cells (IHC, x20).

Histopathologically, the schwannomas have distinctive alternate pattern of Antoni A and B areas and nuclear palisading (Verocay bodies). Immunohistochemically, the protein S-100 is found on the supporting cells of both central and peripheral nervous systems, therefore allows the use of immunohistochemical S-100 in identifying the tumors arising from nerve sheath Schwann cells.^[13]

Differential diagnosis of schwannoma tongue includes neurofibroma, inflammatory fibrous hyperplasia, tumors of salivary gland origin, leiomyoma, rhabdomyoma, lymphangioma, hemangioma, granular cell tumor, (epi) dermoid cysts, lipomas, inflammatory lesions and lingual thyroid.^[18]

Therapeutically, complete surgical excision or enucleation is the treatment of choice.^[19] Recently, the use of CO₂ laser for excision of a base of tongue schwannoma has also been reported. Schwannomas are highly radio-resistant, therefore radiation therapy is not indicated. The prognosis is excellent due to the benign nature of this tumor. The recurrence rate as well as its malignant potential is quite low as reported by some authors.^[20]

Conclusion

Schwannoma of the tongue is an extremely rare benign tumor, especially in children. Given the rarity of this lesion, it generally remains masked or unrecognized by the treating physicians, creating diagnostic dilemmas. Hence, tongue schwannoma should be included as one of the

important differential diagnosis among the tongue lesions irrespective of the age group. Though, a combination of detailed clinical history, physical examination, cytological and radiological assessment aids in its diagnosis but it is histopathology that is confirmative.

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Competing Interests

None declared

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