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Case Report (Open Access)

SCHWANNOMA OF THE TONGUE: A RARE ENTITY

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ABSTRACT
Schwannomas, also known as neurilemmomas are uncommon neoplasms, derived from the schwann cells. They are benign, encapsulated, slow growing and usually solitary tumors. We report a case of schwannoma arising from right lateral border of tongue with an unusual presentation as a fungating mass. Because schwannomas are quite rare in the oral cavity, they are often not immediately included in the differential diagnosis of oropharyngeal masses, causing delay in identification and treatment. The definitive diagnosis requires histopathological examination.

Keywords: *Schwannoma, Tongue, Histopathology*

INTRODUCTION

Schwannoma or neurilemmoma is a rare, benign neurogenic neoplasm composed of Schwann cells (nerve sheath). It is usually slow growing, solitary, well demarcated and encapsulated (Wall and Snow, 1998; Neville *et al.*, 2003). Approximately 25-45% of all Schwannomas occur in the head and neck (Katz *et al.*, 1971). Of these approximately 1-12% occur intra orally (Lopez and Ballestein, 1993; Das Gupta *et al.*, 1969), tongue being the most common site. They originate more frequently from sensory nerves and can affect all cranial nerves, except the olfactory and optic (Lollar *et al.*, 2010). These tumors usually appear between the second and fourth decade of life, with no predilection for gender or race (Sawhney *et al.*, 2008). The size and locations of lesions determine the presence and intensity of symptoms. The goal of treatment is complete excision, which results in low rates of recurrence. Here, we present a case of schwannoma arising from right lateral border of tongue as a fungating mass.

CASE

An 18-year-old boy presented to the surgery opd for evaluation of a lesion on the right side of tongue (ventral part) slow growing since 1 1/2years. There was no history of tobacco consumption and past

Figure 1: Operative Pictures

medical history was unremarkable. Clinical examination revealed a 2cm, firm, fungating mass on the right lateral border of tongue (Fig1). The rest of the oral cavity and head and neck examination was unremarkable.

CT scan revealed a well-defined irregular mass lesion arising from the anterior aspect of right lateral margin of tongue measuring 2.5*2.0*1.4cm (AP*CC*TR) in size. On contrast administration, the lesion showed enhancement and appeared to be supplied by the lingual artery. A few sub cm sized non necrotic L.N's were noted in both sides of neck at levels II, III, IV (Fig 2).

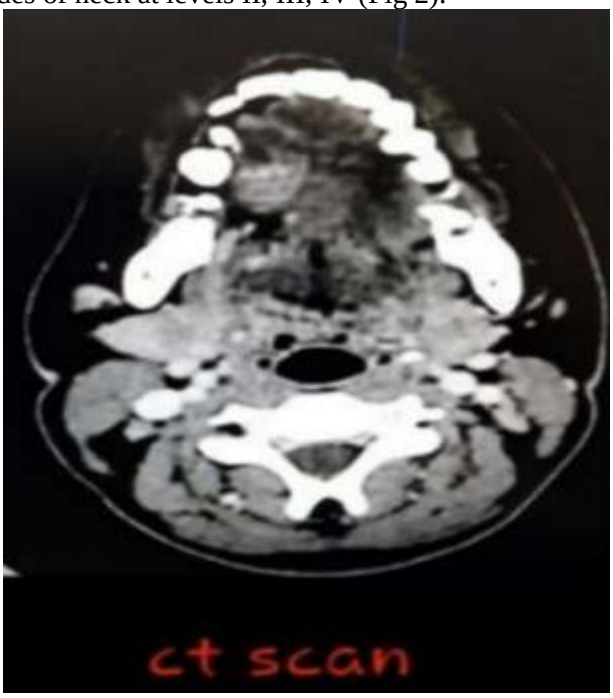

Figure 2: CT Findings

Punch biopsy taken from the lesion revealed hyperplastic and ulcerated squamous mucosa with underlying granulation. There was presence of spindle cell tumor with verocay body formation in the submucosa (Fig 3). Tumor was diffusely positive for S100 and negative for AE1/AE3, p40 and Desmin (Fig4). Findings suggestive of nerve sheath tumor – schwannoma of tongue. A wide local excision of the tumor was done (Fig1). Patient had an uneventful recovery and was discharged on the 2nd post op day.

Figure 3: Histopathology

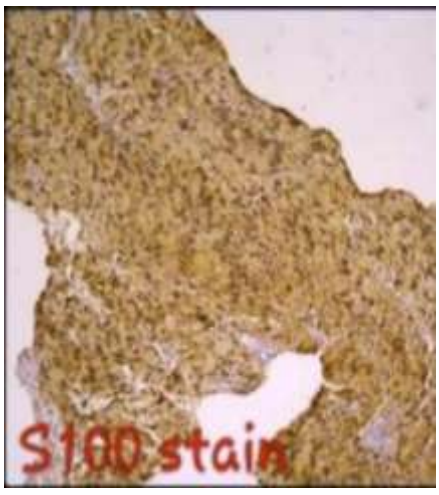

Figure 4: Histopathology

DISCUSSION

The Schwannoma also known as neurilemmoma or perineural fibroblastoma is a rare benign neural tumor arising from the neural sheath Schwann cells of the peripheral, cranial (except the optic and olfactory nerves) or the autonomic nerves, Lollar *et al.,* 2010). The etiology is unknown but it is postulated that the lesion arises by proliferation of the Schwann cells at one point inside the perineurium. Approximately 25-45% of all Schwannomas occur in the head and neck (Katz *et al.,* 1971). Of these approximately 1-12% occur intra orally (Lopez and Ballestein, 1993; Das Gupta *et al.,* 1969), tongue being the most common site. Other common locations for oral cavity are buccal mucosa, intra medullary bone of maxilla or mandible, floor of mouth, palate, gingival, lips, and vestibular mucosa, in that order (Gallo *et al*., 1977).

Schwannomas usually present as a solitary lesion. When multiple, however, they can be associated with neurofibromatosis. The differentiation between schwannoma and neurofibroma is essential because an apparently "solitary" neurofibroma may be a manifestation of neurofibromatosis, Sardinha *et al.,* 2005). Approximately 15% of patients with neurofibromatosis will have malignant transformation in one or more lesion, which is in marked contrast to the typical behaviour of a schwannoma, Bansal *et al.,* 2005).

Histologically, schwannomas display several features. Virtually, all of these tumors are encapsulated. Beneath this capsule, two main patterns are seen intermingled but sharply defined from each other. The first pattern is referred to as *Antoni type A* which consists of closely packed Schwann cells that form bundles or are arranged in rows with palisading, elongated nuclei. Free bands of amorphous substance between rows of nuclei constitute the *Verocay bodies.* The second pattern is known as *Antoni type B* and is composed of very loosely arranged Schwann cells set in a meshwork of reticulum fibers and microcysts, (Karaca *et al.,* 2010). In addition to these characteristic patterns, diagnosis is aided by immunohistochemical markers, S-100 and Leu 7, which support the Schwann cell nature of these tumors, (Sardinha *et al.,* 2005).

Several cases of malignant transformation of head and neck schwannomas have been reported (Kragh *et al.,* 1960), including one occurrence in the tongue, (Piatelli *et al.,* 1984).

All reported cases of schwannomas of the tongue have been treated by surgical excision. The most common approach was the transoral route.

CONCLUSION

Schwannomas although rare should be included in the differential diagnosis of well circumscribed slow growing tongue masses. A fungating presentation may mislead towards malignant diagnosis however the definitive diagnosis requires histopathological evaluation.

Transoral resection allows for removal of this tumor in a manner that precludes recurrence, avoids causing morbidity of tongue function, and remains the standard approach for treatment of the vast majority of these tumors. The chance of malignant transformation of these tumors is exceedingly unlikely.

Conflicts of Interest: None

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