

Tongue schwannoma: A case report and review of the literature

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ABSTRACT

Schwannoma is a benign common tumour of the head and neck. However, intraoral location is very infrequent (only 1-12% of all head and neck schwannomas). Tongue schwannoma are encapsulated, slow-growing and usually solitary neoplasms that arise from Schwann cells of the peripheral nerve sheath. We report a rare case of lingual schwannoma in a 37-year-old male who presented with an asymptomatic, slow-growing mass over the dorsal surface of the oral tongue with two months of evolution. The patient underwent MRI examination and fine-needle aspiration biopsy. Complete transoral surgical excision was conducted under general anaesthesia. Histopathological and immunohistochemical examination confirmed the diagnosis. No recurrences were observed on clinical follow-up. Schwannoma of the tongue, although rare, should be considered as a differential diagnosis for painless slow-growing lingual mass.

Keywords: Schwannoma, Tongue, Oral cavity, MRI, Surgery

INTRODUCTION:

Schwannomas, or neurilemmomas, are benign, encapsulated and slow-growing neoplasms that arise from Schwann cells surrounding the peripheral nerve sheaths [1].

Approximately, 25–45% of all schwannomas are seen in the head and neck. Of these, approximately 1–12% occurs intraorally with the tongue being the most common site [2,3]. But overall, lingual cases of schwannoma remain infrequent.

Although the etiology is unknown, some causative factors such as spontaneous growth, external injury, chronic irritation, or exposure to radiation have been hypothesized [4].

Schwannomas are usually sporadic, but in some cases they are associated with neurofibromatosis type II or schwannomatosis, with a latency of up to 50 years [5].

Schwannomas of the tongue most commonly occur between the second and fourth decades of life and display no gender predilection (52.8% female vs 47.2% male) and often present as a painless mass (69.6%) [1]. Intensity of symptoms is determined by the location and size of the tumour [6]. The treatment of choice is complete surgical excision of the tumor via the transoral approach. The recurrence rate is very low and malignant transformation is very rare [6].

We present a case of lingual schwannoma in a 37-year-old male patient, with MRI and histopathological correlation, in order to report the the diagnostic and therapeutic management elements with review of the literature.

OBSERVATION

A 37-year-old male complained of a slow growing and painless bump on the dorsum of the tongue, first noticed two months ago. The patient was otherwise healthy and non-smoker. No significant personal or family medical history was reported. There was no history of surgery or trauma to the area.

Intraoral clinical examination revealed the presence of a nodule (1,5 × 1cm in size), firm, well-defined, nontender and nonulcerated, with no contact bleeding, on the dorsal surface of the oral tongue (at the junction of anterior one-third and posterior two-third) close to the right border (Figure 1). Cervical lymphnodes were not palpable and the remaining clinical examination was normal.



Figure 1: nodule, firm, well-defined, nontender and nonulcerated, with no contact bleeding, on the dorsal surface of the oral tongue close to the right border



A MRI was performed to characterize the lesion, revealing a well-circumscribed nodule in the oral tongue, uniformly isointense to muscle on T1 weighted images and homogeneously hyperintense on T2 weighted images, showing homogeneous hypervascular enhancement in the contrast dynamic study (Figure 2).

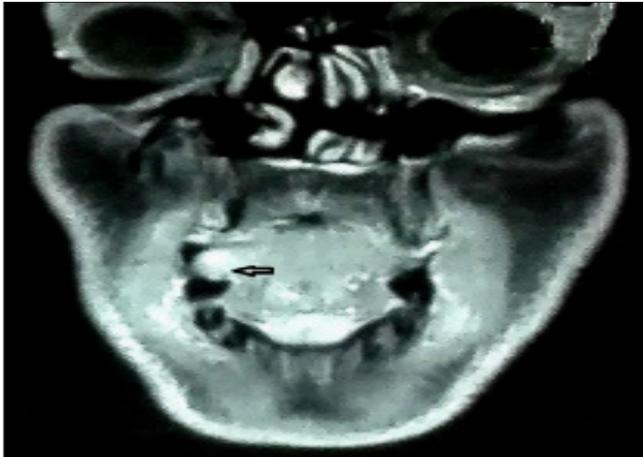


Figure 2: A well-circumscribed nodule of the oral tongue (arrow) showing homogeneous hypervascular enhancement in the contrast dynamic study on T1-weighted coronal view.

Fine-needle aspiration biopsy was performed suggesting the diagnosis of schwannoma. Complete transoral surgical excision was conducted under general anaesthesia. Grossly the mass was well encapsulated and measuring 1,5cm in diameter. The histopathological and immunohistochemical examination confirmed the diagnosis of tongue schwannoma.

The patient has not shown any recurrence with a follow up period of two years.

DISCUSSION:

Schwannomas are slow-growing, benign neurogenic neoplasms arising from Schwann cells in the nerve sheath which surround cranial, peripheral, or autonomic nerves, except the optic and olfactory nerves. The aetiology is unknown. Detecting the nerve of origin, in oral location, is often challenging [7].

Approximately 25–45% of all schwannomas are seen in the head and neck. The most commonly affected nerve in the head and neck is the vestibulocochlear nerve (90%). Approximately 1–12% of the head and neck schwannomas occur intraorally [3].

In the oral cavity, the tongue is most commonly affected, followed by the roof of the mouth, the floor of the mouth, the buccal mucosa, the gingiva, the lips and the vestibular mucosa [8].

Approximately 150 cases of schwannoma of the tongue have been previously documented in the literature, predominantly as single case reports or small series [9]. Lingual schwannoma can affect all age groups, being most commonly found between 10 and 40 years of age, without gender predisposition. In this site, they usually

appear as slow-growing, progressive nodules, showing with symptoms that, when present, vary according to their size and location.

In the tongue, two-thirds of cases involve the oral portion, and about one-third involve the base.

When in the base of the tongue, these tumours cause symptoms such as ~~sore throat, dysphagia, sleep apnoea~~, bleeding, infection and pain in about 75% of cases [10].

The vast majority of schwannomas present as solitary, sporadic lesions. Multiple schwannomas can be a manifestation of neurofibromatosis type II or schwannomatosis, which have to be considered as separate entities. The hallmark signs of neurofibromatosis type II are vestibular schwannomas, whereas in case of schwannomatosis, these tumours are absent [6].

Complaints of pain are usually identified in patients with schwannomatosis rather than sporadic cases. Nerve oedema and myxoid changes have been correlated to this symptom [11].

Two types of schwannomas can be seen in the oral cavity. The most common is the submucosal type, which presents itself as well defined, encapsulated, with firm consistency, and cyst-like appearance. The other is the non encapsulated type, located below the basal layer of the mucous membrane [12].

Thompson et al reported, in a recent clinicopathologic Study of 19 Cases of tongue schwannoma, a variable tumor encapsulation with nearly half of the lesions lacking a well-defined fibrous capsule. This observation is consistent with the frequent finding of capsular absence in mucosal based schwannomas of other oral cavity sites [9].

The diagnosis is suggested by fine-needle aspiration biopsy and confirmed via histopathological examination, by observation of a well-defined, encapsulated tumor formed of spindle shaped cells that can be arranged in two patterns. The “Antoni A areas”, which are more cellular and contain the characteristic Verocay bodies, formed of elongated cells with no atypia; the cell nuclei are arranged in line and their cytoplasm is fused into eosinophilic masses. The “Antoni B areas” contain randomly distributed cells in a myxomatous stroma. Schwannomas are characterized by strong and diffuse immunoreactivity for S-100 protein, which is the clue for the diagnosis [13].

Imaging has become an integral part of evaluation for tongue lesions, and thus, a systematic imaging approach should be considered. On MRI, schwannoma is typically a well-defined lesion which demonstrates hypointensity on T1WI, hyperintensity on T2WI, and intense post contrast enhancement. Furthermore, helpful radiological MRI signs include split fat sign (thin peripheral rim of fat on T1WI), target sign (central low signal within the lesion), and a fascicular sign (multiple internal small ring-like structures) [14].

The clinical differential diagnosis includes those lesions that present as a well-encapsulated tumor: granular cell



tumor, salivary gland tumor, leiomyoma, neurofibroma, lymphangioma, hemangioma, cyst, lipoma, and others [13]. Although most extracranial and intracranial schwannomas are benign, malignant schwannomas are reported in the literature and account for 5% of all soft tissue sarcomas. Of these, only 9–14% are located in the head and neck [15].

Malignant transformation of schwannoma is, in contrast to neurofibroma, an exceptionally rare event; reported cases often occurred in the setting of von Recklinghausen's disease [16].

Treatment of tongue schwannomas is surgical and is more complex in those tumors situated on the base of the tongue. Several surgical techniques are employed to remove schwannomas of the tongue due to varying levels of surgical difficulty based on tumor size and location. Traditional surgical techniques include transoral excision, suprahyoidpharyngotomy, submandibular approach, and mandibulotomy with lip splitting. The transoral surgical approach is the most common and preferred method. The utilization of CO₂ laser treatment in tumor excision is an emerging alternative to traditional surgical methods [1].

Schwannomas are not responsive to radiotherapy, and incomplete surgical excision may result in recurrence, although recurrence is uncommon following complete resection [17].

CONCLUSION:

Tongue schwannomas are rare benign neoplasms of the oral cavity. Nevertheless, they should be considered in the differential diagnosis of lingual masses, especially since prognosis is favourable, when this condition is correctly identified. The imaging modality of choice is MRI. It allows a precise localization and measurement of tumour size, as well as a better visualization of the relations to other structures.

The final diagnosis is done after histopathological and immunohistochemical examination. Transoral resection is the standard treatment approach in most cases. Although more often solitary, association with neurofibromatosis type 2 should be kept in mind. Despite a very low recurrence rate and an exceptional malignant transformation, a regular follow-up should be considered.

Compliance with ethical standards

Conflict of interest: The authors stated that there is no conflict of interest.

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