Case Report

A rare clinical case: schwannoma of tongue

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ABSTRACT

Schwannoma are slow growing tumours, which can arise from any peripheral nerve. 10% of schwannomas that occur in the head and neck region mostly originate from the vagus or sympathetic nervous system. Extracranial schwannomas in the head and neck region are rare neoplasm. Intraoral schwannoma are only 1% of all head and neck tumours. Diagnosis is established by imaging studies such as magnetic resonance imaging or computed tomography, while FNAC is used to rule out other conditions. We report a rare case of lingual schwannomas generally present as a painless lump schwannoma of the tongue in a 27-year-old male complaining of asymptomatic swelling over a posterolateral surface of the tongue, treated by complete surgical excision. The diagnosis was established on the basis of clinical, histopathological, and immunohistochemical examination. We report a rare case of schwannoma over the posterolateral surface of tongue. Prognosis is good for the patient when this condition is correctly diagnosed as the condition rarely recurs after complete resection.

Keywords: Schwannoma/neurilemmoma, Peripheral nervous system neoplasms, Tongue diseases

INTRODUCTION

Schwannomas also known as neurilemmomas or neurinomas are benign nerve sheath tumors deriving from Schwann cells that occur in the head and neck region in 25-45% of cases.1 About 10% of schwannomas that occur in the head and neck region mostly originate from the vagus or sympathetic nervous system.2,3 However, they are quite rare in the oral cavity, accounting just over 1% of benign tumors.4 In the oral cavity, tongue is the most common location followed by palate, floor of mouth, buccal mucosa, and mandible.5 In more than 50% of the intraoral tumors, it is difficult to differentiate between tumors of the lingual, hypoglossal, and glossopharyngeal nerves. Interestingly, if these lesions arise from a large nerve, the nerve fibers are spread out over the capsule surface and not integrated within the mass itself with the parapharyngeal space being the most common site.6,8 Preoperative imaging studies such as magnetic resonance imaging (MRI) and computed tomography (CT) are used to distinguish its location and origin. The treatment of schwannoma is primarily surgical resection.9,10 Malignant transformation and recurrence following complete excision are rare.11 We report a rare case of schwannoma over the posterolateral surface of tongue for several years.

CASE REPORT

A 27-year-old Indian man presented with a slow-growing swelling over posterior part of left side of tongue that was first noticed three years before. Initially it was small in size, gradually progressed to current size, not associated with pain, with no difficulty in swallowing, chewing and speaking during this period. Oral cavity examination showed a mass of 5 × 4 cm on the left lateral aspect of the tongue, with normal-appearing overlying mucosa. Palpation revealed a well circumscribed, smooth, firm, nontender, noncompressible, nonreducible, nonfluctuant
mass with well defined borders, involving the lateral tongue on left side (Figure 1). Tongue mobility was normal. Simple gustatory testing to sweet, sour, and salt yielded normal results. The remaining oral examination was unremarkable and there were no cervical lymph node enlargements. He had no coexisting disease or any exposure to relevant health hazards. The clinical differential diagnosis included benign tumors such as fibroma, lipoma, and neurofibroma.

Routine hematological and urine examination were normal. Pre-operative magnetic resonance imaging (MRI) showed, a solitary well-circumscribed focal enhancing lesion in the left half of oral tongue, splaying the tongue musculature and midline raphe without extension across midline reaching up to the ipsilateral, floor of mouth, but sparing the tip, root and base of tongue, along with lingual and hypoglossal nerves. The imaging features favored a benign or low grade neoplasm like minor salivary gland neoplasm (Figures 2-4). Lesion size 5.3 cm horizontal by 4.4 cm anterioposterior by 3.1 cm wide, with a lesional volume of 36.2 cc (Figure 5, 6). The midline raphe was splayed along the periphery of the lesion with minimum distance between the lesion and midline raphe being 2.8 mm. Spares the tip, root and BOT, RMT, FOM and lingual – hypoglossal nerves. Surgical excision via trans-oral approach was performed. Histopathology reports of the excised surgical specimen on low-power imaging revealed Antoni A areas with ill-defined fascicles of spindle-shaped cells (Figure 7), while high-power imaging found fewer cellular Antoni B areas, with loosely arranged cells (Figure 8). Immunohistochemistry was positive for S-100 protein, confirming schwannoma as the diagnosis (Figure 9).

Figure 1: Preoperative image of schwannoma over the left posterior part of the tongue.

Figure 2: Axial T2 TSE through the left border of the tongue shows a circumscribed mass centered left of midline in the posterior base of the tongue. The tumor has a smooth, well defined border, with no invasion into adjacent muscle.

Figure 3: Coronal T2 TSE through the left border of the tongue shows a circumscribed mass centered left of midline in the posterior base of the tongue. The tumor has a smooth, well defined border, with no invasion into adjacent muscle.

Figure 4: Sagital T2 TSE through the left border of the tongue shows a circumscribed mass centered left of midline in the posterior base of the tongue. The tumor has a smooth, well defined border, with no invasion into adjacent muscle.
DISCUSSION

Majority of the schwannomas occurs in the head and neck region, and intraoral schwannomas account for approximately one percent of all schwannomas. This rarity of schwannomas in the oral cavity makes it difficult for the clinicians to diagnose it promptly. A brief review of literature published by Lee et al reported only 85 cases of lingual schwannoma since 1959. On conducting a Google scholar search for lingual schwannoma cases after 2017, we could identify six more cases till date. Two of these cases, one pediatric case from United States and other adult patient from Japan, were associated with neurofibromatosis type 2 (NF2). They both were histologically confirmed and treated with surgical excision. Earlier to these reported cases, only three cases of lingual schwannomas in patients with NF2 have been reported, all of which were single, well-circumscribed, nodular lesion on the dorsal surface of the tongue. Due to the medical history of NF2, the tongue lesions are often
misdiagnosed as neural tumors associated with NF2. However, only two of the reported cases were histologically diagnosed as localized amyloidosis and amyloid tumors. The third case was clinically diagnosed as neurofibroma or schwannoma but without histological confirmation. These cases highlight the importance of conducting careful oral examinations in patients with NF2. It may also provide an opportunity to diagnose NF2 in patients with lingual schwannoma, which may have been missed earlier.

Two cases have been reported from India. One case reported by Nair et al, was a 19 year old male, diagnosed with mucocoe in the posterior part of the tongue based on the clinical and cytological findings. Cut section revealed mucinous material and histopathological examination revealed both hypocellular and hypercellular areas with cystic areas. However, on immunohistochemistry, the cells were strongly positive for S100, which led to the final diagnosis of cystic schwannoma of the tongue. Histological appearance of schwannoma has been well described in the literature, like the presence of alternating pattern of hypercellular Antoni A and hypocellular Antoni B areas, nuclear palisading, and Verocay bodies. However, cystic schwannomas after secondary changes may become difficult to diagnose even on histopathology. In such cases, immunohistochemistry plays a pivotal role in confirming the diagnosis, as the treatment of mucocoe and schwannoma are different. The other case reported from India by Rana and Ohri was of a 17 year old male presenting with a small swelling on the base of the tongue. Initially diagnosed with fibroma, transoral excision biopsy revealed schwannoma.

Diplan and colleagues reported a case of a 40 year old Dominican woman who presented with a painless, slow, and progressive growing tumor of the oral tongue with a discreet margin at the tip of the tongue. She complained of slurred speech and dysphagia. An anterior midline glossotomy approach with total excision and primary closure was done along with a temporary tracheotomy because of the tumor size, involvement of the base of the tongue and the risk of edema. Inoue et al reported a case of 61-year-old male presenting with a one year history of tongue atrophy and dysarthria. Imaging revealed a dumb-bell shaped giant mass in the right extrapleural hypoglossal canal invading intradurally with a large extension to the high cervical region. The authors used a single-stage radical surgical procedure using the anterolateral extreme lateral infraglottic transcondylar exposure approach. Postoperative assessment revealed no neurologic deficit except for the pre-existing tongue atrophy and histological examination confirmed schwannoma.

Majority of the reported cases of lingual schwannomas have been treated by surgical excision, most commonly by the transoral route. Several other approaches have been described including submandibular, suprathyroid pharyngotomy, and transhyoid approaches. More recently, the use of CO2 laser for excision of a base of the tongue schwannoma has also been reported. The goal of surgical therapy is to complete resection. Complete resection is very rarely associated with recurrence. Safety and efficacy of gamma knife radiosurgery for the treatment of lingual schwannoma is still unclear. Elsharkawy et al and Suri et al reported satisfactory results with the use of gamma knife surgery.

CONCLUSION

Schwannoma of the tongue is a relatively rare tumor and is often misdiagnosed. Though histopathology should confirm the diagnosis in majority of cases, immunohistochemistry examination may be required to support the diagnosis. Trans-oral excision is the most commonly reported treatment but it will finally depend on the location and size of the schwannoma. Refined micro-surgery skills are required for complete resection, after which recurrence is rare.

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