

Schwannoma of tongue- A case report

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ABSTRACT

Introduction: Schwannoma is a benign, encapsulated, slow growing solitary tumor, derived from Schwann cells of the peripheral nerve sheath. Approximately, 25-48% of cases are seen in the head and neck region of which only 1% occurs in oral cavity.

Case Presentation: Here we report a case involving left ventrolateral aspect of tongue of a 48 years old male patient which was diagnosed as schwannoma, based on clinicopathological evaluation.

Management and Prognosis: Incisional biopsy was performed from the representative site of the lesion under local anesthesia. After proper diagnosis, the case was sent to Oral and Maxillofacial Surgery Department for further treatment and management.

Conclusion: Schwannoma of the tongue is a relatively rare tumor of head and neck region. Following complete surgical excision recurrence is relatively rare. So proper clinical and histopathological diagnosis is very essential to treat this benign neoplasm.

Keywords: Nuclear palisading, Neurilemmoma, Schwann cell, Tongue, Verocay bodies.

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INTRODUCTION

Schwannomas or Neurilemmomas are benign nerve sheath tumors, usually arising from the Schwann cells which are an integral component of myelin sheath, ensheathing the peripheral nerves^{1,2}. They account for 25-40% occurrence in the head and neck region³ while others involve the flexor surfaces of upper and lower extremities, posterior mediastinum and retroperitoneum.^{4,5} 1-2% schwannomas can occur intraorally, the most common site being tongue, followed by palate, floor of mouth, buccal mucosa and rarely it may occur centrally within the mandible.^{3,5} The usual age of occurrence is between 2nd – 5th decades of life without any definite gender predilection⁴. Clinically, schwannomas present as non tender, slow growing, soft to firm, globular, expansile submucosal masses being sessile or pedunculated, varying in size from 1-4 cm in diameter. Depending on the location of tumor and its mass effect on nerve involvement, patient may experience pain, hoarseness of voice, dysphagia, cranial nerve neuropathy and even Horner's syndrome.² Tongue base schwannomas may cause pain, swelling, fasciculations, loss of tongue control and weight loss, although most are asymptomatic until the tumor reaches a significant size.^{1,2}

Histopathologically, they are composed of collagenous products, especially consisting of numerous proliferating spindle shaped cells with elongated nuclei, showing a typical biphasic patterns namely Antoni A and B. Nuclear palisading is usually present in Antoni A areas, forming "Verocay bodies"; whereas Antoni B areas are paucicellular with small round cell within a myxoid stroma.^{7,8,9,10} Immunohistochemically, the neoplastic cells showed intense immunopositivity for S100. Focal positivity for SOX 10 indicates neural crest origin of the cell and immunonegativity to SMA helps us to confirm the diagnosis. In general, this neoplasm does not undergo malignant transformations⁵.

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Schwannomas are not responsive to radiotherapy. It is usually treated by complete surgical excision through the transoral approach. Following complete excision the recurrence is relatively rare. More recently carbon dioxide laser has also been introduced to treat tongue schwannoma².

Based on the above clinico-pathological, radiological, histopathological and immunohistochemical findings, a case of schwannoma involving the left ventrolateral side of tongue was diagnosed and has been discussed herewith.

CASE REPORT

A 48 years old male patient from a semiurban area reported to the Department of Oral and Maxillofacial Pathology of Guru Nanak Institute of Dental Sciences and Research, Panihati, Kolkata, with a chief complaint of a growth involving the left lateral border of tongue for about 5-6 months. There had been a gradual increase in size of the growth to attain the present dimension over a period of 2-3 months without causing any



Fig 1 (A,B) Intraoral photographs showing well localized, multilobulated, round to ovoid, non tendered, soft to firm growth involving left ventrolateral aspect of tongue.



Fig 2 Contrast enhanced MRI of oral cavity revealed an ill-defined altered signal intensity lesion along the left lateral border of the tongue in the sublingual space. It appears isointense to muscle in T1W1 and heterogeneously hyperintense in long TR sequences

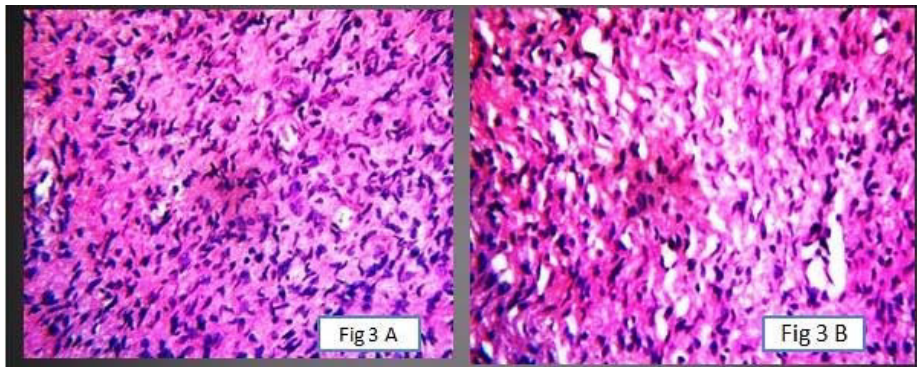


Fig 3 (A,B) High power photomicrographs (H&E X40) revealed the presence of diffuse arrangement of neoplastic spindle shaped cells within a loosely arranged myxoid stroma.

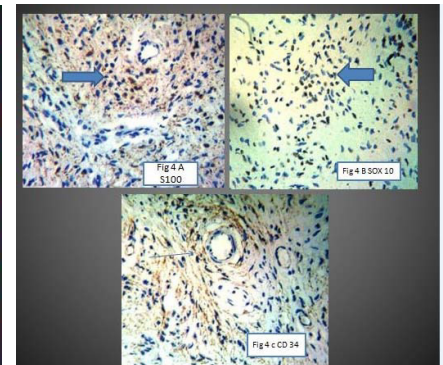


Fig 4(A,B,C) High power photomicrographs showing intense immunopositivity of neoplastic cells towards S100, focal positivity to SOX 10 and CD 34.

pain. Patient also complained of discomfort during mastication, phonation and tongue movement. Medical history of the patient revealed hypertension which was well controlled with medication and no pertinent family history was reported.

Intraoral examination revealed the presence of a well localized, multilobulated, round to ovoid, non tendered, non pulsatile, non compressible, sessile, soft to firm growth measuring about 4.5x5.5 cm, involving the left ventrolateral border of tongue and floor of the mouth (Figure 1). Oral hygiene of the patient was very poor without any lymphadenopathy.

Based on above clinical findings, the case was provisionally diagnosed as benign soft tissue neoplasm. Differential diagnoses of Neurofibroma, Schwannoma, Granular cell myoblastoma, Giant cell fibroma, Focal fibrous hyperplasia and Lymphangioma were suggested. Then the patient was advised to perform MRI of the affected site. Contrast enhanced MRI of oral cavity revealed an ill-defined altered signal intensity lesion along the left lateral border of

the tongue in the sublingual space. It appears isointense to muscle in T1W1 and heterogeneously hyperintense in long TR sequences (Figure 2). The lesion was seen to cause mild compression over the left genioglossus muscle medially, situated above the mylohyoid muscle and there was no extension of the lesion into the floor of the mouth.

Considering all clinical and radiological features, an incisional biopsy was performed from the representative site of the lesion after taking the written consent from the patient. The light microscopic features revealed the presence of nonencapsulated tumor mass with the tumor parenchyma consisting of numerous proliferating spindle shaped cells having elongated nuclei, scattered in disorderly manner within a relatively myxoid stromal background along with multiple vascular spaces of varying size. The tumour consist of both hypocellular and hypercellular areas at places (Figure 3). The overall histopathological features were suggestive of benign spindle cell tumor. Differential diagnoses of Neurofibroma,

Leiomyoma, Fibroma, Fibroepithelial polyp, Granular cell tumor and Lipoma were considered. To confirm the histopathological diagnosis Immunohistochemistry was performed. The neoplastic cells showed intense immunopositivity to S100, focal positivity to SOX 10, CD 34 and immunonegativity to SMA (Figure 4). These findings helps us to confirmly diagnose the case as 'Schwannoma'.

The patient was then referred to the Department of Oral and Maxillofacial Surgery for further treatment and management. However, he had refused to undergo any surgical intervention in our institution because of his poor socio-economic condition. Hence, any sort of follow-up treatment was not be possible.

DISCUSSION

The peripheral nerve sheath tumors are divided into two groups:- benign and malignant. The benign category includes two most common and closely related tumors –schwannoma and neurofibroma. Schwannomas are benign encapsulated nerve sheath neoplasm composed of Schwann cells, first discovered by Verocay in the year of 1908. This tumor can arise from any nerve covered with Schwann cell which includes the cranial nerves, (except for optic and olfactory) the spinal nerves and the autonomic nerves.^{1,2,3,7,8,9}

Schwannomas are usually solitary but multifocal lesions have also been reported. Multiple lesions occur in: 1-multiple localized neurilemmomas, 2- in association with neurofibroma in Von Recklinghausen's disease, 3- in Schwannomatosis, a non hereditary disease characterized by multiple subcutaneous and intradermal schwannomas along with a variety of intracranial tumors. The patient under discussion was a 48 years old male, having a well localized, multilobulated, non tendered swelling involving the left ventrolateral border of tongue leading to mild discomfort during phonation and mastication. All of these clinical findings are similar with the observations reported by various authors in different studies^{1,2,3,5,6}.

Magnetic resonance imaging (MRI) scans can be performed to determine extent of the lesion and the characteristic findings revealed a homogenous enhancing lesion without evidence of infiltration into the adjacent normal^{3,4,7}. The MRI finding of our case revealed an ill defined altered signal intensity lesion along the left lateral border of tongue in the sublingual space causing mild compression over left genioglossus muscle without any extension of the lesion in the floor of mouth.

Because of uncommon occurrence and non specific clinical presentation, the diagnosis is usually confirmed by histopathological and immunohistochemical evaluation.

Microscopically, most of the tumors are well demarcated from the adjacent normal by a fibrous capsule and the basic cellular constituents are proliferating neoplastic spindle shaped cells characterized by the presence of elongated nuclei. These tumor cells are arranged in two distinct patterns: Antoni type A and Antoni type B which are pathognomic for the lesion. Antoni A pattern revealed parallel rows of palisaded nuclei of Schwann cells arranged around the centrally located round to ovoid

eosinophilic acellular areas i.e. Verocay body where as Antoni B pattern characterized by the randomly arranged cells having oval, sometimes elongated nuclei within a loose myxoid stroma- this pattern is less cellular, less organized than Antoni A. Antoni B sometimes associated with degenerative changes like hemorrhage, cellular, nuclear pleomorphism and nuclear atypia –hence they are sometimes confused with malignancy. In accordance with previous histopathological findings as reported in published case reports,^{1,2,3,4,5,7,8,9,10} the present case revealed Antoni B pattern comprising of haphazardly arranged spindle shaped cells in a loosely arranged myxoid stroma interspersed with numerous vascular spaces of varying sizes. Immunopositivity for S100, SOX 10 and CD 34 supports the Schwann cell origin of this neoplasm.

Neurofibromas consist of interlacing bundles of spindle cells exhibiting wavy nuclei and associated with delicate collagen bundles in variable myxoid stromal background. Special stain with Toluidine Blue reveals presence of mast cells. Immunohistochemistry suggests scattered positivity for S100 while negativity to SOX 10.

On the contrary, Schwannomas reveal Streaming fascicles of spindle cells with elongated nuclei arranged in Antoni A and Antoni B patterns; however our case revealed only Antoni B areas. Immunohistochemistry reveals intense diffuse positivity for S100 and SOX 10 which determines the Schwann cell lineage. This guided us to the confirmatory diagnosis of schwannoma.

CONCLUSION

Schwannoma of the tongue is a relatively rare tumor of head and neck region. Following complete surgical excision recurrence is relatively rare. So proper clinical and histopathological diagnosis is very essential to treat this benign neoplasm.

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