CASE REPORT

Exploring Schwannoma of the Tongue: Two Distinct Case Reports

Sabu Paul, Latha Mary Cherian, Aparna Thomas, Niva P, M.S Aparna, Srevya Rajeev

Abstract

Schwannomas are benign encapsulated nerve sheath tumor of schwann cell origin. The preferred site of occurrence is head and neck region in 25-45% cases, among which oral cavity accounts for less than 1%. This case report discusses two cases of schwannoma, that exclusively occurred on the tongue.

Case 1: A 41 year old female with a swelling on the left lateral border of tongue with well defined border and no history of surface changes. Case 2: A 21 year old male with a swelling on right anterior two-third of the tongue, which was well circumscribed and firm. Both the patients underwent successful surgical resection and the lesions were confirmed for schwannoma histopathologically. The post operative recovery was uneventful with no signs of recurrence during follow up.

Schwannoma is a benign tumor with good prognosis. These tumors are usually treated by transoral resection, which makes it possible to remove the tumor in a way that prevents recurrence and impairment of tongue function.

Keywords: Benign lesion, Schwannoma, Soft tissue tumor, Schwann cell origin

Introduction

Virchow made the initial identification of schwannoma in 19081. Any nerve coated by a Schwann cell sheath, such as the spinal nerves, the autonomous nervous system, and the cranial nerves (except from the optic and olfactory nerves), can develop these tumours[1,2,3]. Studies on tissue cultures conducted by Murray and Stout, who grew this tumor in vitro, support the hypothesis that Schwann cells are the source of origin[4]. Though the tongue is most frequently affected, other mouth regions that are also affected by intraoral schwannoma include the palate, mouth floor, buccal mucosa, gingiva, lip, and vestibule[5]. The tongue schwannoma is most commonly seen in the second to fourth decades of life[6]. The lesion is slow growing, and thus, its onset is usually long before presentation. The role of sex is not important in its pathogenesis[7]. In addition, some systematic review studies have reported that about 70% of tongue schwannomas are painless[5]. Clinically, the schwannomas may be indistinguishable from other encapsulated benign tumors, therefore biopsy and histological examination are essential to formulate a correct diagnosis.

Case Report

CASE 1: A 41-year-old female patient presented to the Department of Oral Medicine and Radiology with a chief complaint of swelling on the left lateral side of the tongue, since past eight months. On intra-oral examination, a firm, sessile swelling of size 1.5 x 2 cm on left lateral border of tongue was noted, with mild tenderness on palpation. Color of swelling was similar to adjacent mucosa and no induration was noted (Figure 1).

On extra oral examination, there was no detectable facial asymmetry. Bilateral submandibular lymph nodes were soft, non-tender and not fixed. Her past medical history, personal history, and family history were non-contributory. A provisional diagnosis of lipoma or traumatic fibroma or vascular lesionwas considered. Upon ultrasonic examination of the lesion, a welldefined hypoechoic lesion, with no evidence of internal or peripheral color uptake/ vascularity was found. The lesion was excised and sent for histopathological examination.

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CASE 2: A 21-year-old male patient reported with a chief complaint of swelling on the right side of tongue since 2 months. The patient had no difficulty in chewing, swallowing, or phonation and also no sensory or taste abnormalities. On intra-oral examination a well circumscribed sessile growth of size 1x0.8 cm over right anterior two-third of tongue was present (Figure 2). On palpation the swelling was non-tender, non fluctuant, non-indurated and firm in consistency. On extra-oral examination, lymph nodes were non-palpable and non-tender. Past medical history, personal history, and family history were non-contributory. A provisional diagnosis of fibroma was considered. The lesion was excised and sent for histopathological examination.

The hematoxylin and eosin-stained sections of Case 1 and Case 2 revealed stratified squamous epithelium overlying the moderately collagenousconnective tissue stroma. Connective tissue stroma revealed a well encapsulated area consisting of numerous spindle cells. Combination of hypocellular and hypercellular areas noted. Streaming fascicles of palisading schwann cells constitute the Antoni A areas (Figure 3). Few hypocellular areas consisting of spindle cells were randomly arranged in the loose myxoid stroma, which constituted the Antoni B areas (Figure 4). Vascularity was moderate.

Immunohistochemical staining were done for Case 1 and 2 for confirmatory diagnosis. Schwann cells were S100 positive (Figure 5). With this, a confirmatory diagnosis of Schwannoma was given for both cases. Patients were followed-up for recurrence.

Discussion

In 1908, Virchow made the initial identification of schwannomas[1]. Also known as neurilemmoma, neuroma, neurinoma, or perineural fibroblastoma is a very frequent tumor originating from Schwann cells[8]. Any nerve coated by a Schwann cell sheath, such as the cranial nerves (except from the ocular and olfactory nerves), the spinal nerves, and the autonomous nervous system, can develop these tumors[2,3]. It can be challenging to find the nerve sourceintongue schwannomas between the lingual, glossopharyngeal, and hypoglossal nerves due to their close proximity[9]. Murray and Stout's tissue culture research, fostered the in vitro growth of this tumor, which indicated that Schwann cells are the cells of origin[4,7].

According to reports, between 25% and 50% of Schwannomas are located in the head and neck area, and between 1% and 12% are discovered in the oral cavity[2,3,5,6,7]. Even though

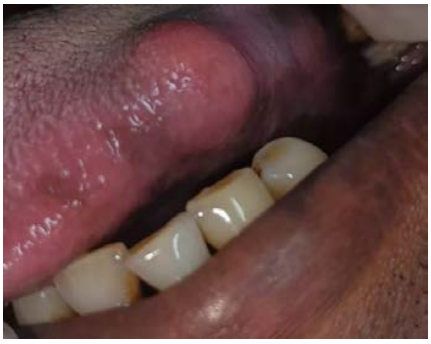

Fig. 1: Clinical examination showing swelling of size 1.5x2 cm on left lateral border of tongue.

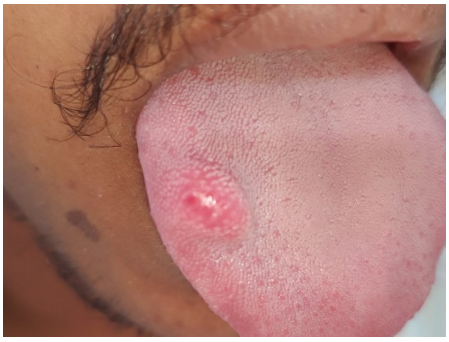

Fig. 2: Clinical examination showing swelling of size 1xo.8 cm on right anterior two-third of tongue

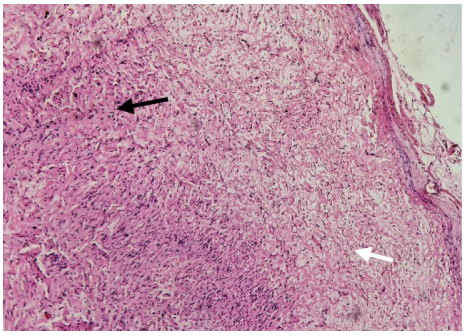

Fig. 3: shows combination of hypercellular and hypocellular areas in connective tissue stroma with streaming fascicles of palisading Schwann cells constituting Antoni A (black arrow) and randomly arranged Antoni B (white arrow) areas. (H&E stain, original magnification x10).

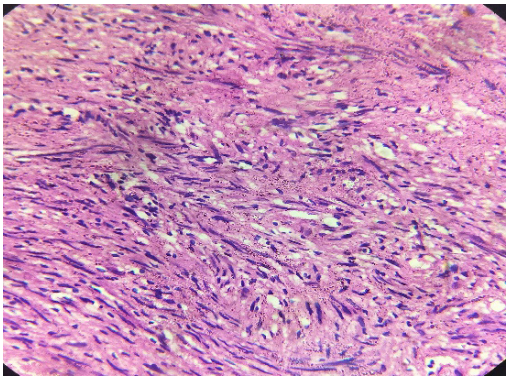

Fig. 4: Spindle cells arranged randomly in loose myxoid stroma, constituting Antoni B areas. (H&E stain, original magnification x40).

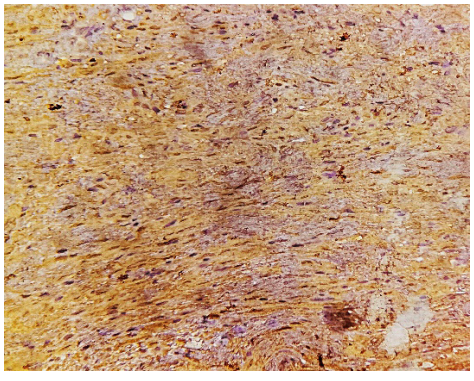

Fig. 5: shows S100 positive tumor cells.(x40)

the exact cause of Schwannoma is unknown, some pertinent risk factors, such as radiation exposure, oxidative stress, external injury, chronic irritation, and hereditary vulnerability, have been documented in the literature[6]. The neurilemmoma has developed in a number of oral and paraoral sites, including the tongue, palate, floor of the mouth, vestibule, lip, gingiva, and buccal mucosa. Additional domains concerning are the retropharyngeal, maxillary sinus, and salivary glands, nasopharyngeal and retrotonsillar regions. Furthermore, it has been documented that neurilemmoma occur as a central lesion within bone, primarily the mandible, that appears to originate from the mandibular nerve[4,3].

Schwannomas are more common in people between the ages of thirty and fifty. The majority of cases of tongue schwannoma occur in the second and fourth decades of life. Studies have also shown that it is more common in women (52.8%) than in males (47.2%), suggesting that gender has little to no role in the disease's aetiology[6,7].

The loss of merlin activity, either directly through a genetic alteration involving the NF2(neurofibromatosis 2) gene on chromosome 22 or indirectly through merlin inactivation, is a key factor in the pathophysiology of these tumors. Number of studies with mice lacking NF2 gene have showed a marked impairment in nerve regeneration. The inability of Schwann cells to redifferentiate is a primary cause of this drastically reduced regeneration. Tumor formation eventually comes from persistent cell proliferation caused by a lack of Schwann cell re-differentiation[10,11].

Schwannomas are well-circumscribed, benign tumors made up of clonal populations of Schwann cells, which frequently experience degenerative and cystic changes. The majority of occurrences are sporadic, but a small percentage are linked to NF2(3%), schwannomatosis(2%), Carney's complex (5%) with meningiomatosis with or without NF2, and also bilateral vestibularschwannomas are diagnostic of NF2[3,12,13].

The heterogeneity of schwannomas was initially described by the Swedish neurologist Nils Ragnar Eugene Antoni, who separated the tumor tissue into two sections with different architectures[11]. They contain regions made up of fascicles of Schwann cells with the morphology of spindle cells (Antoni A pattern), which can abruptly show transition to other areas which are microcystic and myxoid (Antoni B pattern) or merge with them. Schwannomas exhibit regions of palisading or nuclear alignment; these regions frequently form Verocay bodies or parallel nuclear arrays, which were described by Jose Juan Verocay in 1910[4,14,15].

Histological variants of schwannoma includeancient schwannoma, cellular schwannoma, plexiform schwannoma, schwannoma with pseudoglandular elements, cystic schwannoma and psammomatous schwannoma. Schwannomas classified as ancient exhibit pronounced degenerative nuclear atypia. Cyst formation, calcification, bleeding, and hyalinization caused by a high number of siderophages and histiocytes are examples of degenerative processes. Large, hyperchromatic, and frequently multilobed, Schwann cell nuclei lack mitotic features. Cellular schwannoma is a crucial variant to identify because of its high cellularity, fascicular growth pattern, enhanced mitotic activity, and occasionally locally destructive behavior, which frequently raises the possibility of malignancy. It lacks Verocay bodies and its Antoni B pattern growth is limited to very focal areas (less than 10% of the tumor size). Plexiform variant is characterized by intraneuralnodular pattern of growth. It may be linked, to disorders such as NF2 and schwannomatosis. These tumors might not even have a capsule. Melanotic schwannoma is characterized by epithelioid cells with variable-sized nuclei and a considerable buildup of melanin in the neoplastic cells and melanophages[14,16,17].

Histologically, schwannomas resemble neurofibromas which contain a mixture of Schwann cells, fibroblasts, and perineurial-like cells embedded in a collagenous matrix. Unlike schwannomas, neurofibromas lack Antoni A, Antoni B areas and Verocay bodies[18]. Other differential diagnosis include malignant peripheral nerve sheath tumor and solitary fibrous tumor, both of which exhibit negative S100 immunostaining.[19]

Strong and diffuse S-100 protein positivity is a hallmark of Schwannomas, despite the fact that S-100 protein production is typically reduced in the Antoni B regions, immunostaining for this protein is so reliable and intense that it is a valuable diagnostic technique. Immunohistochemically, neurofibromas are also positive for S-100, but the staining is less intense and less uniform compared to schwannomas. Another effective marker for schwannomas is nuclear staining for SOX10, marker of neural crest development. These tumors also express CD57 and, on occasion, glial fibrillary acidic protein (GFAP)[18,20,21,22,23].

The usual course of treatment for schwannomas is complete excision. Malignant transformation and recurrence are quite uncommon. In young individuals, tongue lesions should always be biopsied or excised since, malignancy can occur, even though rare[2,7].

The uniqueness of these cases lies in the rare presentation of tongue schwannoma occurring in two distinct age groups, mimicking more common lesions and necessitating histopathology and immunohistochemistry for a definitive diagnosis.

Conclusion

Schwannomas of the tongue, while rare, represent a unique subset of peripheral nerve sheath tumors that can be challenging to diagnose due to their nonspecific clinical presentation. Early recognition, accurate diagnosis via imaging and histopathological analysis, and surgical excision are crucial for successful management. These cases underscore the importance of considering schwannomas in the differential diagnosis of tongue masses, ensuring appropriate treatment and favorable outcomes with minimal recurrence risk.

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Consent for Publication

Signedconsent has been obtained from the patient.

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