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Case Report

Base of tongue schwannoma on a 22 year old lady: a rare case

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ABSTRACT

Schwannomas are benign tumours arising from the Schwann cells. Around 25-45% schwannomas have been reported occured in head and neck region, but only 1% located intraoral. We report a case of 22 year old woman who presented with foreign body sensation in throat, dysphagia, change of voice, and snoring. There was a slowly growing, solitary, encapsulated, painless, mass on the tongue base. Microscopically, it had the typical features of schwannoma, the diagnosis being supported by a strong positivity for S-100 protein. Ki67 staining showed low proliferative index (<5%) indicated benign schwannoma. CT-scan of oropharynx demonstrated a well-circumscribed soft tissue density mass, oval shape, connected with the left portion base of tongue, homogenous, and strong enhancement mass. The entire mass was removed in general anesthesia with conservative surgical excision via trans-oral route, histopathologic reported consistent as schwannoma. The patient remained disease-free, without any residual problems. Although the rarity of schwannoma on the tongue base, this case highlights the importance of including it in the differential diagnosis of mass from the tongue base. To the best of our knowledge, our case is only the twentieth of schwannoma at the base of tongue that reported in the literature.

Keywords: Schwannoma, Neurilemmoma, Base of tongue, Lingual

INTRODUCTION

Schwannomas (neurilemmomas) are relatively uncommon, solitary, encapsutaled, slowly growing benign lesions which originate from the peripheral neural sheath of any myelinated nerve.[1] Schwannoma is first reported by Verocay in 1908, who called this benign neurogenic tumour as neurinoma. In 1935, it is proposed that these tumours arise from nerve sheath elements and they are termed neurilemmomas.[2,3] Schwannomas can arise throughout the body, that can originate in any peripheral, autonomic, or cranial nerve, other than the olfactory and optic nerves.[4] Twenty five to forty five percent of schwannomas have been reported to occur in the head and neck region.[5] Only 1% of the schwannomas are located intraoral, with the tongue being the commonest site, followed by buccal mucosa, intra medullary bone of maxilla and mandible, floor of mouth, palate, gingiva, and lips.[1,3,6,7] The tongue as a whole is the most common location for intraoral schwannomas to occur, however, it is quite rare to form schwannomas at the tongue base.[8]

CASE REPORT

A 22 year old woman presented to the Department of Otorhinolaryngology Head and Neck Surgery at Dr. Sardjito Hospital in Yogyakarta for evaluation of foreign body sensation in throat. There was a slowly growing mass on the tongue base for the past two years, which followed by difficulty on swallowing of solid food, but not liquids. The patient denied pain or loss of taste sensation. There was change in voice, which became muffled. The patient also complained snoring while

sleeping. The patient denied daytime sleepiness, irritability or short temper, morning headache or forgetfulness. The patient denied bleeding, difficulty in breathing, and weight loss. The patient also denied other masses, history of injury, chronic irritation, or exposure to radiation.

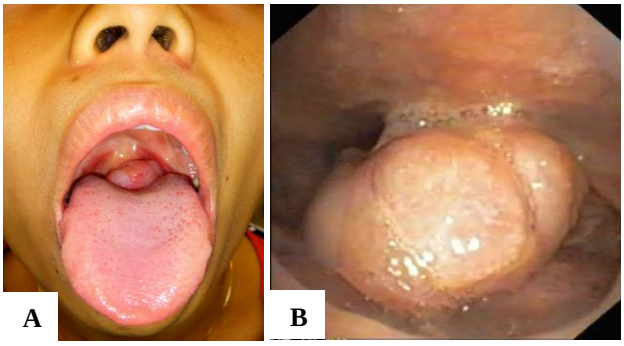

Figure 1: (A) Clinical appearance of schwannoma on the tongue base; (B) Direct flexible laryngoscopy view.

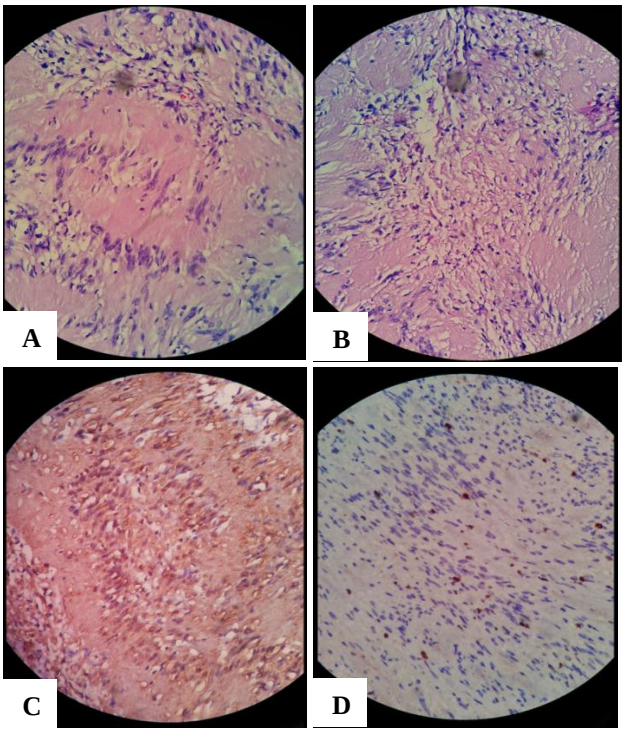

Figure 2: Photomicrography showing the typical characteristic of the schwannomas. (A) Antoni A tissue with palisading nuclei and Verocay bodies (hematoxylin & eosin stain, 100 X); (B) Antoni B tissue with disorderly arranged cells (hematoxylin & eosin stain, 100 X); (C) Immunostaining of the S-100 protein- strong positivity in the tumour cells (100 X); (D) Ki67 staining- positivity in tumour cells nucleus <5% from all tumour cells (100 X).

ENT examination revealed a solitary, painless, lobulated mass that was round smooth bulging arisen from the base of tongue and filled the oropharynx. No bleeding to touch, nor attachment to surrounding tissue. The tongue movement was unrestricted. The adjacent mucosa was normal (Figure 1). Simple gustatory testing to sweet, sour, salt, bitter yielded normal results. The dentition and rest of oral cavity was normal. The appearance and movement of the vocal folds were normal. There was no palpable cervical lymphadenopathy or cranial nerve palsy.

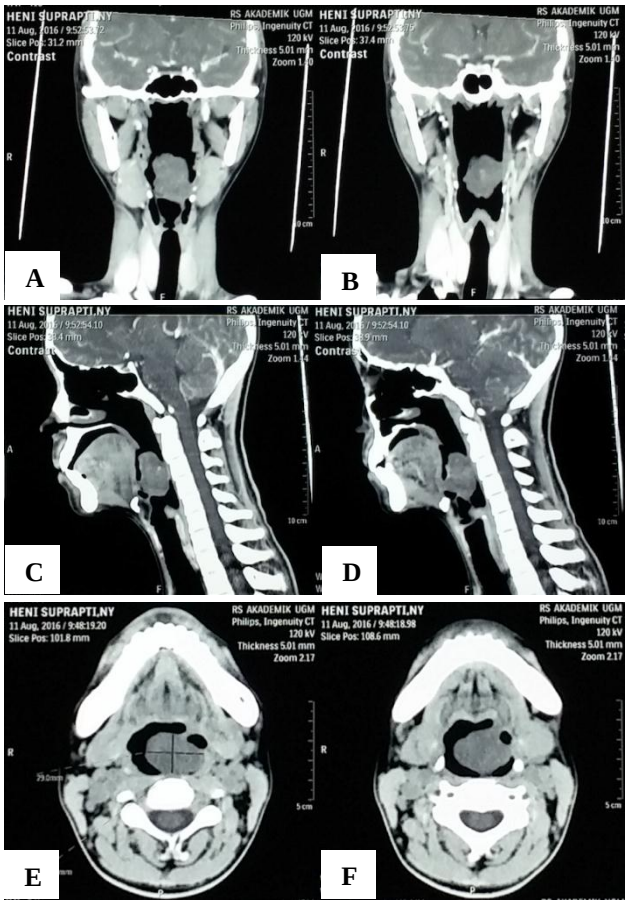

Figure 3: CT-scan of oropharynx. (A, B) Coronal view; (C, D) Sagital view; (E, F) Axial view.

Histopathologic examination from the mass showed outer aspect of capsule coated by stratified squamous epithelial cell with well circumscribed mesenchymal tumour, consisted of two different patterns: Antoni A and Antoni B areas. Hypercellular areas (Antoni A) contained monomorf cells, medium size, oval shape, spindle, smooth chromatin. In between nuclear palisading constitute verocay bodies. Hypocellular areas (Antoni B) contained loose stroma, microcystic. No sign of malignancy. S-100 protein immunohistochemical staining showed strong positivity in the tumour cells. Ki67 staining showed positivity in tumour cell nucleus <5% from all tumour cells. The histologic examination and immunohistochemistry confirmation with S-100 concluded that it was schwannoma (Figure 2).

Contrast-enhanced computed tomography (CT) of oropharynx demonstrated a well-circumscribed soft tissue density mass, oval shape, connected with the left portion

base of tongue, homogenous, and strong enhancement mass (27.5 HU to 50.4 HU). The capsule was well delineated and intact (Figure 3).

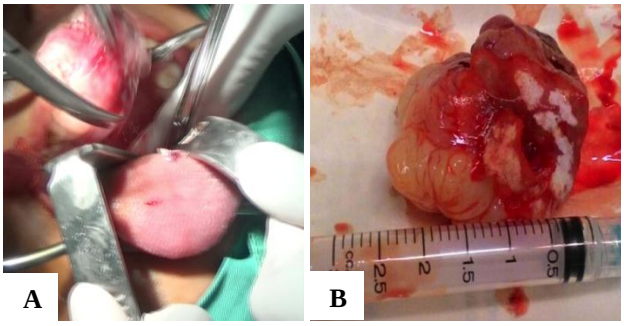

Figure 4: (A) Trans-oral excision of the mass, (B) Gross appearance of the mass.

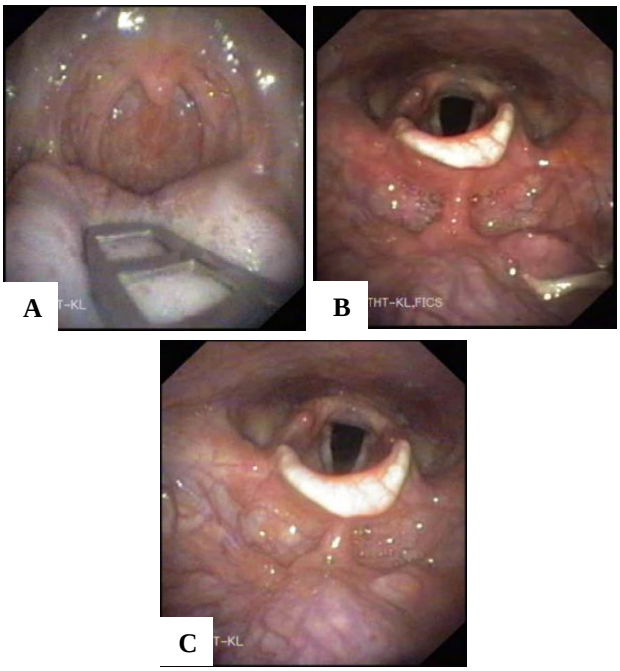

Figure 5: (A, B) Follow up 1 week post-operative, (C) Follow up 6 months post-operative.

The management was conservative surgical excision via trans-oral route with general anesthesia. The mass was rubbery, pedunculated, well circumscribed, and encapsulated. The tumour was seen to be attached to the left tongue base by a pedicle. The mass measured 4.5×4.0×3.0 cm (Figure 4). After it was removed, the wound was sutured with primary closure, without any flap. The specimen was sent for histopathological and immunohistochemical examination, then reported consistent as schwannoma. After the operation, she was completely relieved of all foreign body feeling in the throat, dysphagia, muffled voice, and snoring symptoms. She was discharged home one day postoperative. After 6 months post operatively, the patient remained disease-free and without any residual problems (Figure 5).

DISCUSSION

Nerve sheath tumours which originate from peripheral nerves are two types, neurofibroma and schwannoma. It can originate in any peripheral, autonomic, or cranial nerve, except the olfactory and optic nerves. Neurofibroma is benign neoplasm derived from fibroblasts of the perineurium, which composed of neurites, Schwann cells, transitional cells, and fibroblasts with collagenous and myxoid matrix, whereas schwannoma originate from Schwann cells of nerve sheath which cover myelinated nerve fibre.[1] Schwann cells form a thin barrier around each extracranial nerve fiber of motor and sensory nerves and wrap larger fibers with myelin sheath to enhance nerve conductance.[9,10]

The possible etiologies of the tongue schwannoma are spontaneous growth, external injury, chronic irritation, or exposure to radiation; but the certain cause of the tongue schwannoma is unknown.[7,11] Loss of function of the merlin protein (coded by gene *NF2*) appears to be the cause of the appearance of schwannomas. This protein has complex functions that are essential to various pathways in the nucleus and in the cell membranes. Loss of merlin function in the nucleus leads to an increased expression of membrane proteins, including integrins and growth factor receptors. These growth factors are usually inhibited by cell-cell contact. The absence of merlin favors their activation, stimulating mitogenic and survival pathways, and favoring a lack of cell polarization. These alterations cause schwannoma cells to be unable to bind to an axon.[12] As schwannomas grow larger, they tend to outgrow the blood supply and may undergo cystic degeneration in some areas.[13]

Schwannomas can occur at any age, often seen between 2nd and 6th decades of life, very rare in below 10 years of age, seen equally in both sexes.[2,14,15] They can reach up to considerable size, although they usually remain small. Twenty five to forty five percent of schwannomas have been reported to occur in the head and neck region.[5] These tumours are found with the highest frequency in the superior vestibular division of the VIII nerve, and more than 90% of intracranial schwannomas involve the VIII cranial nerve. After vestibular schwannoma, parapharyngeal space is the most common site. In parapharyngeal space it can arise from IX, X, XI, XII and 3rd division of V nerve.[1] Schwannomas do not arise in the olfactory (I) and optic (II) nerves because these nerves lack Schwann cells.[4] Only 1% of the schwannomas are located intraoral, with the tongue being the commonest site, followed by buccal mucosa, intra medullary bone of maxilla and mandible, floor of mouth, palate, gingiva, and lips.[1,3,6,7] The tongue as a whole is the most common location for intraoral schwannomas to occur, however, it is quite rare to form schwannomas at the tongue base.[8] Two-thirds of tumours arose in the tongue, with the remaining one-third affecting the tongue base.[7] So far, only about 19 cases of schwannomas have been reported

at the base of tongue to the best of our knowledge.[2,3,5,7,11,14,16]

Schwannomas initially appear as slowly growing, generally asymptomatic tumours. Neurogenic deficits do not always correlate with the nerve from which the neoplasm arises, and many patients are asymptomatic. The average schwannoma size in the asymptomatic groups was 18.2 versus 33.0 mm in the symptomatic group, such as throat discomfort, dysphagia, and voice changes.[14] Patients become symptomatic when a mass affects a vital position within a nerve, causes pain and paresthesia, or when it becomes large enough to cause dysphagia or other physical symptoms.[4] Moreover, if the schwannomas arises in the posterior two-thirds of the tongue, they are more likely to produce debilitating symptoms.[14] This patient presented with foreign body feeling in the throat, dysphagia, change of voice, and snoring.

The definitive diagnosis of schwannoma requires histopathologic examination. Cytology may help in differentiating benign and malignant tumours of soft tissue, but rarely accurate in the diagnosis of neural tumours.[17] Histologically, schwannomas are well circumscribed and encapsulated, may show cystic degeneration.[1] Depending on the morphological variant, schwannomas can be classified into conventional, cellular, plexiform, and melanotic.[12,18] Diagnostic features include Antoni A and Antoni B areas. Antoni A areas are solid with Schwann cell lying in rows resulting in palisading of nuclei. Free bands of amorphous substance between rows of nuclei constitute the verocay bodies which under electron microscope, appear to be composed of thin cytoplasmic processes with small amount of collagen and basal laminar material showing frequent redoubling. In Antoni B areas, spindle or oval cells are arranged haphazardly in a loose matrix, with interfibrillar vacuolated nuclei.[1] These findings clinched the diagnosis of schwannoma in this patient.

In addition to these characteristic patterns, the diagnosis is aided by immunohistochemical markers. The protein S-100 is found on the supporting cells of both central and peripheral nervous systems.[8] Schwannomas usually showed intense immunostaining for S-100 (particularly Antoni A areas), which may help to distinguish peripheral nerve sheath neoplasms from other tumour.[3,14,19] In this patient, immunohistochemistry showed strong S100 positivity in the tumour cells. Hence, the diagnosis schwannoma was confirmed. Ki67 staining showed low proliferative index (<5%). It showed a benign schwannoma arising on the base of tongue.[2]

Contrast-enhanced computerised tomography is helpful in the differential diagnosis, contrast-enhanced MRI is especially useful in cases of intracranial and intraorbital involvement.[4] High resolution CT was recommended to determine the size and extent of the tumour, to demonstrate degree of tumour vascularity, and to differentiate between benign and malignant lesions.[5] The majority of schwannomas present as a well circumscribed, homogeneous soft-tissue mass that exhibits low density on CT images, high signal intensity on T2-weighted images, and strong enhancement on CT and MR images, as some of the schwannomas are very vascular. According to previous case reports, Schwannomas arising in the base or floor of the tongue tend to show homogeneous enhancement patterns, while those arising in the tongue tend to show heterogeneous enhancement patterns. The differences in the imaging features of the tumours at these two sites may be caused by the compositions of the Antoni A and B areas, it was suggested that the composition of Antoni A and B areas depends on the location of the tumour. The tumour at the floor of tongue showed abundant Antoni A areas compared with the tumour of the tongue.[20,21] When the CT scans of schwannomas show inhomogeneous areas, a malignant change in the neurogenic tumour may be suspected.[16] In this patient, contrast-enhanced computed tomography (CT) of oropharynx demonstrated a well-circumscribed soft tissue density mass, oval shape, connected with the left portion base of tongue, homogenous, and strong enhancement mass.

The origin of nerve was difficult to identify. When the nerve of origin is small, its association with the tumour may be difficult to demonstrate. On the other hand, if the nerve of origin is large, then the nerve fibers are splayed out over the outer aspect of the capsule rather than being incorporated in the tumour mass.[3,14] Only 50% of schwannomas have a direct relation with a nerve.[7] In more than 50% of intraoral lesions, it is not possible to differentiate the nerve origin whether from glossopharyngeal, hypoglossal, or lingual nerve.[2,7,11] In our case, the origin of schwannoma on base of the tongue could not be determined in consideration of the outer aspect of capsule coated by stratified squamous epithelial cell without splayed out of nerve fiber, the tongue movement and taste sensation was normal.

The schwannoma is clinically undistinguishable from other reactive and neoplastic swelling, which can sometimes mislead the diagnosis. The differential diagnosis of a soft tissue submucosal lesion in the oral cavity is extensive, its includes neurofibromas, mucoceles, fibroepithelial polyps, fibromas, lipomas, leiomyomas, rhabdomyomas, granular cell tumours, retention cysts, hemangiomas, lymphangiomas, pyogenic granulomas, and benign salivary gland tumours.[13,14,22,23] The differential diagnosis must be made relative to malignant tumours based on the speed of growth and clinical appearance, includes squamous cell carcinoma and angiosarcoma.[1,2,10,15,18] When the lesion presents as an exophytic mass, the possibility of a papilloma, aberrant thyroid tissue, and hypertrophied lingual tonsil tissue must also be included.[22] Histologically, the differential diagnosis includes all spindle cell lesions at base of tongue including sarcamatoid squamous cell carcinoma, smooth muscle, fibroblastic, lipomatous, and

rarely salivary gland tumours.[2] A detailed clinical history, physical examination, histopathology, and radiological assessment help in differentiating schwannomas in such cases.[1]

Schwannomas extremely rare undergo malignant transformation.[4,5,15] In malignant schwannoma, the tumour contains unequivocal malignant foci as manifested by the presence of increased cellularity, numerous mitoses, pleomorphism of spindle cells, anaplastic cells, and invasiveness.[3,14,24] Malignant etiologies should always be considered, but are unlikely to present with the slow clinical course that is typical of a schwannoma.[3,14] To date, no cellular schwannoma has metastasized.[18]

The primary objective management of schwannomas is complete resection.[1,16] The non-encapsulated form requires a margin of normal tissue and careful separation from the involved nerve which necessary to preserve normal function.[2] The surgical approach of trans-oral excision is the most common preferred, especially in tongue base schwannomas, with trans-hyoid, suprahyoid, and submandibular approach can be used in large tongue base tumours if needed. The choice of which is based on tumour size and location. CO_2 laser, coblation, bipolar cautery, or conventional tools may be used for excision intraorally.[14,16,23] Radiation therapy is not indicated because schwannomas exhibit a high degree of radio-resistance.[1,4,5,16] In this patient, the entire mass was removed with conservative surgical excision via trans-oral route.

To avoid recurrence, the surgeon should perform a complete excision, which is usually simple in view of the encapsulated nature of this tumour.[4,16,18] Incomplete removal result in recurrence which should be treated with repeat excision.[1] The postoperative prognosis is very good. The length of follow-up remains debatable, long-term follow-up is not necessary if clearance margins are tumour-free, but if clearance is inadequate for any reason, the patient should annually review.[4]

CONCLUSION

In conclusion, although the rarity of schwannoma on the tongue base, this case highlights the importance of including a nerve sheath tumour in the differential diagnosis of mass from the tongue base. To the best of our knowledge, our case is only the twentieth of schwannoma at the base of tongue to be reported in the literature.

Most of the tongue base tumours are silent early until the mass large enough to cause symptoms. In our case, the patient initially had foreign body sensation in the throat, which progressed to dysphagia, change of voice, and snoring. ENT examination revealed a solitary round smooth bulging, painless, lobulated mass that was arisen from the base of tongue. The final diagnosis done after histopathological examination, aided by immune-histochemical analysis. Trans-oral excision allows for complete removal of this tumour 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 tumours. The chance of malignant transformation of these tumours is exceedingly unlikely.

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