CASE REPORT

Ancient Schwannoma of the tongue

Mahima V. Guledgud¹, Saikrishna Degala², Karthikeya Patil¹, Meghana Maruthi¹

¹Department of Oral Medicine & Radiology, J.S.S. Dental College & Hospital, J.S.S. University, Mysore, Karnataka, India, ²Department of Oral & Maxillofacial Surgery, J.S.S. Dental College & Hospital, J.S.S. University, Mysore, Karnataka, India

Abstract

Ancient Schwannoma is a rare Schwannoma variant which is a painless, slow-growing benign neoplasm of Schwann cell origin. It is relatively a rare tumor in the head and neck region. We report a case of ancient Schwannoma of the tongue in a 38-year-old female patient, who presented with a slowly progressing painless mass over the tongue. Investigatory impression of ultrasonography and fine-needle aspiration cytology of this lesion was suggestive to be a benign tumor of muscle/nerve origin. An excisional biopsy of the mass with primary closure was done. A final diagnosis of ancient Schwannoma was given based on histopathological examination.

Keywords: Ancient Schwannoma, neurilemmoma, Schwannoma, tongue

Introduction

Schwannoma, otherwise known as neurilemmoma, is a benign neoplasm originating from Schwann cell.¹ It can originate from any nerve covered with Schwann cell sheath, which includes cranial nerves (except for the first and second cranial nerves), spinal and autonomic nervous system.²,³ It is relatively uncommon although about 25-48% of all cases manifest in the head and neck region.¹ The most common type is acoustic neuroma affecting the vestibulocochlear nerve.⁴ However, about 1-2% occurs intraorally with tongue being the most common site followed by the palate, floor of mouth, oral mucosa, and mandible.⁵,⁶

Ancient Schwannoma is an uncommon Schwannoma variant which is a slow growing encapsulated painless benign neoplasm.³

Case Report

A 38-year-old known hypotensive female patient presented to us with an asymptomatic slow growing swelling on the tip of the tongue crossing the midline, of 20 years duration. Initially, it appeared as a pea sized swelling when the patient was 2 months pregnant at the age of 18 years and gradually increased to the present size. It was painless and there was a loss of clarity of speech. History of occasional bleeding from the swelling was elicited.

Clinical examination revealed a conical soft tissue mass measuring about 5 cm × 3 cm × 2.5 cm in its greatest dimension on the dorsal tongue extending inferiorly onto the ventral surface. Dorsally, it appeared pink and was velvety in texture, firm, and hard in consistency except for the tip of the mass which was smooth and soft. Ventrally, the mass was smooth, lobulated, firm, and cystic not fixed to the overlying tissue with dilated vessels [Figures 1 and 2]. It was non-tender, uncompressible, and irreducible on palpation. There was no significant weight loss and lymphadenopathy. There was no evidence of similar type mass elsewhere in the body.

Routine blood investigation revealed, hemoglobin and red blood corpuscle count to be 9.8 g% and 2.9 million cells/mm³, respectively, giving an impression as moderate anemia. Ultrasonography [Figure 3] and fine-needle aspiration cytology (FNAC) were suggestive of benign tumor possibly of muscle/nerve origin. The patient underwent excisional biopsy of the mass under general anesthesia, and the specimen was sent for histopathological examination.

The patient was taken up for the excisional biopsy under general anesthesia. Nasoendotracheal intubation was done. The demarcation of the incision was performed with methylene blue. The tongue was pulled out by placing two black braided silk...
sutures 3-0 which was fixed to the surgical field, and symmetry of both the sides was maintained. Local infiltration of lignocaine 2% with adrenaline in the ratio of 1:80,000 was infiltrated around the mass. As the lesion was present on the anterior one-third of the tongue extending from the dorsal to the ventral surface, Harris, Blair, Hendrick incision was used which allowed the excision of the mass as a whole. Full thickness incision was made by the blade No. 15. Electrocautery was used for better hemostasis. The mass was excised, and the rest of the tongue was sutured by absorbable Vicryl 3-0 round body in a layered fashion. During the surgical procedure, copious amount of saline irrigation was used.

The excised specimen [Figure 4] was sent for histopathological examination and revealed spindle cells with elongated wavy nuclei and fibrillar cytoplasm. Prominent nuclei, palisading with verocay bodies among Antoni A and B tissues were seen with evidence of myxoid degeneration and cystic changes. Thereby, a final diagnosis of ancient Schwannoma was given [Figure 5].

The patient was reviewed for 3 weeks post-operatively and showed no signs of paresthesia of tongue or lip. Tongue shape was maintained [Figure 6]. Improved motor movements, speech and swallowing were observed. No alteration in taste sensation.
was noted. The patient was referred for phono-audiological evaluation for speech rehabilitation.

Discussion

Schwannomas are encapsulated nerve sheath benign neoplasms composed of Schwann cells. Theodor Schwann (1810-1882), a German anatomist, physicist and co-founder of the cell theory discovered these cells which are derived from ectomesenchymal neural crest cells during the 4th week in utero. In 1908, José Verocay discovered Schwannoma. Etiology is said to be unknown; however, it is assumed that the lesion arises from the propagation of Schwann cells at a point inside the perineurium, which causes compression and displacement of the surrounding normal nerve. It is most often difficult to find out the nerve of origin. It is not possible to differentiate between tumors of lingual, hypoglossal, and glossopharyngeal nerves in about 50% of intraoral cases. The tumor can be seen associated with Von Recklinghausen’s disease.

Intraoral Schwannomas account to about 1-2% of all head and neck tumors. It can present at any age, however, most often presents at second to the fifth decade of life with no sex predilection. The most common site involved is the tongue followed by the palate, floor of mouth, oral mucosa, and mandible. Schwannomas are usually asymptomatic lesions which are slow growing, smooth surfaced, solitary, and of variable size. They usually present as a painless mass, although pain and paresthesia can occur. Other symptoms such as dysphagia, disturbances in mastication and phonation, and Tinel’s sign (an electric like shock when the affected area is touched) might be present depending on the size of the tumor and location. In our case, the tumor was large in size located on the tongue, and there was a disturbance in phonation.

Considering the above-mentioned features, neurofibroma, traumatic neuroma of long standing duration, fibroma, lipoma, fibrolipoma, rhabdomyoma, and leiomyoma can be considered in the differential diagnosis. In some cases, squamous cell carcinoma, sarcomas, and glandular malignant processes may present with similar clinical features.

Diagnostic investigations include ultrasonography, computed tomography, magnetic resonance imaging (MRI) scan, and FNAC. On ultrasonography, tumor presents as well-defined hypoechoic nodules which are oval or lobulated with posterior acoustic enhancement, and on color Doppler ultrasonography it appears hypervascular. MRI scan reveals Schwannoma of the tongue to be isointense to the muscles on the T1-weighted images, whereas homogeneously hypointense on the T2-weighted images. Other features on MRI are smooth, well-demarcated borders without invasion into the surrounding muscles. In our case, we did not perform MRI because the mass was well circumscribed and demarcated. This feature was confirmed on ultrasonography. Peripheral smear of FNAC appears as a benign mesenchymal tumor.

For excision of the tongue Schwannomas, various kinds of incisions can be used. Each can lead to a different degree of exposure, infection rates, residual tongue function, and post-operative appearance. Incisions such as Pichler incision, Kole, Davalbakta, and Lambert incision, Morgan et al. and Kacker et al. incision could be used but due to the size and extent of the lesion being reported, the Harris, Blair, Hendrick “V” shaped incision was employed. It gave easy access for complete excision of the mass with minimum removal of the uninvolved tongue tissue. Along with the mass, tongue muscles such as transverse and inferior longitudinal muscle fibers were also removed which are present at the tip of the tongue and at the anterolateral border of the tongue. In cases where only tip of the tongue is involved, Pichler incision is commonly employed and for tongue mass which is extending anteroposteriorly, the Kole, Davalbakta, and...
Lamberty incision and Morgan et al. and Kacker et al. incision is used.\(^\text{[12-15]}\)

Histopathological examination is the only definitive key for final diagnosis. The tumor tissue comprises of Antoni A and Antoni B type cells. Streaming fascicles of spindle-shaped Schwann cells is characteristic of Antoni A cells which palisade around central, acellular eosinophilic areas known as Verocay bodies. Antoni B cells are less cellular and less organized, and spindle cells are seen within the loose, myxomatous stroma.\(^\text{[1]}\)

The term “ancient Schwannoma” was coined by Ackerman and Taylor in 1951. It is an extremely rare Schwannoma variant which is a slow growing tumor of Schwann cell origin of a long duration. Although it cannot be clinically differentiated from Schwannoma but histopathologically shows evidence of degenerative changes such as hemorrhage, hemosiderin deposits, hyalinization, inflammation, fibrosis, and nuclear atypia which precisely indicate aging of the tumor. They are benign, slow growing with rare malignant transformation.\(^\text{[3,10]}\)

The treatment of choice is complete excision of the tumor. The recurrence rate of Schwannoma of the tongue is said to be low; however, the recurrence rate of vestibular Schwannoma has been reported to be about 0.05% for enlarged translabyrinthine approach, 0.7% for retrosigmoid approach and 1.8% for middle cranial fossa approach.\(^\text{[16]}\)

Conclusion

To conclude, the clinical appearance of ancient Schwannoma in this case was one of its kinds - a 38-year-old female patient had this slow growing tumor in her oral cavity for 20 years! Ancient Schwannomas are relatively rare tumors of long duration occurring in the oral cavity which can present between second to fifth decades of life with no sex predilection with the most common site being the tongue. It can be differentiated with Schwannoma only by histopathologically examination showing degenerative changes. Complete excision of the tumor is the definitive treatment with good prognosis. It has been reported to have low recurrence rate and least incidence of malignant transformation.

References


