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A rare case of LINGUAL Schwannoma

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Tumor; Intaoral; Hemangioma

1. Abstract

1.1. Background: Schwannoma of tongue is a benign nerve sheath tumor arising from Schwann cells. Only 1-12% of all Schwannomas shows intaoral occurrence, tongue being the commonest site in the region.

1.2. Case Report: A 22-year-old female presented with a swelling of 3*2*3 cm over the left lateral border of tongue. Movement of tongue was normal. Cytological evaluation revealed a benign mesenchymal lesion and CE-MRI showed a well circumscribed lesion of benign etiology. Transoral excision was done. Histopathological examination revealed a lingual Schwannoma.

1.3. Conclusion: Lingual schwannomas are rare tumors which presents with varying location and clinical presentations, so should be considered a differential diagnosis while evaluating a tongue mass.

2. Introduction

Schwannoma also known as neurilemma is a benign nerve sheath tumour. It was first identified by Virchow in 1908. These tumours can emerge from any nerve covered with a myelin sheath, including the cranial nerves (with the exception of optic and olfactory nerves), the spinal nerves and the autonomous nervous system [1] About 25-45% of all schwannomas occur in the head and neck [2]. Around 1-12% of these occur intraorally [3,4] with the tongue being the most common site [4,5]. Clinically schwannomas are usually benign, painless solitary encapsulated masses. They present as a diagnostic dilemma because of its rare occurrence and variable location. The clinical presentation of intraoral schwannoma varies from an asymptomatic mass to dysphagia, pain, dysphonia, paraesthesia and snoring. We are reporting this case so that such a rare differential is kept in mind while evaluating a tongue mass by a clinician. It will further aid in timely and appropriate management of such cases.

3. Case Report

A 22-year-old female presented with a swelling over the left lateral border of tongue since 10 years. No history of pain, difficulty in swallowing, weight loss, trauma or loss of taste disturbance was present. On Intraoral clinical examination a smooth globular swelling of size 3x2x3cm involving left lateral border of tongue was present which was non tender, firm in consistency with well-defined margins (Figure 1).

Tongue movements were normal. No cervical nodes were palpable. Rest of the ENT examination was normal.

Based on the clinical finding, differential diagnosis of lipoma, lymphangioma, benign capillary hemangioma, papilloma, fibroma, benign mesenchymal lesion and neurofibroma were made.

It was further evaluated with FNAC which was suggestive of a benign mesenchymal lesion.

The CEMRI of tongue showed a well-defined oval soft tissue lesion of approx size 28 x 32 x 20 mm in the tongue on left side, appearing hyperintense on T2WI and hypointense on T1WI. Lesion was seen displacing the lingual septum towards the right side without any evidence of invasion of floor of mouth and showed homogenous post contrast enhancement. The features were likely suggestive of a benign etiology (Figure 2).

After pre anaesthetic check-up patient was planned for transoral excision of the mass under general anaesthesia. Incision was given on the ventral surface of the lesion. Mucosal flap was raised. Mass was dissected from surrounding attachments (Figure 3) and

removed in toto (Figure 4) Haemostasis was attained and incision was closed in 2 layers.

Histopathological examination of the surgical specimen revealed a well circumscribed biphasic spindle cell tumour with hyper and hypo cellular areas with verocay bodies. Immunohistochemistry showed positivity for S100 protein, confirming the diagnosis of schwannoma (Figure 5 and 6).

Patient is under follow up and doing well (Figure 7). There is no sign of recurrence.

Figure 1: Mass on left lateral border tongue

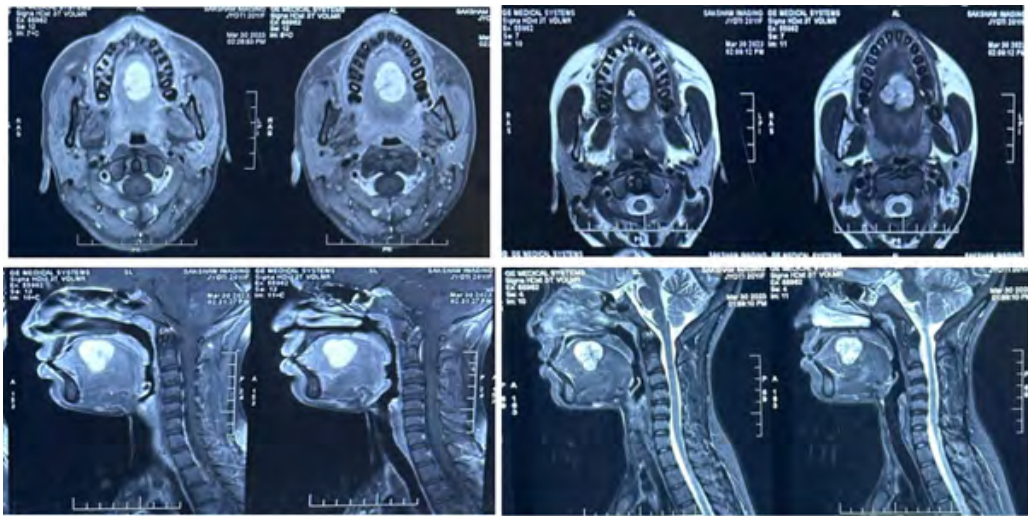

Figure 2: Axial and Sagittal cut of CEMRI Scan showing the tumor

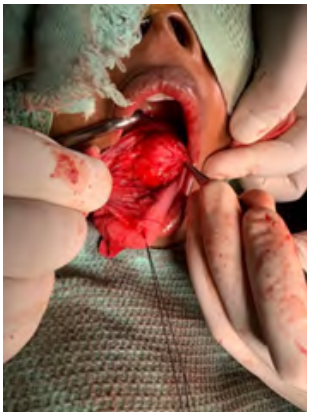

Figure 3: Mass dissected from attachments

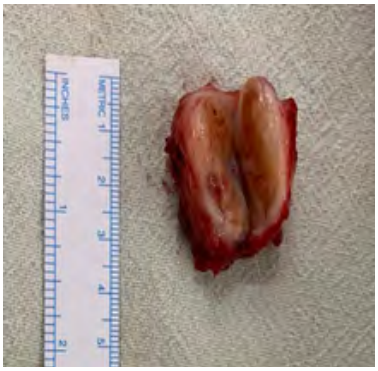

Figure 4: Specimen delivered in toto

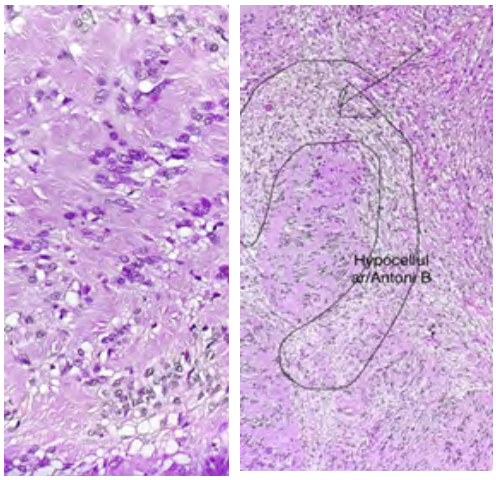

Figure 5 and 6: Microscopic examination showing elongated nuclei and hypercellular and hypocellular areas

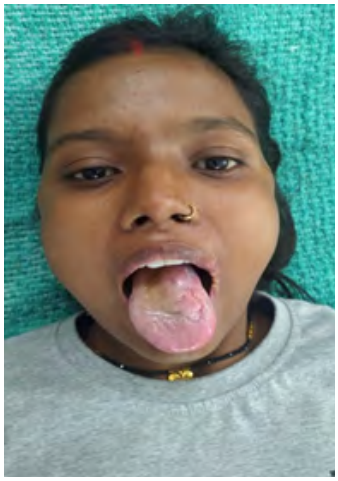

Figure 7: Post-op follow-up after 6 weeks

4. Discussion

Schwannomas arise from Schwann cell which are responsible for formation of myelin sheath.

25-45% tumors arise in the head and neck out of which 1-12% occur intra orally with tongue being the most common site, followed by the palate, floor of mouth, buccal mucosa, gingiva, lip, and vestibule [7,8 ,9]. Lingual schwannoma shows no gender predisposition but high incidence has been found between second and fourth decade. In our case too the patient presented in the third decade [10]. The tongue is innervated by various nerves like lingual, hypoglossal and glossopharyngeal however, in 50% of the cases it is not possible to identify nerve of origin[21]. In our case since mobility of tongue and taste sensation was not affected hence the nerve involved could not be identified.

The literature reveals that patients with lesion size upto 2.4 cm are generally asymptomatic. However, when the mass is located in posterior one third of tongue or exceeds 3 cm in size anteriorly, they are usually symptomatic [16]. In our case patient had no complains as the size of lesion was 3*2*3cm present along the left lateral border.

MRI is the imaging modality of choice for tongue lesions. It allows for accurate estimation of mass size and its location in relation to surrounding structures. Usually, schwannoma does not undergo malignant transformation [14, 15]. In our case too MRI was suggestive of a benign lesion and helped us to rule out further differentials.

Schwannomas are usually treated by surgical excision [17]. Transoral excision is the most commonly used approach. In our case too patient underwent transoral excision under general anesthisa.

More recently, CO2 laser excision has also been used to treat base of tongue Schwannomas [16]. On the other hand, if a mass is located at the posterolateral base, open techniques (such as lip split and submandibular approach) can be used [3, 18]. Recurrences are rare after complete surgical excision [19]. Confirmatory diagnosis is made by histopathological examination.

Histologically, all schwannomas are encapsulated. Two main patterns are observed,

Antoni type A and Antoni type B, both are composed of elongated Schwann cells exhibiting a palisading nuclear pattern and a less dense myxoid pattern respectively.

In our case the biopsy report showed well circumscribed spindle cell biphasic tumour with compact hyper and hypo cellular areas with vertical bodies and elongated nuclei, which gave the impression of a schwannoma. Immunohistochemistry showed diffuse membranous positivity for S100.

Schwannomas carry good prognosis. Our Patient is under regular follow up and has shown no sign of recurrence.

5. Conclusion

Lingual schwannoma is a relatively rare tumor of the head and neck region. Therefore, they should be considered in the differential diagnosis while evaluating intra oral mass. Transoral excision is the preferred treatment. It carries a good prognosis with recurrence being rarely reported.

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