Case Report

Giant Vascular Hamartoma of the Tongue

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Abstract

We present one of the largest lingual hamartomas of the tongue base to have been reported, along with a review of the current literature and a description of the management of this case, as well as insights into the histopathology of the lesion. A 21-year-old woman presented with a mass on the base of her tongue, extending to the vallecula. The mass was found to be over 4 cm and enhancing on computed tomography. The size, vascularity, and site of the lesion merited its excision using the suprahyoid pharyngotomy approach. Histopathology confirmed the mass to be a vascular hamartoma. In reviewing the literature, we encountered 61 reported cases of lingual hamartomas, which are described with a number of pathological variants and sites of occurrence and with different methods of surgical excision. The size, vascularity, and site of the lesion we found merited a different approach from the conventional transoral approach that was used in all of the previous reports. Also, our study agrees with current world literature that histopathological examination plays an important role in the final diagnosis.

Keywords: *lingual, hamartomas, tongue, vascular*

Introduction

Some "tumour-like lesions," such as pyogenic granulomas, hemangiomas, and hamartomas, can involve the anterior tongue and the tongue base. These lesions occur in different age groups and can clinically mimic a malignancy. However, hemangiomas and hamartomas are benign proliferative lesions with no infiltrative or metastatic properties and, unlike true neoplasms, they do not exhibit any unchecked monoclonal tissue proliferation. Hamartomas are composed of a mixture of locally derived mature tissue in differential proportions. The relative abundance of a particular tissue dictates its subtype, behavior, and appearance. These lingual lesions continue to be relatively rare. Indeed, to our knowledge, only two retrospective studies have been published, one describing 18 cases (1) and the other involving 25 oral neurovascular hamartomas (2) of which 13 were found to be on the tongue. Other than these, there are only isolated case reports. Based on the results of a Pubmed search using the keywords "tongue" and "hamartoma", the total number of lingual hamartomas that have been reported in English literature is, to the best of our knowledge, just 61. This includes a case of lingual hamartoma associated with tuberous sclerosis (3). However, all cases associated with Cowden's disease were excluded from the search because this disease exhibits a different set of syndromic manifestations. This report highlights the diagnosis and management of isolated lingual vascular hamartomas. In our case, the size of the lesion was found to be larger than any previously reported hamartomas that involved the base of the tongue with extension to the vallecula. The lesion was excised using the suprahyoid pharyngotomy approach.

Case Report

A 21-year-old female presented in the otolaryngology out-patient department with complaints of dysphagia and a sensation she had had for the last six months that there was a foreign body in her throat. Laryngoscopy revealed a smooth mucosa-covered mass involving the left side of the base of her tongue and her vallecula, and occupying almost all of the oropharynx. Computed tomography (Figure 1) revealed a solid mass measuring 3.9 × 4.5 cm, which arose from

the left posterior third of the tongue, extending to the vallecula, and pushing the epiglottis and the left pharyngeal wall. The mass was well defined and enhancing in delayed venous scans on computed angiography; it was seen to derive its blood supply from the facial and lingual arteries. A biopsy was performed under general anesthesia after a tracheotomy had been performed for anesthetic considerations. During the biopsy, the lesion was found to be particularly vascular because of an intra-operative hemorrhage that occurred. However, no blood vessels were seen on the lesion. The histopathology suggested the possibility of a pyogenic granuloma. The mass was to be excised under general anesthesia and, because of the size of the tumor and its vascularity, the suprahyoid pharyngotomy approach (Figure 2) was chosen. The entire mass was excised and sent for histopathological examination. The diagnosis of a vascular hamartoma was confirmed through the presence of an admixture of mature adipose tissue and muscle, with a predominance of blood vessels of various sizes, all of which were endogenous to the affected region (Figure 3). It took the patient two weeks of intensive swallowing therapy to return to a normal diet and to being able to swallow properly. The patient was successfully decannulated and discharged, and she remains symptom free after one year of treatment.

Discussion

Hamartomas are considered to be benign tumour-like malformations or non-neoplastic developmental anomalies caused by endogenous tissues that are proliferating in a disorganised manner. Most of the lingual hamartomatous lesions that have been reported have been within the pediatric population (1,4–6,10), with a few in the adult population, mostly among females (2, 7–9).

In a review done in 1989 by Takimoto et al. (4), most of these types of lesions were found to be at the base of the tongue in the midline, while the pathological review by Kreiger et al. (1) reported that the dorsal anterior tongue was the most commonly affected area.

Most lingual hamartomas are asymptomatic in nature or associated with only minor symptoms, such as dysphagia or globus sensation. Pediatric cases are often discovered because of feeding difficulties. The symptoms and the reasons for referral to an otolaryngologist often depend on the site and size of the lesion.

Hamartomas are pathologically sub-

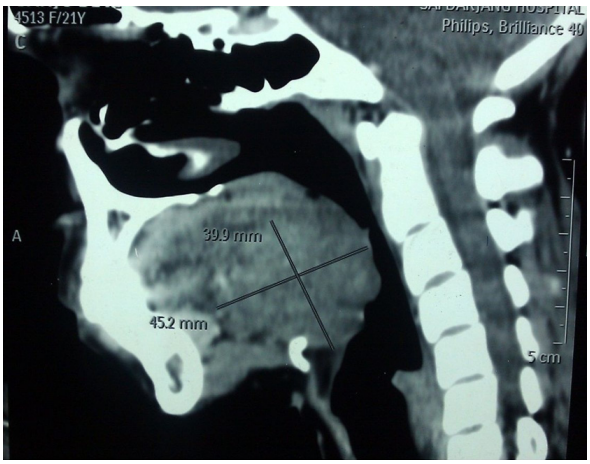

Figure 1: Sagittal computed tomography showing mass at base of tongue.

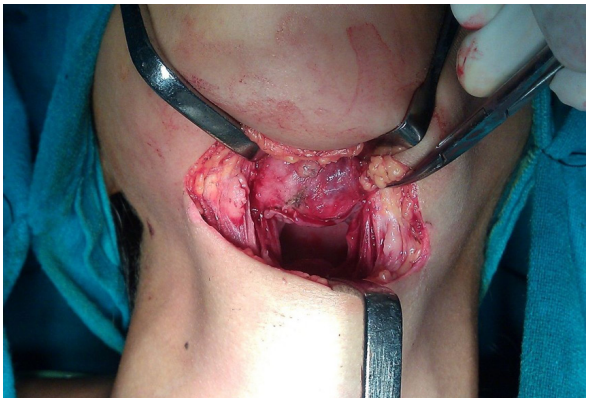

Figure 2: Mass excision using suprahyoid pharyngotomy.

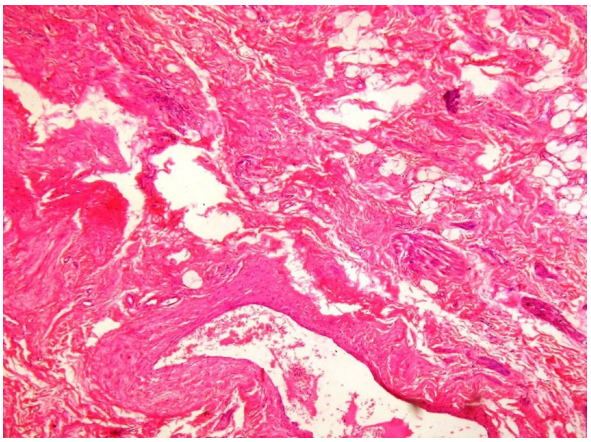

Figure 3: Hematoxylin & eosin stained section (100× magnification) of the lesion showing an admixture of numerous vessels of varying calibres, mature adipose tissue and muscle.

classified, depending upon the relative abundance of a particular endogenous tissue, and the variants described include vascular (1), muscle predominant (1,7,9,10) and adipose tissue predominant (1), as well as the intramuscular capillary variants (8) where numerous thin-walled mature capillaries are interspersed between and around muscle bundles. Takato et al. (6) reported two cases of lingual hamartomas in the pediatric age group and commented on their histological similarities with benign mesenchymomas.

There could be a propensity for the posterior tongue and the tongue base to develop these lesions, and this might be attributed to the embryological development of the posterior tongue (5), which develops from the fusion of two "analges" in the midline. The foramen cecum, which represents the vestige of the fusion of these two "analges," is a slit-like opening called the sinus arcuatus. This opening closes in the developmental stage, but remains prone to multiple factors that can cause irritation and developmental alterations in the area of the tongue base.

Tongue base hamartomas can pose a diagnostic dilemma because they may have various clinical appearances and biopsies from non-representative sites may prove to be misleading or inconclusive, as in our case. The final histopathological diagnosis rests on a thorough examination of the entire specimen and visualization of all the individual elements inside the mass. In addition, in our case, the diagnosis changed from a pyogenic granuloma to that of a vascular hamartoma after examination revealed vessels of varying calibres, adipose tissue and muscle in the resected specimen.

Most hamartomas that have been reported in the literature have been smaller in size, less than 3 cm, and have therefore been removed transorally, either by using conventional methods (5) or by lasers (4). In a report by de Faria et al. (9) of a giant leiomyomatous hamartoma, the maximum dimension noted was 4 cm. We believe that a large hamartoma involving the tongue base and the vallecula can be safely removed using a suprahyoid pharyngotomy approach. This approach provides good exposure to the entire base of the tongue, the vallecula and the epiglottis, and also allows for better vascular control, which is particularly important in lesions that are very vascular. Aside from suprahyoid pharyngotomy, a mandibulotomy with lip-split can provide adequate exposure of the entire oropharynx, but involves a significantly increased risk of morbidity and longer hospital stays. Although major tongue base surgeries, even those for benign lesions, can lead to significant swallowing disturbances and temporary aspiration, these surgeries are amenable to conservative management in the form of dedicated swallowing rehabilitation measures. Complete surgical excision remains the treatment of choice, and recurrences are unlikely and have not been reported.

Conclusion

This case report describes one of the largest lingual hamartomas involving the tongue base and vallecula that has been reported. The size and vascularity of this lingual hamartoma merited a different surgical approach, in the form of suprahyoid pharyngotomy.

Large vascular lesions of the tongue base that are not amenable to being excised transorally can be safely removed using suprahyoid pharyngotomy, which provides for adequate exposure and vascular control.

This paper is in agreement with the current world literature concerning the definite need for the histopathological assessment of an entire surgical specimen, in order to determine the nature of the hamartoma and to achieve a cure through complete surgical excision.

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Conflict of Interest

None.

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Authors' Contributions

Conception and design: NNM, AV, GK
Analysis and interpretation of the data: NNM, AV, GK, SRC
Drafting of the article: NNM, AV, SRC
Critical revision of the article for the important intellectual content and final approval of the article: NNM, AV
Collection and assembly of data: SRC

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