Case Report of a Congenital Leiomyomatous Hamartoma: New Epidemiological Findings and a Review of the Literature

1Arjuna B. Kuperan, MD, 2Neena Mirani, MD, 1Huma A. Qurashi, MD

Abstract

Educational Objective: At the conclusion of this presentation, the participants should better understand the epidemiology of lingual leiomyomatous hamartomas and the differential diagnosis of lesions in the same anatomic location. Participants should also understand the radiologic and histologic findings of this rare lesion and its surgical management.

Objectives: To present a unique case of a lingual leiomyomatous hamartoma, a current review of the literature on this previously unreported lesion.

Methods: Case report and review of the literature.

Results: The differential diagnosis of a congenital midline tongue mass near the foramen cecum includes thyroglossal duct cyst, lingual thyroid, and rarely, lingual hamartoma. We describe an interesting case of a 5-month-old male who presented with a posterior midline tongue mass; ultrasound imaging showed a normal thyroid gland without any lingual component. The mass was excised under general anesthesia, with the defect closed primarily. The patient tolerated the procedure well and was discharged the following day without any complications.

Conclusions: Congenital lingual leiomyomatous hamartomas are thought to be quite rare; however, a comprehensive literature review indicates that they are twice as common as currently reported. It is therefore important to consider this lesion in the differential of congenital midline tongue masses.

Discussion

Limited radiologic data exists regarding imaging of lingual hamartomas; however, a few studies discuss magnetic resonance imaging (MRI) and computed tomography (CT) findings. One study found a leiomyomatous hamartoma to have hypointense signal on T1 weighting and heterogeneous signal on T2 weighting; the corresponding CT of the neck showed minimal contrast enhancement. A report found a lingual hamartoma of unknown subtype to have hypointense signal on T1 and hyperintense signal on T2.1 3

A CT of a lingual hamartoma in conjunction with tuberous sclerosis was found to have intermediate attenuation on postcontrast CT.1 12 The conclusion is that imaging findings are non-specific.

The histopathologic picture of a leiomyomatous hamartoma is described as a proliferation of unencapsulated smooth muscle within the subepithelial region. These muscle bundles are organized irregularly and can be mixed with connective and vascular tissue; in addition, salivary gland or adipose tissue may be present. On greater magnification the presence of fusiform, eosinophilic cells with oval, elongated nuclei is easily recognized.2 9

Chemical staining with hematoxylin and eosin (H&E) (Image 3 and 4) is a standard preparation. Special stains such as Masson’s trichrome (Image 5) can be used to further analyze the smooth muscle. Immunohistochemical staining aids in confirming histopathologic findings; positive staining for muscle-specific actin, smooth muscle actin, and S-100 protein, and desmin is a common feature of these smooth muscle hamartomas.2 11

Once the diagnosis of a leiomyomatous hamartoma is made, the next definitive step is surgical excision. In the comprehensive literature review performed here, there are no reported cases of recurrence after excision.2 11

The largest series reviewing lingual leiomyomatous hamartomas describes a fusiform resection of centrally located lesions with an incision along the long lingual axis; peripheral lesions were excised in a wedge fashion. For small lesions exceeding 2 cm, primary closure is generally acceptable.2 With no reported recurrences the argument for even small margins is weak despite the unencapsulated nature of these lesions.2 11

Conclusions

A leiomyomatous hamartoma is a rare lesion but must be considered in the diagnostic workup of midline tongue masses near the foramen cecum. The 8 reported cases we report here is the largest number histologically confirmed leiomyomatous lesions to date, with an equal gender predilection in contrast to previous reports. CT and MRI findings are highly variable and non-specific.

The limited data that exists. Histopathologic confirmation of this lesion is generally considered with standard H&E histochemical staining but can be confirmed when in doubt with immunohistochemical stains for smooth muscle actin and S-100 protein. Definitive management is surgical excision with a primary closure of the defect, with no reported recurrences to date.

References