Leiomyomatous Hamartoma of the Tongue in an Infant: A Case Report

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ABSTRACT

A case of leiomyomatous hamartoma in the postmedian region of the dorsum of the tongue in a 3-year-old boy is reported. This lesion had been noticed at about 1 year of age but was left untreated. The intensity of the inner region of the mass was homogenous and similar to that of the surrounding tongue muscle on both T1- and T2-weighted images of an MRI. The mass was diagnosed as benign tumor of the tongue and resected. Histopathologically, nodular overgrowth of spindle cells containing eosinophilic cytoplasm was noted in the submucoepithelial connective tissue. In immunohistochemical staining, spindle cells were negative for S-100 protein and positive for vimentin and -SMA, suggesting that these cells were derived from smooth muscle. In the 10 months after surgery, there has been no recurrence of the lesion. To our knowledge, only 26 cases (including the present case) have been diagnosed histopathologically as leiomyomatous hamartoma in the oral cavity between 1945 and 2009. Clinical features, differential diagnosis, and treatment are discussed herein.

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KEYWORDS: LEIOMYOMATOUS HAMARTOMA, TONGUE, INFANT

Hamartoma is a congenital mixed abnormality in which one or several tissue components grow excessively in a normal organ.[1] This condition can occur in many body regions, including the lung, spleen, liver, pancreas, and kidney,[2,3] but the incidence in the oral cavity is low,[1-3] and leiomyomatous hamartoma rarely develops in the tongue.[3,4] We encountered a rare case of leiomyomatous hamartoma in the postmedian region of the dorsum of the tongue in a 3-year-old boy. The purpose of this paper was to report a case of leiomyomatous hamartoma of the tongue and to review the associated literature.

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CASE REPORT

A 3-year-old Japanese boy was brought to the Department of Oral Surgery at the hospital of Mie University School of Medicine, Mie, Japan in April 2009, with a chief complaint of a mass in the tongue. The patient had been receiving orally admistered treatment for asthma since she was 2 years old (Kipres Fine Granules 4 mg, once a day). A mass close to the median terminal sulcus of the tongue was noticed at approximately 1 year old, and the infant was examined by a pediatrician, but no symptoms were observed and the mass was left untreated. It did not remit, however, and the infant was referred to our department for close examination and treatment. A whole body examination revealed no problems. A painless mass with a clear boundary measuring approximately 10 mm was present medially and anterior to the dorsal lingual terminal sulcus. The tumor was pediculated, elastic, and soft, and the surface was smooth and was the color of the mucosa (Figure 1). No eating or speech disorder was noted.

An MRI revealed a mass of 12 mm in diameter in the corresponding region. The intensity of the mass' inner region was homogenous and similar to that of the surrounding tongue muscle on both T1- and T2-weighted images (Figure 2). The position of the thyroid gland was normal.

The mass was diagnosed as a benign tumor of the tongue and resected under general anesthesia. The excised specimen measured 13×12 mm and was elastic and soft. The cross-sectional surface was solid and whitish-yellow with no capsule (Figure 3). Histopathologically, the superficial layer was covered with stratified squamous epithelium. Nodular overgrowth of spindle cells containing eosinophilic cytoplasm was noted in the submucoepithelial connective tissue, and fibrous, adipose, and minor salivary gland tissues were present around the lesion. There were no findings, however, that suggested tumorous proliferation (Figure 4). Viewed with immunohistochemical staining, spindle cells were negative for S-100 protein and positive for vimentin and α-SMA, suggesting that these cells were derived from smooth muscle (Figure 5). Based on these findings, leiomyomatous hamartoma was diagnosed. In the 10 months after surgery, the patient has had no speech, eating, or swallowing disorders and there has been no recurrence of the lesion.

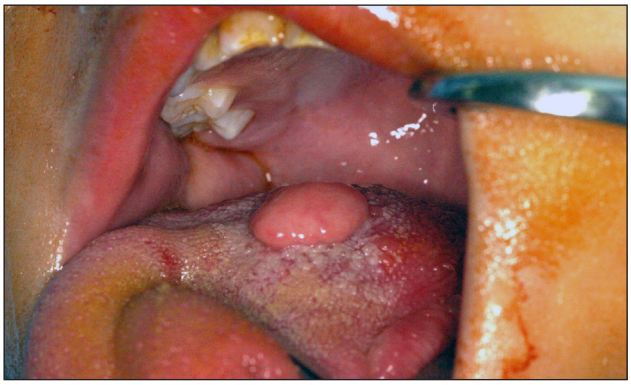

Figure 1. Photograph of the oral cavity of a patient with leiomyomatous hamartoma. A painless mass of approximately 10 mm with a clear boundary was present medially and anterior to the dorsal lingual terminal sulcus. The tumor was pediculated, elastic, and soft, and the surface was smooth and had the color of the mucosa.

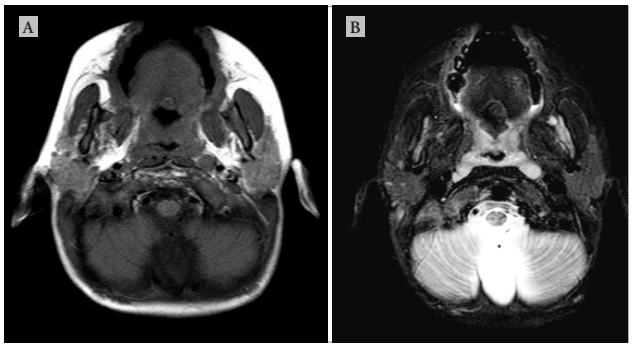

Figure 2. (A) T1-weighted and (B) T2-weighted MRI images of the leiomyomatous hamartoma. The intensity of the mass inner region was homogenous and similar to that of the surrounding tongue muscle.

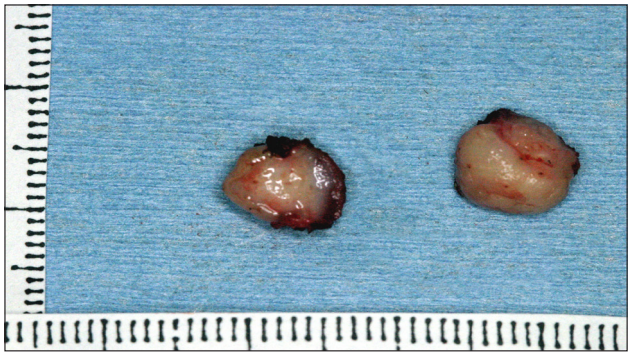

Figure 3. Photograph of the excised leiomyomatous hamartoma. The specimen measured 13x12 mm, was elastic and soft, and had a solid and whitish-yellow cross-sectional surface.

DISCUSSION

Hamartoma is defined as a congenital mixed abnormality in which one or several tissue components grow excessively in a normal organ. This condition was initially reported by Albrecht in 1904.[1] A lesion formed by overgrowth of external tissue that was originally absent at the site of development is referred to as a choristoma.[5] Although these lesions should be distinguished from hamartoma,[6] differentiation is difficult since the tissue aberration cannot be clarified with certainty. Thus, cases of choristoma have recently been interpreted as hamartoma in a broad sense.

Hamartoma is pathologically classified based on the main tissue component.[7] In addition to leiomyomatous hamartoma, cases of lipomatous,[8] cartilageous,[3] hemangiomatous,[3] myofibromatous,[7] and ectodermal[2] hamartoma of the oral cavity have been reported, but leiomyomatous hamartoma of the tongue is rare.[3,4] In our patient: overgrowth of smooth muscle cells was dominant;

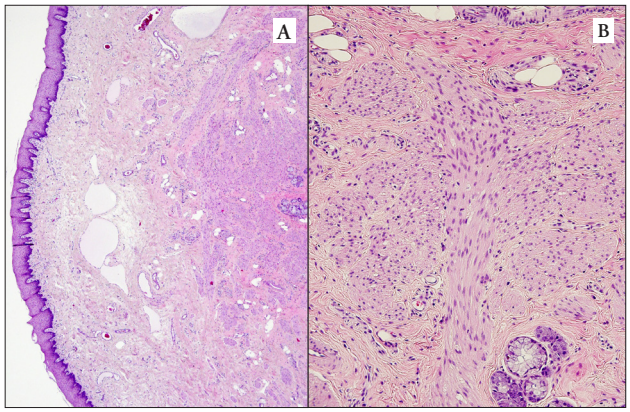

Figure 4. Histopathologic images of leiomyomatous hamartoma (A) hematoxylin and eosin staining (40x) and (B) hematoxylin and eosin staining (200x). The superficial layer was covered with stratified squamous epithelium, and nodular overgrowth of spindle cells containing eosinophilic cytoplasm was noted in the submucoepithelial connective tissue. There were no findings, however, that suggested tumorous proliferation. Fibrous, adipose, and minor salivary gland tissues were present around the lesion.

fibrous, adipose, and minor salivary gland tissues were present around the lesion; and no capsule or tumorous proliferation were noted. These findings led to a diagnosis of leiomyomatous hamartoma. Confirmation of α-SMA positivity using immunostaining was required for definite identification of the smooth muscle as the origin of the main overgrowing cells.

To our knowledge, only 26 cases (including the present case) have been diagnosed histopathologically as leiomyomatous hamartoma in the oral cavity between 1945 and 2009. Of the 25 previous cases, 17 occurred in the tongue, followed by 6 in the gingiva, 1 in the hard palate, and 1 in which the location was unclear. In the 17 cases in the tongue, the site of development was in the posterior region of the tongue (as in our patient) in 11 cases (65%), and the male:female ratio was 11:6, showing a higher incidence in males. The age at the first examination varied from birth to 61 years old. The lesion was found at birth in 12 cases, and all except 2 were found by 1 year old; 1 case was discovered at 34 years old, and the age at the time of discovery was unclear in 1 case.

Ten cases underwent resection by 3 years old. The size of the lesion was 0.1 to 4.0 cm, and the mean size (excluding unclear cases) was about 1.5 cm (Table 1). In our patient, the lesion was found at 1 year old, which was slightly later than in most other reported cases. This may have been due to the small lesion size (1.0 cm), the absence of an eating disorder, and the parents' attention to the child's oral cavity.

The cause of tongue root hamartoma is unclear, but it is thought to be associated with the embryonic developmental mechanism of the tongue.[3] The body of the tongue is formed as the bilateral lateral lingual swellings extend to cover the tuberculum impar and

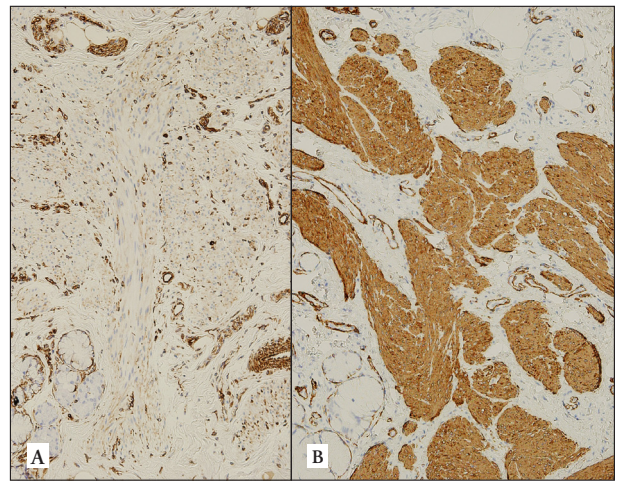

Figure 5. Immunohistological staining of the leiomyomatous hamartoma for (A) vimentin and (B) α-SMA. Spindle cells in the lesion were positive for both vimentin and α-SMA.

Table 1. Reported Cases of Leiomyomatous Hamartoma of the Tongue

Reference	Year	Age	Age at discovery	Gender	Region	Size (cm)
Stamm et al.[8]	1945	At birth	At birth	F	Posterior	Unclear
Perri et al.[9]	1956	34 ys	32 ys	F	Posterior	2
Hinshaw et al.[10]	1963	4 ys	At birth	F	Posterior	2
Demuth et al.[11]	1981	1 y, 6 mos	2 mos	M	Posterior	Unclear
Becker et al.[12]	1984	At birth	At birth	M	Posterior	2
Goldsmith et al.[13]	1995	1 y, 4 mos	Unclear	M	Anterior	Unclear
Rosa-Garcia et al.[14]	1999	6 ys	At birth	M	Anterior	1.3
Kobayashi et al.[3]	2001	6 mos	At birth	M	Posterior	1.4
Iida et al.[5]	2007	2 ys	4 mos	M	Anterior Posterior	0.2 0.4
Kreiger et al.[6]	2007	8 ds	At birth	M	Posterior	0.1-2
		4 mos	At birth	F	Posterior	0.1-2
		5 mos	At birth	F	Central	0.1-2
		1 y	At birth	M	Posterior	0.1-2
		5 ys	Unclear	M	Central	0.1-2
Nave-Villalba et al.[6]	2008	5 ys	At birth	M	Posterior	0.7
Rogerio de Faria et al.[1]	2008	61 ys	At birth	F	Posterior	4
Present case	2009	3 ys	1 y	M	Posterior	1

fuse. The root of the tongue is formed as the copula formed by fusion of the ventromedial region of the second branchial arch is covered by the mesenchymal tissue-derived hypobranchial eminence in the ventromedial region of the third and fourth branchial arches. Finally, the body and root of the tongue fuse at the terminal sulcus and form the tongue. At this point, a proliferative error may occur and induce hamartoma. The relatively high incidence of hamartoma in this region compared to that in other regions may be due to the complex developmental mechanism that involves many tissue types.

Diseases to be differentiated in the clinical diagnosis of leiomyomatous hamartoma include fibroma of the tongue, ectopic thyroid gland, and thyroglossal duct cyst,[15] and pathological diagnoses include leiomyoma, leiomyosarcoma, and benign mesenchymoma.[4] In our patient, ectopic thyroid gland was excluded because the thyroid gland was present at the normal position on an MRI. A thyroglossal duct cyst is found in the tongue in very rare cases, and can be differentiated based on its clinical feature of a mobile mass with palpable waves and on MRI findings. Within pathological diagnoses, the absence of a capsule, no infiltration in the surrounding tissue, and no tumorous proliferation were noted, based on which leiomyoma and leiomyosarcoma were excluded.

Benign mesenchymoma contain 2 or more types of mesenchymal tissue, in addition to fibrous connective tissue, and are very similar to hamartoma. The points of differentiation are a tendency for infiltration in the surrounding tissue, a larger size than a hamartoma lesion, and the absence of tissues from the salivary gland and epithelial system.[16] Differentiation was easy in our case, because there was no tendency for infiltration, and salivary gland and epithelial system tissues were included in the lesion.

Surgical resection is the first choice of treatment for hamartoma. Although the lesion does not grow rapidly during course observation,[3] early resection should be performed when symptoms such as speech, eating, and swallowing disorders are present. Even when no symptoms are present and the disease cannot be diagnosed clinically, early resection is desirable for differentiation from other diseases. In addition, the tumor does not become malignant or recur, because it is composed of differentiated tissue and has the normal tissue features.[6] In fact, no malignant or recurrent case of hamartoma in the oral cavity has been reported. Malignant hamartoma of the lung has been reported,[17] however, suggesting the need for sufficient follow-up after resection.

In conclusion, when a mass is present in the postmedian region near the terminal sulcus of the dorsum of the tongue in neonates and infants, the possibility of hamartoma should be kept in mind in diagnosis and treatment, even though the incidence is rare.

REFERENCES

1. de Faria RP, Batista JD, Durigueto AF, et al. Giant leiomyomatous hamartoma of the tongue. J Oral Maxillofac Surg 2008;66:1476-80.
2. Ishii T, Takemori S, Suzuki J-I. Hamartoma of the Tongue. Arch Otolaryng 1968;88:79-81.
3. Kobayashi A, Amagasa T, Okada N. Leiomyomatous hamartoma of the tongue: Case report. J Oral Maxillofac Surg 2001;59:337-40.
4. de La Rosa-Garcia E, Mosqueda-Taylor A. Leiomyomatous hamartoma of the anterior tongue: Report of a case and review of the literature. Int J Paediatr Dent 1999;9:129-32.
5. Barnes L, Eveson JW, Perichart P, Sidransky D. Pathology and genetics of head and neck tumors. World Health Organization Classification of Tumours. International Agency for Research on cancer; 2005:348.
6. Nave-Villalba M, Seamanduras-Pacheco A, Aldape-Barrios BC. Leiomyomatous hamartoma: Report of two cases and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008; 105:e39-e45.
7. Iida S, Kishino M, Senoo H, Okura M, Morisaki I, Kogo M. Multiple leiomyomatous hamartoma in the oral cavity. J Oral Pathol Med 2007;36:241-4.
8. Kriger PA, Ernst LM, Elden LM, Kazahaya K, Alawi F, Russo PA. Hamartomatous tongue lesions in children. Am J Surg Pathol 2007;31:1186-90.
9. Stamm C, Tauber R. Hamartoma of the tongue. Laryngoscope 1945;55:140.
10. Perri FA. Myoepithelial hamartoma of the tongue. Arch Otolaryngol 1956;64:289.
11. Hinshaw CT Jr. Unusual lesions of the tongue. Hamartoma. J Kans Med Soc 1963;64:154.
12. Demuth RJ, Johns DF. Recurrent aspiration pneumonitis in a cleft palate child with hamartoma of the tongue. Cleft Palate J 1981;18:299.
13. Becker GD, Ridolfi R, Ingber C. Tongue hamartoma in a newborn. Otolaryngol Head Neck Surg 1984;92:357.
14. Goldsmith P, Soames JV, Meikle D. Leiomyomatous hamartoma of the posterior tongue: A case report. J Laryngol Otol 1995;109:1190-1.
15. Owen G, Berry J. Hamartoma of the tongue. J Laryngol Otol 1993;107:363-7.
16. Herzog S, Bressman J, Giglio JA. Case 61: Tongue mass in an infant. J Oral Maxillofac Surg 1986;44: 463-6.
17. Basile A, Gregoris A. Malignant change in a benign pulmonary hamartoma. Thorax 1989;4:232-3.