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Case Report

Ear, Nose and Throat Section

Congenital Midline Tongue Base Mass in An Infant: Lingual Hamartoma

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ABSTRACT

Lingual hamartoma is a rare finding of congenital midline posterior tongue mass. The lesion may be seen as a single anomaly or maybe associated with syndrome especially the Oral Facial Digital Syndrome (OFDS). Here, we report an otherwise normal and healthy two-month-old boy with a congenital midline base of tongue mass presented with snoring and episodic vomiting since the age of 1 month. Tumour excision from the area of foramen of caecum recovered a pinkish pedunculated tumour. Histopathology examination confirmed the diagnosis of leiomyomatous lingual hamartoma. Differential diagnosis, especially for midline tongue mass and other paediatric tongue lesions are discussed. We also discuss the epidemiology, histopathologic features, treatment and prognosis of lingual hamartoma based on the literature review.

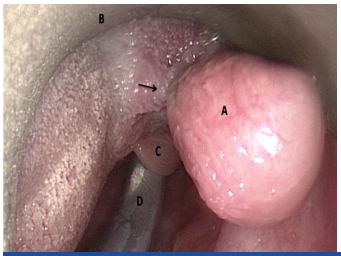
Keywords: Excision, Oral facial digital syndrome, Pediatric

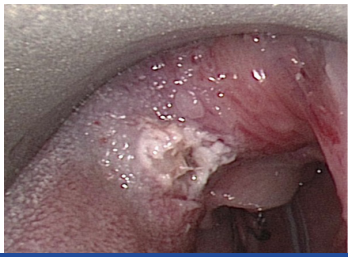
CASE REPORT

A one and half-month-old boy was referred from outpatient of a private clinic to our medical centre with complaint of snoring and episodic vomiting since 1 month. The mother also observed an intraoral mass since birth. The mother noticed it since day 3 post natal period, but never sought treatment for the problem. The swelling did not increase in size and never caused a problem until the age of 1 month. The mother noted the child to snore especially when he slept in supine position and the sounds were louder when the child had cough and runny nose. Otherwise, the child was tolerating breast feeding and there was no choking, nor cyanotic spells and he was thriving well. There was no history of perinatal insults. The child was born full-term with birth weight of 3.7kg. His birth was uneventful and his immunization was up to age.

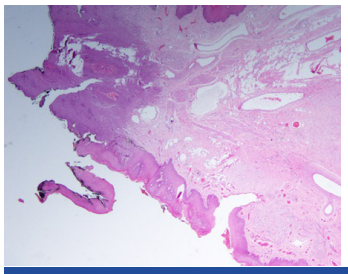
When examined in our clinic, the child was comfortable, neither in stridor nor respiratory distress. A flexible scope showed a pinkish, pedunculated mass, originating from the tongue base. No abnormalities were observed over the larynx and the vocal cords were mobile. The results of thyroid function test were within normal limits and ultrasound of neck showed a normal thyroid gland in its usual position in the neck. There were no pre-operative imaging done prior to the surgery.

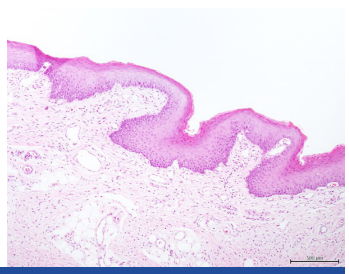
After discussion with the parents, they agreed and consented for the surgery. At 2 months, he underwent transoral tumour excision. Airway was secured with an endotracheal tube by a senior anaesthetist. Examination under anaesthesia using Lindholm laryngoscope (Karl Storz, Germany) showed a pedunculated, firm mass, measuring 1.5x0.6x0.5 cm, arising from midline base of tongue [Table/Fig-1]. The mass was excised using cold instrumentation and surrounding mucosa with a thin layer of tongue musculature was removed without complications [Table/Fig-2]. Histopathology examination confirmed the diagnosis of smooth muscle predominant lingual hamartoma [Table/Fig-3,4]. The patient tolerated the procedure well and was discharged the following day after uneventful airway observation. There was no recurrence of the lesion to date.





[Table/Fig-1]: A: pedunculated tongue base mass, B :Lindhom laryngoscope C :Lingual surface of epiglotis, D: Endotracheal tube, **[Table/Fig-2]:** Post excision of tongue base lesion.





[Table/Fig-3]: A section showing a polypoidal lesion with thin stalk lined by stratified squamous epithelium. **[Table/Fig-4]:** A section showing mature stratified squamous epithelium with underlying stroma composes of predominantly unencapsulated smooth muscle fibers intermingle with occasional adipose tissues and vascular channels.

DISCUSSION

Hamartoma is a benign tumour-like overgrowth of mature tissue which is native to the organ of its origin [1]. Lingual hamartoma is a rare tongue mass, primarily diagnosed in childhood and was first described in 1945 by Stamm and Tauber [2]. It is a congenital lesion, which is usually asymptomatic and detected as incidental tongue mass [1]. From literature review by Nava-Villalba et al., they described specific clinical characteristics of hamartoma in the oral cavity which include: 1) small lesion (less than 1.5 cm in largest diameter); 2) pink pedunculated/polypoid lesion; 3) tendency of occurrence in the midline of the palate(anterior region) or the midline of the tongue [3].

Lingual hamartoma may present as an isolated anomaly or as part of syndrome especially Oral Facial Digital Syndrome (OFDS) which is a group of hereditary disorders comprising of oral abnormality, facial dysmorphism and hand/foot malformations [4]. It was described that lingual hamartomas presenting as an isolated anomaly are extremely rare [5]. However, a previous review of 135 cases involving paediatric tongue lesions, reported 14 out of 18 lingual hamartomas to occur in otherwise normal non-syndromic children [1].

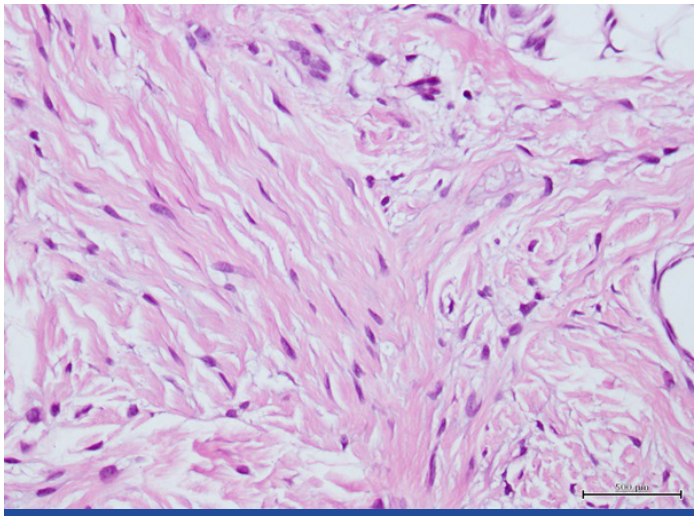
Differential diagnosis of midline posterior part of tongue lesion in children often is thought to be of a lingual thyroid gland or a thyroglossal duct cyst, as this lesion most commonly occurs in

the area of foramen caecum. The presence of a normal-appearing thyroid in the lower neck also may not exclude lingual thyroid.

Nevertheless, lingual hamartomas may also occur as solitary lesions on the dorsum of the tongue, usually posterior and midline in the area of the foramen cecum [5] and should be considered as one of the differential diagnosis in the present case, although the occurrence is rare. The position of these lesions to occur in the midline posterior part of the tongue may be explained on its location as a fusion region during embryological development [5].

Some other lesions that may be present as congenital midline tongue mass includes choristoma, leiomyoma and benign mesenchymoma. Other causes of paediatric tongue lesions irrespective of the site of occurrence would be vascular or lymphatic lesion (lymphangioma, pyogenic granuloma, capillary haemangioma, vascular malformations), mucus extravasation phenomenon, reactive or traumatic lesions (fibroma and neuroma), cystic lesion, benign and malignant neoplasm [1].

Most of the lingual hamartomas demographically occur in young aged children, less than 2 years [1,3] with female preponderance [1,5]. According to definition, all lesions are present at birth but age of presentation for excision ranges between 1 month to 61 years [1,3]. Summary of the literatures is presented in [Table/Fig-5].



[Table/Fig-6]: Fusiform eosinophilic cells with elongated tapered end nuclei.

Haematoxylin and Eosine (H&E) examination, features of leiomyomatous hamartoma compose of smooth muscles bundles which are unencapsulated in the submucosal region and haphazardly arranged intermingled with variably sized vessels, adipose tissues,

Authors	Number of case(s)	Gender	Symptoms/ presentations	Associated syndrome	Appearance	Site	Age at surgical excision
Kreiger et al., 2007 [1]	18	6 males, 12 females	All asymptomatic mass, except one with choking and cyanotic spells	4 cases associated with OFDS	Range in size of 0.1-2 cm; and nodular, polypoid or pedunculated gross appearance	All at dorsal part of tongue except 1 case at the ventral part of the tongue	Less than 2 years old: 15 cases. 3 cases at age 3, 5, 16 years old respective
Stamm and Tauber, 1945 [2]	1	Female	Feeding difficulties	None	Sessile tumour	Base of tongue, more to the left	Day 9 of life
Nava-Villalba et al., 2008 [3]	1	Male	Congenital asymptomatic tongue mass	None	7 mm, polypoidal pedunculated lesion	Posterior third, dorsum of tongue	5 months old
Hsu et al., 2012 [4]	1	Female	Incidental finding of granular lesion at the tongue	None	Pedunculated 1.5x0.6x0.5 cm	Base of tongue, more to the left	1 year 5 months old
Goold et al., 2003 [5]	1	Male	Congenital asymptomatic tongue mass	None	Lobulated sessile mass 2x 2cm	Base of tongue, near foramen caecum	5 months old
Valdyanathan et al., 2011 [6]	1	Female	Congenital asymptomatic tongue mass	None	1.5 cm sessile lesion	Left lateral border of tongue	1year 2 months old
de Faria et al., 2008 [7]	1	Female	Congenital asymptomatic tongue mass, rapid enlargement at 61 years old with dysphagia, dysphonea, dyspnoea for 2 weeks duration	None	"Giant leimyomatous hamartoma"4 cm bilobulated, pedunculated lesion	Midline, posterior region of the tongue	61 years old
Kuperan et al., 2012 [8]	1	Male	Congenital asymptomatic tongue mass	None	0.5 cm pedunculated mass	Midline posterior part of the tongue	5 months old
Wang et al., 2013 [9]	1	Male	Painless slow growing mass on the tongue since childhood	Having bifid tongue with tongue tie, but not associated with OFDS	2x1.5cm, pedunculated mass	Anterior midline of the tongue, near the apex of the bifid tip	29 years old
Noraziana et al., 2016	1	Male	Congenital tongue mass with snoring and episodic vomiting	None	1.5 x 0.6 x 0.5 cm, pedunculated mass	midline base of tongue	2 months old

[Table/Fig-5]: Summary of literature review.

Typically, lingual hamartoma occur in the submucosal connective tissue, superficial to the tongue muscles [1]. The type of hamartoma is classified largely on the predominant cell type in the histological specimen namely, leiomyomatous, rhabdomyomatous, neurovascular, fatty, glial and so forth [5]. In routine histologic nerves tissues or salivary glands. On higher magnification, the presence of spindle shape cells, with elongated and tapered end nuclei can readily be seen [Table/Fig-6]. Immunohistochemistry (IHC) staining may help to confirm leiomyomatous hamartoma by identification of smooth muscle actin, muscle-specific actin, S-100

protein and desmin [3]. The fact that it differs from neoplasm is from the absent of pleomorphism of the nuclei in the tumour [3].

In our case ultrasonography of neck was performed to confirm presence or absence of cervical thyroid tissue as to rule out lingual thyroid or ectopic thyroid tissue. Preoperative CT scan or MRI was not performed prior to operation as the lesion has benign characteristics on the clinical appearance i.e. small lesion with pedunculated stalk. The radiological feature is not well described in literature, as majority of the cases were operated without preoperative imaging. However, from limited reports with pre-operative imaging of CT scan and MRI have shown inconsistent findings with no specific characteristic [4].

Definitive step of management is excision of the tumour. According to literature review, the prognosis is good and there are no cases of recurrence following complete surgical excision of lesion [3-9].

CONCLUSION

In general although it is rare, lingual hamartoma should be considered as a differential diagnosis of a congenital midline base of tongue lesion in children apart from lingual thyroid and thyroglossal duct cyst. Having a history of OFDS may suggest the possibility of having this lesion. However, we should also consider lingual hamartoma as one of the differential diagnosis, taking into account majority of cases occur in a normal and non syndromic child. Standard histologic H&E confirmed the diagnosis of leiomyomatous hamartoma, and IHC staining may aids in the diagnosis when in doubt. The treatment of choice is complete surgical excision of the lesion. It has a good prognosis, as no recurrence of the lesion after excision is reported in literature, to date.

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