Lingual bronchogenic cyst in a young child: A case report

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**ABSTRACT**

Choristomas, or foregut duplication cysts, are benign embryonic tumors characterized by normal tissue appearing in abnormal sites. If they are lined with respiratory epithelium they are termed bronchogenic cysts. When present, these rare lesions are usually intra-thoracic. We here present a case of a 4-year-old boy diagnosed with a bronchogenic cyst in the tongue.

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**Introduction**

Bronchogenic cysts are benign congenital malformations. They are part of a group of tumors called choristomas, or heterotopic cysts, or foregut duplication cysts. These are embryonic tumors characterized by normal tissue in abnormal sites. Choristomas can be lined with respiratory epithelium, i.e. ciliated pseudostratified squamous epithelium comprising mucous secreting cells, smooth muscle and cartilaginous tissue. They are then referred to as bronchogenic cysts. Choristomas can also be lined with gastrointestinal epithelium and are then termed alimentary cysts. There are even reports of cysts lined with a mix of these two types of epithelia [1,2]. Bronchogenic cysts are usually intra-thoracic, and extra-thoracic sites are rarely seen [3]. Only an extremely limited number of lingual bronchogenic cysts have been previously described [1,3–5]. We here present a case of a young boy with a lingual bronchogenic cyst.

**Clinical summary**

A 4-year-old boy with a previously diagnosed IgA deficiency, but otherwise healthy, was remitted to our Ear, Nose and Throat Department from the Pediatric Department because of an enlargement of the mobile part of the tongue. It had been noticed since birth that he tended to rest his tongue outside his mouth, and that his tongue was somewhat enlarged. He also had a tendency to bite his tongue when eating. No impairment of speech or swallowing had been noticed. The boy had been stated to have very strong opinions on what type of food to eat, though neither his parents nor his pediatricians had had any suspicion that this was due to his tongue anatomy. His dentist had expressed concerns it would negatively affect his tooth development. According to the parents the enlargement did not vary in size and there was no history of bleeding or ulcerations.

On examination was noted a submucosal, bilobular and consistent tumor, directly posterior to the tip of the boy’s tongue. In size it was similar to that of two adjacent grapes, thus visually almost filling the complete anterior aspect of the tongue. The tumor was transparent under illumination. The overlying mucosa showed no sign of involvement. There were no obvious problems with opening or closing of the mouth. Further examination of the mouth and pharynx was without any remarks.

This enlargement was primarily suspected to be a lymphatic malformation (LM). A magnetic resonance imaging (MRI) was performed and showed a lobulated cystic tumor, measuring $20 \times 23 \times 16$ mm, with a thin capsule. In conclusion, it was described as best compatible with a LM, alternatively a lingual bronchogenic cyst (Figure 1).
Sclerotherapy under anesthesia was considered, the first-line treatment for LM in this area. However, due to the clinical findings in combination with the MRI results, the decision was made to treat this tumor surgically. This decision was reached since, albeit rare, a lingual bronchogenic cyst could reasonably be the more probable diagnosis in this instance, as well as taking into consideration risks and expected post-treatment morbidity following sclerotherapy.

A few months after the patient’s fifth birthday surgery was performed, with a submucosal total extirpation of the tumor. The tumor capsule was found to be relatively stable, in contrast to what was to be expected from a LM, and surrounding nerves could be identified and dissected from the tumor. The tumor was completely removed without any complications (Figures 2 and 3), and the patient was extubated directly postoperatively. The patient received phenoxymethylpenicillin prophylaxis. The postoperative period was free from complications, and 2 days after surgery the boy was discharged home, feeling relatively well. Histological examination of the tumor showed a $20 \times 10 \times 15$ mm cyst, with a smooth cystic capsule and a ciliated respiratory epithelium, but without any signs of gastrointestinal epithelia. This confirmed the diagnosis of a benign bronchogenic cyst. Two months after surgery the boy returned for follow-up examination. Both subjectively and objectively the function of the tongue was normal, hypoglossal nerve function was intact, and its appearance was that of a normal tongue, apart from minor scar tissue on the ventral aspect (Figure 4). The patient denied any sensorineural or taste disturbances. There were no other problems or symptoms reported.

Discussion
A bronchogenic cyst in the tongue is an extremely rare congenital malformation. The hypothesis is that these cysts are formed due to a developmental derangement
of the foregut, hence are they sometimes called ‘intralingual cysts of foregut origin.’ The definite pathogenesis is however not known [6]. These tumors are often diagnosed shortly after birth, either as asymptomatic lingual masses, or when an infant experiences feeding difficulties. If the tumor is sufficiently large, it can present at prenatal ultrasonography [5,7]. However, as these tumors are often asymptomatic, if they are small they can go undiagnosed until later in childhood. At that time the diagnosis is often made when the child shows speech disturbances, or problems with eating or mouth closure [1]. An exception to childhood diagnosis was presented by Kim et al., who reported a case of a 27-year-old male presenting with a sublingual bronchogenic cyst [8].

These tumors can measure up to 9 cm in diameter, although they usually measure less than 3 cm. They, like most congenital deformities have a male predilection [9]. Surgical excision is considered the treatment of choice in this condition. It should be performed with certain urgency if patients present with dysphagia and dyspnea, otherwise as soon as is reasonable due to age and general condition. Significant post-operative swelling in the operated area may develop, therefore these patients might be candidates for intensive care in the immediate post-operative period [7].

MRI is the examination of choice in the work-up for a lingual tumor suspected of being a bronchogenic cyst; this however does not give a definite diagnosis. For diagnostic certainty, excisional biopsy is required [6]. Differential diagnosis to cystic lingual tumors in infants include LM, dermoid cysts, ranula (or mucocele), thyroglossal duct cyst, venous malformations, ectopic thyroid, etc. Malignant tumors of the tongue are very rarely seen in children. However, cases of adenocarcinoma arising from benign bronchogenic cysts have been reported. Olsen et al. [10] reported a case of an adenocarcinoma arising from an intra-thoracic bronchogenic cyst, and Volchok et al. [11] reported a case of adenocarcinoma arising from a previously undiagnosed lingual foregut cyst in an adult.

Albeit a rare diagnosis as well, LM is the main differential diagnosis to lingual choristoma. Sclerotherapy, preferably with OK-432, is the first line of treatment of LM, though with the location in this case it would be associated with risks for airway obstruction, and consent for a temporary tracheostomy would be needed preoperatively. Surgery for LM is difficult, partly because of the ill-defined borders of the disease and partly because of the weak structure of the cyst walls, making the tumor difficult to handle. In the present case, the tumor, in accordance to the diagnosis, was relatively easy to handle, making radical surgery possible without hampering nerve function or normal lingual tissue. This, in combination with less morbidity than sclerotherapy and the simultaneous handling of the small risk for malignancy, further stresses the advantage of surgery in these rare cases. Regarding the timing of surgery, findings and symptoms should be put against the risks related to general anesthesia at a low age.

Conclusions
Lingual bronchogenic cysts are a rarely seen congenital lesion. When suspected in a patient we recommend surgical excision as a first line of treatment, but recommend measures being taken to be prepared for any possible postoperative swelling.

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Disclosure statement

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References

[1] Manor Y, Buchner A, Peleg M, et al. Lingual cyst with respiratory epithelium: an entity of debatable histogenesis. J Oral Maxillofac Surg. 1999;57:124–127. Feb discussion 128-9.
[2] Joshi R, Cobb AR, Wilson P, et al. Lingual cyst lined by respiratory and gastric epithelium in a neonate. Br J Oral Maxillofac Surg. 2013;51:173–175. Mar
[3] Kun-Darbois JD, Breheret R, Bizon A, et al. Bronchogenic cyst of the tip of the tongue: report of two cases. Eur Ann Otorhinolaryngol Head Neck Dis. 2015;132:49–51. Feb
[4] Fortier A, Boyer C, Ducasse H, et al. Bronchogenic cyst of the tongue in an infant. Rev Laryngol Otol Rhinol (Bord). 2013;134:157–159.
[5] Burkart CM, Brinkman JA, Willging JP, et al. Lingual cyst lined by squamous epithelium. Int J Pediatr Otorhinolaryngol. 2005;69:1649–1653.
[6] Eaton D, Billings K, Timmons C, et al. Congenital foregut duplication cysts of the anterior tongue. Arch Otolaryngol Head Neck Surg. 2001;127:1484–1487. Dec
[7] Akyol MU, Orhan D. Lingual tumors in infants: a case report and review of the literature. Int J Pediatr Otorhinolaryngol. 2004;68:111–115. Jan
[8] Kim YS, Ahn SK, Lee SH. Sublingual foregut cyst. J Dermatol. 1998;25:476–478.
[9] Morgan WE, Jones JK, Flaitz CM, et al. Congenital heterotopic gastrointestinal cyst of the oral cavity in a neonate: case report and review of literature. Int J Pediatr Otorhinolaryngol. 1996;36:69–77.
[10] Olsen JB, Clemmensen O, Andersen K. Adenocarcinoma arising in a foregut cyst of the mediastinum. Ann Thorac Surg. 1991;51:497–499.
[11] Volchok J, Jaffer A, Cooper T, et al. Adenocarcinoma arising in a lingual foregut duplication cyst. Arch Otolaryngol Head Neck Surg. 2007;133:717–719.