

Bronchogenic cyst in an unusual location

Cisto broncogênico de localização não habitual

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ABSTRACT

This article reports the case of a patient with lingual bronchogenic cyst and presents a brief review of discrepancies in the nomenclature of lingual cysts. A 2-year-old male patient was admitted to the pediatric surgery outpatient clinic of a university hospital due to the presence of a 2 cm mass on the dorsal surface of the tongue. The tumor was excised and the anatomopathological report revealed a foregut cyst, bronchogenic subtype. Lingual cysts are rare, especially when compatible with bronchogenic formations. Their pathogenic process is not clear; one of the possibilities is the separation of cells from the primitive gut, before the separation between the esophagus and the trachea. The treatment usually consists of surgical excision.

Key words: bronchogenic cyst; congenital abnormalities; choristoma.

RESUMO

Este artigo relata o caso de um paciente com cisto broncogênico lingual e apresenta uma breve revisão das discrepâncias de nomenclatura para cistos linguais. Paciente do sexo masculino, 2 anos de idade, foi admitido no ambulatório de cirurgia pediátrica de um hospital universitário devido à presença de uma massa de 2 cm na face dorsal da língua. O tumor foi excisionado; o laudo anatomopatológico revelou foregut cyst, subtipo broncogênico. Cistos linguais são raros, especialmente quando compatíveis com formações broncogênicas. Seu processo de patogênese é incerto, e uma das possibilidades é a separação de células do intestino primitivo antes da separação entre esôfago e traquéia. O tratamento, comumente, consiste em excisão cirúrgica.

Unitermo: cisto broncogênico; anormalidades congênitas; coristoma.

RESUMEN

Este artículo relata el caso de un paciente con quiste broncogénico lingual y presenta una breve revisión de las discrepancias de nomenclatura para quistes linguales. Paciente de sexo masculino, de 2 años de edad, acudió a cirugía pediátrica de un hospital universitario con una masa de 2 cm en la cara dorsal de la lengua. El tumor fue extirpado y el informe anatomopatológico reveló quiste de duplicación, subtipo broncogénico. Quistes linguales no son frecuentes, sobre todo cuando concomitantes con formaciones broncogénicas. Su patogénesis es incierta: una de las posibilidades es la separación de células del intestino primitivo antes de la separación entre esófago y tráquea. El tratamiento, en general, consiste en escisión quirúrgica.

Palabras clave: quiste broncogénico; anormalidades congénitas; coristoma.

INTRODUCTION

Lingual cysts are rare and are generally classified by the type of epithelium they present⁽¹⁾. The most common types of epithelium present are those of the gastrointestinal and respiratory tract, and the presence of both may occur. There is no consensus on nomenclature, and multiple terms are used, such as foregut cyst, lingual choristoma, enteric duplication cyst, heterotopic gastrointestinal cyst of oral cavity, among others⁽²⁾. This variety masks the true incidence of this type of lesion, since it makes it difficult to search for and review the literature. The term foregut cyst is the most commonly used and includes bronchogenic cysts, esophageal duplication cysts, and enteric duplication cysts⁽³⁾. Its pathogenesis is not clear; therefore the possibility of derivation from a sprout of cells separated from the primitive foregut is indicated⁽⁴⁾. Such a process would explain why the cyst may present both digestive and respiratory epithelium, since both originate from the foregut-derived epithelium.

In this report we present a case of lingual cyst in a two-year-old boy with a history of mass in the anterior region of the tongue, since birth. After evaluation of the lesion by computed tomography (CT), surgery was performed for the excision and the material was sent for anatomopathological examination. The result revealed pure respiratory epithelium, raising the hypothesis of a foregut cyst, bronchogenic subtype, in an unusual location.

DESCRIPTION OF THE CASE

Two-year-old male patient with history of a mass in the middle and anterior third of the tongue, in dorsal face, since birth. According to the mother, it presented volume variation, with spontaneous increase and decrease. The CT showed a cystic rounded image, with well-defined contours, with no significant contrast-enhancement, located in the midline of the tongue, measuring about $18 \times 15 \times 15$ mm. Ultrasonography (US) of the thyroid revealed the presence of toxic thyroid. After excision of the lesion (**Figures 1-3**), the diagnosis by hematoxylin and eosin (HE) examination was bronchogenic cyst (**Figure 4**). Immunohistochemistry (IHC) (**Figure 5**) of the surgical specimen was performed, which was positive for the cytokeratin 7 (CK7) markers and negative for the cytokeratin 20 (CK20), S100 and smooth muscle actin markers. The anatomopathological diagnosis associated with the immunohistochemical profile was conclusive for foregut cyst of bronchogenic subtype.

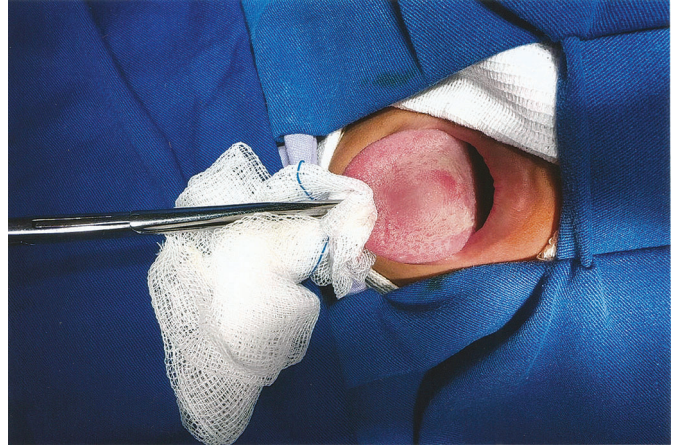


FIGURE 1 – Image of the lesion before the procedure

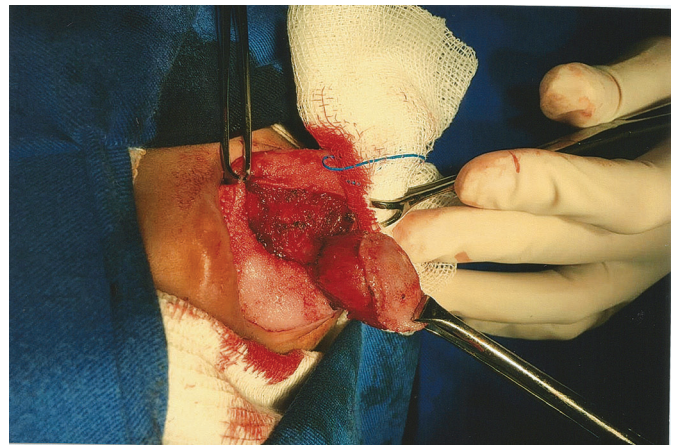


FIGURE 2 – Surgical removal of the lesion

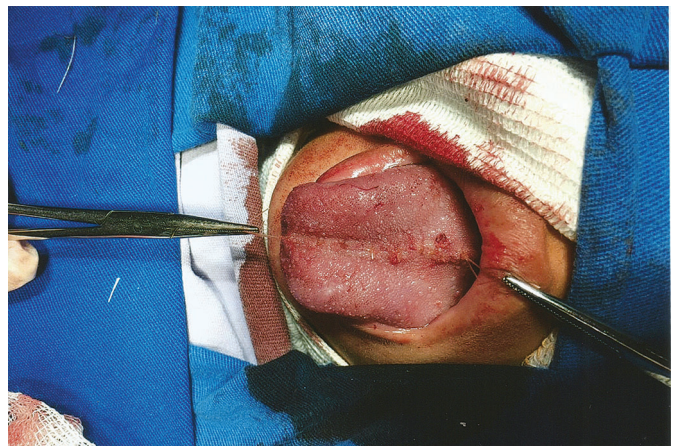


FIGURE 3 – Tongue after surgical wound suture

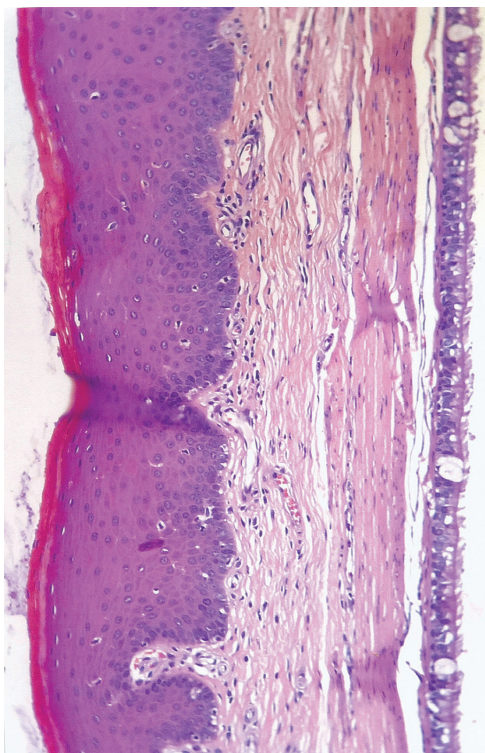


FIGURE 4 – HE stained sections of lesion
HE: hematoxylin and eosin.

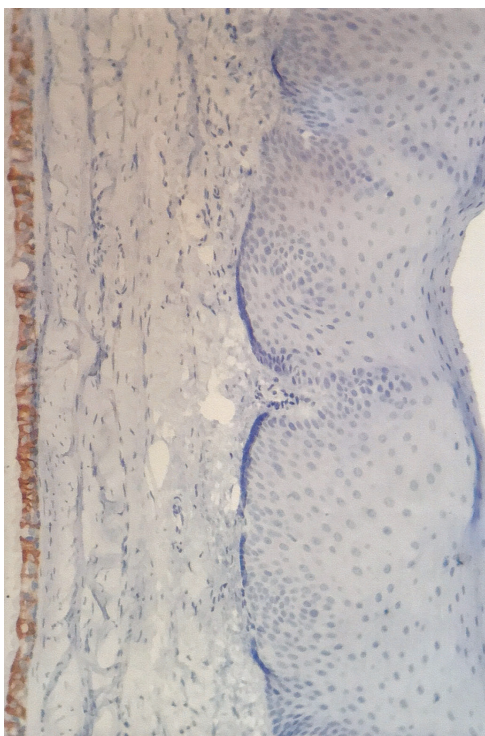


FIGURE 5 – Immunohistochemical profile of lesion section

DISCUSSION

Oral foregut cysts are rare congenital choristoma, which arise in the oral cavity during the embryological development from the remnants of foregut-derived epithelium⁽²⁾. In the literature, these types of cystic lesions are called heterotopic cysts or foregut duplication cysts⁽⁴⁾, because they derive from the primitive foregut. Differences in nomenclature make it difficult to determine the incidence of these lesions, since they may be called median lingual cyst, foregut cyst, heterotopic cyst and enterocystoma^(1, 2, 4). These formations comprise two groups: bronchogenic cysts and foregut duplication cysts.

The diagnosis of foregut cyst follows three basic criteria: to be covered by a layer of smooth muscle, to contain remnants of foregut-derived epithelium and to be attached to a portion of the foregut⁽³⁾. The term foregut cyst encompasses the bronchogenic cyst and the esophageal duplication cysts, among others. The reported case shows, through histology, foregut-derived epithelium (ciliated pseudostratified – respiratory-type), attached to the tongue, meeting enough criteria to be classified as a foregut cyst. The macroscopic characteristics are also consistent with this hypothesis, since the foregut cyst usually presents as a cystic mass, with well-defined and well-circumscribed contours⁽²⁾, as demonstrated by the patient's CT.

The subclass of the foregut cyst is predominantly determined by the histological type found in the cyst⁽¹⁾, so the anatomopathological analysis of the surgical specimen is required.

Bronchogenic cyst is the nomenclature to cystic formations that contain classically respiratory ciliated pseudostratified epithelium, goblet cells, smooth muscle cells and cartilaginous tissue. Its most common location is parabronchi, and can occur anywhere in the tracheobronchial tree. The bronchogenic cyst represents about 60% of all mediastinal cysts⁽⁵⁾. However, there are reports of bronchogenic cysts in unusual regions, such as the tongue⁽⁴⁾. Despite the absence of cartilaginous tissue, due to the typical histological characteristics of respiratory epithelium, the most appropriate subclass for the case at issue is bronchogenic cyst.

The most conclusive way to determine the type of epithelium observed is the IHC, and it is possible to detect specific markers of certain tissues⁽²⁾. In the case presented, the final determination of tissue type was made by the IHC study. Cytokeratins are the main markers for the immunohistochemical profile of the oral foregut cysts⁽⁵⁻⁷⁾. Among them, the most used are CK20 and CK7. CK20 is found in skin cells and in normal gastrointestinal tract epithelium⁽⁸⁾. Whereas CK7 is found in simple and transitional epithelia and in the luminal surface of the

pseudostratified respiratory epithelium⁽⁸⁾. In the case presented, IHC corroborated the findings observed in the histology, with CK20 negative and CK7 positive, a typical result of respiratory epithelium. Other markers used, including in this report, are S100 protein and smooth muscle actin. S100 protein is found in cells derived from the neural crest and in conjunctive cells, such as chondrocytes⁽⁹⁾, and may be positive in bronchogenic cysts; however, in our case, it was negative because there was no cartilaginous tissue.

CONCLUSION

The divergences in the nomenclature of cysts related to structures from the foregut make it difficult to establish a specific case series, because they hinder the real estimated number and characteristics of the existing cases. Studies aimed at reviewing the multiple cases in the literature in order to search for patterns to classify these lesions would be relevant for both the teams involved in the treatment and the patients' quality of life.

REFERENCES

1. Azanero WD, Mazzonetto R, León JE, Vargas PA, Lopes MA, de Almeida OP. Lingual cyst with respiratory epithelium: a histopathological and immunohistochemical analysis of two cases. *Int J Oral Maxillofac Surg.* 2009; 38(4): 388-92. PubMed PMID: 19217261.
2. Rosa AC, Hiramatsu DM, de Moraes FR, Passador-Santos F, de Araújo VC, Soares AB. Oral foregut cyst in a neonate. *J Craniofac Surg.* 2013; 24(6): 2158-60. PubMed PMID: 24220427.
3. Kieran SM, Robson CD, Nosé V, Rahbar R. Foregut duplication cysts in the head and neck: presentation, diagnosis, and management. *Arch Otolaryngol Head Neck Surg.* 2010; 136(8): 778-82. PubMed PMID: 20713753.
4. Kün-Darbois, JD, Breheret R, Bizon A, Paré A, Laccourreye L. Bronchogenic cyst of the tip of the tongue: Report of two cases. *Eur Ann Otorhinolaryngol Head Neck Dis.* 2015; 132(1): 49-51. PubMed PMID: 24993780.
5. Joshi R, Cobb AR, Wilson P, Bailey BM. Lingual cyst lined by respiratory and gastric epithelium in a neonate. *Br J Oral Maxillofac Surg.* 2013; 51(2): 173-5. PubMed PMID: 22554694.
6. Horie N, Shimoyama T, Kawano R, Ide F. Lateral lingual cyst with respiratory epithelium: a rare case. *Oral Surg* [Internet]. 2008. Available at: <https://onlinelibrary.wiley.com/doi/full/10.1111/j.1752-248X.2008.00027.x>.
7. Woolgar JA, Smith AJ. Heterotopic gastrointestinal cyst of oral cavity: a developmental lesion? *Oral Surg Oral Med Oral Pathol.* 1988; 66: 223-5. PubMed PMID: 2459648.
8. Chu PG, Weiss LM. Keratin expression in human tissues and neoplasms. *Histopathology.* 2002; 40(5): 403-39. PubMed PMID: 12010363.
9. Dabbs DJ, editor. *Diagnostic Immunohistochemistry.* 2nd ed. Churchill Livingstone/Elsevier; 2006. pp. 75; 165-6.

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