Head and Neck Pathology
https://doi.org/10.1007/s12105-021-01362-0

SINE QUA NON CLINICOPATHOLOGIC CORRELAT

Lingual Choristoma with Gastric Epithelium Revisited

Christina M. Ombres[1] · Isabelle Lestadi[2] · Juan Putra[3,4]

Received: 18 May 2021 / Accepted: 8 July 2021

Abstract
The term 'choristoma' refers to normal appearing tissue in an abnormal location. We describe a case of choristoma with gastric epithelium of the dorsal tongue in a pediatric patient. Lingual choristomas are uncommon cystic or solid lesions which may demonstrate different types of tissue (e.g. gastric epithelium, respiratory epithelium, osseous and neuroglial tissue) histologically. Choristomas with gastric epithelium, also known as heterotropic gastric mucosa or foregut duplication cysts, are thought to arise from pluripotential cells of the embryonic foregut. They most frequently involve the anterior two-thirds of the tongue. Most patients are asymptomatic, but larger lesions may lead to feeding and breathing difficulties. Pathologic evaluation and surgical excision remain the mainstay of diagnosis and treatment, respectively. The pathologic characteristics of other congenital tongue lesions are also discussed.

Keywords Lingual choristoma · Gastric heterotropia · Foregut duplication cyst · Lymphatic malformation · Hamartoma · Tongue · Pediatric

History

An otherwise healthy 5-year-old Asian boy presented with a slowly enlarging tongue lesion; the nodule was painless and initially noted by the parents when he was 1-year-old. It was suspected that the lesion was present at birth. There was no history of previous trauma to the region or pertinent family medical history.

Clinical Findings

Clinical examination demonstrated an 0.8 cm pedunculated nodule arising on the dorsal midline of the tongue (Fig. 1a). The surface was pink and smooth without ulceration. The patient did not show any dysmorphic features, and no additional lesions were identified. He subsequently underwent surgical excision of the lesion.

Diagnosis

Gross examination of the tongue lesion revealed a tan-pink and smooth cut surface. Histologically, the lingual mucosa comprised of relatively unremarkable squamous mucosa. The lesion was characterized by columnar epithelium with focal erosion (Fig. 1b); the subepithelial layer showed packed mucinous glands, reminiscent of pyloric-type glands of the stomach (Fig. 1c). Focal reactive atypia was identified in association with acute (neutrophilic) inflammation (Fig. 1d). Other types of epithelium were absent. There was no evidence of dysplasia or malignancy. The pathologic findings were those of a lingual choristoma with gastric epithelium. The patient was doing well with no evidence of disease recurrence at the time of this writing (3 years of post-excision follow-up period).

✉ Juan Putra
Juan.Putra@sickkids.ca

[1] Department of Pathology and Cell Biology, University of South Florida, Florida, Tampa, USA

[2] Department of Pathology, Harapan Kita National Centre for Women and Children's Health, Jakarta, Indonesia

[3] Division of Pathology, Department of Paediatric Laboratory Medicine, Hospital for Sick Children, 555 University Ave Rm. 3119, M5G 1X8 Toronto, Ontario, Canada

[4] Department of Pathobiology and Laboratory Medicine, University of Toronto, Toronto, Ontario, Canada

Published online: 14 July 2021

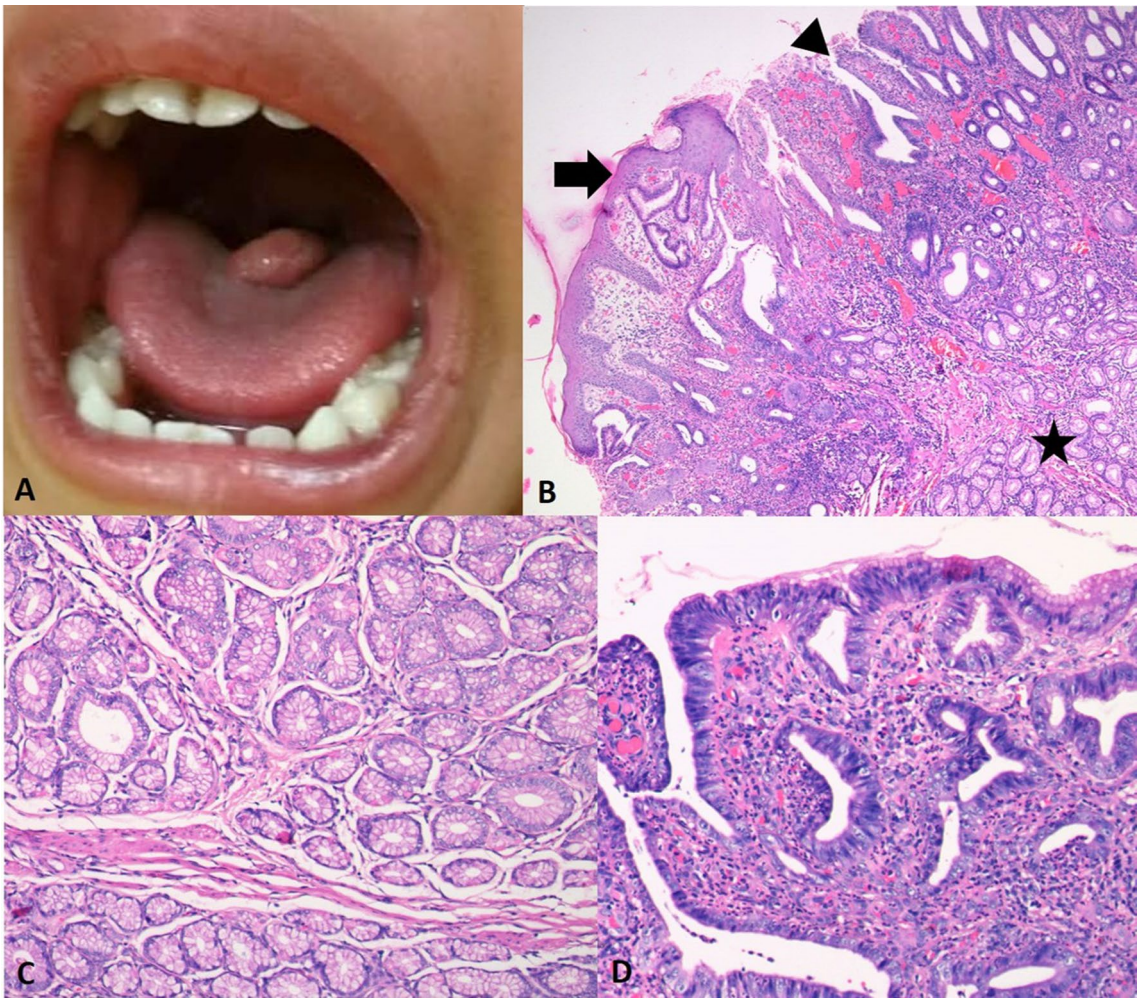

Fig. 1 Lingual choristoma with gastric epithelium **a** physical examination reveals a well-demarcated, smooth surfaced mass on the dorsal midline of the tongue. **b** Histologically, the lesion is partially lined by squamous mucosa (arrow); focal erosion (arrowhead) is noted, while the deep layer is occupied by back-to-back glands (asterisk) (Hematoxylin and eosin, 4x). **c** These mucinous glands are consistent with pyloric-type glands (Hematoxylin and eosin, 20x). **d** Focal reactive atypia is identified in the area showing acute inflammation (Hematoxylin and eosin, 10x)

Discussion

A choristoma represents histologically normal tissue in an abnormal location [1]. Other terms, including ectopia and heterotopia, have been used to describe the same pathologic process. These entities should be distinguished from hamartoma, a lesion composed of an excessive proliferation of disorganized tissues indigenous to the site [1].

Choristomas with gastric and/or intestinal epithelium, also known as heterotropic gastrointestinal mucosa or foregut duplication cysts, may manifest as a solid or cystic lesion. Lingual lesions account for 0.3% of foregut duplication cysts, and the most common locations include the anterior tongue and the floor of mouth [2, 3]. Lingual choristomas with gastric epithelium are believed to represent derivatives of entrapped endodermal precursors that would subsequently form the stomach, which are in close proximity to the pharyngeal arches containing the developing tongue [1, 4].

These lesions demonstrate a slight male predisposition (male to female ratio of 3:2) [2]. Although lingual choristomas with gastric epithelium are generally diagnosed in young children, they have rarely been reported in adults [4]. The lesions are usually asymptomatic, but a subset of cases may lead to increased salivation, altered speech, feeding difficulties, and airway compromise [5, 6]. MRI is the preferred imaging technique due to its excellent soft tissue contrast resolution. The lesions are usually hyperintense and nonenhancing on T2-weighted MRI, while the T1-weighted signal intensity is more variable [7].

Table 1 Clinicopathologic characteristics of congenital tongue lesions

Entity	Common tongue location	Histologic characteristics
Lingual choristomas	Anterior two-thirds	Solid/cystic lesions with gastric or intestinal epithelium (foregut duplication cyst), and/or respiratory epithelium (bronchogenic cyst). Others: osseous and neuroglial tissue.
Thyroglossal duct cysts	Posterior one-third	Cysts lined by nonkeratinizing stratified squamous and/or respiratory epithelium, lymphocytic inflammation, with/without thyroid follicles.
Ectopic lingual thyroid	Posterior one-third	Normofollicular thyroid tissue grows in between skeletal muscle and minor salivary glands.
Lingual hamartomas	Posterior one-third	The lesions may include vascular, nerve, skeletal, smooth muscle, adipose, or salivary gland tissue.
Vallecular cysts	Base of tongue or lingual surface of epiglottis	Cysts with squamous epithelial lining, may contain respiratory epithelium and mucous glands.
Lymphatic malformations	Anterior two-thirds	Malformed lymphatic channels, usually ill-defined and microcystic.
Dermoid cysts	Anterior two-thirds	Cysts lined by stratified squamous epithelium with associated appendages.

Histologically, lingual choristomas may be lined by stratified squamous, respiratory-type, gastric foveolar, and/or intestinal epithelium [1]. Other tissue types, including smooth muscle, adnexal structures, salivary gland, and neural elements, may be present. If representative tissues from ectoderm, mesoderm, and endoderm are identified, a diagnosis of congenital teratoid cyst can be rendered [1]. A more descriptive diagnosis (e.g. lingual choristoma with gastric epithelium, lingual choristoma with respiratory epithelium, lingual osseous choristoma, etc.) is recommended because of the variable diagnostic terms.

Pathologic evaluation is important because of the overlapping clinical and radiologic features of tongue lesions in the pediatric population. Kreiger et al. reviewed 135 tongue lesions in children, and reported vascular/lymphatic anomalies, mucus extravasation phenomenon, and hamartomatous lesions as the most common entities in their cohort [8]. Table 1 summarizes the clinicopathologic characteristics of selected congenital tongue lesions. In the appropriate clinical setting, neoplastic conditions such as schwannoma, granular cell tumor, alveolar rhabdomyosarcoma, alveolar soft part sarcoma, and Burkitt lymphoma, should also be considered [7, 8].

Surgical excision with removal of the mucosal lining is the treatment of choice for lingual choristoma with gastric epithelium, while disease recurrence is generally uncommon [5, 6]. Complications of lingual choristoma containing acid-producing gastric epithelium include ulceration and bleeding [6]. While the lesions are generally considered benign, Volchok et al. reported adenocarcinoma arising in a long-standing lingual foregut duplication cyst of an elderly man [9].

In conclusion, we presented a classic case of lingual choristoma with gastric epithelium. Pathologic evaluation is necessary for diagnostic confirmation, as more common entities, such as lingual hamartomas and lymphatic malformations, should be excluded. Surgical excision remains the treatment of choice for these lesions. Clinicians and pathologists should be familiar with this rare entity and include it in the differential diagnosis of congenital tongue lesions.

Funding The authors have no funding to declare.

Declarations

Conflict of interest The authors have no conflicts of interest to disclose.

Consent for publication Written informed consent cannot be obtained because the patient and his family have relocated since the time the personal information was originally collected. The clinical image and information does not contain identifiable characteristics.

Ethical approval This article does not involve the use of human or animal subjects and has complied with the ethical standards as outlined by the publisher.

Informed consent Written informed consent cannot be obtained because the patient and his family have relocated since the time the personal information was originally collected. This case report involves no more than minimal risk to the participant.

References

1. Ozolek JA, Tekkesin MS. The "-OMAS" and "-OPIAS": targeted and philosophical considerations regarding hamartomas, choristomas, teratomas, ectopias, and heterotopias in pediatric otorhinolaryngologic pathology. Head Neck Pathol. 2021;15:25–40.
2. Chapman MC, Soares BP, Li Y, Shum DJ, Glenn OA, Glastonbury CM, et al. Congenital oral masses an anatomic approach to diagnosis. Radio Graphics. 2019;39:1143–60.
3. Rosa RR, Burghgrave GS, Seixas AM, Padilha WSM, Siqueira CS, de Faria PR, et al. Heterotopic gastrointestinal mucosa of the tongue. J Pediatric. 2015;167:1161-1.e1.
4. Patel P, Branstetter BF, Myers EN. Lingual foregut duplication in a middle-aged adult. Am J Neuroradiol 2011;32:E40-1.

5. Jorquera JPC, Rubio-Palau J, Cazalla AA, Rodríguez-Carunchio L, Choristoma. A rare congenital tumor of the tongue. Ann Maxillofac Surg. 2016;6:311–3.
6. Chai RL, Ozolek JA, Branstetter BF, Mehta DK, Simons JP. Congenital choristomas of the oral cavity in children. Laryngoscope. 2011;121:2100–6.
7. Haber MA, Jaimes C, Lee EY, Juliano AF. Pediatric tongue lesions: an often-overlooked but important collection of diagnoses. Am J Roentgenol. 2020;214:1008–18.
8. Kreiger 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. Volchok J, Jaffer A, Cooper T, Al-Sabbagh A, Cavalli G. Adenocarcinoma arising in a lingual foregut duplication cyst. Arch Otolaryngol Head Neck Surg. 2007;133:717–9.