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Heterotopic Gastrointestinal Cyst of the Oral Cavity Radiology–Pathology Correlation

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Abstract
Heterotopic gastrointestinal cyst of the oral cavity is a rare congenital lesion that may arise from ectopic undifferentiated endodermal cells. Imaging, particularly MRI, is useful for surgical planning. On MRI, the cysts typically demonstrate high signal on T2-weighted sequences and variable signal on T1-weighted sequences, which can resemble other conditions, such as dermoids. On histology, the appearance of these lesions can be variable, and may include stratified squamous, simple and ciliated columnar, as well as foveolar and intestinal-type epithelia, often surrounding by smooth muscle. Complete surgical excision is the treatment of choice.

Keywords Oral cavity · Foregut duplication cyst · Radiology · Pathology

History

The patient is an ex-37 week female infant born via repeat C-section who was identified in utero as having a mass in the floor of the mouth. There was a reasonable nasopharyngeal airway and the trachea was filled with amniotic fluid, so an exit procedure was not performed. Delivery was complicated by breech presentation and the patient did not cry spontaneously at birth.

Radiographic Features

MRI performed shortly after birth demonstrated a large, well-defined, homogeneous midline oral cavity cyst with thinning of the oral tongue, but no erosions of the mandible (Fig. 1). The imaging findings were not particularly specific and the differential diagnoses based on the imaging included lymphatic malformation, inclusion cysts, thyroglossal duct cysts, ranula, and heterotopic gastrointestinal cyst. However, there were no septations, fluid levels, or enhancing components within the cyst, as might be encountered with lymphatic malformations, no restricted diffusion, as might be encountered with epidermoid cysts, no fat components, as might be encountered with dermoid cysts, and no significant extension into the tongue base, as might be encountered with thyroglossal duct cysts. In addition, the midline location was not typical of ranulas. The imaging findings were otherwise compatible with heterotopic gastrointestinal cyst.

Diagnosis

Grossly, the specimen appeared as a smooth-walled mass that measured approximately 3.7 cm in greatest dimension (Fig. 2). The lesion was comprised of a unilocular cyst filled with gray-white, cloudy, and viscous fluid. The internal lining of the cyst was smooth and a 0.6 cm wide area of raised pale-tan tissue was apparent. Representative sections were submitted including the entire raised area. Histologic examination of the tissue sections revealed a cyst lined in most areas by non-keratinizing stratified squamous epithelium with flaky keratinaceous debris occupying the cyst space (Fig. 3a). The wall was composed of fibrous tissue with vessels and lymphatics, as well as skeletal muscle fibers and seromucinous glands. Focally, corresponding to the grossly raised area, the cyst wall was lined

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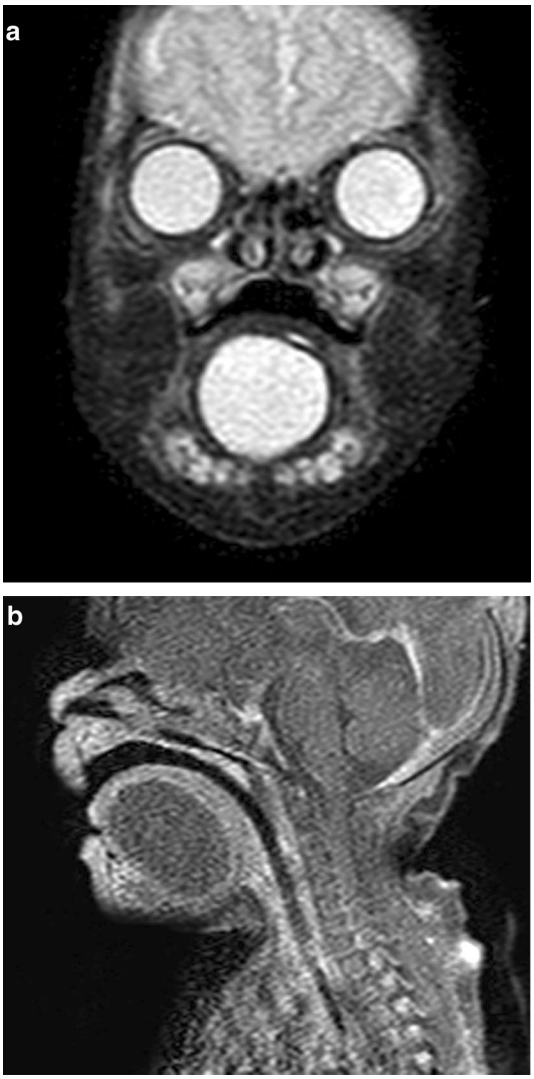

Fig. 1 Coronal STIR (**a**) and sagittal fat-suppressed post-contrast T1-weighted (**b**) MR images show a well-defined, unilocular, midline oral cavity cyst without enhancement. The cyst stretches and thins the oral tongue and does not extend below the floor of mouth or into the tongue base

by columnar cells with pale eosinophilic cytoplasm, consistent with gastric foveolar epithelium with intraluminal mucin secretion (Fig. 3b). Associated with this lining were glands analogous to gastric mucin pits containing chief and parietal cells. Focally, the foveolar lining exhibited goblet cells, consistent with intestinal metaplasia (Fig. 3c). These features are consistent with ectopic gastrointestinal mucosa lining a simple cyst. Therefore, a diagnosis of heterotopic gastrointestinal cyst of the oral cavity was rendered.

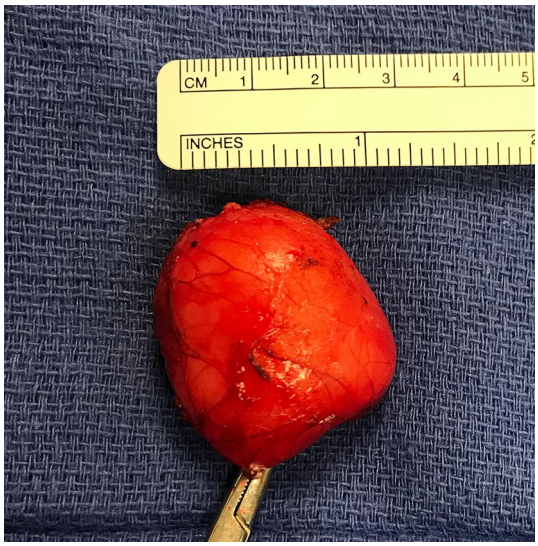

Fig. 2 Gross specimen photograph shows a smooth-walled mass

Discussion

Heterotopic gastrointestinal cyst of the oral cavity, also referred to as gastric cystic choristoma or enterocystoma, is a rare, congenital lesion that may result from ectopic undifferentiated endodermal cells in the developing stomodeum [1, 2]. This type of cyst is analogous to foregut duplication cysts, which are more commonly located in the thorax or abdomen, such as bronchogenic cysts, esophageal duplication cysts, or enteric duplication cysts [3].

Diagnostic imaging, particularly MRI is useful for preoperative planning of oral cavity cysts [4]. The lesions may be located in the midline or laterally and tend to appear as unilocular cysts without enhancing components [5]. On MRI, the cysts typically demonstrate high signal on T2-weighted sequences and variable signal on T1-weighted sequences. The differential diagnosis for congenital cystic lesions in the oral cavity includes dermoid and epidermoid cyst, thyroglossal duct cyst, lymphatic malformation, and mucocele or ranula [6]. Although these lesions may be indistinguishable from heterotopic gastrointestinal cysts of the oral cavity, dermoids can sometimes be characterized by the presence of fat contents, epidermoids may display restricted diffusion, thyroglossal duct cysts would be expected to have a component in the tongue base, lymphatic malformations can be multilocular and multispatial and may enhance, and ranulas tend to be lateralized and may extend below the floor of mouth [6].

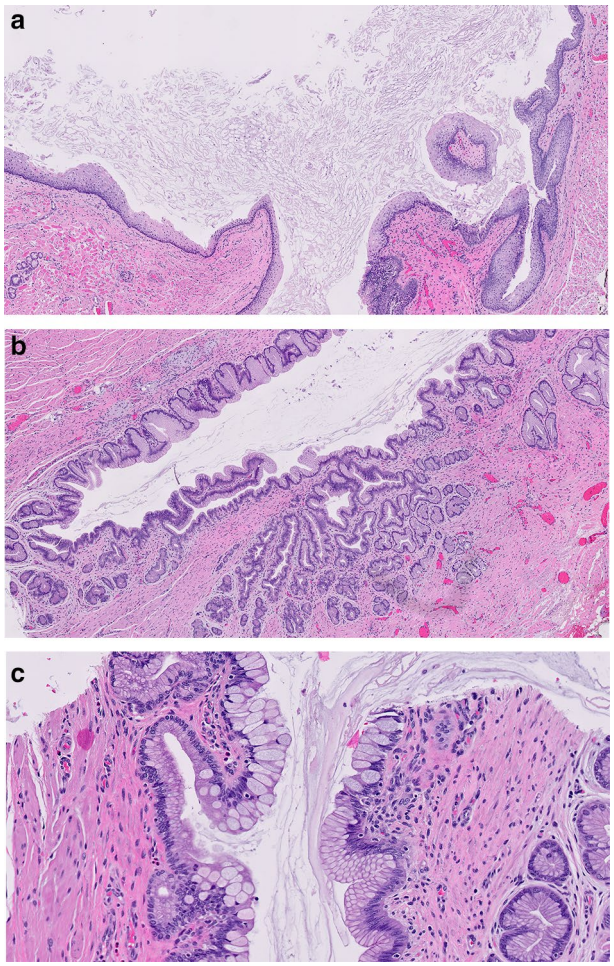

Fig. 3 Hematoxylin and eosin stained photomicrographs (**a**–**c**) show a cyst lined by stratified squamous epithelium and flaky keratinaceous debris occupying the cyst space. The cyst wall is composed of columnar cells with pale eosinophilic cytoplasm, consistent with gastric foveolar epithelium with intraluminal mucin secretion. Associated with this lining are glands analogous to gastric mucin pits containing chief and parietal cells, as well as goblet cells, consistent with intestinal metaplasia

Ultimately, the diagnosis of heterotopic gastrointestinal cyst of the oral cavity is made based on histopathology. The mucosa in the cyst may correspond to the normal mucosa at the anatomic level of the cyst and there are often different mucosal types within a single cyst [4]. Indeed, the histologic appearance of these lesions is variable, and may include stratified squamous, simple and ciliated columnar, as well as foveolar and intestinal-type epithelia. Smooth muscle is usually identified surrounding the cyst [4].

Patients do not necessarily experience respiratory compromise, despite the large size of the cyst [3]. Excision with complete removal of the cyst lining is generally curative, with recurrence only documented following incomplete excision [2].

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Gorlin RJ, Jirasek JE. Oral cyst containing gastric or intestinal mucosa. An unusual embryological accident or heterotopia. Arch Otolaryngol. 1970;91:594–7.
2. Daley TD, Wysocki GP, Lovas GL, et al. Heterotopic gastric cyst of the oral cavity. Head Neck Surg. 1984;7:168.
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;136:778–82.
4. Eaton D, Billings K, Timmons C, Booth T, Biavati JM. Congenital foregut duplication cysts of the anterior tongue. Arch Otolaryngol Head Neck Surg. 2001;127:1484–7.
5. Martone CH, Wolf SM, Wesley RK. Heterotopic gastrointestinal cyst of the oral cavity. J Oral Maxillofac Surg. 1992;50:1340–2.
6. Edwards RM, Chapman T, Horn DL, Paladin AM, Iyer RS. Imaging of pediatric floor of mouth lesions. Pediatr Radiol. 2013;43(5):523–35.