Congenital Foregut Duplication Cysts of the Anterior Tongue

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Objective: To review our experience with foregut duplication cysts of the anterior tongue, an unusual and rarely encountered mass in this location.

Design: A retrospective review of patients with anterior tongue foregut duplication cysts identified between 1990 and 2000.

Setting: Academic, tertiary care children’s medical center.

Patients: Six pediatric patients (5 boys and 1 girl) ranging in age from birth to 8 months at diagnosis.

Intervention: Three patients underwent preoperative magnetic resonance imaging (MRI). All 6 patients underwent excisional biopsy.

Main Outcome Measures: Clinical description of foregut duplication cysts, ability to make the diagnosis preoperatively, and recurrence rates.

Results: No patient presented with respiratory compromise, despite the large size of the anterior tongue masses (range, 1.5-2.4 cm). An MRI study was performed in 3 patients, all given a presumptive diagnosis of dermoid cyst based on the radiographic findings. No patient was diagnosed correctly prior to surgical excision. All patients underwent surgical excision, and the average time from birth to surgical excision was 11 months (range, 3 days to 3.7 years). Surgical pathologic findings were reported as a foregut duplication cyst (enterocystoma) in all patients, with 3 specimens containing foci of gastric mucosa. No recurrence has occurred at 1-month follow-up.

Conclusions: Foregut duplication cysts rarely present in the anterior tongue and are easily misdiagnosed preoperatively. An MRI study is helpful in preoperative planning, although all lesions were radiologically indistinguishable from dermoid cysts. These masses may be an underappreciated entity in the differential diagnosis of congenital anterior tongue masses.


FOREGUT (enteric) duplication cysts are classified as choristomas containing heterotopic islands of gastrointestinal mucosa. They consist of a cystic wall composed partly of stratified squamous epithelium and partly of gastrointestinal (columnar) epithelium. Variable amounts of parietal or chief cells, goblet cells, argentaffin cells, and Paneth cells are present. Smooth muscle is usually identified surrounding the cyst. Though enteric duplication cysts occur anywhere from the oral cavity to the rectum, they are rare in the oral cavity; only 21 cases have been reported. Most present asymptomatically shortly after birth, but the location of these lesions has the potential to cause respiratory and feeding difficulties. Magnetic resonance imaging (MRI) is the study of choice, but the diagnosis is made only with excisional biopsy. Treatment is surgical excision, with no reported recurrences. The cases presented here demonstrate the difficulty in making the diagnosis preoperatively.

RESULTS

The Table summarizes the pretreatment and histopathologic patient data. Five patients presented at birth with a mass in the anterior tongue/floor of the mouth (FOM) region (Figure 1). Two of these patients were asymptomatic, and 3 presented with difficulty feeding. The sixth patient was noted at birth to have a large protruding tongue, but an anterior FOM cyst was not recognized until age 8 months. No patient presented with respiratory compromise, despite the large size of the anterior tongue masses (range, 1.5-2.4 cm).

The differential diagnosis included dermoid cyst, ranula, cystic hygroma, he-
PATIENTS AND METHODS

This was a retrospective medical record review of children who underwent excision of foregut duplication cysts of the tongue. Between 1990 and 2000, 6 patients at Children's Medical Center in Dallas, Tex, underwent excision of anterior tongue cysts determined postsurgically to be enterocystomas.

Hospital, clinic, and surgical records were reviewed for age at diagnosis, presenting symptoms, sex, site of lesion, age at surgical excision, histopathologic and preoperative imaging findings, complications, and recurrences. All procedures were performed in the operating room at Children's Medical Center in Dallas. All patients were extubated postoperatively and had no postoperative complications. No recurrences were identified at 1-month follow-up.

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age at Diagnosis/Sex</th>
<th>Symptom</th>
<th>Examination Findings</th>
<th>Imaging Findings</th>
<th>Size/Pathologic Diagnosis</th>
<th>Age at Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Birth/M</td>
<td>Asymptomatic</td>
<td>Purple right FOM cyst</td>
<td>MRI: anterior tongue cyst (dermoid ?)</td>
<td>1.8 × 1.5 × 0.6-cm enteric duplication cyst with gastric mucosa</td>
<td>10 mo</td>
</tr>
<tr>
<td>2</td>
<td>Birth/M</td>
<td>Difficulty feeding</td>
<td>FOM mass</td>
<td>MRI: anterior sublingual space mass (dermoid ?)</td>
<td>2.4 × 2.2 × 0.3-cm FOM enteric duplication cyst with gastric mucosa and 1.6 × 1.2 × 0.4-cm sublingual dermoid</td>
<td>6 d</td>
</tr>
<tr>
<td>3</td>
<td>Birth/M</td>
<td>Difficulty feeding</td>
<td>Enlarged distal third of the tongue, pigmented (hemangioma?)</td>
<td>None</td>
<td>2.0 × 1.7 × 0.5-cm enteric duplication cyst</td>
<td>1.5 mo</td>
</tr>
<tr>
<td>4</td>
<td>Birth/M</td>
<td>Difficulty sucking</td>
<td>Anterior FOM mass</td>
<td>Lateral nasopharynx film: ranula in oral cavity?</td>
<td>2.4 × 1.8 × 1.1-cm enteric duplication cyst with gastric mucosa</td>
<td>3 d</td>
</tr>
<tr>
<td>5</td>
<td>8 mo/M</td>
<td>Asymptomatic, protruding tongue</td>
<td>Large protruding tongue due to FOM cystic mass</td>
<td>MRI: dermoid within intrinsic tongue muscle?</td>
<td>2.0 × 1.3 × 1.0-cm enteric duplication cyst</td>
<td>10 mo</td>
</tr>
<tr>
<td>6</td>
<td>Birth/F</td>
<td>Asymptomatic</td>
<td>2 Cystic lesions: tip of tongue and FOM</td>
<td>None</td>
<td>1.5 × 1.0 × 0.9-cm enteric duplication cyst</td>
<td>44 mo</td>
</tr>
</tbody>
</table>

*FOM indicates floor of mouth; MRI, magnetic resonance imaging.

The mucosa observed in the cyst may not correspond to the normal gastrointestinal mucosa at the anatomic level of the cyst, and mixed mucosal types within a single cyst are common. The endodermal derivation of the respiratory mucosa is reflected in the frequent presence of ciliated columnar epithelium in these cysts. Thus, although pure esophageal, gastric, and intestinal duplications are seen, the cysts often defy anatomic categorization and are more conveniently designated by the generic name of foregut duplication or foregut cyst. Broncho-

**COMMENT**

Enteric duplication cysts are cystic lesions containing a gastrointestinal mucosal lining. Some consist of only a mucosal lining, and others have a multilayered wall of mucosa, submucosa, and muscularis propria (Figure 6 and Figure 7). Three lesions in this series contained foci of gastric mucosa. They may be multiple and occur anywhere from the oral cavity to the anus, though they are rare in the oral cavity, with only 0.3% reported in the tongue.9

![Figure 1. Patient is a 2-day-old boy with a cystic mass on the anterior floor of the mouth.](https://jamanetwork.com/)

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genic cysts belong to this same group, being distinguished mainly by the presence of hyaline cartilage in the wall.

Figure 2. Midline sagittal T1-weighted image of a 10-month-old boy. A well-defined mildly and uniformly hyperintense mass is present within the anterior floor of the mouth (arrow).

Figure 3. Midline sagittal T1-weighted image of a 2-day-old boy. A predominantly isointense mass is present within the anterior tongue, and a focus of increased signal (arrow) appears within the anterior aspect of the mass, corresponding to dermal elements found pathologically.

Figure 4. Intraoperative dissection showing a well-defined, cystic mass on the floor of the mouth.

Figure 5. Gross specimen of a congenital foregut duplication cyst, cut open, revealing smooth mucosal lining. The units indicated on the ruler are centimeters.

Figure 6. Gastric foveolar epithelium (hematoxylin-eosin, original magnification ×10).

Figure 7. Respiratory epithelium, mucous glands, inner layer of smooth muscle, and outer layer of skeletal muscle (hematoxylin-eosin, original magnification ×4).

The embryology of duplications in the tongue and FOM region is unclear, and several theories on the pathogenesis have been proposed. Tongue development begins in the fourth week of gestation with the formation of the tuberculum impar (median tongue bud) and lateral lingual swellings from the first branchial arch mesenchyme. The lateral lingual swellings ultimately over-
grow the tuberculum impar and form the anterior two thirds of the tongue. The posterior one third of the tongue is formed from third and fourth branchial arch mesenchyme. The mucosa of the anterior two thirds of the tongue is of ectodermal origin, whereas the posterior one third is of endodermal origin.4

Four main theories have been proposed to explain how gastric mucosa (endoderm) becomes incorporated into the tongue. Gorlin and Jirasek1 suggested in 1970 that heterotopic gastric mucosa may be derived from entrapped embryonic gastrointestinal epithelium or ectoderm from the primitive stomodeum. This theory fails to explain the presence of esophageal, intestinal, or colonic mucosa anterior to this area.

Another theory, developed by Daley et al5 in 1984, proposed that endodermal cell rests of the stomodeum became trapped by the lateral lingual swellings and were subject to inductive influences causing differentiation into gastrointestinal epithelia. In 1988, Woolgar and Smith6 performed a mucin histochemical study of an enteric duplication cyst of the tongue and found well-differentiated columnar and goblet cells. These did not correspond precisely to normal gastrointestinal epithelium. This supported the theory that these cysts arose from primitive endodermal gastric mucosa subjected to inductive influences.

Veeneklaas7 initially proposed the currently accepted explanation in 1952, when he observed vertebral clefts and rib anomalies in association with intestinal duplications. He suggested that a disturbance in the development of the notochord and surrounding structures accounted for misplaced segments of gastrointestinal mucosa. In this situation, adherent endodermal cells became caught during the infolding of the notochordal plate. This split notochord syndrome is now the favored developmental theory.

The differential diagnosis includes dermoid cyst, cystic hygroma, hemangioma, ranula, and thyroglossal duct cyst. Most are diagnosed in asymptomatic infants, but they have the potential to go undetected for years if the cyst is small and asymptomatic.8 Difficulty with feeding is not unusual, as reported in our series. Large cysts in the newborn may initially cause airway obstruction, and needle aspiration may be used as a temporary measure to maintain the airway. Prenatal ultrasound has led to early diagnosis, and 2 cases of intraoral enteric duplication cysts have been detected in utero.2 In both cases, the cysts were initially decompressed and the airway secured prior to division of the placental cord. These patients were later treated with definitive surgical excision.

Magnetic resonance imaging and/or computed tomographic scans are helpful in determining the extent of the cyst. An MRI study, with its lack of ionizing radiation and superior soft tissue resolution, is the imaging study of choice. It allows an accurate assessment of the extent of tumor infiltration for preoperative planning. As demonstrated in this case series, enteric duplication cysts are often indistinguishable from dermoids on MRI because of the presence of proteinaceous fluid. They appear as cystic lesions that do not enhance with intravenous contrast. They may be hyperintense on T1-weighted images, depending on the amount and distribution of proteinaceous contents. They uniformly appear hyperintense on T2 and short T1 inversion recovery sequences.

CONCLUSIONS

The rare occurrence of foregut duplications in the oral cavity mandates vigilance with respect to the airway. An MRI study is a recommended part of the preoperative evaluation but cannot be relied on for definitive diagnosis because these lesions appear similar to dermoids. Complete cyst excision with removal of the mucosal lining is the treatment of choice. Aspiration alone results in recurrence, and the functional mucosa continues to secrete mucus if left intact. Also, there is a risk of ulceration and bleeding in the presence of acid-secreting gastric mucosa. The long-term prognosis is excellent, with no recurrences reported and complete recovery expected. This report comprises the largest series of these unusual lesions at a single institution.

Accepted for publication July 17, 2001.

Presented as a poster at the American Society of Pediatric Otolaryngology meeting, Scottsdale, Ariz, May 9-11, 2001.

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REFERENCES