Foregut duplication cysts are benign developmental anomalies that occur along the foregut-derived alimentary tract. As defined by Rickham et al, foregut duplications must (1) contain intramural smooth muscle, (2) be attached to a structure formed by the foregut, and (3) contain epithelium and/or mucosa derived from the foregut.1–2 Developed from heterotopic rests of the foregut, they may contain many different types of epithelium and mucosa including squamous, gastric, respiratory, and cuboidal. These lesions may also contain muscularis mucosa, submucosa, muscularis propia, ganglion cells, and pancreatic tissue. Their nomenclature varies, and depending in part on their particular anatomic location or histologic components, foregut duplication cysts have been variously called lingual choristoma, lingual cyst, enterogenous cyst, gastric mucosa choristoma, esophageal duplication cyst, bronchial duplication cyst, and alimentary tract cyst.2–3

Foregut duplication cysts, regardless of location, have an incidence of 1 in 4500 and most commonly arise in the thorax and abdomen, with only 0.3% of cases presenting in the head and neck. The oral cavity, specifically the oral tongue, accounts for 60% of the head and neck cases, and the majority of these are asymptomatic at presentation, with a smaller percentage presenting with feeding difficulties and, more seldom, respiratory distress.4 In this report, we describe the prenatal diagnosis, multidisciplinary antenatal planning, and management of a large cystic tongue lesion causing respiratory distress. Its imaging and pathologic features are also detailed herein.

Report of a Case

A 4.16-kg female was born at 39 weeks 2 days gestation to a 20-year-old healthy mother in her first pregnancy. The mother had been referred to our institution’s fetal care center for concern over a cystic tongue lesion. A prenatal screening ultrasound performed at 28 weeks gestation had shown a thick-walled introral cyst that measured 3.1 × 2.6 × 2.5 cm, deformed with swallowing and chewing motions, and protruded through the fetal mouth. Fetal magnetic resonance imaging (MRI) at that time showed a similar-sized lesion localized to the anterior tongue (Figure 1A).

A follow-up MRI examination conducted at 34 weeks gestation (not shown) showed slight interval growth of the tongue lesion and protrusion of the tongue out of the mouth but...
no involvement of the tongue base or compression of the oropharynx, hypopharynx, or trachea. Amniotic fluid was identified within the oral cavity, oropharynx, hypopharynx, trachea, esophagus, and stomach, and there was no polyhydramnios, indicating normal swallowing function.

At this stage, the otolaryngology, radiology, and obstetric teams discussed the possible need for an ex utero intrapartum treatment (EXIT) vs a planned cesarean delivery. On the basis of prenatal imaging results, the otolaryngology team was confident that intubation would be feasible at the time of birth or, at worst, that the tongue mass could be bypassed with a nasal airway and positive pressure. The teams concluded that a planned cesarean delivery with the otolaryngology team in the birth suite was most appropriate. The baby was born via a planned cesarean delivery with the otolaryngology team at the bedside. Apgar scores were 8 and 9 at 1 and 5 minutes, respectively. Because the newborn showed some signs of airway obstruction while supine and given arterial oxygen saturations of 80% to 85%, the decision was made to secure the airway. Direct laryngoscopy showed a large protuberant tongue mass, as well as the oropharynx, hypopharynx, and larynx.

Figure 1. Fetal and Neonatal Magnetic Resonance Imaging

A, Sagittal fast imaging using steady state acquisition image of the fetus at 28 weeks gestation showing a markedly bright, circumscribed lesion (white arrowhead) within the anterior aspect of the fetal tongue. The black arrowheads point to amniotic fluid within the oral cavity, nasopharynx, oropharynx, and hypopharynx. B, Postnatal sagittal T2-weighted image showing a macrolobulated, well-circumscribed cyst within the anterior tongue (arrowhead) without internal architecture, septations, or fluid-fluid levels. C, The postcontrast coronal T1-weighted spin echo with fat saturation image shows mild enhancement of the superior wall of the cyst (arrowhead).

Figure 2

The postnatal MRI examination performed on the first day of life showed a 3.7 × 2.3 × 1.6-cm unilocular, macrolobulated cyst within the anterior tongue. The cyst was hypointense on T1-weighted images; hyperintense on T2-weighted images; and was without internal soft-tissue components, flow voids, fluid-fluid levels, or restricted diffusion. On postcontrast imaging, mild rim enhancement of the lesion was also seen (Figure 1B and C). The clinicoanatomic differential diagnoses included foregut duplication cyst and less likely ranula, dermoid cyst, teratoma, cystic hygroma, or lingual thyroid.

The patient remained orally intubated in the neonatal intensive care unit until surgical excision of the tongue lesion was performed on the sixth day of life. Endoscopic findings prior to excision were unchanged, and the intubation was switched from oral to nasal. A ventral, anterior, midline incision was made in the tongue, and dissection was taken down to the capsule. A thick cyst wall was identified, and blunt dissection was used to define its anterior aspect. Bipolar electrocautery was used to tease the intrinsic tongue musculature from the capsule wall. As the posterior oral tongue was approached, the cyst was decompressed with an 18-gauge needle, revealing thick mucoid fluid. An incision was made through the anterior aspect of the cyst to allow probing within the lesion and to facilitate better definition of its posterior extent (Figure 3). The cyst was entirely confined to the oral tongue and did not approach the tongue base and was completely excised. After hemostasis and irrigation, the tongue musculature was reapproximated with interrupted absorbable sutures and a Penrose drain was left in place.

Gross examination of the resected specimen revealed a 3.5 × 2.1 × 0.6-cm cyst. Microscopically, it showed a mucosa lined by squamous, gastric foveolar, ciliated columnar, and cuboidal epithelium (Figure 4). A smooth muscle wall resembling muscularis propria of variable thickness was also seen.

Postoperatively, the patient was transferred to the neonatal intensive care unit intubated, sedated, and with a nasogastric tube for feeding. The patient was extubated on postoperative day 2, after which oral feedings were tolerated and there was no airway obstruction. The patient was discharged on postoperative day 8 (14th day of life).

Discussion

In this report, we describe the antenatal planning, birth, and airway management of a newborn who received a prenatal diagnosis of a large anterior cystic tongue mass, as well as the
surgical and postoperative management. Several case reports and case series have reported the diagnosis and management of foregut duplication cysts of the head and neck in children, but diagnosis prior to birth is rare (to our knowledge, only 5-6 cases in the English literature). Prenatal diagnosis of these lesions allows for a thoughtful multidisciplinary collaboration to formulate a careful delivery plan that ensures airway protection in a neonate at risk for upper airway obstruction.1,2,5,6

Preoperative imaging aids in narrowing the differential diagnoses of tongue masses and plays an important role in presurgical planning. Diagnostic considerations for a cystic lesion located within the anterior tongue and floor of mouth in a fetus include a foregut duplication cyst, dermoid cyst, ranula, teratoma, hemangioma, and venous or lymphatic malformation. Prenatal ultrasound and MRI aid in characterizing a fetal tongue lesion and help determine its precise location, its relationship to the airway, and the level and degree of airway obstruction. The presence of polyhydramnios, decreased swallowing, and tongue protrusion suggests airway obstruction.7,8 In regard to postnatal imaging, assessment with MRI is preferred over computed tomography because this modality provides superior soft-tissue contrast and ionizing radiation is avoided. On MRI, foregut duplication cysts can be of variable signal intensity on T1-weighted images, which is related to the protein content of the cyst fluid. On T2-weighted images, the cyst is typically bright and without internal septations or soft-tissue nodularity. Following the administration of contrast medium, the cyst wall may or may not be enhanced. The absence of restricted diffusion on diffusion-weighted imaging helps distinguish a foregut duplication cyst from a dermoid cyst.

The oral cavity, specifically the tongue, is the most common site for foregut duplication cysts in the head and neck. The most common presenting symptoms in older children are odynophagia, feeding difficulties, stridor, tongue edema, and speech difficulties. Neonates rarely present with respiratory distress secondary to airway obstruction. Only 1 of 31 patients in studies describing foregut duplication cysts in the head

Figure 2. Neonatal Photograph

Newborn intubated and sedated just after birth. Note the protruding tongue without obvious mucosal involvement.

Figure 3. Intraoperative Photographs

A, A ventral incision has been made along the anterior tongue. Dissection was taken down to the capsule. The surrounding musculature has been gently peeled away anteriorly. B, The anterior aspect has been opened and contents drained. C, The cavity closed down with a Penrose drain left in place.
Large Foregut Duplication Cyst of the Tongue

Conclusions

Given the extensive differential diagnosis of cystic head and neck lesions in neonates, imaging is recommended in all patients to localize and characterize the lesion and assess its extent because management of these lesions may differ substantially. For cases that are diagnosed antenatally, coordination of a multidisciplinary fetal care team and early discussions can optimize the predelivery workup and provide clear delivery and airway management plans. The oral cavity, specifically the tongue, is the most common site for foregut duplication cysts in the head and neck, with the majority of these lesions being asymptomatic. Early airway securement can ease transport considerations and facilitates additional imaging during the operative planning process. On the basis of our experience and review of the literature, we recommend complete surgical excision of oral foregut duplication cysts in the perinatal period to prevent subsequent complications such as feeding difficulties, infection, and ulceration.
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REFERENCES


