Management of a Large Antenatally Recognized Foregut Duplication Cyst of the Tongue Causing Respiratory Distress at Birth

Eric A. Gantwerker, MD, MS; Amy Lawrason Hughes, MD; V. Michelle Silvera, MD; Sara O. Vargas, MD; Reza Rahbar, MD, DMD

IMPORTANCE Foregut duplication cysts are benign developmental anomalies occurring along the foregut-derived portion of the alimentary tract. Several cases of foregut duplications in the head and neck region have been reported, most without airway symptoms. This case is an antenatally recognized anterior tongue lesion leading to respiratory difficulties at birth that was successfully managed by a coordinated fetal care team.

OBSERVATIONS We describe a 4.16-kg female born at full term whose anterior tongue lesion was noted on routine prenatal ultrasound. An airway management plan was developed by a multidisciplinary fetal care team, and the airway was controlled at the time of cesarean delivery. The lesion was completely excised on the sixth day of life without complications.

CONCLUSIONS AND RELEVANCE Given the extensive differential diagnosis of cystic head and neck lesions in neonates, imaging is recommended to localize and characterize the lesion 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. We recommend complete surgical excision of oral foregut duplication cysts in the perinatal period to prevent complications such as feeding difficulties, infection, and ulceration.

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.²³ 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 propria, 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.²³

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, is the most common site. 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, 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 (Figure 2) and a normal palate, supraglottis, glottis, and subglottis. A grade 2 laryngeal view and intubation were easily achieved with a Parson’s laryngoscope and rigid endoscope.

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 clinicoradiographic differential diagnoses included foregut duplication cyst and less likely ranula, dermoid cyst, teratoma, lymphatic malformation, and 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 anteriormost 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.
and neck presented with respiratory distress, and in that case, the lesion involved the vallecula and not the anterior tongue.\textsuperscript{1-6}

Prenatal diagnosis of these lesions permits anticipation of airway difficulty and enables creation of an airway management algorithm based on the location of the lesion. In a case series of 6 children from Children’s Medical Center in Dallas, Texas, prenatal ultrasound suggested the diagnosis of a foregut duplication cyst in 2 patients. In these newborns, the cysts were decompressed and the airway was secured prior to division of the placental cord. None of their 6 patients presented with airway obstruction.\textsuperscript{6}

In a review of 22 children with head and neck foregut duplication cysts treated at Boston Children’s Hospital, in only 1 patient was the cystic lesion diagnosed by means of antenatal ultrasound. This patient was stable at birth and did not require airway intervention. Of the 3 patients who received a diagnosis after birth in the perinatal period, only 1 presented with respiratory distress.\textsuperscript{1}

Antenatal diagnosis of an oral foregut duplication cyst presents a dilemma with regard to delivery management and requires a decision whether to perform an EXIT procedure. The perinatal airway management depends greatly on the location and size of the lesion. Our patient was managed with a planned cesarean delivery because of the anterior position of the mass within the tongue, the intact posterior airway, and the lack of tongue base involvement. Rousseau et al\textsuperscript{7} describe similar prenatal management considerations of these “enteric” duplication cysts and also emphasize the importance of the coordination of care of the obstetrical, pediatric, and surgical teams prior to delivery. At our institution, the perinatal care of these children often involves a fetal care team consisting of pediatric specialists from multiple disciplines including obstetrics, neuroradiology, otolaryngology, anesthesia, genetics, general surgery, and neonatology. Together, these specialists formulate a treatment plan including the best method and location of delivery, whether a fetal EXIT procedure is warranted, and what the best form of airway secure-

ment might be. At our institution, EXIT procedures are typically reserved for patients in whom airway securement is expected to be greatly compromised.

Early and complete surgical excision of foregut duplication cysts is recommended for the purpose of diagnosis and treatment. A complete resection can prevent the development of failure to thrive, potential size increases of the cyst from trauma or hemorrhage, and potential airway compromise.\textsuperscript{2} The surgical approach from the ventral tongue has the advantage of limited scarring and better restoration of intrinsic tongue musculature but does run the risk of tethering the tongue if the incision is taken too close to the floor of the mouth. Care must also be taken to avoid the sublingual salivary ducts in this location. Division of the cyst during dissection assists in identifying the posterior extent of the lesion and helps to avoid important neurovascular structures nearby. Bipolar cautery is useful in the excision for hemostasis of these lesions, as noted by El-Bitar et al.\textsuperscript{2}

Although clinical observation of a foregut duplication cyst may be a reasonable management option in the asymptomatic patient, it carries the risk of complications from subsequent infection, bleeding, trauma, enlargement, and ulceration if functioning gastric mucosa is present. Furthermore, without surgical excision or biopsy, a definitive diagnosis cannot be obtained. Needle aspiration as the sole treatment method for these lesions is not recommended because of the high recurrence rate and the potential for complications similar to those seen with observation. In the acute setting, needle decompression may aid in airway securement and facilitate feeding as a temporizing measure.\textsuperscript{1,6} Clinicians should be cautioned, however, that needle decompression can result in bleeding into the airway and therefore should be reserved for emergent situations. Marsupialization as a treatment option has been shown to be ineffective because of a high recurrence rate, and treatment with sclerotherapy is also of limited efficacy.\textsuperscript{3,5}

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.
ARTICLE INFORMATION
Submitted for Publication: May 30, 2014; accepted August 9, 2014.
Published Online: October 9, 2014. doi:10.1001/jamaoto.2014.2331.

Author Contributions: Dr Gantwerker had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.
Study concept and design: Gantwerker, Hughes.
Acquisition, analysis, or interpretation of data: Gantwerker, Silvera, Vargas, Rahbar.
Drafting of the manuscript: Gantwerker, Hughes, Rahbar.
Critical revision of the manuscript for important intellectual content: Gantwerker, Hughes, Silvera, Vargas.
Statistical analysis: Rahbar.
Administrative, technical, or material support: Gantwerker, Hughes.

Study supervision: Gantwerker.

Conflict of Interest Disclosures: None reported.

REFERENCES