Journal of Pediatric Surgery Case Reports 89 (2023) 102546

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: www.elsevier.com/locate/epsc

Successful management of a large lingual foregut duplication cyst with an ex-utero intrapartum treatment procedure

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ARTICLE INFO

Keywords:
Foregut duplication cyst
Ex-utero intrapartum treatment
Lingual cyst
Airway obstruction
Prenatal management

ABSTRACT

Foregut duplication cysts are rare congenital choristomas that sometimes cause feeding and respiratory problems, depending on their size and location. Prenatal ultrasonography revealed a fetus with a large oral cystic mass and polyhydramnios. Differential diagnoses included ranula, lymphangioma, and thyroglossal duct cysts. A multidisciplinary fetal care team devised an airway management plan. We performed ex-utero intrapartum treatment (EXIT) because the origin and patency of the fetal airway could not be confirmed using fetal magnetic resonance imaging. At delivery, a lingual cyst occupied the oral cavity and protruded from the mouth. Oral intubation was performed after the aspiration of the cyst during the EXIT. On day 16, the cyst was completely excised. Histopathological examination revealed a foregut duplication cyst lined by the respiratory and gastric epithelium. Lingual foregut duplication cysts pose a risk for airway obstruction. For prenatally diagnosed cases, coordination of a multidisciplinary fetal care team and early discussions can optimize the plan for prenatal management, including the EXIT procedure.

Foregut duplication cysts are rare congenital choristomas that sometimes cause feeding and respiratory problems, depending on their size and location. Here, we present a case of a prenatally identified large oral cystic mass that caused respiratory difficulties at birth.

The mass was successfully managed using an ex-utero intrapartum treatment (EXIT) procedure.

1. Case report

Prenatal ultra-sonography at 24 weeks gestation showed a female fetus with a large oral cystic mass and polyhydramnios. Fetal magnetic resonance imaging (MRI) at 26 weeks showed a cystic mass that occupied the oral cavity and extended to the pharynx (Fig. 1). Differential diagnoses included ranula, lymphangioma, and thyroglossal duct cysts. A multidisciplinary fetal care team devised an airway management plan. We performed an EXIT procedure because the origin and patency of the fetal airway could not be confirmed using fetal MRI.

EXIT was performed at 35 weeks of gestation (Table 1). At delivery, a lingual cyst occupied the oral cavity and protruded from the mouth (Fig. 2).

Oral intubation is a safe procedure after the aspiration of a cyst during EXIT. The fluid drained from the cyst was a pale milky-brown liquid containing numerous inflammatory cells, mainly neutrophils. After puncture and aspiration, the tongue cyst returned to a size that occupied the oral cavity within a few days, and ultrasonography revealed a low echoic cyst that occupied the entire tongue. MRI on day 7 showed a 2.5 × 2.1 × 1.6 cm tongue cyst with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (Fig. 3). It had clear borders and no septum and was suspected to be a thyroglossal duct cyst or ranula. On day

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https://doi.org/10.1016/j.epsc.2022.102546
Received 22 November 2022; Accepted 13 December 2022
Available online 14 December 2022

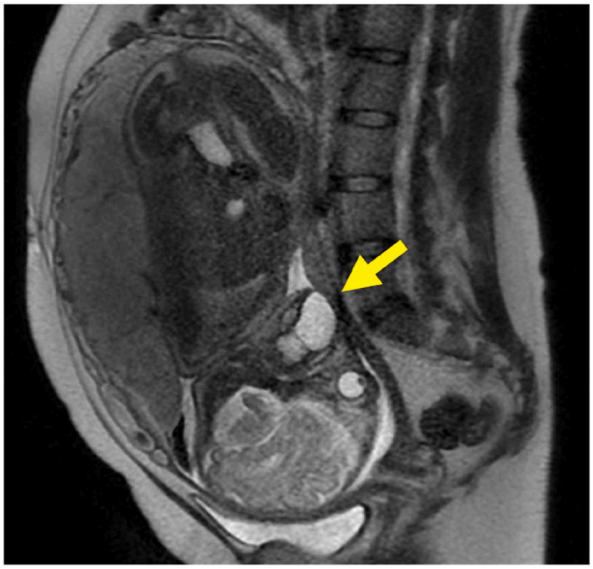

Fig. 1. Fetal MRI: MRI at 26 weeks' gestation showing a cystic mass protruding from oral cavity (arrow).

Table 1
Details about EXIT procedure in this case.

EXIT procedure	
Gestational age	35 weeks
Birth weight	2245 g
Time on placental support	8 min
Time to intubation	1 min
Maternal blood loss	720 m
Autologous blood transfusion	820ml

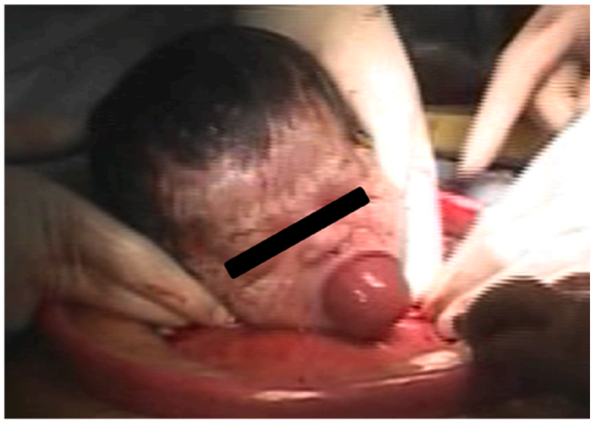

Fig. 2. Exposure of fetal head during an EXIT: Photograph taken immediately after exposing the fetal head from the uterus while maintaining placental blood flow with EXIT.

16, the cyst was completely excised. The cyst had a circumscribed wall and was easily separated from the tongue (Fig. 4). Histopathological examination revealed a foregut duplication cyst lined by respiratory and gastric epithelium (Fig. 5). She was discharged on day 34 following an uneventful postoperative course and oral feeding. During infancy, she had a history of involuntary mouth opening, but she could close her mouth and had no dysarthria. As she grew older, her mouth opened less frequently. She is now seven years old without any significant issues.

2. Discussion

Ex utero intrapartum treatment (EXIT) has been successfully used to manage various forms of fetal airway obstruction, allowing airway control at birth. Advances in prenatal diagnosis have enabled the detection of various congenital disabilities that can poten-

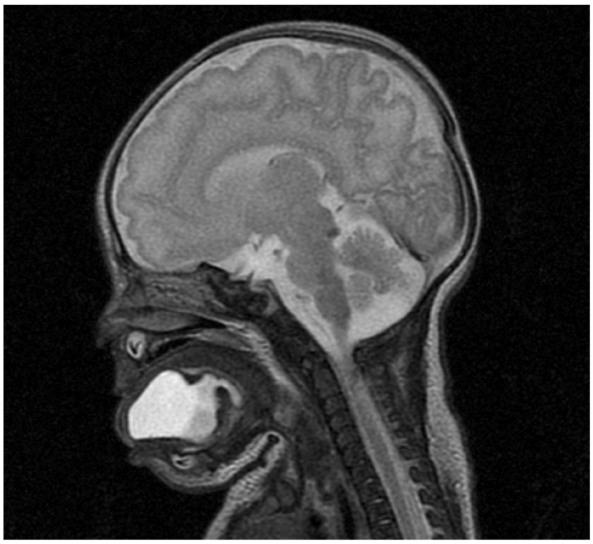

Fig. 3. MRI on day7: Postnatal T2-weighted MRI showing oral cystic mass measuring 2.5 × 2.1 × 1.6cm.

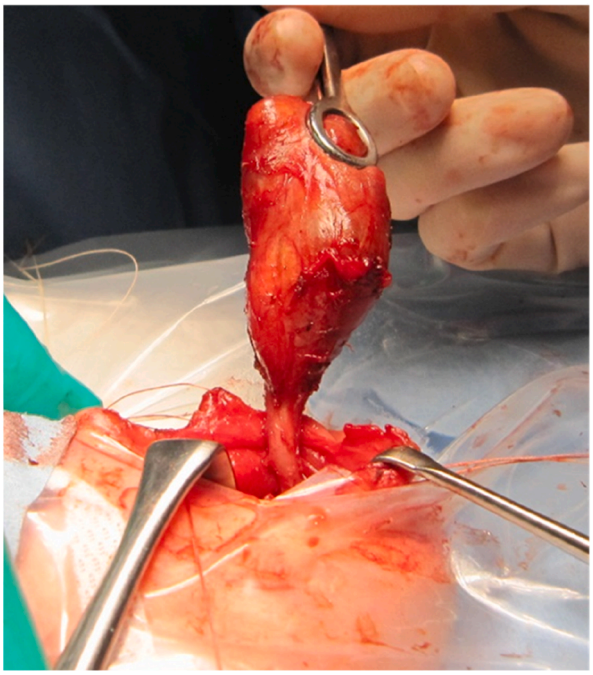

Fig. 4. Intraoperative photograph: The cyst was completely excised on day 16 of life. The cyst wall appeared circumscribed and was easily separated from the tongue.

tially obstruct the fetal airway or impede ventilation at birth [1–3]. The EXIT procedure is characterized by hysterotomy and partial delivery of the head and upper torso of the fetus while maintaining uteroplacental gas exchange through the umbilical cord. When the procedure is performed for an obstructed or difficult airway, the umbilical cord is clamped and cut for full delivery of the neonate once the airway is secured and adequate ventilation is established. The EXIT procedure may be performed to secure an airway in cases of airway obstruction or aberrant airway anatomy, head and neck mass resection, placement of ECMO cannulas, or separation of conjoined twins [2–4].

Foregut duplication cysts are benign developmental anomalies containing foregut derivatives. Traditionally, three criteria must be met to diagnose foregut duplication cysts: they must (1) be covered by a smooth muscle layer, (2) contain epithelium derived from the foregut, and (3) be attached to a portion of the foregut. Foregut duplication cysts feature a wide variability of mucosal linings, typically involving gastric, squamous, intestinal, or respiratory epithelia or any combination of these epithelia. Foregut duplication cysts occur most commonly in the chest and abdomen. In the head and neck, the most common site of foregut cysts is the oral cavity, especially in the anterior aspect of the tongue and on the floor of the mouth [5,6]. The etiology remains unclear owing to the variability in

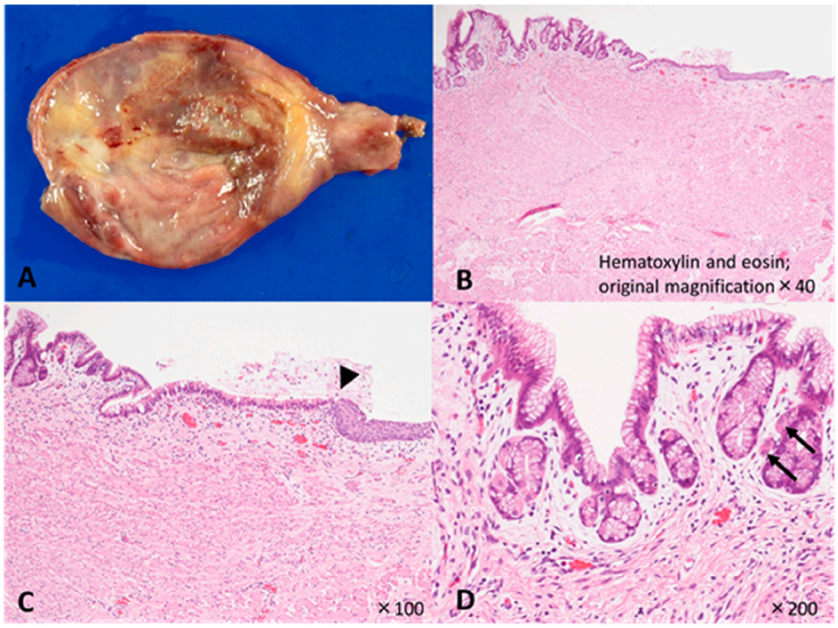

Fig. 5. Pathological findings of the lingual cyst: A: Macroscopic findings of the lingual cyst. B: Low magnification view of the cyst showing several types of epithelium. C: Transition from respiratory-type stratified squamous epithelium to intestinal-type epithelium (arrowhead). D: Gastric foveolar epithelium including parietal cells (arrows).

mucosal linings and locations across reported cases. The observation of gastric, squamous, respiratory, or intestinal epithelium is consistent with foregut cells and suggests an endodermal origin. Perhaps the most widely accepted and frequently cited theory discussing foregut cyst formation was first elucidated by Veeneklaas in 1952 [7]. They proposed that derangement of notochord development and surrounding structures can lead to cyst formation due to endodermal cell entrapment or adherence during notochord plate infolding. The Veeneklaas theory can potentially be used to describe most, if not all, cases of cyst development distal to the oral cavity, which includes the majority of these cysts as a category but does not sufficiently describe the presentation of lingual cysts [8,9].

Tongue development begins in the fourth week of gestation, with the formation of the tuberculum impar (median tongue bud) and lateral lingual swelling from the first branchial arch mesenchyme. The lateral lingual swelling eventually overgrows the tuberculum impar and forms the anterior two-thirds of the tongue. The posterior one-third of the tongue is formed by the 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 [9]. Foregut duplication cysts of the tongue may thus form small inclusions of epithelium that are retained within the substance of the tongue along the lines of fusion of the various primordia that contribute to organ formation. In the presence of epithelial islands, mesenchymal components may be subsequently induced to form an organoid structure that is evident to a variable extent in these lesions. However, given this hypothesis, the presence of mucosal types that differ from those typically found in the mouth is difficult to reconcile [9].

Diseases like dermoid cysts, lymphangiomas, teratomas, ranula, and foregut cysts can make the prenatal diagnosis of oral cysts challenging. Giant cystic lesions in the oral cavity are associated with a risk of postnatal respiratory compromise; therefore, a multidisciplinary prenatal delivery plan is necessary. Imaging studies, such as fetal MRI, are useful in determining airway patency, but if airway patency is not certain, a delivery plan, including EXIT, should be developed. In our case, we decided to secure the airway under EXIT because there was no prenatal diagnosis, and we considered that there was a risk of airway obstruction. After the child's head was extracted, the tongue was punctured under echocardiographic guidance, and the child was easily intubated. The same procedure may have been possible with a conventional cesarean section. There are reports of tongue foregut cysts successfully delivered by conventional cesarean section, avoiding EXIT because fetal MRI confirmed that the airway was passable [10,11]. However, because the child's condition was not fully predicted before birth, EXIT allowed us to secure the airway, ensuring more time. EXIT is also associated with risks such as maternal hemorrhage. Therefore, it is important to discuss treatment plans in a multidisciplinary manner prior to birth.

Since the tongue cyst, in this case, returned to its original large size shortly after the puncture, cystectomy was performed in the neonatal period under intubation. The histopathological findings were consistent with a foregut duplication cyst. Most foregut duplication cysts of the tongue are asymptomatic, with a smaller percentage presenting with feeding difficulties, odynophagia, stridor, tongue edema, and speech difficulties [6]. There have been reports of large cysts with no respiratory compromise or respiratory control with only nasal continuous positive airway pressure [12]. However, large cysts in newborns can potentially cause airway obstruction, so it has been reported that the patient should be intubated after temporarily securing the airway with puncture aspiration [13] or using a bronchoscope [10,14]. Most of these lesions can be treated by surgical excision, with a good postoperative course and a low recurrence rate.

Although foregut cysts of the tongue are extremely rare, MRI in reported cases, including our case, commonly shows a well-circumscribed cyst within the anterior tongue [8,10–12,14,15]. A foregut duplication cyst should be considered in the differential diagnosis when similar findings are seen on fetal or postnatal MRI.

3. Conclusion

Airway obstruction can occur because of a lingual foregut duplication cyst. Coordinating a multidisciplinary fetal care team and having early discussions can optimize the plan for prenatal management, including the EXIT procedure, for cases diagnosed in utero.

Funding

No funding or grant support.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Patient consent

Parental informed consent was obtained for this presentation.

Ethics statement

Ethical approval was waived by the institutional review board because this study is a case report.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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