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Review article

The delivered promise of prenatal imaging and a challenge to the utility of sildenafil for severe lymphatic malformations

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ABSTRACT

We report the case of an infant with severe lymphatic malformation necessitating ex-utero intrapartum treatment (EXIT) procedure and examine recent advances in high resolution ultrasonography and magnetic resonance imaging, which allow for improved prenatal diagnosis of lesions that cause critical airway obstruction in the neonate. Treatments for lymphatic malformations including surgical resection, sclerotherapy, coblation, and sildenafil are discussed. Our patient did not have any reduction in the size of the lymphatic malformation from sildenafil as suggested in another series.

Contents

1. Introduction

Recent advances in high resolution ultrasongraphy (US) and magnetic resonance imaging (MRI) allow for improved prenatal diagnosis of lesions that may cause airway obstruction in the neonate. Despite these advances, determining the specific pathology of obstructive neonatal lesions prior to delivery remains difficult. Subtle clues can suggest partial versus complete airway obstruction.

This case highlights the sequential use of imaging techniques to assess prenatal airway pathology, and the use of laryngeal mask airway (LMA) as a temporizing bridge to allow for surgical airway intervention. It also demonstrates an instance in which medical management with sildenafil was not beneficial contrary to previously reports [1,2].

2. Case

A 31-year-old G1P0 female underwent fetal ultrasound at 22-weeks gestation, which demonstrated a large multi-septated

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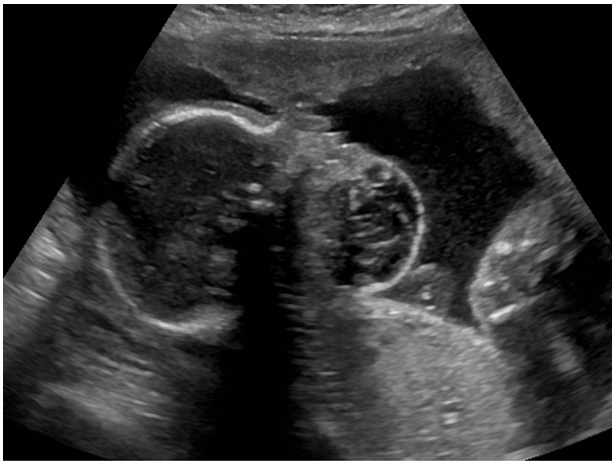

Fig. 1. A large multiseptated cystic mass located within the fetus's anterior neck.

cystic mass located within the fetus's anterior neck (Fig. 1). There were no calcifications seen, and vascularity and solid components were minimal. Amniotic fluid volume was normal. A fetal magnetic resonance imaging (Fetal MRI) was then performed at 28 and 34 weeks gestation. Fetal MRI revealed progressive growth ($8.7 \times 6.5 \times 6.0$ cm) with multilocular fluid levels. Polyhydramnios had developed and the lesion obstructed the esophagus, larynx and trachea, with extension into the anterior mediastinum (Fig. 2). In addition, in this fetus, there were small amounts of fluid in the cervical and thoracic trachea on Fetal MRI imaging, suggesting partial airway patency.

The increasing size of the mass and progressive narrowing of the glottic region on follow up Fetal MRI exams combined with the development of polyhdyramnios made EXIT a prudent choice. Tracking of the amniotic fluid index (increased to 34), which provided an indicator of airway involvement, was abnormally increased during her third trimester.

The EXIT procedure was planned for 40 weeks gestation but the mother delivered prematurely at 35 weeks, which is common when polyhydramnios is present [3]. An EXIT procedure was performed and our infant was intubated after direct laryngoscopy confirmed an obstructed airway (Fig. 3). Our neonate, however, became more hypoxic and bradycardic despite successful intubation because of fluid within the lungs; as a result, we substituted a laryngeal mask airway (LMA) for the intubation, followed by a tracheotomy (Fig. 4). The neonate was stabilized in the operating room and transferred to NICU for further care without neurological sequelae.

The child has undergone 25 serial treatments including over 20 episodes of sclerotherapy with doxycycline/bleomycin (beginning at 1 week of age), coblation of base of tongue and floor of mouth, and 3 cervical surgical debulkings with partial resection. Additionally, she has undergone CO_2 laser resection of laryngeal lesions. Her expected results have been satisfactory but she continues to have a need for tracheostomy tube for respiration in addition to gastrostomy tube. She is able to phonate and take sips of water but does not tolerate tracheotomy capping or oral food intake. The patient was offered a protocol with sildenafil, which did not show any reduction in size but did show slight subjective softening, during a trial of 12 months at 1.0 mg/kg of body weight administered three times daily. The infant is now 3 years of age and healthy but remains tracheostomy and gastrostomy tube dependent (Fig. 5). Her family is currently contemplating treatment with rapamycin [4].

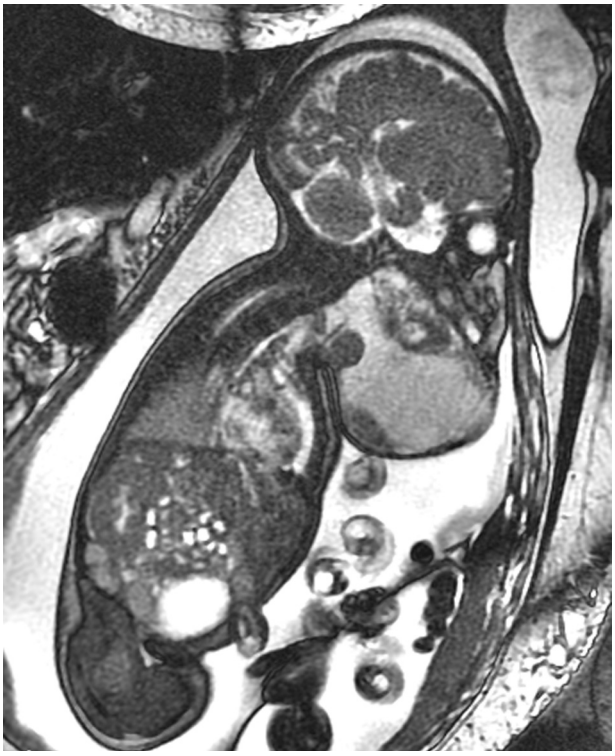

Fig. 2. T2—weighted sequences demonstrate high signal intensity from fluid filled structures of the oropharynx, trachea and bronchi. In this case there is obstruction of the fluid-filled fetal trachea.

3. Discussion

Advances in ultrasonography (US) and magnetic resonance imaging (MRI) have resulted in improved prenatal diagnosis of lesions that cause airway obstruction. This imaging provides essential information that allows the clinician to determine the best method to secure the airway at birth. Complex asymmetric cystic masses, which violate tissue planes such as the one described in our patient, are most often LMs. LMs may undergo rapid enlargement from internal bleeding into the lymphatic channels during birth trauma, and/or infection, which highlights the importance of early recognition and management. In severe cases, LMs can lead to fatal airway obstruction at delivery [5].

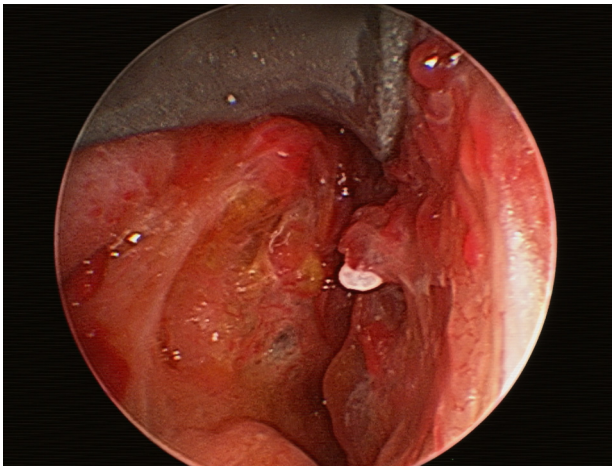

Fig. 3. Direct laryngoscopy revealing near complete obstruction of the larynx.