MR imaging and ultrasound of fetal cervical cystic lymphangioma: utility in antepartum treatment planning

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ABSTRACT
We present a case of fetal cystic lymphangioma that was initially diagnosed by ultrasonography and further evaluated by prenatal MR imaging. MR imaging findings aided in improved delineation of the neck mass. T2-weighted MR images revealed partial compression of the airway by the neck mass. This information was useful in the decision to use ex utero intrapartum treatment (EXIT) and helped surgeons in planning their approach to establish airway control during delivery.

Key words: • lymphangioma, cystic • fetus • magnetic resonance imaging • ultrasonography

Ultrasonography (US) has been used as the primary screening method for prenatal diagnosis. However, because US reveals limited anatomical information in certain anomalies, magnetic resonance (MR) imaging has found a more important role in prenatal imaging in recent years (1, 2). For example, fetal neck masses should be documented in detail and US may not present necessary detailed anatomical information such as the relationship between the mass and the airway of the fetus. Fetal MR imaging, however, may provide additional detailed anatomical information that could aid in planning the procedures necessary for airway access (3, 4).

We present a case of fetal cervical cystic lymphangioma diagnosed by US and further evaluated with prenatal MR imaging for antepartum treatment planning.

Case report
A 19-year-old gravida 2, para 1 woman was referred for evaluation of a fetal neck mass that had been identified on US at 12 weeks' gestation. Serial US examinations confirmed a mixed solid and cystic mass located within the anterior neck region, with dimensions of approximately 9 × 4 × 8.5 cm (Figure 1). No other fetal abnormalities were identified on US. US-guided amniocentesis and cytogenetic analysis revealed no chromosomal abnormality.

Following informed consent, fetal MR imaging was performed at 28 weeks' gestation. Fast spin-echo T2-weighted images were obtained in axial, sagittal, and coronal planes with a 1.5 Tesla MR imaging unit. T2-weighted images demonstrated a large mixed solid and cystic mass, arising from the region of the mandible and extending into the thoracic inlet, involving the entire anterior neck (Figure 2). Two areas of hypointensity suggesting calcifications were identified. These findings were thought to be most consistent with a cystic teratoma although cystic lymphangioma/hemangioma was also included in the differential diagnosis. MR imaging also demonstrated narrowing of the larynx and partial compression of the trachea, as well as the development of polyhydramnios.

The infant was delivered by cesarean section with EXIT (ex utero intrapartum treatment) procedure to aid in protection of the infant’s airway and immediately underwent partial resection of the mass. Additional surgery was anticipated due to incomplete resection of the mass lesion. Pathological study of the mass revealed cystic lymphangioma involving the mastoids, the right parotid gland, and the bilateral large vessels of the neck.

Discussion
Cystic lymphangiomas (hygromas) are rare, benign congenital tu-
mors that arise from sequestered lymphatic sacs that fail to communicate with the main lymphatic channels and represent as much as 6% of the benign tumors of children (5). These lesions are located in the neck and the lower face region in 75%-80% of the cases, and are usually within the posterior cervical triangle. In 10% of cystic hygroma cases, there is intrathoracic extension. As the mass enlarges, it may violate tissue planes, leading to airway compression (6, 7). Complete surgical excision is necessary for cure. However, complete surgical resection is often difficult, as the masses tend to be infiltrative. Interestingly, spontaneous regression is seen in 10%-15% of the cases (8).

Fetal lymphangiomas are rare, but are associated with high morbidity and mortality if airway compression occurs. Hence, affected infants need relief of airway compression immediately after birth. For this reason, it is of paramount importance to identify airway obstruction prenatally. In cases involving large hygromas presented during late stages of the gestation, perinatal coordination between various medical teams (particularly radiology, obstetrics, and pediatric surgery) may be necessary for successful delivery and to prevent infantile respiratory obstruction (6).

Recently, with the therapeutic option of operating on placental support (the EXIT procedure), as much detailed anatomical information as possible is crucial for planning of the procedure (4). Prenatal US reveals the mass and may also reveal an airway obstruction (by secondary signs such as polyhydramnios, decreased swallowing, or tongue protrusion) (6,7). However, assessment of the extent of the mass and the direct visualization of both the larynx and trachea is difficult on US. As such, the surgeons may be uncomfortable in planning an intervention with the low anatomical detail expressed by obstetric US (7). Fetal MR imaging allows enhanced global imaging of these masses. The ability of MR imaging to visualize the fluid-filled airway is most critical in estimating the severity of airway obstruction as well as planning an approach for possible tracheostomy and resection of the mass (7). Anatomical relationships and tissue characteristics are demonstrated by MR imaging with superior detail, except for calcifications (which are better noted with US) (7). Of particular significance in the use of MR imaging is the high signal intensity of the fetal airway (from the amniotic fluid on T2-weighted images), which provides excellent contrast resolution and allows assessment of the extent of airway displacement or compression. MR imaging also allows enhanced evaluation of possible extension into the thoracic cavity (7) and the relation of the mass to the neck vessels (9).

In our patient, MR imaging provided comprehensive information about

Figure 1. Axial ultrasonography image demonstrates a heterogenous mass with cystic and solid components in the anterior cervical region (arrows).

Figure 2. a-c. a, b. Axial T2-weighted MR images (a: superior level, b: inferior level) demonstrate heterogenous mass in the anterior neck region (arrows). Note the partial compression of the airway by the mass (arrowhead, a). c. Sagittal T2-weighted MR image demonstrates a heterogenous mass with cystic and solid components (arrowheads) extending from the mandible through the thoracic inlet (arrows) into the superior mediastinum.
both the anatomy and extension of the tumor and the compression of
the airway. Of the imaging examinations performed prior to the EXIT
procedure, MR imaging was believed to provide the surgeon with better
detail concerning the size and position of the mass, as well as its relationship
to the airway in comparison to imaging obtained with US. The infant was
intubated and a tracheostomy was placed during the delivery. Subse-
quently the patient underwent partial resection of the tumor, which was
planned antepartum.

In conclusion, MR imaging provided excellent in utero visualization of a large
fetal neck mass. The fetal MR imaging was essential in planning the mode of
delivery and establishing control of the airway in the delivery room.

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