Case Report

Fetal Neck Myofibroma

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Magnetic resonance imaging (MRI), as an adjunct to ultrasonography, has become a promising tool in prenatal diagnosis and therapy. In this report, the authors described a case of giant solid mass arising in the fetal neck region diagnosed by prenatal sonographic examination at the gestational age of 33 weeks'. MRI was used to confirm the diagnosis, and to assist fetal airway assessment. Due to the concern of fetal airway compromise, the ex utero intrapartum treatment (EXIT) was strategically planned with help from specialists in the according fields. This allowed the authors to secure the fetal airway before fetomaternal circulation was disconnected. It was performed successfully through Cesarean section at the time of birth. Histopathology revealed infantile myofibroma, which is a rare form of such a tumor arising on the fetal head and neck region diagnosed prenatally.

Keywords: *Fetal MRI, Infantile myofribroma, EXIT, Fetal therapy*

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Case Report

A case of a Thai pregnant woman was referred to the Maternal Fetal Medicine Unit, Faculty of Medicine Siriraj Hospital at the menstrual age of 33 weeks' due to an increasing abdominal discomfort for 2 weeks, along with large-for-date uterine size. She was a 34-year old woman, gravida 2 para 1, with a history of an uncomplicated vaginal delivery of an otherwise healthy baby girl seven years ago. Her past history was significant for hemoglobin E trait. She had had her routine anatomy scan at the menstrual age of 18 weeks', where no anomalies were found. Her antenatal history was otherwise unremarkable.

She began her prenatal care at the menstrual age of 27 weeks' complaining of an abdominal tightness. Physical examination was unremarkable at that time, except that the baby was in the breech position. She was scheduled for the next visit in 2 weeks, where her uterine size was found to be too large for date. The clinical impression was polyhydramnios, and hence the consultation was made for a detailed anatomy scan.

The ultrasonography revealed a single fetus in vertex position, with fetal parameters consistent with menstrual age of 35 weeks'. Amniotic fluid index (AFI) was 41 centimeters. There was a homogeneous solid mass, measuring 108×92 millimeters on the anterior of the fetal neck, extending onto the anterior chest wall, as shown in Fig. 1. This suggested a rapid growth of this tumor, since it was not found on the previous anatomy scan at 18 menstrual weeks'. Anatomy of the neck and chest was difficult to evaluate due to the acoustic shadowing of this mass. Borderline cardiomegaly was also seen from the scan. Otherwise, the baby appeared to be active. Amniodrainage of 2 liters was performed to alleviate the mother's respiratory distress symptom.

Compression of the esophagus was suspected due to the presence of polyhydramnios. Due to the concern that this mass might compromise the airway as well, and if so, the baby is at risk of fatal asphyxiation at the time of birth when the umbilical cord is clamped and cut. Patency of the trachea was not adequately evaluated from sonographic images. Decision

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was, therefore, made to perform a fetal MRI for a better assessment of fetal airway. Balanced Turbo Fields Echo (B-TFE) technique was used on multiple planes with maternal breath holding for demonstrating fetal anatomy. Maternal sedation or fetal paralysis was not required. MRI demonstrated a huge low signal intensity mass measuring 110 × 100 millimeters in size at the anterior neck extending onto the upper chest wall. The feeding vessels were demonstrated. Obstruction of the esophagous was suspected due to the lack of fluid-like signal intensity in the esophageal canal, as shown in Fig. 2 and 3. This explained the presence of polyhydramnios. The normal high signal intensity airway structure was seen posterior to the esophagous, indicating patent trachea with no evidence of airway obstruction. The fetal neck was apparently fixed in extension position. At this point, the discussion was made to continue the pregnancy to term. Cesarean section was considered due to malposition of the fetal head, which might obstruct the normal mechanism of labor.

The patient spontaneously ruptured her membranes at the menstrual age of 35 weeks. Given the size and location of this mass, the airway patency of this fetus at birth was still of concern. Therefore, a multidisciplinary team approach was set up to perform fetal intubation through the Cesarean section wound. The team composed of the obstetricians, neonatologists, and anesthesiologists, studied the fetal MRI to plan a strategy for fetal intubation. Fetal trachea diameter measured from the MRI was approximately 5 millimeters, which is consistent with the neonatal endotracheal tube number 3.5 Fr. Otolaryngology specialists were also consulted for a possible tracheostomy.

At the time of surgery, the fetus was in vertex position. The fetal head was partially delivered, with the mass and the rest of the body still *in utero*. This reduced the chance of umbilical cord compromise and placental separation during the EXIT. The fetus was then successfully intubated using direct laryngoscopy with an endotracheal tube number 3.5 Fr through the Cesarean section wound, as shown in Fig. 4. The baby girl was then delivered, with Apgar scores of 3, 6, and 8 at 1, 5, and 10 minutes, respectively. Her birth weight was 2,760 grams, and the body length and head circumference were 50 and 33 centimeters, respectively. Physical examination revealed a huge neck mass consistent with that observed from the prenatal MRI, as shown in Fig. 5. Postnatal echocardiography showed secondum atrial septal defect, patent ductus arteriosus, and mild pulmonary hypertension. Postnatal CT scan revealed a 9.9 × 5.9 centimeter soft tissue mass at the neck, extending on the anterior chest wall, and attached to the sternum and manubrium. This mass showed heterogenous density with minimal calcification at the periphery.

The mother was doing well postoperatively. The baby was stabilized in the NICU for 8 days before being transferred to the pediatric surgery intensive

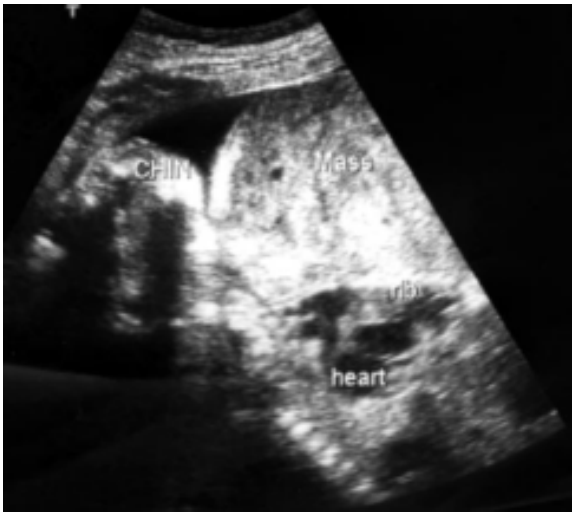

Fig. 1 Sonographic picture showing a huge solid mass on the anterior portion of the fetal neck, extending onto the chest

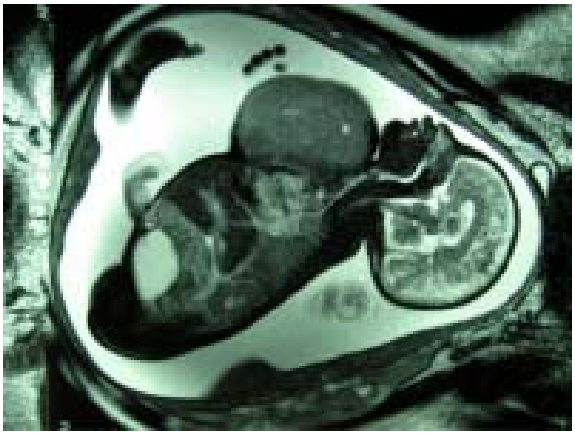

Fig. 2 Magnetic resonance imaging of the fetus in midsagittal plane shows a huge solid mass on the anterior portion of the neck and chest. The fetus was apparently in fixed extending position. Nasopharynx, oropharynx and trachea were intact and filled with amniotic fluid

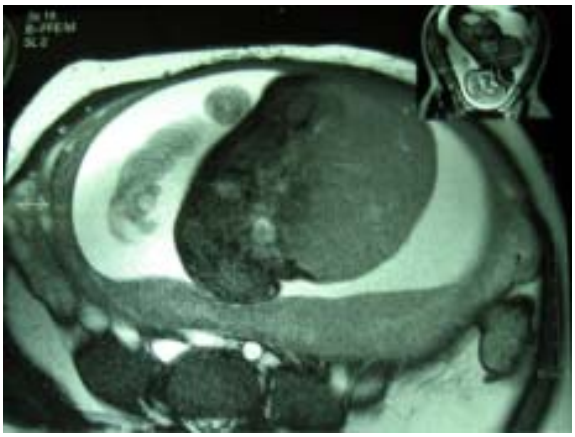

Fig. 3 Axial cut of magnetic resonance imaging of the fetus shows a huge solid mass on the anterior portion of the neck and chest

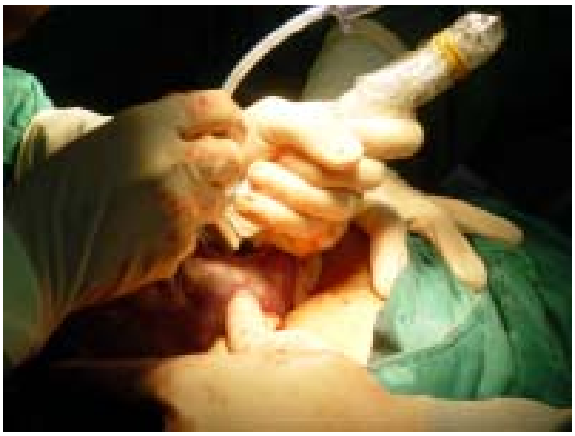

Fig. 4 The fetus was receiving an ex-utero intrapartum treatment (EXIT). Endotracheal intubation was performed through the Cesarean section wound before the baby was delivered and the umbilical cord was clamped and cut

Fig. 5 Picture of the baby with a huge neck mass

care unit. The tumor was surgically removed when the baby was 11 days of age, and histopathological examination revealed solitary infantile myofibroma. No adjuvant therapy was needed. Her latest follow-up at 7 months of age did not show any signs of recurrence. She, however, developed a rectal polyp. It was surgically removed, and histopathological examination showed an inflammatory pseudopolyp with granulation tissue. This had no relationship to her infantile myofibroma.

Discussion

Congenital abnormalities involving the fetal neck are rare. The nature of these masses are varied in terms of both physical appearance (solid or cystic) and the aggressiveness, nevertheless they could cause asphyxia because of airway obstruction at the time of delivery. Prenatal imaging techniques, including ultrasonography and MRI, even though may not always provide definite diagnosis, could be of tremendous help to clearly identify the location of the mass and its effect on the adjacent structures. Therefore, prenatal management, as well as fetal intervention could be properly carried out.

Until now, ultrasonography has been considered a cost-effective venue of real-time fetal imaging. Newer ultrasonographic models have a high resolution capacity and safety index to the mother and the fetus. However, MRI is used as a complimentary modality on the selected circumstances, especially in the rare abnormalities where the sonographic findings are equivocal. This case benefited from the MRI due to its ability to determine tissue layers in the neck region, which were previously obscured by the acoustic shadow of the mass. The authors were able to demonstrate clearly fetal airway from multiplanar MR images. These MRI findings were helpful for the team to make a plan of management, and to perform the EXIT procedure safely.

A few technical issues involving the fetal MRI are worth mentioning. The Balanced Turbo Fields Echo (B-TFE) technique allows for a scan time of less than 1 second. This significantly reduces the motion artifacts, therefore maternal/fetal sedation was no longer required. Lack of fluid signal in fetal esophagous suggested an esophageal compression, leading to

symptomatic polyhydramnios in the presented case. The rapid growth of neck mass suggested its malignant characteristics. Differential diagnosis of malignant tumors on the head and neck region of the fetus should include rhabdomyosarcoma, malignant teratoma, and other sarcomatous or carcinomatous tumors arising in the organs of the anterior neck region, i.e. thyroid gland[1,2]. The mortality associated with upper airway obstruction could be as high as 20%, therefore EXIT was planned[3]. The MRI, in contrast with the sonogram, can show the relationship of the tumor to the airway and gives an impression of the degree of displacement and compression[4].

Despite the rapid growth of tumor in the presented case, the histopathology revealed a benign infantile myofibroma. Giant fetal neck mass diagnosed on the fetal MRI and treated successfully with EXIT has previously been reported[5]. However, the authors reported here the use of MRI to guide the direction of fetal intubation, as well as the size of the endotracheal tube needed to enhance the successfulness of EXIT. In addition, this is a rare case of prenatally diagnosed infantile myofibroma arising on the fetal head and neck region, which showed locally invasive characteristics.

In conclusion, if a neck mass is detected in the fetus by prenatal ultrasonography, then a strategic plan for these types of cases should be developed as early in the gestation as possible[6]. The airway management plans should be tailored to each individual case. MRI should also be just as useful if fetal surgery for a fetal neck mass is considered[7]. Coordination and the expertise of obstetricians, neonatologists, radiologists, otolaryngologist, and anesthesiologists are needed to manage these complex situations.

Acknowledgement

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เนื้องอกที่ลำคอของทารกในครรภ์

ตวงสิทธิ์ วัฒกนารา, โสภาพรรณ เงินฉ่ำ, รัชดา กิจสมมาตร, พรพิมพ์ เฟื่องธารทิพย์

นอกจากการตรวจด้วยคลื่นเสียงความถี่สูงแล้ว การตรวจวินิจฉัยด้วยภาพจากคลื่นสะท้อนพลังแม่เหล็กเริ่มมีบทบาทมากขึ้นเรื่อย ๆ ในการวินิจฉัยและรักษาทารกในครรภ์ ในรายงานฉบับนี้ ผู้นิพนธ์ได้บรรยายถึงทารกในครรภ์รายหนึ่งที่มีก้อนขนาดใหญ่บริเวณลำคออันตรวจพบจากการตรวจด้วยคลื่นเสียงความถี่สูงเมื่ออายุครรภ์ 33 สัปดาห์ การตรวจเพิ่มเติมด้วยภาพจากคลื่นสะท้อนพลังแม่เหล็กยืนยันการวินิจฉัยดังกล่าว รวมทั้งสามารถประเมินทางเดินหายใจของทารกได้อย่างชัดเจน จากขนาดของก้อนและท่าทางของทารกในครรภ์ทำให้มีความเป็นไปได้ที่ทารกจะมีภาวะอุดกั้นทางเดินหายใจเมื่อแรกคลอด หลังจากการวางแผนร่วมกันระหว่างทีมแพทย์ หลายสาขา โดยอาศัยข้อมูลจากภาพคลื่นสะท้อนพลังแม่เหล็กแล้ว จึงตัดสินใจที่จะให้การรักษาทารกในครรภ์โดยการใส่ท่อช่วยหายใจในช่วงของการผ่าตัดคลอดก่อนที่ทารกจะคลอด การรักษาดังกล่าวทำให้คณะแพทย์สามารถรักษาทางเดินหายใจของทารกได้ก่อนที่สายสะดือจะถูกตัด ทารกดังกล่าวได้รับการผ่าตัดรักษาไม่นานหลังคลอด และพบว่าเนื้องอกดังกล่าวเป็นชนิด infantile myofibroma ซึ่งเป็นเนื้องอกที่พบไม่บ่อยบริเวณศีรษะและลำคอของทารกที่ได้รับการวินิจฉัยได้ก่อนคลอด