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Prenatal diagnosis of a giant fetal cervical teratoma by magnetic resonance imaging: a case report

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Abstract

Fetal cervical teratomas are the rare forms of congenital teratomas with a high risk of perinatal morbidity and mortality. Imaging plays an essential role in the management of cervical teratoma and also helps in counselling parents. Ultrasound may be inadequate in the prenatal diagnosis of cervical teratoma due to large tumor size and fetal position. Magnetic resonance imaging could be useful in the work-up of tumours detected by ultrasound. We reported a 29-year old pregnant woman referred to our hospital with a finding of giant solid mass at the fetal neck. Ultrasound examination revealed a right-side mass sized 87x64x51 mm that extended from mandible to the anterior thoracic wall. Fetal magnetic resonance imaging provided additional information regarding exact anatomical location and extent of the mass. Thus, we found that fetal magnetic resonance imaging is a complementary diagnostic modality to antenatal ultrasound examination in the differential diagnosis of cervical teratoma.

Key words: Fetal, Cervical, Teratoma, MRI, Prenatal diagnosis

Introduction

Fetal teratomas are congenital tumors containing more than one embryonic germ cell layer. Although teratomas are the most common fetal tumors, those of cervical origin constitute less than 5% of all teratomas with an incidence of 1/35000-200000 live births (1). The prenatal diagnosis of cervical teratoma is critical as it is associated with polyhydramnios, non-immune hydrops, cardiac failure and preterm birth (2). Ultrasound is the preferred primary imaging technique for the fetal neck tumors, but its ability to detect nature of the mass is limited. However, magnetic resonance imaging (MRI) could be a valuable tool for differential diagnosis of cervical teratoma and accurate identification of location and extension of the tumor (3). Together with ultrasound, MRI may thus be useful in selection of foetuses that require an ex utero intrapartum treatment procedure (EXIT). Here, we report prenatal diagnosis of a giant cervical teratoma diagnosed by sonographic and MRI findings.

Case

A 29 year old gravida 3, para 2 woman was consulted to perinatology clinic with a fetal neck mass which was identified on routine ultrasound examination at 16 weeks of gestation. The woman has two healthy children and her past medical history is unremarkable. On ultrasound examination, there was a right-side mass sized 87x64x51 mm that extended from mandible to the anterior thoracic wall (Figure 1).

It was composed of solid and cystic components with no calcifications. The border between normal tissue and mass was not clearly visualized. Colour Doppler flow revealed that mass was not particularly vascular. There were no accompanying anomalies and amniotic fluid was normal. We considered that the tumor was probably neuroblastoma, goiter or teratoma. For differential diagnosis, half-Fourier acquisition single-shot turbo spin-echo sequence in T2-weighted MRI of the fetal neck was performed. MRI revealed that mass was extending from mandible through supraclavicular level and compressing tracheoesophageal structures without thoracic infiltration (Figure 2).

The epicentre of the lesion was thought to be the cervical canal. Amniocentesis was performed and fetal karyotype was obtained normal 46 XY. Ultrasound and MRI findings suggested that the mass was most likely a cervical teratoma.

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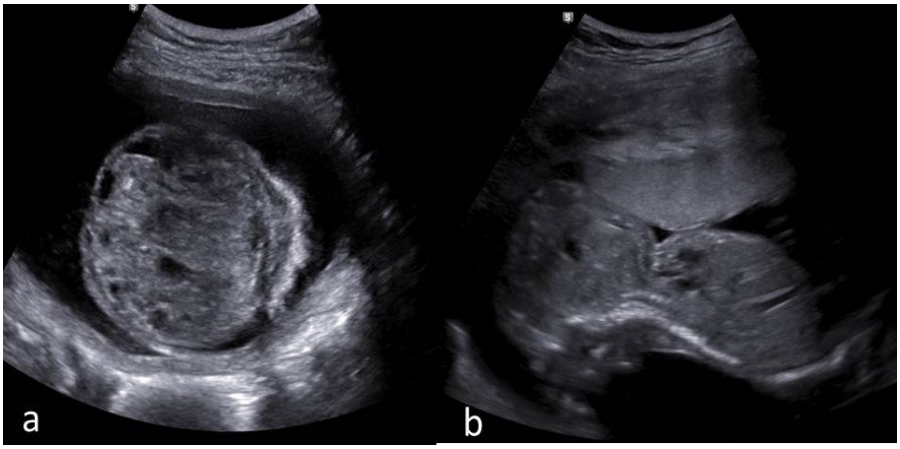

Figure 1. Ultrasound images showing solid-cystic mass extending from anterior neck on axial (a) and sagittal (b) planes.

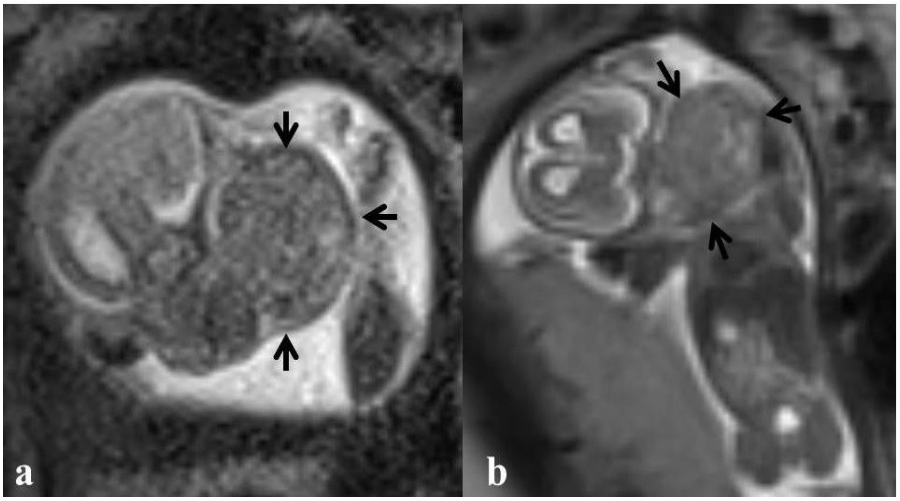

Figure 2. T2-weighted MRI is demonstrating tumor-like mass which compress adjacent structures on axial (a) and sagittal (b) planes.

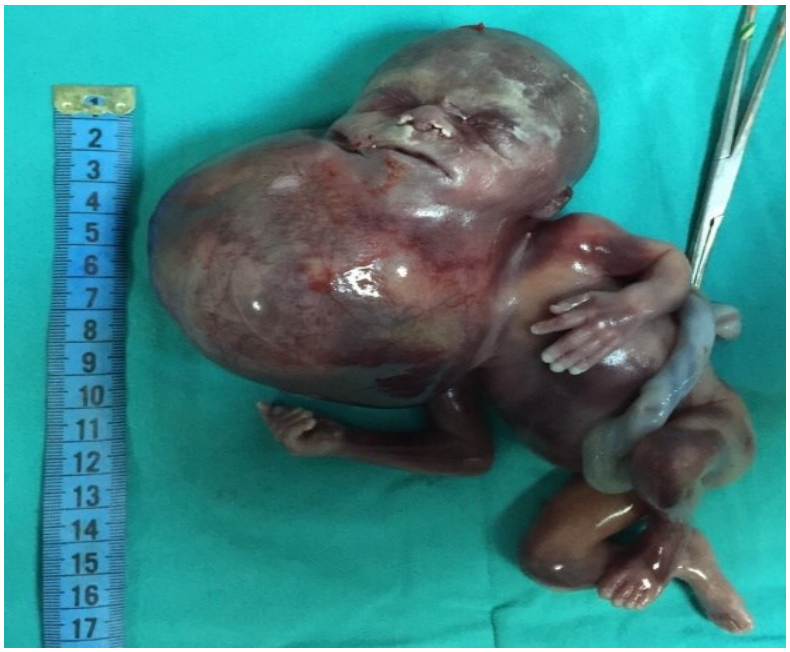

Figure 3. Postnatal findings of infant with cervical teratoma.

The MRI findings, prognosis and treatment options were discussed with consultants from pediatric surgery. Due to large tumor size, poor perinatal outcome and lack of experience with EXIT procedure, termination of pregnancy was offered to the parents. Since the patient had a caesarean section six months ago, pregnancy was terminated at 20 weeks by cesarean section. Postpartum examination revealed 10x7x6 cm well-defined mass originating from anterior portion of the neck (Figure 3). Histologic examination has showed grade 3 teratoma with immature neuro-ectodermal elements. No other structural anomalies were identified.

Discussion

Teratomas are tumors composed of abnormal tissues derived from three germ cell layers (mesoderm, ectoderm, endoderm) and may originate from nasopharyngeal, sacrococcygeal, facial or cervical region (1). Fetal cervical teratoma constitutes only 5% of all teratomas. Malignant transformation of cervical teratoma, which relies on the presence of primitive undifferentiated cells, has been reported in less than 5% of the cases (4). Fetal cervical teratoma is usually considered benign tumor, however, it may result in significant perinatal morbidity and mortality by causing fetal airway obstruction and extensive soft tissue distortion.

Although rare, fetal cervical teratoma may be associated with other structural malformations including imperforate anus, cystic fibrosis, chondro dystrophy fetalis, and maxillar deformity (5-7). It is also reported that teratomas are associated with chromosomal abnormalities such as gene mutations, trisomy 13, ring X-chromosome mosaicism and gonosomal pentasomy 49, whereas cervical teratoma often display normal karyotype (1). In this case, both prenatal and postpartum examinations revealed isolated teratoma and karyotyping turned out to be normal.

Perinatal complications of cervical teratoma include polyhydramnios (from impaired fetal swallowing of amniotic fluid), non-immune hydrops (from arteriovenous shunting) and pulmonary hypoplasia (due to mass effect) (8, 9). Obstruction of circulation and arteriovenous shunting through the tumor may also lead to high-output cardiac failure and, ultimately, fetal demise. Giant cervical teratoma and polyhydramnios increase the risk for preterm labor and may give rise to difficult labor. However, these complications are infrequent findings in the first half of pregnancy, as in our case.

Most of the congenital cervical teratomas can be diagnosed with antenatal ultrasound between 15 and 29 gestational weeks, but the tumor can develop later in pregnancy (4). Typical ultrasound features are well defined, solid or mixed solid-cystic masses extending along the midline with calcifications in about half of the cases. Colour Doppler flow imaging may reveal varying degrees of blood flow within the mass. In this case, the striking feature was presence of poorly vascularized large mass on the anterior neck region without calcifications.

Differential diagnosis of cervical teratoma includes cystic hygroma, haemangioma, goiter, cervical neuroblastoma and nasopharyngeal tumor (10). Cervical teratoma is generally seen at the anterior and midline region of the neck while cystic hygroma, lymphangioma, haemangioma and bronchogenic cyst are usually seen at the posterior and lateral region. However, differential diagnosis of nasopharyngeal tumors, congenital cervical neuroblastomas and goiter are often difficult (11).

MRI could be helpful as a complementary diagnostic tool in the differentiation of these fetal neck masses. MRI enables different image contrasts and provides high spatial resolution and large field of view as compared to conventional ultrasound. Nemec et al. demonstrated the ability to visualize tumors on prenatal MRI in a study of 18 fetuses with tumors (12). In that study, MRI findings changed 50% of suspected ultrasound diagnosis and postpartum histopathology examination confirmed 73% of MRI diagnosis (12). Fetal MRI has shown the potential to provide significant additional information in terms of tumor extent, composition and complications caused by the tumor (3). It is found that fetal MRI was a reliable diagnostic method in the classification of some congenital tumors (7). In our case, MRI clarified the diagnosis of cervical teratoma by identifying the exact anatomical location and extent of the mass.

The recommended therapy for cervical teratoma is surgical excision of tumor mass following establishment of the airway by EXIT procedure. The EXIT procedure involves partial delivery and airway assessment of infant while uteroplacental circulation and gas exchange is maintained. If EXIT procedure is not performed, fetuses with cervical teratoma have high mortality rates from respiratory distress immediately postpartum (6). The EXIT procedure provides time to secure the airway, administer resuscitation medications and resect cervical masses. Hedrick et al. have been used EXIT procedure in the delivery of 10 fetuses with cervical teratomas. They managed to intubate all neonates and long-term survival was reported as 80% (13).

During antenatal surveillance, it is essential to identify fetuses that require EXIT procedure at the time of delivery. Thus, MRI may be of critical importance to assess the relationship between cervical teratoma and structures of the airway and may aid in the selection of fetuses requiring respiratory support. Our MRI studies allowed us to better understand the difficulty in surgical resection of entire mass. Therefore, huge size of the tumor at this stage of gestation period, poor perinatal prognosis and lack of experience with EXIT

procedure were discussed at our institution with the family and termination of pregnancy was offered.

Conclusion

In conclusion, this case report demonstrated that fetal MRI can contribute to a more reliable evaluation in the differential diagnosis of cervical teratomas, particularly in cases where ultrasound is technically unable to establish site of origin and tumor extension.

Conflict of Interest: The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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