A Challenging Delivery by EXIT Procedure of a Fetus With a Giant Cervical Teratoma

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Abstract

Background: Congenital giant neck teratomas are rare tumours associated with high perinatal mortality. Recent advances in prenatal diagnosis and delivery by ex utero intrapartum treatment (EXIT) have improved perinatal outcome.

Case: An otherwise healthy 32-year-old woman, gravida 3, para 2, was referred to our institution at 25 weeks' gestation with a diagnosis of a fetal giant cervical teratoma. Ultrasound and magnetic resonance imaging (MRI) findings suggested airway obstruction in the fetus. An EXIT procedure was attempted but did not result in survival of the baby, despite extensive preoperative planning and the best efforts of a multidisciplinary team.

Conclusion: Despite prenatal detection and diagnosis of airway compromise in a fetus with a giant neck teratoma, securing the fetal airway can be challenging. This is because massive teratomas can completely distort normal tissue and anatomy.

Résumé

Contexte : Les tératomes cervicaux géants congénitaux sont des tumeurs rares qui sont associées à un taux élevé de mortalité périnatale. Des percées récentes dans les domaines du diagnostic prénatal et de l'accouchement par traitement intrapartum *ex utero* (EXIT) ont entraîné une amélioration de l'issue périnatale.

Cas : Une femme de 32 ans, gravida 3, para 2, autrement en santé a été orientée vers notre établissement à la 25e semaine de gestation en raison d'un diagnostic de tératome fœtal cervical géant. Les résultats de l'échographie et de l'imagerie par résonance magnétique (IRM) laissaient entendre la présence d'une obstruction des voies respiratoires chez le fœtus. Une intervention EXIT a été tentée mais ne s'est pas soldée par la survie de l'enfant, et ce, malgré une planification préopératoire exhaustive et les meilleurs efforts d'une équipe multidisciplinaire.

Conclusion : Malgré la détection et le diagnostic prénatal de problèmes affectant les voies respiratoires d'un fœtus présentant un tératome cervical géant, la libération des voies respiratoires fœtales peut s'avérer difficile. Cela s'explique du fait que les tératomes considérables peuvent entièrement déformer l'anatomie et les tissus normaux.

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INTRODUCTION

Congenital neck teratomas account for 5% of all congenital teratomas.[1] Teratomas are rare benign tumours consisting of tissues originating from all three embryonic layers; however, when they are located in the neck, perinatal mortality is high as a result of upper airway obstruction.[2] Recent advances in prenatal diagnosis, anticipation of neonatal upper airway obstruction, and delivery by EXIT have resulted in improved perinatal outcome.[3,4]

During an EXIT procedure, the fetus is partially delivered from the uterus while controlled uterine hypotonia is used to maintain the utero-placental circulation. This allows maintenance of fetal oxygenation while diagnostic and/or therapeutic procedures are performed on the fetal airway. Deep maternal anaesthesia is required to maintain uterine relaxation; however, there is a theoretical risk of postpartum hemorrhage from uterine atony. Other inherent risks include a potential need for future Caesarean section and, despite precautions, the potential inability to secure the airway with consequent demise of the newborn.

Prenatal diagnosis aims to select patients who will most benefit and not be exposed to additional risks from the procedure. A coordinated multidisciplinary team approach involving perinatologists, anaesthesiologists, neonatal intensivists, otolaryngologists, and nurses is required for planning and execution.[5]

We present a case of a giant congenital cervical teratoma in which the EXIT procedure was attempted but was unsuccessful, despite extensive preoperative planning and the best efforts of the team.

THE CASE

An otherwise healthy 32-year-old woman, gravida 3, para 2, was referred to our institution at 25 weeks' gestation for assessment and delivery by EXIT procedure. Her previous two pregnancies had been uncomplicated and resulted in spontaneous vaginal deliveries at 36 weeks. With the index pregnancy, nuchal translucency at 12 weeks was within normal limits, and integrated prenatal screening was negative. A routine anatomical scan at 18 weeks, however, revealed an avascular, largely cystic neck mass measuring 7 cm × 5 cm × 6 cm, with solid components. There was hyperextension of the fetal neck and a small stomach suggestive of esophageal obstruction, but the amniotic fluid index was normal.

Follow-up 2D and 3D ultrasound examinations (Figures 1 and 2) at 24 weeks showed an increase both in the size of the neck mass and in the amniotic fluid index. Fetal MRI concurred with ultrasound findings and also showed amniotic fluid in the mouth and upper gastrointestinal tract, but not below the level of the mass, suggesting obstruction to the trachea and esophagus. A provisional diagnosis of a large cervical teratoma was made by the referring institution.

In anticipation of the potential difficulty in establishing an airway at birth, the patient was referred to our centre for delivery via EXIT procedure. Consultations with the anaesthesiology, neonatalology, and otolaryngology services helped establish a multidisciplinary plan for delivery at 38 weeks. Serial weekly ultrasound examinations assessed fetal and tumour development and the progression of polyhydramnios. At 28 weeks' gestation the mother's abdomen became very tense, causing significant discomfort and concern about possible preterm labour.

The patient accepted the offer of therapeutic amnioreduction after receiving detailed counselling about the risks of prolonged preterm rupture of membranes and preterm labour possibly arising from amniocentesis. At the time of amniocentesis, approximately 250 mL of serosanguineous fluid was concurrently aspirated from the cervical mass. Cytological analysis of this fluid revealed no malignant cells. As there was progressive reaccumulation of amniotic fluid and return of abdominal discomfort, amnioreduction was repeated at 33 weeks. At

Figure 1. Two-dimensional ultrasound showing giant cervical teratoma with mixed echogenicity and polyhydramnios

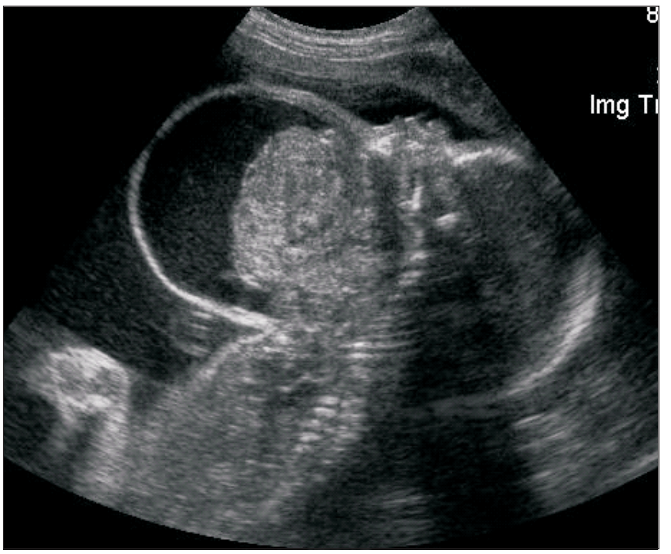
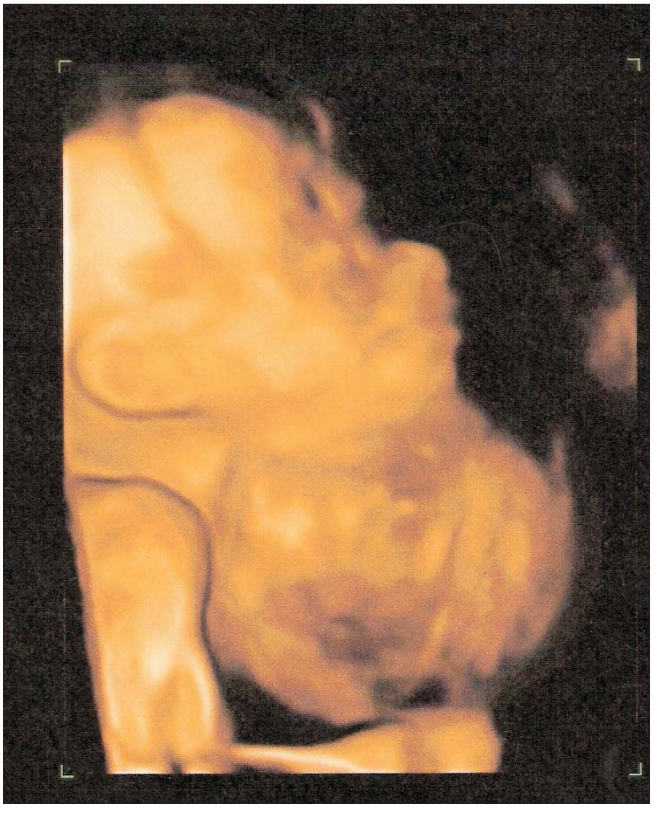


Figure 2. Three-dimensional surface rendered ultrasound shows giant cervical teratoma



ABBREVIATIONS

2D	two dimensional
3D	three dimensional
EXIT	ex utero intrapartum treatment
MRI	magnetic resonance imaging