

Management of Giant Cervical Teratoma with Intracranial Extension Diagnosed in Utero

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Abstract

Cervical teratomas are rare germ cell tumors affecting the fetus that are associated with significant morbidity and mortality due to an increased risk of airway obstruction at delivery. These tumors can commonly produce polyhydramnios that results from the fetus' impaired ability to swallow amniotic fluid. Improved rates of prenatal diagnosis through comprehensive evaluations and imaging have dramatically impacted the perinatal management of infants with this condition. Here, we report a patient diagnosed with polyhydramnios whose fetus was discovered to have a giant cervical teratoma on imaging studies. The child underwent surgical resection after having the airway secured under the uteroplacental support as part of an ex utero intrapartum treatment procedure performed at 37 weeks. The following gross pathological and magnetic resonance images demonstrate this condition and its currently accepted treatment.

Keywords

- ▶ cervical teratoma
- ▶ EXIT procedure
- ▶ perinatal management
- ▶ polyhydramnios

Introduction

Cervical teratomas have an estimated incidence of 1 per 20,000 to 40,000 live births and represent the second most frequent perinatal germ cell tumor.^{1–3} Most of these tumors are benign, and therefore, complete resection at birth can be curative. However, the location of these tumors importantly can lead to airway distortion and obstruction.³ Furthermore, by compressing the hypopharynx and esophagus, giant teratomas can also prevent the swallowing of amniotic fluid resulting in polyhydramnios. Preterm labor, premature rupture of membranes, and placental abruption secondary to this polyhydramnios can complicate the surgical management of these tumors.⁴ As the incidence of polyhydramnios in previous large series has been reported as high as 82%, this finding is critical not only for

diagnosis but proper preoperative planning.¹ This planning can be particularly useful in determining whether to proceed with a controlled delivery technique such as an ex utero intrapartum treatment (EXIT) procedure. We present a case whereby polyhydramnios on ultrasound prompted the discovery of an impressive lesion on maternal magnetic resonance (MR) imaging; we then describe the unique surgical treatment method that followed.

Case Report

A 32-year-old primigravid woman was diagnosed at 21 weeks gestation with polyhydramnios during routine screening. Her fetus was found to have a giant fetal neck mass by antenatal ultrasound. MR imaging was subsequently performed in

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utero revealing a cystic and heterogeneous-appearing mass with extension into the middle and posterior cranial fossae consistent with a giant cervical teratoma. At 32 weeks gestation, imaging was repeated (►Fig. 1A). Given the tumor's encroachment on the airway and esophagus, an EXIT procedure was planned at 37 weeks gestation to first secure the airway and then resect the mass following stabilization.^{5,6}

At the time of delivery, a cesarean delivery with the partial delivery of the fetus was undertaken. The child's head and upper body were exteriorized via hysterotomy guided by ultrasound. While attached to the placenta via the umbilical cord, anesthesia was administered. A combination of desflurane, propofol, and remifentanyl were provided.⁷ An endotracheal tube was placed under direct laryngoscopy to secure the airway. Once secured, the umbilical cord was cut, and resuscitation was performed before surgical resection. The ensuing resection revealed that the tumor was extradural. Following a

craniotomy, one portion of the tumor was resected from the middle fossa and was then followed through Kawase triangle into the posterior fossa.⁸ Enlargement and entry of the jugular foramen allowed for ligation of the jugular vein.

Grossly, the tumor was found to be protruding from the right ear canal and contained several pieces of abnormal bone (►Fig. 1B, C). These sections and the large cervical portion were removed en bloc. Pathology findings indicate a cystic teratoma containing scant immature tissue and more prominent mature tissue, consisting of skin, fibroadipose connective tissue, bone, cartilage, skeletal and smooth muscle, respiratory and gastrointestinal-type epithelium, pancreas-like glandular tissues, and lymphoid tissue. Postoperative MR imaging demonstrates near-total resection from the right middle cranial fossa, right posterior fossa, and right neck with little residual tumor (►Fig. 1D). Despite having a right-sided hearing deficit and swallowing dysfunction necessitating a gastrostomy tube,

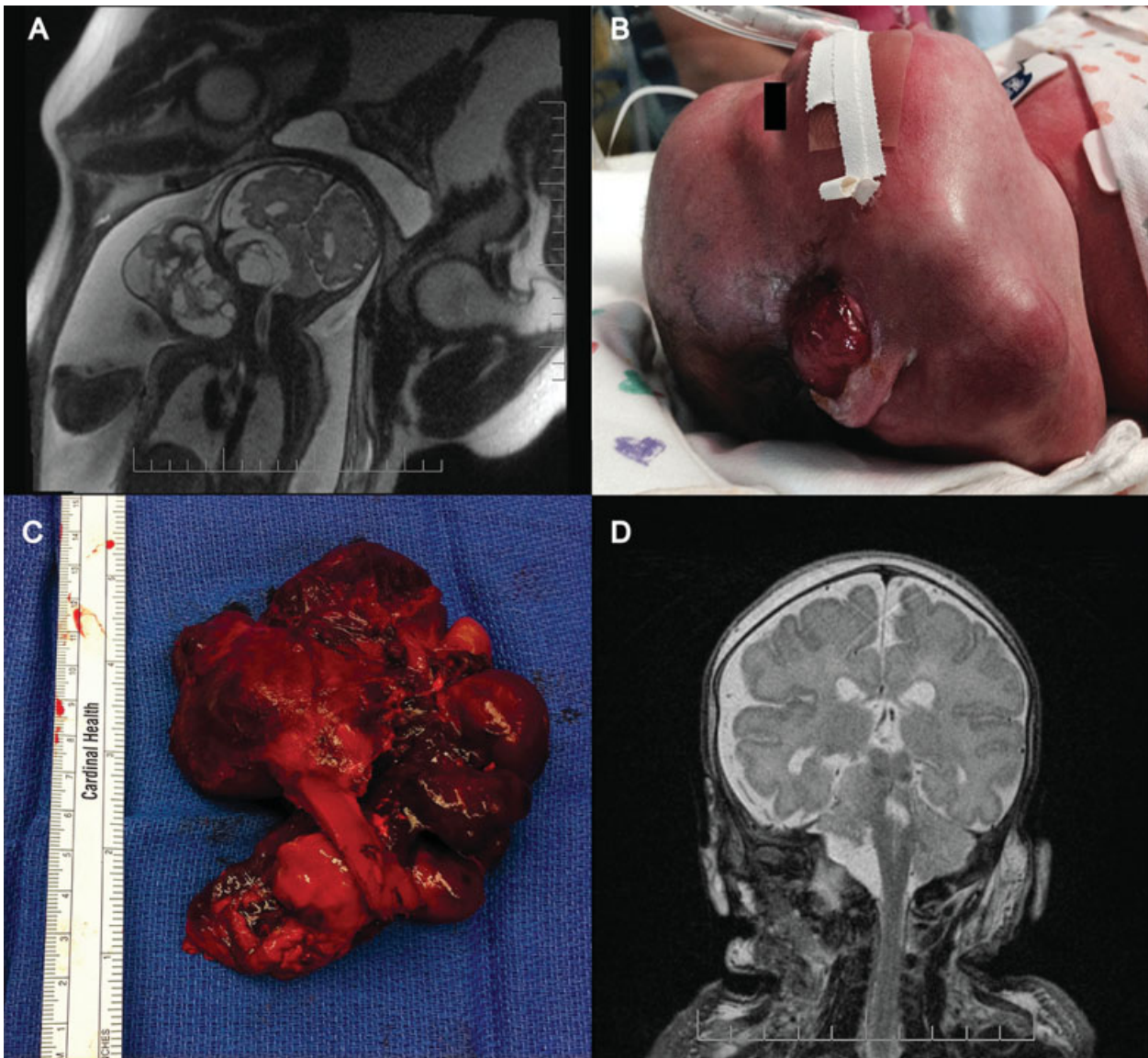


Fig. 1 (A) T2-weighted maternal MRI, 32 weeks gestation. A large fetal neck mass is seen with mass effect on the adjacent cerebellar hemisphere and brainstem. (B) Intraoperative view showing the mass protruding from the external ear. (C) Gross specimen with tissues including bone and hair. (D) Postoperative MRI at 3 months age. MRI, magnetic resonance imaging.

at the time of this writing (6 months postoperatively), the child is breathing without a tracheostomy and meeting motor and cognitive milestones.

Discussion

Kelly et al first introduced the concept of operative management of a fetal cervical mass under uteroplacental support.⁹ The EXIT procedure was described and refined by Dr. Adzick, Dr. Harrison, and others in the 1990s.^{5,10,11} This fetal surgical procedure allows for safe delivery and the avoidance of airway obstruction, which carries with it a high risk of mortality in neonates.¹² During the procedure, the head, neck, and shoulders of the fetus are delivered through a hysterotomy, thereby maintaining uterine volume and minimizing uterine contractions. Before the hysterotomy, intraoperative ultrasound is utilized to accurately map the placental edges to avoid placental abruption. Uteroplacental blood flow and gas exchange, as well as uterine relaxation, are also maintained through the use of inhalational agents, β -adrenergic agonists, and nitroglycerin.¹ Movement of the fetus and breathing during the procedure is restricted to the use of vecuronium, atropine, and fentanyl, as previously described.⁵ In addition to the time needed for surgical resection, this provides adequate time for techniques necessary to secure the airway, including surfactant administration, direct laryngoscopy, bronchoscopy, and tracheostomy procedures. The EXIT procedure provides up to 1 hour of uteroplacental support in fetal patients with giant cervical teratomas.⁵ Possible complications relating to the EXIT procedure that were fortunately not encountered in this case include uterine atony, uterine rupture, and placental injury during hysterotomy.⁵

Previous reports of teratomas of the head and neck region have emphasized the importance of a skilled multidisciplinary team of physicians and nurses specializing in fetal, neonatal, and maternal care.^{1,5,13} Indeed, the rarity of these tumors and the expertise required to minimize complications necessitate careful planning and coordination to ensure the success of the EXIT procedure. Given the cervical and intracranial location of tumors such as that described, pediatric neurosurgeons remain integral to minimizing the morbidity associated with surgical treatment and in following patients through infancy and childhood.

Conclusion

Although rare, cervical teratomas can be diagnosed in the antenatal stage and treated safely using the EXIT procedure. Ultrasonography, by facilitating early diagnosis, MR imaging, by accurately illustrating anatomical relationships, and EXIT, by providing a mode of treatment, have all contributed complementary and beneficial roles in reducing mortality of cervical masses. This procedure allows for the partial delivery of the fetus to prevent airway compromise, a complication closely associated with infant mortality. Surgical resection of cervical teratomas in this manner can thus be used to minimize morbidity and mortality.

Competing Interests

None.

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