CASE REPORT

Ex-utero Intrapartum Treatment (EXIT) Procedure for Giant Fetal Epignathus

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Received: February 05, 2015;
Initial review: April 14, 2015:
Accepted: July 15, 2015.

Background: Large fetal oropharyngeal tumors are rare, and have the potential to cause airway obstruction during birth. **Case characteristics**: A 35-year-old woman with antenatally diagnosed large heterogenous mass in fetal neck displacing trachea and filling up the orophanygeal space. **Intervention**: The infant was delivered at 31 weeks of gestation by ex-utero intrapartum therapy procedure to secure the airway. **Outcome:** Tumor was resected successfully on day 8 of life. Histopathology revealed mixed teratoma. **Message**: *Ex-utero* intrapartum therapy for fetuses with severe upper airway compromise may prove life-saving.

Keywords: *Antenatal diagnosis, Fetus, Orolpharynx, Teratoma.*

Epignathus, a teratoma arising from the oral cavity or pharynx, is rare in neonates. The estimated incidence is 1:35,000 to 1:200,000 live births with a female preponderance [1,2]. Arising from pluripotent cells, they contain all three germ cell lines, and range from benign, well-differentiated cystic lesions to those that are solid and malignant [3]. Large oropharyngeal epignathi have the potential to cause life-threatening airway obstruction and even death during or shortly after birth [1]. Antenatal diagnosis gives the managing team an opportunity to perform an *ex-utero* intrapartum treatment (EXIT) procedure to secure the airway. We report a case of giant fetal epignathus delivered successfully by EXIT procedure at 31 weeks of gestation.

CASE REPORT

A 35-year-old woman was referred to us at 26 weeks of gestation with threatened labor. Fetal scans were unremarkable at the referral center. Ultrasound scan revealed a homogeneous mass 4.8 x 4.4 x 3.7 cm in the fetal neck along with polyhdramnios (amniotic fluid index of 24 cm) necessitating an amnioreduction. Fetal magnetic resonance imaging revealed a large heterogeneous mass in the anterior part of the fetal neck posterior to the trachea, displacing it to the left, filling up the oropharyngeal space, and infiltrating the floor of the mouth (***Fig.* 1a**). This was suggestive of a teratoma with significant airway compromise. There was no intrathoracic or intracranial extension. The case was discussed at the multidisciplinary birth defect meeting and the need for an EXIT procedure was felt. A team having obstetrician, neonatologist, otolaryngologist, pediatric surgeon and anesthetists was formed. They planned to secure the airway by EXIT procedure and resect the mass after stabilization following imaging to delineate the anatomy at 34 weeks of gestation. Serial scans showed a gradual increase in mass size. A second amnioreduction was needed at 29 weeks.

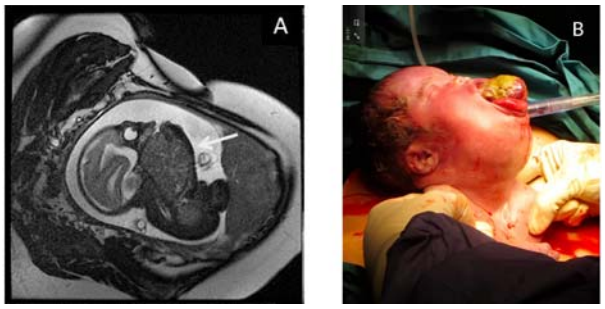

FIG. 1 *(a) T2-weighted sagittal MRI image showing a large mass in the anterior part of the fetal neck, displacing the trachea (arrow); and (b) the neonate after delivery of the head and shoulders showing the presence of the large oral teratoma necessitating a tracheostomy.*

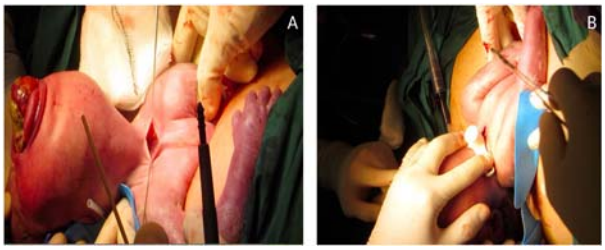

FIG. 2 *The neonate undergoing tracheostomy during the EXIT procedure.*

An emergent delivery was needed at 31 weeks due to rupture of membranes. The EXIT team was mobilized. A caesarean section was done under deep general anesthesia. The fetal head and shoulders were delivered. A giant solid mass protruding through the oral cavity was noted (***Fig.* 1b**), making any attempt at intubation impossible. With the placental circulation still intact, the otolaryngologist performed a tracheostomy (***Fig.* 2**). Time to tracheostomy from delivery of the head was 24 minutes. After suctioning the trachea, surfactant was administered and the umbilical cord was clamped. The infant weighed 2000 g. A computed tomography of the neck showed a large heterogeneously enhancing mass distending the oral cavity and oronasopharynx and abutting against the under surface of the bony hard palate. Serum alpha-fetoprotein (AFP) level was markedly elevated. At surgery on day 8 of life, the tumor was noted to be attached to the anterior surface of the posterior put of tongue by a 3 cm stalk with no other oropharyngeal attachments. There was pressure necrosis of the tip of the anterior part of tongue. The tumor was vascular and composed of hair and cartilage. It was excised and a hemiglossectomy was performed. A cleft palate was noted.

The resected tumor was lobulated pale-tan to hemorrhagic and weighed 100 g. Histologically, the tumor was composed of derivatives from all three germ cell layers including prominent immature neuroectoderm, focal melanin pigmentation, choroidal epithelium, cartilage, smooth and skeletal muscles, mucinous epithelium with focal ciliation, squamous islands and liver tissue. The histological grade was 3 out of 3. No malignant germ cells were identified. Serial AFP levels showed a declining trend postoperatively.

The patient was discharged at 2 months corrected age with tracheostomy mask and tube feeds. At 5 months, there was a recurrence at the base of the tongue, which was excised. Histopathology report showed a mixed teratoma. Screening for metastasis was negative. Gastrostomy was performed at 4 months of age to facilitate feeding. Repair of the cleft palate was also done. Persistent sialorrhea was controlled with scopolamine patches and botulinum toxin injections. At 15 months, he has mild gross motor delay with normal fine motor and social skills. He has speech delay which is anatomical because of glossectomy. Vision and hearing tests are normal. Discussions are underway to assess the feasibility of tongue reconstruction.

DISCUSSION

EXIT procedure was originally designed to remove tracheal occlusion devices in fetuses with congenital diaphragmatic hernia [4]. It can however be used to secure the fetal airway prior to complete delivery whenever airway compromise is anticipated at birth [5]. During EXIT, a cesarean section is done under general anesthesia. This procedure differs from the usual caesarean section in that the mother receives deep anesthesia and enough time is given for the anesthetics to pass via the placental circulation to the fetus. The goals are to maintain uterine relaxation and volume to maintain the uteroplacental circulation, delay fetal-neonatal transition, achieve a surgical level of fetal anesthesia without cardiac depression and maintain normal maternal blood pressure during the period of deep anesthesia [4]. Only the fetal head and shoulders are delivered and the airway is secured by intubation or tracheostomy while the placental circulation is intact. Once the airway is secured, the fetus is delivered and the cord is cut.

In a large case series reported by Hedrick, securing the airway was the most common indication for EXIT in fetuses with large neck masses or congenital high airway obstruction syndrome [6]. Other indications of the procedure are [4,7,8] for insertion of ECHO cannular when severe hypoxia is anticipated in the delivery room due to pulmonary or cardiac abnormalities; and for rapid control of the airway when twin separation in conjoined twins is indicated for survival.

Our patient had a large oropharyngeal mass with suspected airway compromise. She presented at 26 weeks which is beyond the legal limit for medical termination of pregnancy. The team discussed offering comfort care *versus* delivery by EXIT to the parents, highlighting that this procedure had never been done in our country, and the parents opted for an EXIT procedure.

Early radical excision is the treatment for large head and neck teratomas without intracranial extension [9]. These are mostly benign, and recurrence is rare [10]. Higher mortality in giant oral teratomas is invariably due to the inability to secure airway leading to death at or shortly after birth. The concept and expertise of EXIT procedure is limited to only a few tertiary centers in the world, and needs to be expanded.

Acknowledgements: Adjunct A/Prof Teo Eu-leong Harvey James, Department of Diagnostic Imaging, and Dr Anicete Rosslyn Capuno, Department of Otolaryngology for their help in providing the images for this report. Ms. Siti Nurfadilah, Department of Neonatology for her assistance in formatting the text and images.

Contributors: PAJ: Review of literature and manuscript preparation, involved with antenatal planning, delivery and managing the case; SC: Manuscript preparation and literature review; BS: Manuscript preparation and involved with antenatal planning, delivery and managing the case; KTEC:

Provided the histopathology slides with descriptions and interpretation.

Funding: None; *Competing interests*: None stated.

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