Management of a Prenatally Diagnosed Huge Teratoma Arising from the Soft Palate

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Teratomas arising from the oral cavity are relatively rare and can cause life-threatening airway obstruction immediately after birth. We report a case of a huge teratoma arising from the soft palate detected prenatally. To save the patient, a caesarean section and ex utero intrapartum treatment (EXIT) were planned at 29 weeks of gestation. However, during EXIT, the patient's cardiopulmonary status deteriorated due to unstable cord blood flow secondary to uterine contractions. EXIT was abandoned, the patient was delivered and an emergency tracheotomy performed. The tumour was successfully excised 4 hours after tracheotomy. The tumour weighed 1,591 g and was 20 × 22 × 12 cm. The patient, a female, weighed 715 g. Histopathology showed Grade II teratoma. The postoperative course was uneventful and she is now 5 years old with no neurological sequelae. EXIT is indicated in patients who have a high risk for airway obstruction immediately after birth. However, if EXIT must be abandoned, as in this case, urgent tracheotomy is mandatory. Planned prevention of airway obstruction at delivery is indispensable for successful outcome and requires excellent coordination among obstetricians, neonatologists, anaesthesiologists and paediatric surgeons. [Asian J Surg 2006;29(3):212–5]

Key Words: prenatal diagnosis, soft palate, teratoma

Introduction

A teratoma is a type of germ-cell tumour derived from pluripotent cells and made up of elements of different types of tissues from one or more of the three cell layers.1 Fetal teratomas arising from the oral cavity are relatively rare,2 but they can cause life-threatening airway obstruction immediately after birth.

There have been outstanding advances in prenatal diagnostic technology and subsequently, major changes in the surgical skills required for treatment during the fetal and neonatal periods, which strongly influence perinatal management.3-7 For instance, ex utero intrapartum treatment (EXIT) was designed to provide time to secure the airway while uteroplacental gas exchange is preserved.3

Herein, we report a case of a huge teratoma arising from the soft palate, which was diagnosed prenatally; EXIT was attempted for excision of the teratoma.

Case report

A huge tumour around the face of the fetus was detected by ultrasonography (US) at 22 weeks of gestation (Figure 1). Fetal magnetic resonance imaging (MRI) also showed a huge mixed intensity tumour, which was strongly suggestive of a teratoma arising from the oral cavity (Figure 2). Polyhydramnios was noted. At this time, the perinatal team decided that the fetus should be delivered by caesarean section and managed prior to ligation of the umbilical cord using EXIT because there was an extremely high...
risk of airway obstruction immediately after birth. Various other scenarios were also prepared for in case EXIT failed; for instance, emergency tracheostomy. Umbilical cord blood flow was found to be decreasing on US at 29 weeks' gestation, so a caesarean section was performed under general anaesthesia and the tumour was to be excised using EXIT. Vecuronium bromide, fentanyl citrate and atropine were administered to control uterine contractions, and intraoperative US was performed to identify the best position for uterotomy. A vertical incision in the body of the uterus was made, based on the location of the fetal head and tumour. Great care was taken during incision, but the tumour proved to be so large that it could not be delivered through the initial incision. The caesarean incision was extended, but only the tumour and mouth of the patient could be successfully delivered. The uterus began to contract when the tumour was delivered, probably because of the sudden change in size of the uterine cavity as the tumour was much larger than the patient, and umbilical blood flow decreased. EXIT was abandoned, the cord was clamped and divided, and the patient delivered. Preoperatively, the perinatal team had tried to predict various outcomes and preparations had been made for most scenarios. Intubation was attempted, but it was next to impossible because the pharynx and larynx could not be visualized at all. It took only 8 minutes from incision of the uterus to tracheal intubation. On gross inspection, the tumour was large and contained a mixture of solid and cystic parts, with obvious hair and appendages covered partially by skin, arising from the soft palate (Figure 3). The patient appeared to have no other deformities. At 3 hours after delivery in the NICU, the infant's cardiovascular status began to compromise, probably because blood was being trapped within the tumour. At this point, the patient's family was consulted about further intervention because the mandibular joint on the side of the tumour seemed to be dislocated and an intracranial origin for the tumour could not be excluded. The family was eager to save their baby, so a tourniquet was placed around the base of the tumour, which was found to arise from the soft palate, and the base was ligated using double Maxon sutures and the tumour excised. Bleeding points in the stump

Figure 1. Prenatal ultrasound shows a huge tumour around the face.

Figure 2. Fetal magnetic resonance imaging shows a huge mixed intensity tumour arising from the oral cavity.

Figure 3. Gross inspection of the tumour and baby after tracheostomy.
were controlled. The patient tolerated the procedure well and was stable following excision. The tumour measured $20 \times 22 \times 12$ cm in size and weighed 1,591 g. Residual tumour resection was performed later by neuro- and plastic surgeons. Histopathology showed Grade II teratoma consisting of fat, muscle, bone, intestine and immature neural tissue.

After excision of the tumour, the patient weighed 715 g. On the 90th day of life, she was extubated. Her mandibular joint that was initially dislocated due to the tumour was found to be intact, and she started to drink milk when she was 4 months old. She is now 5 years old (Figure 4) with no neurological sequelae and no evidence of recurrence.

**Discussion**

The successful outcome of our case clearly illustrates the value of accurate prenatal diagnosis and carefully planned perinatal management. Previously, Jordan and Gauderer studied over 200 cases of cervical teratoma and reported that the death rate for cases with respiratory distress at delivery was 43%. However, perinatal management of tumours of the neck has changed greatly since then due to advances in prenatal diagnosis.

Successful management of prenatally diagnosed massive teratomas located in the head and neck or intraorally using EXIT was reported in the 1990s. The EXIT procedure maintains uteroplacental blood flow and fetal gas exchange by keeping the uterus relaxed through the use of inhalational agents and the uterine volume stable by only partially exposing the fetus. It has been reported that relatively normal cord blood gas values can be maintained for up to 50 minutes on uteroplacental support. The EXIT procedure allows time to perform multiple procedures such as laryngoscopy, bronchoscopy and tracheotomy for securing the airway and providing ventilation. However, premature labour is detrimental during EXIT because of increased uterine contractility, and prolonging EXIT can increase the risk of uterine haemorrhage.

In our case, we attempted to use EXIT, but failed. To the best of our knowledge, reports of tumours managed successfully using EXIT have all been much smaller than ours. In this case, EXIT was abandoned because uterine contractions and cord blood saturation could not be controlled after the tumour was delivered through the caesarean incision because of a sudden decrease in the volume of the intrauterine cavity, which had been occupied almost entirely by the tumour. Extracorporeal membrane oxygenation (ECMO) is probably an alternative method of maintaining neonatal oxygenation, although ECMO is associated with an increased risk of bleeding. In the end, the airway was secured within 8 minutes from incision of the uterus by performing an emergency tracheostomy in a completely prepared operating theatre. Fortunately, the patient has demonstrated no neurological sequelae thus far.

In conclusion, EXIT is indicated in neonates who have a high risk of airway obstruction immediately after birth. However, if EXIT must be abandoned, as in our case, urgent tracheotomy is mandatory. Careful prenatally planned management is mandatory to save those neonates with airway obstruction. We have no doubt that the excellent coordination among the obstetricians, neonatologists, anaesthesiologists and paediatric surgeons involved with this patient was indispensable for the successful management of this patient.

**References**


