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Cervical rhabdomyosarcoma and EXIT procedure; case report☆

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ABSTRACT

A 37 year-old primipara women was referred for management of a large solid tumor found in the neck of her fetus, showing the trachea to be compressed by the mass, and intubation through the ex utero intrapartum treatment (EXIT) was planned. After the mother was anesthetized using a Fentanyl with Isofluorane, the head and left arm of the baby were delivered through a standard lower segment cesarean section, and the baby was intubated with a 3.0 mm tracheal tube before clamping and dividing the umbilical cord, then transferred to a resuscitation table. Time from partial delivery to transfer was 9 min, and the baby weighed 3300 g and quickly achieved acceptable oxygen saturations. A tracheostomy and biopsy of the mass were performed on day 1, and Rhabdomyosarcoma was diagnosed. Multiagent chemotherapy was commenced without any reduction in size (82 × 80 × 70 mm), and radical resection was performed 8 weeks after EXIT. Preoperatively, arterial feeders to the mass arising mainly from the right external carotid artery, were selectively embolized, and surgical excision was performed. After additional chemotherapy and radiotherapy, the child is doing well without recurrence after 3 years of follow-up.

Solid fetal neck masses are rare conditions that may cause life-threatening airway obstruction. Prenatally, a fetus with complete airway obstruction can survive because of placental gas exchange but will die once delivered. Diagnosing the condition before birth may allow palliation at delivery and may allow an EXIT (ex utero intrapartum treatment) procedure to be performed such as delivering just the head of the baby to maintain placental gas exchange until adequate ventilation can be secured, whereupon the rest of the baby is delivered. Rhabdomyosarcoma (RMS) is the most common soft tissue malignancy of childhood, but may occur extremely rarely in the neonatal period or in the fetus [1,2]. Application of EXIT to cervical RMS has not been reported, and this case report could contribute to understanding of the applications of EXIT and highlight how to manage fetal cervical RMS.

1. Case report

A 37 year-old primipara Japanese women was referred to Shizuoka Children's Hospital at 32 weeks gestation for management of a solid mass found in the neck of her fetus. On routine ultrasonography, a solid tumor, 70 mm in diameter was observed in the anterior neck of her fetus, and magnetic resonance imaging (MRI) showed the trachea to be compressed by the mass (Fig 1). Polyhydramnios was also present. The mass grew 5 mm per week and serial amnioreductions were performed. Because of predicted respiratory distress during delivery, EXIT was proposed to the parents and intubation was planned just after the head was delivered in order not to compromise placental circulation [3]. Informed consent was obtained from both parents.

At 36 weeks' gestation, the mother was anesthetized using Fentanyl with Isofluorane to aid uterine relaxation. Initially, the head and left arm of the baby were delivered through a standard lower segment cesarean section taking great care to ensure there was no stretching or kinking of the umbilical cord. An Atropine sulfate (0.1 mg) and Musculax (0.6 mg) were given to the baby

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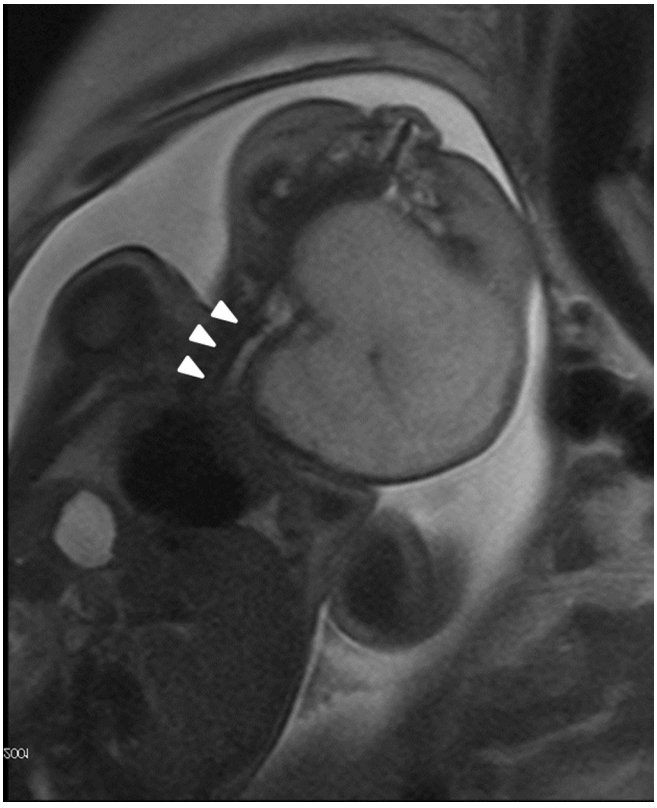

Fig. 1. Prenatal MRI showing the trachea being compressed by the mass (arrowheads) in the anterior neck of the fetus.

through an intramuscular injection. One surgical assistant was assigned specifically to monitor hemodynamic stability by palpating the umbilical artery. The baby was intubated with a 3.0 mm tracheal tube before clamping and dividing the umbilical cord (Fig 2), then transferred to a resuscitation table. Time from partial delivery to transfer was 9 min. The baby weighed 3300 g and quickly achieved acceptable oxygen saturations in the low 90 s using low pressure ventilation. A tracheostomy and biopsy of the mass were performed the day after delivery, and RMS was diagnosed (Fig 3). MRI revealed that the mass measured 82 × 80 × 70 mm and was located at the bottom of the oral cavity, displacing the tongue upward and the pharynx to the left (Fig 4). Multiagent chemotherapy, including vincristine, actinomycin, cyclophosphamide, cisplatin etc., was commenced without any reduction in size, and radical resection was performed 8 weeks after EXIT. Preoperatively, arterial feeders to the mass arising mainly from the right external carotid artery were selectively embolized using gelatin sponges by a neurosurgical team. Surgical excision of the mass was performed through a 5 × 3 cm T-shaped incision made over the apex of the mass. Dissection was easy except for a fibrous band connected to the right submandibular gland, which was resected with the mass, and some cervical muscle was also resected with the submandibular nerve; the thyroid gland and hyoid bone were preserved intact. Total intraoperative blood loss was 60 mL necessitating intraoperative blood transfusion (Fig 5). After additional chemotherapy and radiotherapy, the child is doing well without recurrence after 3 years of follow-up.

2. Discussion

The most common cause of fetal solid neck mass is teratoma, and the incidence of fetal teratoma ranges from 1 in 20,000–40,000 live births [4]. Cervical teratoma causing airway obstruction has been managed successfully with EXIT [3,5]. In contrast to teratoma, congenital RMS is extremely rare, although RMS is the most common soft-tissue tumor in children. Embryonal RMS, the most common type of RMS, generally responds very well to chemotherapy [6], and good response to chemotherapy allows surgery to be less aggressive if it is required [7]. According to a past report from the Intergroup Rhabdomyosarcoma Study, of 3217 patients with RMS only 14 were less than 1-month-old at diagnosis [8]. There are no reports of RMS being treated using EXIT, although there is one case report [9] where an EXIT procedure was planned for the treatment of one of a pair of twin fetuses with cervical RMS before hydrops fetalis developed, however the parents declined to give consent because of concern about delivering the normal fetus prematurely and the extent of local invasion of the mass which would make successful resection difficult.

In the 1960's, less than one-third of children with RMS survived, but cure rates are now approximately 70%; great improvement in survival has been made during the past 40 years especially for patients with localized disease [10]. Embryonal RMS, the most common type of RMS, generally responds very well to chemotherapy [6], and good response to chemotherapy allows surgery to be performed less aggressively [7]. However, there was no reduction in size of the tumor in our case after chemotherapy and radical surgical resection was finally chosen for treatment at 2 months of age following preoperative embolization for arterial feeders to the tumor by a neurosurgical colleague. Thus, for the best chance of survival with lowest morbidity, it is paramount that the indications and timing of surgical intervention be appropriate [10].

Treatment of neonatal RMS requires a multidisciplinary approach, where surgery and chemotherapy both have their own specific roles

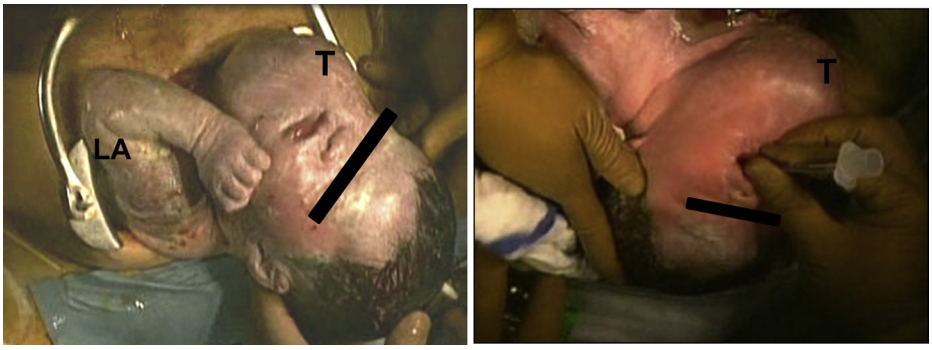

Fig. 2. Exit procedure; initially, the head and left arm (LA) of the baby were delivered through a standard lower segment cesarean section, then the baby was intubated with a 3.0 mm tracheal tube before clamping and dividing the umbilical cord. T: tumor.