A Multidisciplinary Approach for a Newborn with a Large Cervical Teratoma and Hypoplastic Right Heart Syndrome

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Introduction

Congenital cervical teratoma and hypoplastic right heart are life-threatening congenital anomalies which require a multidisciplinary approach for optimal outcome. Our patient presented one day prior to delivery, requiring a rapid, coordinated plan for managing these conditions.

Case Report

A 20 year old G2P1 at 36 weeks 4 days estimated gestational age was admitted to labor and delivery from clinic upon diagnosis of fetal neck mass and complex congenital heart defect. She had received no prior prenatal care. Fetal ultrasound showed a neck mass measuring 10.5 x 10.4 x 9.5cm as well as a single ventricle with L-transposition of the great arteries. MRI showed that the mass invaded the airway at the level of the larynx. Plans were made for cesarean section to be performed the following week with ex-utero intrapartum treatment (EXIT) procedure for management of the newborn’s airway. The day after admission, the mother experienced preterm labor; the decision was made to proceed emergently with cesarean section. Cesarean section was performed under general anesthesia with high-dose Desflurane® of approximately 2 MAC. After uterine incision and before separation from placental circulation, the newborn was intubated by Pediatric ENT using a Parsons laryngoscope®. A size 2.5 endotracheal tube was placed after three attempts. Following this, the infant was delivered and was resuscitated. The infant was taken to the neonatal intensive care unit intubated and in stable condition. At five days of age, under general anesthesia, the newborn had pulmonary artery banding by pediatric cardiac surgery immediately followed by resection of the neck mass by two ENT surgeons. At 13 days of age, the infant returned to the OR for controlled extubation in the presence of pediatric anesthesia and pediatric ENT. The infant tolerated extubation, was later discharged home, and was in good condition at most recent follow-up. Biopsy of the cervical mass confirmed the suspected diagnosis of benign teratoma.

Discussion

Congenital cervical teratoma is a rare condition, with an incidence of approximately 1 in 20,000 to 40,000 live births (1). Although typically a benign lesion, proximity to the upper airway may cause life-threatening airway obstruction. The EXIT procedure has been established for initial management of a variety of fetal conditions and has significantly improved survival rates for congenital cervical teratoma. In the EXIT procedure, uterine incision is made, followed by delivery of the infant’s head and shoulders. The infant is intubated while placental circulation is still intact. The infant is fully delivered only after the airway has been secured. Immediate survival rates for congenital cervical teratoma have been reported at up to 100% with use of The EXIT procedure (2). A concomitant heart defect is rare in congenital cervical teratoma (3). In a typical cesarean section, the goal is to minimize fetal exposure to anesthetic agents and preserve uterine tone to decrease maternal hemorrhage. In contrast, in an EXIT procedure, high levels of inhalation anesthetics are used to maximize uterine relaxation to preserve uteroplacental gas exchange (4). Our infant had a predisposition to the deleterious effects of volatile anesthetics on myocardial contractility and heart rate, yet tolerated high-dose Desflurane® without hemodynamic compromise after delivery. This infant underwent successful orotracheal intubation, delivery, and two complex surgeries, with effective coordination of care that allowed discharge from the hospital.

References