Case report: Giant congenital pulmonary airway malformation initially managed and resected on ECMO

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1. Case report

Our patient is a female fetus who was diagnosed via prenatal ultrasound with a giant right thoracic CPAM at 18 weeks. Due to signs of hydrops, a thoracentesis was performed at 20 + 6 weeks and a significant decrease in the CPAM volume ratio (CVR) from 2.03 to 0.50 was achieved. Fetal ultrasound at 21 + 4 weeks revealed that the CVR increased to 1.36. In addition, skin edema and ascites were noted. Due to the size of the cystic lesions, re-accumulation of fluid, and signs of hydrops, the fetus was treated with a series of shunt decompression, with the insertion of seven shunts on six occasions (21 + 5, 23 + 6, 25 + 5, 27 + 4, 29 + 6, and 30 + 1 weeks). During the course of the gestation, the CVR reached a maximum of 4.96 at 36 + 4 weeks. After initial drainage, signs of hydrops were minimal and were primarily confined to small pleural effusions. To further characterize the extent of this CPAM, a fetal MRI was obtained and demonstrated a multicystic CPAM that filled the entire right chest with significant mediastinal shift into the left chest.

The prenatal course was further complicated by premature rupture of membranes at 30 3/7 weeks, which was treated with antibiotics and bed rest. Courses of beclomethasone were administered on multiple occasions. The infant was delivered by Cesarean section at 36 5/7 weeks. The intrapartum course was uncomplicated. The child exhibited Apgar scores of 4 at 1 min, 5 at 5 min, and 7 at 10 min. The child was intubated for respiratory distress and transferred to the NICU. The child was initially ventilated with oscillatory ventilation and required increasing ventilator support. A cardiac echo demonstrated elevated but not supra-systemic right sided pressures, decreased right sided ventricular function and normal left sided ventricular function. Due to worsening of the child’s ventilation and oxygenation on maximal ventilator support, she was placed on venoarterial ECMO through a right cervical approach. ECMO cannulation and initiation was uneventful.

Due to persistent expansion of the cystic areas of the CPAM and the sheer size of this deformity, we judged that pulmonary recruitment strategies would be ineffective while the lesion remained in situ. Therefore, we proceeded with right lower lobectomy on day two of the ECMO run. The operative course was uncomplicated. The right lower lobectomy was accomplished in an operative time of 69 min with and estimated blood loss of 50 mL. Hypoplastic lung on the affected side was evident. The multicystic CPAM intraoperative measures to minimize blood loss were undertaken, including liberal use of electrocautery during incision and dissection, and the use of adjunctive hemostatic agents such as Surgicel, Thrombin, and Gelfoam. Activate clotting times were maintained between 160 and 180 for the duration of the operation and for the following 24 h. Amicar was not administered. Platelets (30 mL) were transfused intraoperatively to maintain a platelet count greater than 100.

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Postoperatively, chest tube output was minimal and removed on postoperative day number 3. No additional transfusion was required related to operative blood loss. The ECMO run was uncomplicated. Ventilatory management was focused on gentle ventilator strategies to recruit additional functional lung, and the child was maintained on oscillatory ventilation. The ECMO run was terminated on day 8, and decannulation was uncomplicated.

After ECMO decannulation, the strategy of gentle ventilation was continued. Ventilation was changed to conventional tidal ventilation on post-resection day eleven. A gradual ventilator wean was accomplished over 15 days, and the child was extubated from CPAP. Feedings were gradually advanced, and the child was discharged to home 27 days after termination of the ECMO run.

In postoperative follow-up, the child was found to be having difficulty with feeding due to tiring with oral intake, and significant gastroesophageal reflux. A Nissen fundoplication with gastrostomy tube placement was performed when the child was 71 days old, and feedings have subsequently been uncomplicated. She continues to thrive and meet her developmental milestones.

2. Discussion

Congenital cystic adenomatoid malformation (CCAM), also known as congenital pulmonary airway malformation (CPAM), is a type of thoracic abnormality that makes up 30–40% of congenital lung malformations. Prenatal ultrasound is generally the most relied upon method of diagnosing antenatal CPAM and five classes of CPAM exist, with class identification based on cyst size, type (fluid/air-filled or solid), and origin [1]. CPAMs develop within the first six weeks of gestation and may result in improper pseudoglandular stage bronchiolar structure development [2]. Although it is recognized that CPAM may result in fetal hydrops [3] or pulmonary hypoplasia [4,5] and lead to associated respiratory distress, the specific pathogenesis remains unclear. CPAMs are treated surgically through thoracocentesis [6], thoracoamniotic shunt placement [6], or lobectomy [7] and benefits of surgery while on extracorporeal membrane oxygenation (ECMO) continues to be a highly contested topic [8,9].

Major operations can be performed while anticoagulated on ECMO, though they are associated with a higher risk of bleeding complications. Other authors have reported resection of CPAM while on ECMO [10–12]. There are also several reports of ECMO initiated after pulmonary lobectomy due to the development of pulmonary hypertension [13]. In our case, pulmonary lobar resection while anticoagulated on ECMO was performed because of progressive pulmonary failure likely due to a combination of pulmonary hypertension, right ventricular failure, and pulmonary hypoplasia all secondary sequelae of this massive CPAM occupying the majority of the right chest.

CPAM can cause fetal hydrops and pulmonary hypoplasia. These findings were evident in the case presented, and were aggressively managed prenatally, with thoracentesis and thoracoamniotic shunt placement. After delivery, the child experience respiratory failure due to difficulties with pulmonary hypoplasia and with ventilator mechanics related to the size of the CPAM. We opted for ECMO support rather than primary resection, due to concerns about the degree of pulmonary hypoplasia given the massive size of the CPAM. Aggressive prenatal and postnatal management by a multidisciplinary team resulted in a good outcome for this child.

References