Multidisciplinary Management of a Large Microcystic Congenital Pulmonary Airway Malformation

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Abstract

Introduction: Congenital Pulmonary Airway Malformations (CPAMs) are rare sporadic lesions frequently associated with poor fetal prognosis. Type 3 CPAMs are characterized by small hyperechogenic cysts (<5 mm). Hydrops often develops secondarily, and the fetal survival rate is approximately 5% in this setting.

Case Report: We present a case of a large type 3 CPAM complicated by fetal hydrops. The lesion was detected at 19 Gestational Weeks (GW) and confirmed by fetal MRI at 29 GW. At 22 GW, a course of maternal steroids was given as a possible treatment of type 3 CPAM. Peritoneal-amniotic shunt was placed twice to reduce fetal ascites, with unsatisfactory results. Similarly, polyhydramnios was relieved by two amnioreductions, but redeveloped soon after. A baby girl was delivered spontaneously at 33 GW, and received a two-stage partial lobectomy in the first three months of life. She now approaches one year of age, with near-to-normal growth and developmental milestones.

Discussion: The survival of this infant can be attributed to prenatal management, accurate diagnostic imaging, and early postnatal surgical intervention. Her outcome is unexpectedly positive and she may expect a good quality of life. Multidisciplinary vigilance and collaboration with frequent specialist follow ups was the key to success for both mother and child.

Keywords: CPAM; CVR; Peritoneal-amniotic shunt; Polyhydramnios; Fetal hydrops

Introduction

Congenital Pulmonary Airway Malformations (CPAMs) are the most common Congenital Lung Malformations (CLMs). With a prevalence of 1/11,000 to 1/33,000 live births, they are rare overall [1]. Twenty-five to 30% of neonates with antenatally-diagnosed CPAMs will present with respiratory distress and infection, and perinatal mortality may reach 50% [1]. Furthermore, the development of fetal hydrops is a devastating complication that is associated with <5% survival [2-4]. With improvements in the quality of fetal Ultrasonography (US), CPAMs are increasingly detected in the prenatal period [5], leading to the evolution of management strategies that continue to improve peri- and postnatal prognosis [6].

Case Presentation

A 23-year-old primigravida presented at the clinic in April 2020. Her routine US at 19 Gestational Weeks (GW) revealed a fetus with ascites and a large hyperechogenic, homogenous lesion (5.25 cm × 3.58 cm) that occupied the left lung. The contralateral lung was hypoplastic (1.0 cm × 1.5 cm) and extreme cardiac dextroposition was visualized. Fetal US revealed typical features of type 3 CPAM (Figure 1). Routine screening for cytomegalovirus, toxoplasmosis, rubella, and parvovirus B19 was negative. Intrauterine paracentesis was performed and 80 mL of fetal peritoneal fluid was aspirated. Amniocentesis revealed a normal karyotype (46, XX). At 22GW, due to the re-accumulation of peritoneal fluid, 80 mL was drained in a second fetal paracentesis. A course of steroids (Dexamethasone 4 mg × 6 mg) was given as a possible treatment of type 3 CPAM. The ultrasonographic presentation of CPAM did not change and there were no signs of fetal heart failure. The CPAM Volume Ratio (CVR) was 3.02. At 25GW, due to increasing ascites, a peritoneal-amniotic shunt was placed twice but was displaced both times, and the effect was thus unsatisfactory. At the same time, polyhydramnios was detected (Amniotic Fluid Index [AFI] 33cm) and amnioreduction (2000 mL) was performed. At 28 GW, the patient was readmitted.
Fetal hydrops was diagnosed at this time with the presence of ascites, mild hydrothorax and subcutaneous edema (Figure 2). There were no signs of fetal heart insufficiency. A second amnioreduction (2000 mL) was also performed, but polyhydramnios redeveloped soon after (AFI 36 cm). Fetal MRI was performed at 29 GW to confirm the diagnosis of type 3 CPAM. At 33 GW, soon after premature rupture of membranes, the patient naturally delivered a baby girl (2,814 g, Apgar 2,6,6).

The baby presented with respiratory distress and was placed immediately on invasive oscillatory ventilation. General poor condition with oxygen delivery 1.0-0.4 necessitated total parenteral nutrition. Later, poor sucking and swallowing – attributed to the displacement of the diaphragm and aberrant thoracic anatomy – justified PEG placement. Plain film radiography and Computed Tomography (CT) were obtained (Figure 3, 4). CT confirmed the absence of arterial systemic feeding vessels and she was qualified for two-stage partial left lobectomy. The first surgery was performed on day 14, removing the lower 45% of the left upper lobe (Figure 5). The baby girl demonstrated significant desaturations, and it was necessary to balance the challenging ventilation of a cystic and noncompliant upper left lobe, an atelectatic lower left lobe, and a hypoplastic right lung. The second surgery was performed after two months to remove necrotic portions of the left upper lobe. Plain films from February 2021 revealed significant anatomic improvement (Figure 6). Histopathology of the excised tissue confirmed CPAM Stocker type 2 (cysts <2 cm). At the time of writing, the baby has achieved her first year of life. Her developmental milestones are delayed by approximately 3 months, attributed to poor abdominal tone, leading to difficulties with crawling and sitting. Tracheostomy and the use of a ventilator have also caused speech delay.

**Discussion**

**Natural history of CPAMs**

Typically, CPAMs peak in size at 28 GW to 30 GW [7]. Spontaneous prenatal regression occurs in approximately 8% to 49% [8] during the third trimester. The size of our lesion remained generally stable, measuring 5.25 cm × 3.58 cm at 23 GW and 5.84 cm × 4.44 cm at 31 GW. Approximately 15% of CPAMs regress after birth [7,9]. Fetal clinical presentation varies according to lesion size, and affected fetuses may suffer from hydrops, polyhydramnios, and fetal death [3]. Large symptomatic lesions are associated with high in...
results were unsatisfactory. A course of maternal steroids was given at 22 GW. Other than stimulating the growth of the hypoplastic right lung, steroids have been shown to significantly increase fetal survival by reducing the size of microcystic (but not macrocystic) CPAMs (CVR>1.6), which may lead to the resolution of hydrops [17,28]. The pharmacomechanism of steroids on CPAMs is unknown [17] and steroid-prescribing practices among obstetricians vary widely [8]. In our patient, the therapeutic effect of steroids was only minimal [17].

Postnatal care

Surgical resection is the standard of care for symptomatic neonates in order to prevent lung infections, allow for compensatory lung growth and reduce future complications [3,29]. The prognosis after surgery appears to be good [30]. Ben-Ishay et al. [7] performed life-saving pneumonectomies for a series of giant CPAMs, but lobectomy via thoracoscopic (including video-assisted thoracoscopic surgery) or thoracotomy remains the most prevalent surgical method [31]. In our baby girl, respiratory distress and desaturation prompted immediate surgical qualification for a partial lobectomy. The first surgery was performed on day 14. Despite the initial plan to remove the tumor in its entirety, during the procedure it became clear that the cystic lobe did contain viable lung tissue; the closure of the left upper lobe bronchus led to significant desaturation. Thus, only the lower 45% of the left upper lobe could be safely removed. The problem of the restricted thoracic volume (for the lungs and heart to grow and return to anatomic position) remained. Ventilation between the two stages was a significant challenge for neonatal intensivists. The only way to avoid severe barotrauma was to place our patient on oscillatory ventilation with deep sedation and use of paralytic agents. There are no neonatological guidelines dictating the long-term use of ventilation and sedation in such a unique situation. This strategy appeared to be effective, and it was possible to reduce oxygen delivery from 100% to 40% during this period. The baby completed her two-stage procedure in 2 months. Because of the large size of the CPAM, the anatomic improvement was immediately appreciable (Figure 6). As a result, her growth and developmental milestones are only slightly delayed. She is followed up frequently in specialist centers including otolaryngology, physiotherapy and speech therapy.

Conclusion

Type 3 CPAMs complicated by fetal hydrops are associated with high perinatal mortality. Antenatal interventions are typically ineffective. The CVR of 3.02 was a significant predictor of neonatal respiratory distress and prompted the consideration of early surgical intervention. Partial lobectomy allowed for the compensatory growth of healthy lung tissue. Baby’s positive outcome can be attributed to prenatal management and early postnatal surgical intervention; furthermore, she may expect a good quality of life. Multidisciplinary vigilance and collaboration with frequent specialist follow ups was the key to success for both mother and child.

References


