

CASE REPORT

Successful Thoracoscopic Excision of Type 0 CPAM in a Newborn

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ABSTRACT

Thoracoscopic excision of a Type 0 congenital pulmonary airway malformations (CPAM) is rarely described in literature. A 1-day-old female neonate presented with prenatal diagnosis of right lung CPAM. Mother had undergone fetoscopic thoracoamniotic shunt placement which led to radiographic resolution of large CPAM cyst. Soon after delivery, the patient developed multiple premature ventricular contractions (PVCs) that were thought to be secondary to retained shunts. Thoracoscopic excision of CPAM performed.

Key words: Congenital pulmonary airway malformations; CPAM; Neonate; Prenatal diagnosis; Fetal intervention

INTRODUCTION

Congenital pulmonary airway malformation (CPAM) is rare congenital anomaly with a prevalence of 1 in 10,000-35,000 pregnancies.[1] The pathophysiology involves nonfunctioning cystic abnormal tissue in lungs preventing formation of normal lung parenchyma. Five different CPAM types have been described in the literature.[2] Type 0 CPAM, also known as acinar dysplasia or dysgenesis, is the most rare and comprises less than 2% of CPAM types and is usually associated with pulmonary hypoplasia and poor prognosis.[3,4] To our knowledge, this is the first report on an atypical CPAM type 0 presenting without pulmonary hypoplasia.

CASE REPORT

One-day-old female neonate presented with prenatal diagnosis (24th week) of right upper lobe CPAM. Due to progressive increase in cyst size, she underwent fetoscopic thoracoamniotic shunt placement at 28-week gestation. Procedure was complicated by internalization of first shunt requiring placement of second shunt. Rest of the pregnancy was uncompli-

cated with delivery at 39th week of gestation. Shortly after birth, she developed intermittent premature ventricular contractions (PVCs) that were thought to be secondary to retained shunts. She was subsequently transferred to our institution for management of CPAM and PVCs. Chest radiography confirmed retained shunts (Fig.1). Decision was then made to take patient to the operating room for diagnostic thoracoscopy with removal of prenatal shunts.

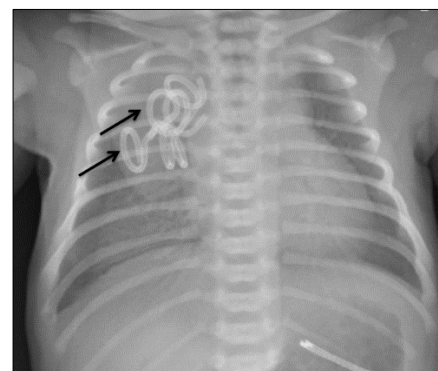


Figure 1. Preoperative chest radiograph showed two shunts (black arrows) coiled in the right upper thorax with a rounded opacity surrounding the shunts.

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After induction of anaesthesia the patient was placed in the left lateral decubitus position. Three 3 mm ports were used to get access to right chest cavity. Pneumothorax was created using 5 mmHg and 3 L/min. Upon entry a large subpleural structure was visualized. The cyst appeared to be separate from all three lobes and did not involve the parenchyma. The pleura was then opened using a combination of sharp dissection and 3 mm sealer device. The cyst was then partially enucleated and appeared to contain two thoracoamniotic shunts. Using a combination of the 3 mm sealer, blunt dissection and sharp scissors, we were able to further skeletonize, which was noted to arise from the trachea (Fig.2). With further skeletonization the lesion was liberated from the trachea. No air leak was observed using underwater testing. The pneumothorax was evacuated and the lesion was removed and opened in the back table to extract the two drains (Fig.3). Finally, we performed rigid bronchoscopy and no tracheal injury was observed.

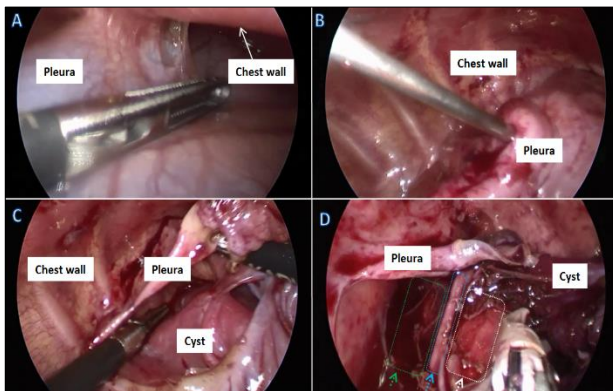


Figure 2. Pleura being separated from chest wall (A). Needle drainage of cyst cavity (B). Pleura opened to visualize collapsed cyst (C). Dissection at the level of proximal trachea showing posterior to anterior, esophagus (green arrow), vagus nerve (blue arrow) and trachea with cyst attachment (white arrow) (D).



Figure 3. CPAM specimen with the two thoracoamniotic shunts.

The cystic mass was sent to pathology and reported as a type 0 CPAM. She was extubated on postoperative day 1 and chest tube was removed next day. She was discharged 8 days after the procedure.

DISCUSSION

We present a Type 0 CPAM in the newborn period, managed successfully with a thoracoscopic approach. Today with the availability of more enhanced imaging techniques including prenatal ultrasound, more congenital airway defects are being diagnosed.[3] Despite the increase in diagnosis, it is often hard to predict if the patients will develop symptoms right after birth. Current literature suggests that both symptomatic and asymptomatic cases of CPAM should be managed with surgical excision in infancy to prevent risk of symptom development or malignancy later in life.[5,6]

The prenatal management of CPAM involves maternal steroid therapy for the maturation of the fetal lungs, and thoracoamniotic shunting in severe cases.[7] Thoracoamniotic shunts are novel strategies in the management of CPAM, however, this procedure is not without risks, and there are reports of shunt dislodgement that resulted in refractory tension pneumothorax or required retrieval in the neonatal period.[8,9] Our patient developed PVCs presumably secondary to the retained thoracoamniotic shunts. There are only two reports of thoracoamniotic shunt catheter dislodgement that required thoracoscopic removal in the neonatal period. Inoue et al, reported three cases of shunts placement for congenital chylothorax and subsequently dislodged, causing massive pleural effusions. Thoracoscopic removal of the dislodged shunts performed subsequently.[10]

Although the management of CPAM in general is surgical resection, treatment for type 0 CPAM was usually supportive, as up until now it has been considered a lethal diagnosis in the neonatal period.[6] There is one case report of an infant who survived an incomplete form of acinar dysplasia; managed with supportive care without surgical excision. Chow et al, reported a patient with severe respiratory distress at 1 minute after birth and subsequently put on mechanical ventilation and veno-venous ECMO. She improved and was discharged at 4 months of age. Open and thoracoscopic excision of CPAM is performed. In the index case, we removed CPAM thoracoscopically.

Conclusion:

The dearth of literature on CPAM type 0 can be partially attributed to its rarity, and as such, a particular effort must be made to continue characterizing the nature and presentation of these lesions. VATS help delineate and differentiate the origin of CPAM types and allows for a safe exploration and operation, especially in the face of an atypical presentation of CPAM type 0. It is our recommendation that

CPAM type 0 should be removed thoracoscopically and can be safely done right after birth if clinically indicated.

Consent: Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

Author Contributions: All the authors contributed fully in concept, literature review, and drafting of the manuscript and approved the final version of this manuscript.

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