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Congenital Cystic Adenomatoid Malformation in a Premature Newborn: Case Report

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Abstract: ***Congenital cystic adenomatoid malformation (CCAM) is a rare congenital condition affecting the respiratory systems development. This case report presents a preterm newborn at 32 weeks gestation who experienced respiratory distress. A persistent radiolucent image in the lung base led to imaging studies, diagnosing CCAM, which required surgical intervention. The case highlights the complex management required for such malformations and the successful outcome following a lobectomy of the right lower lobe. This report emphasizes the need for early diagnosis and multidisciplinary care.***

Keywords: cystic adenomatoid malformation, pulmonary hypertension, lobectomy, Congenital pulmonary airway malformation

1. Introduction

Congenital cystic adenomatoid malformation (CCAM) is defined as a benign anomaly characterized by an abnormally developed pulmonary mass, which can be cystic, solid, or mixed (1,2). It is often detected through imaging studies during pregnancy (2,3).

The incidence of CCAM is variable with estimated ranging from 1 in 10,000 to 35,000 live births. The variability in these figures may be attributed to differences in diagnostic practices and the quality of prenatal ultrasounds [2,3,4].

In 1977, Stocker proposed a classification that divided congenital adenomatoid malformation into three main types based on the histological and radiological appearance of the lesions (3). This classification was later modified in 2002 to include types 0 and 4 and to change the name to Congenital Pulmonary Airway Malformation (CPAM) (3), see Table 1.

Table 1: Modified Stocker Classification

Modified Stocker Classification	Characteristics	Clinical aspects
Type 1	Large cysts >2 cm (which can be single or multiple).	Favorable prognosis: they can be asymptomatic, but if symptoms or complications arise, they may require surgery.
Type 2	Proliferation of numerous small cysts (which can affect a large area of the lung) and give a spongy appearance.	Significant respiratory problems due to extensive lung tissue involvement. Surgical intervention is often required.
Type 3	Solid mass containing cystic areas, with a high risk of malignancy.	Less favorable prognosis: it can cause pulmonary hypertension. Surgical resection is often necessary.
Type 4	Adenomatoid Malformation with Chronic Inflammatory or Infectious Components.	It often requires antibiotic or anti-inflammatory treatment in addition to surgical intervention.
Type 0	It originates at the tracheobronchial level, representing a severe alteration in the development of the functional units of the lung.	Extremely severe prognosis, generally incompatible with life.

Modified from Dehner, L. P., Schultz, K. A. P., & Hill, D. A. (2023). Congenital pulmonary airway malformations with a reconsideration and current perspective on the Stocker classification. Pediatric Developmental Pathology, 26(3), 241-249.

The diagnosis of CCAM typically begins during pregnancy through prenatal ultrasound (2,3,4), which can reveal unusual pulmonary masses; however, confirmation is required after birth through extensive imaging studies such as computed tomography (CT) or magnetic resonance imaging (MRI). These studies identify the cystic condition and evaluate the extent of the malformation [4,5].

Complications associated with CCAM can be life-threatening and vary depending on the type and severity of the malformation. Common complications include recurrent pulmonary infections, pulmonary hypertension, and, in severe cases, chronic respiratory failure (5,6). Management often requires a multidisciplinary approach, involving collaboration among neonatology, pediatric surgery, and pulmonology [4, 5, 6].

The treatment of CCAM varies depending on the classification and severity of the malformation (5,6). In asymptomatic cases, or when the malformation is small and not causing significant problems, regular monitoring without

immediate surgical intervention may be considered (5,6). It is important to conduct periodic examinations and imaging studies to monitor the size and progression of the condition, as well as to provide respiratory care, including supplemental oxygen if necessary, and management of any respiratory symptoms (5). Surgery is considered when the malformation causes significant symptoms, compromises pulmonary function, or presents a risk of complications. Factors that may lead to surgery include respiratory distress, recurrent infections, pulmonary hypertension, an increase in size affecting pulmonary function or compressing adjacent structures, and concerns that the malformation may transform into a neoplasm. Surgery typically involves resection of the affected tissue; see Table 2 (5,6).

Table 2: Surgical Procedure

Surgical Procedure	*Definition*
Segmental Resection	Removal of a portion of the affected lung.
Lobectomy	Removal of an entire lobe of the lung if the malformation is localized to a single lobe.
Pneumonectomy	Removal of the entire affected lung.

Modified from Hall, N. J., & Stanton, M. P. (2017). Long-term outcomes of congenital lung malformations. Seminars in Pediatric Surgery, 26(5), 311-316.

The prognosis varies depending on the type of malformation and the intervention performed. Type I malformations generally have a good prognosis, while types III and IV may have a more reserved prognosis. Long-term follow-up is required to monitor pulmonary function, detect potential late complications, and assess the patient's growth and development (1,3,5,6).

2. Methodology

The methodology of this case report involves a comprehensive review of the patient's medical history presented during the prenatal period, in addition to the concepts of each specialty that provides treatments, all of the above combined with a relevant review of the literature in databases such as PubMed, Medline, Embase, to explain the findings found.

3. Case Report

A preterm newborn at 32 weeks gestation by Ballard, delivered via cesarean section due to acute fetal distress, required immediate admission to the neonatal intensive care unit. This was secondary to respiratory distress from grade II hyaline membrane disease (two doses of exogenous pulmonary surfactant were supplied) and initial pulmonary hypertension of 70 mmHg, managed with high-frequency oscillatory ventilation (HFOV) and nitric oxide at 20 ppm for 48 hours, to which the newborn responded well, with a transthoracic echocardiogram showing pulmonary pressure at 25 mmHg (7).

Despite improving the pulmonary hypertension, the patient remained on conventional invasive mechanical ventilation (IMV) without improvement in ventilatory parameters. A chest X-ray revealed a consolidative image in the right lower lobe, initially suspected to be pneumonia, and was managed with antibiotic therapy. After one week, a follow-up showed microcystic images in the right lower lobe (Figure 1). Further diagnostic studies were required, and a chest CT scan reported clustered cystic lesions in the right lower lobe suggestive of congenital cystic adenomatoid malformation (CCAM) (Figure 2). Pediatric surgery service indicated the need for surgical intervention.

At 36 days of life, a lobectomy of the right lower lobe was performed, along with closure of the fistula, with intraoperative findings of a multicystic mass (multiple cysts of varying sizes) and a bronchopleural fistula. A biopsy was collected for pathological study, which reported cystic adenomatoid transformation of the right lower pulmonary lobe, without malignancy.On the third day post-operative, he developed a pleural effusion that resolved spontaneously. He continued to show favorable progress and improvement in ventilatory parameters, leading to a successful extubation. At 58 days of life (40.2 weeks corrected gestational age), he tolerated oral feeding and had good respiratory mechanics. Medical discharge was decided, with ongoing management including inhaled beta-2 agonists and corticosteroids due to the context of bronchopulmonary dysplasia.

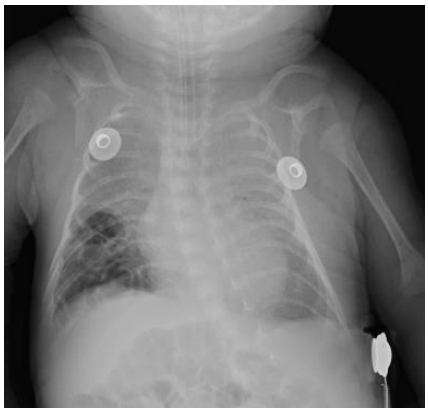

Figure 1: Chest X-ray AP (microcystic images in the right lower lobe)

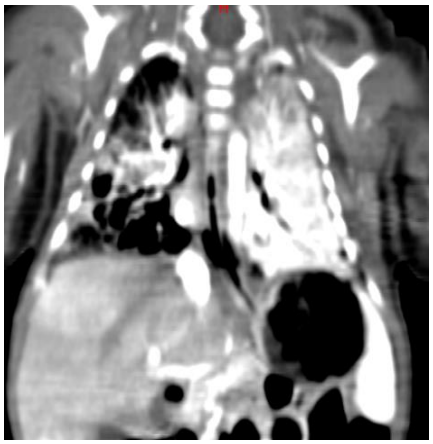

Figure 2: Chest CT (clustered cystic lesions in the lower pulmonary lobe)

4. Discussion

Congenital pulmonary airway malformation (CPAM), formerly known as congenital cystic adenomatoid

malformation (CCAM), is an uncommon anomaly of the lower respiratory tract development (1,2,3). Data suggests an incidence of congenital pulmonary cysts ranging from 1 in 8,300 to 35,000 live births (8,9). It encompasses a variety of cystic and non-cystic lung lesions resulting from an early disruption in the embryonic development of the airways. More than 70% of cases are asymptomatic at birth, and spontaneous regression has been observed in some instances without the need for intervention. This condition was first described in the English literature in 1949 by Chiang and Tang (10), and the first prenatal diagnosis via ultrasound was made by Garret and colleagues in 1977 (11).

CPAM may be associated with the presence of neonatal pulmonary hypertension due to increased vascularization of the pulmonary arteries and heightened reactivity of the pulmonary vascular bed to vasoconstrictive stimuli such as hypoxia and acidosis, a finding observed in our patient (12).

The diagnosis can be prenatal or postnatal. The prenatal diagnosis of CPAM is most based on ultrasound findings first detected in the second trimester; approximately 25% of cases are symptomatic. Some ultrasound predictors of respiratory distress at birth include mediastinal shift, polyhydramnios, and ascites. Hydrops associated with poor cardiac function is linked to a high risk of perinatal death (8, 13).

In the case of a postnatal diagnosis, imaging techniques used include chest X-rays, which in our case allowed us to observe numerous homogeneous microcysts. Another significant diagnostic tool is chest CT, which identifies small cysts with greater precision, providing information on size, content, and other important characteristics. These examinations facilitate diagnosis, establish the exact topography, visualize vascular supply, and exclude possible associated anomalies (13).

Congenital cystic adenomatoid malformations (CCAM) are distributed evenly between the right and left lungs and can appear in any lobe (14). They are generally uni-lobular, as the case of our patient, located in the right lower lobe. According to its histopathology, our case falls into type 2, which comprises 15 to 20 percent of CPAM cases. It consists of multiple cysts ranging from 0.5 to 2 cm in diameter and solid areas that merge with adjacent normal tissue and have no risk of developing malignancy (8). Generally, patients are asymptomatic; in our case, typical signs of respiratory distress were present, including tachypnea, increased respiratory effort, muscle retractions, and generalized cyanosis (8). We highlight the difficulty in diagnosing our patient due to the condition of prematurity, which masked the clinical characteristics, primarily at the respiratory level, leading us to initially consider more prevalent pathologies for the gestational age.

The severity of these anomalies can be explained by how well conventional treatment responds to pulmonary hypertension and congenital malformations. Available treatments include high-frequency ventilation, induced alkalosis, nitric oxide, sedation, and paralysis (12). Once the patient is stabilized, surgical resection is considered, which is indicated to prevent future pulmonary complications such as recurrent infection, compression of adjacent structures, and malignant transformation. In our case, a lobectomy of the right lower lobe was performed via open thoracostomy; however, thoracoscopy appears to yield better results (8). The definitive diagnosis is established through histopathological examination, demonstrating the type of epithelium lining the malformation (lung parenchyma with formation of multiple cysts lined by cuboidal epithelium and with an adenomatoid appearance) (8,9,12).

5. Conclusion

Congenital cystic adenomatoid malformation CCAM is a rare but significant congenital anomaly that poses complex clinical challenges, particularly in preterm infants. Early diagnosis and surgical intervention, as demonstrated in this case, are critical to managing the condition effectively and ensuring favorable longterm outcomes. This case highlights the importance of comprehensive prenatal evaluation and multidisciplinary management to address potential complications.

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