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Case Report

From prenatal diagnosis to postnatal stability: a case of isolated type II congenital pulmonary airway malformation

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ABSTRACT

Congenital pulmonary airway malformation (CPAM) is the most common congenital lung anomaly. CPAM is characterized by multiple cysts of varying diameter, of embryological origin. With advances in prenatal imaging, it is frequently detected antenatally. We report a male neonate with antenatal diagnosis of CPAM in the lungs at 22 weeks of gestation. The baby was delivered at 39 weeks via planned cesarean section, weighing 2425 gm, and had mild transient tachypnea after birth. Postnatal imaging confirmed multiple thin-walled cysts in the right lung, without compression of adjacent structures. No associated anomalies were found. The neonate was managed conservatively and discharged with multidisciplinary follow-up. This case highlights the possibility of a benign course in asymptomatic CPAM type II, underscores the role of prenatal and postnatal imaging, and discusses current controversies regarding conservative versus surgical management.

Keywords: Congenital pulmonary airway malformation, Congenital cystic adenomatoid malformation, Alveolar developmental abnormality, Stocker's classification

INTRODUCTION

Congenital pulmonary airway malformation (CPAM) is a rare developmental disorder, resulting from abnormal branching morphogenesis of the tracheobronchial tree during fetal life. The condition encompasses a spectrum of cystic and solid lesions arising from failure of normal alveolar development and airway differentiation. Reported incidence varies from 1 in 7,200 to 1 in 35,000 live births, with differences largely attributed to advances in prenatal ultrasonography and increased surveillance during the second-trimester anomaly scan.[1-3]

Previously known as congenital cystic adenomatoid malformation (CCAM), the term was changed to CPAM to better reflect its airway origin and microscopic features. Stocker's classification divides CPAM into five types (0-IV) based on cyst size, histology, and embryologic origin.[4,5] Type I (most common) has large cysts; type II (15-20% of cases, like the present case) has multiple small cysts under 2 cm; type III is a solid microcystic mass; type IV has large peripheral cysts; and type 0, the rarest, arises from tracheobronchial tissue and is usually fatal.

Exact pathogenesis is poorly understood, but there are a variety of genes implicated in this process. All of these genes have a role in cell proliferation or apoptosis, leading to the various types of malformations under the CPAM classification.[2,6] Another theory suggests that blockage of a fetal airway causes the distal airways to enlarge and form cysts.

CPAM is usually detected by prenatal ultrasound between 18-24 weeks of pregnancy, which shows a cystic or solid-looking mass in the fetal chest. Lesions can stay the same size, grow, or shrink; up to half disappear on their own, often in the third trimester.[7] Larger lesions, mediastinal shift, and signs of hydrops fetalis predict a worse outcome.[8] The CPAM volume ratio (CVR)-lesion

volume divided by head circumference-over 1.6 signals higher hydrops risk.[7,9] Infection is the most common complication of CPAM, usually appearing in the first few years of life in infants who remain untreated. The main malignant complication is pleuropulmonary blastoma (PPB), with a higher risk in type 4 CPAM, multifocal or bilateral cysts, family history of PPB, or history of pneumothorax. Type 1 CPAM is linked to bronchioloalveolar carcinoma.[3,10]

After birth, a chest radiograph is usually the first test, showing cystic or overly bright lung areas. High-resolution CT scans are the most accurate for diagnosis and surgery planning because they show detailed lesion shape, cyst size, and spread.[11] Treatment depends on symptoms and lesion features. Babies with symptoms need quick surgery, usually lobectomy. For symptom-free cases, there is debate-some doctors recommend surgery to prevent later problems, while others choose careful monitoring.[3,12]

CASE REPORT

A male newborn, with an antenatal ultrasound scan at 22 weeks of gestation, showing cystic changes in the right lung, suggesting CPAM, was delivered at 39 weeks of pregnancy to a primi mother by planned cesarean section. Child cried immediately after birth, weighing 2425 gm at birth. On admission child had mild respiratory distress in form of tachypnea. Other vital parameters were normal. He was admitted to the neonatal intensive care unit (NICU) for observation but remained well, with stable vital signs and normal physical examination. After initial supportive treatment, the distress settled within few hours, most likely to be transient tachypnea of newborn. The baby had no further signs of breathing difficulty.

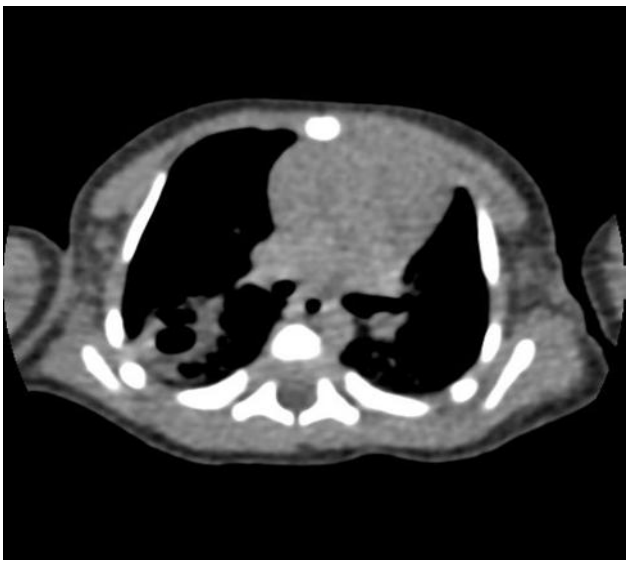

Figure 1: Axial non-contrast HRCT image demonstrating multiple cystic lucencies in the affected lung parenchyma with intervening soft-tissue attenuation.

Chest radiograph showed multiple small, well-defined cystic areas in the right lung. A CT scan confirmed several thin-walled cysts less than 2 cm in size, localized to the right lung (Figure 1 and 2), consistent with CPAM type II, without any compression of nearby structures. Ultrasound of the abdomen, neurosonogram, and 2D echocardiogram were performed to assess for associated malformations. No other congenital abnormalities were detected during initial screening. Child was discharged, and Parents were counseled regarding benign condition of the lung malformation, presently, and requiring regular paediatric surgeon and pulmonologist follow-up. On the latest follow-up at 2 years of age, child's development and physical growth were normal, and no recurrent respiratory complaints.

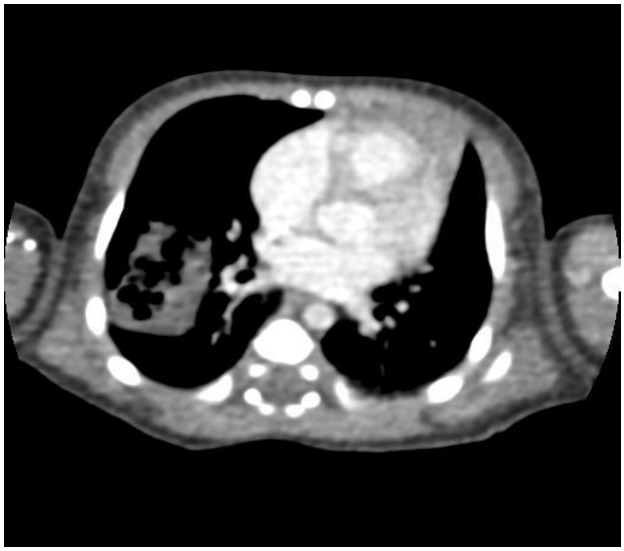

Figure 2: Axial contrast-enhanced HRCT image showing better delineation of the cystic components and their relationship to adjacent vascular structures.

DISCUSSION

This case presents an antenatally detected, isolated CPAM type II presenting with minimal postnatal symptoms, confirmed on imaging, which was successfully managed conservatively. The prenatal detection allowed for planned delivery in a facility with neonatal intensive care, ensuring immediate access to specialized care if symptoms had progressed.

The diagnosis of CPAM type II carries specific prognostic and management implications, because these are associated with the highest rate of concurrent congenital anomalies among CPAM types, occurring in up to 60% of cases.[4,5] The absence of associated anomalies in this patient significantly reduced the likelihood of complex postnatal management. Postnatal CT scanning confirmed the lesion morphology and excluded complications such as mass effect or mediastinal shift, supporting the decision to defer surgery.

Various case reports have established positive findings in chest radiograph for the diagnosis of CPAM.[13,14] CT remains the reference standard for postnatal diagnosis, providing critical input for planning between conservative and surgical management.[11]

Management of asymptomatic CPAM continues to be debated. A systematic review by Kapralik et al found no clear superiority of surgical over conservative management in asymptomatic patients, though about one-quarter of observed cases later developed symptoms.[12] Infections are the most common complication, often prompting eventual surgery. Proponents of early elective resection argue that surgery is technically easier before infection or inflammation causes adhesions, and that removal eliminates future malignant potential.[2,15] Conversely, others highlight that many CPAMs remain asymptomatic and stable, making the risks of surgery and anesthesia unjustifiable in low-risk cases.[4,12,15]

In our patient, the small, non-compressive lesion, stable course, and absence of associated anomalies favored observation. This approach requires structured follow-up, typically including periodic clinical assessment and imaging during infancy and early childhood. Parents must be educated about warning signs such as recurrent respiratory infections, wheezing, or acute respiratory distress.

The long-term outlook for conservatively managed CPAM type II is generally favorable. Compensatory lung growth in early life allows children who undergo later resection to maintain near-normal pulmonary function.[12] Those managed non-operatively often achieve normal function as well, although subtle deficits may appear in some studies, emphasizing need for ongoing surveillance.

This case also reinforces the importance of multidisciplinary collaboration. Optimal outcomes rely on coordinated care among radiologists, neonatologists, surgeons, and pulmonologists, particularly in tailoring decisions to individual patient risk profiles. As molecular and genetic understanding of CPAM advances, future risk stratification may become more precise, potentially guiding the timing and necessity of intervention.

CONCLUSION

Isolated CPAM type II diagnosed antenatally and asymptomatic at birth can be managed conservatively with careful follow-up. Imaging plays a central role in confirming diagnosis and guiding management. Multidisciplinary care ensures that decisions are individualized, balancing the low risk of malignancy against the potential complications of surgery and anesthesia in infancy.

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