ECRONICON OPEN ACCESS

EC PAEDIATRICS
Case Report

Pneumonia Secondary to Congenital Pulmonary Adenomatoid Malformation, Complicated by Methicillin-Resistant *Staphylococcus aureus* Sepsis

Sarah Aslam, Mamoona Hashim*, Munazza Noor Mangi, Syed Zafar Mehdi, Hafiza Laila Hashim Khan, Summera Hashim

Liaquat College of Medicine and Dentistry, Pakistan

***Corresponding Author:** Mamoona Hashim, Liaquat College of Medicine and Dentistry, Pakistan.

Received: June 26, 2025; **Published:** July 10, 2025

Abstract

Congenital pulmonary adenomatoid malformation (CPAM) is an infrequent developing lung abnormality. We document the case of a 14-day-old term infant who had escalating respiratory distress, repeated pneumothoraxes, and substantial pleural effusion before being diagnosed with bilateral CPAM with superimposed infection. The case exemplifies the diagnostic obstacles given by CPAM when masked by acute respiratory disease and the significance of sophisticated imaging and interdisciplinary care in such complex patients.

Keywords: *Cystic Adenomatoid Malformation Of Lung; Congenital; Neonate; Pneumothorax; Pleural Effusion; Respiration*

Abbreviations

CPAM: Congenital Pulmonary Adenomatoid Malformation; CCAM: Congenital Cystic Adenomatoid Malformation

Introduction

Congenital Pulmonary Adenomatoid Malformation (CPAM), previously known as Congenital cystic Adenomatoid Malformation (CCAM) is a rare developmental anomaly of the lung. Although uncommon, it affects approximately 1 in 10,000 to 35,000 live births. CPAM constitutes 95% of congenital cystic lung disease and 25% of all congenital lung malformations. The case mortality rate ranges from 9% to 49% [1]. CCAM was first described by Chin and Tang and first reported in the English medical literature in 1949. Presentation to CPAM is usually with acute respiratory distress. In the absence of involvement of the alveoli, CCAM is thought to result from an over proliferation of tubular-bronchial structures in a regulated and non-regulated fashion. Single-lobar involvement is four times more frequent than multi-lobar diseases, but both the left and right lungs are affected with equal frequency [2]. It is due to malformations of the bronchopulmonary foregut resulting from the interrupted development of the bronchial tree in early pregnancy. The majority of CPAM are diagnosed in the first 2 years of life, presenting with symptoms like respiratory infections and poor development. Although isolated CPAM are most common, they may also present with other anomalies, including skeletal malformation, renal agenesis or dysgenesis, gastrointestinal atresia, and cardiac defects [3].

Case Presentation

A 14-day-old male neonate, weighing 3.0 kg and born at term via elective lower segment caesarean section to a G3P2+0 mother, was admitted to the NICU via OPD on October 7, 2023. He presented with a 3-day history of cough and respiratory distress, in addition to one day of documented temperature peaking at 102°F that was momentarily alleviated by paracetamol. There were no vomiting, convulsions,

diarrhoea, or use of non-recommended neonatal treatments. The baby was entirely breastfed and had natal immunisations, and no substantial familial or socioeconomic issues were identified.

Upon examination, the infant was sluggish and febrile, with signs of respiratory distress. On room air, oxygen saturation was 88%, respiratory rate 64/min, pulse 170 bpm, and capillary refill time (CRT) more than 3 seconds. The auscultation revealed decreased air entry on the left lower chest.

Given the clinical picture, late-onset neonatal sepsis and pneumonia were evaluated. A chest X-ray and laboratory assays were performed as part of the first inquiry (Figure 1). The newborn was treated with B-CPAP, IV fluids, intravenous ampicillin (200 mg/kg/day, 6 hourly), gentamicin (4 mg/kg/day), and inotropic support.

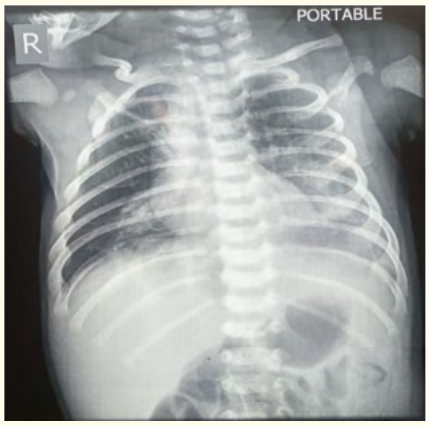

Figure 1: *Initial chest x-ray.*

The patient became severely desaturated on the second day of admission. A left-sided tension pneumothorax was discovered during the evaluation and verified with a radiograph (Figure 2A). An immediate needle thoracostomy followed by chest tube insertion was done (Figure 2B). The infant was placed under mechanical ventilation, and antibiotic coverage was boosted to IV meropenem (20 mg/kg every 8 hours) and vancomycin (10 mg/kg every 8 hours).

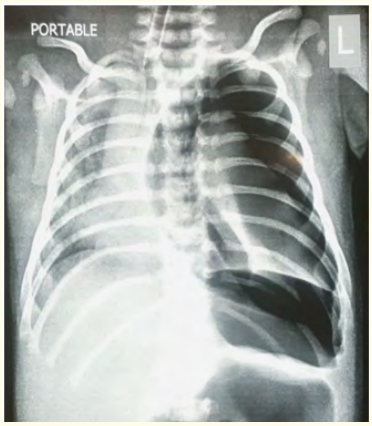

Figure 2A: *Chest X-ray showing a left-sided tension pneumothorax with mediastinal shift.*

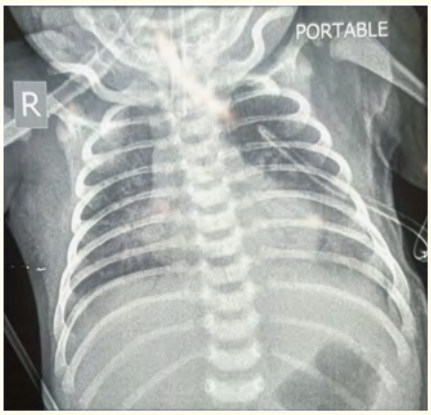

Figure 2B: *Chest X-ray post left thoracostomy demonstrating lung re-expansion and chest tube in situ.*

During the first 72 hours of ventilation, sepsis indicators were substantially raised, demanding the administration of cryoprecipitates and extra FFP. As ventilator settings were gradually weaned, the baby was prepared for extubation; however, extubation was postponed due to imaging that indicated a new right-sided pneumothorax (Figure 3A), mandating right chest tube installation. Figure 3B shows the post-right chest tube x-ray.

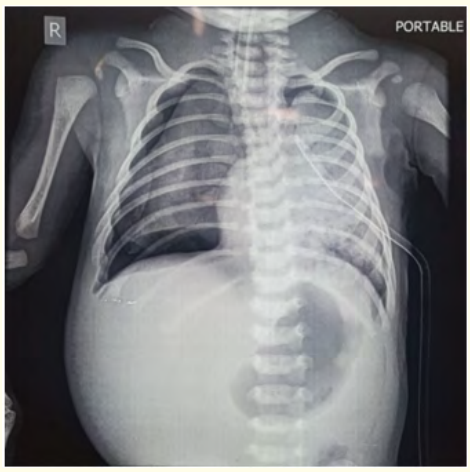

Figure 3A: *Chest X-ray showing right-sided pneumothorax with absent peripheral lung markings on the right and visible pleural line.*

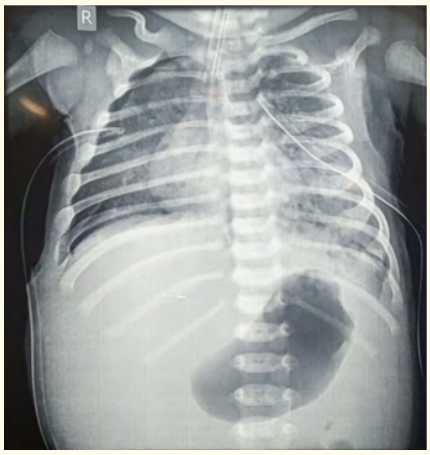

Figure 3B: *Post right chest tube insertion with re-expanded right lung.*

After four days, clinical and biochemical developments enabled successful extubation to B-CPAP. However, after 48 hours, the neonate developed significant respiratory distress. A follow-up X-ray disclosed a left-sided large pleural effusion and right-sided lung consolidation (Figure 4A). The left chest tube was adjusted, and the child was reintubated. The pleural aspirate was exudative; it was sent for culture, which returned negative, but MRSA was discovered in blood cultures (covered by continuous vancomycin). Figure 4B shows the radiographic presentation of post-reintubation resolving left-sided pleural effusion.

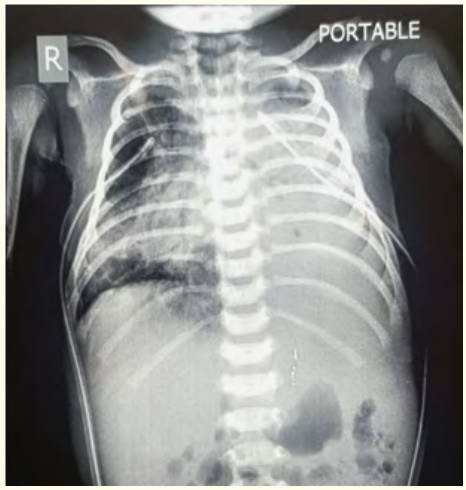

Figure 4A: *Left-sided massive pleural effusion and right-sided consolidation.*

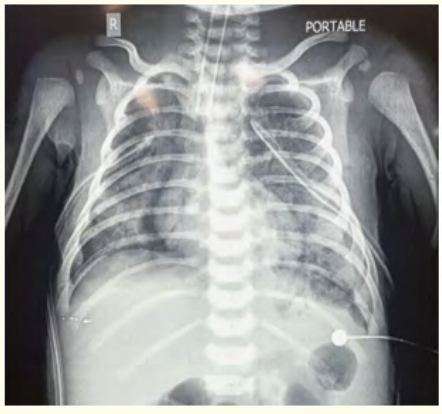

Figure 4B: *Resolving left-sided plural effusion.*

Following re-intubation and clamping of the right chest tube, the baby acquired a massive right-sided pneumothorax that subsided with de-clamping, which can be seen in figure 5A and figure 5B, respectively. This sequence sparked suspicions about an underlying structural abnormality. The patient progressively got better and was successfully extubated using B-CPAP.

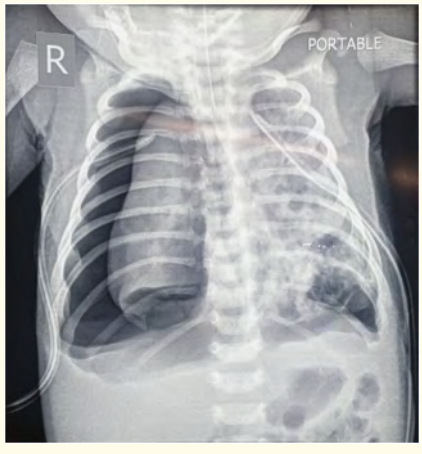

Figure 5A: *Depicts the right side of the massive pneumothorax following the clamping of the right chest tube.*

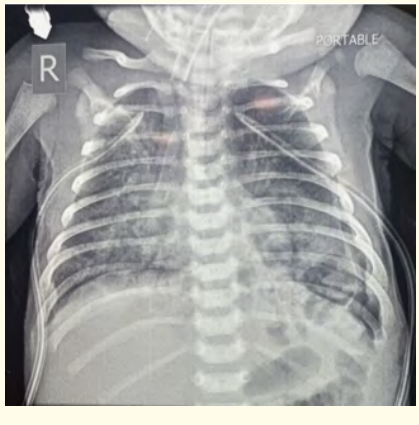

Figure 5B: *Depicts the resolution of a right-sided pneumothorax following the declamping of the right chest tube.*

Additional imaging, comprising a chest X-ray and a CT scan with IV contrast (Figure 6A and Figure 6B), revealed bilateral multiloculated cystic lesions. The left lower lobe had multiple fluid- and air-filled cysts with loss of normal architecture, while the right lower lobe had comparable lesions as well as consolidation and atelectasis. The outcomes correlated with bilateral congenital pulmonary airway malformation (CPAM), which was exacerbated by reaffirmed pneumothoraxes and infection. Our patient was operated on for CPAM but was found to develop a bronchopleural fistula on the third postoperative day and required thoracotomy and repair. We sent him home in a stable condition with advice to attend follow-up, and the surgical result was satisfactory. The child is one year old now and has achieved milestones at an appropriate age.

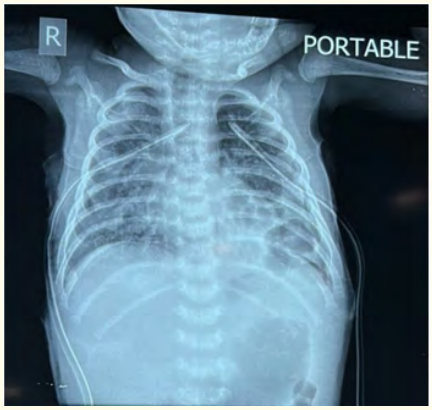

Figure 6A: *Chest X-ray showed cystic lesions in the lower zones of the lung.*

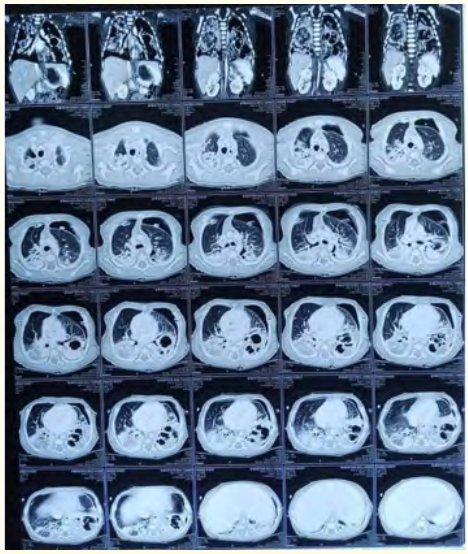

Figure 6B: *Depicts a CT scan with IV contrast that revealed bilateral multiloculated cystic lesions.*

Discussion

Our case illustrates diagnostic and management difficulties associated with CPAM, particularly relapsing pneumothoraxes and infective complications. This case is consistent with several previously reported cases from different parts of the world, thereby emphasizing the varied manifestations and diagnostic delays commonly seen in CPAM. Likewise, a 2013 publication from Sudan reported a 40-day-old male infant who had respiratory difficulty and poor feeding since day 3 of life. An initial imaging was non-specific, whereas CT confirmed CCAM [1]. A 2016 Indian case report described a 5-month-old child with left-sided pneumothorax and cystic lung lesion on CT. Despite empiric antibiotics for suspected staphylococcal pneumonia, there was little improvement until the discovery of CPAM [2]. CPAM is a rare hamartomatous lesion of the lung of unknown aetiology. It is not hereditary [4]. Cases of bilateral CPAM lesions may be related to genetic expression of abnormal cell signalling, increasing the likelihood that a CPAM or other areas of the lung go on to develop cancer [5]. Hydrops fetalis and microcystic CPAM are known risk factors for poor prognosis [6]. According to CCAM Stocker's classification, Type I (large cysts > 2 cm) has a favourable outcome, Type II (multiple smaller cysts < 1 cm) is commonly associated with other anomalies, and Type III is very rare and has tiny macroscopic cysts (<0.5 cm) which present a spongy parenchyma. The recent CPAM classification factors in the stages of developmental arrest. They are categorized from tracheal (Type 0) to acinar (Type IV) based on the size of cysts and histological findings. Type IV is characterized by the absence of cartilage and an association with pleuropulmonary blastoma [7]. Bailey., *et al*. proposed that CCAM is caused by a primary disorder of alveolar development [8]. Prenatal diagnosis of CCAM is possible with ultrasound and confirmed postnatally with radiology and MRI studies. The treatment of choice is surgical excision to avoid complications such as recurrent infections, pneumothorax, and the risk of malignant transformation. Medical treatment with antibiotics for pneumonia-associated cases. Supportive care including oxygen and, when indicated, mechanical ventilation [9].

Another report from Saudi Arabia described antenatally suspicious CPAM in a baby who was clinically well at birth but represented febrile in the neonatal period and had cystic changes on imaging. CT revealed a right lower lobe type 2 CCAM. The patient underwent resection with a good result [10]. This highlights the role of postnatal monitoring in suspected CPAM, even for asymptomatic neonates. Unlike this case, the case we observed rapidly progressed and occurred with bilateral involvement, multiple pneumothoraxes, pleural effusion, and MRSA sepsis.

Conclusion

CCAM is a rare congenital lung malformation that may present with signs and symptoms suggestive of neonatal pneumonia or sepsis, occasionally resulting in delayed diagnosis. This case brings attention to the value of early imaging in the setting of failed conventional therapy and the role of multidisciplinary care. Definitive treatment is surgical excision, especially in uncomplicated cases.

Patients' Consent

Informed consent was obtained from all patients.

Conflict of Interest

None.

Bibliography

1. Rahman Sirageldin MK Abdel., *et al*. "Congenital cystic adenamatoid malformation: A case report". *Sudanese Journal of Paediatrics* 13.2 (2013): 57-60.

2. Ansari Mohammed Junaid., *et al*. "Congenital cystic adenomatoid malformation: case report". *JMSCR* 4.3 (2016): 9729-9732.

3. Walvir Nazia Manzoor., *et al*. "A complete spectrum of congenital cystic adenomatoid malformation of the lung deceptive clinical presentations and histological surprises; a single institutional study from a tertiary care hospital in North India". *Lung India* 40.5 (2023): 423-428.

4. Laanait Radia., *et al*. "Congenital cystic adenomatoid malformation in a toddler: Unusual presentation with pleural effusion". *Radiology Case Reports* 19.11 (2024): 5013-5017.

5. Gosain Ratna and Rohini Motwani. "Congenital cystic adenomatoid malformation of the lung: a case report with embryological review". *Anatomy and Cell Biology* 55.2 (2022): 264-268.

6. Laberge JM., *et al*. "Outcome of the prenatally diagnosed congenital cystic adenomatoid lung malformation: a Canadian experience". *Fetal Diagnosis and Therapy* 16.3 (2001): 178-186.

7. Salerno Monica., *et al*. "Congenital cystic adenomatoid malformation (CCAM) type II: A rare case of sudden infant death". *Children* 9.12 (2022): 1830.

8. Kuga Takayuki., *et al*. "Congenital cystic adenomatoid malformation of the lung with an esophageal cyst: report of a case". *Journal of Pediatric Surgery* 36.6 (2001): E4.

9. Green Thomas P and D Jonathan. "Finder, Congenital Disorders of the Lung". Richard E. Behrman, Robert Kliegman, hal B-Jenson. Nelson Textbook of Paediatrics 17th edition. New Delhi, Elsevier 1424 (2004).

10. Alshamiri Kamal M and Hatem B Abbod. "Congenital cystic adenomatoid malformation". *International Journal of Pediatrics and Adolescent Medicine* 4.4 (2017): 159-160.

Volume 14 Issue 8 August 2025