Case Report

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Congenital Pulmonary Airway Malformation in a Child: A Case Report

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ABSTRACT

Background: Congenital Pulmonary Airway Malformation (CPAM) is a rare developmental lung anomaly that may present with respiratory distress in neonates or recurrent respiratory tract infections in older children.This case report aims to describe a case of delayed diagnosis of Congenital Pulmonary Airway Malformation (CPAM) in a 4-year-old child presenting with recurrent pneumonia and to highlight the importance of clinical suspicion, imaging and timely surgical intervention in improving outcomes, particularly in resource-limited settings.

Case Presentation: We describe a 4-year-old girl with recurrent chest infections, growth failure, and chest deformity. Clinical evaluation and imaging confirmed CPAM of the right lung. She underwentsurgical lobectomy with favorable recovery.

Conclusion: CPAM should be suspected in children with recurrent pneumonia unresponsive to conventional therapy. Early surgical intervention prevents recurrent infections and long-term complications.

KEY WORDS

Congenital pulmonary airway malformation; Delayed Diagnosis; Lobectomy; Pediatric lung anomaly; Recurrent pneumonia.

INTRODUCTION

Congenital Pulmonary Airway Malformation (CPAM) previously known as congenital cystic adenomatoid malformation, is the most common congenital lung anomaly, with an estimated incidence of 1 in 25,000–35,000 live births.[1] Most cases are detected prenatally via ultrasonography, however, delayed diagnosis beyond infancy is not uncommon, particularly in resource-limited settings.

Children presenting later often exhibit recurrent respiratory infections, chronic cough, failure to thrive, or chest deformities. Symptomatic CPAM typically necessitates surgical resection due to the risk of recurrent infections and potential malignant transformation.[2-5] This case highlights a delayed diagnosis of CPAM presented with repeated history of respiratory infection in a 4-year-old child, emphasizing the need for clinical suspicion and timely surgical management.

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CASE PRESENTATION

A 4-year-old female, the fourth child of non-consanguineous parents from a low socioeconomic background, admitted in Pediatrics Department of Chittagong Medical College Hospital on 3rd July 2025, presented with persistent cough for two months and intermittent fever. She had a history of repeated respiratory infections requiring four hospital admissions, previously treated as bronchopneumonia. Despite no known tuberculosis contact, she was clinically suspected of pulmonary tuberculosis, Mantoux test and GeneXpert on gastric lavage were negative.

- **Growth Parameters:** Severe wasting and moderate stunting.
- **History:** Born by LSCS due to maternal gestational diabetes, fully immunized per EPI schedule, no tuberculosis exposure.
- **Examination:** Ill-looking, cachectic, pale, digital clubbing (Figure 3) right-sided chest bulging, trachea shifted to left, apex beat displaced to left 5th ICS lateral to MCL, percussion revealed dullness, auscultation showed absent breath sounds over the affected area, mild hepatomegaly.

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Investigations

- **Chest X-ray (PA view):** Multiple air-filled cystic lesions in the right lower lung zone with mild mediastinal shift to the left (Figure 1).
- **High-resolution CT thorax:** Coronal images showed Multiple thin-walled, variable-sized cystic lesions involving the right lung, consistent with CPAM (Figures 2).
- **Routine labs:** Normal except mild anemia.
- MT- Negative.

Treatment

The patient was initially stabilized with nutritional support, broad-spectrum antibiotics for suspected secondary infection and physiotherapy. A multidisciplinary team recommended surgery. She underwent right lower lobectomy without complications.

Outcome and Follow-up

The postoperative course was uneventful. The patient showed gradual clinical improvement with resolution of respiratory symptoms. At follow-up, she gained weight, had no recurrent infections and was more active compared with preoperative status.

Figure 1 Chest X-ray showing multiple air-filled cystic lesions in the right lower lung zone with mild mediastinal shift to the left, consistent with adenomatoid malformation of the lung (CPAM).

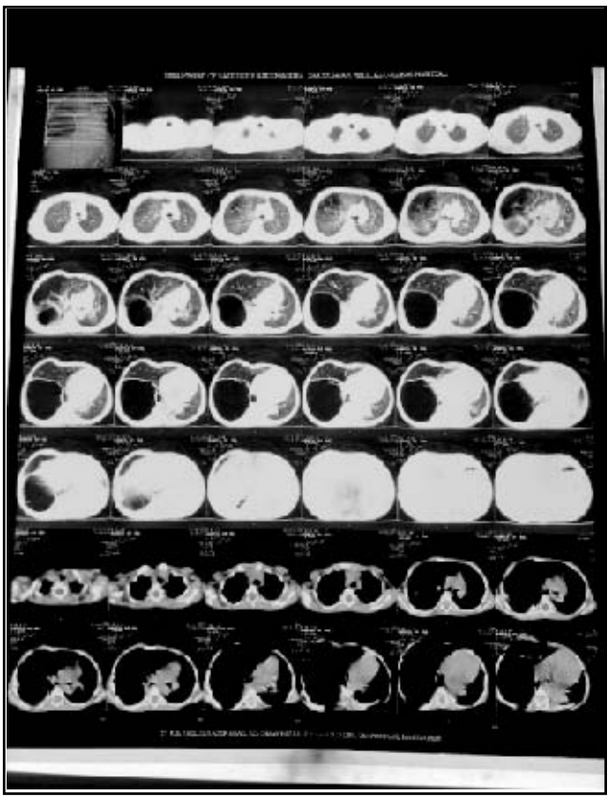

Figure 2 HRCT thorax (Coronal reconstruction) demonstrating Multiple thin-walled, variable-sized cystic lesions involving the right lung, with a few showing air–fluid levels Mild mediastinal shift to the left, finding consistent with Congenital Pulmonary Airway Malformation (CPAM) likely Type I. Features also suggest superimposed infection within some cysts

Figure 3 A photograph of the patient showing clubbing of toe

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DISCUSSION

This case demonstrates a delayed presentation of CPAM in a resource-limited setting. The child had recurrent pneumonia since infancy, repeatedly misdiagnosed as bronchopneumonia. Recognition was delayed until advanced imaging revealed CPAM.

In children, recurrent or persistent pneumonia localized to the same lung region should raise suspicion for congenital lung malformations. HRCT remains the diagnostic gold standard.

Surgical resection (Lobectomy or segmentectomy) is the treatment of choice for symptomatic CPAM. Recent studies advocate early surgical intervention even in asymptomatic cases to prevent recurrent infections and reduce the risk of malignant transformation, including pleuropulmonary blastoma.[2-5] Our patient's clinical improvement following lobectomy supports this approach. Clinician awareness and prompt recognition are essential, particularly in settings where delayed diagnosis is more likely.

Patient's Perspective

"We were worried as our daughter was always sick and not growing like other children. After the operation, she is healthier, eating better and has not fallen sick again. We are very relieved and grateful."

CONCLUSION

- Recurrent pneumonia in the same lung region in children warrants evaluation for congenital lung malformations.
- HRCT chest is essential for accurate diagnosis and surgical planning.
- Early surgical intervention prevents recurrent infections, improves growth and reduces malignancy risk.
- Clinician awareness is crucial, particularly in low-resource settings.

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DISCLOSURE

All of the authors declared no competing interests.

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