

# Pyopneumothorax with Stocker type III congenital cystic adenomatoid malformation in a 5-month-old infant

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## ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) is a rare, developmental, hamartomatous abnormality of the lung characterized by a cessation of normal bronchiolar maturation, resulting in cystic overgrowth of the terminal bronchioles. We report one such case of CCAM in a 5-month-old female infant who was in perfect health until she suffered from spontaneous pyopneumothorax with type III CCAM of the lung and recovered after lobectomy.

**KEY WORDS:** Congenital cystic adenomatoid malformation, lobectomy, pyopneumothorax

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## INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM) is a rare, nonhereditary, developmental abnormality of the lung with incidence of 1 in 25,000 to 1 in 35,000 pregnancies and represents 25% of congenital lung malformations and 95% of congenital lung lesions; males and females are equally affected.<sup>[1,2]</sup> It is characterized by cystic overgrowth within the lung that stems from abnormal embryogenesis at the expense of normal alveoli.<sup>[3]</sup> CCAM was classified into three subtypes in 1977, and later it was reclassified into five types by Stocker in 2002.<sup>[4,5]</sup> It is usually discovered in neonates because of respiratory distress and may occasionally be discovered in older children or adults who have repeated chest infections.<sup>[2,6]</sup> We present a case of a 5-month-old infant suffering from pyopneumothorax with type III CCAM who underwent lobectomy.

## CASE REPORT

The 5-month-old previously normal, well-growing

[weight = 5.3 kg, length = 60 cm, weight/height = -1 SD to -2 SD as per the World Health Organization (WHO) growth chart, head circumference = 40.5 cm] immunized female child presented with fever since 6 days followed by fast breathing for 2 days. Roentgenogram of the patient's chest [Figure 1] showed left-sided large pneumothorax with shift of the mediastinum on the right and was referred to our center for further management.

On admission, she was eutermic, irritable [heart rate = 168/min, respiratory rate = 68 bpm, blood pressure = 98/60 mmHg, peripheral capillary oxygen saturation (SpO<sub>2</sub>) = 88% on room air] with reduced chest expansion on the left side. Her breath sounds were decreased on the left side with a hyperresonant note. She was started on high-flow oxygen and was shifted to the pediatric intensive care unit (PICU). On admission to PICU, the left-sided intercostal drainage tube was placed [Figure 2], which drained gush of air followed by thick, white-colored pleural fluid that was exudative. The

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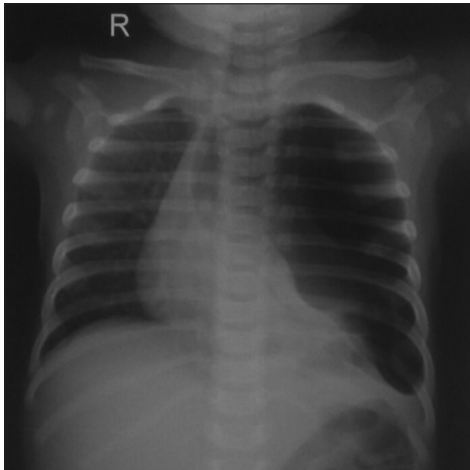
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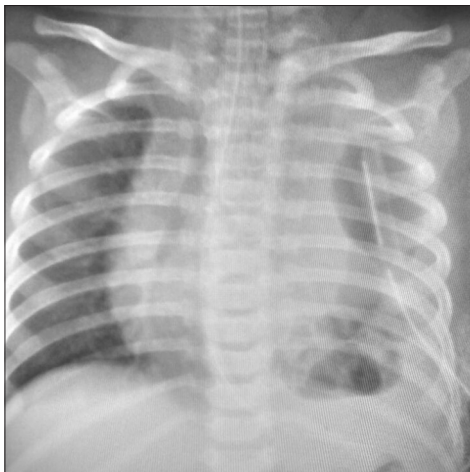
complete blood picture of the patient showed hemoglobin level = 10 gm%, total leukocyte count (TLC) = 37,400/mm<sup>3</sup> (P 80% and L 15%), and C-reactive protein (CRP) level was positive (18.3 mg/dL), human immunodeficiency virus enzyme-linked immunosorbent assay (HIV ELISA) was negative. Ceftriaxone injection, Ampiclox, and Clindamycin were started, along with other supportive measures.

Gradually, her respiratory distress was alleviated over a period of next 24 h but roentgenogram [Figure 3] of the chest showed incomplete expansion of the left side with multiple cavitations even after 5 days, hence computed tomography (CT) scan of the chest [Figure 4] was done that showed collapse consolidation of the left lung with multiple cavitations and some cavity in the lower lobe communicating with pleural cavity and suspected to be CCAM. Left thoracotomy with decortication with lower lobe lobectomy was done in view of suspected CCAM. Gross surgical sample [Figure 5] revealed a wedge of lung measuring 10 cm × 8 cm × 5 cm. Pleural surface was

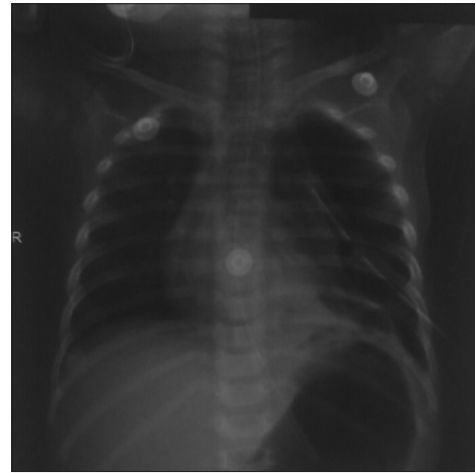
congested and cut surface was firm with no or little normal looking lung parenchyma. Microscopic examination [Figure 6] of the patient showed signs of pleuritis with underlying suppurative inflammation of the lung indicative of abscess. Adjacent lung tissue showed vague lobulated architecture with interlobular fibrosis, definite increase in the number of bronchiolar structures, and small alveolar spaces lined by plump cuboidal epithelium. Overall symptoms in correlation with clinical and radiological findings were consistent with type III CCAM.



**Figure 1:** Roentgenogram of the chest: Large pneumothorax with collapse of the left lung and a normal right lung



**Figure 3:** Roentgenogram of the chest: After 5 days of ICD tube insertion with multiple cavitations



**Figure 2:** Roentgenogram of the chest: Post-ICD insertion with incomplete expansion of the left lung



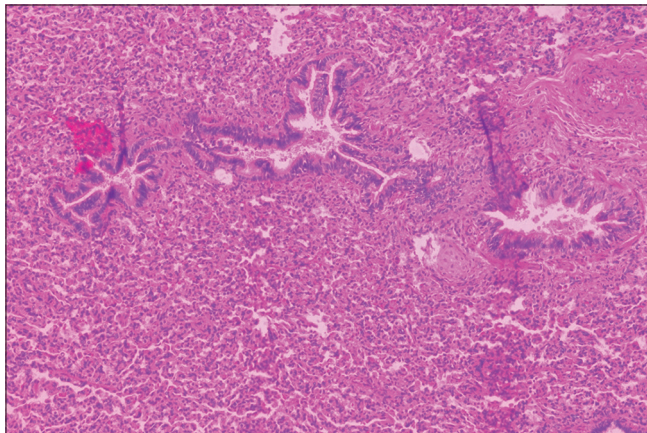
**Figure 4:** CT of the chest: Collapse-consolidation of the left lung with multiple cavitations, with some cavity in the lower lobe communicating with pleural cavity. Moderate left pleural effusion with multiple air foci and drain tube *in situ*

Later, she became hemodynamically stable with improved air entry to the left side of the chest. Roentgenogram of the chest [Figure 7] showed resolution of the pneumothorax following which the implantable cardioverter

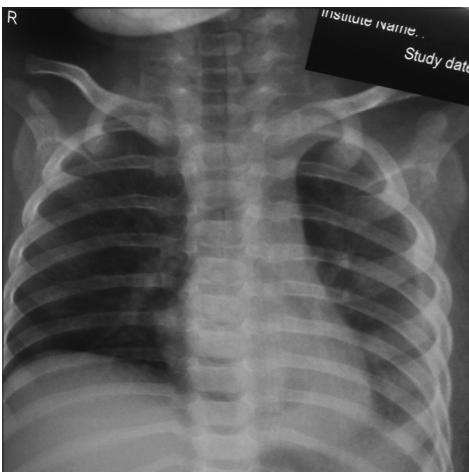
defibrillator (ICD) tube was removed. Later blood and pleural fluid cultures were sterile; hence, the patient was started on oral antibiotics and was discharged after pneumococcal and flu vaccination. On follow-up after 4 weeks, her growth and respiratory functions were found to be normal.



**Figure 5:** Postlobectomy gross appearance of the cavitations and infected pleura



**Figure 6:** Solid type CCAM with increased bronchial structures and cysts are not discernible (type III)



**Figure 7:** Postlobectomy roentgenogram of the chest

## DISCUSSION

CCAM is an unusual congenital, unusual, congenital developmental, nonhereditary anomaly of the lung characterized by proliferation of the terminal bronchiole like interconnecting structures with the formation of cysts of varying sizes. This lesion was first described by Chi'n Tang in 1949.<sup>[7]</sup> It is usually unilateral and restricted to a single lobe but can involve one or both the lungs.<sup>[2,8]</sup> Most postnatally identified cases of CCAM occurring in the newborn period present with respiratory distress (80%) secondary to mass effect and pulmonary compression or hypoplasia and severe cases present with air trapping. Beyond the neonatal period, cases of CCAM present with recurrent or persistent pneumonia, pneumothorax, rarely hemopneumothorax or pleural effusion, and pneumatocele.<sup>[9,10]</sup> Roentgenogram of the chest, CT scan, and magnetic resonance imaging (MRI) are helpful in diagnosis.<sup>[11]</sup> In a case series of seven cases, four were diagnosed to be having CCAM over 1 year where the presenting features were either persistent or recurrent respiratory infections but investigations revealed a congenital malformation of lungs.<sup>[9]</sup>

Stocker initially classified CCAM into three different categories (types I-III) based on clinical and pathological features and later in 2002, he modified the classification by adding two more types (types 0 and IV) [renamed the lesion as congenital pulmonary airway malformation (CPAM)].<sup>[2,4,5]</sup>

Type 0 is of tracheobronchial origin, has solid appearance with small and firm lungs, and microscopically shows bronchiolar type airway with cartilage, smooth muscle, and glands separated by abundant mesenchymal tissues.

Type I constitutes of 50-70% and it is composed of single or multiple large cysts (>2 cm) lined by flattened, cuboidal cells frequently producing mediastinal herniation.

Type II constitutes of 15-30% and it is composed of multiple small cysts (<2 cm) lined by ciliated cuboidal to columnar epithelium, its structure resembles that of respiratory bronchioles, and distended alveoli are present between the epithelium lined cyst. This type is usually associated with other systemic anomalies.

Type III constitutes of 5-10% and it is usually composed of large bulky noncystic lesions producing mediastinal shift. Bronchial-like structures are lined by ciliated cuboidal epithelium and separated by masses of alveolus-sized



structures by nonciliated cuboidal epithelium. Our patient was histopathologically diagnosed with type III CCAM.

Type IV is of distal acinar origin, has peripheral cystic type, and is composed of large cysts (>10 cm) lined by flattened epithelium and resting on loose mesenchymal tissue.<sup>[10]</sup>

The new classification is not much used as type 0 lesions are very difficult to differentiate from bronchogenic cyst and the similarities between type IV cyst and pleura pulmonary blastoma may cause problem(s) in diagnosis.<sup>[11]</sup>

CCAM is treated with lobectomy in symptomatic cases.<sup>[1,11]</sup> Treatment can be postponed if the patient is asymptomatic and the cyst is resolving. Long-term outcome of this treatment is very good, after the treatment the children lead normal lives and their lung volume gets decreased.<sup>[12]</sup>

Prognosis also depends on Stocker type, with type I lesions carrying an overall good prognosis. In type II lesion, it is the associated anomalies that determine the prognosis. Type III lesions carry bad prognosis and they are usually large with associated cardiovascular compromise. Overall bilateral involvement associated with hydrops and associated congenital anomalies carry poor prognosis.<sup>[8]</sup>

## CONCLUSION

Extra vigilance and alternative diagnosis are required in children when they present with infective chest conditions unusual for their age and in the absence of immunocompromised status. Necessary imaging studies followed by appropriate interventions help in the diagnosis of congenital lung malformations.

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## Conflicts of interest

There are no conflicts of interest.

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