

## Case Report

# Congenital Pulmonary Airway Malformation (Cpam). A Diagnostic Dilemma!

Anand M<sup>1</sup>, Rahul Singh R<sup>2\*</sup>, and A Bhaskaran<sup>2</sup>

<sup>1</sup>Department of Pediatric Surgery, Sri Devaraj Urs Medical College, India

<sup>2</sup>Department of General Surgery, Sri Devaraj Urs Medical College, India

## \*Corresponding author

Rahul Singh R, Sri Devaraj Urs Medical College, Tamaka, Kolar, Karnataka, India

Submitted: 28 June 2017

Accepted: 31 August 2017

Published: 02 September 2017

Copyright © 2017 Rahul Singh et al.

ISSN: 2373-9819

OPEN ACCESS

## Keywords

- Congenital pulmonary airway malformation
- Diaphragm
- Lung parenchyma

## Abstract

Congenital pulmonary airway malformation (CPAM) of the lung is a rare lesion that typically manifests as neonatal respiratory distress, secondary to progressive expansion of the affected lung. Three distinct types have been described, based on the size of the cysts and the microscopic appearance. Type II lesions may carry a poor prognosis because of associated renal and cardiac anomalies. Malformations can involve the lung parenchyma, airways, pulmonary or systemic arteries, pulmonary or systemic veins, fistulas with the gastrointestinal tract, or defects in the diaphragm, and various combinations of these. We present a case of one month old baby with CPAM, which led to initial diagnostic difficulty, but was later managed appropriately, at this institution, located in a backward rural area, of Karnataka.

## INTRODUCTION

Congenital pulmonary airway malformation (CPAM) is a rare congenital abnormality that affects mostly the newborn and presents with acute progressive, respiratory distress [1]. The newer terminology, congenital pulmonary airway malformation (CPAM) has largely replaced congenital cystic adenomatoid malformation [2]. CPAM is a multi cystic lung mass, resulting from a proliferation of terminal bronchiolar structures, with an associated suppression of alveolar growth. The presenting features of CPAM resemble a host of other conditions, such as congenital lobar emphysema, bronchogenic cyst, pulmonary sequestration, pleuropulmonary blastoma, intra parenchymal lymphangioma, for which it can be mistaken [3].

## CASE REPORT

A month old male baby, presented with history of cough - 5 days duration, refusal of feeds and progressive respiratory distress of one day duration. On admission, baby was found to be tachypnoeic, with a respiratory rate of 74 cycles per minute, cyanosis and an O<sub>2</sub> saturation of 65 % on room air. A chest radiograph was taken and was interpreted as Right pneumothorax (Figure 1) by the duty general surgeon and an intercostals drainage tube was placed. However, there was no clinical improvement and follow up radiograph showed, no resolution of hyperlucency in the right hemi thorax (Figure 2).

A pediatric surgeon opinion was sought, who advised a CT scan of the chest, which showed a large hyperlucent area involving right upper lobe with few thin internal septae, suggestive of over inflated lobe of right lung (Figure 3). Intercostal drain tube was seen through right 5th intercostal space with its tip abutting



**Figure 1** Initial chest radiograph showing large hyperlucency in right hemi thorax, crossing the midline and extending to involve partly left hemi thorax. The mediastinum including trachea and cardia have been shifted to the left.



**Figure 2** Follow up chest radiograph after placement of intercostal drainage (ICD) tube at 5th intercostal space on right side. Note that there is no interval resolution of hyperlucency in the right hemithorax, suggesting that the condition could be due to pulmonary pathology.

posterior chest wall. The CT findings suggested, the possibility of congenital lobar emphysema/over inflation involving right upper lobe.

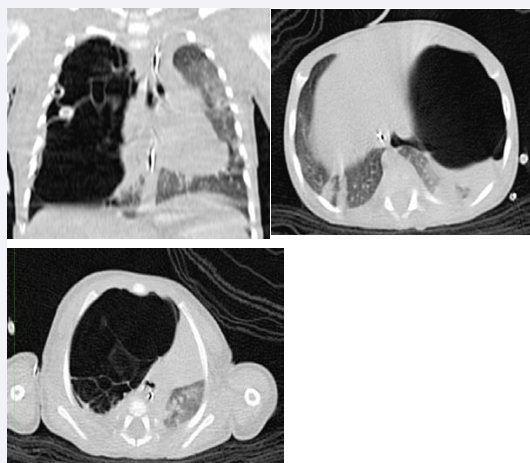
Figure 3 Coronal and axial sections of CT thorax showing large hyperlucent area involving right upper lobe with thin internal septations. Note significant mass effect in the form of mediastinal shift to the left and compressive atelectasis of right middle and lower lobes. There is sub segmental collapse/consolidation of superior and posterior basal segments of left lower lobe with compression of rest of the left lung.

Patient was prepared and taken up for right thoracotomy. Peroperatively, a large air filled lesion without any obvious lung parenchyma was seen, involving the entire upper lobe. The middle and lower lobes appeared normal, though compressed by the mass lesion. Right upper lobectomy was done. Baby was extubated after 24 hours and made a steady progressive recovery with good lung expansion by the 4<sup>th</sup> postoperative day (Figure 4 a,b,c) and (Figure 5). The alert, active baby was discharged on 9<sup>th</sup> post-operative day, in near normal condition. Periodic follow up has been carried since and the baby's clinical condition has remained satisfactory.

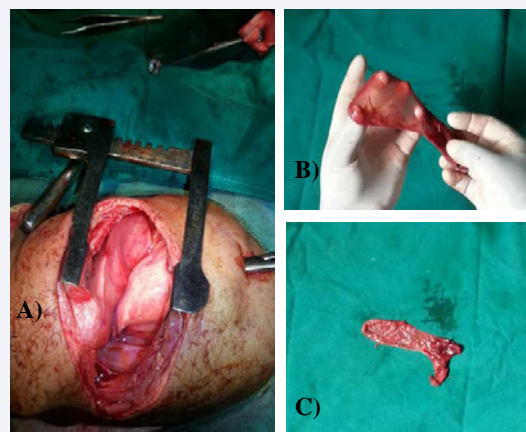
Figure 6 Photomicrograph (hematoxylin-eosin stain) showing cystic spaces lined by cuboidal to columnar type of epithelium showing cilia at some areas. No definitive alveoli/cartilage. There is evidence of foci of fibrosis and macrophages with hemosiderin pigment. Surrounding lung tissue is congested and areas of hemorrhages are noted. Features consistent with congenital adenomatoid malformation.

## DISCUSSION

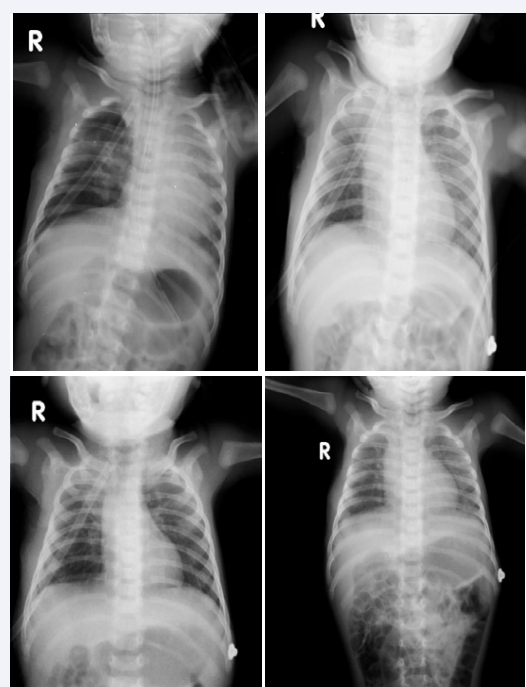
CPAM is a multicystic lung mass resulting from a proliferation of terminal bronchiolar structures with an associated suppression of alveolar growth. It usually presents in immediate neonatal period, with increasing respiratory distress, cyanosis and grunting. Physical examination reveals a prominent ipsilateral chest, decreased breath sounds and hyper resonance on the involved side and distant and shifted heart sounds. Large and severe lesions can produce polyhydramnios and features of fetal



**Figure 3** Coronal and axial sections of CT thorax.



**Figure 4** a: Intraoperative images showing remaining lungs after right upper lobectomy. The cystic lesion was deflated prior to lobectomy. b: Wall of the cystic lesion. c: Deflated cystic lesion.

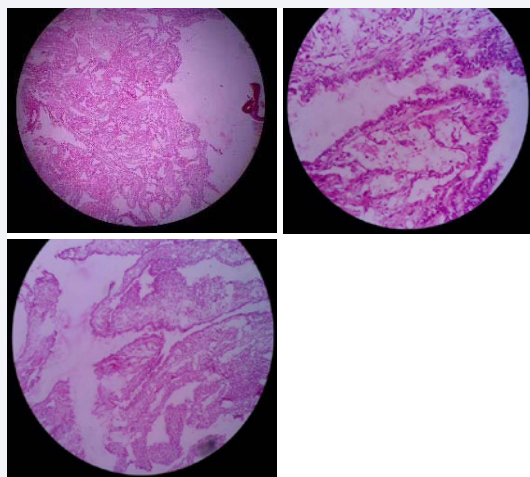


**Figure 5** Serial follow up chest radiographs taken on 1st, 2nd, 3rd and 4th post-operative days successively. Near complete resolution of right lung hyperlucency noted in previous radiographs with progressive lung expansion and normalization of mediastinal shift.

hydrops, due to compression of venacava and impaired cardiac contractility, by pressure effect of the lesion. Rarely, patients may present beyond the neonatal period, with symptoms of respiratory distress. Vomiting, failure to thrive, chest pain, and recurrent pneumonia.

CPAM is classified into three types radiologically and into 5 types histopathological based on modified Stocker classification [3-7].

Type I cystic adenomatoid malformation, the large-cyst type, contains one or more cysts varying from 2 to 10 cm in diameter,



**Figure 6** Histopathology revealed it to be a congenital cystic adenomatoid malformation of the lung.

surrounded by multiple smaller cysts. The larger cysts are lined with ciliated pseudo stratified columnar epithelium (i.e., typical “respiratory” epithelium) and often display a papillary or polypoid appearance because of the presence of elastic tissue beneath the epithelium. A unique finding of the type I lesion is the presence of clusters of mucogenic cells amid the lining of the large cysts or within alveoli adjacent to the cysts. Type I lesions have the appearance of an expanded lobe or portion of a lobe. The lobe may be deformed by different-sized underlying cysts in various degrees of expansion.

The type II lesion, the intermediate cyst type, contains cysts that are simpler and rarely exceed 2 cm in diameter. The cysts, lined by cuboidal to columnar epithelium, resemble dilated terminal and respiratory bronchioles organized in a back-to-back configuration. Mucogenic cells are not seen in the type II lesion. A subset of type II cystic adenomatoid malformation contains, in addition to the bronchiole like structures, strands of skeletal muscle fibers. Type II lesions may be associated with other severe malformations (eg renal agenesis or dysgenesis). In addition, within 15%-25% of extra lobar pulmonary sequestrations, focal areas with microscopic features of cystic adenomatoid malformation type II can be found. Type II lesions are characterized by multiple small uniform cysts that do not exceed 2cm in diameter. The cyst-containing portions of the lesion blend with the adjacent normal lung parenchyma.

Type III cystic adenomatoid malformation, the small-cyst type, this large bulky lesion is seen primarily in newborn or stillborn male infants, whose gestation was associated with significant polyhydramnios. The lesion usually involves an entire lobe and occasionally the entire lung. Type III lesions have a solid appearance representing epithelial-lined alveolar like (“adenomatoid”) structures separated by dilated bronchiole like structures that may be large enough (0.3-0.5 cm) to be visible at gross inspection.

In about 25% cases of CPAM, an associated systemic/pulmonary arterial supply is noted.

## Diagnosis

Antenatal Ultrasound (AU) can usually diagnose CPAM during the 2<sup>nd</sup> trimester, as a solid or cystic mass in the fetal hemithorax. It can also detect findings consistent of Fetal Hydrops (FH), such as fetal ascites, fetal anasarca, placental edema, polyhydramnios, as well as other associated congenital anomalies. Type 1 lesions may exhibit single, multi septate or multiple large cysts, Type 2 lesions, demonstrate multiple small cysts of uniform size, while Type 3 lesions result in solid, usually bulky, echogenic lesions of the fetal thorax. Serial US examinations are helpful in the follow up of fetuses with Fetal Hydrops. Improvements of FH, resolution of polyhydramnios, and even decrease in the size of lung lesions, have been described.

Antenatal Ultrasound also helps in differentiating CPAM, from other congenital lesions of lung such as Congenital Diaphragmatic Hernia, Pulmonary sequestration, Congenital Lobar Emphysema, bronchogenic cyst etc. Finally it can help in planning prompt surgical intervention after birth, and may allow selection of candidates for intra uterine surgical intervention.

As most of the lesions can be detected antenatally, the management depends on the antenatal severity of the disease. Presence of bilateral disease and hydrops is associated with poor outcome. Prenatally, approximately 10% of cases. Need fetal intervention. CPAM babies with Fetal Hydrops are candidates for fetal therapy. Babies with Microcystic disease where hydrops develops before 32 weeks of gestation are treated with maternal steroids to reverse the hydrops, where as those in which hydrops develops after 32 weeks are treated with Ex utero-intrapartum (EXIT) and neonatal resection, Macrocytic lesions which cause hydrops are treated with Thoraco Amniotic shunts of the dominant cyst.

## Postnatal Diagnosis

**Plain X-Ray chest:** Type 1 lesion; typically show a unilateral air filled, multicystic lesion in the hemithorax with mediastinal displacement, and depression of ipsilateral hemidiaphragm. The abnormally inflated lung may herniate across the midline. The uninvolved ipsilateral and contralateral portions of the lung may be atelectatic or hypoplastic, due to compression.

Type 2 lesions may appear as heterogenous areas of uniform small cysts.

Type 3 lesions, are usually large and homogenous, having the appearance of parenchymal consolidation or a mass, rather than that of a cystic lesion.

**CT scan:** In equivocal cases CT Scans helps not only in diagnosis but also in differentiating it from other neonatal conditions, which present with respiratory distress, such as Diaphragmatic hernia, bronchogenic cyst, congenital lobar emphysema, pulmonary sequestration etc. CT depicts the internal structure of the affected lung, such as the presence of air- or fluid filled cysts. The extent and distribution of disease can be assessed and the amount of mass effect, on the uninvolved lungs and mediastinal structures, evaluated.

Type 1 and 2 lesions, appear as multiple thin walled, rounded air filled cystic areas in the lung. Air fluid levels can be identified and occasionally fluid filled cysts can be seen.

Congenital Diaphragmatic Hernia (CDH), Congenital Lobar Emphysema (CLE), Bronchogenic Cyst, Pulmonary Sequestration is some of the conditions that need to be differentiated from CPAM.

CDH can be excluded by visualization of bowel loops, below the diaphragm. Bronchogenic cysts, usually manifest as mediastinal masses and are rarely seen in the neonatal period. CLE can be distinguished by the absence of internal linear opacities, which are commonly seen in CPAM. Pulmonary sequestration manifests in neonatal period or early infancy, as solid intrathoracic masses in a patient with respiratory distress, mimicking Type 3 CPAM. CT will help in differentiation.

Surgical management of CPAM is indicated in the form of thoracotomy with affected lobectomy, which can be done 3/6 months postnatally, in asymptomatic cases to avoid risk of subsequent respiratory tract infections or malignant transformation.

Postnatally, surgical resection remains the mainstay of treatment in symptomatic patients with CPAM [8]. In an asymptomatic patient however, conservative management can be tried. However, results are not very encouraging and some patients may finally undergo surgery. Surgical resection in these individuals, has shown lower risk of future complications [8,9].

Errors in differentiating CPAM from pneumatocele/tension pneumothorax can result in incorrect chest tube insertion. This not only results, in worsening of respiratory distress, increased rates for complications such as pneumothorax, hydro pneumothorax, subcutaneous emphysema and infections but also may delay accurate diagnosis [10].

Chest CT is not routinely recommended in all cases of suspected pneumothorax. Atypical radiograph findings and respiratory distress which, either fails to improve/or worsens with chest tube insertion, are indications, that the pathology is not restricted to simple pneumothorax. In these cases, it is important to diagnose other causes, such as large CPAM/CLO [10].

The prognosis of patients with CPAM depends on the size of the lesion, the degree of development of the adjacent uninvolved lung, and the presence or absence of other congenital anomalies. Patients with large lesions that interfere with normal pulmonary development have a poor prognosis. Severe pulmonary hypoplasia and gross fetal hydrops, have a uniformly fatal outcome. Without prompt antenatal intervention, these infants may be stillborn or may die in the immediate neonatal period. Neonates can tolerate lobectomy well [11]. The residual lung tissue is able to re expand postoperatively, resulting in near-normal clinical function. Type II lesions may carry a poor prognosis because of associated renal and cardiac anomalies [12].

## CONCLUSION

CCAM is a potentially life threatening and reversible, rare congenital disorder which frequently presents with respiratory distress [4,12]. This case is presented to highlight the various clinical conditions which can mimic each other and lead to diagnostic difficulties and incorrect management.

The present case was misdiagnosed for a pneumothorax and hence treated with intercostals drain placement, which could not serve the conceived purpose. Awareness about other conditions, such as congenital lobar emphysema, pneumatocele, pleuropulmonary blastoma, intra parenchymal lymphangioma, helps in proper investigation, which in turn leads to correct diagnosis and management.

## REFERENCES

1. Rosado-de-Christenson ML, Stocker JT. Congenital cystic adenomatoid malformation. *Radiographics*. 1991; 11: 865-886.
2. Paterson A. Imaging evaluation of congenital lung abnormalities in infants and children. *Radiol Clin North Am*. 2005; 43: 303-323.
3. Donoghue V, Watson TA, Garcia-Pena P, Owens CM. The neonatal and paediatric chest. In Adam A, Dixon AK, Gillar JH, Schaefer-Prokop CM editors. *Grainger & Allison's Diagnostic radiology*. 6th edition Chapter 76. Churchill Livingstone Elsevier. 2014; 1778-803e02.
4. Walker J, Cudmore RE. Respiratory problems and cystic adenomatoid malformation of lung (hener). *Arch Dis Child*. 1990; 65: 649-650.
5. Jana M, Gupta AK. Radiologic evolution of congenital cystic adenomatoid malformation in a neonate. *Indian J Pediatr*. 2010; 77: 212-234
6. Biyyam DR, Chapman T, Ferguson MR, Deutsch G, Dighe MK. Congenital lung abnormalities: embryologic features, prenatal diagnosis, and postnatal radiologic-pathologic correlation. *Radiographics*. 2010; 30:1721-1738.
7. Nadeem M, Elnazir B, Greally P. Congenital pulmonary malformation in children. *Scientifica*. 2012: 209896.
8. Marshall KW, Blane CE, Teitelbaum DH, van Leeuwen K. Congenital cystic adenomatoid malformation: Impact of prenatal diagnosis and changing strategies in the treatment of asymptomatic patient. *AJR Am J Roentol*. 2000; 175: 1551-1554.
9. Lakhoo. Management of congenital cystic adenomatous malformations of the lung. *Arch Dis Child Fetal Neonatal Ed*. 2009; 94: 73-76.
10. Ceran S, Altuntas B, Sunam GS, Bulut I. Congenital lobar emphysema: is surgery routinely necessary? *Afr J Paediatr Surg*. 2010; 7: 36-37.
11. Prabhu SM, Choudhury SR, Solanki RS, Shetty GS, Agarwala S. Inadvertent chest tube insertion in congenital cystic adenomatoid malformation and congenital lobar emphysema-highlighting an important problem. *Indian J Radiol Imaging*. 2013; 23: 8-14.
12. Stocker, Kagan-Hallet K. Extralobar pulmonary sequestration: analysis of 15 cases. *Am J Clin Pathol*. 1979; 72: 917-925.

### Cite this article

Anand M, Rahul Singh R, Bhaskaran A (2017) Congenital Pulmonary Airway Malformation (Cpam). A Diagnostic Dilemma! *JSM Clin Case Rep* 5(3): 1135.