Case Report

Congenital Pulmonary Airway Malformation-Two Case Reports and Diagnosis Challenges in a Resource Limited Setting

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ABSTRACT

Congenital Pulmonary Airway Malformation (CPAM) is a rare abnormality of pulmonary airway malformation and may remain undiagnosed until it is discovered as an incidental finding later in life. Reported here are two cases of CPAM, a seven-month-old infant and a two-year-old toddler. A seven-month-old ex-premature male infant presented with recurrent pneumonia and failure to thrive. He had an unresolving consolidation on chest radiograph and was eventually treated as pulmonary tuberculosis with no response. Computerised tomography scan (CT-scan) chest revealed bilateral CPAM of lungs. A 2-year-old female toddler presented to the University Teaching Hospital, Department of Paediatrics & Child Health, as a referral from a second level hospital with a long-standing history of recurrent symptoms and signs of pneumonia with failure to thrive. She was commenced on antituberculous treatment with no improvement. A CT-scan of the chest revealed bilateral CPAM of lungs. These two cases highlight clinical, diagnostic and treatment challenges in children with CPAM in a resource limited setting like Zambia.

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INTRODUCTION

CPAM is a rare lung congenital abnormality but it remains the most common malformation of lung development[1]. Its incidence is 1 in 25 000 to 35 000 pregnancies with a prevalence of 9 per 100 000 live births[2]. The exact cause is not well established. One theory described maladjustment in pseudo glandular phase of lung development before 16 weeks gestation which may be caused by cessation of bronchial maturation along with overgrowth of mesenchymal elements which give rise to multiple cysts at the terminal bronchioles with various sizes and locations[3].The clinical manifestations are respiratory distress and recurrent chest infections[4].The diagnosis of CPAM can be made early in utero by prenatal ultrasonography and postnatally by imaging radiology and MRI[5]. CPAM is often misdiagnosed or diagnosis is usually made late[6].

Key words: *CPAM, Infant and Toddler, Clinical Profiles, Zambia*

We report two cases of an early and a later presentation of CPAM in a seven month old male infant and a two year old female toddler respectively. They both received several courses of antibiotics and antituberculous therapy (ATT) before the diagnosis of CPAM was made by CT imaging. The two cases will raise clinician's awareness on CPAM in young children presenting with recurrent pneumonia and failure to thrive in the face of obvious and common conditions like pulmonary tuberculosis in a resource limited setting.

CASE PRESENTATION

Case Report 1

A two month old male infant presented to the emergency paediatric admissions ward with history of fever, cough and excessive crying and was breast feeding well. He was a twin delivery at seven months of gestation by Caesarean Section, weighed 1.580 kg and the other twin was a still birth. He was admitted to the neonatal intensive care unit for four days and discharged home and remained well for the rest of the neonatal period.

On clinical examination he was found to be tachypnoeic with subcostal recession and the chest was clinically clear. The axillary temperature was 37.1°C. The infant was thriving and weighed 3.2kg at 2 months. Other systems were essentially normal, and a diagnosis of pneumonia was made, and the infant was commenced on first line antibiotics of crystalline penicillin and gentamicin with oxygen administered by nasal catheter. Chest radiography showed infiltrations on the right lung and the cardiac silhouette was normal. The infant remained tachypnoeic for the first one week of admission and later was weaned off oxygen with saturation of 95% and was discharged home on day 12 post admission.

The infant was readmitted five months later at the age of 7 months and weighed 3.7 kg (corrected age, 5 months) with a history of fever for three days and the mother was concerned about the poor weight gain. Past medical history revealed that the infant was admitted at a private hospital for pneumonia on two occasions and two more admissions at a General Hospital within Lusaka city for which he needed imipenem as he was not responding to the second line antibiotics of cephalosporins. Due to the recurrent episodes of pneumonia, the infant was commenced on ATT from the age of about four months and at the time of the second admission to University Teaching Hospital the infant was on ATT for three months with no improvement. A Chest CT-scan revealed bilateral multiple cysts, CPAM. The paediatric surgeons were consulted and the infant died before they could operate on him.

Table 1: Evolution of symptoms and management of an infant with CPAM

Since birth	Asymptomatic
2 months old	Admitted at First Level Hospital with Respiratory distress: Treated for pneumonia
4 months old	Admitted at First Level Hospital with Respiratory distress: Treated for PTB in view of recurrent pneumonia
5 months old	Admitted at First Level Hospital with Recurrent cough, treated for pneumonia with PTB
7 months	Admitted at University Teaching Hospital with Respiratory distress fever, FTT on ATT **Investigations** Haemoglobin-11.3g/dl, WBC-5.0:lymphocytes69.9%,granulocytes-22.4%,platelets-317,HCT-32.8%,MCV-84,MCH-29.0pg,MCHC-34.3g/dl HIV- negative **Chest Xray:** Infiltrates in the right lung, normal cardiac silhouette **CT-scan chest:** Bilateral cystic adenoid of the lungs CT images shown below in figure 1a,1b

Case Report 2

A two year old female toddler presented to UTH, department of paediatrics and child health with a longstanding history of recurrent pneumonia. She was born at term with a weight of 2.6kg and her neonatal history was unremarkable. She was fully immunised. She was well until the age of 10 months when she presented to the local hospital with recurrent pneumonia and treated successfully initially with intravenous antibiotics. At the time of presentation to UTH, she had been treated for unresolving pneumonia with a chronic cough associated with night sweats and difficulties in breathing of one week duration, failure to thrive and congestive cardiac failure. She had fever at the onset of the illness.

On examination, she was fully conscious, in obvious respiratory distress with nasal flaring, respiratory rate of 80/minute. Her temperature was 36.3^{O}C. She had no clubbing and no cyanosis. She was small for age, weight of 7.5kg, had muscle wasting and the weight for age at less than -2Zscore. Respiratory system examination revealed reduced air entry on the left side with bilateral crepitations, cardiovascular system tachycardia with heart rate of 120/min and abdominal examination had a tender liver 6 to 7 cm with a reducible umbilical hernia of 5cm x 6 cm. A diagnosis of congestive cardiac failure with superimposed bacterial pneumonia possibly due to corpulmonale was made and she was commenced on Cefotaxime at 375mg IV QID, Lasix 7.5 mg IV OD, Spironolactone 12.5mg PO OD and Folate 5 mg PO OD

After one week of no response to treatment given, the patient was commenced on ATT and an urgent chest CT-scan was ordered which revealed small multiple cystic lesions of the left lung - CPAM. Paediatric Surgeons were consulted but could not intervene as the patient was graded ASA 4. Thepatient remained oxygen dependent and died after 4 weeks post admission.

Table 2: Evolution of symptoms and management of a Toddler with CPAM

Since birth	Asymptomatic
10 months old	Admitted at First Level Hospital with Respiratory distress: Treated for pneumonia
13 months old	Admitted at First Level Hospital with Respiratory distress: Treated for pneumonia
13months to 2 years	Admitted at First Level Hospital with Recurrent cough with respiratory distress and FTT: Treated for pneumonia
2 years onwards	Admitted at General Hospital with Cough, fever, respiratory distress, FTT, night sweats : Treated for PTB
	Admitted at University Teaching Hospital with Respiratory distress: **Investigations** Haemoglobin-8.7g/dl:WBC -11500/Ul:Lymphocytes62.1%: Polymorphs-31.7%), Platelets-378 000/ul, HCT-28.4%: MCV-89fl, MCH-27.8pg, MCHC-31.1g/dl Urea & Electrolyte: Normal: Blood culture: No growth: HIV - Negative **Chest Xray:** Homogeneous opacity entire left lung field and heterogenous opacities in the right lung **Echo**: Situssolitus, Interventricular septum (IVS) intact, Puff of Tricuspid Regurgitation with pressure of 35 mmHg Dilated RA/RV, Dilated PA, No PE, Good contractility, EF 62%, Conclusion-Pulmonary Hypertension due to CPAM **CT-scan chest:** Small multiple cystic lesions of the left lung, left pleural thickening is present, No pleural effusion Infiltrations in the right lung Conclusion: findings suggestive of CPAM of the left lung and pneumonia in the right lung CT images shown below in figure 2a,2b

Figure 1a: Thoracic CT showing cystic lesions in the lung of the infant

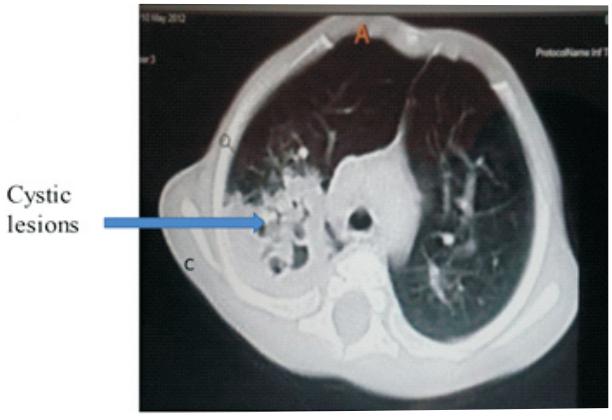

Figure 1b: Thoracic CT scan showing a right upper and middle lobe and left upper lobe CPAM in a 7-month -old infant. The most prominent features are cystic and solid areas in the right lung

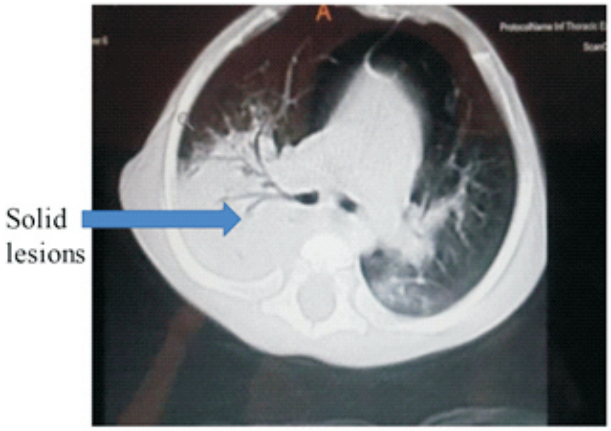

Figure 2a: Chest CT showing multiple cysts in the left lung in the toddler

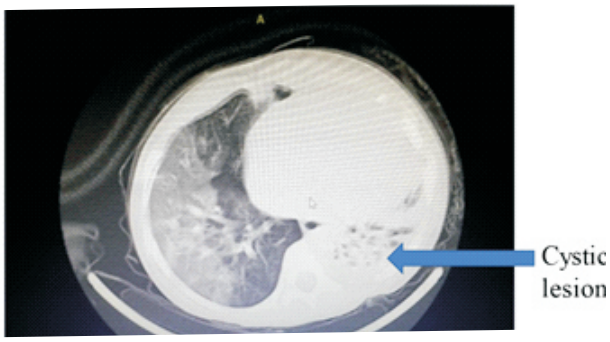

Figure 2b: Chest CT showing multiple cysts with hyperlucency of the left lung in the toddler

DISCUSSION

CPAM is a foetal pulmonary developmental abnormality caused by airway dysgenes is characterised by cystic or adenomatous lesions in the terminal bronchus[6]. Respiratory distress is the commonest presentation of symptomatic CPAM in the neonatal period and respiratory chest infections is common in infancy and childhood[7].Postnatally CPAM is generally asymptomatic and may be complicated and manifests as fever, cough or respiratory distress related to emphysema, pulmonary hypoplasia or respiratory infection[2]. These symptoms are common before the age of 2 years in about 80% to 85% cases[1].Our patients had CPAM which was symptomatic in infancy period from the age of 2 months and presented with recurrent pneumonia and the toddler had symptoms from the age of 10 months. The two patients presented with recurrent pneumonia which was not resolving despite several courses of antibiotics.

Chest CT-scan remains useful and accurate in differentiating CPAM from other respiratory infections causing recurrent respiratory distress. Therefore, it becomes very useful to understand the radiological appearances to help improve early detection to facilitate early diagnosis and early intervention.

Five main types of CPAM have been described radiologically[6]. Type 0 is a rare formand has severe

presentation with small cysts and is lethal. Type 1 is common, accounts for 60-65% casesand comprises small multiple cysts lined by ciliated pseudostratified or columnar epithelium arising from the bronchus and is associated with excellent prognosis[6].Type 2 occurs in 20% case arising from the terminal bronchus comprising small cysts associated with solid areas with cuboidal or columnar epithelium and is associated with other congenital anomalies such as renal agenesis[6]. Type 3 arise from acinar like tissue and comprise macrocytic cysts associated with poor prognosis[6]. Type 4 contain large cysts and has potential for malignant transformation like pulmonary blastomas, bronchoalveolar carcinoma[6].

Our seven month old infant had CPAM comprising cystic and solid cysts in the right lung (figures 1a, 1b) consistent with type 2 CPAM. The toddler had multiple cysts with hyperlucency of the left lung (figures 2a, 2b) consistent with type 1 CPAM, which has an excellent prognosis but demised due to the development of pulmonary hypertension. Prenatal imaging allows for early detection of the pulmonary tract defects during pregnancy and prenatal ultrasound monitoring allows for prenatal care planning.[4]This is rarely the case in underdeveloped countries where there are fewer experienced practitioners.

CPAM may be associated with renal and cardiac complications[8]. Echocardiography is required in all patients with CPAM to rule out any coexisting cardiac lessions[8].It may also provide evidence of persistent pulmonary hypertension although pulmonary hypertension has not been commonly associated with CPAM[8]. The toddler patient had cardiac findings suggestive of pulmonary hypertension. One possible explanation is the combined hypoxia and hypercarbia as evidenced from the recurrent respiratory distress stimulate pulmonary vasculature and the development of pulmonary hypertension which increase pulmonary vascular resistance and a decrease in systemic vascular resistance with further effects on the heart[8].

The effective management of CPAM largely depend on early (prenatal ultrasound) diagnosis. In resource limited countries like Zambia, where prenatal ultrasound is not routine, diagnosis is likely to be late. Post-natal management of symptomatic CPAM patients involve resection of CPAM in all children to remove the risk of direct complications, such as recurrent infection and pneumothorax[9]. The presence of pneumonia may indicate the need for more extensive pulmonary resection[9].The main stay of management of CPAM is surgery and pulmonary resection during infancy is associated with low mortality rates and prevents late complications such as pulmonary compression, infections and malignancy transformation[10]. The diagnosis of CPAM in our two patients was made late and therefore definitive surgical interventions were delayed. It remains very important for all paediatricians to consider the diagnosis of CPAM especially after several presentations of signs and symptoms of pneumonia before a diagnosis of pulmonary tuberculosis is considered.

CONCLUSION

Paediatricians should be aware of CPAM, a rare but potentially fatal condition in order to make early diagnosis and avoid recurrent use of antibiotics and ATT. Recurrent pneumonia in infancy in the absence of immunodeficiency should warrant Chest CT-scan.

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Conflict of interest

The authors declare that they have no conflict of interest.

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