Case Report

Spontaneous Resolution of a Congenital Multicystic Lung Lesion in a Newborn

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INTRODUCTION

Congenital cystic adenomatous malformation (CCAM) also called congenital pulmonary airway malformation (CPAM) is a rare hamartomatous disorder of variably sized cysts occurring in a range of 1/35,000 live births. CCAM occur sporadically. Their formation is not related to maternal factors such as race, age, or exposures. Some authors have reported a slight male preponderance. The lesions are benign characterized by overgrowth of terminal bronchioles with a reduction in alveoli. CCAM is usually discovered in neonates because of respiratory distress due to pulmonary hypoplasia, mediastinal shift, spontaneous pneumothorax, or pleural effusion. It may occasionally be discovered in older children or adults due to a recurrent chest infection. CCAM may be associated with stillbirth or bronchial atresia. Large lesions may be associated with the development of hydrops fetalis in as many as 40% of cases which is thought to arise from compression of the inferior vena cava.

The mainstay of CCAM treatment is surgical excision that prevents complications such as recurrent chest infections, pneumothorax, and malignancy. Medical treatment includes the use of antibiotics for cases complicated by pneumonia and supportive care ranging from oxygen supplementation to mechanical ventilation when indicated. However, cases of spontaneous resolution of this lesion are rare in the literature; we, therefore, report that of a 21-day-old female term baby managed in a semi-urban tertiary health facility.

CASE REPORT

The index case was a female term neonate delivered to a 35-year-old para 7 by spontaneous vagina delivery at home. She presented to the special care baby unit at 21 days of life with complaints of cough, nasal discharge, and fever of 5 days. Fast breathing associated with refusal to feed of 2 days’ duration. There was no history suggestive of cyanosis, abnormal body movements, perinatal hypoxic insult, or meconium stained liquor at delivery. Antenatal history of the mother was not adversely eventful.

Examination findings include weight of 3.5 kg, length of 51 cm, and occipitofrontal circumference of 35 cm (which were all normal for age). No dysmorphic features were identified. She was febrile (38.6°C), in respiratory distress with flaring of ala nasi, subcostal recession, tachypnoeic with a respiratory rate of 78/min.
There was decreased air entry on the left side of the chest, with widespread crepitation which were more on the left hemithorax. The baby was also tachycardic with a heart rate of 182 beats/min, the apex was shifted to 5th right intercostal space at the right sternal border, and the heart sounds were 1st, 2nd, and 3rd with a gallop. Abdominal findings revealed hepatomegaly of 5 cm below the right costal margin.

A preliminary diagnosis of late-onset neonatal sepsis with severe bronchopneumonia complicated with heart failure was made; the differential diagnoses entertained were acyanotic congenital heart disease, congenital diaphragmatic hernia, and dextrocardia. Investigations done include; Chest X-ray (anteroposterior and lateral) which showed mediastinal shift to the right, multiple oval cystic lesions on the left mid lung zones. There was also a homogeneous opacity seen in the left lower lung obliterating the ipsilateral costophrenic sulci [Figure 1a and b]. An abdominal ultrasound scan showed normally sited solid organs, and their sizes were within normal limits except for hepatomegaly. The diaphragmatic shadow appeared continuous and normal. No sonographic evidence of eventration was seen. Others include random blood sugar of 5.9 mmol/L, pack cells volume-51%, white blood cell-28.0 × 10⁹, neutrophil-66%, lymphocytes-28%, monocytes-6%, platelets-202 × 10⁹/l, retroviral screen was nonreactive, serum EUCr; Na⁺-135 mmol/l, K⁺-5 mmol/l, HCO₃⁻-25 mmol/l, Cl⁻-101 mmol/l, Urea-4.3 mmol/l, Creatinine-46 µmol/l. The treatment offered include nil per oral, intranasal oxygen at 2 l/min, intravenous fluid; pediatric saline at 120 ml/kg/day and antibiotics (cefuroxime and gentamicin).

The baby was referred to a more equipped facility; however, caregivers declined due to financial constraint. She continued to receive care which she responded with a decline in respiratory rate (64/min), heart rate (136/min), and temperature normalized (37.2°C). The duration of hospital stay was 2 weeks. She was discharged and was seen at a follow-up clinic 2 weeks after discharge with a chest scanogram, which showed resolution of initial findings [Figure 2]. Noncontrast computerized tomography scan of the chest also showed normal finding except for differential lung expansion in favor of the right lung [Figure 3a-d]. The baby is now thriving well with normal anthropometric measurements.

**Discussion**

CPAM cases are typically identified prenatally by routine ultrasonography screening. Most postnatally identified cases present in the newborn period. Our patient’s mother did not receive any antenatal care; hence, the lesion was not detected prenatally. CPAM may present in older child and adult as an incidental finding or secondary to repeated chest infection. The left lung is involved as often as the right lung with single lobe disease observed four times more often than...
multi-lobe disease[10] as was the case in our patient. The most common mode of presentation is acute respiratory distress secondary to the cyst expanding and compressing its surrounding structures. The distress occurs through a ball-valve mechanism leading to air trapping. This mode of presentation is common during the neonatal period.[11] This patient presented with cough, fever, respiratory difficulty, and poor feeding in the neonatal period with the cystic lesion affecting only the left lung. Other common presentations are recurrent chest infection, hemoptysis, dyspnea, cough, fever, failure to thrive and on examination, tachypnea, pneumothorax, cyanosis, accessory muscle use, and grunting may be present.[11] Complications such as fetal death, premature delivery, pneumothorax, pulmonary hypertension, recurrent pneumonia, haemothorax, and malignant change can occur.[12,13] Imaging studies such as chest radiography, computed tomography (CT) scanning, magnetic resonance imaging, prenatal ultrasonography and renal, cerebral ultrasonography, and echocardiography in newborns may be done as indicated.[11]

The treatment of symptomatic CPAM is always surgical as soon as the diagnosis is made. Thoracotomy and delivery of the hyperinflated lobe would bring immediate relief of the ventilatory and circulatory embarrassment. Lobectomy is usually necessary, but segmental resection is occasionally feasible. There is agreement among surgeons regarding the treatment of symptomatic patients; however, controversy exists about the management of asymptomatic neonates and infants with CPAM with respect to the decision and timing of excision.[14]

Complete spontaneous postnatal resolution of CPAM is said to be rare, and its existence even questioned by some.[15] However, in a 7 years retrospective study by Butterworth and Blair,[15] 2 out of the 56 cases reviewed showed spontaneous resolution at 5 and 37 months of age, respectively. This is similar to the index patient through the resolution occurred earlier. The patient was followed monthly for 3 months with no history suggestive of pneumonia, and she was thriving well.

**Conclusion**

Based on the presentation of this case, clinically, its manifestation cannot be distinguished from other causes of respiratory difficulty. Therefore, a higher index of suspicion should be key so that such condition is not missed. We conclude that the patient is unique as there was a spontaneous resolution of symptoms and signs of CPAM within 1 month of presentation with a radiological corroboration.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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