Congenital pulmonary airway malformation with spontaneous regression in an extremely preterm baby from Sri Lanka

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Introduction

Congenital pulmonary airway malformation (CPAM) is a rare congenital malformation of the lung with an incidence of 1 in 25,000–35,000 live births1. It accounts for 25% of all congenital lung anomalies and 95% of congenital cystic pulmonary disease2. CPAM is characterized by single large or multiple smaller cysts usually involving a single lobe with ipsilateral pulmonary tissue compression and pulmonary hypoplasia1. It is classified histopathologically into five types1. Type I is the commonest and accounts for 60% of CPAM1. Resection of malformed lung fragment is the first-line treatment, and this is recommended before 10 months of age1. We report spontaneous resolution of CPAM in a baby born at 25 weeks of gestation with a birth weight of 520g, the smallest baby reported so far to undergo spontaneous resolution and the first such case reported from Sri Lanka.

Case report

A non-asphyxiated baby girl with a birth weight of 520g was delivered by emergency caesarean section at 25 weeks of gestation due to pregnancy induced hypertension and pre-eclampsia. Mother was 38 years of age. This was the second offspring of non-consanguineous parents with a healthy 8-year-old son. Anomaly scan done at 22/52 +6 was reported as normal.

Baby was admitted to the neonatal intensive care unit due to extreme prematurity. She required surfactant therapy and mechanical ventilation to manage neonatal respiratory distress syndrome (NRDS). Initial chest x-rays were compatible with NRDS. She was stable till day 10 when the oxygen saturation started fluctuating. Chest x-ray (CXR) showed a hyperexpanded left lung with multiple cysts and right mediastinal shift with right lung collapse (Figure 1).

Opinion from a thoracic surgeon was sought at this stage and the decision was to manage conservatively if weaning off from the ventilator is possible. Baby was successfully weaned off to non-invasive positive pressure ventilation (NIPPV) at one month of age. She was discharged from hospital at 4 months of age after achieving satisfactory growth.

Figure 1: Hyperinflated left lung with multiple cysts

Radiological diagnosis of CPAM of the left lung was made. On Day 15, there were desaturating episodes and reduced air entry on the left side which improved with needle aspiration. CXR showed a left sided tension pneumothorax which was managed with intercostal tube insertion under water drainage. Contrast computed tomography of the chest was performed at this stage and the diagnosis of left sided CPAM with a pneumothorax was confirmed (Figure 2).

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The baby was closely followed up at the well-baby clinic. There were no respiratory symptoms, wheezing episodes or lower respiratory infections since discharge. Chest x-rays gradually improved. Contrast enhanced computerized tomography (CECT) which was planned at one year of age was postponed at parent’s request during the Covid-19 pandemic. The CECT lung, done at 3 years of age, showed a well expanded left lung without pulmonary hypoplasia and a few thin-walled cysts which are clinically insignificant. (Figure 3).

Baby is currently four years of age, gaining weight with normal developmental milestones.
Discussion

CPAM, previously known as congenital cystic adenomatoid malformation (CCAM), is a rare developmental anomaly of the lower airways which occurs due to excessive proliferation of tubular bronchial structures during pseudo glandular and saccular periods of lung development. Males and females are equally affected. CPAM is usually unilateral and restricted to one lobe but may involve all lobes of a lung or both lungs. There is still uncertainty concerning pathophysiology, natural history of the disease and the best treatment approach.

A 4% rate of spontaneous resolution has been reported for this condition postnatally. Spontaneous resolution of CPAM has also been reported from India. However, spontaneous resolution of CPAM in an infant born prematurely has only rarely been reported in the literature. This is also the first time that such a case has been reported from Sri Lanka.

CPAM can be diagnosed antenatally when ultrasound of fetal lung reveals cystic or solid lesions accompanied by polyhydramnios and fetal oedema. CPAM usually present in the neonatal period or early childhood with respiratory distress, recurrent chest infection, haemoptysis, or failure to thrive; 90% patients are diagnosed within the first two years of life. The presence of bilateral disease and hydrops fetalis are indicators of poor outcome in the absence of prenatal intervention, while early detection, polyhydramnios and mediastinal shift are not considered poor prognostic signs. Up to one third of antenatally diagnosed cystic lesions resolve before birth.

The sensitivity of CXR in the postnatal period is only 60%. However, the sensitivity of computed tomography (CT) or magnetic resonance imaging (MRI) is 100% in detecting the malformation in the postnatal period. Surgical excision of symptomatic lesions is relatively straightforward, but management of asymptomatic lesions is controversial. Surgical excision is favoured because of the risk of infection and malignant transformation if the CPAM remains in situ. When newborn presents with symptoms such as respiratory distress, or imaging signs like mediastinal shift, polyhydramnios or hydrops, prompt intervention by a paediatric surgeon to excise the lesion is advised. Recent advances in thoracic surgery and anaesthesia have minimized the mortality of new-born babies and infants with congenital cystic pulmonary lesions. In recent years, thoracotomy has been sometimes replaced by minimally invasive thoracoscopy. Due to the potential of malignant transformation, children should have long term follow up.
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