Unilateral pulmonary agenesis—a rare cause of respiratory distress in infancy

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INTRODUCTION

Congenital malformations of the lung are rare and vary widely in their presentation and severity.¹ Pulmonary agenesis is defined as complete absence of the lung and its bronchus.² Cases presenting later on in infancy or childhood may be missed and other common illnesses like foreign body, collapse and/or consolidation may be mistakenly diagnosed. A high index of suspicion is needed to diagnose an underlying malformation at such times. We report a case of unilateral pulmonary agenesis which was initially erroneously diagnosed as left lung collapse.

CASE REPORT

A two-month-old female infant presented with history of fever, cough, and coryza of four days duration followed by poor feeding of one day's duration. She was a product of a non-consanguineous marriage after premature home delivery at 32 weeks period of gestation to a primi mother. The baby was admitted immediately after delivery to a private hospital for pre-term care for 15 days; the birth weight was noted to be 1.5 Kg. There was no respiratory distress noted and baby was feeding well at the time of discharge. No other abnormalities were detected there. Chest radiograph was not taken. At admission to our hospital, the child was lethargic, centrally cyanosed and tachypnoeic with respiratory rate of 74/min and severe bilateral subcostal and intercostal recessions. Breath sounds were absent over the left hemithorax. The trachea and apical impulse were shifted to the left, with apex beat at anterior auxiliary line in fifth intercostal space. A continuous grade III 'machinery', murmur was heard in left second intercostal space in pulmonary area. SpO₂ in air was 78%, after oxygenation by hood rose to 92%.

Chest radiograph showed homogenous opacity in left hemithorax, a hyperinflated right lung, trachea was deviated towards the left side and parahilar haziness with mediastinal shift to the left (Figure 1). Two-dimensional echocardiography with colour Doppler showed a normal heart displaced more towards the left side, with a patent ductus arteriosus with flow from aorta to pulmonary artery and non-visualisation of left pulmonary artery. Based on clinical, radiographic and echocardiographic findings, a provisional diagnosis of complete obstruction of left main bronchus with collapse/consolidation of the left lung was made. A possibility of persisting ductus arteriosus with left to right shunt, an enlarged pulmonary artery with cardiomegaly leading to compression of the left main bronchus was thought of.

Bronchoscopy showed non-visualisation of the left main bronchus. High resolution computed tomography (HRCT) thorax (Figure 2) showed a homogenous opacity involving left hemithorax which was totally unaerated, along with mediastinal shift to left, non-visualisation of left main stem bronchus and non-homogeneous infiltrates in peribronchial region on right side, suggestive of agenesis of left lung with right sided compensatory emphysema and bronchopneumonia.

Figure 1 Chest radiograph showing homogenous opacity in left hemithorax devoid of any bronchovascular markings, a hyperinflated right lung extending into the upper portion of left hemithorax, trachea is deviated towards the left side and parahilar haziness with mediastinal shift to the left, cardiac shadow masked by compensatory emphysema of right lung and blending with the left homogenous unaerated left hemithorax.

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Figure 2 High resolution computed tomography thorax showing homogenous opacity involving left hemithorax (totally unaerated) with severe mediastinal and tracheal shift to left side, non-visualisation of left main stem bronchus, and non-homogeneous infiltrates in peribronchial region on right side, suggestive of agenesis of left lung with right-sided compensatory emphysema and bronchopneumonia. The non-visualisation is clearly seen in the scout film during high resolution computed tomography.

Patient was managed conservatively with antibiotics and chest physiotherapy. The cyanosis persisted and the bronchopneumonia did not resolve adequately. The patient went on discharge against medical advice after two weeks and did not return for follow-up.

DISCUSSION

Pulmonary agenesis entails the absence of bronchial stump or carina with absence of lung. Bilateral pulmonary agenesis is incompatible with life, presenting as respiratory distress and failure.³ Parental consanguinity through an autosomal recessive mode of inheritance, may be a predisposing factor. Left sided lung agenesis is slightly more common and sometimes associated with congenital heart disease such as tetralogy of Fallot, patent ductus arteriosus and septal defects. Right sided lung agenesis may cause more problems from mediastinal shift. This results in obstruction of major vessels and airways, and is more frequently associated with congenital heart disease, such as obstructed anomalous pulmonary venous drainage, ventricular septal defect and coarctation of aorta.⁴ Pulmonary agenesis is often seen with VACTERL sequence.³

The exact incidence of this condition is not precisely known. It ranges between 3.4 and 9.7 per 100,000 live births.¹ About 200 cases of unilateral pulmonary agenesis have been documented in the current literature. Only up to one-third cases are diagnosed when the patient is alive. Most of them are associated with additional congenital abnormalities involving the skeletal, cardiovascular, gastrointestinal, and genitourinary systems.^{2,5} In our patient only cardiovascular abnormalities were seen, namely patent ductus arteriosus with flow from aorta to pulmonary artery and non-visualisation of left pulmonary artery. There were no gastrointestinal tract and genitourinary system abnormalities seen clinically and with sonography.

Monaldi divided the maldevelopment of lung into four groups. Group I: no bifurcation of trachea; group II: only rudimentary main bronchus; group III: incomplete development after division of main bronchus; and group IV: incomplete development of subsegmental bronchi and small segment of the corresponding lobe.⁶ According to Boyden, there are three degrees of maldevelopment: (i) agenesis, in which there is complete absence of lung tissue, (ii) aplasia, in which rudimentary bronchus is present but no lung tissue is present, and (iii) hypoplasia, in which all the normal pulmonary tissues are present but are under-developed.⁶ The present case belongs to the first group of Monaldi classification, and agenesis according to Boyden's classification.

Most cases present with respiratory distress in the early neonatal period. It may present as recurrent respiratory symptoms in early childhood or be discovered incidentally.² Clinical appearance may mimic total left main bronchus obstruction by a mucous or meconium plug. Sometimes the presentation is akin to that of a case of congenital heart disease. It has to be differentiated from atelectasis, diaphragmatic hernia, adenomatoid cystic malformations, and sequestrations. Chest radiograph shows homogenous opacity in place of lung in one hemithorax. Ribs may be crowded and mediastinal shift towards that side is seen. Chest computed tomography scan is necessary to confirm the absence of lung tissue on one side. Bronchoscopy shows absence of ipsilateral main bronchus which may be confirmed by bronchography.⁴ The best method for precise diagnosis of pulmonary agenesis is pulmonary angiography which confirms the absence of ipsilateral pulmonary vessel.¹ This could not be done in our case due to limited facilities and the patient was lost to follow-up. However, there was indirect evidence by 2D-echo with colour Doppler which showed non-visualisation of the left pulmonary artery.

If affected infants survive beyond neonatal period, they are likely to have recurrent pneumonia. Reactive airway disease

may develop and so they should be meticulously followed up and evaluated with pulmonary function testing and echocardiography.¹ Treatment is mainly conservative. Pulmonary infections are to be treated energetically and chest physiotherapy may be of some benefit. Asymptomatic cases when not associated with any other congenital anomalies do not require any treatment.⁷

CONFLICTS OF INTEREST

None identified.

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