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Case Report

Unilateral pulmonary agenesis in a child with recurrent respiratory tract infections

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ABSTRACT

Pulmonary agenesis is a rare congenital anomaly defined as complete absence of lung parenchyma with its bronchial tree and vasculature. It usually presents in early childhood and may be associated with other congenital abnormalities. Chest X-ray gives a suspicion of this condition which can be further confirmed by computed tomography scan of thorax. Oher investigations like echocardiography and ultrasonography of whole abdomen must be done to rule out associated congenital anomalies. We describe here a case of one month old male who presented with cough and shortness of breath. Chest X-ray showed right sided homogenous opacity which on contrast enhanced computed tomography revealed right sided pulmonary agenesis. Echocardiography showed situs solitus with dextroposition of heart. The child was treated with intravenous antibiotics, moist oxygen and nebulisation with bronchodilators following which there was clinical resolution of symptoms. In conclusion, congenital pulmonary agenesis should be included in the differential diagnosis in a child presenting with dyspnoea and homogenous opacity of unilateral lung field in Chest X-ray for early diagnosis, management and prevention of recurrent respiratory infections and consequently to reduce childhood mortality.

Keywords: Congenital abnormalities, Hypoplasia, Respiratory tract infections

INTRODUCTION

Pulmonary agenesis is a rare congenital anomaly. It occurs due to failure of development of the primitive lung bud which normally arises from the ventral wall of the foregut at 4th week of gestation.[1] Prevalence of pulmonary agenesis is about 34 per 1,000,000 live births.[2] In 1673, De Pozze discovered this anomaly during the autopsy of an adult female.[1] In India, Muhamed first reported this condition in a medicolegal autopsy in 1923.[1] It is usually diagnosed during infancy, but sometimes patients are incidentally detected during adulthood.[2] Almost 50% patients with unilateral condition die within 5 years of age while patients with bilateral pulmonary agenesis fail to survive.[2] Oyamada et al reported the oldest case in a 72 year old patient.[3] This report describes a case of a one month old boy, who presented with respiratory distress and eventually was diagnosed as right sided pulmonary agenesis.

CASE REPORT

A one-month-old male child was brought to our emergency with complaint of dry cough and severe respiratory distress. The mother gave history of recurrent respiratory tract infections since day 10 of birth. The baby was born to non-consanguineous parents by lower uterine caesarean section at 39 weeks of gestation. His birth weight was 2.3 kg. The antenatal and early neonatal periods were uneventful. On examination, the child had tachypnoea, nasal flaring, subcostal suction without any cyanosis. His chest was normal shaped with decreased movement on right side. Apex beat was situated on right 4th intercostal space along anterior axillary line. Percussion note was dull on right side and resonant on left side of the chest. Auscultation of chest revealed absent breath sounds on right side, wheeze on left side and normal rhythmic heart sound with tachycardia on right side. Other system examinations were normal.

Chest X-ray showed homogenous opacity on right hemithorax, hyperinflation of left lung, deviation of trachea towards right side, absent cardiac shadow on left side (Figure 1). Contrast enhanced computed tomography (CECT) of thorax showed complete absence of right sided lung parenchyma with its bronchial tree and vessels, hyperinflation of left lung with trans-mediastinal herniation towards right with cardio-mediastinal shifting towards right (Figure 2). Echocardiography showed dextroposition of heart with normal right to left concordance with situs solitus. Ultrasonography of whole abdomen was normal.

Figure 1: Chest X-ray showing homogenous opacity on right hemithorax, hyperinflation of left lung, deviation of trachea towards right side, absent cardiac shadow on left side.

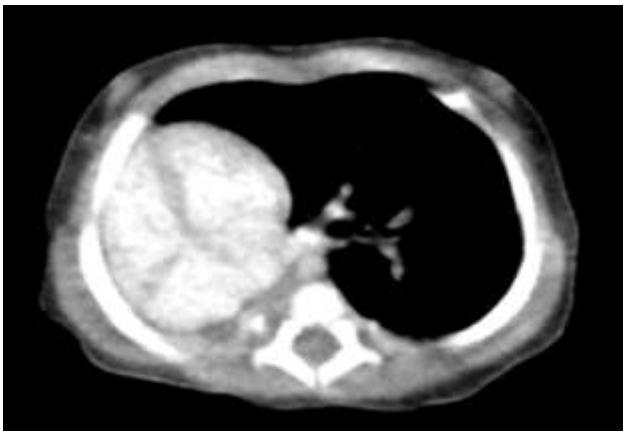

Figure 2: CECT thorax (axial view) showing complete absence of right sided lung parenchyma with its bronchial tree and vessels, hyperinflation of left lung with trans-mediastinal herniation towards right with cardio-mediastinal shifting towards right.

The child was treated with intravenous antibiotics, moist oxygen and nebulisation with bronchodilators following which there was clinical improvement. The mother was counselled about the existing condition and prognosis of the child. She was also advised for follow up in our outpatient department and to immunize the child as per national immunization schedule.

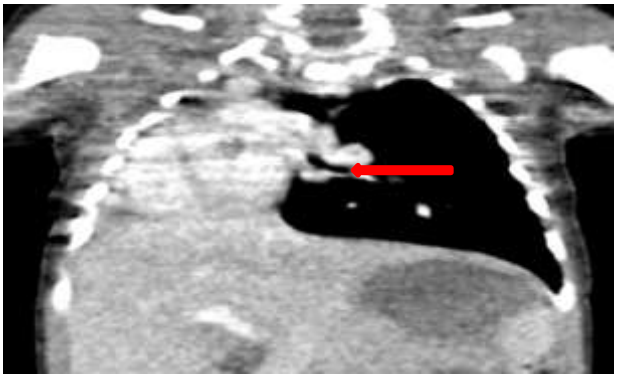

Figure 3: CECT thorax (coronal view) showing complete absence of right sided lung parenchyma with its bronchial tree and vessels, hyperinflation of left lung with trans-mediastinal herniation towards right with cardio-mediastinal shifting towards right. Red arrow shows left bronchus. Red arrow shows left bronchus.

DISCUSSION

Schneider classified pulmonary agenesis at first, whereas Boyden modified it into three groups according to the stage of primitive lung bud development.[1]

In type 1 (agenesis), unilateral lung parenchyma is completely absent along with its bronchus and vasculature; in type 2 (aplasia), complete absence of unilateral lung parenchyma with a rudimentary bronchus; and in type 3 (hypoplasia), partial existence of unilateral lung parenchyma, its bronchial tree and vessels.

Our case can be classified as type 1 due to complete absence of unilateral lung parenchyma, bronchus and vessels as confirmed by CECT of thorax.

Possible etiologies responsible for this condition are genetic factors (e.g. duplication of distal part of upper arm of chromosome 2), dietary deficiency of vitamin A and folic acid during early pregnancy or some viral agents.[4]

Left sided lung agenesis is more common with a better prognosis. Survival rate in patients with right lung agenesis is worse because it is often associated with excessive cardio-mediastinal shift and malrotation of carina leading to distorted broncho-vascular structures, poor drainage of functioning lung and more susceptibility to respiratory infections.[5] The functioning lung produces almost twice alveoli as a compensatory mechanism.[6]

Approximately 50% patients have associated congenital anomalies of cardiovascular, gastrointestinal, genitourinary and musculoskeletal systems.[1] Few cases of Klippel- Feil syndrome, VACTERL syndrome associated with pulmonary agenesis have been reported till date.[7,8] In our case, the child was thoroughly evaluated by investigations like ultrasonography of whole abdomen, echocardiography to rule out other congenital anomalies.

Right sided homogenous opacity with ipsilateral mediastinal shift on chest X-ray was initially thought to be probably due to right sided whole lung collapse. Chest X-ray showing white-out lung also raised a suspicion of lung agenesis which was finally confirmed by CECT of thorax. Bronchoscopy and CT pulmonary angiogram (CTPA) can also be done to establish the absence of ipsilateral bronchus and pulmonary artery respectively.[2] Prenatal diagnosis of isolated lung agenesis can be done by ultrasonography by showing hyperechoic hemithorax although definitive diagnosis is difficult. Fetal magnetic resonance imaging (MRI) can be done to confirm the diagnosis in utero.[5]

Functional integrity of the remaining lung and presence of associated congenital anomalies are the main factors deciding the prognosis of these patients.[1] Patients with isolated lung agenesis only require conservative management. Surgical measures like diaphragmatic translocation, aortopexy, aortic grafting and resection, slide tracheoplasty are required if respiratory failure occurs due to torsion and compression of the trachea by cardio-mediastinal rotation and displaced aortic arch.[9] Agrawal et al reported a case of right lung agenesis with tracheal stenosis due to complete tracheal rings which had been successfully managed by placement of saline tissue expander into the extra pleural space in a 5 month old male infant.[10]

CONCLUSION

Although a rare entity, all clinicians should suspect congenital pulmonary agenesis in a child with dyspnoea and homogenous opacity of unilateral lung field in chest X-ray for early diagnosis, management and prevention of recurrent respiratory infections and consequently to reduce childhood mortality.

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