Case Report:
Left Lung Hypoplasia with Left-Sided Pulmonary Artery Agenesis and Right-Sided Aortic Arch in A Toddler Girl in Aseer Region, Saudi Arabia

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Abstract

Background: Pulmonary hypoplasia is an uncommon congenital aberration which is often interconnected with other abnormalities.

Objective: To report a rare condition of left pulmonary hypoplasia associated with some great vessels anomalies.

Case Report: A 4-year-old Saudi girl with recurrent chest infections was shown to have left lung hypoplasia along with left sided pulmonary artery agenesis and right sided aortic arch. Her condition improved after medical treatment then she was referred for surgical management.

Conclusions: Lung hypoplasia is a rare disease. However, it should be suspected when children present with recurrent chest infections. Our case had a classical presentation since her infancy period. Early diagnosis of such cases is essential for subsequent management and achieving a good outcome.

Key Words: Pulmonary hypoplasia – Pulmonary artery agenesis – Congenital anomaly – Recurrent pneumonia.

Introduction

PULMONARY hypoplasia is a rare congenital anomaly, in which there is low number or size of the airways, vessels and alveoli. The decrease in both the number of alveoli (up to 67%) and the number of airway generations (up to 50%) results in small fibrotic and nonfunctioning lung [1] with bronchiectatic changes [2].

The associated anomalies involve the urinary system [3], diaphragm, cardiovascular system [4], central nervous system [5], as well as musculoskeletal anomalies of the thoracic cage, Klippel Feil syndrome and Down syndrome [7].

The aim of this case report is to describe a rare condition in a child with lung hypoplasia, absent left pulmonary artery and right-sided aortic arch, who presented with recurrent chest infections.

Case Report

In January 2016, a 4-year-old Saudi girl presented to the Pediatrics' Outpatient Clinic of the Maternity and Children's Hospital, in Abha, Saudi Arabia, with a one-day history of cough, fever and shortness of breath. There was a significant history of recurrent chest infections that started at the age of nine months with frequent hospital admissions.

The patient was a precious baby, born at full term via cesarean section after 10 years of secondary infertility. The mother was on hormonal therapy for a long time and was on regular follow-up during pregnancy. The patient had normal development and had received up-to-date vaccinations.

On physical examination, the child looked ill, in respiratory distress, having unstable vital signs, febrile (temperature 38.7°C), tachypnic (respiratory rate 80 breaths/min), with tachycardia (pulse rate 150 beats/min), normotensive (blood pressure 111/60 mmHg) and an oxygen saturation of 75% on room air. Her growth parameters were normal for her age. Chest examination showed signs of respiratory distress, pectus excavatum, asymmetry of the two sides of the thorax, and decreased air entry in the left side with dullness on percussion.
There were wheezing and crepitation on the right side. The remainder of physical examination was unremarkable.

Nasopharyngeal swab for influenza A (H 1 N 1) was negative. Sepsis screening was unremarkable. The biochemical workup at presentation showed white blood count of $21.9 \times 10^{3}/\mu L$ ($5.5-15.5 \times 10^{3}/\mu L$), neutrophils 68%, lymphocytes 18%, hemoglobin level was 14.5g/dl and platelets count was $311/mm^{3}$ ($250,000-550,000/mm^{3}$), estimated sedimentation rate of 8mm/hour (3-13mm/hour), and C-reactive protein was non-reactive. Venous Blood Gasses (VBG) revealed: pH=7.174 (7.38-7.42), PCO$_{2}$=43mmHg (38-42mmHg), PO$_{2}$=46.9 mmHg (75-100mmHg), HCO$_{3}$=14.2meq/L (22-28 meq/L), and BASE=−11.6. Renal and liver function tests were within normal limits.

Chest X-ray showed loss of lung aeration on the left side and shifted mediastinum to the left side with hyperinflated right lung Fig. (1). Echocardiogram revealed absent left pulmonary artery, right aortic arch and normal cardiac function. Computed Tomography (CT) scan of the chest showed complete agenesis of the main left pulmonary artery with hypoplastic left lung, associated with compensatory hyperinflation of the right lung and shifting of mediastinum to the left side, mild right lower lobe atelectasis/consolidation Fig. (2) and complete agenesis of the left main pulmonary artery Fig. (3).

Lung perfusion scan (VQ) was done with a result of whole cardiac mass completely shifted to the left thorax, normal perfusion to right lung, absent left lung perfusion and no scintigraphic evidence of systemic shunting. Electroencephalography (EEG) report was unremarkable with normal CT brain results.

Based on clinical and radiologic findings, a diagnosis of left lung hypoplasia, complete agenesis of the left pulmonary artery and right aortic arch with bronchopneumonia was reached.

The patient was admitted to the Pediatric Intensive Care Unit (PICU), intubated and connected to mechanical ventilation. She received supportive management and empiric antibiotics. She spent 10 days after admission and showed improvement after which she was transferred to the Cardiac Center for further management.

Axial CT scan with spiral volume acquisition showed disconnected left pulmonary artery, right aortic arch with bilateral Patent Ductus Arteriosus (PDA), anterior closed left PDA from innominate
artery and posterior PDA to the right pulmonary artery. There was a good size and caliber of the trachea and both main bronchial branches bilaterally with no evidence of obstruction.

Surgical intervention was planned for exploration of the left pulmonary artery. Later on, the patient underwent operation for retrieval of intrapulmonary left pulmonary artery and construction of systemic shunt from the left subclavian artery to left pulmonary artery, using a 4mm Gortex tube graft.

The patient was then transferred to PICU and was kept on antibiotics. She showed improvement and was then discharged in a good condition for follow-up.

**Discussion**

In this report we described a rare case of 4-year-old girl with left lung hypoplasia along with left sided pulmonary artery agenesis and right sided aortic arch. Chest X-ray revealed loss of lung aeration on the left side and shifted mediastinum to the left side with hyperinflated right lung. There were also absent left pulmonary artery and right aortic arch.

Fox et al., [8] stated that unilateral pulmonary hypoplasia is usually associated with other congenital abnormalities and presents with early respiratory distress. Chest X-ray shows absent lung with compensatory hyperinflation of contralateral lung.

Bialowas et al., [7] stated that the estimated prevalence of single Unilateral Absent Pulmonary Artery (UAPA) is 1/200,000 patients and there is no predilection for the right or left side. Right-sided aortic arch is a kind of aortic arch variants, in which the aortic arch is on the right side rather than on the left. Kotecha [9] added that several types of right-sided aortic arch are present, the most common ones being right-sided aortic arch with aberrant left subclavian artery and the mirror-image type and it associated with congenital heart disease.

Our case presented with a history of shortness of breath, and recurrent chest infections with frequent hospital admissions. Chest examination showed asymmetry of both sides of the chest, decreased air entry in the left side with dullness on percussion, in addition to wheezing and crepitation on the right side. The diagnosis has been settled by chest X-ray and CT scan of the chest.

Kant [10] stated that, in cases of unilateral lung hypoplasia, clinical findings depend on degree of pulmonary abnormality and presence of other congenital malformations. The patient is usually symptomatic. Physical examination characteristically reveals asymmetry of two sides of thorax, reduction in respiratory movements and absence of air entry on the affected side.

Suzuki [11] noted that there are no clear clinical diagnostic criteria to facilitate the identification and management of lung hypoplasia. However, the condition may be diagnosed incidentally during childhood when complicated by pulmonary infection. He added that the diagnosis can be reached with help of chest X-ray, CT thorax, fiber optic bronchoscopy, and if possible pulmonary angiography and bronchography.

Bouros et al., [12] noted that when lung hypoplasia is an isolated finding, it is compatible with even normal life. In most cases, isolated Unilateral Absence of Pulmonary Artery occurs in the right lung [10,13], probably because it usually occurs on the opposite side of the aortic arch due to embryologic relationships, but in our case, it was on the left side, which has been less frequently reported [14-16].

The definite etiology for lung hypoplasia and the other associated conditions in our case could not be reached. However, Alsadi et al., [16] stated that congenital pulmonary hypoplasia may be regarded as primary (idiopathic) or secondary (when it occurs in association with environmental factors or other congenital anomalies). Moreover, Elhassan et al., [15] postulated that a vascular injury in weeks 5-8 in utero could be responsible for the arrest of lung maturation in utero and might be the main reason for both pulmonary artery thrombosis and pulmonary hypoplasia.

Glasberg et al., [17] added that the incidence of secondary form of pulmonary hypoplasia is difficult to determine. Because of its association with a variety of other abnormalities and the difficulty of pathologic diagnosis in some cases, it is likely to be more common than generally recognized. In more than 90% of the patients with secondary lung hypoplasia, the renal and genitourinary anomalies were the most common anomalies, followed by diaphragmatic hernia and musculoskeletal dysplasia [18].

The associated agenesis of left pulmonary artery in our case is a result of failure in the connection of the sixth aortic arch with the pulmonary trunk.
during embryologic development. Most commonly, it occurs with cardiovascular malformations such as tetralogy of Fallot or cardiac septal defects, but it can also occur in isolated form [13].

Unilateral tracheal stenosis along with, unilateral pulmonary agenesis and patent ductus arteriosus have been mentioned in the literature and can rarely occur as a part of VACTERL sequence [19].

Management of our case was both medical and surgical. Turner et al., [20] stated that treatment of lung hypoplasia is in the form of a medical as well as surgical care, both before and after delivery. Before delivery, patient is treated medically with repeated amniocenteses with or without the use of tocolytics, antibiotics and steroids. After delivery, respiratory support is given ranging from oxygen to mechanical ventilation including extracorporeal membrane oxygenation. Dialysis may be required for support of renal function. Surfactant administration at 4ml/kg improves survival rate. Surgical care consists of intrauterine vesicoamniotic shunts and endoscopic ablation of valves and plug the lung until it grows by fetoscopic tracheal occlusion with a clip. Post-delivery surgery can be done to correct diaphragmatic hernia, cystic adenomatoid malformations and decompresses pleural effusions.

Alsaadi et al., [16] stated children with pulmonary hypoplasia are vulnerable to infections and hence emphasis should be there for consideration of palivizumab prophylaxis, and they should also receive pneumococcal and influenza vaccinations. Palivizumab prophylaxis reduces hospitalization for respiratory syncytial virus significantly. Insult like pneumonia or aspiration to the remaining/healthy lung should be aggressively managed. Severe infections sometimes require lobectomy or pneumonectomy.

Chand et al., [13] noted that management of UAPA consists of routine echocardiographic monitoring of asymptomatic patients for the development of pulmonary hypertension, and vasodilator therapy if pulmonary hypertension is established. Palliative surgical treatments of congenital heart defects associated with UAPA is systemic to pulmonary artery shunt and transluminal balloon pulmonary valvuloplasty in patients with non-severe hypoplasia of the single pulmonary artery. Palliative reconstruction of the right ventricular outflow tract is a more favorable procedure for patients with a severe hypoplasia of the single pulmonary artery. Harkel et al., [21] added that any pulmonary surgery in a patient with UAPA may be complicated by the presence of systemic collaterals.

Patients with left lung agenesis have a better prognosis than those with the right one, and the absence of critical anomalies in patients with left lung agenesis may even be the cause of increased survival in this set of patients [22]. According to Say et al., [23], these patients can present with congestive heart failure or pulmonary hypertension in infancy or combination of symptoms in older age such as recurrent chest infections, chest pain, pleural effusion and dyspnea or exercise intolerance, hemoptysis, and high-altitude pulmonary edema. Pneumothorax and pulmonary hypertension are common serious complications. Pneumothorax often develops spontaneously or secondary to mechanical ventilation in infants who present with severe respiratory insufficiency in the first few hours of life.

In conclusion, lung hypoplasia is a rare disease and has a variety of presentations. However, it should be suspected when children present with recurrent chest infections. Our case had a classical presentation since her infancy period. Early diagnosis of such cases is essential for subsequent management and for achieving a good outcome.

References


