Hypoplasia of left pulmonary artery: A rare congenital heart disease

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Abstract

Unilateral hypoplasia or absence of pulmonary artery is a rare pulmonary vascular anomaly that is usually associated with other cardiac anomalies like Tetralogy of Fallot or cardiac septal defects, but can also occur as an isolated anomaly. We present a case of 6-month-old infant who presented with recurrent respiratory tract infection. On work up, chest X-ray showed ipsilateral small hemithorax with hyperluscency in the left lung field. Findings were confirmed with 2D echocardiography and pulmonary angiography which were suggestive of hypoplasia of left pulmonary artery.

Key words: *Congenital heart disease, Hemithorax, Hypoplasia of left pulmonary artery, Multislice computed tomography angiography*

Unilateral absence or hypoplasia of a pulmonary artery is a rare condition with an estimated prevalence of 1 in 2,00,000 [1]. Isolated hypoplasia involves right lung in about 2/3rd of the cases in which hypoplasia is on the side of the chest opposite to the aortic arch. The prognosis depends upon the associated cardiovascular anomaly and a degree of pulmonary hypertension (PAH). Patients may present with chronic dyspnea and exercise intolerance. Sometimes, it can present with more serious complication such as hemoptysis. Diagnosis of hypoplasia of pulmonary artery can be difficult, but important clues are present in chest radiographs. Patients who develop PAH can be treated medically with vasodilator. Complication like the hemoptysis can be treated with embolization, lobectomy or pneumectomy.

CASE REPORT

A 6-month-old male infant presented with cough, fever and breathlessness for 4 days. He was first born child to non-consanguineous parents. There was history of hospitalization thrice with similar complaints starting from the neonatal period. The general examination and anthropometric measurements were unremarkable. There was no cyanosis or clubbing, and the pulse oximetry showed normal arterial saturation. On systemic examination, respiratory examination showed tachypnea, subcostal and intercostal retraction. Auscultation of the chest showed fine crepitations and wheezing in the right lung field. On cardiovascular examination, he had systolic murmur grade 2/6 in 3rd and 4th left intercostal space and 2nd left intercostal space.

On laboratory workup, hemogram showed mild anemia with polymorphonuclear leukocytosis. Chest X-ray (Fig. 1) was suggestive of asymmetric lung fields, with an ipsilateral small hemithorax with hyperlucency in the left lung field. Considering the recurrence of pneumonia in the same site in all consecutive hospitalizations and presence of the murmur, 2D echocardiography was done, which showed? partial anomalous pulmonary venous connection of right upper pulmonary vein, patent foramen ovale with left to right shunt, moderate tricuspid regurgitation, and mild PAH. For further confirmation of findings, angiography was advised. Computed tomography (CT) thorax was done, which showed bilateral upper lobe consolidation. Multislice CT pulmonary angiography (Figs. 2 and 3) was done, which showed small (atretic) left pulmonary artery with prominent left superior intercostal vein.

The patient was treated with intravenous antimicrobial therapy along with other supportive measures for bronchopneumonia after which he improved gradually with resolution of respiratory symptoms. The cardiothoracic surgeon opinion was sought for the underlying heart condition and was advised regular follow up. The patient was immunized with *H. influenzae* type B, *Pneumococcal* and *Meningococcal vaccines* (in addition to routine immunization) on discharge to protect the child from common pathogens. Parents were counseled regarding the disease, associated complications and prognosis. The child came for follow-up regularly up to 10 months of age, he was growing well till then.

DISCUSSION

Unilateral absence or hypoplasia of a pulmonary artery is a rare condition with an estimated prevalence of 1 in 2,00,000 [1]. Pulmonary artery arises from sixth arch artery and opens into the

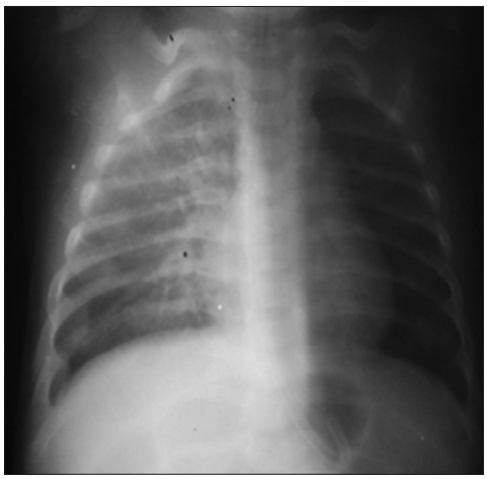

Figure 1: Small hemithorax with hyperlucency in left lung field

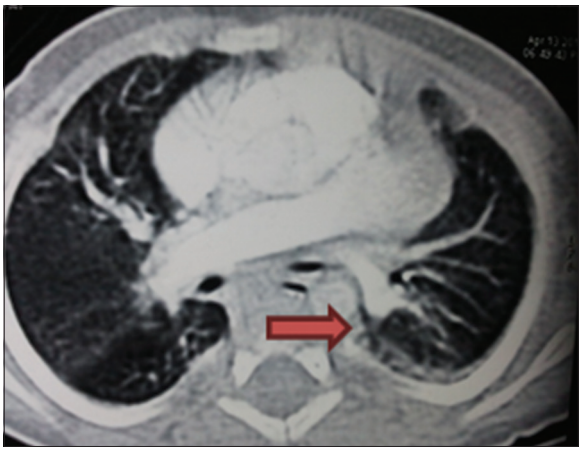

Figure 2: Small (atretic) left pulmonary artery with prominent left superior intercostal vein

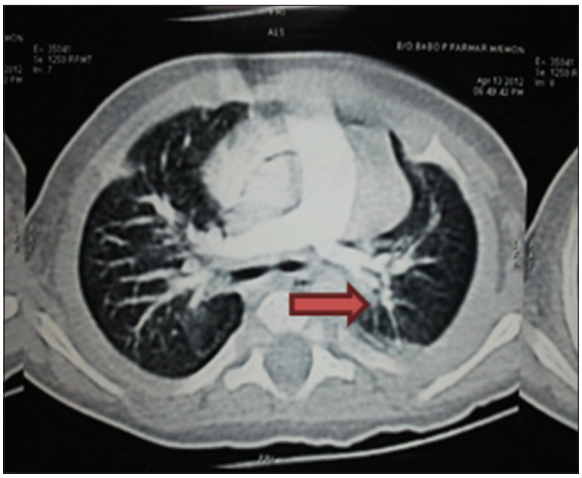

Figure 3: Small (atretic) left pulmonary artery with prominent left superior intercostal vein

dorsal part of the aortic sac. The spiral septum, which is formed in the truncus arteriosus and extends into the aortic sac, fuses with its posterior wall in such a way that blood from the pulmonary trunk arches into the sixth arch artery. Most commonly it occurs in conjunction with cardiovascular anomalies such as Tetralogy of Fallot or cardiac septal defects, but can also occur an isolated anomaly [1,2], as was seen in our patient.

Isolated unilateral absence or hypoplasia of pulmonary artery involves right lung in about 2/3rd of the cases in which hypoplasia of pulmonary artery is on the side of chest opposite to the aortic arch [3-4]. These patients are usually asymptomatic, which may delay the diagnosis. However, they may present with recurrent respiratory infections, chronic dyspnea, exercise intolerance, and sometimes with hemoptysis and high altitude pulmonary edema. The prognosis depends on the associated cardiovascular anomalies and the degree of PAH.

Various mechanisms have been proposed for many of the common sequelae of this condition. PAH may result from blood flow directed away from absent pulmonary artery to remaining pulmonary artery. Increased blood flow leads to sheer stress on the endothelium, which results in the release of vasoconstrictors such as endothelin [5]. Chronic vasoconstriction of the pulmonary arterioles lead to remodeling resulting in increased resistance in the pulmonary vasculature and PAH [5].

The etiology of recurrent infections observed in these patients is likely to be multifactorial. Lack of arterial blood flow to the affected lung may result in poor delivery of inflammatory cells to the sites of inflammation and impaired ciliary function [6]. In addition, poor blood supply to the affected lung may result in alveolar hypocapnea, leading to secondary bronchoconstriction and mucous trapping [6]. Hemoptysis is a serious complication and appears to be caused by large collateral circulation that subject venous systems to unusually high pressures.

Diagnosing unilateral absence or hypoplasia of a pulmonary artery can be difficult, but important clues are present in chest radiographs. X-ray chest may show asymmetric lung fields, with an ipsilateral small hemithorax holding a hyperlucent lung, which was present in our patient [1,7]. Definite diagnosis can be made by CT, magnetic resonance imaging or transthoracic echocardiogram. Pulmonary angiography is considered the gold standard for diagnosis.

Treatment should be based on the symptoms and pulmonary hemodynamic status. Asymptomatic patients should be followed regularly for the development of PAH. Patients who develop PAH can be treated medically with vasodilator therapy, which was not used in our as PAH was mild [3,5]. Alternatively, revascularization of peripheral branches of the affected pulmonary artery to the pulmonary hilum can be attempted [2,8,9]. Hemoptysis may be treated with embolization, lobectomy or pneumonectomy [10,11].

CONCLUSION

Though the disease is rare, we should suspect this disease is patients of recurrent respiratory infections, and chest X-ray showing

abnormality at the same site. Disease should be diagnosed by 2D echocardiography and then managed medically or surgically.

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