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UNILATERAL PULMONARY ARTERY AGENESIS PRESENTING AS LUNG HYPOPLASIA WITH PULMONARY ARTERY HYPERTENSION

Pulmonary Medicine

Dr Gokul Prashanth Post Graduate, Dept Of Pulmonary Medicine, Sri Ramachandra Institute Of Higher Education And Research, Chennai

Dr Aravind Ram* MBBS, MD Assistant Professor, Dept Of Pulmonary Medicine, Sri Ramachandra Institute Of Higher Education And Research, Chennai *Corresponding Author

Dr Hari Prasad MBBS, DTCD Professor, Dept Of Pulmonary Medicine Sri Ramachandra Institute Of Higher Education And Research, Chennai

ABSTRACT

Congenital pulmonary vascular anomalies represent a complex group of developmental abnormalities affecting pulmonary circulation. Among these conditions, unilateral pulmonary artery agenesis (UPAA) is one of the rarest anomalies. UPAA results from failure of development of the proximal segment of the sixth aortic arch during early embryogenesis. This leads to absence of the pulmonary artery on one side, resulting in chronic hypoperfusion of the ipsilateral lung sometimes, may be resulting in a unilateral interstitial lung disease. Rather than complete absence of lung tissue, the affected lung remains present but hypoplastic, relying on systemic collateral vessels for perfusion.Clinical manifestations vary widely, ranging from neonatal respiratory distress to asymptomatic adults discovered incidentally during imaging performed for unrelated conditions. Fewer than 500 cases of isolated UPAA have been reported worldwide, emphasizing both its rarity and the challenges associated with early diagnosis.

KEYWORDS

UPAA, Lung hypoplasia, ILD, Pulmonary Artery Hypertension

CASE REPORT

A 52-year-old female presented with complaints of cough with expecxtoration and worsening shortness of breath for six months. She complaints of recurrent respiratory tract infections since childhood. Patient had decreased saturation at room air, which improved to 94% with supplemental oxygen. Respiratory examination revealed bilateral air entry with diffuse wheeze, more prominent on the right side. Echocardiography revealed moderate pulmonary hypertension (57 mmHg) with mild right atrial and right ventricular dilatation and moderate tricuspid regurgitation. Chest radiography showed heterogeneous opacities in the left lung, along with right lung infiltrates and CT thorax showed absent left pulmonary artery, with arterial supply to the left lung arising from the aberrant left subclavian artery. Additional findings included right-sided aortic arch, double superior vena cava, and dilated main and right pulmonary arteries with reticular bands, interlobular septal thickening, subpleural cystic changes, traction bronchiectasis, and volume loss in the left lung, consistent with unilateral interstitial lung disease associated with left lung hypoplasia.

IMAGE 1 – Chest X ray showing left sided volume loss with pull of trachea to the same side.

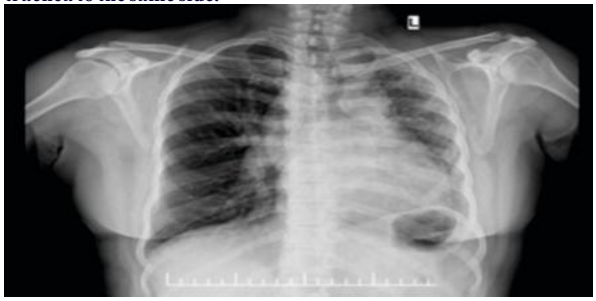


IMAGE 2 – left upper lobe cystic changes with upper lobe volume loss.

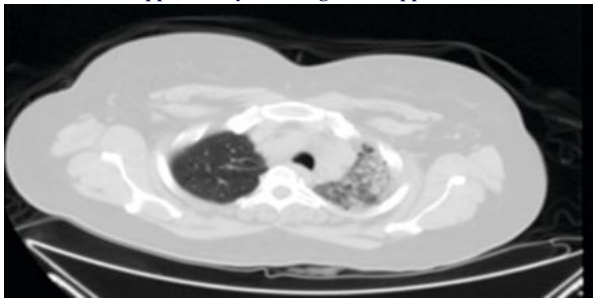


IMAGE 3- traction bronchiectasis of the left lung with rib crowding noted present.

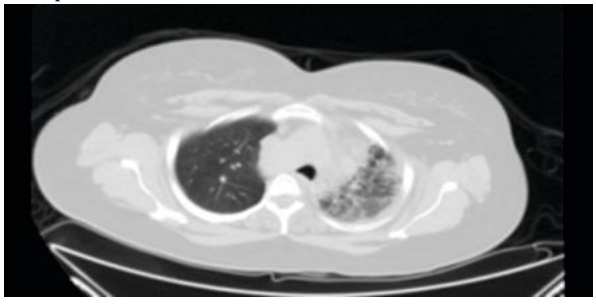
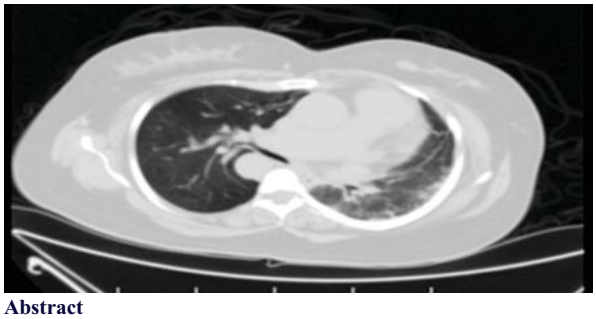


IMAGE 4 – absent left pulmonary artery causing left lung hypoplasia



Abstract

Congenital pulmonary vascular anomalies represent a complex group of developmental abnormalities affecting pulmonary circulation. Among these conditions, unilateral pulmonary artery agenesis (UPAA) is one of the rarest anomalies[1]. UPAA results from failure of development of the proximal segment of the sixth aortic arch during early embryogenesis. This leads to absence of the pulmonary artery on one side, resulting in chronic hypoperfusion of the ipsilateral lung sometimes, may be resulting in a unilateral interstitial lung disease. Rather than complete absence of lung tissue, the affected lung remains present but hypoplastic, relying on systemic collateral vessels for perfusion.

Clinical manifestations vary widely, ranging from neonatal respiratory distress to asymptomatic adults discovered incidentally during imaging performed for unrelated conditions.

Fewer than 500 cases of isolated UPAA have been reported worldwide, emphasizing both its rarity and the challenges associated with early diagnosis.

DISCUSSION

Normal lung development progresses through five stages, and they are embryonic stage, pseudoglandular, canalicular, saccular and the alveolar stage. Pulmonary arterial blood flow stimulates the release of angiogenic factors such as vascular endothelial growth factor (VEGF), which promotes alveolar development. In UPAA, interruption of pulmonary arterial flow during the pseudoglandular and canalicular stages disrupts this vascular-epithelial signaling, resulting in lung hypoplasia. Pulmonary arteries originate from the sixth pair of embryonic aortic arches. During normal development, the truncus arteriosus divides into the pulmonary trunk and ascending aorta, the proximal sixth aortic arches form the right and left pulmonary arteries, and the distal left sixth arch forms the ductus arteriosus. UPAA occurs when the proximal portion of one sixth arch fails to connect with the pulmonary trunk.

UPAA is extremely rare with an estimated prevalence of 1 in 200,000 live births. The right side involvement is about 60-67% and the median age of diagnosis is 14 years with isolated cases are reported in 30% of all and it has no gender predilection. Right-sided agenesis typically occurs in isolation whereas, left-sided agenesis is frequently associated with congenital cardiac anomalies due to its proximity to the aortic arch and ductus arteriosus.

Extra-cardiac anomalies include tracheal stenosis, gastrointestinal abnormalities, and skeletal defects.

The absence of pulmonary arterial perfusion leads to impaired lung growth. Histopathological examination of the hypoplastic lung demonstrates reduced number of alveoli, thickened interalveolar septa, medial hypertrophy of intrapulmonary arteries, abnormal bronchial architecture. These structural abnormalities significantly reduce the effective surface area for gas exchange. In the absence of pulmonary arterial flow, systemic arteries provide compensatory perfusion to the affected lung.

Clinical Manifestations -

Children commonly present with recurrent respiratory infections, exercise intolerance and failure to thrive. Approximately 37% of patients experience recurrent infections. Adults may develop exertional dyspnea, hemoptysis, chest pain and pulmonary hypertension. Approximately 30% of isolated cases remain asymptomatic until adulthood. Typical findings include ipsilateral lung volume loss, associated with ipsilateral interstitial lung disease, as in our case, presence of mediastinal shift toward affected side, absent pulmonary artery shadow and contralateral lung hyperinflation. Pulmonary function testing typically demonstrates a restrictive pattern due to reduced lung volume. Interestingly, diffusion capacity (DLCO) may remain near normal due to compensatory vascular expansion of the contralateral lung. The classical features on chest radiography include a small hemithorax, ipsilateral mediastinal shift, and no identifiable corresponding pulmonary artery[2].

Computed tomography angiography is the gold standard diagnostic modality. CTA provides confirmation of absent pulmonary artery, detailed mapping of systemic collateral vessels and assessment of parenchymal abnormalities. Most of the case, more than 30% usually have a brochiectastasis presentation and the others include ILD and bulla[3]

Echocardiography evaluates pulmonary hypertension, right ventricular function and associated congenital heart defects.

Management depends on patient age and symptom severity. In pediatric patients, revascularization procedures aim to restore pulmonary arterial blood flow to the hypoplastic lung. Early intervention may promote lung growth and reduce the risk of pulmonary hypertension. Medical management in the adult include, management of the complications. Pulmonary hypertension develops in approximately 44% of patients. Common therapies include PDE 5 inhibitors, Endothelin receptor antagonist and soluble guanylate cyclase stimulators. Combination therapy may improve functional capacity and exercise tolerance. The therapeutic approach should be based on symptoms of the patient, pulmonary artery anatomy and associated aortopulmonary collaterals. Treatment options for these patients include partial or total pneumonectomy, closure of selected collateral arteries not solely responsible for pulmonary blood flow or a primary versus staged pulmonary artery anastomosis[4]. Other major complications include pulmonary hypertension, cor-pulmonale, recurrent infections, bronchiectasis and aspergilloma. Aspergillomas may form within bronchiectatic cavities and present with recurrent hemoptysis.

Prognosis depends on presence of pulmonary hypertension, associated congenital anomalies, early diagnosis and treatment. The incidence of pulmonary arterial hypertension has been reported to be between 19% and 25% in different case series. The pathophysiology begins with anterograde blood flow from absent PA to the remaining PA, shearing stress within the endothelium and production of endothelin. This leads to remodeling of pulmonary arterioles and vasoconstriction, resulting in pulmonary hypertension[5]. Overall mortality is estimated to be approximately 7%. Patients without severe pulmonary hypertension may survive well into adulthood with appropriate management. Long-term monitoring is essential.Recommended follow-up includes annual clinical evaluation, periodic echocardiography, imaging if new symptoms occur, functional testing such as the 6-minute walk test.

CONCLUSION

The estimated prevalence of UPAA is 1/200,000 patients[6]. It was first described by Frentzel in 1868, with 30% of patients remained asymptomatic until adulthood. Unilateral pulmonary artery agenesis is a rare congenital anomaly characterized by absence of pulmonary arterial flow, secondary lung hypoplasia, and extensive systemic collateral circulation. Advances in imaging have significantly improved diagnostic accuracy, while modern therapeutic strategies have enhanced survival and quality of life. Early recognition and multidisciplinary care remain critical for optimal management.

Summary:

The estimated prevalence of UPAA is 1/200,000 patients. It was first described by Frentzel in 1868, with 30% of patients remained asymptomatic until adulthood. Unilateral pulmonary artery agenesis is a rare congenital anomaly characterized by absence of pulmonary arterial flow, secondary lung hypoplasia, and extensive systemic collateral circulation. Advances in imaging have significantly improved diagnostic accuracy, while modern therapeutic strategies have enhanced survival and quality of life. Early recognition and multidisciplinary care remain critical for optimal management.

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