Biological and Clinical Sciences Research Journal
eISSN: 2708-2261; pISSN: 2958-4728
www.bcsrj.com
DOI: https://doi.org/10.54112/bcsrj.v2024i1.1135
*Biol. Clin. Sci. Res. J., Volume, **2024**: 1135*
Case Report

CONGENITAL ABSENCE OF PULMONARY ARTERY

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(Received, 04th June 2024, Revised 05th July 2024, Published 30th September 2024)

Abstract: *Isolated unilateral absence of a proximal pulmonary main artery is a rare congenital condition with a prevalence of 1 in 200,000 (1). The first case was reported in 1868, and to date, 350 cases have been reported (2). It usually occurs in conjunction with other cardiovascular anomalies such as tetralogy of Fallot, VSD, truncus arteriosus, etc. (2), and is typically diagnosed in childhood. Survival into adulthood is rare. Symptoms of this condition can include shortness of breath, chest pain, hemoptysis, reduced exercise tolerance, pleural effusion, and recurrent infections. We encountered a case of a 22-year-old female presenting with these symptoms. Initially, the symptoms suggested pulmonary embolism. However, further investigation through CTPA and Echocardiography revealed the absence of the left pulmonary artery. This case report emphasizes that patients showing signs and symptoms indicative of PAH should also be evaluated for the absence of a pulmonary artery." The pulmonary arteries originate from the sixth aortic arch during the sixteenth week of normal embryonic development. The proximal segment of the 6th aortic arch forms the initial portion of the right & left pulmonary arteries, respectively. The link on the arch's right side vanishes, whereas it establishes a connection through the ductus arteriosus on the left side. Pulmonary artery agenesis arises from the aberrant rotation of the afflicted segment's attachment to the aortic arch and dysfunctions during migration (3). Certain publications utilize the phrase "proximal blockage of the pulmonary artery" instead of "agenesis" to describe this anomaly. Although the intraparenchymal distal segment of the pulmonary artery is miniature, it is safeguarded (4). The prevalence of bronchopneumonia is ascribed to multiple causes, such as diminished lung tissue perfusion, impaired mucosal defense, and a possible unbalance of protease-antiprotease enzymes. Moreover, increased collateral blood flow may be associated with the onset of hemoptysis.*

Keywords: Pulmonary artery, congenital anomaly, pulmonary arterial hypertension (PAH)

Case report

A 25-year-old female presented at the emergency room with complaints of shortness of breath, hemoptysis, chest pain, cough, and pain in the right hypochondrium. These symptoms had been ongoing for the last ten years, but her condition had been gradually deteriorating. Upon examination, she was in obvious respiratory distress, with a respiratory rate of 24 breaths per minute. She was tachycardic, pale-looking, and centrally and peripherally cyanosed, with a raised jugular venous pressure. She exhibited grade 4 clubbing. Precordium examination revealed a palpable P2 and left parasternal heave. On auscultation, a pan systolic murmur was heard in the parasternal region on the right side, along with a loud P2. Abdominal examination indicated an increased tender liver span that was moving with respiration (congestive hepatopathy). Her CBC revealed leukocytosis and decreased hemoglobin levels. Viral markers for COVID-19 and Influenza came back negative. An ECG showed tall P waves (P pulmonale), right axis deviation, and right ventricular hypertrophy with strain pattern.

Echocardiography suggested dilated right atrium (RA) and Right ventricle (RV), severe tricuspid regurgitation (TR), and severe pulmonary arterial hypertension (PAH). A chest X-ray indicated pulmonary edema and cardiomegaly, decreased left lung volume, absent hilar markings on the left side, and a raised right hemidiaphragm. Abdominal ultrasound revealed hepatomegaly with dilated intrahepatic channels. A CT pulmonary angiography (CTPA) showed a dilated main pulmonary artery and right pulmonary artery, with an abrupt cut-off of the left pulmonary artery and a hypoplastic left lung, along with some small collaterals arising from the descending thoracic aorta. There was also associated pulmonary edema and hepatomegaly, consistent with right-sided heart overload resulting from the absence of a pulmonary artery. The patient was started on IV antibiotics, diuretics, calcium channel blockers, and endothelin receptor antagonists (bosentan). Her fever was monitored daily, and her oxygen requirement was observed and adjusted accordingly. Her jugular venous pressure (JVP) was also recorded daily. After a few days, her fever subsided, and her symptoms improved. Meanwhile, the pediatric cardiology and cardiothoracic departments were consulted for assistance. The cardiothoracic department

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considered her for pneumonectomy following stabilization.
Reports are as follows.

M-Mode / 2-D Measurements

		Normal			Normal
LVSD (Systolic)	25	mm	LVDD (Diastolic)	38	mm (< 52)
Septal Thickness	09	mm (< 11)	Posterior Wall Thickness	09	mm (< 11)
Left Atrium Diameter	34	mm (< 40)			
RV EDD	40	mm (< 26)			
Ascending Aorta	26	mm (< 36)			

Chambers

Left Ventricle : Normal size left ventricle. Wall thickness is normal. Preserved left ventricular systolic function. Visually estimated ejection fraction is approximately 60 %.

Right Ventricle : Moderately dilated.

Left Atrium : Normal in size.

Right Atrium : Mildly dilated.

LV Diastolic Function: Mitral inflow shows E/A reversal. Left ventricular diastolic dysfunction grade I.

Valve

Mitral : Mild regurgitation.

Tricuspid : Severe regurgitation. PASP is 95 mmHg.

Study Quality: Study quality is adequate.

Pericardium: Pericardium is normal. No pericardial effusion noted.

IAS/IVS: Intact. Intact.

CONCLUSION

CONCLUSION:

DILATED RA / RV

SEVERE TR SEVERE PAH

MILD MR

PRESERVED LV SYSTOLIC FUNCTION.

CPT : CTA PULMONARY WITHOUT CONTRAST

HISTORY : sob, cough, hemoptysis and chest pain.

REPORT : Main pulmonary and right pulmonary artery are dilated measuring 38 and 25.8mm respectively with abrupt cut off of left pulmonary artery.Small collaterals seen from descending thoracic aorta.
Right atrium and ventricle are markedly dilated..
Diffuse ground glass haze with septal thickening is seen involving both lungs with relative sparing of upper lobes along with peripheral pacthy dense alveolar opacities. Bilateral pleural effusion is also seen more so on the right side with associated pericardial effusion.
Included sections of abdomen show hepatosplenomegaly with dilated hepatic veins and Inferior vena cava.

CONCLUSION : Appearances are in keeping with left pulmonary artery occlusion with resultant pulmonary arterial hypertension ,cardiomegally and bilateral pulmonary edema with superimposed infection and its sequelae .

Discussion

The solitary absence of the pulmonary artery was initially documented by Fraentzel in 18681 (1) (2, 5). This illness lacks a genetic component and does not favor a particular sex. The condition typically presents in childhood and is detected at an early age; nevertheless, it has been identified in persons as old as 68. The pulmonary arteries originate from the sixth aortic arch during the sixteenth week of typical embryological development. The proximal segment of the sixth aortic arch forms the initial portion of both the Right & left pulmonary arteries, respectively. The link on the arch's right side vanishes, whereas it establishes an association through the ductus arteriosus on the left side. Pulmonary artery agenesis arises from the aberrant rotation of the afflicted segment's attachment to the aortic arch and dysfunctions during migration (3). Certain publications use the phrase “proximal disruption of the pulmonary artery” instead of “agenesis” to describe this anomaly. Although the intraparenchymal distal segment of the pulmonary artery is miniature, it is safeguarded (4). While around 30% of patients with pulmonary artery agenesis can stay

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asymptomatic throughout their lives, they may exhibit decreased exercise tolerance, congestive heart failure, chronic bronchiectasis, severe necrotizing pneumonia, and significant pulmonary arterial hypertension, as noted by Harkel et al. (6). The etiology is complex. Reduced blood flow in the afflicted lung inhibits the release of appropriate pro-inflammatory cells at the site of inflammation and results in the impairment of ciliary function—furthermore, reduced blood flow results in alveolar hypocapnia and subsequent bronchoconstriction. Chronic infection ultimately leads to bronchiectasis (7, 8).

Shakibi et al. (9). A hemoptysis prevalence of 18–20% was observed. In the compromised lung, pulmonary arteries terminate abruptly at the hilum, with the primary blood supply derived from the primary bronchial arteries, along with intercostal, internal mammary, subclavian, and transpleural branches from the innominate arteries. Haemoptysis transpires when the hypertrophic ectatic collateral circulation hemorrhages into the bronchioles.

A physical examination may reveal a diminished hemithorax with attenuated breath sounds, displacement of the heart & mediastinum, or indicators of pulmonary hypertension. The ECG is typically normal but may have a right-axis deviation. The chest X-ray findings indicate diminished blood flow to a single lung, an absence of density within the central thoracic region, displacement for the heart as well as the mediastinum, a constricted hemithorax with narrowed intercostal spaces, an elevated diaphragm, and a reticular pattern suggesting abnormalities in the bronchial arteries. The perfusion and ventilation scans will demonstrate diminished blood flow to one lung despite normal breathing levels. Pulmonary angiocardiography reveals nonexistent circulatory structures, while aortography may indicate an abnormal blood flow to the diseased lung.

The left and right pulmonary arteries are missing, with roughly similar prevalence. The absence of the left pulmonary artery is predominantly linked to tetralogy of Fallot and may be coupled with a right-sided aortic arch. Unilaterally, the absence of the right pulmonary artery occurs in roughly 50% of instances and is most frequently associated with patent ductus arteriosus when accompanied by an abnormality. Defects linked to the lack of either artery encompass atrial, including ventricular septal defects, coarctation of the aorta, aorticopulmonary window, truncus arteriosus, and in one case, supra cardiac complete anomalous pulmonary venous return.

There is a lack of unanimity on treatment. It is recommended that asymptomatic cases have intermittent ECHO monitoring for the progression of pulmonary hypertension. Should pulmonary hypertension manifest, the prognosis is unfavorable, resulting in significant dyspnoea, weight reduction, and limited physical activity. In the event of right heart failure, signs of insufficiency such as peripheral edema, jugular venous distension, as well as acidosis become prominent. Revascularisation utilizing the artery on the hilus of a side exhibiting arterial agenesis may be a viable therapy option in select circumstances.

The literature study indicated a greater prevalence of solitary absence of both the right and left pulmonary arteries that was previously documented. This may be attributed to the fact that numerous cases were identified in adults, who likely exhibited milder symptoms. Nonetheless, a significant correlation has been demonstrated between the left pulmonary artery's absence and various bulbus and cordis anomalies, including tetralogy of Fallot as truncus arteriosus. A case was published in the American Journal of Medical Sciences by Jesse Roman, MD, et al. (10). Suggests a congenital absence of the left pulmonary artery coupled with ipsilateral emphysema and cancer.

Conclusion

Timely diagnosis of pulmonary artery agenesis is essential, as it may result in severe consequences and mortality. In the presence of common radiological indicators such as necrotizing pneumonia, hemoptysis, pulmonary hypertension, right heart failure, and bronchiectasis, one should suspect pulmonary artery agenesis and utilize suitable radiological diagnostic techniques.

Declarations

Data Availability statement
All data generated or analyzed during the study are included in the manuscript.
Ethics approval and consent to participate
Approved by the department concerned.
Consent for publication
Approved
Funding
Not applicable

Conflict of interest

The authors declared the absence of a conflict of interest.

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