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Isolated unilateral absence of pulmonary artery- an unusual cause of hemoptysis: a case report

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Abstract

Background Isolated unilateral absence of one pulmonary artery (IUAPA) is a rare malformation and a rare cause of hemoptysis. It occurs due to malformation of the sixth aortic arch during embryogenesis. This condition has variable presentations like dyspnoea, reduced exercise tolerance, chest pain, pleural effusion, recurrent bronchopneumonia, pulmonary hypertension, and haemoptysis can be asymptomatic.

Case Presentation We describe a case of a 34-year-old female presenting with recent onset hemoptysis associated with mild dyspnoea and cough. Chest Radiograph revealed unilateral hyperlucent lung field which was later diagnosed as Isolated unilateral absence of pulmonary artery on high resolution computed tomography of chest with angiography.

Conclusion Absent pulmonary artery should be considered as an important differential in cases presenting with hemoptysis. This case emphasises the significance of combining computed tomography with pulmonary angiography to accurately evaluate and define congenital lung abnormalities.

Keywords Collaterals, Hemoptysis, IUAPA, Pulmonary artery

Background

Congenital absence of unilateral pulmonary artery is a rare malformation, with an estimated frequency of 1 in 200,000, [1]. It occurs due to malformation of the sixth aortic arch during embryogenesis. It may occur in isolation or in conjunction with other anomalies. This condition has variable presentations like dyspnoea, reduced exercise tolerance, chest pain, pleural effusion, recurrent bronchopneumonia, pulmonary hypertension, and haemoptysis can be asymptomatic. Delay in the presentation in adults has been recorded despite being primarily a congenital abnormality. We herein report a case of a young female with isolated unilateral absence of pulmonary artery who presented with haemoptysis.

Case presentation

A 34-year-old female presented with haemoptysis for nine days which was also associated with recent onset mild dyspnoea and cough. She had moderate haemoptysis about 80–100 ml of fresh blood per day (2–3 episodes per day). There was no history of any fever, chest, night sweats, orthopnoea, palpitations, trauma, weight loss or lymphadenopathy. She did not have a history of tuberculosis or any other comorbid illnesses. No relevant family history of significant cardiovascular events. There was no history of smoking, alcohol abuse or illicit drug use.

Her vitals were stable on presentation and maintained normal saturation at room air with normal blood

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pressure and her systemic examination revealed no significant finding.

For further evaluation, a chest X-ray was performed for the same. Chest x-ray revealed mild pulmonary asymmetry. The right hemithorax appeared smaller than the left side with mild tracheal and mediastinal deviation to the right with elevation of the right hemidiaphragm. Further, there were decreased vascular markings in the right lung and reduced radiolucency with absent hilar shadow on the right side. A hyperlucent and hyperinflated left lung was also noted (Fig. 1).

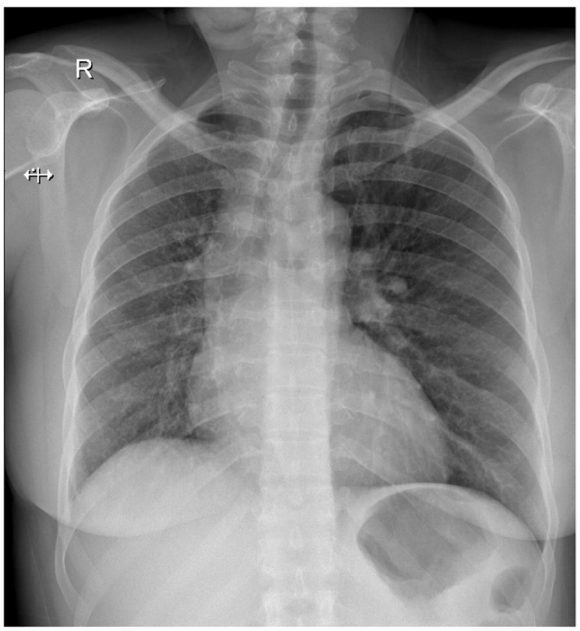

Fig. 1 Radiograph chest PA view showing smaller right hemithorax, with mild tracheal and mediastinal deviation to right and elevation of hemidiaphragm. Mild diffuse haziness and decreased vascular markings over right lung zones. The right hilum is not well visualized

Due to the unspecific nature of the findings on the chest X-ray, the patient was further subjected to multidetector CT (MDCT) chest with bronchial and pulmonary angiography. On plain nonenhanced lung window (Fig. 2), the right lung appeared hypoplastic with a mid mediastinal and tracheal shift towards the right. The left lung showed compensatory hyperinflation (Fig. 2 A, B). Apart from this septal thickening was noted in the right upper lobe and the right oblique fissure shows mild thickening and para septal emphysema (Fig. 2C). Centrilobular ground glass densities were noted in all lung segments. Patchy ground glass density in anterior and medial basal segments of the right lower lobe. These are likely due to hemoptysis.

On MDCT chest with bronchial and pulmonary angiography, main pulmonary trunk measures 2.3 cm, left pulmonary artery measured 2.1 cm with non-visualization of the right pulmonary artery (Fig. 3). In place of the right main pulmonary artery, there were multiple dilated tortuous bronchial arteries encasing the right main stem bronchus (Fig. 4). The right internal thoracic artery and right subclavian artery were also dilated (Fig. 5 A, B). Posterior intercostal arteries in right mid and lower thorax were also prominent as demonstrated in the reconstructed MIP images (Fig. 5 C).

Variant origin of inferior phrenic arteries was seen arising from the aorta just inferior to the celiac artery coursing to the right side along the undersurface of the right diaphragm (Fig. 5 D). Bilateral superior and inferior pulmonary veins however appeared normal. Based on the above-observed findings a diagnosis of isolated unilateral absence of pulmonary artery (IUAPA) was made. 2D echocardiography revealed trivial tricuspid regurgitation and mild pulmonary artery hypertension.Normal left ventricular size and systolic function. Left ventricular ejection fraction- 60%, no regional wall

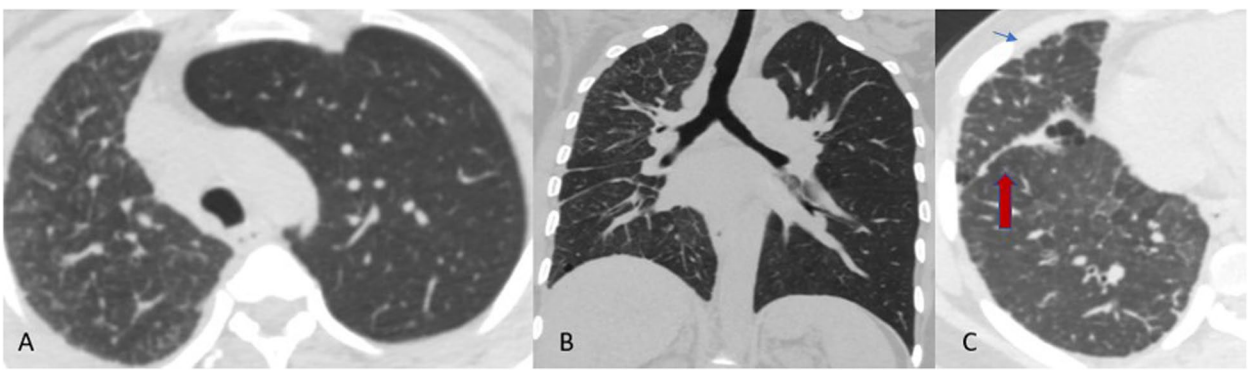

Fig. 2 HRCT thorax (A-axial, B-coronal and C-axial) Lung window reveals small-sized right hemithorax, compensatory overinflation of left lung with mild mediastinal and tracheal shift towards right and elevation of right hemi diaphragm, thickening of right oblique fissure (marked by red arrow) and septal thickening (marked by small blue arrow) and centrilobular ground glass densities in right lung

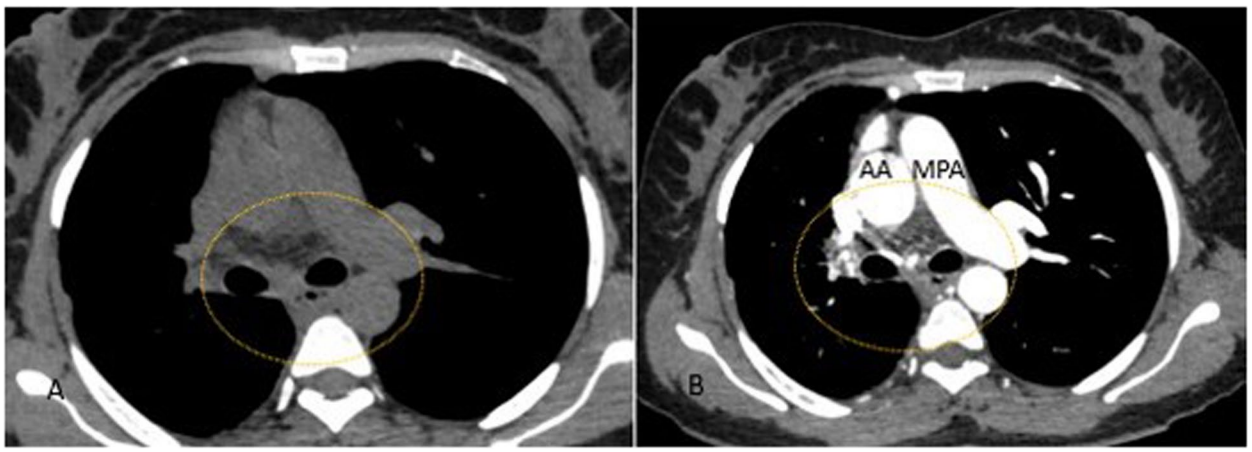

Fig. 3 CT thorax mediastinal window (A- plain study, B- CT Angiography) shows presence of the Main pulmonary trunk and left pulmonary artery and absence of right pulmonary artery. [AA- Ascending aorta, MPA- Main Pulmonary artery]

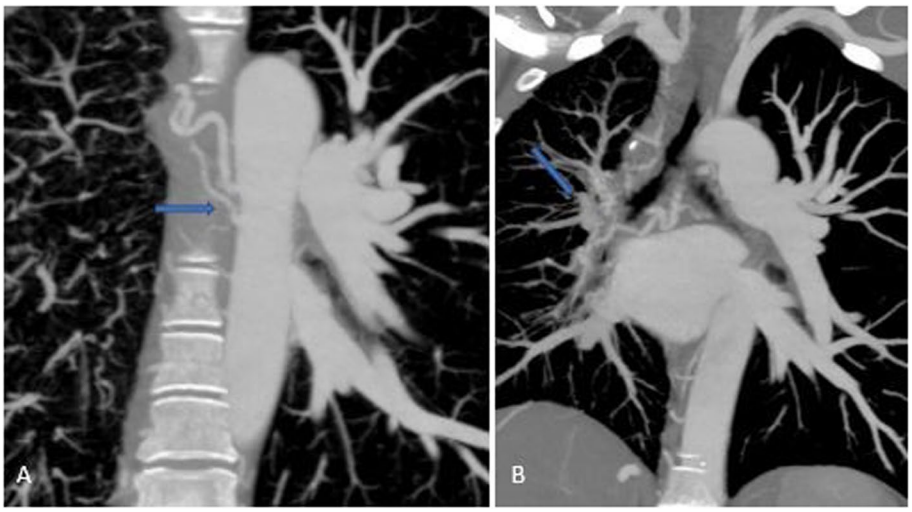

Fig. 4 CT Angiography MIP images coronal view (A, B) showing dilated and tortuous right bronchial arteries arising from the aorta (marked by the blue arrow)

motion abnormality, normal left ventricular diastolic function. No clot/vegetation/pericardial effusion. Pulmonary pressure gradient- 04 mmHg, Aortic pressure gradient-05 mmHg and Tricuspid right ventricular systolic pressure – 33 mmHg.

The patient was advised of bronchial artery embolization which she refused at the time of admission hence supportive treatment was given and the patient was discharged.

Discussion

Frantzel O. Angeborener first characterised unilateral absence of pulmonary artery (UAPA) in 1868, and Madoff and his colleagues later demonstrated it angiographically in 1953 [2].

This rare congenital anomaly occurs due to the persistence of the connection of the intrapulmonary pulmonary artery to the distal sixth aortic arch and involution of the proximal sixth aortic arch. It is also known as pulmonary artery proximal interruption, nonconfluent pulmonary artery or ductal origin of the distal pulmonary artery [3].

The proximal pulmonary artery is absent in isolated unilateral agenesis of pulmonary artery. However, the distal intrapulmonary branches are usually intact. These are supplied by collateral vessels arising from subclavian, internal mammary, bronchial, intercostal, coronary arteries, and subdiaphragmatic arteries [4].

The condition is more prevalent on the right side though there is no preference for the right or left side.

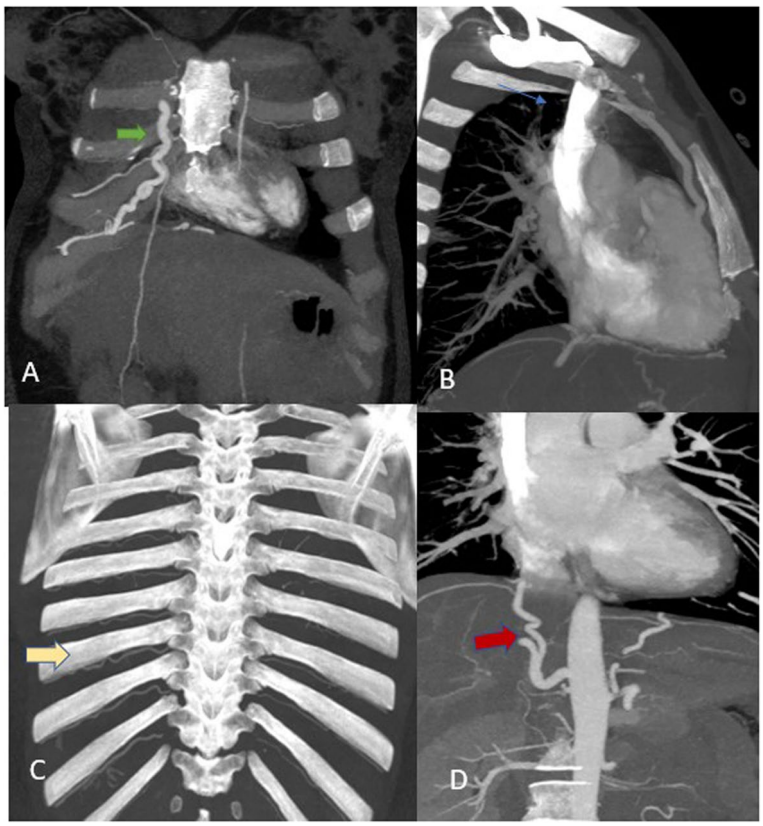

Fig. 5 CT Angiography MIP images (A- coronal, B- sagittal, C-D coronal) showing dilated right internal mammary artery (marked by green arrow), dilated right subclavian artery (marked by blue arrow), dilated posterior intercostal arteries (marked by yellow arrow) along posterior chest wall in right hemithorax and dilated inferior phrenic artery (marked by red arrow)

Left sided pulmonary agenesis is usually associated with cardiac anomalies [5]. Cardiovascular anomalies associated are right sided aortic arch, persistence of ductus arteriosus, septal defects, and tetralogy of Fallot. These are commonly detected in children. It seldom occurs alone and is detected in adults [6].

Patients with absent pulmonary artery can have variable presenting symptoms like dyspnoea, reduced exercise tolerance, chest pain, pleural effusion, recurrent bronchopneumonia, pulmonary hypertension and haemoptysis however the condition can be asymptomatic in up to 15% of cases [1].

Pulmonary hypertension occurs from blood flow directed away from the absent pulmonary artery to the contralateral pulmonary artery. This results in increased blood flow in the contralateral pulmonary artery. As a result, there is sheer stress on the endothelium with the release of vasoconstrictive substances like endothelin. These cause chronic vasoconstriction of the pulmonary arterioles, increased resistance in the pulmonary vasculature and hence pulmonary hypertension [4].

In our patient the major presenting symptom was haemoptysis which indeed can occur in up to 20% of cases, while it is usually self-limiting, it can occasionally lead to significant pulmonary hemorrhage and mortality [7]. Hemoptysis usually occurs due to expansion of collaterals from bronchial, intercostals, internal thoracic and subdiaphragmatic arteries with time supplying the affected lung with eventual rupture [8].

Radiological imaging plays a key role in the diagnosis of unilateral pulmonary artery agenesis. Radiograph findings characteristic of this condition is small hemithorax showing hyperlucent lung with reduced hilar vasculature, contralateral lung hyperinflation which may appear plethoric due to increased blood flow, elevated ipsilateral hemi diaphragm, loss of hilar shadow with cardiac and mediastinal displacement. Pulmonary angiography is considered the gold standard for the diagnosis of IUAPA [9].

Termination of right or left pulmonary artery within 2 cm of its expected origin from the main pulmonary trunk on CT angiography is diagnostic of this entity. The affected lung receives blood supply from systemic

collaterals. These systemic collaterals arise from subclavian, internal thoracic, bronchial, intercostal and subdiaphragmatic and rarely coronary arteries. With time, these collaterals hypertrophy and may rupture resulting in massive hemoptysis [10]. Lung parenchymal findings can occur due to long standing pulmonary hypoperfusion and dynamic changes in the pulmonary blood flow. These include interstitial changes like septal thickening, honeycombing, bronchiectasis, and cystic spaces in the affected lung. Recurrent respiratory infections can occur due to bronchiectasis in the affected lung. Alveolar hypocapnia can lead to bronchial constriction and impairment of bronchial mucociliary clearance. This in turn results in impaired delivery of blood bone inflammatory cells. As a result, mucous trapping, chronic bronchitis, and bronchiectasis can occur.

Various treatment options available are selective embolization of collateral artery (SECA), anti pulmonary hypertension medication and pneumonectomy. SECA is less successful in adult patients with isolated UAPA in controlling hemoptysis. Recurrent hemoptysis can occur after embolization within a week. This occurs due to failure of occlusion of all abnormal vessels during embolization [11]. Pneumonectomy is useful in cases of recurrent massive hemoptysis and refractory respiratory infections.

Conclusion

Isolated unilateral absence of one pulmonary artery is an unusual cause of hemoptysis. A high index of suspicion is needed for its diagnosis on a chest radiograph. 2D echocardiography can detect pulmonary hypertension and rule out associated cardiac abnormalities. Multidetector computed tomography of the chest with bronchial and pulmonary angiography can accurately diagnose this condition. Long-term outcomes are improved by early diagnosis, supportive care, and more modern surgical and interventional methods.

Abbreviations

IUAPA	Isolated unilateral absence of one pulmonary artery
CT	Computed Tomography
CECT	Contrast-enhanced computed tomography
UAPA	Unilateral absence of pulmonary artery
SECA	Selective embolization of collateral artery
MIP	Maximum intensity projection
MDCT	Multidetector computed tomography

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Authors' contributions

SMK and DK analysed and interpreted the patient data regarding the clinical history and the imaging modality. SMK, DK and RK were major contributors in data collection, literature search and writing of the manuscript. All authors have read and approved the manuscript.

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Availability of data and materials

The case file and diagnostic reports with relevant details are available in the medical record section of our institute.

Declarations

Ethics approval and consent to participate

Our institution does not require ethics approval for reporting individual case reports.

Consent for publication

The authors certify that written informed consent was obtained from the patient for the publication of this case report, and accompanying images.

Competing interests

The authors declare that they have no competing interests.

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