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Right Pulmonary Artery Agenesis Revealed by Chest CT: A Case Report

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Abstract | **Case Report**

Background: Unilateral pulmonary artery agenesis (UPAA) is a rare congenital vascular anomaly resulting from an embryologic failure of the sixth aortic arch to connect with the pulmonary trunk. Its clinical presentation is variable, ranging from severe respiratory symptoms in childhood to incidental discovery in asymptomatic adults. We report the fortuitous detection of this condition in an adult male who presented with grade II dyspnea and chest pain lasting for one week**.** Contrast-enhanced chest CT revealed absence of the right pulmonary artery with ipsilateral pulmonary hypoplasia. This case underscores the key role of CT scan in identifying rare vascular anomalies, even in mildly symptomatic patients.

Keywords: Right pulmonary artery agenesis; Congenital vascular anomaly; Pulmonary hypoplasia; CT scan; Chest imaging.

INTRODUCTION

The unilateral agenesis of a pulmonary artery represents an exceptionally rare congenital malformation, with a reported prevalence of about 1 per 200,000 individuals. [1]. This vascular anomaly was originally described by Frentzel in 1868 [2].

Although unilateral pulmonary artery agenesis can occur as an isolated defect, it is more commonly accompanied by various cardiovascular anomalies [3,4].

CASE PRESENTATION

A 40-year-old male presented with grade II dyspnea and a one-week history of chest pain. He had no significant past medical history and no history of cardiovascular disease. Physical examination revealed normal vital signs, with slightly reduced breath sounds over the right lung.

A contrast-enhanced chest CT scan was performed. Imaging revealed absence of the right pulmonary artery (Figure 1), associated with significant mediastinal and subpleural collateral circulation arising from the bronchointercostal trunk, right internal mammary and phrenic arteries, as well as a subdiaphragmatic collateral originating near the origin of the right renal artery. There was also hypoplasia of the right lung, which demonstrated areas of perfusion abnormalities. No associated cardiac anomalies were identified.

The findings were consistent with right pulmonary artery agenesis with extensive compensatory collateral circulation. The patient was advised regular clinical and functional follow-up to monitor respiratory status and overall pulmonary function.

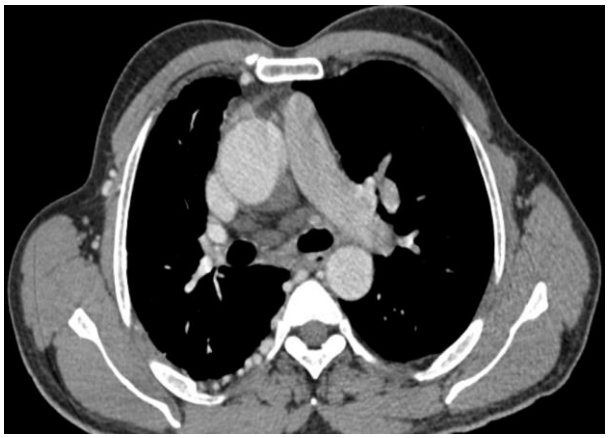

Figure 1: Axial contrast-enhanced chest CT demonstrates absence of the right pulmonary artery.

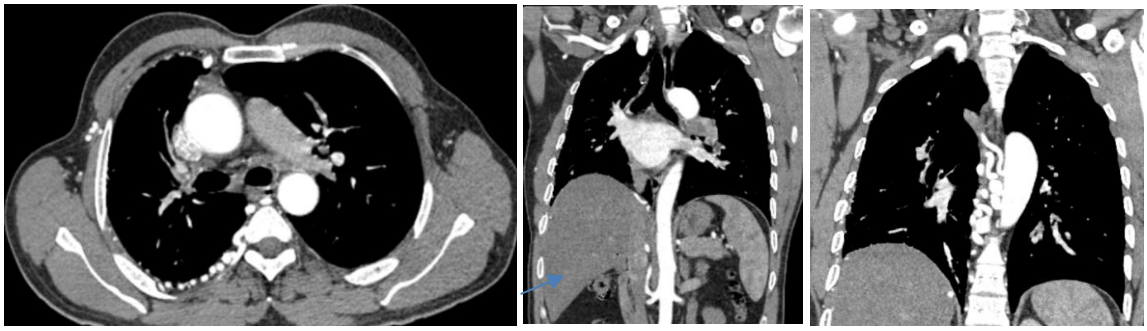

Figure 2: Contrast-enhanced chest CT in axial and coronal planes demonstrates mediastinal and subpleural collateral circulation arising from the right internal mammary artery, the bronchointercostal trunk, and a subdiaphragmatic collateral originating near the origin of the right renal artery (arrow)

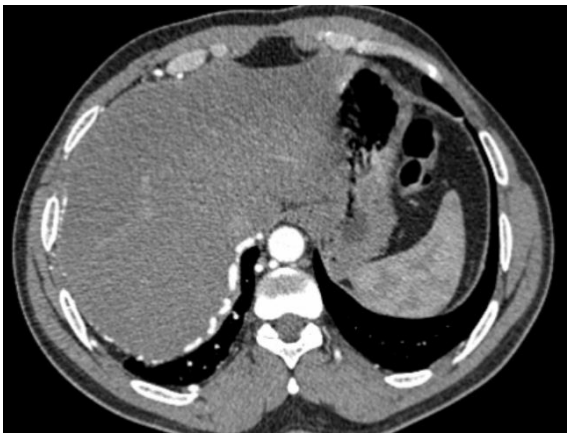

Figure 3: Axial contrast-enhanced chest CT showing collateral circulation arising from the right inferior phrenic artery

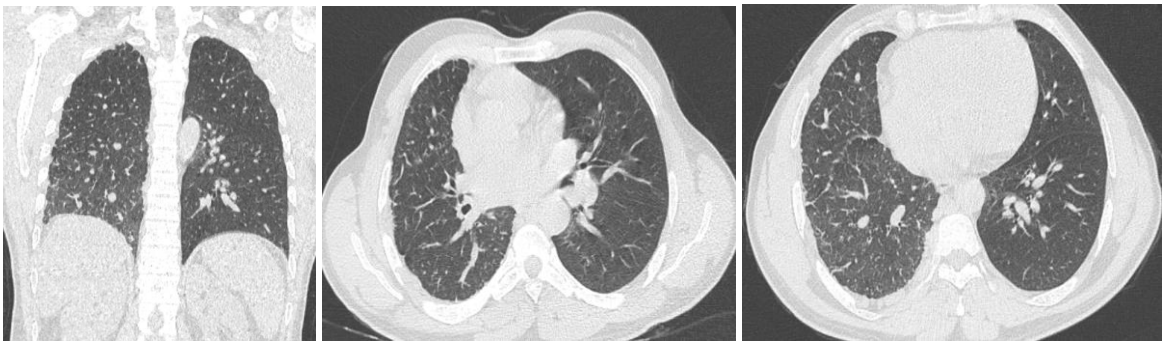

Figure 4: Chest CT in the parenchymal window, axial and coronal planes, showing reduced volume of the right lung with thickening of septal and non-septal lines, associated with perfusion abnormalities

DISCUSSION

Unilateral pulmonary artery agenesis constitutes a rare congenital anomaly, representing only approximately 1% of congenital heart malformations [5].

Embryologically, pulmonary artery agenesis results from a failure of migration and rotation of the primitive sixth aortic arch. Right-sided agenesis is generally not associated with cardiac anomalies and is therefore considered an isolated form of unilateral pulmonary artery agenesis [6,7].

In contrast, left-sided agenesis is frequently accompanied by congenital cardiac abnormalities, making it a non-isolated variant and often necessitating early diagnosis and surgical correction during childhood [6,7].

Congenital pulmonary artery agenesis accompanied by pulmonary hypoplasia has been attributed to a range of factors, including chromosomal anomalies, maternal vitamin A deficiency, intra-uterine infections, and environmental exposures [8].

The clinical presentation of pulmonary artery agenesis is highly variable. Most patients present with recurrent pulmonary infections, reduced exercise tolerance, and exertional dyspnea. Hemoptysis and signs

of pulmonary hypertension are observed in approximately 20% of cases [3].

Pulmonary hypertension commonly emerges early in the disease course and is identified in up to 25% of patients at the time of initial assessment. Its presence constitutes a significant adverse prognostic marker, being strongly associated with reduced long-term survival [5].

Hemoptysis in patients with unilateral pulmonary artery agenesis is attributed to the development of extensive systemic collateral circulation.

Notably, 13–30% of patients may remain asymptomatic, leading a benign course and only being diagnosed in adulthood [3, 11].

Multiple diagnostic modalities are therefore frequently required to establish a definitive diagnosis. On chest radiography, variable features may be observed, including absence of the hilar shadow, reduced pulmonary vascular markings, a small ipsilateral hemithorax, and displacement of the heart and mediastinum toward the affected side, often accompanied by elevation of the ipsilateral hemidiaphragm [12]. Echocardiography can assist in establishing the diagnosis and in evaluating the presence and severity of associated pulmonary hypertension. Both CT and MRI provide detailed anatomical characterization of unilateral pulmonary artery agenesis.

CT angiography remains the gold-standard modality and the preferred technique for confirming the diagnosis of unilateral pulmonary artery agenesis. It also enables detailed visualization of collateral vessels originating from the descending thoracic aorta via bronchial, intercostal, and subdiaphragmatic branches, as well as from the subclavian or coronary arteries. In most cases, the affected lung is predominantly perfused by the bronchial arteries [13].

The radiographic differential diagnosis of pulmonary artery agenesis includes chronic pulmonary thromboembolism, Swyer-James syndrome, large-vessel vasculitis (such as Takayasu arteritis or giant cell arteritis), Behçet's disease, lobar atelectasis, post-lobectomy status, and pulmonary agenesis. Among these, the exclusion of unilateral chronic thromboembolism is particularly critical in adults, as it can also present with ipsilateral pulmonary artery stenosis and the development of collateral circulation [14].

In terms of management, asymptomatic patients typically do not require any intervention. However, those presenting with recurrent hemoptysis or pulmonary infections may necessitate surgical treatment, such as lobectomy or pneumonectomy [15].

CONCLUSION

Right pulmonary artery agenesis is an uncommon congenital anomaly that may remain clinically silent and be discovered incidentally in adulthood. Contrast-enhanced chest CT plays a crucial role in establishing the diagnosis and assessing associated pulmonary and vascular changes. Awareness of this rare entity is essential for accurate interpretation of chest imaging and appropriate patient management.

REFERENCES

1. Kruzliak P, Syamasundar RP, Novak M, Kovacova G, Hlubocká Z, Varga I, et al. Unilateral absence of pulmonary artery: pathophysiology, symptoms, diagnosis and current treatment. *Arch Cardiovasc Dis.* 2013; 106:448–54. doi: 10.1016/j.acvd.2013.05.004.
2. Fraentzel O. Ein Fall von anormer Communication der aorta mit der arteria pulmonalis. *Arch Pathol Anat.* 1868;43(3):420–26. doi:10.1007/BF01915623.
3. Ten Harkel AD, Blom NA, Ottenkamp J. Absence unilatérale isolée d'une artère pulmonaire. Un rapport de cas et une revue de la littérature. *Chest.* 2002; 122:1471–7. doi:10.1378/chest.122.4.1471.
4. Welch K, Hanley F, Johnston T, Cailes C, Shah MJ. Absence unilatérale isolée de l'artère pulmonaire proximale droite : réparation chirurgicale et suivi. *Ann Thorac Surg.* 2005 ;79:1399–402. doi: 10.1016/j.athoracsur.2003.10.037.
5. Griffin N, Hamilton M, et al. Imaging features of isolated unilateral pulmonary artery agenesis presenting in adulthood: a review of four cases. *Clin Radiol.* 2007.
6. Reading DW, Oza U. Unilateral absence of a pulmonary artery: a rare disorder with variable presentation. *Proc (Bayl Univ Med Cent).* 2012; 25:115–8. doi:10.1080/08998280.2012.11928802.
7. Komatsu Y, Hanaoka M, Ito M, et al. Unilateral absence of the pulmonary artery incidentally found after an episode of hemoptysis. *Intern Med.* 2007; 46:1805–8. doi:10.2169/internalmedicine.46.0257.
8. Sunam G, Ceran S. Pulmonary artery agenesis and lung hypoplasia. *Eur J Gen Med.* 2009; 6:265–7.
9. Chaudhry A, Rathore M, Banavaliker JN. Isolated right pulmonary artery agenesis with agenesis of right upper lobe and bronchiectasis of right lower lobe with anomalous arterial supply from celiac axis with normal venous drainage. *Indian J Chest Dis Allied Sci.* 2014; 56:49–52.
10. Shakibi JG, Rastan H, Nazarian I, Paydar M, Aryanpour I, Siassi B. Isolated unilateral absence of the pulmonary artery: review of the world literature and guidelines for surgical repair. *Jpn Heart J.*
11. Bouros D, Pare P, Panagou P, Tsintiris K, Siafakas N. The varied manifestation of pulmonary artery agenesis in adulthood. *Chest.* 1995; 108:670–6.
12. Catala FJ, Marti-Bonmati L, Morales-Marin P. Proximal absence of the right pulmonary artery in

the adult: computed tomography and magnetic resonance findings. *J Thorac Imaging.*

13. Hayek H, Palomino J, Thammasitboon S. Right pulmonary artery agenesis presenting with uncontrolled asthma in an adult: a case report. *J Med Case Rep.* 2011; 5:353.
14. Simonneau G, Torbicki A, Dorfmüller P, Kim N. The pathophysiology of chronic thromboembolic pulmonary hypertension. *Eur Respir Rev.* 2017; 26:160112.
15. Jariwala P, Maturu VN, Christopher J, Jadhav KP. Congenital isolated unilateral agenesis of pulmonary arteries in adults: case series and review. *Indian J Thorac Cardiovasc Surg.* 2021;37(Suppl 1):144–54.