

Case Report

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A rare vascular combination in an adult: Hypoplasia of the right pulmonary artery and veins

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Abstract

Combined unilateral hypoplasia of the pulmonary artery and veins is an extremely rare congenital anomaly, mostly described in children and exceptionally seen in adults. It is usually manifested by ipsilateral lung hypoplasia, reduced lung vascularization, and compensatory contralateral vessel hyperplasia. Contrast-enhanced Computed Tomography (CT) typically reveals a reduction in pulmonary artery caliber, absent or hypoplastic pulmonary veins, and lung parenchymal changes such as interlobular septal thickening and ground-glass attenuation. Patients may present with recurrent respiratory infections or remain asymptomatic until adulthood. We report a case of an adult patient with combined hypoplasia of the right pulmonary artery and veins, diagnosed incidentally during an evaluation for chronic respiratory symptoms. Angio-CT confirmed the diagnosis. This case highlights the importance of recognizing such rare vascular anomalies to ensure accurate diagnosis and appropriate clinical management.

Keywords: Pulmonary artery hypoplasia; Pulmonary vein hypoplasia; CT angiography; Congenital malformation.

Introduction

Unilateral hypoplasia of the pulmonary artery and veins is an extremely rare congenital anomaly caused by embryologic developmental failure [1]. This condition often leads to ipsilateral lung fibrosis [2,3]. While isolated hypoplasia of the pulmonary artery or veins has been more frequently reported, the combination of both anomalies is exceptionally uncommon, particularly in adults [4,5].

Clinically, patients may present with nonspecific symptoms that can mimic other pulmonary conditions such as tuberculosis or interstitial lung diseases [1]. Imaging, especially angio-CT, is essential for the diagnosis, showing both vascular and parenchymal abnormalities. A thorough analysis is necessary to eliminate differential diagnoses such as chronic thromboembolic pulmonary hypertension, Scimitar syndrome, and fibrosing mediastinitis [4].

This report highlights a case of combined hypoplasia of the right pulmonary artery and veins diagnosed incidentally during the evaluation of chronic respiratory symptoms. It aims to underline the clinical and imaging features crucial for identifying such rare anomalies and preventing diagnostic delays.

Case report

A 58-year-old female presented with chronic dry cough and progressive dyspnea over the past five years. The patient reported recurrent respiratory infections since early adulthood, treated with antibiotics. There was no history of hemoptysis, chest pain, or pulmonary embolism.

On physical examination, the patient was afebrile, and her oxygen saturation was normal. Laboratory tests, including a complete blood count and inflammatory markers, were within normal limits. Sputum analysis for tuberculosis was negative.

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Initial chest CT showed reduced right lung size with interlobular reticulation (Figure 1). A follow-up non-contrast thoracic CT demonstrated signs of diffuse subpleural and basal fibrosis in the right lung, characterized by interlobular septal thickening, traction bronchiectasis, and subpleural honeycombing (Figure 2). The diagnosis of Usual Interstitial Pneumonia (UIP) was considered.

Due to persistent symptoms despite symptomatic treatment, a contrast-enhanced CT was performed. The angio-CT revealed the following:

- The main pulmonary artery and left pulmonary artery were normally opacified.
- The right pulmonary artery was only opacified at its origin, with an abrupt cutoff and complete absence of distal branches (Figure 3).
- Hypoplasia of the right pulmonary veins was observed. The right posterior pulmonary vein measured 3 mm near its insertion into the left atrium, while the right anterior vein was visualized only at its insertion, measuring 2 mm in diameter (Figure 4).
- Significant collateral circulation from hypertrophied bronchial arteries and systemic veins was noted. Diffuse tracheobronchial calcifications and small mediastinal lymph nodes were also identified. 3D reconstructions provided a comprehensive overview of the vascular anomalies (Figure 5).
- The findings suggested unilateral pulmonary fibrosis secondary to vascular hypoplasia. A multidisciplinary approach involving pulmonology and cardiology was adopted. Respiratory function tests revealed a restrictive ventilatory pattern. The patient's significant collateral circulation explained the delayed onset of symptoms.

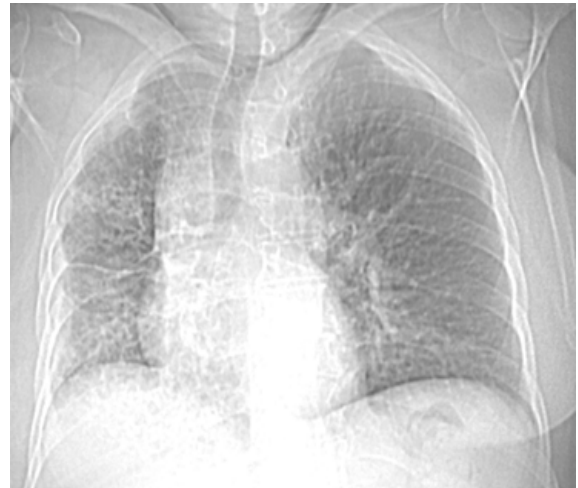


Figure 1: Frontal chest X-ray showing an asymmetry in size and opacity between the two lungs, with a retracted right lung displaying scattered reticulations.

Discussion

The simultaneous hypoplasia of the right pulmonary artery and veins is an exceptionally rare condition, particularly in adults. It results from embryologic developmental failure. The right pulmonary artery originates from the sixth aortic arch during embryogenesis [2,5]. Improper separation of the truncus arteriosus can cause hypoplasia or interruption of the artery. Hypoplasia of the pulmonary veins is due to incomplete incorporation of the common pulmonary vein into the left atrium [8].

Radiological evaluation is essential for diagnosis. Chest radiographs often show asymmetry in lung volumes, with the affected side exhibiting volume loss and mediastinal shift. Angio-CT is the gold standard for evaluating vascular anomalies [6].

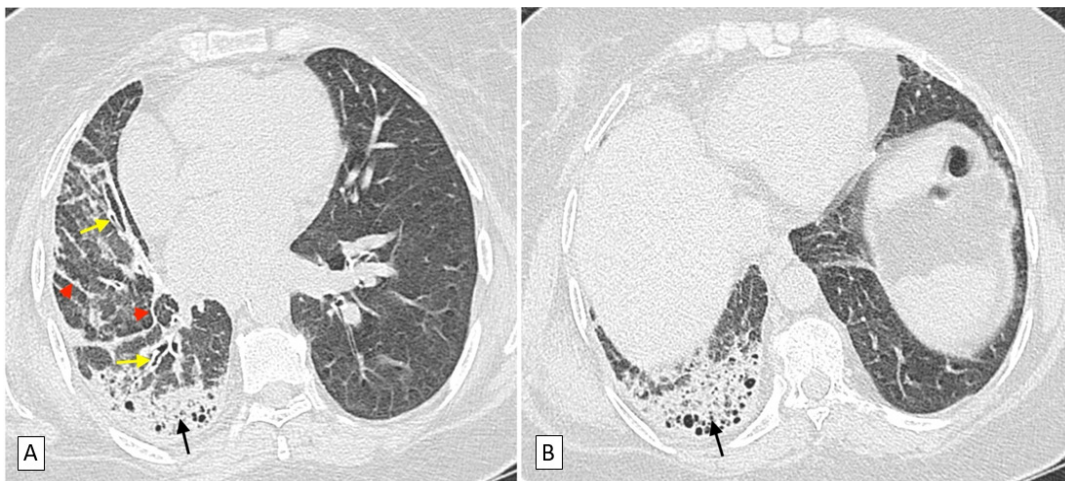


Figure 2: Thoracic CT scan, axial slices in lung window. (A) Interstitial infiltrate with interlobular septal thickening (red arrowheads), associated with traction bronchiectasis (yellow arrows), and subpleural cystic images forming a honeycombing pattern (black arrow). (B) This pattern is more pronounced in the basal regions (black arrow).

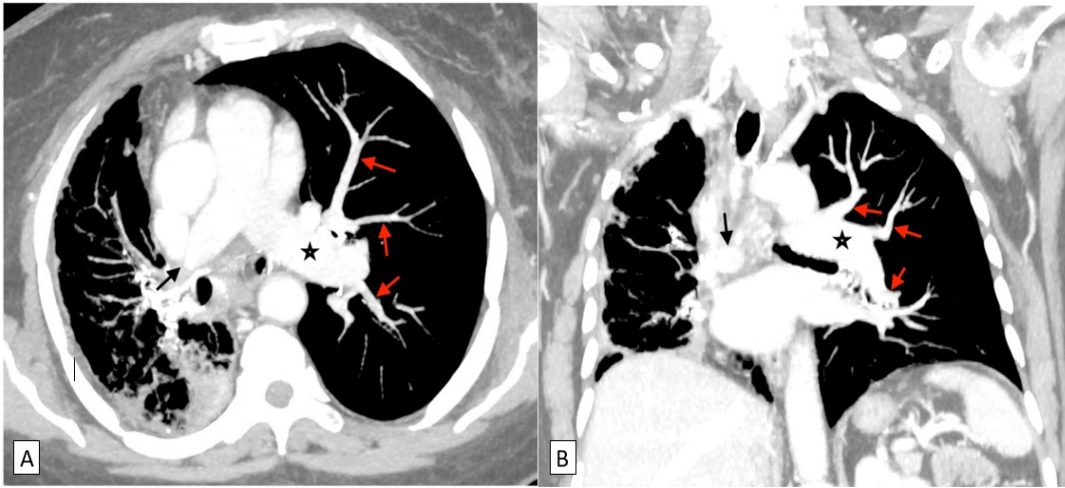


Figure 3: CT pulmonary angiography (CTA) with MIP reconstruction in axial (A) and coronal (B) views. The main pulmonary artery and its left branch (star) are of normal caliber, with normal-appearing lobar and segmental branches (red arrows). The right pulmonary artery arises with a normal caliber but demonstrates an abrupt cutoff of contrast opacification (black arrow), with no visualization of downstream branches, consistent with hypoplasia of the pulmonary artery.

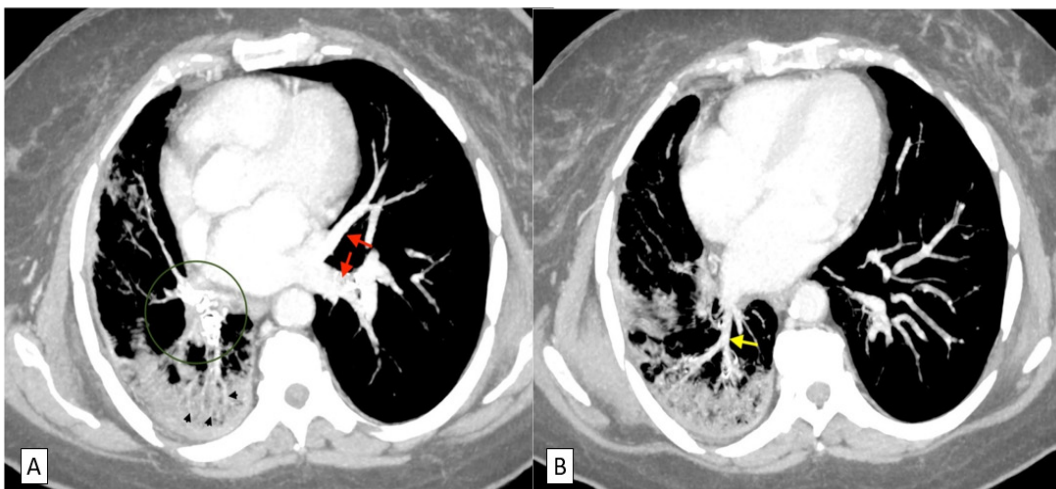


Figure 4: CT pulmonary angiography (CTA) with MIP reconstruction in axial view: (A) The left pulmonary veins are of normal caliber, with opacification of distal branches (red arrows). Prominent collateral circulation is observed on the contralateral side (circle). (B) On the right side, the posterior right pulmonary vein is hypoplastic (yellow arrow), with poor opacification of distal branches compared to the contralateral side. The anterior right pulmonary vein is not opacified.

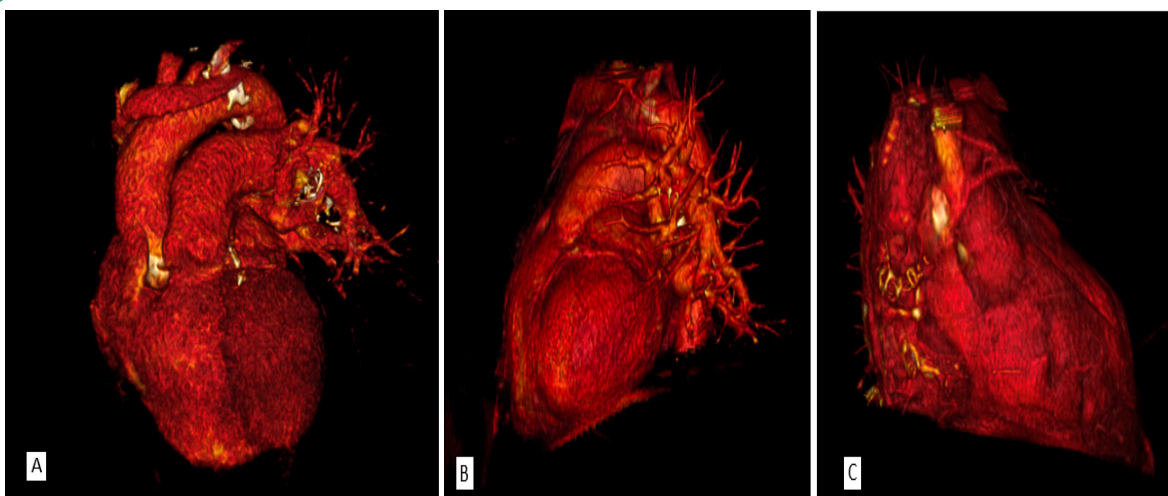


Figure 5: (A,B) Three-dimensional reconstruction showing normal-caliber pulmonary veins and arteries on the left side, (C) with hypoplasia of pulmonary vessels on the right side.

Differential diagnoses include chronic thromboembolic pulmonary hypertension, scimitar syndrome, and fibrosing mediastinitis. Management depends on symptom severity. Conservative treatment is suitable for asymptomatic patients. Symptomatic cases may require bronchial artery embolization, pneumonectomy, or lung transplantation [4,8].

Conclusion

The rare combination of right pulmonary artery and vein hypoplasia highlights the importance of a multidisciplinary approach for accurate diagnosis and management [8]. Advances in imaging modalities, particularly CT angiography and three-dimensional reconstructions, have significantly improved our understanding of this condition, enabling precise characterization and tailored treatment strategies [4]. Further studies are warranted to explore long-term outcomes and optimal management protocols for these patients [6].

Declarations

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Ethics approval: Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent: Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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