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Severe pulmonary hypertension associated with unilateral pulmonary artery hypoplasia: A case report

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Abstract

BACKGROUND: Pulmonary hypertension (PH), defined as mean pulmonary arterial pressure >20 mmHg, has diverse etiologies and high morbidity and mortality, necessitating early, accurate diagnosis.

METHODS: A 42-year-old woman, misdiagnosed with chronic thromboembolic pulmonary hypertension (CTEPH) and treated with anticoagulation for four years, had a rare congenital vascular anomaly. She presented with cough, hemoptysis (~200 mL of fresh blood two hours before admission), progressive dyspnea, and chest discomfort. Examination revealed a loud P2, suggesting right ventricular strain. Echocardiography showed elevated pulmonary artery pressure with right ventricular dysfunction. Chest CT angiography revealed abnormal vascular anatomy; pulmonary angiography confirmed unilateral pulmonary artery hypoplasia (UPAH) as the cause of PH. Anticoagulation was stopped, and embolization was performed successfully. At the two-month follow-up, symptoms had resolved and hemoptysis had not recurred.

RESULTS: UPAH is a rare congenital cause of PH that can mimic CTEPH, leading to misdiagnosis and inappropriate prolonged anticoagulation. Symptoms often overlap with common conditions such as pulmonary thromboembolism. In this case, lack of treatment response prompted reevaluation. Careful reassessment of prior imaging, history, and rare differential diagnoses was essential. This case highlights the importance of reconsidering the diagnosis when the course deviates from expectations and the need for individualized management in uncommon PH etiologies.

Keywords: Right Ventricular Dysfunction; Congenital Cardiovascular Malformations; Pulmonary Circulation Disorders; Hemoptysis; Embolization, Therapeutic; Diagnostic Imaging; Misdiagnosis

Introduction

As a potentially life-threatening disease, pulmonary hypertension (PH) is characterized by a mean pulmonary artery pressure (mPAP) of >20 mmHg caused by the remodeling of the pulmonary vasculature[1]. Various conditions can precipitate the onset of this pathology. Worldwide, left-sided heart and chronic lung diseases are the leading factors contributing to PH. Pulmonary arterial hypertension (PAH) and chronic thromboembolism pulmonary hypertension (CTEPH), along with several miscellaneous diseases, constitute additional etiologies[1,2]. However, regardless of the underlying pathology, the development of PH is associated with a significant risk of morbidity and mortality[2]. Thus, prompt diagnosis and initiation of therapy are crucial, necessitating a comprehensive understanding of characteristic clinical manifestations[3]. Concerning this, we present a very rare case of severe pulmonary hypertension related to isolated unilateral pulmonary artery hypoplasia in a 42-year-old female who had been previously diagnosed and treated for pulmonary thromboembolism several times.

Case Presentation

A 42-year-old female, previously diagnosed with chronic thromboembolism pulmonary hypertension (CTEPH) and receiving anticoagulant treatment for almost 4 years, presented to our emergency department with a chief complaint of cough and hemoptysis. The patient experienced progressive dyspnea during the last month, resulting in dyspnea at rest since the preceding week (Function Class IV). Meanwhile, she also had several episodes of retrosternal chest discomfort, described as pressure, with radiation to both upper extremities. The chest pain episodes lasted 15–20 minutes, and the pain was non-pleuritic and relieved with walking. Two hours prior to admission, she experienced sudden-onset cough accompanied by approximately 200 milliliters of fresh blood. The patient reported no weight loss, orthopnea, paroxysmal nocturnal dyspnea, fever, chills, or sweating. During the initial admission, the patient reported that she had experienced the same event several times over the past 4 years.

The patient had been on a regimen consisting of 5 mg of Warfarin once a day, 50 mg of Sildenafil three times a day, and 125 mg of Bosentan twice a day for 4 years. She reported an absence of notable family history, smoking, alcohol, or illicit drug abuse, as well as no recent travels or exposure to individuals with illnesses. She was a housekeeper.

Vital signs were as follows: blood pressure of 112/75 mmHg, heart rate of 95 beats per minute, respiratory rate of 24 breaths per minute, oral temperature of 36.9°C, and a room air O_2 saturation of 84%, which improved to 97% with the administration of 5 liters/min oxygen via nasal cannula.

On physical examination, the patient appeared awake, alert, and responsive, with no signs of cardiac or respiratory distress. A cardiac examination revealed loud P2 at the apex. The thorax was symmetrical and adequately expanded without external deformities or muscle retraction. Percussion of the lungs indicated resonance, while auscultation revealed diffuse rhonchi in the left lung. Extremities were warm, without edema, clubbing, or calf tenderness, and with normal, symmetric pulses.

The initial laboratory results indicated a partial thromboplastin time (PTT) of 55.7 seconds, a prothrombin time (PT) of 45 seconds, and an INR of 5.65. Additionally, the patient exhibited mild respiratory acidosis, while her complete blood count, inflammatory markers, and troponin levels remained within normal limits. The electrocardiogram did not reveal any changes consistent with ischemic-related abnormalities; however, an rsr' pattern was noted in leads V1–V2, along with the presence of P pulmonale.

Given the volume of hemoptysis and the absence of pleuritic or acute chest pain, there was low clinical suspicion of CTEPH; hence, we decided to explore other possible underlying causes.

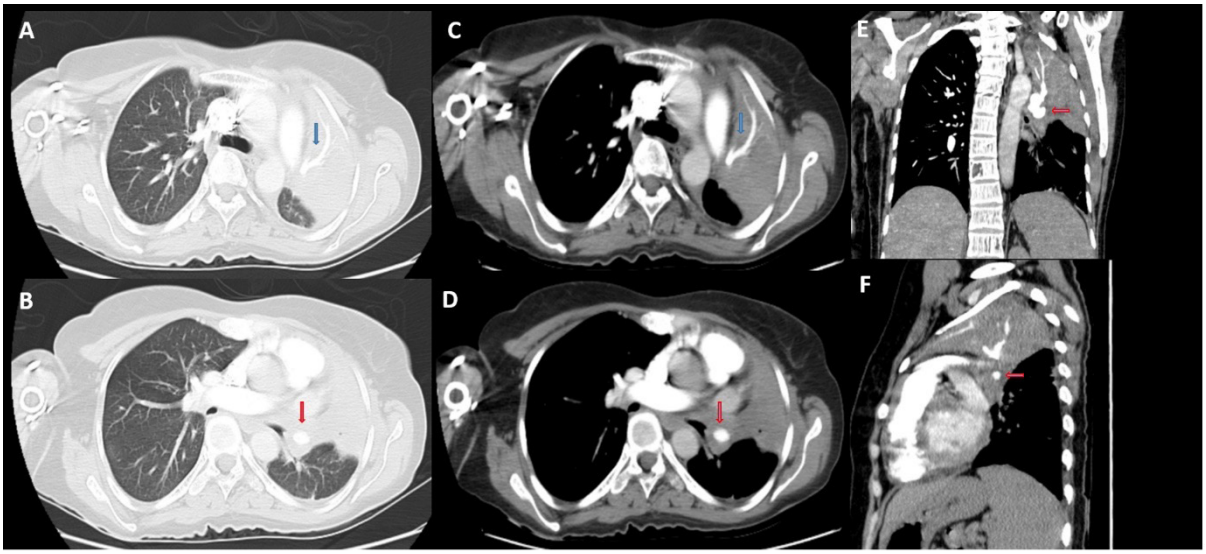

Figure 1. The Multidetector computed tomography angiogram

Echocardiography revealed an ejection fraction of 55%, along with severe pulmonary hypertension, severe tricuspid regurgitation, and moderate dilation of the right ventricle. No congenital heart anomalies were detected.

Multidetector Computed Tomography Angiography (hemoptysis protocol) was performed (Fig. 1). There was evidence of pulmonary artery dilation compared to the ascending aorta. The left lower pulmonary artery was abruptly terminated with a convex margin, accompanied by leftward mediastinal and heart shift and some parenchymal bands. The hypoplastic left lower pulmonary artery measured approximately 10 mm in diameter, terminating at the hilum without distal arborization. Thorough evaluation by obtaining a comprehensive health history and carefully analyzing the patient's prior lung perfusion scans revealed unilateral severe reduction or almost complete absence of radiotracer perfusion in the lower lobe of the left lung, which had not changed after four years. The perfusion scan showed near-complete absence of radiotracer uptake in the left lower lobe, consistent with non-functional pulmonary perfusion. Pulmonary embolism accounts for only a minor portion of these instances; thus, it is crucial to investigate alternative potential causes to avoid misdiagnosis, including mediastinal masses and fibrosis, parenchymal and pleural diseases, as well as vascular and congenital anomalies[4,5].

After a cardiology consultation, to establish a definite diagnosis, pulmonary artery angiography was performed (Fig. 2). This revealed left ascending and left descending pulmonary artery interruption, consistent with a right ventricle systolic pressure of 103 mmHg and a systolic pulmonary artery pressure (sPAP) of 103 mmHg. Thus, a diagnosis of pulmonary hypertension caused by unilateral pulmonary artery hypoplasia was made for our patient.

The patient was informed about her diagnosis, and Warfarin therapy was discontinued. She was also counseled for embolization. Moreover, she continued to do well after a week of hospitalization and was subsequently discharged with a recommendation for ongoing follow-up. At a two-month follow-up appointment, she reported no hemoptysis recurrence. Although only two months of follow-up are currently available, the patient remains under regular surveillance, with ongoing monitoring for recurrent hemoptysis or worsening hemodynamics. She was referred to her former treating physician for further follow-up.

Discussion

As a rare congenital abnormality, pulmonary artery interruption (PAI) is defined by the

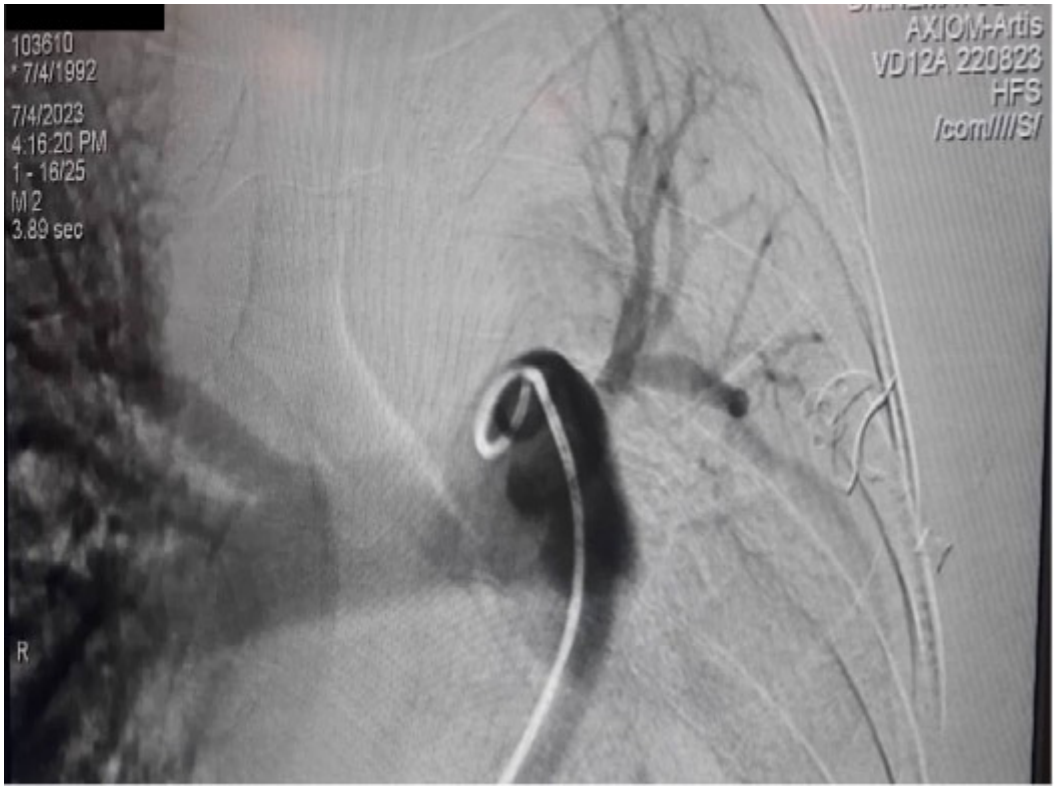

Figure 2. Pulmonary artery angiogram.

absence of a pulmonary artery caused by the failure in connection of the pulmonary trunk to the sixth aortic arch[5,6]. In the majority of cases, PAI is accompanied by other congenital cardiovascular defects and typically presents in early life due to the manifestations of the concomitant abnormalities[7]. However, in isolated cases (PAI without any concurrent defects), making a diagnosis might be delayed until adulthood, when the abnormality is discovered as an incidental finding[8]. Nevertheless, PAI might present with nonspecific signs and symptoms, including dyspnea, hemoptysis, chest pain, limited exercise tolerance, recurrent pulmonary infection, and sometimes high-altitude pulmonary edema, causing a diagnostic challenge[6,7]. Hence, as a result of misdiagnosis with some other cardiopulmonary conditions, PAI patients might stay undiagnosed for a median duration of 5 years after the onset of symptoms[7].

While unilateral emphysema, bronchiectasis, absent lung, coarctation of the pulmonary artery system, pulmonary vasculitis, and old tuberculosis are among the differential diagnoses, PAI shares several misleading characteristics with pulmonary thromboembolism (PTE), introducing a possibility of diagnostic errors[7,9].

First of all, one of the most common symptoms observed in adult patients with isolated PAI is hemoptysis, resulting from a large systemic collateral supply derived from the bronchial arteries. This characteristic is shared with chronic PTE and primary pulmonary hypertension, causing recurrent episodes and reduced transradiancy on chest X-rays obtained from the affected site[7,10].

Secondly, it is crucial to recognize the limitations of imaging in accurately distinguishing between PAI and PTE, as the similarities in imaging characteristics between these conditions can often lead to diagnostic challenges and errors[11]. In this regard, chest radiography of PAI patients can demonstrate an absent shadow of one pulmonary artery along with the underdevelopment of the affected

lung. Furthermore, ipsilateral hemidiaphragm elevation, mediastinal shift, and hyperinflation of the contralateral lung can be observed[6,10,12]. On the other hand, PTE can produce a chest X-ray showing an absent pulmonary artery shadow, reduced lung volume, and diminished vasculature[11].

In CT scans of PAI, an absent pulmonary artery, hyperinflated contralateral lung, and collateral circulation can be observed, while PTE can produce mosaic perfusion and a mural thrombus[13]. However, although the blockage of the main pulmonary artery by a thrombus can be similar to pulmonary artery agenesis, it may precipitate sudden-onset symptoms and make the patient unstable. Hence, CT scans should also be used carefully and interpreted regarding the clinical condition. A ventilation-perfusion scan also cannot differentiate these phenomena completely, as they both demonstrate absent lung perfusion and normal (or reduced) ventilation[13].

Despite ongoing anticoagulation therapy, the individual reported no sudden-onset or pleuritic chest pain over the past 4 years, and her dyspnea and episodic hemoptysis persisted, leading to further diagnostic evaluation due to atypical presentation.

In similar case reports, Cogswell et al.[14] described a patient with 3 days of hemoptysis, along with dull chest pain and a ventilation-perfusion mismatched radionuclide scan in the left lung, which could be in favor of large PTE in an appropriate clinical setting; however, the patient showed no dyspnea and no perfusion defects in the right lung. They decided to evaluate the possibility of a congenital pulmonary circulation anomaly. A pulmonary angiogram confirmed the absence of the left pulmonary artery along with the smooth contour of the main pulmonary artery in the expected location of the branch.

In contrast to our report and those of Cogswell et al., Moser et al.[11] described three cases of suspected PAI that were diagnosed as chronic PTE through thromboendarterectomy or angioscopy. They reported potential factors to consider PTE as an alternative diagnosis for PAI. In the first case, a previous lung scan demonstrated no perfusion defect in the suspected lung. Similarly, in another case, a previous series of chest radiographs did not reveal any agenesis-related findings of the pulmonary artery. These findings were suggestive of PTE instead of PAI. Another case described a long history of deep vein thromboembolism, while the pulmonary angiography suggested pulmonary artery agenesis. Thus, Moser et al. believed that X-ray, computed tomography, and angiography of the pulmonary vasculature cannot differentiate between thromboembolism and interruption of the pulmonary artery.

In conclusion, accurately differentiating between chronic thromboembolic pulmonary hypertension (CTEPH) and rare congenital vascular anomalies such as unilateral pulmonary artery hypoplasia (UPAH) remains a clinical challenge. Misdiagnosis can lead to prolonged inappropriate therapy, such as anticoagulation, which in patients with UPAH may increase the risk of massive hemoptysis.

Several key factors contributed to the correct diagnosis in our case:

- The patient's chronic symptoms, including episodic hemoptysis and lack of response to anticoagulation, were inconsistent with typical CTEPH.
- Review of prior ventilation-perfusion scans revealed preserved ventilation and complete absence of perfusion in the left lung, consistent with non-functional pulmonary vasculature (Fig. 3).
- Imaging findings, combined with clinical reevaluation and catheter-based angiography, confirmed unilateral pulmonary artery hypoplasia.

Consistent terminology, clinical suspicion, and integration of imaging modalities are essential in approaching such atypical cases. Given the potential harms of misdiagnosis, including unnecessary anticoagulation and missed therapeutic opportunities, early recognition of UPAH is critical.

This case also highlights the importance of reevaluating presumed diagnoses when the

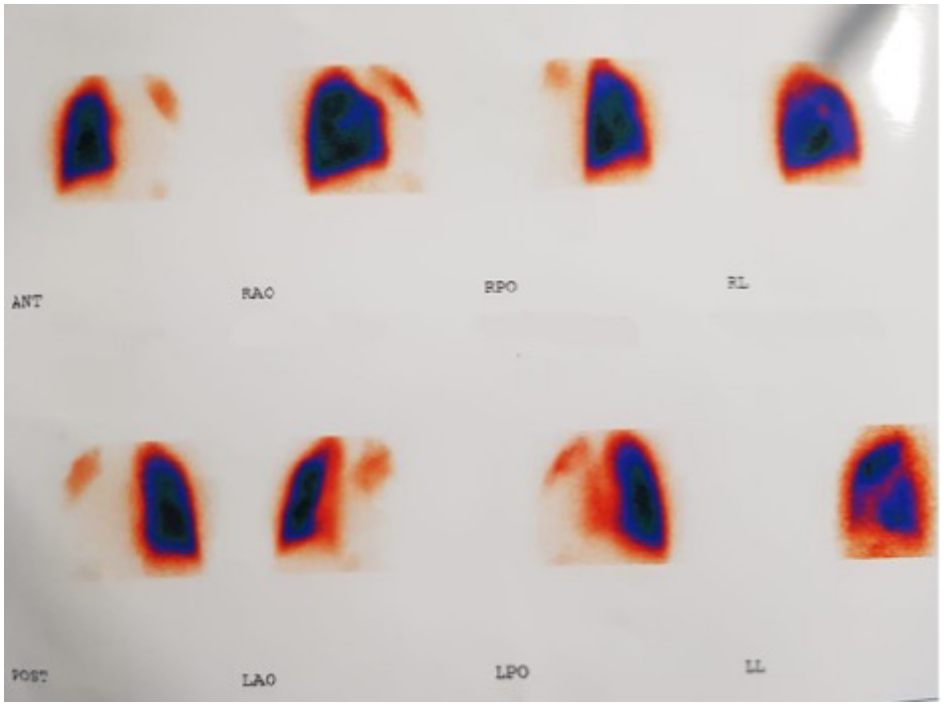

Figure 3. Ventilation-perfusion scans.

clinical course deviates from expectations. Pulmonary thromboembolism remains a frequent initial consideration and, as this case shows, may act as "the Great Masquerader" in pulmonary vascular disease (Camera et al. 2012)[10].

Conflict of interests

The authors declare no conflict of interest.

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Author's Contributions

Study Conception or Design: EK, FK
Data Acquisition: EK, HM, MM, AN
Data Analysis or Interpretation: -
Manuscript Drafting: HM, MM
Critical Manuscript Revision: EK, FK, AN
All authors have approved the final manuscript and are responsible for all aspects of the work.

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