



Case Study

Massive Hemoptysis Revealing Isolated Right Pulmonary Artery Absence

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Unilateral absence of the pulmonary artery (UAPA) is a rare congenital anomaly that may remain undiagnosed until adulthood, especially in the absence of associated cardiac defects. We report a 32-year-old male presenting with massive hemoptysis. He had a history of recurrent respiratory infections since childhood. Chest radiograph showed cystic changes and reduced vascular markings in the right lung. CT pulmonary angiography revealed complete absence of the right pulmonary artery with systemic collateral supply and associated cystic bronchiectasis. Echocardiography showed mild pulmonary hypertension. Infectious causes, including tuberculosis, were ruled out. The patient was managed conservatively and remained stable. This case highlights isolated UAPA as a rare cause of massive hemoptysis and emphasizes the role of CT pulmonary angiography in diagnosis and the importance of recognizing associated bronchiectasis as a contributing factor.

Keywords: unilateral absence of pulmonary artery, hemoptysis, bronchiectasis, CT pulmonary angiography.

INTRODUCTION

Unilateral absence of the pulmonary artery (UAPA) is a rare congenital malformation (1) which is often associated with cardiac malformation (2). In the absence of associated cardiac malformation, the condition may be asymptomatic until adult age (3,4). We present a case with isolated UAPA, which was diagnosed upon the occurrence of hemoptysis, and angiography was subsequently performed.

Case report:

A 32-year-old male manual labourer presented to the emergency department with a sudden episode of massive hemoptysis, expectorating around 500 mL of fresh frank blood over a short duration. There was no history of fever, trauma, chest pain, or constitutional symptoms such as weight loss or night sweats.

On detailed medical history patient revealed similar episode during early childhood at the age of five years, when he had hemoptysis approximately 50–100

mL. In addition, he reported recurrent respiratory tract infections throughout childhood, requiring multiple courses of antibiotics but no definitive diagnosis was established. On presentation, patient was conscious, alert and oriented, but was anxious. His vital signs showed tachycardia with heart rate, blood pressure of 110/70 mm Hg and respiratory rate of 24 breaths per minute and oxygen saturation of 92 % on room air and He was afebrile. No evidence of pallor, icterus, cyanosis, clubbing or lymphadenopathy.

Laboratory investigations complete blood count, renal and liver function tests, coagulation parameters were within normal limits, effectively ruling out systemic infection, coagulopathy, or metabolic causes of bleeding. Sputum smear for acid-fast bacilli and CBNAAT was negative for mycobacterial tuberculosis. Sputum gram stain and bacterial culture did not reveal pathogenic organism. These findings effectively ruled out pulmonary tuberculosis and active bacterial infection as the cause of hemoptysis.

Chest xray PA view (figure 1) showed evidence of multiple cystic lucencies in right mid and lower zones, suggestive of cystic bronchiectatic changes. The right hilum appeared less prominent compared to the left, with relative paucity of vascular markings on the right side. There was mild ipsilateral mediastinal shift and compensatory hyperinflation of the left lung. These findings were suggestive of chronic structural and vascular abnormality involving the right lung, requiring further evaluation.

We proceeded with CT pulmonary angiography (Figure 2) demonstrated complete absence of the right pulmonary artery with multiple systemic collateral vessels supplying the right lung. Associated cystic bronchiectasis and volume loss of the right lung were noted. The left pulmonary artery and left lung were normal. These findings explain the patient's presentation with massive hemoptysis.

The patient was managed conservatively with bed rest, hemostatic agents, and supportive care. Broad-spectrum antibiotics were administered in view of underlying bronchiectasis. The patient remained hemodynamically stable, and no further episodes of hemoptysis occurred during hospitalization. He was advised regular follow-up and smoking cessation.

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 (5)

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 (6).

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 (7)

With regard to symptoms, Ten, et al. reported dyspnea or exercise limitation in 40% of cases, frequent respiratory infections in 37%, and hemoptysis in 20%. In addition, 15% to 24% of cases were reported to be asymptomatic (3,4). Hemoptysis, which was also seen in our case, is presumed to be caused by excessive collateral circulation and fragility of the vessels including the capillary bed. (8)

Angiography is suitable for evaluation of the collateral circulation, and also contributes greatly to treatment. Although hemoptysis is likely to resolve naturally (9) in cases where repeated hemoptysis is observed, angiography is used to evaluate the collateral circulation, and the blood vessels responsible for the hemoptysis are embolized. (10) If hemoptysis recurs after the embolization, pneumonectomy may be performed. Evaluation of the collateral circulation using angiography will also provide useful information for handling blood vessels during surgery. (11)

Ten, et al. reported that pulmonary hypertension was found in 44% of isolated UAPA cases. Inability of the decreased pulmonary vascular bed to cope with increased blood flow leads to pulmonary hypertension. The reduction in pulmonary arterial blood flow to the affected lung leads to chronic hypoperfusion, impaired mucociliary clearance, and recurrent infections, which may eventually result in structural lung damage such as bronchiectasis. Bronchiectasis is one of the most commonly reported parenchymal abnormalities in patients with isolated UAPA and contributes significantly to morbidity. (3,4)

Our patient had isolated right-sided UAPA with no concomitant heart abnormality, and the right middle and lower lobes were affected by cystic bronchiectasis. The development of bronchiectasis due to persistent hypoperfusion was probably influenced by a history of repeated respiratory infections during childhood. A major contributing factor to the development of extensive hemoptysis was the existence of bronchiectasis and delicate systemic collateral arteries. Early hemodynamic effects of the underlying vascular abnormality were further demonstrated by mild pulmonary hypertension (PASP 35 mmHg). This highlights the significance of

bronchiectasis as a major hemoptysis-causing complication of isolated UAPA.

CONCLUSION:

Isolated unilateral absence of the pulmonary artery is rare cause of massive hemoptysis and may remain undiagnosed until adulthood. Associated bronchiectasis increases the risk of hemoptysis due to brittle collateral circulation. Early diagnosis with CT pulmonary angiography is crucial for appropriate management and prevention of complications.

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