Case Report:
Unilateral Pulmonary Artery Agenesis Presenting with Massive Hemoptysis

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Abstract
Background: Congenital unilateral absence of a pulmonary artery is a rare anomaly most frequently accompanied by other cardiovascular anomalies. It is usually diagnosed during childhood and effectively treated surgically by doing a pneumonec-tomy.

Case Report: A young adult with right pulmonary artery agenesis, who presented in November 2011 to Aseer Central Hospital with massive hemoptysis. He was treated successfully with arterial embolization, as the definitive management plan was an elective pneumonec-tomy after referral to an expert specialized center for the latter form of treatment.

Conclusion: Pulmonary artery agenesis, needs to be considered as a cause of recurrent hemoptysis, especially in a patient with decreased unilateral lung volume. Such patients should be strongly considered for undergoing a pulmonary angiography for confirming this rare pulmonary vascular malformation.

Key Words: Massive hemoptysis — Unilateral agenesis — Pulmonary artery.

Introduction

ISOLATED absence of a pulmonary artery is a rare developmental anomaly due to a failure in the connection of the sixth aortic arch with the pulmonary trunk [1]. Few patients may remain asymptomatic until adulthood. However, an increased incidence of recurrent respiratory tract infections has been noted. Some patients also present with dyspnea on exertion or congestive heart failure [2] and it is a rare cause of massive hemoptysis [3]. While initial stabilization of patient could be accomplished by bronchial artery embolization, a definitive treatment usually includes a pneumonec-tomy [4].

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Case History

During November 2011, a 26-year old Saudi male presented to Aseer Central Hospital, Abha, Kingdom of Saudi Arabia, with a history of hemoptysis of one week's duration. The patient, with no previously reported morbidities, presented with a history of an abnormal sensation in his throat followed by sudden expectoration of large amount of bright fresh blood, which was estimated to be around 200 cc.

There was no associated sputum expectoration, fever or symptoms suggestive of upper or lower respiratory tract infection. Our patient has been a non-smoker with no history of any occupational exposure to organic or inorganic pollutants. Three years earlier, he experienced a single attack of hemoptysis. The family history was unremarkable for a similar problem in the first degree relatives.

On examination, the patient looked well. His vital signs were stable, with a blood pressure of 110/65 mmHg, regular heart rate of 95 beats/minute, normal temperature (37°C), respiratory rate of 20 breaths/minute with an oxygen saturation of 96%, while breathing ambient air. Head, eyes, ears, nose and throat examination was within normal limits with no clinically significant lymphadenopathy. His chest examination revealed tracheal shift toward the right side together with a decreased chest expansion ipsilaterally with a normal (resonant) percussion note and vesicular breath sounds bilaterally with a decreased breath sound intensity in the right hemithorax. The cardiovascular examination revealed normal first heart sound together
with a prominent (loud) pulmonic component (P2) of the second heart sound, with no added sounds, murmurs or rubs. Rest of his physical examination was within normal limits.

His initial investigations showed a normal complete blood count, with white cell count of 10,000/gL, hemoglobin of 15gm/dL, platelet count of 409,000/gL, a normal coagulation profile, serum biochemistry (renal and liver function tests and serum electrolytes) and urine analysis (including a negative urine dipstick for protein, RBCs and nitrites). Electrocardiography revealed a normal sinus rhythm with a prominent P-pulmonale. Chest radiography revealed a smaller right hemi-thorax with a small right hilum and an elevated right hemi-diaphragm along with a right-sided mediastinal shift and hyperlucent left lung field (Fig. 1). Contrast enhanced CT chest scan confirmed the absence of the right pulmonary artery (Fig. 2).

Fiber optic bronchoscopy showed a polyp at the right main bronchus and histopathologically, there was evidence of only chronic inflammation. Pulmonary angiography showed atretic right main pulmonary artery together with several hypertrophied and tortuous bronchial arteries on the same side.

The patient underwent two sessions of bronchial artery embolization and electively the patient was considered for pneumonectomy as the definitive management for his condition and was referred to a higher centre for this therapeutic purpose.

Discussion

Developmental anomalies of the lung can be categorized as bronchopulmonary anomalies, vascular anomalies, or combined anomalies. They are primarily due to aberrant intrauterine development. Pulmonary vascular anomalies include proximal interruption of a central pulmonary artery, an anomalous origin of the left pulmonary artery from the right pulmonary artery, partial or total anomalous pulmonary venous return, pulmonary arteriovenous malformation, and mal-development of lymphatic vessels, such as cystic hygroma.

Blood flow through the central pulmonary arteries (i.e., the main pulmonary artery, right pulmonary artery, and left pulmonary artery) may be interrupted due to pulmonary artery agenesis, pulmonary arteria, or pulmonary artery stenosis. Radiographically, the ipsilateral hilum and lung are small and receive arterial blood from systemic collaterals. The right pulmonary artery is most frequently involved.

Unilateral pulmonary artery agenesis is a rare condition often associated with other congenital abnormalities. In some cases, they occur together with anomalies, mostly with skeletal, heart and diaphragmatic anomalies. Chromosomal abnormalities, vitamin A deficiency, intrauterine infections, and environmental factors has been held responsible for the etiology of congenital lung malformations.

Our patient presented with recurrent hemoptysis although he had no other symptoms. Angiography revealed absence of the right pulmonary artery and tortuous bronchial collaterals. Pulmonary angiography has been used as a main diagnostic tool for this condition. He underwent two sessions of bronchial artery embolization and was considered for pneumonectomy, which is the definitive management for this condition.
Hemoptysis has been described as a clinical symptom in up to 10% of patients, originating from either hypertrophied bronchial collateral vessels or peripheral arteriovenous fistulas ipsilateral to the absent pulmonary artery, as well as from rupture of chronically hyperperfused vessels on the contralateral side [8].

A therapeutic option in adults may be an interventional or surgical occlusion of collateral vessels or pneumonectomy on the affected side to avoid hemoptysis. Heart-lung transplantation may be an option to simultaneously improve blood oxygenation [4].

Rousou et al. [9] reported a case of intrapulmonary hemorrhage and hemoptysis in a patient with left-sided pulmonary artery agenesis. The patient was successfully treated by performing a left pneumonectomy.

Conclusion:

Pulmonary artery agenesis, though a rare condition, needs to be considered as a cause of recurrent hemoptysis, especially in a patient with decreased unilateral lung volume. Such patients should be strongly considered for undergoing a pulmonary angiography, which is a gold standard investigative modality, for confirming this rare pulmonary vascular malformation.

References


