Congenital Bronchial Artery to Pulmonary Artery Fistula Presenting as Hemoptysis

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A 51-year-old male presented with 2 weeks of hemoptysis. Pulmonary angiography was performed and identified a bronchial artery to pulmonary artery fistula of the right upper lobe. Despite angioembolization, the hemoptysis recurred 1 year later. It was hypothesized that the recurrence occurred due to retrograde filling from the pulmonary arterial side of the abnormality. Right upper lobectomy was performed and resulted in resolution of hemoptysis. We present a case report of a rare, congenital bronchial artery to pulmonary artery fistula.

Pulmonary arteriovenous malformation (PAVM) is a broad term used to describe various vascular lesions with abnormal communications within the lungs. Conventionally, PAVMs are between the pulmonary artery and vein, while a small subset connects the systemic vasculature with the pulmonic vasculature. PAVMs were first identified in 1897 as an autopsy finding [1]. Another form of systemic PAVMs is acquired major aortopulmonary collateral arteries found in neonates with pulmonary atresia [2]. They are thought to arise during the maldevelopment of the pulmonary outflow tract as seen in pulmonary atresia or severe tetralogy of Fallot, and represent persistent systemic-pulmonic connections from embryologic periods [3]. Less common, PAVMs may have no clear cause and arise by congenital mechanisms. Two separate case series [4, 5] report that systemically supplied PAVMs are a rare clinical entity, with approximately 4% of congenital PAVMs having systemic arterial supply. Additionally, there have been very few bronchial artery to pulmonary artery PAVMs in the literature [6, 7]. We report the case of a congenital bronchial artery to pulmonary artery PAVM that presented as hemoptysis.

A 51-year-old male presented with 2 weeks of hemoptysis. Bronchoscopy was performed finding only trace amounts of blood arising from the orifice of the right upper lobe; therefore, the patient was referred for angioembolization. Angiography demonstrated a bronchial artery arising from a dilated right intercostal bronchial trunk, feeding an apical PAVM with outflow into the right pulmonary arterial circulation (Fig 1). The patient initially underwent embolization of the supplying bronchial artery with Embozene Microspheres (CeloNova BioSciences, Inc, San Antonio, TX) and Gelfoam (Pharmacia & Upjohn Company, Kalamazoo, MI) resulting in stasis of flow through the malformation. Embolization led to resolution of symptoms. Over the course of outpatient follow-up, the patient complained of occasional right-sided chest pain but no further bleeding. However, the patient had significant recurrence 1 year later.

Repeat embolization was unsuccessful, and it was hypothesized that retrograde filling of the PAVM from the pulmonary arterial side of the abnormality was contributing to the ongoing bleeding. Therefore the patient was taken to surgery. Right video-assisted thoracotomy and right upper lobectomy were performed. Dense apical adhesions were discovered, representing the transpleural connection between the systemic circulation and the pulmonary arterial supply. Radiofrequency energy (LigaSure device; Covidien, Mansfield, MA) was used to hemostatically perform lysis of adhesions and to separate the lung from the chest wall. The patient had an uneventful postoperative course and was discharged home on postoperative day 5.

Pathologic examination revealed clotted blood throughout the bronchial tree. Microscopic examination revealed multiple ectatic blood vessels, containing varying degrees of embolic material. These vessels appeared to represent muscular ramifications of the pulmonary arteriovenous fistula. A 51-year-old male presented with 2 weeks of hemoptysis. Pulmonary angiography was performed and identified a bronchial artery to pulmonary artery fistula of the right upper lobe. Despite angioembolization, the hemoptysis recurred 1 year later. It was hypothesized that the recurrence occurred due to retrograde filling from the pulmonary arterial side of the abnormality. Right upper lobectomy was performed and resulted in resolution of hemoptysis. We present a case report of a rare, congenital bronchial artery to pulmonary artery fistula.

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arteries extending to the level of the respiratory bronchiole as well as associated smaller vessels and veins. They appeared consistent with the evidence of systemically supplied PAVM with prior embolization (Fig 2).

**Comment**

Systemically supplied PAVMs differ in their clinical presentation from their more conventional counterpart [8]. Most cases are asymptomatic as they represent a left-left shunt and therefore do not present with cyanosis. It is also suggested that systemic PAVMs do not have an association with Osler-Weber- Rendu syndrome, as do many PAVMs. Due to their systemic pressures, they are more prone to enlarge or rupture over time, which requires more definitive management. Angioembolization is a useful tool and should be a first-line therapy for hemoptysis. It allows for delineation of anatomy and may also be therapeutic. In our patient it provided one year of symptom control. In the second instance, it confirmed location of the PAVM and additionally helped to reduce systemic flow through the PAVM, which may have helped to mitigate operative blood loss by “depressurizing” the PAVM.

**References**