An Unusual Cause of Hemoptysis: Congenital Absence of the Right Pulmonary Artery in a Young Male

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Unilateral absent pulmonary artery (UAPA) is rare and can occur in association with cardiovascular anomalies or in isolation. Herein, we describe a case of isolated UAPA in a 14-year-old male patient who presented with hemoptysis secondary to abnormal hypertrophy of collateral vasculature. The patient was successfully treated by endovascular embolization with polyvinyl alcohol particles.

Key words  Hemoptysis · Congenital · Pulmonary diseases · Pulmonary artery · Embolization.

INTRODUCTION

Unilateral absent pulmonary artery (UAPA) occurs in approximately 1 in 200000 young adults [1]. UAPA may be associated with a spectrum of cardiovascular anomalies [2], such as tetralogy of Fallot, atrial septal defect, coarctation of the aorta, right aortic arch, and pulmonary atresia. Isolated UAPA involves the right lung in two-thirds of cases [3]. Due to embryonic relationships, UAPA often occurs on the side of the chest opposite the aortic arch [4]. The exact embryologic etiology of UAPA remains uncertain. However, aberrant development of a sixth aortic arch segment is thought to result in a ductal origin of a pulmonary artery that leads to proximal interruption of that vessel when the ductal tissue regresses at birth [4]. Herein, we report a case of a 14-year-old male patient with right UAPA who presented with hemoptysis.

CASE REPORT

A 14-year-old boy with good past health presented with a 2-day history of hemoptysis. No dyspnea, chest pain, or other systemic symptoms were evident. He remained hemodynamically stable. On physical examination, there was right-sided tracheal deviation. The apex beat of the heart was deviated toward the right side and was indistinct. There was no anemia. Intravenous transamin was administered with subsequent resolution of hemoptysis. A chest radiograph (Fig. 1A) revealed an ipsilateral, small, right hemi-thorax and a right-sided, unilateral, hyper-transradiant hemi-thorax. The chest radiograph also showed right-sided tracheal deviation and mediastinal shift. Initial bedside echocardiogram demonstrated a normal-sized main pulmonary trunk, but the right pulmonary artery was not visualized. A Tc-99m macroaggregated albumin lung perfusion scintigraphy/scan (Fig. 1B) showed near-absent right lung perfusion, compatible with absent right pulmonary artery. CT confirmed right UAPA (Fig. 1C). There was hypertrophy (Fig. 1D) of the right bronchial and non-bronchial systemic arteries. No active extravasation or anomalous veins were identified. Endovascular embolization (Fig. 2A and B) was performed due to repeated symptomatic hemoptysis. Polyvinyl alcohol particles (500–710 microns in size) were administered via a 2.7 French (Fr) micro-catheter placed at the proximal part of the right bronchial artery. Post-procedural angiography (Fig. 2C) revealed successful occlusion of the distal branches of the right bronchial artery. There were no reported episodes of hemoptysis in the following two years, and follow-up imaging demonstrated the absence of significant collateral hypertrophy formation in the right bronchial and non-bronchial systemic arterial system.

Relevant patient consent was obtained.
DISCUSSION

UAPA is rare and most commonly diagnosed during adolescence. A literature review by Ten Harkel et al. [3] in 2002 placed the median age of presentation at 14 years, identical to our patient. Adult patients are often asymptomatic and, hence, remain

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**Fig. 1.** Multi-modality imaging for the evaluation of unilateral absent right pulmonary artery. (A) Chest radiograph demonstrated a small right hemi-thorax, right-sided unilateral hyper-transradiant hemi-thorax, tracheal deviation, and mediastinal shift. (B) A Tc-99m macroaggregated albumin lung perfusion scintigraphy scan revealed near-absent right lung perfusion. (C) Axial contrast-enhanced CT scan confirmed absence of the right pulmonary artery. (D) Right bronchial artery hypertrophy and presence of multiple abnormal collateral vessels were observed, with no active contrast extravasation.

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**Fig. 2.** Diagnostic and therapeutic endovascular intervention for unilateral absent right pulmonary artery. (A) Initial aortogram performed with a 5-Fr pigtail flush catheter confirmed hypertrophy of the right bronchial artery. (B) Pre-embolization selective right bronchial angiogram demonstrated hypertrophic collateral vasculature. (C) Post-procedural right bronchial angiogram exhibited successful occlusion of the distal branches of the right bronchial artery after administration of polyvinyl alcohol particles.
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undiagnosed [3]. Patients present with exercise intolerance, recurrent respiratory infections, and hemoptysis, one of the most serious complications of UAPA, resulting from large and excessive collateral circulation that subjects venous systems to abnormally high pressures [5]. The distal intrapulmonary arteries of the affected artery are often supplied by collateral vessel formation arising from the bronchial, intercostal, internal mammary, subclavian, and infra-diaphragmatic arteries [6]. Although hemoptysis is often self-limiting for years, it may ultimately result in massive pulmonary hemorrhage and death [5]. Clinical prognosis depends on the associated cardiovascular anomalies and the degree of pulmonary hypertension, if present. The overall reported mortality for UAPA is 7%.

CT typically demonstrates termination of the absent pulmonary artery within 1 cm of its expected origin from the main pulmonary artery. Pulmonary angiography is recognized as the gold standard for diagnosis, but it is rarely used as a primary diagnostic tool due to rapid advancements in cross-sectional imaging. As in our patient, pulmonary angiography is reserved for hemoptysis that requires therapeutic endovascular intervention. Currently, there is no consensus regarding the treatment for isolated UAPA [5]. Clinical management of these patients should be determined by symptomology, pulmonary arterial anatomy, associated cardiovascular abnormalities, presence of collaterals, and degree of pulmonary hypertension [7]. In our patient, selective embolization was deemed less invasive with lower procedural risk compared to a pneumonectomy and was selected as the treatment modality with the highest likelihood of therapeutic success.

Conflicts of Interest
The authors declare that they have no conflict of interest.

REFERENCES