Cryptogenic hemoptysis caused by isolated aortopulmonary collateral artery formation: a case report

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Abstract
Hemoptysis in children is caused by various factors, the most common of which is basic lung disease or heart disease. Aortopulmonary collateral arteries (APCAs) are blood vessels that originate from the aorta or its branches and provide blood flow to the pulmonary tissues. We herein report a rare case of APCAs without abnormal structures in the heart. The patient was a previously healthy boy with APCAs originating from the descending aorta. He had no history of congenital heart disease and developed repeated episodes of cryptogenic hemoptysis during his school-age years. Arteriography examination facilitated the diagnosis of APCAs. After embolization, the patient developed no further hemoptysis during 10 months of follow-up. Arteriography is of great significance in determining the cause of recurrent cryptogenic hemoptysis.

Keywords
Hemoptysis, cryptogenic hemoptysis, aortopulmonary collateral artery, case report, children, cardiovascular angiography

Date received: 12 May 2021; accepted: 16 August 2021

Introduction
Hemoptysis refers to bleeding of the larynx and any part of the respiratory tract below the larynx. Blood is coughed up either alone or with sputum. When the etiology of hemoptysis cannot be determined by chest
computed tomography (CT) and bronchoscopy, the condition is termed cryptogenic hemoptysis. This type of hemoptysis is caused by vascular malformations. The present report describes a rare case of cryptogenic hemoptysis caused by the formation of isolated aortopulmonary collateral arteries (APCAs).

Case report

An 11-year-old boy was admitted to the Children’s Hospital of Shanghai, China on 20 March 2019 because he had experienced three episodes of hemoptysis within 5 days. The coughed-up blood was bright red and contained blood clots and a small amount of mucus. The patient showed no other abnormalities, such as hematemesis or melena. Moreover, a routine blood examination in another hospital before admission had shown no abnormalities. Chest CT showed possible inflammation in the middle and lower lobes of the right lung; the possibility of a bronchial mucus plug in the middle lobe of the right lung was not excluded, and nodules were observed in the middle and lower lobes of the right lung. The child had been hospitalized in another facility 2 months previously (from 21 December 2018 to 13 January 2019) because of a paroxysmal cough and hemoptysis for 5 days. At that time, chest CT angiography showed no obvious abnormality in either of the two pulmonary vessels, and bronchoscopy revealed a follicle-like protrusion from the tracheal wall in the opening of the right middle lobe with no fresh or altered blood within the trachea. The patient was given anti-infection treatment until discharge. The timeline of the patient’s medical history is shown in Figure 1.

The patient had no history of foreign body inhalation, trauma, familial bleeding or coagulation abnormalities, or vascular malformations. He had been previously healthy, had been delivered at term, and had experienced normal growth and development. No obvious abnormalities were found during the physical examination at the time of admission, such as shortness of breath, cyanosis, or finger clubbing. His oxyhemoglobin saturation was normal on room air, and his appearance was not consistent with anemia. The patient had clear consciousness, smooth respiration, and normal reactions. His thorax was symmetrical with no deformities, and his bilateral

![Figure 1. Timeline of the patient’s medical history. CTA, computed tomography angiography.](image-url)
respiratory movements were symmetrical without dry or wet rales. His heart sounds were strong and regular with no pathological murmurs. No bleeding from the nasal cavity or oropharynx was observed in the laryngopharyngeal examination. Routine blood examination showed that both the leukocyte count and hemoglobin concentration were in the reference ranges. The procalcitonin concentration was <0.01 ng/mL. A tuberculosis infection T-cell spot detection kit (T-SPOT.TB) and purified protein derivative of *Mycobacterium tuberculosis* test were negative. Hence, tuberculosis infection was excluded. No hemosiderin granules were found in the sputum for 3 days; thus, pulmonary hemosiderosis was excluded. Clotting function was normal; thus, bleeding or clotting dysfunction was excluded. No obvious abnormalities were detected in a rheumatic screening, and the results were as follows: perinuclear anti-neutrophil cytoplasmic antibody was weakly positive, cytoplasmic anti-neutrophil cytoplasmic antibody was negative, anti-glomerular basement membrane antibody was negative, and anti-double-stranded DNA antibody was negative. The C-reactive protein concentration was ≤5 mg/L, the erythrocyte sedimentation rate was 24 mm/hour, the complement C3 concentration was 1.42 g/L, and the complement C4 concentration was 0.26 g/L. Hence, vasculitis and rheumatic diseases were excluded after consultation with the renal department. Cardiac CT angiography revealed no obvious abnormalities (Figure 2). Because the cardiac ultrasound findings were normal, congenital heart disease was excluded. The patient was administered anti-infection treatment with cefuroxime, hemostasis treatment with etamsylate, and other symptomatic treatment. On 28 March 2019, cardiac catheterization and cardiovascular angiography were performed. During the operation, a

![Figure 2. Chest computed tomography angiography scans of the patient. No significant abnormality was seen.](image-url)
A collateral vessel with an inner diameter of 1.5 mm was found to originate from the descending aorta. The collateral vessel was divided into left and right branches to supply the left and right lung fields, respectively (Figure 3 and movie clip in Supplemental Material). Spring rings \((3 \times 2)\) were used to embolize the branches of the left and right lung fields, and repeat angiography showed a significant reduction in the shunt. After embolization, bronchoscopic examination showed that no bleeding had occurred during the operation. The diagnosis of APCAs was finally confirmed. The patient developed no hemoptysis during the >10-month follow-up.

**Discussion**

Hemoptysis is rare in children. Previous studies have shown that massive hemoptysis occurs in 13% of children hospitalized for treatment of hemoptysis, especially those with cystic fibrosis.\(^3\) Hemoptysis in children is caused by various factors, the most common of which is basic lung disease or heart disease. It is rarely detected in children with normal cardiopulmonary health. When focal bleeding is suspected, a chest CT scan and bronchoscopy are conducted.\(^4\) When the chest CT scan and bronchoscopy fail to determine the bleeding source, the condition is termed cryptogenic hemoptysis. About 7% to 25% of adult patients with hemoptysis have cryptogenic hemoptysis, which is primarily caused by vascular malformations.\(^2\)–\(^5\)

Systemic-to-pulmonary collaterals, also known as APCAs, are blood vessels that originate from the aorta or its branches and provide blood flow to the pulmonary tissues. APCAs might participate in the pulmonary blood supply either alone or with the inherent pulmonary artery.\(^6\) Furthermore, APCAs can be divided into the following three types according to the connection between the APCAs and the pulmonary artery: APCAs originating from the bronchial artery, APCAs originating from the direct aortopulmonary collaterals, and APCAs originating from the branches of the aorta. APCAs can also be divided into the following three types according to their diameter: large APCAs with a diameter of >2 mm, medium APCAs with a diameter of 1 to 2 mm, and small APCAs with a diameter of <1 mm.\(^7\)
APCAs are usually secondary to cyanotic heart disease and are rare in children without cyanotic heart disease. Most patients have dyspnea and cyanosis soon after birth or in infancy. It is extremely rare for school-aged children to have APCAs without an abnormal cardiac structure but with repeated cryptogenic hemoptysis. APCAs originating from the descending aorta without a history of congenital heart disease have not been previously reported. The vast majority of APCAs without cardiac structural abnormalities are asymptomatic. Only a small number of children with APCAs have hemodynamic manifestations of blood flow from left to right, leading to increased blood flow in pulmonary tissues, increased cardiac load, heart enlargement, respiratory symptoms, and cardiac dysfunction. These children can be misdiagnosed with myocardial disease or patent ductus arteriosus. Patients with recurrent chest pain and dyspnea after activity have been described; their condition might be attributed to APCAs originating from the coronary arteries, causing coronary artery steal and leading to myocardial ischemia. The present case involved a previously healthy boy with APCAs originating from the descending aorta, and he had no history of congenital heart disease but experienced repeated cryptogenic hemoptysis during school age. Arteriography examination facilitated the diagnosis in this case. This report will assist clinicians in diagnosis and treatment when encountering such cases.

The reporting of this study conforms to the CARE guidelines. We have de-identified all patient details.

Ethics approval and consent to participate
This is a case report; therefore, ethics approval was not required. The study was approved for publication by the board of Children’s Hospital of Shanghai, Shanghai Jiaotong University. We treated the patient according to an established regimen, with no complications. Informed consent was obtained from the patient and his family for the operation, treatment, and publication of this case report.

Declaration of conflicting interests
The authors declare that there is no conflict of interest.

Funding
This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Data availability
The datasets generated and/or analyzed during the current study are available from the corresponding author on reasonable request.

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