

Severe Hemoptysis From Dilated Systemic Aberrant Arteries Supplying Normal Lung Segments

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Summary. Hemoptysis is an uncommon presentation in children. It can be caused by several systemic and pulmonary disorders. Bleeding from an anomalous arterial supply to normal lung segments with no underlying pulmonary or cardiovascular disorders was widely reported in adults but is extremely rare in the pediatric age group. Here we describe 4 previously normal children and one girl with chronic lung disease, secondary to bronchopulmonary dysplasia, with acute and significant hemoptysis of variable severity. Extensive clinical and laboratory investigations did not identify any reasonable causes for their symptoms. Cardiac catheterization showed dilated anomalous systemic arteries that supplied a pulmonary arteriovenous malformation in one case and normal basal lung segments in the other 4 cases. Embolization of the anomalous arterial collaterals led to occlusion of these arteries and the cessation of further hemoptysis. **Pediatr Pulmonol.** 2004; 38:477–482. © 2004 Wiley-Liss, Inc.

Key words: hemoptysis; aberrant arteries; arteriovenous malformation.

INTRODUCTION

Hemoptysis is a very frightening and unusual symptom in the pediatric age group.¹ Diffuse pulmonary hemorrhage in all age groups can occur secondary to infection, vasculitis, cardiac failure, cardiovascular anomalies, immune deficiency, clotting disorders, pulmonary hypertension, alveolar injury, and exposure to various drugs and toxins.^{1–7} Idiopathic pulmonary hemosiderosis can be seen even in the absence of other etiologic factors. Lung bleeding due to idiopathic hemosiderosis is usually sporadic, but a subclinical chronic hemorrhage can also occur.¹ Bronchiectasis secondary to cystic fibrosis (CF) is the most common cause of hemoptysis in children in developed countries due to the growth and dilation of bronchial arteries secondary to the chronic inflammatory response in CF lungs.^{6,7}

Bronchial arteries supply a small amount of blood to different parts of the lung.⁷ Pulmonary bleeding from a pulmonary arteriovenous malformation (PAVM) or anomalous systemic or bronchial arteries was widely described in adults but is considered very rare in the pediatric age group.^{8–16} Here, we describe 5 children with severe or persistent hemoptysis. They had extensive but unremarkable laboratory, radiologic, and bronchoscopic evaluations. Cardiac catheterization showed anomalous systemic arteries that perfused normal right lung segments. In one child, these arteries supplied a single PAVM

during the patient's first episode of hemoptysis. Embolization of these arteries led to cessation of hemoptysis and prevention of further bleeding in all these children.

CASE REPORTS

Case 1

This is a 7-year-old white girl who had a choking episode while eating followed by severe coughing, expectoration, and vomiting of few clots of blood. Initially, she had a normal chest roentgenogram (CXR), normal complete blood count (CBC), clotting studies, and urine

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analysis (UA). Two days later, she had a repeat CXR that showed haziness in the right lower lobe (RLL) area. The child had rigid bronchoscopy which showed blood tinged secretions and a large clot in the bronchus intermedius extending to the right middle (RMLB) and right lower lobe (RLLB) bronchi. When the surgeon tried to gently pull the clot from the RMLB, she developed significant bleeding which was eventually controlled. She had a pulmonary and aortic catheterization which showed three systemic anomalous arteries coming off the proximal descending aorta and a fourth one from the right carotid and subclavian junction. All of these arteries were feeding a pulmonary arteriovenous malformation (PAVM) lesion in the RLL area. Coils were placed in all of these anomalous arteries, which led to termination of blood flow in these arteries and disappearance of the PAVM. Five years later, she was admitted again with several episodes of hemoptysis for 1 day. Another aortic angiography revealed several arterial collaterals that included one from the right innominate artery (RIA), two vessels from the descending aorta, and one vessel from the right internal mammary artery (RIMA). Several coils were placed in these anomalous collaterals. Figure 1 shows a repeat embolization of a collateral off the descending aorta. She was seen 15 months after the second episode, and has had no further hemoptysis.

Case 2

This is a 13-year-old African American boy who developed substernal chest pain and hemoptysis of bright red blood after strenuous exercise in the school gymnasium. He had no prior history of pulmonary or cardiac disorders. He had a normal physical examination, normal CBC, normal UA, normal erythrocyte sedimentation rate (ESR), negative antinuclear cytoplasmic antibody serology (ANCA), negative serology for antiglomerular basement membrane antibody (AGBMA), and a negative tuberculin (PPD) skin test. He also had a normal CXR, and normal chest CT. Aortic angiography showed several arterial collaterals originating from various levels of the left internal mammary artery (LIMA) and the midthoracic right intercostal artery which supplied the right lung and the pericardium. Coils were placed in the largest arterial collateral from the LIMA and the branch from the right intercostal artery (Fig. 2). No further hemoptysis episodes were noted following the coil embolization procedure for more than 18 months.

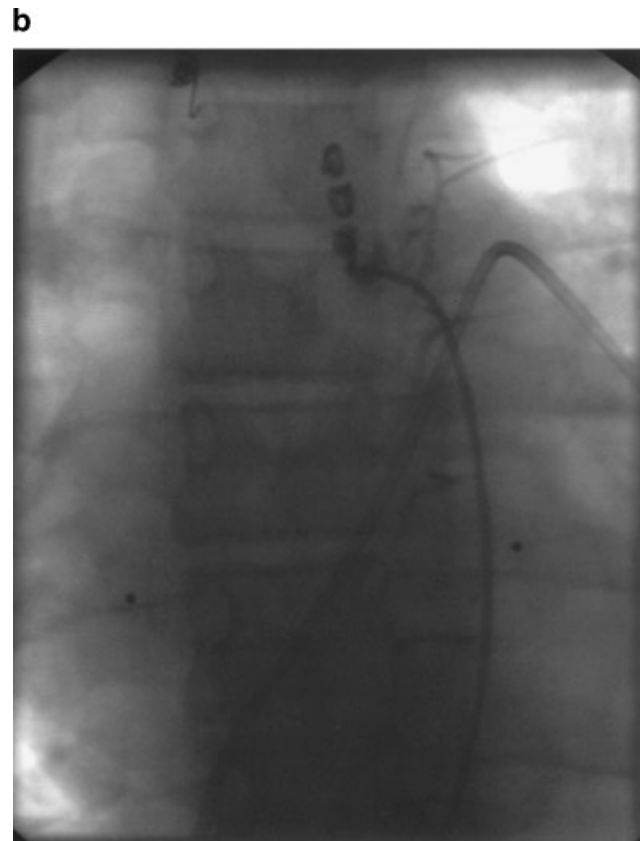


Fig. 1. Pre (a) and post (b) coil embolization of a large aorto-pulmonary systemic collateral arising off proximal descending aorta which perfused primarily right lower lobe segments. This collateral was previously embolized 5 years earlier, with patient redeveloping aorto-pulmonary collaterals proximal to previously placed coils.

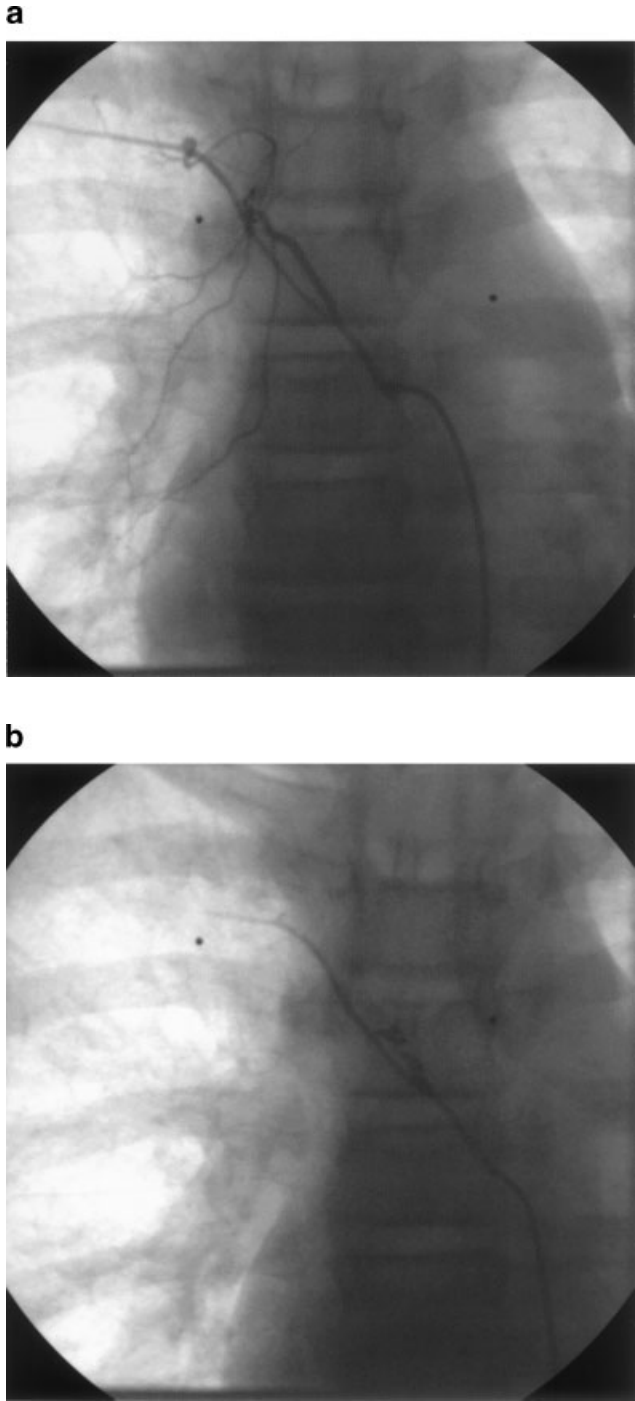


Fig. 2. Pre (a) and post (b) coil embolization of a small aortopulmonary collateral arising off right intercostal artery which perfused superior part of right lower lobe. Following coil embolization of aberrant vessel, right intercostal artery is better visualized.

Case 3

This is a 15-year-old white girl admitted for intermittent hemoptysis for 1 day. She expectorated 1–2 ounces of bright red blood each time. She was on daily nonsteroidal

anti-inflammatory medications (NSAID) for hip pain for about 4 weeks before this admission. Her examination was normal. Her laboratory data on admission included normal CBC, normal immunoglobulin levels, and normal UA. She also had normal serology for ANA, rheumatoid factor (RF), anti-DNA, AGBMA, and ANCA. The initial CXR showed opacity in the right lower lobe area. Spiral CT-scan of the chest showed opacity in the RML and RLL areas, and an irregular filling defect in the right main bronchus suspected to be a clot obstructing the airway. She had a cardiac catheterization which showed several anomalous arterial vessels coming off the RIA, transverse aortic arch, proximal descending aorta, and RIMA. All these vessels were supplying several segments of the RML and RLL areas. Some of these vessels were about 2 mm in diameter. Several coils were placed in the anomalous arteries (Fig. 3). She had a rigid bronchoscopy which revealed a large clot filling the bronchus intermedius, and parts of the clot were extracted. She continued to have coughing and developed wheezing for 2 days subsequently. A spirometry was done which showed reduced forced expiratory volume in 1 sec (FEV_1) and small airway flows indicating the presence of small airway obstruction. She was treated with nebulized albuterol for several days. Repeated CXRs showed gradual resolution of the RLL atelectasis. Sixteen days after the initial presentation, she had a normal CXR. In addition, the repeat spirometry showed normal lung volumes and small airway flows. She had no further hemoptysis for 18 months.

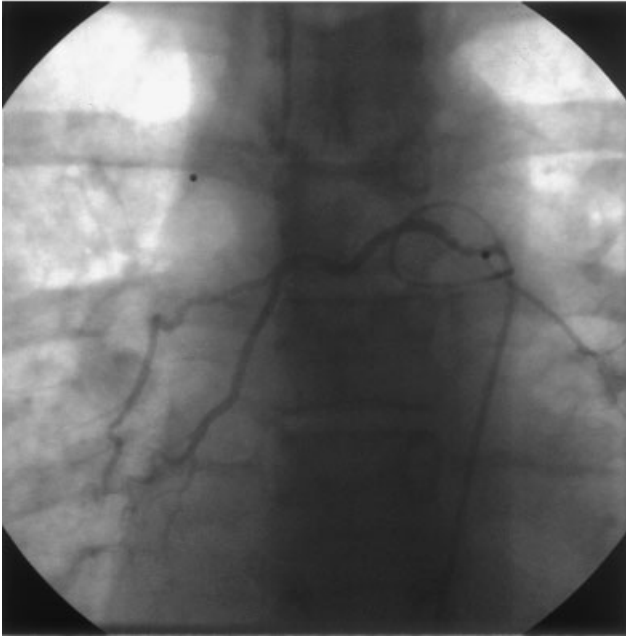
Case 4

A 7-year-old boy presented with episodes of “bringing up blood” intermittently for 6 months. He had about one-half to 1 ounce of blood with every episode. These episodes occurred both at home and in school. He had a history of mild asthma. He had a negative physical examination, normal CBC, normal clotting studies, negative serology for vasculitis, a negative PPD-skin test, a normal CXR, a normal chest CT-scan, and normal cardiac echocardiography. He also had normal serology for ANA, RF, anti-DNA, AGBMA, and ANCA. He had a normal rhinoscopy and upper airway examination, and a normal flexible bronchoscopy done when he was not actively bleeding. An aortic angiogram showed a relatively large anomalous artery arising from the aorta and going toward the right lung. Coils were placed in this artery, with cessation of blood flow afterwards. He did not have any further hemoptysis for 2 years.

Case 5

This child, who was born premature, had severe bronchopulmonary dysplasia and was on home mechanical ventilation for the first 2 years of her life. At 10 years

a



b

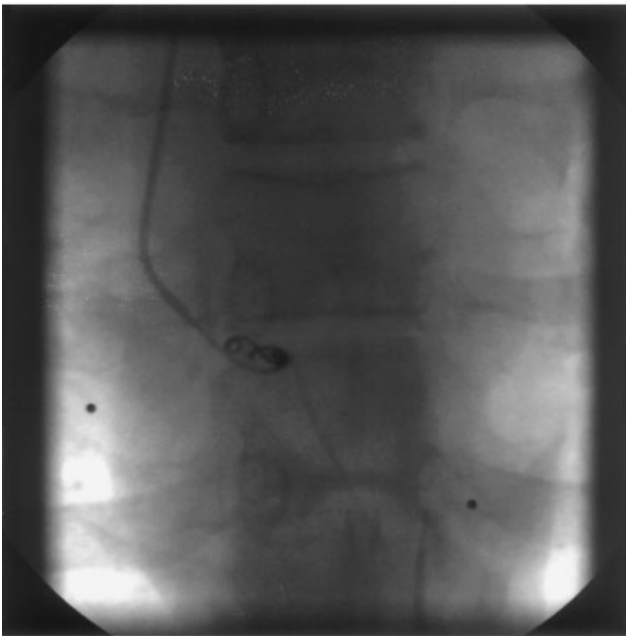


Fig. 3. Pre (a) and post (b) coil embolization of a large aortopulmonary systemic collateral arising off right innominate artery and proximal right subclavian artery. This aberrant vessel perfused right middle and right lower lobes.

of age, she choked suddenly on a piece of chicken and had considerable amounts of fresh red bloody secretions coming out of a small tracheostomy stoma. The child's respiratory status deteriorated rapidly, and she required intubation and mechanical ventilation. She had normal bronchoscopic examinations and a thorough but negative

evaluation for vasculitis, respiratory tract infections, and anatomical anomalies. She had a cardiac catheterization that showed a systemic arterial collateral coming directly from the aorta which was successfully coiled. The child did well after that, had no more pulmonary hemorrhages, and was successfully extubated. Five months later, she developed significant pulmonary bleeding, hypoxemia, and respiratory failure. She was placed again on mechanical ventilation for several days. Another cardiac catheterization was done, and this revealed several small systemic arterial collaterals that required coiling to obliterate their blood flow. The child had no further bleeding up to 4 months after the second episode.

DISCUSSION

The lung is the only organ supplied by two types of arterial and venous systems. Consequently, it receives and drains blood to both sides of the heart. Systemic arterial blood to the lung is provided through the bronchial arteries that accompany airway branching. Bronchial arteries normally receive 1–2% of the cardiac output, and provide oxygenation and nourishment to different parts of the lung and the visceral pleura.⁷

Extrasytemic arterial collaterals to normal lung segments and pulmonary bleeding from these vessels have been well-described in adults but seldom reported in children.^{8–16} These arteries usually arise from the aorta directly or from a few of its branches.¹⁷

Case 1 had a PAVM supplied by systemic arteries during the first episode of hemoptysis only. During the second episode, she had several new systemic arteries that supplied normal RLL with no any other vascular anomalies. She required embolization for both episodes to control her symptoms. PAVM is not a common vascular anomaly, especially in the pediatric age group.^{18–22} The incidence of PAVM is 2–3 per 100,000 population.^{19,20} About 70% of cases of PAVM are associated with hereditary hemorrhagic telangiectasia (HHT), and 15–35% of patients with HHT have PAVM.^{20,23} In 95% of cases, PAVM is supplied by pulmonary arteries, while a connection to the systemic arteries is very rare.²¹ Only a few cases of PAVM were described with a dual pulmonary and systemic arterial supply.²⁴ A PAVM connection to the bronchial arteries may rarely occur with bronchiectasis, or as a reaction to local tissue hypoxia induced by shunting at the malformation or a prior surgical manipulation.^{12,20,25,26}

Symptoms in patients with PAVM occur in about 72% of subjects, either from the PAVM it self or from HHT lesions in other organs. These include epistaxis, dyspnea, cyanosis, clubbing, fatigue, heart failure, cerebrovascular accidents, hemoptysis, or hemothorax.^{10,18,20} PAVM-related symptoms commonly occur between the fourth and sixth decades of life.^{18,20} Hemoptysis is not a common complication of PAVM, especially in children.^{18,20,27}

Patients susceptible to hemoptysis are typically cyanotic and have PAVMs that are either large, multiple, or supplied by systemic arteries. These types of lesions are very rare in the pediatric age group. We are aware of only two case reports of two 11-year-old girls with hemoptysis from a PAVM supplied by a single bronchial artery in each one of them.^{28,29} Embolization of the feeding bronchial arteries led to cessation of hemoptysis in each of them. Case 1 in our report had an unusual presentation and several unique features. She had two episodes of hemoptysis, with the first episode occurring at a younger age than the other 2 previously reported children.^{28,29} Her aortography, during each episode, showed several relatively large systemic arterial collaterals instead of a single enlarged bronchial artery. The vessels identified during her second episode supplied the right lung directly, and they could have originated either from hypertrophy and dilation of preexisting collaterals or as a reaction to vascular manipulation during the first embolization. Revascularization or development of new extensive collaterals may occur in several disorders after bronchial artery occlusion.^{7,11,13} Embolization of the arterial supply to a PAVM was successfully performed in adults since it was first reported by Porstmann in 1977.^{18,20}

The presence of an anomalous arterial collateral supply to normal lung segments is well-reported in adults, but is much less common in children.⁹⁻¹⁵ According to a recent review of the English-language literature that looked at anomalous systemic arterial supplies as the sole blood supply to normal basal segments of the lower lobes, 6 of 12 identified patients were children whose ages ranged from newborn to 6 years.¹⁶ The same authors found 4 more patients who had an anomalous arterial supply to normal lower lobes with an intact and normal pulmonary arterial supply. Two of these patients were 14 months and 3 years in age, while the other 2 were adults. Therefore, according to this report, a total of 8 children with anomalous arterial collaterals to normal lower lung segments were identified.¹⁶ Another article reviewed all pediatric surgical lung specimens and autopsies collected over a period of 29 years with aberrant systemic arteries in normal or abnormal lung segments.³⁰ Fifteen cases were identified, and 9 of these (4 autopsies and 5 specimens) had aberrant systemic arteries to normal lung tissue. These cases had several characteristics that included: young age (3 days to 2 years), predominance of female gender, aberrant arteries arising most frequently from the abdominal aorta and supplying the right lower lobes, and presence of cardiac anomalies in 8 cases. Respiratory distress, pneumonia, and failure to thrive were the main presenting symptoms in affected children.³⁰ The same authors reviewed the literature and identified 13 more pediatric specimens among 22 reported cases of aberrant systemic arterial supply to normal lungs. Adult patients with dilated bronchial or anomalous arteries to normal lung segments were either

asymptomatic or presented with several symptoms, including hemoptysis.^{11,12,16,31} Unlike adults, the most common symptom in affected children is cardiac murmur rather than hemoptysis.^{16,30,31} None of the 9 pediatric cases identified by Holder and Langston had hemoptysis or a clinically relevant pulmonary hemorrhage, even in the presence of hemosiderin-laden macrophages in some biopsies.³⁰ This is further supported by Yamanaka et al.¹⁶ The majority of adults listed in their review had hemoptysis, while all 8 children had heart murmurs without hemoptysis.¹⁶

To our knowledge, only one previously healthy 11-year-old boy was recently reported with hemoptysis, attributed to bleeding from a normal bronchial artery that supplied a normal lung.³² This child's bleeding was from a normal right bronchial artery, and he did not have any other systemic collaterals with a usual origin or distribution. Therefore, the initial presentation of recurrent or significant hemoptysis in our cases is a unique and rarely reported symptom in children. No dilated vessels were seen in our patients' radiographic studies. Although chest CT scans and CT angiography may detect anomalous systemic arterial branching, it is not unusual to have normal chest radiographs and CT-scan studies, even in adults with massive hemoptysis.^{13,14,33}

Case 3 developed wheezing, coughing, and small airway obstruction. These symptoms probably occurred as a result of irritation of the bronchi, causing bronchospasm and production of a large amount of mucus in response to the presence of blood in the affected airways.³⁴

In conclusion, hemoptysis is a very unusual symptom in children. Thorough clinical laboratory evaluations are often required to determine the etiology of the bleeding. A table by Sheikh et al.³² listed the various causes of hemoptysis and required investigations, and the reader can refer to this table for further information. Pulmonary and systemic aortography may be required to identify any significant vascular anomalies or aberrant systemic arteries causing hemoptysis. Hemodynamic data obtained from pulmonary or aortic angiography can provide useful information that may aid in the diagnosis and treatment of pulmonary bleeding. Aortic angiography and embolization of abnormally dilated vessels or vascular malformations can be a safe and effective therapeutic intervention in children.

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