A 7-Year-Old Girl with Hemoptysis for 3 Days

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A 7-year-old girl presented to our unit because of hemoptysis for 3 days. She had hemoptysis one to two times per day with approximately 2 to 3 mL of blood each time. The blood was bright red without clots. Amoxicillin and ethamsylate were administered for 3 days in the local hospital. She had previous episodes of hemoptysis at age 1.5 years and age 4 years. During the second episode of hemoptysis (at age 4 years), on physical examination, she had a normal heart rate and blood pressure but appeared acutely ill. Coarse breath sounds were noted on lung auscultation. A chest radiograph showed increased lung markings in the right lung, and the mediastinum and bronchi were shifted to the right (Figure 1). Doppler echocardiography showed normal structure of the heart and the right pulmonary vein was not clearly seen. Computed tomography angiography (CTA) in the local hospital showed smaller right pulmonary artery and undeveloped right pulmonary vein with normal airways. Bronchoscopy showed manifestations of bronchitis, and bronchoalveolar lavage demonstrated no hemosiderin cells.

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Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pedann@Healio.com.
Hemoglobin, platelets, prothrombin time, thrombin time, activated partial thromboplastin time, fibrinogen, and D-dimer were within the normal range. Screening laboratory tests for urine, stool, C-reactive protein, procalcitonin, erythrocyte sedimentation rate, liver and kidney function, blood gas analysis, and TORCH (Toxoplasmosis, Other diseases including HIV, syphilis, and measles, Rubella, Cytomegalovirus, and Herpes simplex) immunoglobulin M, were all within normal limits or were negative.

Chest CT showed smaller right thoracic cage and hilum of the lung. The density of the right lung field was higher than that of the left. A cloudy, patchy shadow without clear boundary was widely scattered in the right lung. CTA showed absence of the right pulmonary veins, thin right pulmonary artery with fewer branches, dilatation of right bronchial vessels, and collateral circulation (Figure 2). A soft tissue mass was noted in the right middle and rear mediastinum, and the blood supply was mainly from the thoracic aorta. Mild right pleural thickening and enlarged heart were noted.

Etamsylate and aminomethylbenzoic acid were administered for 5 days. Inhaled epinephrine was administered 2 times a day for 2 days. The hemoptysis disappeared on the second day of hospitalization. After the diagnosis was confirmed her parents refused further treatment, and so the patient was discharged on the fifth day of hospitalization.

**DISCUSSION**

Multiple vascular and pulmonary malformations in one patient are rare. In this case, two unilateral vascular malformations (pulmonary vein atresia and artery hypoplasia) and two pulmonary malformations (pulmonary hypoplasia and pulmonary sequestration) were found. Although right pleural thickening, right bronchial vessel and collateral circulation dilatation, and cardiac enlargement were noted in this patient, these findings could be secondary to recurrent infection due to pulmonary malformations and compensatory for the right pulmonary vein atresia and artery hypoplasia. To our knowledge, this is the first report of these four malformations found in one patient.

The mechanism of development of these four main vascular and pulmonary malformations is still unclear. As the patient presented with hemoptysis at age 1.5 years without a history of serious infection, tumors, thrombosis, or radiofrequency ablation, we concluded that the right pulmonary vein atresia and artery hypoplasia were congenital. Because these malformations occurred on the same side, and because pulmonary sequestration is closely associated with an abnormality of blood supply, and because some conditions that affect the pulmonary vascular bed may induce pulmonary hypoplasia, we speculated that pulmonary vascular malformations due to congenital defects of germ or embryo quality protoplasm were the primary abnormalities that then induced the other pulmonary malformations, including pulmonary hypoplasia and pulmonary sequestration.

Unilateral pulmonary vein atresia, artery hypoplasia, and pulmonary hypoplasia without other abnormality may be asymptomatic in childhood. Similar to our patient, hemoptysis is
the chief complaint, and it is caused by the collateral circulation dilation resulting from the pulmonary vein atresia.\textsuperscript{3} However, hemoptysis was not frequent or serious in our patient. We speculate that the less-frequent hemoptysis in our patient occurred because of the associated pulmonary artery hypoplasia and pulmonary hypoplasia, which may delay the hypertension and dilatation of collateral circulation, although dilatation of collateral circulation was confirmed by CTA. Decreased exercise tolerance, and even dyspnea, has also been reported secondary to decreased gas exchange due to pulmonary circulation abnormality, secondary pulmonary hypertension, and pulmonary hypoplasia.\textsuperscript{3} Recurrent lung infection may occur due to pulmonary hypoplasia and pulmonary sequestration, especially in the intralobar type.\textsuperscript{5}

Because there may be few symptoms and only nonspecific physical findings in these conditions, the diagnosis is often missed during infancy and childhood unless there is a high index of suspicion. In our patient, the diagnosis was not made until 5 years after the first episode of hemoptysis. Chest radiography, CT, and magnetic resonance imaging may reveal the sites of these malformations. CTA is a useful method to show the distribution and course of the pulmonary vessel and bronchi vessels, and to demonstrate the systemic vessels supplying the pulmonary sequestration.\textsuperscript{5,6} The therapeutic methods are based on the severity of the clinical presentation. In a patient with asymptomatic or mild symptoms, conservative treatment and close follow up are required to monitor pulmonary hypertension and other complications.\textsuperscript{4} In patients with serious complications, percutaneous balloon valvuloplasty, vascular anastomosis, or pneumonectomy should be considered to prevent the exacerbations.\textsuperscript{4,7}

**CONCLUSION**

Pulmonary vascular malformations may need to be considered for a child with recurrent hemoptysis alone or accompanied by pulmonary hypoplasia or pulmonary sequestration. Further investigation (eg, CTA) is required to confirm the diagnosis.

**REFERENCES**