A ruptured pulmonary arteriovenous fistula

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A 26-year-old pregnant woman presented to our hospital with a 2-month history of progressive dizziness and

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dyspnea. There was no recent history of fever or thoracic trauma. The physical examination revealed low breath sounds in the left lower lung. Contrast-enhanced chest computed tomography showed a massive left pleural effusion with suspicion of a vascular malformation in the left lower lobe (Figure 1, A, asterisks), which needling proved to be a hemothorax. Pulmonary angiography and 3-dimensional reconstruction confirmed the presence of 2 different sizes of pulmonary arteriovenous fistulas (PAVFs) between the lower left pulmonary artery and the pulmonary vein in the left lower lung (Figure 1, B-D, arrows and arrowheads). After the diagnosis was confirmed, an emergency left posterolateral thoracotomy was performed. During the procedure, the larger PAVF was visible on the surface of the lung, and no active bleeding was observed. To avoid recurrence of hemorrhage, the left lower lobe was

FIGURE 1. A, Chest computed tomography scan shows a massive left pleural effusion with suspicion of a vascular malformation in the left lower lobe (asterisks). B-D, Pulmonary angiography and 3-dimensional reconstruction confirm the presence of 2 different sizes of PAVFs between the lower left pulmonary artery and the pulmonary vein in the left lower lung (arrows and arrowheads).

completely resected after the removal of hemorrhage and blood clots in the thorax. Postoperative recovery was uneventful. No recurrence of symptoms was observed during a 5-year follow-up.

PAVFs are rare vascular malformations of the lung. The cause may be congenital or acquired. Although more than half of patients with PAVFs are asymptomatic, the typical symptoms of fatigability, exertional dyspnea, and palpitations are frequently found in some patients. Hemothorax

due to spontaneous rupture of a fistula sac is rare and usually occurs in pregnancy. The increased blood volume, cardiac output, and venous distensibility could be strongly associated with the life-threatening complication during this special period. Therapeutic options include angiographic embolization and surgical excision. However, once the spontaneous rupture of PAVFs is suspected, the posterolateral thoracotomy with wedge resection or lobectomy is the preferential choice because of low mortality and recurrence rate.

EDITORIAL COMMENTARY

Pulmonary arteriovenous malformations: The consequences of bypassing the capillary bed

Sunil J. Ghelani, MD, and Rahul H. Rathod, MD

See related article on pages 716-7.

“Loud roaring pulmonary systolic bruit and very highly accentuated second sound were present during life, with haemoptysis, epistaxis, and dropsy,” Churton described while presenting the clinical findings of the first documented case of pulmonary arteriovenous malformation (PAVM) in a 12-year-old boy in 1897.¹ He went on to describe the autopsy findings: “Four of the secondary branches in one lung, and three in the other, led to aneurysms as large as walnuts, filled with blood clot.”

PAVMs represent abnormal communications between an artery and a pulmonary vein that allow blood to bypass the pulmonary capillary bed. Usually the arterial supply is from 1 or more branches of a pulmonary artery, physiologically leading to a right-to-left shunt, although rarely a systemic artery may feed the malformation. More than 80% of PAVMs are congenital, and their precise etiology is unknown. A strong association is noted between hereditary hemorrhagic telangiectasia (HHT), also known as *Osler-Weber-Rendu disease*, and PAVMs. More than three-

quarters of patients with macroscopic PAVMs have HHT; conversely, about a quarter of patients with HHT have PAVMs.²⁻⁴ Acquired PAVMs are less common, and the 2 common associations are hepatic cirrhosis and surgery for congenital heart disease.⁵ Rarely, PAVMs may be associated with chest trauma, lung parasites (schistosomiasis), metastatic lung disease (thyroid carcinoma), and Fanconi syndrome. Lesions are often classified as macrovascular or microvascular. Microvascular PAVMs are typical for acquired forms, such as those associated with cyanotic congenital heart disease.

Symptoms of PAVMs result from a combination of right-to-left shunting, ventricular volume overload, and associated lung parenchymal disease. The presentation usually occurs when the patient is a young adult, and common presenting symptoms include dyspnea and cough. In rare cases, patients are first seen in overt heart failure with edema and ascites. Children are often initially seen with symptoms related to other manifestations of HHT (epistaxis from nasal telangiectasia or skin lesions) rather than with cardiorespiratory symptoms. Cyanosis, clubbing, and related complications such as brain abscess were commonly reported in older series but are less common in the modern era.

In this issue of the *Journal*, Lin and colleagues⁶ describe an interesting case of a ruptured PAVM in a 26-year-old woman. The PAVM was a large, solitary lesion in the left lower lobe of the lung, which is the most common location for such lesions.⁷ The patient had a successful diagnosis made by means of contrast-enhanced computed tomography and was surgically treated with an excellent outcome.

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