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

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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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FIGURE 1. Proposed management algorithm for suspected macrovascular pulmonary arteriovenous malformations.

Massive hemorrhage in the form of hemothorax or hemoptysis is an uncommon but life-threatening complication of PAVMs. In a series of 143 patients referred for transcatheter embolotherapy, 11 (8%) had a hemothorax requiring hospitalization or involving massive hemoptysis.⁸ In this series, massive hemorrhage was the presenting symptom in 9 of the 11 subjects, similar to the described case. The patient whose case is presented by Lin and colleagues⁶ was also pregnant at the time of presentation. Worsening of PAVM with pregnancy has been reported by multiple studies, and approximately 30% of massive hemorrhagic events occur during pregnancy.⁸⁻¹⁰

Diagnostics tests for PAVMs have evolved during the last few decades.¹¹ The 2011 guidelines from the HHT working group recommend the use of transthoracic agitated saline solution contrast-enhanced echocardiography as the first line test to screen all patients with HHT for presence of PAVMs.¹² The current recommendation is to follow-up the presence of any bubble in the left atrium with

noncontrast thin-cut computed tomography. Although magnetic resonance imaging is not widely recommended as a screening tool for PAVMs, several studies have shown promising results, and it is being increasingly used as a diagnostic test.¹³

Current first-line treatment for PAVMs is transcatheter embolization therapy. An algorithm for management of patients with suspected macrovascular PAVMs is presented in Figure 1. The goals of treatment are symptom relief, prevention of neurologic complications, prevention of cardiac failure, and elimination of hypoxia and related complications. The decision to treat subjects without symptoms should be made on a case-by-case basis, and factors to consider include the size of the lesion and the diameter of the feeding artery. Results of embolization therapy have generally been excellent, with few failures and rare major complications.⁶ Surgical resection is typically recommended for large PAVMs with feeding arteries that are larger than 3 mm in diameter. There is increasing experience

with using newer catheter-based devices and plugs to occlude vessels of larger diameter in the catheterization laboratory.¹⁴

Treatment of diffuse microvascular PAVMs, such as are seen in patients with palliated single-ventricle heart disease, is more difficult. Microvascular PAVMs are often too small to visualize on fluoroscopy, and even if they are seen, they will recur after catheter-based embolization. The impetus for growth of these PAVMs is thought to be exclusion of hepatic venous blood from the pulmonary circulation.⁵ In an attempt to increase pulmonary blood flow and allow delivery of “hepatic factor” to the pulmonary vascular bed, fistulas from the brachial and axillary artery to the venous circulation have been created. These systemic arteriovenous fistulas allow part of the hepatic blood to bypass the systemic capillary bed and have been shown to decrease PAVMs.^{15,16} Alternatively, in the patient with single-ventricle heart disease, modifications to the Fontan procedure allow the symmetric distribution of hepatic blood flow to both lungs.

Lifelong follow-up is recommended for patients with PAVMs, including those who have been treated, because reperfusion of previously embolized vessels can occur. Follow-up should include cross-sectional imaging, because transthoracic agitated saline solution contrast-enhanced echocardiography continues to yield positive results in more than 90% of patients after catheter embolization.¹⁷ In addition, antibiotic prophylaxis is recommended for prevention of brain abscesses. To avoid air embolism, scuba diving is discouraged, and extra caution is recommended when these patients need any form of systemic intravenous access.

To summarize, the case presented by Lin and colleagues⁶ demonstrates the potentially life-threatening nature of PAVMs, especially in a pregnant woman. These lesions have a strong association with HHT. Diagnosis is facilitated by transthoracic agitated saline solution contrast-enhanced echocardiography and computed tomography, although magnetic resonance imaging is increasingly being used. Transcatheter thromboembolization is the treatment of choice in most cases, although

surgical resection is often required in patients with large-diameter feeding vessels.

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