Pulmonary Arteriovenous Malformations and Risk of Stroke

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KEYWORDS

• Pulmonary arteriovenous malformations • Stroke • Risk • Hereditary hemorrhagic telangiectasia • PAVMs

KEY POINTS

- The incidence of pulmonary arteriovenous malformations (PAVMs) is about 2 to 3 cases per 100,000.
- Most PAVMs (50%–80%) occur in patients with hereditary hemorrhagic telangiectasia.
- Hypoxemia and orthodeoxia are some of the more common clinical presentations.
- Complications include polycythemia, paradoxic systemic (septic) emboli, rupture, migraine, and seizures.
- The estimated risk of stroke secondary to PAVMs is as high as 2.6% to 25.0%.
- A combination of chest radiograph and contrast echocardiography is a good screening evaluation in patients with suspected PAVMs, followed by a high-resolution computed tomography scan.
- Transcatheter therapy is the most suitable therapeutic option in most patients with PAVMs, and a variety of devices can be used to occlude the PAVMs.

INTRODUCTION

Pulmonary arteriovenous malformations (PAVMs) are an abnormal communication between pulmonary arteries and pulmonary veins. They are rare, with an incidence of about 3.2 to 4.5 cases per year[1] or about 2 to 3 per 100,000.[2] Most PAVMs are hereditary; about 50% to 80% occur in patients with hereditary hemorrhagic telangiectasia (HHT), whereas about 10% to 30% of patients with HHT develop PAVMs.[3,4] PAVMs can also present as a congenital malformation or be acquired secondary to liver disease or associated with lack of hepatic flow to the pulmonary circulation, such as in patients with congenital heart disease who have undergone a Glenn procedure. It has been suggested that the absence of a liver-derived inhibitor of vascular proliferation is responsible for some of the acquired PAVMs.[5] Pathologically, PAVMs often appear as dilated vascular channels with abnormalities in the makeup of the vessel wall and may occur either as an isolated malformation (Fig. 1) or as multiple (Fig. 2) or diffuse (Fig. 3) anomalies; about 50% to 70% are located in the lower lobes.[5,6] They may be recurrent, in particular in patients with HHT.

CLINICAL FINDINGS AND DIAGNOSIS

The spectrum of clinical presentations is wide. Hypoxemia and orthodeoxia are some of the more common clinical presentations, and it is not unusual for a PAVM being diagnosed as part of a workup for a suspected patent foramen ovale (PFO) with right-left shunting. Depending

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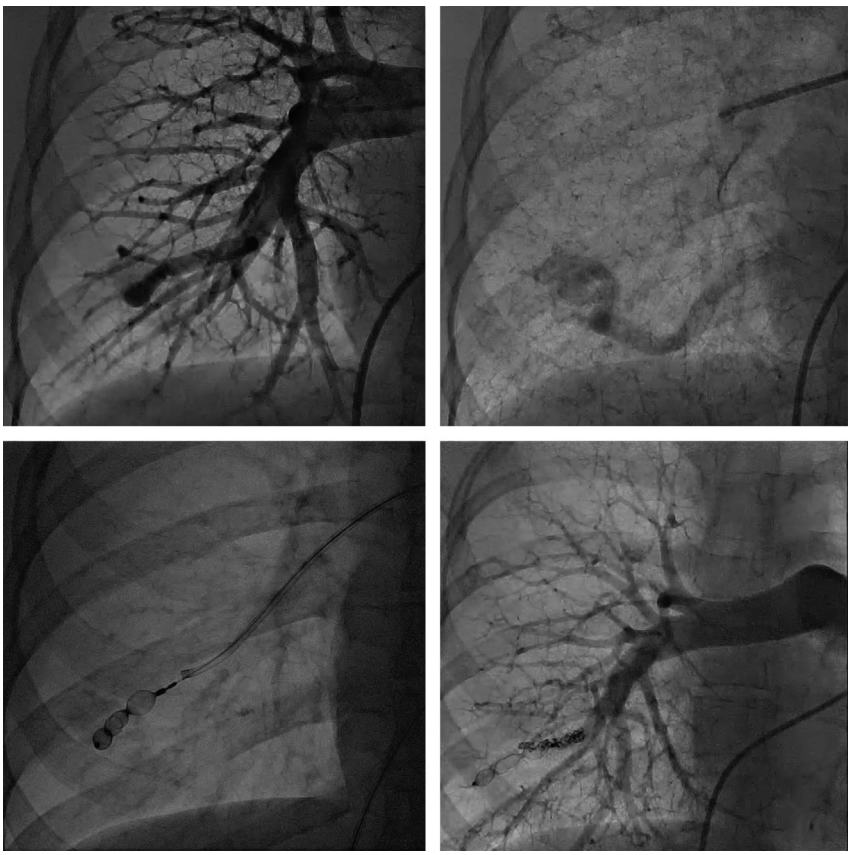

Fig. 1. Isolated PAVM. Isolated PAVM in the right lower lobe draining to the right lower pulmonary vein. The feeder vessel was occluded by placing initially an Amplatzer vascular plug II distally, followed by placement of several nondetachable coils. (*Top left*) Feeder vessel of the PAVM in the right lower lobe. (*Top right*) Draining vessel to the right lower pulmonary vein. (*Bottom left*) Vascular plug II deployed in the feeder vessel. (*Bottom right*) Right pulmonary artery angiography after release of the plug and placement of several additional nondetachable coils. (*Courtesy of* John Cheatham, MD, Columbus, OH.)

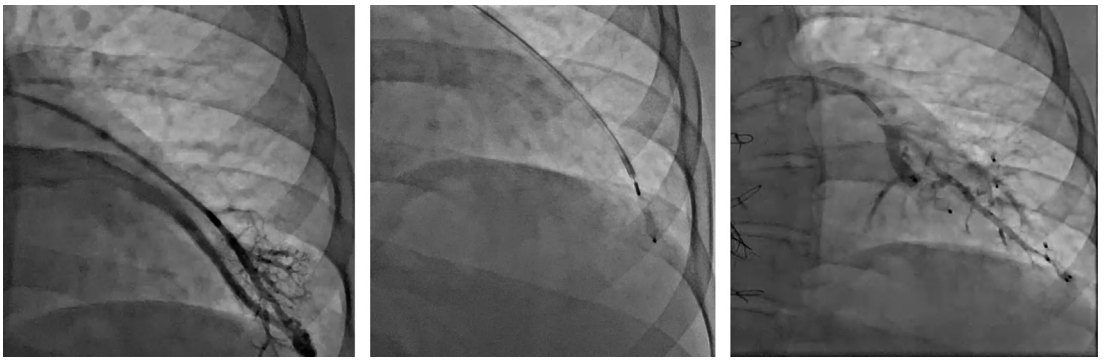

Fig. 2. Multiple PAVMs. Multiple PAVMs in a patient with cyanotic congenital heart disease. (*Top*) One of the PAVMs shown on angiography to the left lower lobe and draining to the left lower pulmonary vein. (*Middle*) Deployment of an Amplatzer vascular plug type IV. (*Bottom*) Angiography after occlusion of multiple AVMs, documenting the various vascular plugs (type II and IV) in different segments of the left lower lobe. (*Courtesy of* John Cheatham, MD, Columbus, OH.)