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IMAGES IN INTERVENTION

Transcatheter Closure of Pulmonary Arteriovenous Malformation to Facilitate Treatment of Pulmonary Arterial Hypertension

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A 60-year-old Caucasian woman was admitted to our institution with new onset progressive dyspnea for 2 to 3 weeks and worsening of chronic lower extremity edema or abdominal distension. She had a history of cirrhosis (significant alcohol intake and hepatitis C), portal hypertension, and an intracranial aneurysm. Her physical examination was significant for tachypnea, jugular venous distension, a parasternal heave, holosystolic murmur at the left lower sternal border, and marked pitting edema in lower extremities. Her oxygen saturation (sAO_2) was 70% while on room air—an arterial blood gas analysis done with 100% fraction of inspired oxygen revealed oxygen saturation of 89% and a partial pressure of oxygen of 64 mm Hg, thus giving a Qs/Qt shunt fraction of 26%. A chest radiograph was unrevealing except for a "fluffy" nodule in the left lower lobe (**Figure 1A**). This was revealed to be a large pulmonary arteriovenous malformation (PAVM) originating from the left lower lobe branch of the pulmonary artery (PA) and feeding into the left lower pulmonary vein by computed tomography angiogram (**Figures 1B and 1C**). An echocardiogram revealed a dilated right ventricle with a markedly positive bubble study for an intrapulmonary shunt with bubbles noted in the left atrium 4 cycles after the right atrium (RA) (**Figure 1D**). The patient then underwent a hemodynamic study, revealing severe pulmonary hypertension with PA pressure 56/36 mm Hg (mean 47 mm Hg), RA 22 mm Hg, pulmonary capillary wedge pressure 12 mm Hg, pulmonary vascular resistance (PVR) 6.8 WU, and Fick cardiac index 2.54 l/min. The Qp/Qs was 0.60. Pulmonary angiography corroborated the computed tomography findings of a PAVM in the left lower lobe (**Figure 1E**, Online Video 1). Thus, the patient had pre-capillary (World Health Organization group 1) PA hypertension (PAH) likely secondary to cirrhosis. There was no family history of pulmonary hypertension, and other etiologies including connective tissue disease, chronic thromboembolic pulmonary hypertension, sleep apnea, and human immunodeficiency virus were excluded. The significance of the PAVM was unclear, as it might have enlarged secondary to PAH or the increased flow through it and the resultant hypoxemia may be a contributory factor for PAH. The presence of severe baseline hypoxemia and high oxygen requirements made the use of pulmonary vasodilators challenging, given the potential for worsening right-to-left shunting with vasodilators.

Thus, we elected to close the PAVM with an Amplatzer vascular plug II (St. Jude Medical, Plymouth, Minnesota). After the device was deployed, we observed the hemodynamics for an extended period of time (**Figure 1F**, Online Video 2): the PA pressures decreased slightly, the right atrial pressure remained stable, and there was only a slight drop in cardiac output after 30 min (PA pressure 55/27 mm Hg with a mean of 39 mm Hg, RA 20 mm Hg, pulmonary capillary wedge pressure 9 mm Hg, PVR 6 WU, Fick cardiac index 2.50 l/min); the device was then released. Oxygen saturation quickly improved to >95% on 27% fraction of inspired oxygen. She was initially started on tadalafil and ambrisentan but with marked reduction of PA pressures at 3 months

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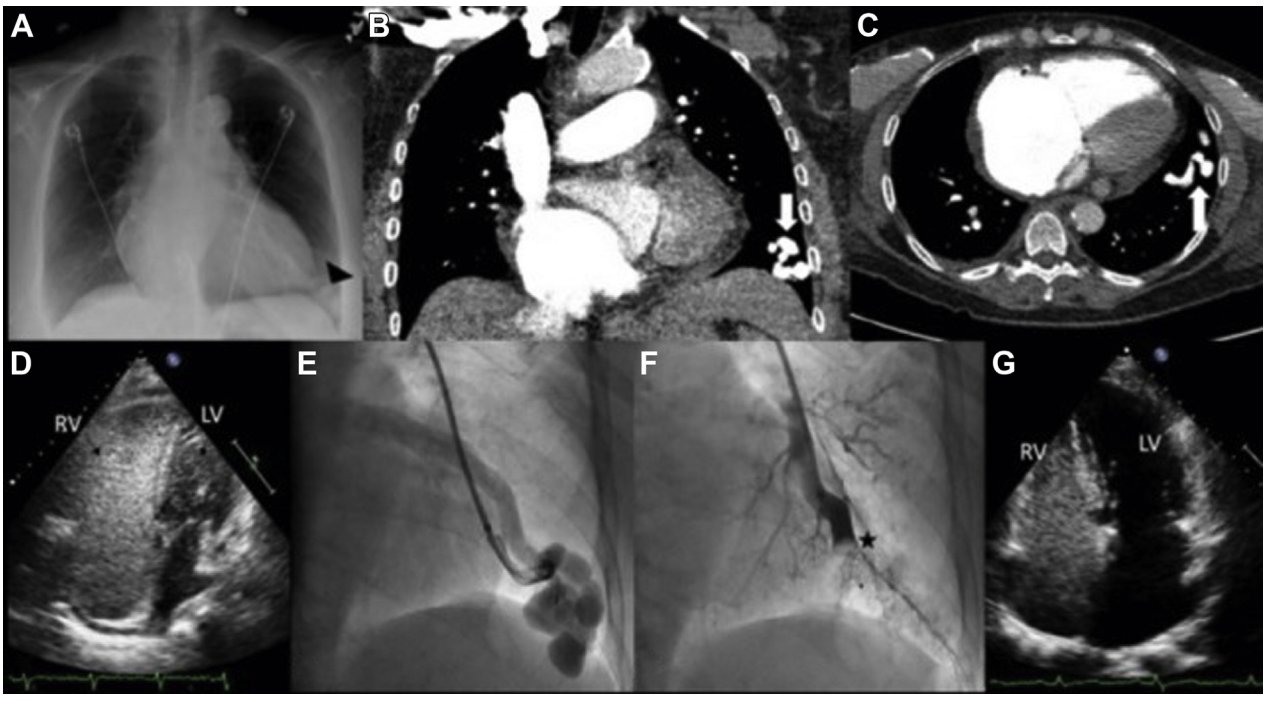
FIGURE 1 Transcatheter Closure of PAVM in a Patient With Severe Pulmonary Arterial Hypertension



(A) Chest x-ray with left lower lobe nodule **(black arrowhead)**. **(B, C)** Computed tomography angiogram showing left lower lobe pulmonary arteriovenous malformation (PAVM) **(white block arrows)**. **(D)** Contrast echocardiogram showing markedly positive right-to-left shunting and dilated right ventricle (RV). **(E)** Selective pulmonary angiogram delineating the PAVM (Online Video 1). **(F)** Thrombosed PAVM after deployment of an Amplatzer vascular plug II **(asterisk)** device (St. Jude Medical, Plymouth, Minnesota) (Online Video 2). **(G)** Contrast echocardiogram 5 months after closure showing improved RV size and no residual shunting. LV = left ventricle.

(PA pressure 35/8 mm Hg, RA 3 mm Hg, pulmonary capillary wedge pressure 6 mm Hg, PVR 2.9 WU, Fick cardiac index 2.8 l/min) by right heart catheterization, ambrisentan was discontinued and she did not need supplemental oxygen any more. The right ventricular function also improved remarkably, with no evidence of residual intrapulmonary shunting (**Figure 1G**).

PAH coexists with PAVM in a minority of patients only and is commonly associated with hereditary hemorrhagic telangiectasia syndrome (1). Our patient did not appear to be at high likelihood of hemorrhagic telangiectasia syndrome because of the absence of any nosebleeds or mucocutaneous telangiectasias or other AVMs or family history. Significant PAH has been traditionally considered a contraindication to closure of PAVM because of reports of worsened PAH and mortality with attempts at such cases (2). A large recent series of PAVM embolization cases thus also excluded patients with severe PAH with the same rationale (3). Although that is certainly a possibility, our case demonstrates that placement of a retrievable device with careful extended hemodynamic monitoring before release may be considered in select patients and may actually facilitate PAH treatment.

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KEY WORDS pulmonary AVM, pulmonary hypertension, transcatheter closure

APPENDIX For supplemental videos, please see the online version of this paper.