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CASE REPORT

ADVANCED

CLINICAL CASE

Retrograde Transseptal Pulmonary Vein Transcatheter Plug Closure for Pulmonary Arteriovenous Malformation

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ABSTRACT

Pulmonary arteriovenous malformations (PAVMs) may manifest with bleeding or embolic events necessitating intervention. Transcatheter coil embolization through the pulmonary artery (PA) is an established approach. We present a case of recurrent PAVMs despite numerous PA coil embolizations. PAVM occlusion was achieved through plug placement by a transseptal and pulmonary venous approach. **(Level of Difficulty: Advanced.)** (J Am Coll Cardiol Case Rep 2022;4:150–153)

HISTORY OF PRESENTATION

A 60-year-old man experienced 2 consecutive episodes of left-sided hemiplegia associated with chest tightness, cough, and migraine with visual aura for which he presented to the emergency department for evaluation. On presentation, the patient was asymptomatic, physical examination was unremarkable, and vital signs were stable, as follows: temperature, 36.3 °C; blood pressure, 121/70 mm Hg; heart rate, 64 beats/min; and respiratory rate, 15 breaths/min.

LEARNING OBJECTIVES

- To summarize the current practice and limitations of transcatheter device deployment for PAVMs.
- To recommend the role of transseptal retrograde PV plug placement.
- To promote the efficacy of a multidiscipline approach in transseptal PV plugging for PAVMs.

PAST MEDICAL HISTORY

The patient had a history of Hodgkin lymphoma, hypertension, hyperlipidemia, and hereditary hemorrhagic telangiectasia (HHT) with multiple pulmonary arteriovenous malformations (PAVMs) requiring 31 coil embolizations. Although his symptoms initially improved following coil embolization, they would recur with the formation of new PAVMs.

DIFFERENTIAL DIAGNOSIS

The episodes of left-sided hemiplegia mirrored the patient's previous symptoms of paradoxical embolism in the setting of PAVM, thus making recanalization of his existing PAVM the most likely diagnosis. Alternative diagnoses included arterial transient ischemic attack or stroke and migraine without an embolic event.

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INVESTIGATIONS

Results of computed tomography (CT) of the head were negative for acute disease. Results of CT angiography of the chest were negative for new PAVM, although imaging was limited by the presence of coils. There were multiple acute segmental and subsegmental right lower lobe pulmonary emboli in the absence of acute deep vein thrombosis, likely related to the recent coiling procedures, given his contraindication to anticoagulation. Transesophageal echocardiography results were consistent with large right upper and moderate left upper pulmonary vein (PV) intrapulmonary shunts, findings supporting incomplete embolization or recanalization of the recent coil-embolized right upper lobe PAVM.

MANAGEMENT

As previously, transcatheter pulmonary artery (PA) coil embolization was attempted, but we were unable to achieve satisfactory positioning, and no coil was deployed. Accordingly, a retrograde approach using transseptal puncture and PV access was considered to occlude flow in this segment with a transcatheter plug device. Access was obtained in the right internal jugular vein to facilitate PA pigtail catheter placement for visualization, right femoral vein 14-F access for transseptal puncture and plug deployment and 10-F left femoral vein access for 10-F intracardiac echocardiography guidance. A transseptal puncture was performed with a medium-curve Agilis introducer (Abbott) and a 98-cm BRK needle (St Jude Medical) guided by intracardiac echocardiography and biplane fluoroscopy. An 8-F multipurpose guide was advanced through the steerable guide catheter sheath and used to carry out selective angiography of the right upper PV. The 6-F pigtail in the right PA was exchanged for an H1 catheter (Torcon NB, Cook Medical) which was navigated through the PA toward the nested coil in the large right upper lobe PAVM to enable selective angiography. Simultaneous arterial and venous injections were performed to localize the superior PV exiting the PAVM around the coils, with a STORQ wire (Cordis) used to navigate through the PAVM from the PV side. The steerable guide catheter was then advanced and positioned in the venous ostium of the PAVM. The wire and multipurpose guide were then removed. A 12-mm Amplatzer Vascular Plug II plug (Abbott) was deployed on the venous aspect of the existing coils with mild residual flow through the device. Accordingly, we elected to place an additional Micro Vascular Plug 9-mm device (Medtronic) on the PV aspect of the previous plug. Sequential arterial and venous contrast injections demonstrated successful occlusion of flow through the area (**Figures 1A to 1L,** Video 1).

DISCUSSION

HHT, or Osler-Weber-Rendu syndrome, is a multisystem vascular dysplasia associated with a 70% incidence of PAVMs.[1] Although these PAVMs are often asymptomatic, they can lead to spontaneous hemothorax, hemoptysis, and embolic events, necessitating intervention.[2] Symptomatic PAVMs are challenging to treat. Treatment of PAVMs includes both transcatheter intervention (typically coil embolization) and surgical resection.[2]

Surgical approaches are limited by postoperative recovery, recurrence, worsening pulmonary hypertension, and enlargement of previously undetected PAVMs during the postoperative period.[3] Embolization is the favored approach, and multiple strategies are available. The most commonly used are coils and plugs. Coil and plug therapy is performed by selective angiography of a PAVM with device deployment through the catheter within the PAVM to ensure that flow to the PAVM has subsided.[4] Limitations of this strategy include recanalization rates of approximately 20% and 5% for coils and plugs, respectively.[2]

We present a case of symptomatic PAVMs persisting despite multiple coil embolizations managed with transcatheter plug placement, from a retrograde approach from the PV aspect. This procedure was accomplished through a multidisciplinary approach, including interventional radiology, interventional cardiology, and expert imaging. This patient had undergone 31 previous coil embolizations in response to recurrent recanalization of his PAVM. Comparisons of PAVM management yielded that vascular plugs had lower PAVM persistence rates than isolated coil embolization.[5] Typically, these devices are deployed on the PA aspect of the PAVM given the ease of access. However, PV access is regularly performed in the setting of cardiac ablation and PV interventions.[6-8] Although it requires transseptal puncture,[9] PV access can be readily achieved, and it provides an alternative approach to either delivering occlusion devices on the PV aspect or facilitating PA delivery by creation of a rail.[10] The transseptal approach is an efficacious means of achieving successful closure of PAVMs.

ABBREVIATIONS AND ACRONYMS

CT = computed tomography

HHT = hereditary hemorrhagic telangiectasia

PA = pulmonary artery

PAVM = pulmonary arteriovenous malformation

PV = pulmonary vein

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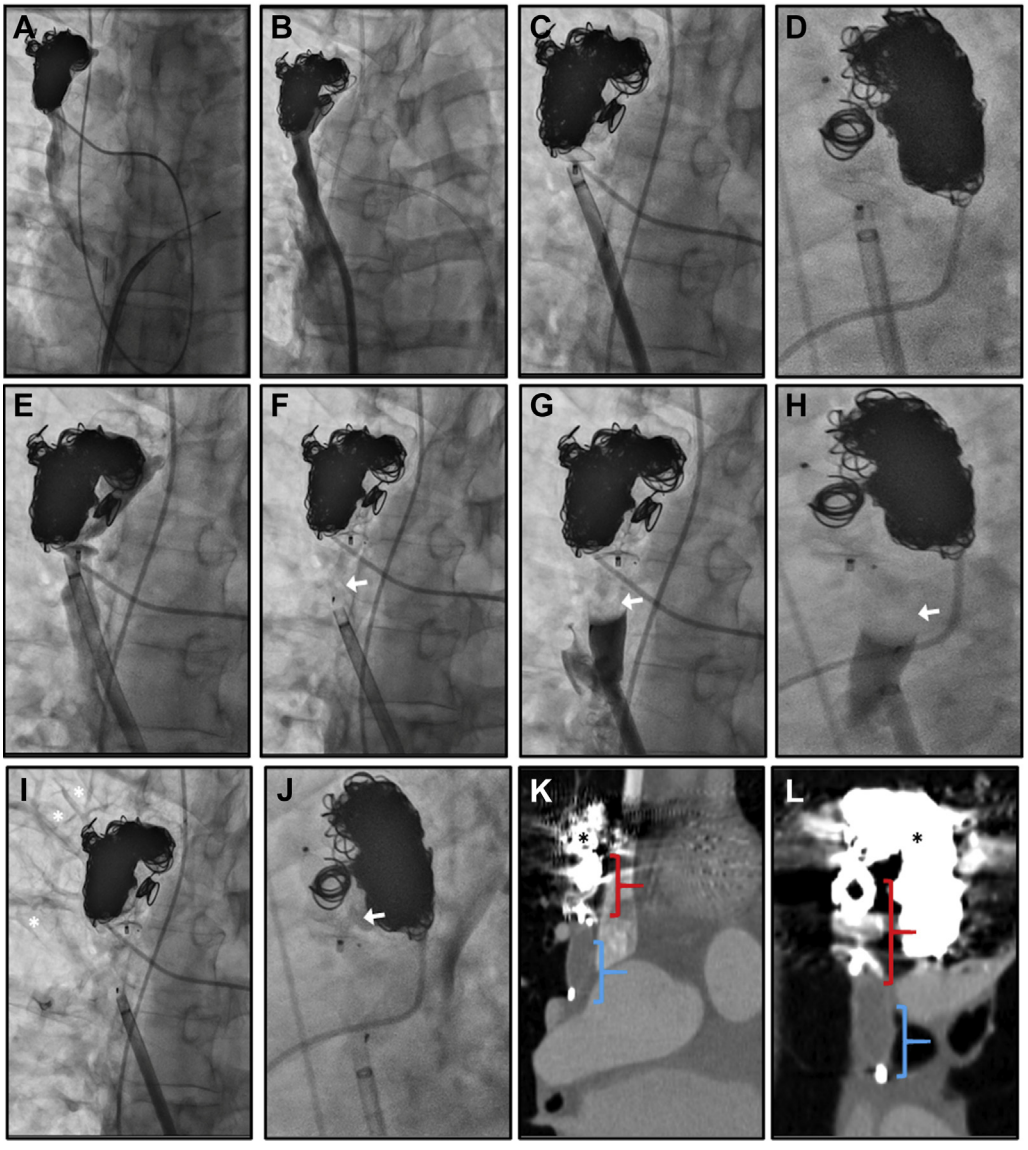
FIGURE 1 Transseptal Pulmonary Venous Plug Deployment



(A) Selective pulmonary artery angiography through an H1 catheter demonstrating persistent flow around the existing coils into the right upper pulmonary vein back into the left atrium. **(B)** Selective interrogation of the right upper pulmonary vein delineated in **(A)** with a steerable guide catheter through a transseptal approach positioned just distal to the existing coils with pulmonary vein contrast injection confirming correct branch selection. **(C)** Anteroposterior and **(D)** lateral projections of the deployed Amplatzer Vascular Plug II (AVP-II, Abbott) 12-mm device on the pulmonary vein aspect of the existing pulmonary arteriovenous malformation. **(E)** Persistent flow from the pulmonary artery to the pulmonary vein through the pulmonary arteriovenous malformation and the AVP-II device following pulmonary artery injection. **(F)** Deployment of an additional 9-mm microvascular plug (Micro Vascular Plug [MVP-9], Medtronic) **(arrow)** on the pulmonary vein aspect of the initial AVP-II 12-mm device. **(G)** Anteroposterior and **(H)** lateral pulmonary vein angiography demonstrating cessation of flow into the pulmonary arteriovenous malformation at the site of MVP-9 deployment **(arrow)**. **(I)** Anteroposterior and **(J)** lateral pulmonary artery angiography demonstrating flow into the surrounding pulmonary artery **(asterisks)** and within the AVP-II device **(arrow)** but without flow beyond the device into the pulmonary vein where the steerable guide catheter is situated, thus confirming successful occlusion. **(K)** Anteroposterior and **(L)** lateral projections on computed tomography angiography demonstrating successful occlusion with the AVP-II device **(red bracket)** and MVP-9 device **(blue bracket)** denoted on the background of previous embolization coils **(asterisk)** with significant artifact as a result.

However, this approach should be considered by a multidisciplinary team, including cardiologists skilled at transseptal puncture, to ensure that the complication rates (including pericardial effusion, iatrogenic cardiac injury, or embolic events) remain low, as noted in published reports.[6,9]

FOLLOW-UP

At 47 days postprocedurally, repeat CT chest angiography demonstrated stable device position (**Figures 1K and 1L**) and persistent successful occlusion. Moreover, repeat lung perfusion studies demonstrated persistent cessation of abnormal PAVM flow. At 11 months postprocedurally, our patient had no further evidence of paradoxical embolism, with a right-to-left shunt of 3.4% compared with 12.8% before plug deployment on a nuclear medicine cardiac shunt study. Moreover, a chest radiograph showed stable embolization coils and venous plugs.

CONCLUSIONS

Transcatheter treatments of PAVMs are limited by recanalization rates of 5% to 20%, requiring repeat interventions. Transseptal PV access offers an alternative approach to closure of PAVMs, thus facilitating either additional wire rail support or retrograde delivery of closure devices to aid PAVM occlusion. A retrograde PV approach provides an efficacious alternative for transcatheter PAVM closure.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS PAVM, plug, pulmonary arteriovenous malformation, transseptal

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