

# Pulmonary Arteriovenous Malformation in Combination With Stroke.

Jung Guen Cha

Kyungpook National University School of Medicine

Jihoon Hong (✉ [blushain@gmail.com](mailto:blushain@gmail.com))

Kyungpook National University School of Medicine <https://orcid.org/0000-0003-3389-244X>

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## Research Article

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# Abstract

## Background

As pulmonary arteriovenous malformation (PAVM) include a right-to-left shunt, it can be accompanied by fatal complications such as stroke and brain abscess due to paradoxical embolism. There are few PAVM patients accompanied by concurrent pulmonary embolism, so the treatment sequence has not been established.

## Case presentation

A 62-year-old female patient was transferred to our hospital with a simple PAVM and concurrent bilateral pulmonary embolism (PE). The acute thromboembolus was extended to the proximal segmental artery where the feeding artery originated. Anticoagulation was started, but on the fifth day of admission, the patient complained of left side weakness and brain magnetic resonance imaging revealed an acute infarction in the right lateral thalamus. This situation could lead to a dilemma between the risk of thrombus migration during catheter manipulation in PAVM embolization and another embolic event due to the late occlusion of the shunt during anticoagulation. This complex situation was successfully managed with delayed endovascular embolization for PAVM after four months of anticoagulation. The cause of PE in this patient was eventually diagnosed as antiphospholipid syndrome (APS).

## Conclusion

The authors reported a rare case of APS-induced PE in the PAVM feeding artery course that led to stroke during hospitalization. We successfully treated this patient with delayed endovascular embolization for PAVM after anticoagulation for PE and stroke. It is thought to be valuable in deciding for a treatment plan for this rare condition.

## Background

Pulmonary arteriovenous malformations (PAVMs) can cause paradoxical embolism such as stroke through a right-to-left shunt (Tellapuri et al., 2019). The occurrence of venous thrombosis (VT) or pulmonary embolism (PE) in patients with PAVM is a strong predisposing factor for a systemic embolism (Hewes et al., 1985). This likelihood will increase if the location of emboli is a part of PAVMs. Although there are no disagreements about anticoagulation as a first-line management for this situation, the timing of shunt occlusion with endovascular treatment has not been clearly established. On the other hand, antiphospholipid syndrome (APS) is an autoimmune disorder that causes recurrent VT and PE (Farmer-Boatwright et al., 2009), and if accompanied by a PAVM shunt, it can be a life-long threat of stroke.

## Case Presentation

A 62-year-old female patient was transferred to our hospital with massive PE discovered during hospitalization due to enteritis. She had a history of seven spontaneous abortions and complained of worsening dyspnea for 6 days. Her vital sign was stable, and 97% oxygen saturation was noted on 2 L/min nasal cannula. On chest computed tomography (CT), multifocal PE was observed in the bilateral lobar pulmonary arteries and its segmental branches (Fig. 1A, 1B). In addition, a simple PAVM with a 3-mm diameter feeding artery was identified in the right lower lobe superior segment. The acute thromboembolus was extended to the proximal segmental artery where the feeding artery originated (Fig. 1C). Using enoxaparin, the patient's dyspnea gradually improved, but on the fifth day of hospitalization, she complained of a sudden onset of left side weakness. Brain magnetic resonance imaging revealed an acute infarction in the right lateral thalamus and occlusion of the right posterior cerebral artery P2 segment. In such complicated situation, if PAVM embolization was immediately performed, the possibility of thrombus migration during catheter manipulation was expected to be high. After a multidisciplinary discussion, it was decided to embolize the PAVM after sufficiently resolving the PE with anticoagulants. After four months of anticoagulation with warfarin and rivaroxaban, a follow-up CT showed a complete resolution of PE (Fig. 1D). There were no new-onset neurologic complications during this period. An endovascular treatment was decided, and the patient was referred to the angio suite. After a right common femoral venous access, a 6-Fr guiding catheter (Flexor Shuttle Guiding Sheath; Cook Medical, Bloomington, Indiana) and 5-Fr catheters (Torcon NB Advantage, MPA and Headhunter type; Cook Medical, Bloomington, Indiana) were used to select the right main pulmonary artery. On angiography, the PAVM with a feeding artery arising from the segmental artery of the right lower lobe was observed, and no residual PE was noted (Fig. 2A). Then, a microcatheter (Masters Parkway Soft; Asahi Intecc, Tokyo, Japan) was advanced into the venous sac (Fig. 2B), and 10 mm- to 4 mm-sized 11 detachable coils (Concerto; Medtronic, Minneapolis, Minnesota) were used to embolize the venous sac and feeding artery. Completion angiography showed an occlusion of the shunt flow (Fig. 2C). The 5-month follow-up CT showed a significant reduction in the draining vein and feeding artery diameter, and no recurrence of PE (Fig. 3). With the suspicion of APS because of the patient's history, antibody testing was performed during hospitalization and outpatient follow-up, and she was finally diagnosed with APS, with a strong positivity for anti-β2 glycoprotein 1 IgM.

## Discussion

The acute PE in PAVM is a predisposing factor for life-threatening complications, such as stroke, and there have been few reports of cases in which they were found at the same time and how they were treated (Graves et al., 2009; Serra et al., 2015). Although hereditary hemorrhagic telangiectasia itself, a disease related to PAVM, promotes a hypercoagulable state (Shovlin et al., 2007), patients with both PAVM and PE are very rare. This is a devastating situation, as PE itself can cause hemodynamic instability, and it is a risk for the paradoxical embolism of thromboemboli.

According to Graves et al (Graves et al., 2009), the acute on chronic PE in a patient with multiple-infarct dementia due to a large PAVM was successfully treated with concurrent anticoagulation and endovascular embolization. Considering the nature of PE, they judged that the risk of migration through

minimal catheter manipulation was not high, and the benefit from blocking the shunt was greater. In a report of Serra et al. (Serra et al., 2015), asymptomatic PE was found in the feeding artery course of residual PAVM during a follow-up consultation after multiple PAVM embolization. The remaining PAVM was treated with delayed embolization after one week of anticoagulation. It seems that a safer method was chosen because the burden and clinical significance of PE in this patient were not large.

However, in the current case, there was a large volume of acute PE that seemed unstable on CT based on studies on factors affecting the stability of thrombi (Jeong et al., 2019). This induced thalamic infarction due to paradoxical embolism during hospitalization. In addition, the PAVM feeding artery was more than 3 mm, suggesting the possibility of another embolic event (Shovlin et al., 1999). In a situation where anticoagulation could not be stopped due to stroke, the bleeding risks, such as PAVM rupture, associated with the procedure could not be ignored. Therefore, it was determined that inducing thrombus fragmentation and migration during catheter manipulation would be riskier than slowly dissolving the thrombus through anticoagulation.

APS is an autoimmune disorder that promotes a hypercoagulable state and causes vascular thrombosis like VT, and its most common pulmonary manifestation is PE (Farmer-Boatwright et al., 2009; Sarinc Ulasli et al., 2021). Although it has been reported that an APS-induced PE rarely appears in a massive and life-threatening form (Sarinc Ulasli et al., 2021), if it is accompanied by PAVM, as in this case, paradoxical embolism can occur at any time during the patient's life, so an early diagnosis and appropriate treatment seems to be more important.

## Conclusion

The authors reported a rare case of APS-induced PE in the PAVM feeding artery course that led to stroke during hospitalization. The management of this complex situation has not been established, but we successfully treated the patient with delayed endovascular embolization for PAVM after anticoagulation for PE and stroke. It is thought to be valuable in deciding for a treatment plan for this rare condition.

## Abbreviations

PAVM: Pulmonary arteriovenous malformation

VT: Venous thrombosis

PE: Pulmonary embolism

APS: Antiphospholipid syndrome

CT: Computed tomography

## Declarations

## Ethics approval and consent to participate

For this type of study formal consent is not required. This study was approved by the Institutional Review Board

of Kyungpook National University Hospital.

## Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

## Availability of data and material

Not applicable.

## Competing interests

The authors declare that they have no competing interests.

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## Authors' contributions

JGC performed the chart review and was major contributors in writing the manuscript. JH and JGC were main performers of the procedure. JH and JGC were major contributors in revision of the manuscript. JH was major contributor in mentoring the creation of this manuscript. All authors read and approved the final version of the manuscript.

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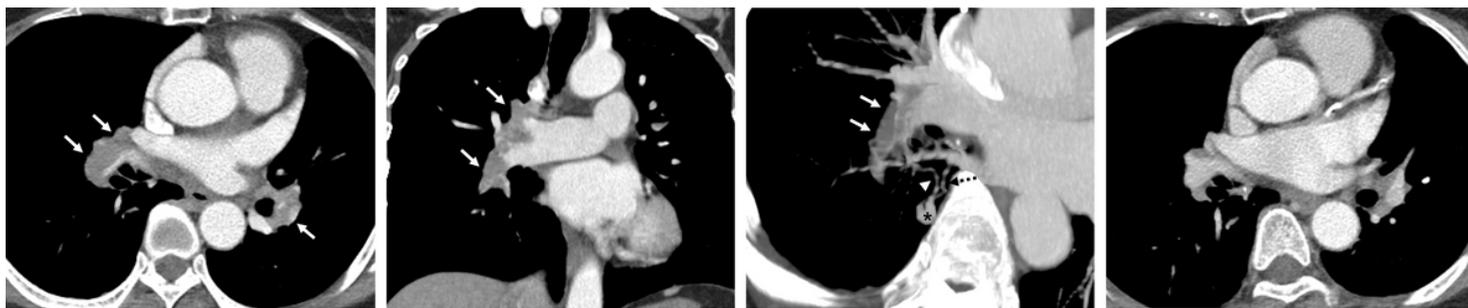
Not applicable

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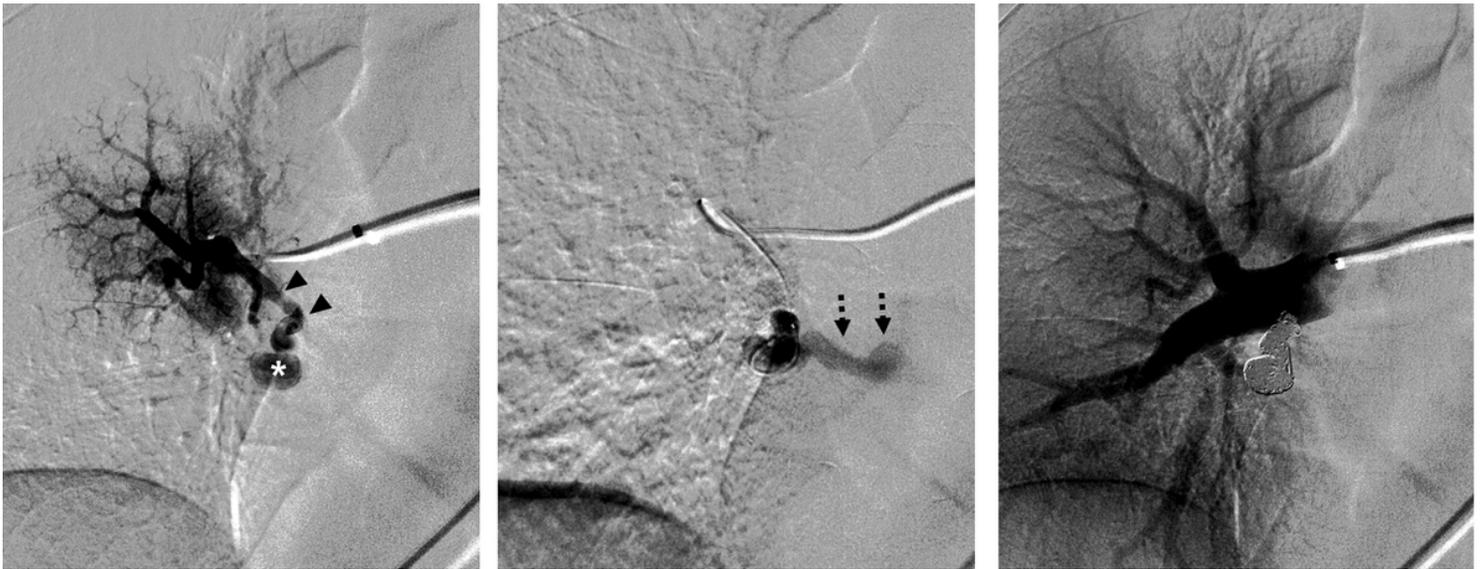
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## Figures



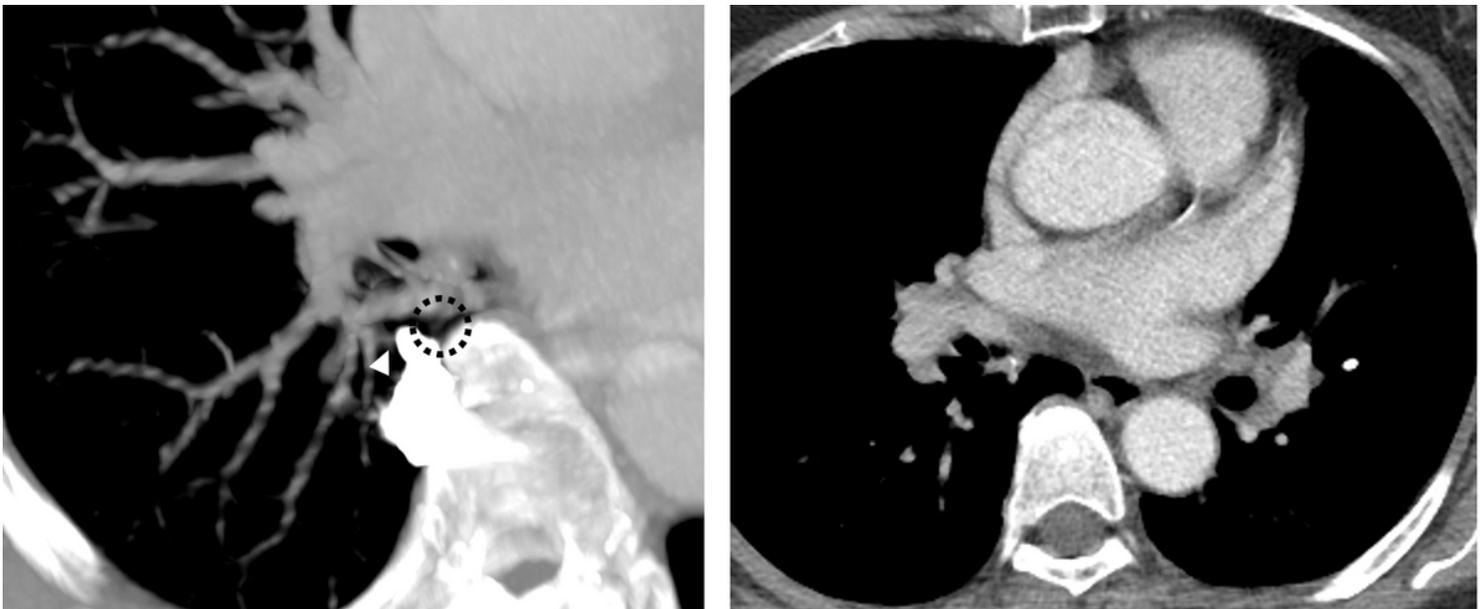
**Figure 1**

Initially performed axial (A) and coronal (B) computed tomography (CT) images show acute multifocal pulmonary embolism involving the bilateral lobar and segmental branches of the pulmonary artery (arrows). C Multiplanar reconstructed CT with maximum intensity projection shows the angioarchitecture of the PAVM consisting of the feeding artery (arrowhead), venous sac (asterisk), and draining vein (dashed arrow). Thromboembolisms that spread to the segmental artery from which the feeder originate are also observed (arrows). D After four months of anticoagulation, all pulmonary embolisms were resolved based on a follow-up CT.



**Figure 2**

A Selective right lower lobe superior segmental pulmonary arteriography shows an acute-angled and tortuous feeding artery (arrowheads) and venous sac (asterisk) without an evidence of residual thromboembolism. B The angiography performed on the venous sac of PAVM using a microcatheter shows engorged draining vein (dashed arrows) that directly drained to the left atrium. C After the venous sac embolization using multiple coils, completion angiography shows no residual shunt flow of the PAVM.



**Figure 3**

A After five months of PAVM embolization, the venous sac packed with coil nest and a significant reduction in the size of the feeding artery (arrowhead) and draining vein (dashed circle) are observed on reconstructed CT with MIP. B No recurrence of pulmonary embolism was noted.