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Multiple Pulmonary Arteriovenous Malformations in a Young Patient Presenting with Stroke

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Abstract

Pulmonary arteriovenous malformations are abnormal vascular communications between the pulmonary arteries and veins, resulting in an extracardiac right-to-left shunt. Pulmonary arteriovenous malformations are most commonly hereditary and associated with hereditary hemorrhagic telangiectasia. In the current literature, pulmonary arteriovenous malformations have been found to cause hypoxia, ischemic stroke, brain abscess, and hemothorax. Due to these potential complications, transcathether embolization is recommended as a treatment of pulmonary arteriovenous malformations. Here we describe a case of a 25-year-old male with multiple bilateral pulmonary arteriovenous malformations presenting with stroke due to right-to-left shunting.

Keywords: Angiography, arteriovenous malformation, CT scan, embolization, hereditary hemorrhagic telangiectasia

Introduction

Pulmonary arteriovenous malformations (PAVMs) are abnormal vascular connections between pulmonary arteries and veins, resulting in an intrapulmonary right-to-left shunt.[1] Pulmonary arteriovenous malformations have been reported to cause dyspnea, polycythemia, stroke, migraine, hemoptysis, brain abscess, or seizures.[2-4] A recent study has estimated the prevalence of PAVMs to be higher than previously suggested at approximately 38 per 100 000 with a female predominance.[5-6] Pulmonary arteriovenous malformations are most commonly associated with hereditary hemorrhagic telangiectasia (HHT), an autosomal dominant condition characterized by vascular abnormalities in the skin, mucous membranes, and visceral organs.[1,7] Approximately 80%-95% of patients with PAVM are diagnosed with HHT.[6] Due to serious complications that can occur with PAVMs, therapy through transcatheter embolization, the gold standard for treatment, is recommended.[2,8-9] In the following case, a patient presenting with stroke was found to have multiple bilateral PAVMs which were successfully treated with transcatheter embolization.

Case Presentation

A 25-year-old male with no significant past medical history presented to an emergency department (ED) due to 1-day duration of acute left upper extremity weakness and trouble gripping objects. He denied any other symptoms. A physical exam showed clubbing of the fingers, and laboratory values were significant for polycythemia with a hematocrit of 58.7 and hemoglobin of 20.5. Though the patient denied dyspnea, his oxygen saturation levels were in the 80's when standing and he was placed on 2 L of oxygen. The patient's family history revealed HHT and PAVM in his mother and maternal grandparents. He reported frequent childhood epistaxis which decreased in frequency in recent years, now occurring approximately once a week.

Due to the acute onset of unilateral weakness, computed tomography (CT) brain without contrast was obtained. Imaging revealed a subacute hypodense area in the right corona radiata extending to the centrum semiovale. Magnetic resonance imaging of the brain was significant for bilateral chronic cerebellar ischemia, right basal ganglia, and right motor strip infarcts. A chest radiograph showed a large 5.2 cm round opacity in the left lower lobe, prompting a CT scan of the chest which revealed numerous bilateral PAVMs, with the largest measuring 3.6 × 4.7 cm in the left lower lobe (Figure 1).

Interventional radiology was consulted for a pulmonary angiogram with embolization of the dominant left lower lobe PAVM (Figures 2 and 3). Given the multiple PAVMs, outpatient staged

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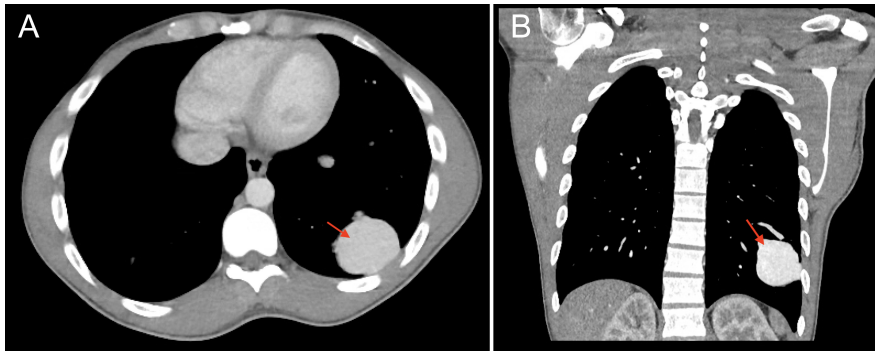

Figure 1. Pre-procedural contrast-enhanced computed tomography images of the chest showing the largest pulmonary arteriovenous malformation of 3.6 × 4.7 cm (arrow) in the left lower lobe in (A) axial and (B) coronal views.

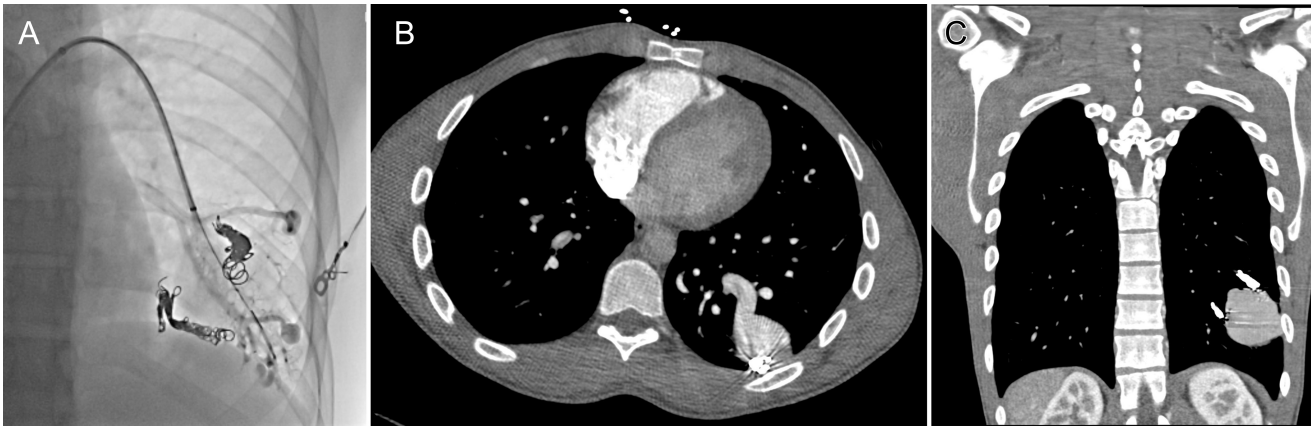

Figure 2. (A-C) Post-procedural images. (A) Pulmonary angiography showing embolization of 2 feeding arteries of the largest left lower lobe pulmonary arteriovenous malformation (PAVM) using a combination of Ruby® and Penumbra Occlusion Device® (POD) packing coil. (B, C) Contrast-enhanced computed tomography angiography of the chest revealing embolization of the left lower lobe PAVM in the (B) axial and (C) coronal views.

procedures were planned for further management. However, 2 days after embolization, the patient required increased oxygen support, and repeat CT chest showed persistently patent left lower lobe PAVM. Patient was scheduled for repeat embolization, which was treated for 8 PAVMs on the left and 1 PAVM on the right (Figure 4). Oxygen saturation improved immediately afterward, and no complications were observed. Patient was seen in the outpatient follow-up at 2 and 5 months with no change in remaining PAVMs or recanalization.

Main Points

- Pulmonary arteriovenous malformations (PAVMs) are abnormal vascular connections between pulmonary arteries and veins that result in an extracardiac right-to-left shunt, most often seen in hereditary hemorrhagic telangiectasia, an autosomal dominant condition.
- Pulmonary arteriovenous malformations are often found incidentally and can be asymptomatic, but serious complications such as ischemic strokes can occur in young patients if not treated.
- The gold standard for diagnosis of a PAVM is a computed tomography angiography; however, initial screening with contrast transthoracic echocardiography and anteroposterior chest radiograph has a sensitivity of 100%.
- Transcathether embolization is the recommended treatment for PAVMs and should be performed in cases in whom embolization is feasible regardless of the size or presentation of the PAVM due to the serious complication that can occur if not treated.

Discussion

A majority of PAVMs are congenital and are most commonly associated with HHT, an autosomal dominant condition resulting in multisystem arteriovenous abnormalities affecting 1 in 5000-8000 people worldwide.[1,6,7,10-12] Hereditary hemorrhagic telangiectasia can be diagnosed clinically by applying the Curacao criteria which state that HHT can be diagnosed if 3 of the following 4 criteria are present: (1) spontaneous and recurrent epistaxis, (2) multiple telangiectasias on skin and mucous membranes, (3) visceral arteriovenous malformations, and (4) first-degree relative with HHT.[7,12] Our patient was clinically diagnosed with HHT due to his history of recurrent epistaxis occurring since childhood, an extensive family history of confirmed HHT, and the presence of multiple bilateral PAVMs. He was encouraged to receive genetic testing and was referred to an HHT clinic for further management.

While many PAVMs are asymptomatic and discovered incidentally, serious consequences can occur if they are not recognized and treated.[2,4] The most commonly seen complications are neurologic, such as stroke and brain abscesses, in which

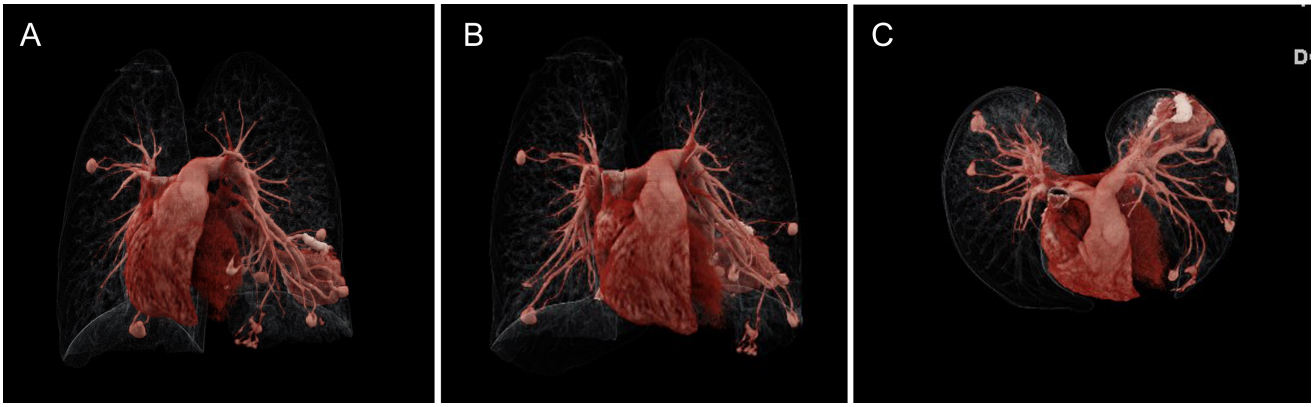

Figure 3. Post-procedural volume rendering computed tomography pulmonary angiography revealing the (A) oblique, (B) anterior, and (C) superior view of multiple pulmonary arteriovenous malformations with the largest located in the left lower lobe.

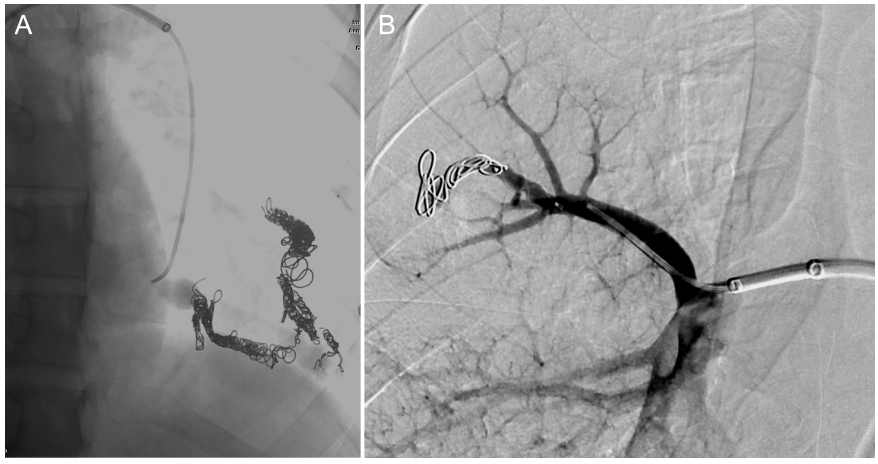

Figure 4. Pulmonary angiography showing successful embolization of multiple pulmonary arteriovenous malformations (PAVMs): (A) successful complete re-embolization of previously treated left lower lobe PAVM and (B) successful treatment of right upper lobe PAVM through embolization of the pulmonary arterial branch.

30% of patients are affected.[1] Recent literature has documented PAVMs as a rare cause of ischemic stroke due to paradoxical emboli resulting from a right-to-left shunt.[13,14] Therefore, a systematic approach is imperative when searching for a cause of stroke in young patients, and PAVMs should always be considered.

The gold standard for confirmation of PAVMs is computed tomographic angiography (CTA).[4] However, contrast transthoracic echocardiography (TTE) is recommended for initial screening to avoid unnecessary radiation and is known as the most sensitive test.[3,10,11] When combined with an anteroposterior chest radiograph, TTE had a sensitivity and negative predictive value of 100%.[11] In this case, our patient received a chest radiograph and CT chest before the TTE while in the ED. However, a TTE was obtained later to rule out intracardiac causes of stroke. This revealed early right-to-left shunting with opacification of the left-sided chamber at 3 cycles, suggesting a pulmonary cause and supporting the known diagnosis of PAVM in this patient.

Historically, embolization of PAVMs was recommended only for feeding vessels with a diameter greater than 3 mm, due to assumed increased risk of cerebral stroke or abscess.[9] However, recent works have suggested otherwise, reporting that PAVMs with smaller feeding arteries can cause paradoxical emboli, stroke, and absecess.[12,15] Therefore, it is now recommended to treat PAVMs in both symptomatic and asymptomatic patients, regardless of the size.[8,12] Transcathether embolization is the gold standard of treatment for PAVMs and is seen to significantly decrease the risk of neurological complications after treatment (2.6%-25% vs. 0%-2%).[8,9,12] There are various embolic materials available for use; however, coils and vascular plugs are utilized most frequently.[8] In this study, we initially embolized the largest PAVM with coils. However, due to consistent desaturation, the patient underwent re-embolization of the largest PAVM in addition to 7 smaller ones. Treatment was successful, and the patient saw immediate improvement of SpO_2 (90%-99% on 6 L of oxygen). At the time of follow-up, CTA showed no recanalization of previously embolized PAVMs, and the patient remained stable on room air with no further complications.

Conclusion

This case supports the current literature that PAVMs should be considered a cause of paradoxical emboli in young patients presenting with stroke. Our case is interesting due to the diffuse amount of PAVMs the patient presented with bilaterally. We agree with the recommendation that PAVMs should be

treated through transcatheter embolization, regardless of size and presentation, due to increased risk of cerebral and pulmonary adverse events.

Informed Consent: Informed consent was not required for this case report due to non-identifiable patient information.

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Declaration of Interests: The authors have no conflict of interest to declare.

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