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CASE REPORT

Sporadic Pulmonary Arteriovenous Malformation with a History of Stroke/Cerebrovascular Ischaemia Successfully Treated with Coil Embolisation: Two Case Reports

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CASE PRESENTATIONS

Case 1

A 64-year-old female, who had been a chronic smoker for 47 years smoking one to five cigarettes per day, had a history of ischaemic stroke at the age of 39. In 2019, she was referred to the medical outpatient clinic of our institution due to a 2-month history of cough, and a chest X-ray revealed an incidental finding of a 2-cm opacity at the right lower zone (Figure 1). Subsequent contrast computed tomography (CT) of the thorax revealed a 2-cm avid arterial-enhancing lesion at the right lower lobe near the costophrenic angle, corresponding to a previous chest radiograph–detected lesion. With the presence of a single hypertrophic tortuous pulmonary artery supply and single hypertrophic early draining pulmonary vein (Figure 2), a diagnosis of pulmonary arteriovenous malformation (PAVM) was made.

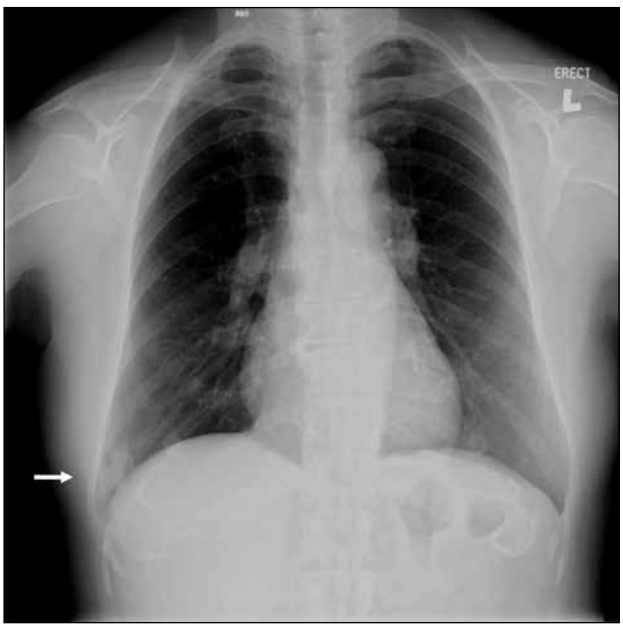

Figure 1. Case 1. Chest X-ray showing a 2-cm circumscribed non-calcified mass (arrow) at the right lower lobe near the costophrenic angle.

The patient was treated with coil embolisation to the PAVM in the right lower lobe, performed via a right

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Ethics Approval: This study was conducted in accordance with the tenets of the Declaration of Helsinki. Informed consent was obtained from the patients for the purpose of the study.

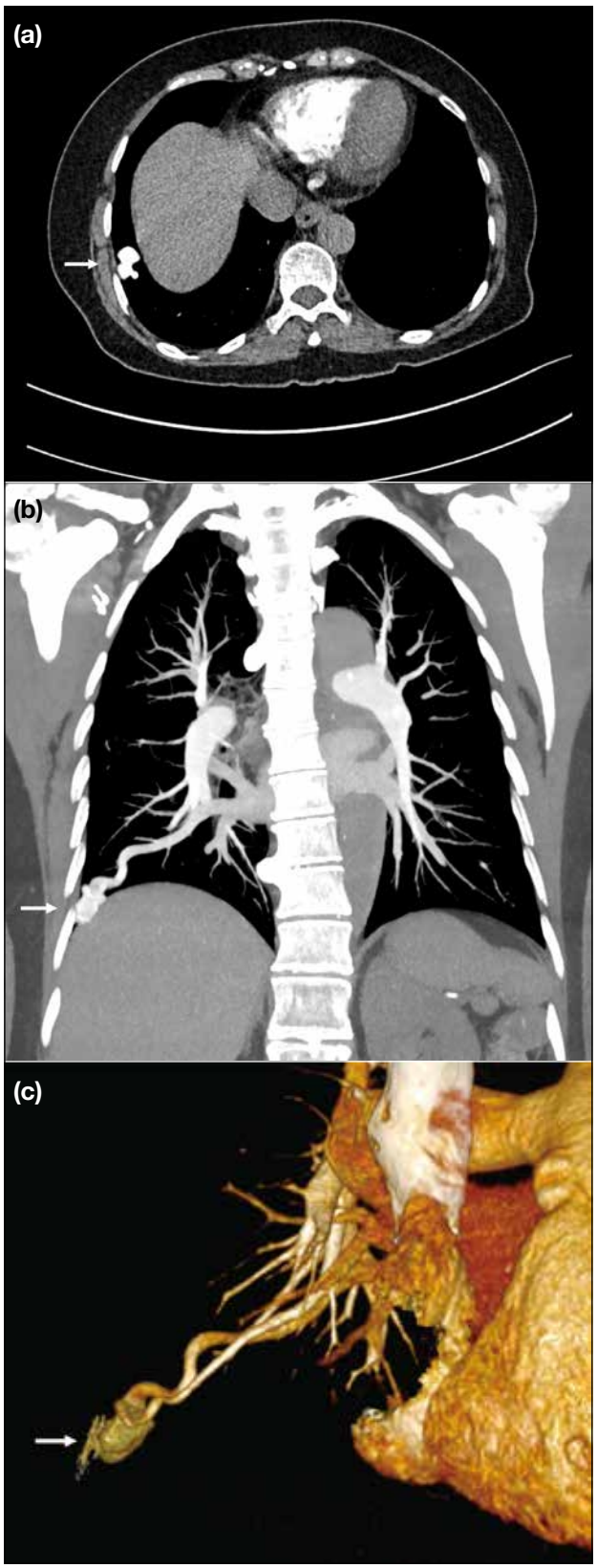

Figure 2. Case 1. (a) Axial, (b) coronal, and (c) three-dimensional reconstruction of contrast computed tomography of the thorax showing simple pulmonary arteriovenous malformation (arrows) in the right lower lobe with single hypertrophic tortuous right lower lobe segmental artery supply and single hypertrophic draining vein.

antegrade common femoral vein approach. The right lower lobe pulmonary artery was cannulated with a 5-Fr multipurpose angiographic (MPA) catheter (Cordis Corporation, Miami Lakes [FL], US). Digital subtraction angiography (DSA) showed a dumbbell-shaped simple PAVM with single hypertrophic right lower lobe segmental pulmonary artery (6 mm in diameter) and single hypertrophic draining right lower lobe pulmonary vein (Figure 3a). The PAVM was selectively cannulated with a 3-Fr Rebar Reinforced Microcatheter (Micro Therapeutics Inc, Irvine [CA], US) through the 5-Fr MPA catheter, and coil embolisation was performed with 28 detachable coils (EV3 Concerto detachable coils; Micro Therapeutics Inc, Irvine [CA], US). Post-embolisation DSA revealed complete occlusion of the PAVM (Figure 3b). No complication was encountered and the patient was discharged home on the day of the procedure. Nonetheless she defaulted her follow-up 2 months after embolisation.

Case 2

A 73-year-old female with controlled hypertension was referred to the medical outpatient clinic of our institution in 2019 for non-exertional chest pain. She had a history of transient ischaemic attack at the age of 64. Chest radiograph was unremarkable (Figure 4). CT coronary angiogram incidentally revealed a cluster of tortuous serpiginous vasculature at the anteromedial retrosternal region of the right upper lobe, suspicious of PAVM. Contrast CT of the thorax subsequently confirmed this diagnosis (Figure 5).

The patient underwent coil embolisation to the PAVM in the right upper lobe via a right antegrade common femoral vein approach. The right upper lobe pulmonary artery was cannulated with a 5-Fr MPA catheter. DSA showed a 2.5 cm × 1.8 cm simple PAVM at the anteromedial retrosternal region of the right upper lobe (Figure 6a and b). The PAVM was selectively cannulated with a 3-Fr microcatheter through the 5-Fr MPA catheter, and coil embolisation was performed with 21 detachable coils (EV3 Concerto detachable coils).

Post-embolisation DSA showed complete occlusion of the PAVM (Figure 6c). No complication was encountered and the patient was discharged home the next day. At 2-month follow-up, she remained asymptomatic and had no exertional shortness of breath, ankle oedema or haemoptysis. Follow-up radiological imaging was still pending.

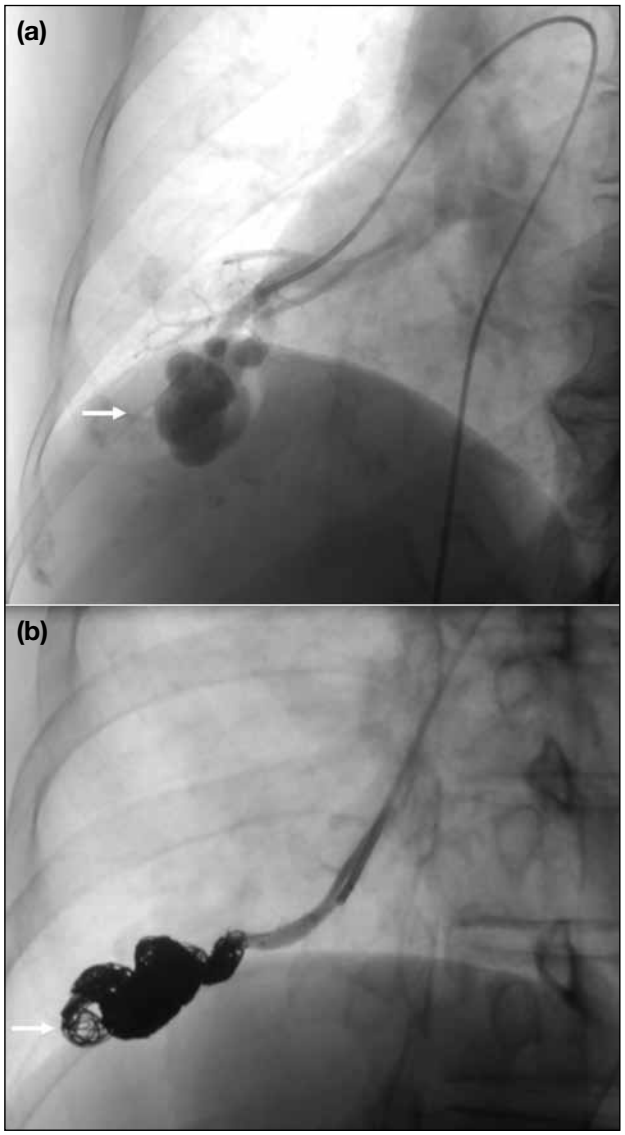

Figure 3. Case 1. (a) Digital subtraction angiography (DSA) of the right lower lobe segmental pulmonary artery showing the dumbbell-shaped simple pulmonary arteriovenous malformation (PAVM) [arrow] at the right lower lobe near the costophrenic angle, with single hypertrophic right lower lobe segmental pulmonary artery (6 mm in diameter) and single hypertrophic drainage pulmonary vein. (b) Post-embolisation DSA showing the PAVM (arrow) was completely occluded by detachable coils with no draining vein being opacified.

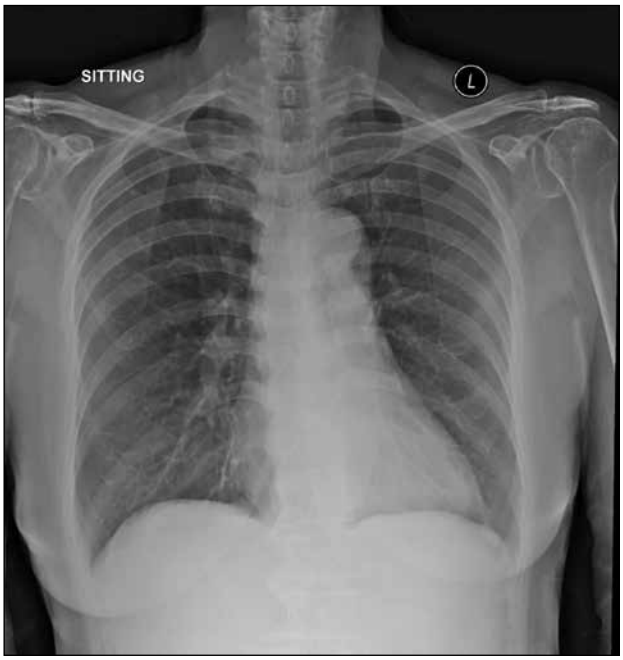

Figure 4. Case 2. Chest radiograph was unremarkable. No focal lung mass lesion was seen.

DISCUSSION

PAVM is a rare disease. It is a high-flow low-resistance fistulous communication between the pulmonary artery and pulmonary vein without interposition of a capillary bed. Limited prevalence data suggest that PAVM may affect up to 1 in 2600 individuals.[1,2] The majority (80%-90%) of PAVMs are congenital with concomitant hereditary haemorrhagic telangiectasia (HHT), while the rest are sporadic. Sporadic PAVM is rarely reported in the literature. According to Albitar et al,[3] sporadic PAVMs occur more commonly in females, are most often simple and single, have lower lobe predominance, and are associated with a high rate of neurological complications. For our two patients, they had no recurrent spontaneous epistaxis, mucosal telangiectasia or family history of HHT, but with only visceral AVM. Therefore, they had fewer than two of four Curaçao diagnostic criteria, making HHT unlikely. They were thus considered sporadic PAVM.

A PAVM can be asymptomatic or present with right-to-left shunt symptoms, including dyspnoea, cyanosis, haemoptysis and polycythaemia. The classic triad of dyspnoea on exertion, cyanosis, and clubbing should alert the clinician to the possibility of a PAVM. Untreated PAVMs can cause complications such as stroke, brain abscess, rupture causing haemoptysis or haemothorax. Both our patients had a history of stroke/cerebrovascular ischaemia.

The preferred treatment for most patients with PAVM is transcatheter embolisation. This has largely replaced surgery because of its minimal invasiveness and short recovery time. Surgical resection is reserved for PAVMs not amenable to embolisation.

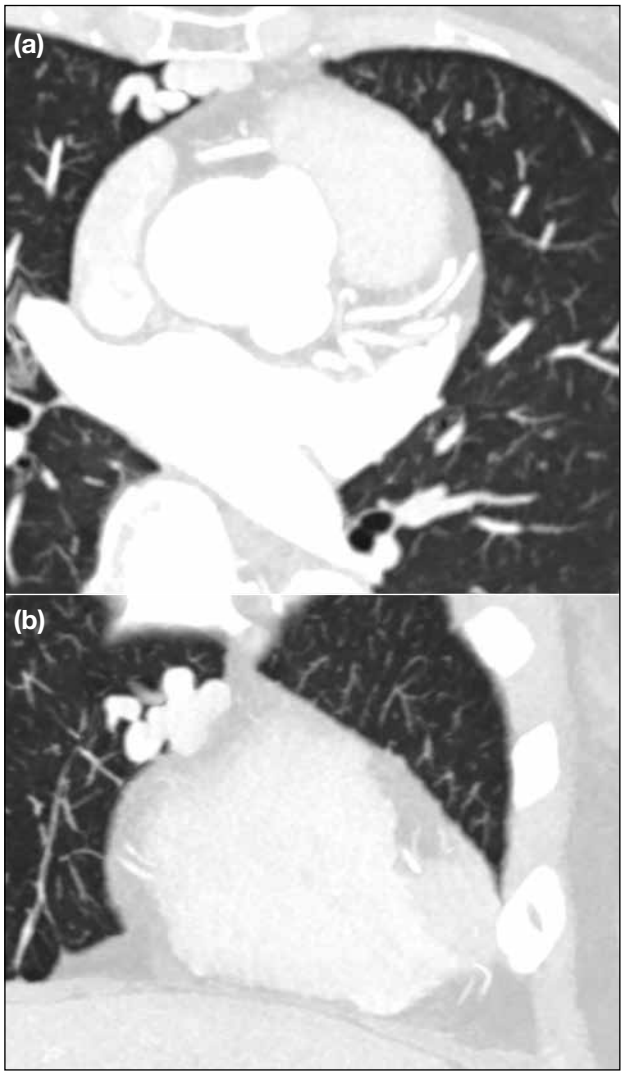

Figure 5. Case 2. (a) Axial and (b) coronal contrast computed tomography of the thorax showing a cluster of tortuous serpiginous vasculature at the anteromedial retrosternal region of the right upper lobe, with early arterial enhancement and supply from a single right upper lobe segmental pulmonary artery, draining to a single right upper lobe segmental pulmonary vein entering the left atrium.

Treatment of PAVM can help prevent cerebral complications such as transient ischaemic attack, stroke or brain abscess; it can also help prevent pulmonary complications such as lung haemorrhage, or decline in exercise tolerance. A general rule may be to treat PAVMs with feeding arteries >3 mm although more dedicated centres will offer intervention when the feeding artery exceeds 2 mm.[4] Nevertheless intervention is obviously indicated when patients are symptomatic or when the PAVM is enlarging.

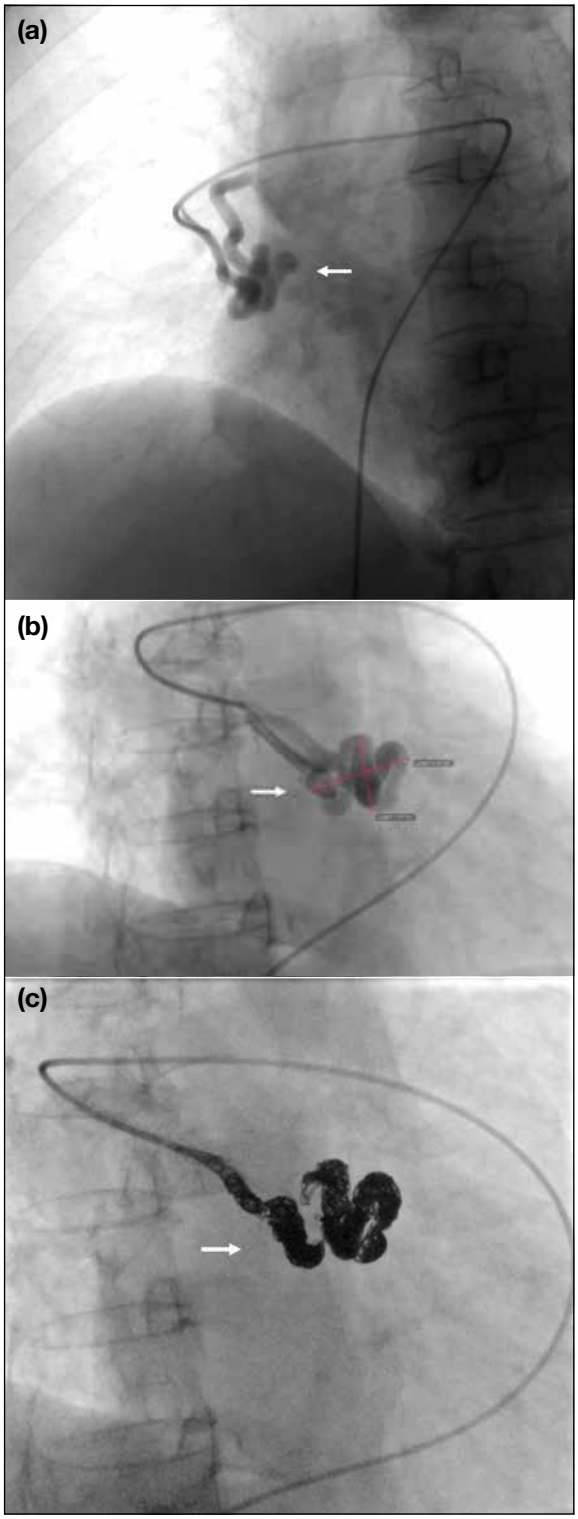

Figure 6. Case 2. (a) Anterior-posterior view and (b) pulmonary arteriovenous view of digital subtraction angiography (DSA) of the right upper lobe segmental pulmonary artery showed the tubular tortuous simple pulmonary arteriovenous malformation (PAVM) [arrows] at the anteromedial retrosternal region of the right upper lobe with single hypertrophic segmental artery (3 mm in diameter) and single hypertrophic drainage vein. The PAVM measures 2.5 cm × 1.8 cm. (c) Post-embolisation DSA showing the PAVM (arrow) was completely occluded by detachable coils with no draining vein opacified.

A Grollman catheter is a right-angled pigtail catheter with the curve on the reverse side of the angle. This shape is most ideal to navigate through the heart or to perform pulmonary angiogram. A reverse curve catheter such as the Omni Flush catheter can also be used to navigate through the right heart, but subsequent exchange to an appropriate catheter for intervention is needed. A steerable guiding sheath is another option. As these catheters were not available at our centre, a 5-Fr MPA catheter was used and we were able to successfully navigate through the heart.

In DSA, we measure the diameter of arteriovenous malformation to determine coil or plug size. An undersized device should be avoided due to the risk of distal migration. We may oversize the coil or plug by 20% to 50% relative to the feeding vessel.

The choice of embolic agents includes detachable metallic coils, Amplatzer plug occluding device and microvascular plug (MVP). Detachable metallic coils enable more precise control. Coils should not be placed directly into a PAVM because of the risk of paradoxical coil embolisation. Instead, the tip of the coil should be placed in a small side branch proximal to the target. This allows it to prolapse into the feeding vessel. For an Amplatzer plug occluding device, an appropriately sized sheath is placed into the feeding vessel and the device is introduced via the sheath to the feeding vessel proximal to the target. After confirming the position with angiography, the device can be deployed by retracting the sheath. For MVP, the MVP-5 system is advanced into the feeding artery and the MVP plug is unsheathed to occlude the target vessel. The position is confirmed by selective angiogram with contrast injected via the rotator haemostatic valve, followed by deployment of the MVP using the detachment system.

Our patients were treated with detachable metallic coils. Heparin was given during the procedure to minimise the risk of periprocedural paradoxical emboli, estimated at 1%.[4]

Complications of embolisation of PAVM are classified as immediate, early or late. Immediate complications include vascular injury and cardiac arrhythmia, but these are rare. Air embolism is becoming rarer, probably due to advances in technique and equipment. Precaution is also taken to double-flush all catheters to avoid air bubbles and clots. To avoid the complication of catheter entrapment in a heart valve, extra care should be exercised when withdrawing the catheter along the guidewire passing via the heart valve post-embolisation. Pleurisy is considered the most common early complication, occurring in up to 13% to 31% of patients.[5] It is usually self-limiting. Other rarer early complications include pulmonary infarcts and migration of embolic material. Late complications include recanalisation and growth of new feeding arteries due to collateralisation.[6] For both patients, they had no post-embolisation complication detected.

After treatment of PAVMs, patients require long-term follow-up imaging, preferably with a baseline scan 3 to 6 months after treatment to check if there is successful sac involution. Follow-up imaging every 3 to 5 years is necessary to capture any late collateralisation or recanalisation.[4]

CONCLUSION

Sporadic PAVM is rare and unlike congenital PAVM, it is not associated with HHT. With embolisation being minimally invasive and enabling faster recovery, it has replaced surgery as the first-line treatment. We present two cases of sporadic PAVM in middle-aged women with a history of cerebrovascular accident. Both patients had a single pulmonary artery supply and single pulmonary vein drainage, and they were treated successfully with coil embolisation without immediate complications.

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