CASE REPORT

Stroke resulting from an isolated pulmonary arteriovenous malformation

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SUMMARY

Isolated pulmonary arteriovenous malformations (PAVM) are rare, but can be responsible for paradoxical embolic events. It is important to recognise these situations early and intervene quickly to attain the best outcome. We describe the case of a 45-year-old woman with a long-standing history of migraines with aura who presented to the emergency department with left-sided weakness and slurred speech. Work-up was unclear and she was diagnosed with cryptogenic stroke. Follow-up transoesophageal echocardiogram was suggestive of an atrial septal defect, though no clear defect was evident. Pulmonary angiography revealed an isolated PAVM arising from the right lower lung segmental artery with a 2 mm feeding artery leading into 3–4 mm of dysplastic vessel that connects to the vein. The vessel was embolised successfully using a combination of a 4 mm Amplatzer Vascular Plug in the distal feeding artery and a 4 mm Nester coil proximally.

BACKGROUND

The rare event of pulmonary arteriovenous malformation (PAVM) causing a stroke in young women highlights the increasing prevalence of PAVMs in the population and the need to consider them as differential diagnoses in young patients with cerebrovascular accidents (CVA).

CASE PRESENTATION

A 45-year-old woman with a history of long-standing migraines with aura presented to the emergency department (ED) with complaints of weakness on her left side and blurriness in her right eye associated with slurred speech. Her medical history was significant for classic migraines three to four times per month since her teenage years for which she was prescribed Fioricet. She smoked about 1 pack of cigarettes per month.

INVESTIGATIONS

Biochemical and haematological profiles were negative.

Brain MRI without contrast performed soon after was significant for a 12 mm infarct of the right thalamus area.

Transeoophageal echocardiogram (TEE) was suggestive of an atrioseptal defect (ASD).

Transcranial Doppler bubble study found severe shunting at rest and with Valsalva.

Intracardiac echocardiogram (ICE) was performed resulting in no ASD present, but clearly

delayed (five cardiac cycles) positive bubble study, suggesting a right-to-left shunt likely at the pulmonary level.

CT angiography was performed and significant for a dilated vascular structure at the lower lobe of the right lung suspected to represent an arteriovenous shunt.

Selective pulmonary angiography showed a small simple PAVM arising from the right lower lung segmental artery with a 2 mm feeding artery leading into approximately 3–4 mm of dysplastic vessel that connects to the vein.

DIFFERENTIAL DIAGNOSIS

Head CT scan demonstrated no acute changes; however, MRI performed soon after was significant for a 12 mm infarct of the right thalamus area. Transthoracic echocardiogram was performed during this admission and was unremarkable.

Interval head and neck MR angiography, carotid ultrasound and hypercoagulable profile were all negative. However, the patient continued to have symptoms of intermittent dizziness and visual deficits, so she was referred to cardiology for TEE, which was suggestive of an ASD in the setting of a positive bubble study although no clear defect was noted (figure 1). Subsequently, a transcranial Doppler bubble study found severe shunting with Valsalva (figure 2). A preprocedural chest X-ray demonstrated no acute intrathoracic process, and ICE was performed for further structural investigation. No ASD was present, but a clearly delayed (five cardiac cycles) positive bubble study was seen suggesting a right-to-left shunt likely at the pulmonary level. A CT angiography was performed and significant for a dilated vascular structure at the lower lobe of the right lung suspected to represent an arteriovenous shunt. She then underwent selective pulmonary angiography which showed a small simple PAVM arising from the right lower lung segmental artery with a 2 mm feeding artery leading into approximately 3–4 mm of dysplastic vessel that connects to the vein (figure 3).

TREATMENT

The patient was initially discharged from the ED on enteric-coated aspirin 81 mg daily, Lipitor 10 mg daily, and daily subcutaneous Lovenox injections with follow-up in about 1 month with a diagnosis of cryptogenic stroke.

For definitive treatment, the dysplastic vessel was embolised using a combination of a 4 mm Amplatzer

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Figure 1 Positive bubble study with evidence of bubble contrast in left atrium (yellow arrow) and right atrium (red arrow) simultaneously. Interatrial septum (blue arrow).

Vascular Plug version 4 in the distal feeding artery and a 4 mm Nester coil proximal to that, resulting in complete occlusion of the PAVM (figure 4). Closure of the arteriovenous malformation was confirmed with a transcranial Doppler bubble study which found no evidence of shunting with Valsalva (figure 5).

OUTCOME AND FOLLOW-UP

The patient has not had any lasting effects of her CVA and on follow-up has no further shunting, central nervous system events, as well as a documented reduction in her migraines.

DISCUSSION

PAVMs are uncommon communications between pulmonary arteries and veins, leading to the potential of a right-to-left shunt. PAVMs are either sporadic or congenital, and the most common aetiology is hereditary haemorrhagic telangiectasia (HHT). PAVMs are the presenting sign of HHT 15%–50% of the time.¹ Without a diagnosis of HHT, PAVMs were initially considered to be uncommon in the general population. However, current reports suggest PAVMs are increasingly diagnosed in the general population with most recent prevalence estimated to be about 1 in 2600 patients.² Clinical manifestations common to congenital or sporadic PAVMs may include dyspnoea, asymptomatic

Figure 3 Chest CT showing pulmonary arteriovenous malformation (black arrow) arising from right lower lung segmental artery with 2 mm feeding artery leading into approximately 3–4 mm of dysplastic vessel that connects to the vein.

hypoxaemia and paradoxical emboli leading to stroke, transient ischaemic attack, brain abscess, massive haemoptysis and spontaneous haemothorax.^{1–3} In this report, the patient was initially thought to have suffered a paradoxical embolus through an atrial septal defect. The aetiologies of stroke in patients under 50 years old are vast and include vasculitis, prothrombotic states, illicit drug use and arterial dissection.⁴ However, paradoxical emboli are associated with a narrower set of aetiologies and especially

Figure 2 Transcranial Doppler showing severe shunting by bubble contrast with Valsalva.

Figure 4 CT chest showing embolised arteriovenous malformation with 4 mm Amplatzer Vascular Plug version 4 in the distal feeding artery and a 4 mm Nester coil proximal to that, resulting in complete occlusion of the pulmonary arteriovenous malformation.

Figure 5 Transcranial Doppler postarteriovenous malformation closure with Valsalva with no evidence of shunting.

congenital cardiac abnormalities. Under this presumption, TEE was performed and suggestive of an ASD. However, at the time of ASD closure, there was no evidence of ASD, but a delayed positive bubble study was present. Imaging showed a solitary right lower lobe PAVM with a 2mm feeding artery diameter. The prevalence of an isolated PAVM as an aetiology of ischaemic stroke is reported to be 0.5%.⁵ As described by Shovlin,² the increased prevalence of PAVMs is associated with their incidental identification through the use of CT. However, most of the PAVMs will remain asymptomatic, and the occurrence of paradoxical emboli is still a rare occurrence. Another uncommon occurrence is the development of strokes in young adults. Young adults make up 10%–15% of all stroke victims.⁶ The young and old adult populations maintain the same modifiable risk factors for stroke; however, data are limited to the extent that these modifiable risk factors would have on precipitating a stroke in a patient with a PAVM. In this case, the only identifiable risk factor excluding the PAVM is the patient's smoking history of 1 pack per month. Some reports^{4,5} describe patients with previous neurological symptoms such as transient ischaemic attack and altered facial sensation. However, another case reported by Dubrava *et al*⁷ had features similar to our patient such as smoking history and chronic migraines. The differences and similarities in these cases highlight the variability and challenge of predicting which patients might be at risk for CVA through a paradoxical embolism. According to Moussouttas *et al*,⁸ brain infarction most likely occurs with PAVMs that have feeding arteries greater than 3mm in diameter. As previously reported by Todo *et al*,⁹ this is another unique case of a less than 3mm feeding artery resulting in a brain infarction. Although the incidence of an isolated PAVM and subsequent stroke is a rare clinical phenomenon, their diagnosis and treatment is understood.

PAVMs are not noticeable on physical exam or on chest X-ray. Thus, when a right-to-left shunt is suspected the gold standard for diagnosis is considered to be contrast enhanced TEE.⁷ If the TEE is positive for a right-to-left shunt without any evidence of other aetiologies of such a shunt, the diagnosis of PAVM can be confirmed via CT angiogram. Amplatzer vascular plugs or coils are used for embolisation of PAVMs, which is a safe and effective treatment to prevent recurrent strokes.^{2,10} The uniqueness of this case is observed in the patient's age of CVA, isolated PAVM, and the size of the feeding artery and lack of any prior symptomatology until her CVA in February 2016.

Learning points

Expanding the differential diagnosis of younger patients who present with cryptogenic stroke.
Transcranial Doppler can be used to augment diagnostic accuracy for right-to-left shunting.
Consider definitive therapy in a young patient who presents with cryptogenic stroke.

Contributors JS, HA and SSK all equally participated in researching, writing and editing of this case report.

Competing interests None declared.

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