CASE REPORT

Recurrent minor strokes/TIA with a right to left shunt

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SUMMARY

A patient came to our institution for evaluation and closure of suspected patent foramen ovale actively manifesting as cerebrovascular accident. Through further studies, we found the presence of a pulmonary arteriovenous fistula, which was subsequently and successfully closed through non-invasive percutaneous selective segmental pulmonary artery embolisation.

BACKGROUND

Pulmonary arteriovenous fistulas (PAVFs) are abnormal communications between pulmonary veins and arteries, allowing shunting of blood bypassing pulmonary circulation. They are either congenital or acquired and associated with vascular or neurological complications including cyanosis, clubbing, pulmonary haemorrhages, strokes, transient ischaemic attacks and seizures. Acquired PAVFs are usually associated with trauma, infection or malignancies whereas congenital PAVFs, which represent about 80% of total PAVFs, are usually associated with hereditary haemorrhagic telangiectasias (HHT).

Only a few cases of spontaneous congenital PAVFs have been reported where the main presentation is hemiplegia related to paradoxical brain emboli.

Here we present a 47-year-old woman with paradoxical brain emboli to the left middle cerebral artery (MCA), who came to our institution for closure of a suspected patent foramen ovale (PFO) and who was found to have a PAVF through subsequent studies. This is significant because it highlights the importance of formulating a broad differential diagnosis when a right to left shunt is present or suspected. It highlights the most efficient way to detect a PAVF through the use of CT pulmonary angiography. Most importantly, it highlights the importance of closing the shunt to prevent further neurological damage and eliminate the risk of further embolisation. This paper will be of interest to readers in the areas of interventional cardiology and general cardiology.

CASE PRESENTATION

A 47-year-old woman with no history of cardiovascular disease, presented to our institution for closure of a PFO diagnosed at an outside facility. Approximately 8 months prior, the patient presented to her local hospital with right hemiplegia and dysarthria. She underwent transoesophageal echocardiogram (TEE) with bubble contrast, which was positive for a right to left shunt and a PFO was suspected. The patient was treated with aspirin and plavix.

However, she continued to experience frequent recurrent episodes of headaches with transient neurological deficit (TND); thus, a percutaneous PFO closure was planned. Due to family relocation, she moved closer to our institution, requesting further intervention. She presented with mild signs of residual left MCA stroke, including right hemiparesis and a right facial droop. More importantly, the patient continued to have episodes of severe headache followed by TND symptoms, which occurred several times per month.

INVESTIGATIONS

CT brain and MRI brain confirmed an old left middle cerebral artery distribution infarct.

Hypercoagulable state workup (including protein S, protein C, factor V Leiden mutation, antithrombin III and antiphospholipid antibodies) was negative.

A percutaneous PFO closure was attempted, but a PFO was not identified. A PFO was not able to be crossed despite the use of several different catheters. A repeat TEE was then performed which revealed no PFO, secundum atrial septal defect, sinus venosus or anomalous pulmonary venous drainage. However, a delayed appearance of agitated saline bubbles was noted in left atrium after three to four cardiac cycles (figure 1).

Computed tomographic angiogram of pulmonary arteries and selective pulmonary angiogram were then performed which confirmed a PAVF (figures 2 and 3).

DIFFERENTIAL DIAGNOSIS**Intracardiac right to left shunt**

- ▶ PFO.
- ▶ Persistent truncus arteriosus (minimal cyanosis).
- ▶ Transposition of great vessels.
- ▶ Tricuspid atresia.
- ▶ Total anomalous pulmonary venous return.
- ▶ Eisenmenger syndrome.
- ▶ Tetralogy of Fallot.

Non-cardiac (pulmonary) right to left shunt

- ▶ PAVF or malformation.

TREATMENT

Patient underwent successful percutaneous closure of an AV pulmonary fistula with an 8 mm Amplatzer vascular plug device 8 mm (figure 4).

OUTCOME AND FOLLOW-UP

After the AV closure, patient recovered well and was sent home in stable condition. Currently and 4 years

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Figure 1 Transoesophageal echocardiogram showing appearance of bubbles in the left atria after four cardiac cycles. AA, aorta artery; LA, left atrium; PAVF, pulmonary arteriovenous fistula; RA, right atrium.

after the PAVF closure, patient remains asymptomatic with no more migraine headaches and no further neurological deficits.

DISCUSSION

PAVFs are abnormal communications between pulmonary veins and arteries allowing shunting of blood bypassing pulmonary circulation.¹ PAVFs are rare and found twice as often in women compared with men.² The aetiology is not well known, but it is estimated that at least 80%–90% of cases are congenital,^{3,4} and of those, up to 80% are associated with HHT.^{5,6} Spontaneous (isolated) PAVFs are uncommon but when they are present, their mechanism for cerebrovascular accident and central nervous complications are related to paradoxical embolisms.⁵

Pathogenesis is not well understood but it has been postulated that it could be related to a defect in pulmonary artery loops, which cause dilatation and formation of thin walled capillary sacs.⁷ Others have postulated that fistulas are related to incomplete separation of arterial and venous septae during fetal development.⁸

Figure 2 CT angiogram of pulmonary arteries showing a pulmonary AVF. AVF arteriovenous fistula; LLL, left lower lobe.

Figure 3 Selective pulmonary angiography confirming a pulmonary AVF. AVF, arteriovenous fistula; LLL, left lower lobe.

The size of PAVFs vary reaching up to 10 cm in diameter but most are within 1–5 cms.⁹ More than 80% are classified as simple, which entails a single artery and a single vein. The rest are complex, which includes more than one pulmonary artery and/or more than one pulmonary vein.^{10–12}

Symptoms are directly associated with the size of the fistula,¹³ where a single fistula <2 cm in diameter is usually asymptomatic.^{13,14} The most common symptoms are epistaxis, dyspnoea, cyanosis and polycythemia. The most common complication involves the central nervous system, presenting as strokes, transient ischaemic attacks, migraine headaches, brain abscesses and seizures.^{13,15–18} It has been postulated that paradoxical emboli by formation of thrombus in the fistula is the main mechanism for these complications. Other postulated mechanisms for development of neurological complications are thromboembolism caused by polycythemia and lower extremity deep vein thrombosis.¹⁵ Once a PAVF is suspected, contrast echocardiography is a good initial study to evaluate intrapulmonary shunts, whereas TEE is the study of choice.^{19–21} If an intrapulmonary shunt is

Figure 4 Pulmonary angiogram showing successful percutaneous closure of pulmonary AVF. AVF, arteriovenous fistula; LLL, left lower lobe.

detected, angiography or CT angiography are performed for definitive diagnosis.⁹

Indications for treatment of PAVFs are based on whether PAVFs are progressively enlarging, or if there are symptoms or complications as consequences of PAVFs.^{13 16} More recent indications are based on PAVF characteristics and size.^{12 22–24} In general, if PAVFs are symptomatic and >2 cm in diameter, they likely require intervention. If the feeder pulmonary artery is >3 mm in diameter, intervention to close the shunt should be performed to avoid neurological complications.^{11 12} Up until 1977, surgery was the only method of treating symptomatic PAVFs via lobectomy, whereas conservative management and observation were the preferred choice for those that were asymptomatic or those with small diameter fistulas. Surgical resection, though effective, was associated with increased morbidity related to the procedure, as well as prolonged hospitalisations.⁹ In 1977, transcatheter embolisation therapy came along as an alternative successful method through the use of steel coils and/or detachable balloons.²⁵ More recently, since the use of balloon therapy or coils have shown to be expensive and time consuming, a new approach has been developed using vascular plugs, which are both easy to deploy and efficient. Since their introduction in the 1980s, transcatheter embolisation has been the treatment of choice for the management of PAVFs, avoiding prolonged hospital stays, general anaesthesia and lung resection among others.^{11 26–32} Success rate after embolisation has been reported as 96% evidenced by CT imaging showing shunts as either undetected or reduced in size.³³ The mechanism for this success is thought to be related to thrombosis and involution of the aneurysmal sac after successful embolisation.^{31 33}

To date, no experimental studies or randomised control clinical trials have supported the fact that embolisation should be the mainstream treatment for PAVFs. Therefore, our case will hopefully encourage the development of further registry studies to improve the management, safety and outcome of PAVFs interventions.

Learning points

- This case demonstrates the importance of formulating a broad differential diagnosis when a right to left cardiac shunt is suspected as the cause of a paradoxical embolism.
- It highlights the most efficient way to detect pulmonary arteriovenous fistula through the use of CT pulmonary angiography.
- The pulmonary arteriovenous shunt can be closed percutaneously with great efficacy and safety.

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Contributors MC contributed with data collection gathering, designing the case report, data analysis and interpretation. Also contributed by writing the initial and the subsequent drafts after being revised by all the authors involved, as well as submitting the final case report. MWL (deceased) contributed with data analysis and interpretation as well as drafting and critically revising the article. As primary attending, he evaluated patient, made the diagnosis of pulmonary arteriovenous fistula (PAVF) and was the primary operator during the pulmonary angiography as well as the main operator for the closure of the PAVF as described in the manuscript. GSM contributed with data analysis and interpretation as well as drafting and critically revising the article. He approved the final and submitted version of the case report. All authors critically reviewed, edited and approved the manuscript.

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