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An isolated fistula between the right pulmonary artery and the right pulmonary vein: An unusual cause of stroke in a young female

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Abstract

We describe the case of a 29 year-old female who presented with right sided hemiparesis with global aphasia. She had a history of transient ischemic attack with migraine headaches. Diagnostic workup revealed a right to left cardiac shunt. An isolated right pulmonary artery to left pulmonary vein fistula was diagnosed on pulmonary angiogram. The fistula was occluded successfully by cardiac catheterization. Early recognition and intervention is indicated to prevent further complications. (Cardiol J 2011; 18, 1: 73–76)

Key words: stroke, pulmonary arterio venous fistula, transcatheter closure

Introduction

Pulmonary arterio venous fistula (P-AVF) in Rendu-Osler-Weber disease (ROW) or hereditary hemorrhagic telengiectasia (HHT) with right to left shunt causing paradoxical embolus and stroke, is well known and its incidence is approximately 14% for single and 27% for multiple P-AVF [1–3].

However, isolated P-AVF (without HHT) causing paradoxical brain embolism is uncommon. There are few cases reported in the literature with evidence of asymptomatic deep vein thrombosis, septic emboli, and varicose veins as the source of thrombus formation leading to stroke and brain abscess [4–6]. We report an isolated P-AVF in a young female in the absence of physical findings, without clinical features of HHT, negative work-up for any source of thrombosis, and P-AVF as a plausible source of paradoxical embolism and stroke.

Case presentation

A 29 year-old female from Poland, visiting her sister in the US, was brought to the emergency room in an unresponsive state. On examination she was found to have right sided flaccid paralysis with subtle right facial palsy and global aphasia. An initial computed tomography scan was negative for a cerebral bleed. Magnetic resonance imaging revealed a huge infarct in the middle cerebral artery territory. Chest X-ray was normal and electrocardiogram did not show any arrhythmia. On further questioning it was found that she had a transient ischemic attack seven years ago, and suffered chronic migraine headaches. She has never smoked and had no excessive alcohol, or any illicit drug, use. She was not on any medications including oral contraceptive pills except the over-the-counter Motrin or Naproxen for her migraine. She had no history of

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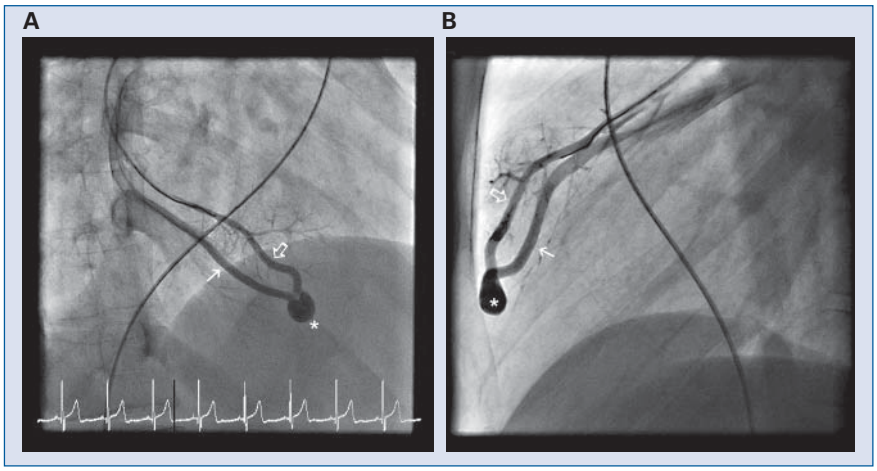

Figure 1. Isolated pulmonary arterio venous fistula from right lower pulmonary artery (RLPA) to right lower pulmonary vein (RLPV); **A.** Anterio-posterior view right anterior oblique projection; **B.** Lateral projection with selective RLPA angiogram demonstrating an afferent feeding artery (open arrow) entering an aneurysmal sac (*) of the fistula and the efferent vessel (arrow) draining into the RLPV.

telangiectasia, recurrent epistaxis, or family history of the ROW disorder.

She had normal physical examination and normal oxygen saturations in both supine and standing positions. Other than positive neurological signs, there was no cyanosis, no clubbing, no carotid bruit, no irregular rhythm nor heart murmur. Her lab work-up revealed neither arterial hypoxia nor polycythemia. Her complete coagulation work-up, carotid ultrasound, and peripheral Doppler studies for asymptomatic deep vein thrombosis were negative. Her cardiac evaluation was negative for structural heart disease. Transthoracic echocardiogram showed a positive bubble study which was confirmed by transesophageal echocardiogram. It was unclear if she had a delayed or early positive contrast bubble study. Patent foramen ovale (PFO) was suspected. Further evaluation by a cardiac catheterization demonstrated no PFO and selective pulmonary angiography revealed a right to left shunt with a fistulous connection from the anteromedial aspect of the right lower pulmonary artery (RLPA) to the right lower pulmonary vein (RLPV).

She was started on anti-coagulation and referred for trans-catheter closure of the fistula. Using a 6 Fr sheath in the right femoral vein, a venous catheter was advanced into the right and left pulmonary arteries. An agitated contrast saline injection under transesophageal echocardiogram guidance showed positive contrast study in the right lung. A 4 Fr JB glide catheter was advanced to the feeding RLPA and a selective angiogram was performed. This demonstrated an isolated P-AVF. The feeding (afferent) RLPA coursed antero-inferiorly; entering an aneurysmal sac and an efferent vessel curved acutely coursing posterio-superiorly and drained into the RLPV. The afferent feeding artery measured 3mm, central aneurysmal sac was 7 mm × × 8 mm in size, and the efferent vessel measured 3 mm (Fig. 1A, B). The JB catheter was advanced to the tip of the aneurysmal sac over a 0.018# Flex-T guide wire. An 8 mm × 5 cm Gianturco coil (Cook Inc., Bloomington, IN, USA) was advanced using a 0.038" glide wire and deployed into the aneurysmal sac of P-AVF successfully. Post coil delivery angiogram showed complete occlusion of the fistula without residual flow (Fig. 2A, B). Repeat right and left pulmonary artery angiograms were negative for any additional P-AVF and agitated saline bubble contrast study in both the branch pulmonary arteries were also negative, indicating complete occlusion and no additional lesions. She was discharged home on the same day and treated with oral Plavix for two months and placed on oral Coumadin for a total of six months. She improved with physical and occupational therapy post stroke, traveled back to Poland and is leading an active lifestyle. At the last follow-up over the phone, she had made a complete physical recovery, except for a mild speech impairment. She continues to receive speech therapy.

Discussion

Pulmonary arterio venous fistula are vascular malformations which can be congenital or acquired. More than 80% of P-AVF are congenital, and 45–

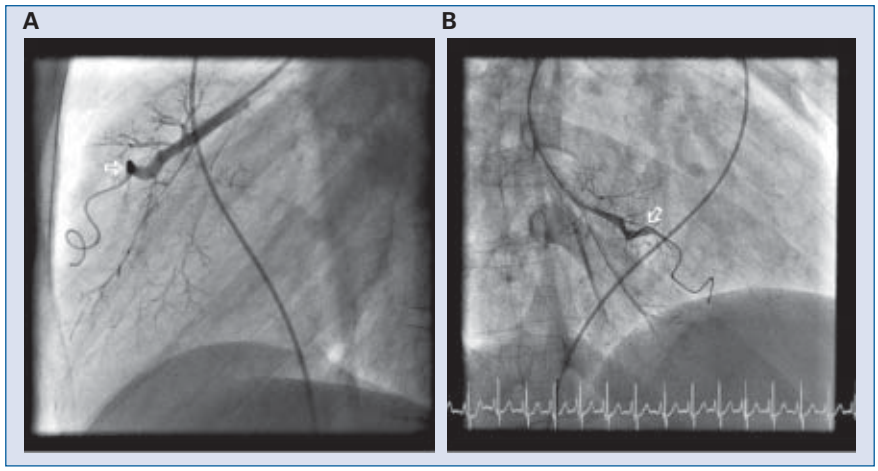

Figure 2. Successful coil occlusion of the pulmonary arterio venous fistula; **A.** Lateral projection; **B.** Anterio-posterior view showing selective right lower pulmonary artery angiogram following successful coil occlusion (open arrow) of the feeding artery without residual leak.

–80% are associated with ROW or HHT. Its associated central nervous system complications include migraine 43%, transient ischemic attack 37%, stroke 18%, abscess 9%, and seizure 8% [1–3]. In the absence of ROW or HHT manifestations in our patient, we report an isolated P-AVF with presentation of cryptogenic stroke.

Isolated P-AVF is rare and most remain asymptomatic except for an occasional presentation of pulse oxygen desaturation [7]. Our patient had normal oxygen saturation and negative auscultatory findings due to small amount of shunting through the fistula. Isolated P-AVF as a basis of acute ischemic stroke is reported to be about 0.5% [5]. Paradoxical embolism due to right to left extra cardiac shunting of peripheral venous origin as the mechanism for neurological event has been described. Kimura et al. [5] reported four patients with isolated P-AVF with cryptogenic stroke. All patients had asymptomatic deep vein thrombosis and three had pulmonary embolism as a source of paradoxical embolism. Similarly, Reguera et al. [8] reported a 37 year-old male with large lower extremity varicose veins as a likely source for paradoxical cerebral embolism with isolated P-AVF. There are two case reports of isolated P-AVF with septic paradoxical emboli causing brain abscess requiring intravenous antibiotics and surgical decompression [4, 6]. In contrast, similar to our patient there are only a few case reports to date of cryptogenic stroke from isolated P-AVF in the absence of cardiac or extra-cardiac source of embolus. Peters et al. [9] reported a patient with recurrent stroke after PFO closure with Helix device. Further evaluation with agitated saline contrast transesophageal echocardiogram study was positive, raising a suspicion of a residual PFO defect and the patient was placed on anticoagulation. Despite medications he presented with recurrent transient ischemic attack in six months prompting cardiac catheterization. There was no residual PFO and pulmonary angiography revealed isolated P-AVF in the left lower lobe that was successfully coil occluded.

In our patient, due to lack of another cause of embolism and the presence of isolated P-AVF with a central dilated aneurysmal sac, the potential for flow stasis, and thrombus formation, exists. However, there was no evidence of thrombus in the aneurysmal sac. Accordingly, a credible explanation for paradoxical embolus is purely speculative. In the absence of physical findings, contrast echocardiography with agitated saline either by transthoracic or transesophageal echocardiogram with or without valsalva maneuver will be particularly helpful for diagnosis [6, 9].

Transcranial Doppler has also been used successfully for diagnosis, especially in aphasic patients [5]. The timing of contrast appearance in the left atrium after injecting agitated saline is critical. Immediate opacification following the same or subsequent heart beat confirms direct interatrial communication (PFO). A delayed opacification suggests presence of P-AVF where the contrast bypasses the capillary bed and arrives in the left atrium 3–5 beats later. Furthermore, during catheterization contrast injection in the right atrium and pulmonary arteries under transesophageal echocardiogram guidance would help differentiate PFO and P-AVF. Nonethe-

less, pulmonary angiography by catheterization remains the gold standard for diagnosis, precise location and definition of the fistula. Similarly, in our patient, agitated saline contrast study in the right pulmonary artery followed by pulmonary angiography confirmed the presence of isolated P-AVF.

Following diagnosis, most isolated P-AVF closure can be accomplished by interventional catheterization using various coils for small to moderate sized fistula [5] and devices such as Amplatzer vascular plugs for larger fistulas [10]. Rarely, surgical resection or lobectomy for P-AVF which are multiple complicated or for failure of trans-catheter therapy is required [11]. A placement of the device such as vascular plug in the afferent vessel might have excluded the distal aneurysmal sac with potential for thrombus formation and embolus post-closure. Therefore, we opted to deploy the coil in the aneurysmal sac with proximal extension into the feeding artery (Fig. 2A, B). Complications or recurrence of stroke following post coil or device occlusion of isolated P-AVF have not been reported. However, recurrent neurological complications in HHT patients due to appearance of new or recurrent P-AVF are not uncommon [3]. Nevertheless, anticoagulation with antiplatelet agents or warfarin to prevent thrombus from residual distal sac for a short duration may be acceptable.

Conclusions

In conclusion, our case calls attention to the prospect of right to left shunt from isolated P-AVF as a potential source of paradoxical embolism causing stroke. Most importantly, absence of physical findings and any source of thrombus necessitate additional investigation with contrast echocardiography and cardiac catheterization for the diagnosis of P-AVF. Early recognition is imperative to ensure timely definitive trans-catheter treatment of this rare lesion to prevent further complications.

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