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A Case Report on Pulmonary Arteriovenous Fistula with Recurrent Cerebral Infarction

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ABSTRACT

Patent foramen ovale (PFO) and pulmonary arteriovenous fistula (PAVF) have been both proposed as a mechanism for cerebral infarction. However, there are only a few reports on how to distinguish the role of the two factors in cerebral infarction.

CASE REPORT

A 52-year-old female patient suffered from recurrent cerebral infarction three times. Five years ago, she had sudden syncope with unknown cause, with the loss of consciousness, the disorder of speech, and an inability for physical activity. Brain magnetic resonance imaging (MRI) showed hemorrhagic cerebral infarction in the junctional zone of the left occipital lobe, but had no sequelae after symptomatic treatment. She had left hemianopsia 3 years ago, which was verified by brain MRI indicating hemorrhagic cerebral infarction in the right occipital lobe. She then received symptomatic treatment with no occurrence of sequelae. Additional sudden syncope occurred 3 months ago, with aggravated left hemianopsia, and large-area cerebral infarction at the right side as shown in brain computed tomography (CT). Although receiving symptomatic treatment, she complained of slurred speech and impaired sight, hearing and memory. The patient had a history of stripping for the varicose right great saphenous vein which occurred 10 years ago.

Findings of physical examination were as follows: blood pressure of 120/80 mmHg (1 mmHg = 0.133kPa), clear sound of bilateral breathing, heart rate of 80 beats/min, regular cardiac rhythm, no edema in lower limbs. Findings of neurological examination were as follows: clear consciousness, partial mixed aphasia, the shallow left nasolabial groove, the deviation of tongue, the paresis of right limbs, negative results in the other parts. Findings of other examinations were as follows: normal results in the routine blood test and lipid assay; no atherosclerotic plaque or stenosis found in carotid ultrasound; curtain microembolic signals detected by the contrast transcranial Doppler (cTCD) within 10s after the resting state and Valsalva maneuver (VM); the patent foramen ovale (PFO) shown in transesophageal echocardiography (TEE); by contrast transthoracic echocardiography (cTTE), numerous micro-bubbles were visualized in the resting state in the left heart within the second cardiac cycle (turbid throughout the chambers of the heart).

With no exclusion of recurrent cerebral infarction induced by PFO-paradoxical embolism, the PFO closure procedure was attempted with a PFO size of 0.2 mm by selective ovalis fossa angiography. The catheter failed to pass through the PFO in our attempts. Considering the smaller size of the PFO in this patient, which causes a lower risk of paradoxical embolism, the contrast echocardiography of the right pulmonary artery was intraoperatively performed, indicating a large amount of the right-to-left shunt (RLS); but no RLS was observed in the contrast echocardiography of the left pulmonary artery; and the pulmonary arteriovenous fistula (PAVF) was shown in selective right pulmonary angiography (Figure 1). This suggests a high possibility of recurrent cerebral infarction induced by PAVF. Therefore the procedure

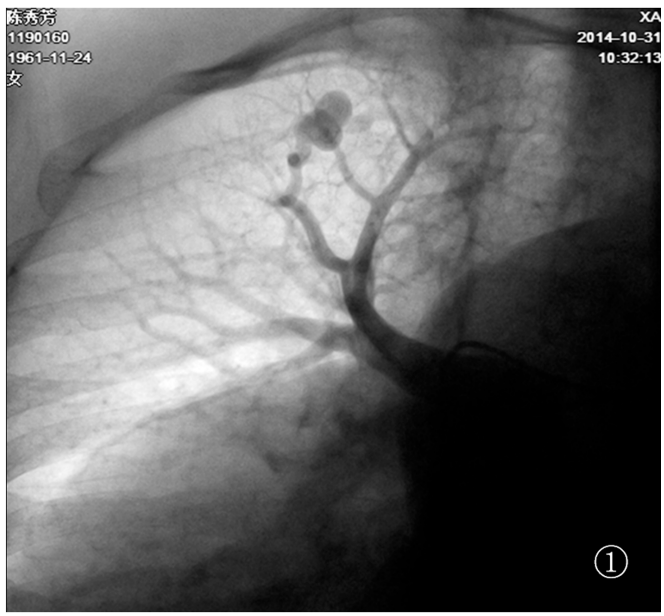

Figure 1. A huge, abnormal expansion (with a diameter of about 5 mm) at approx. 17 mm from the origin of the right pulmonary artery, with blood flowing back to the left atrium through two abnormal branches.

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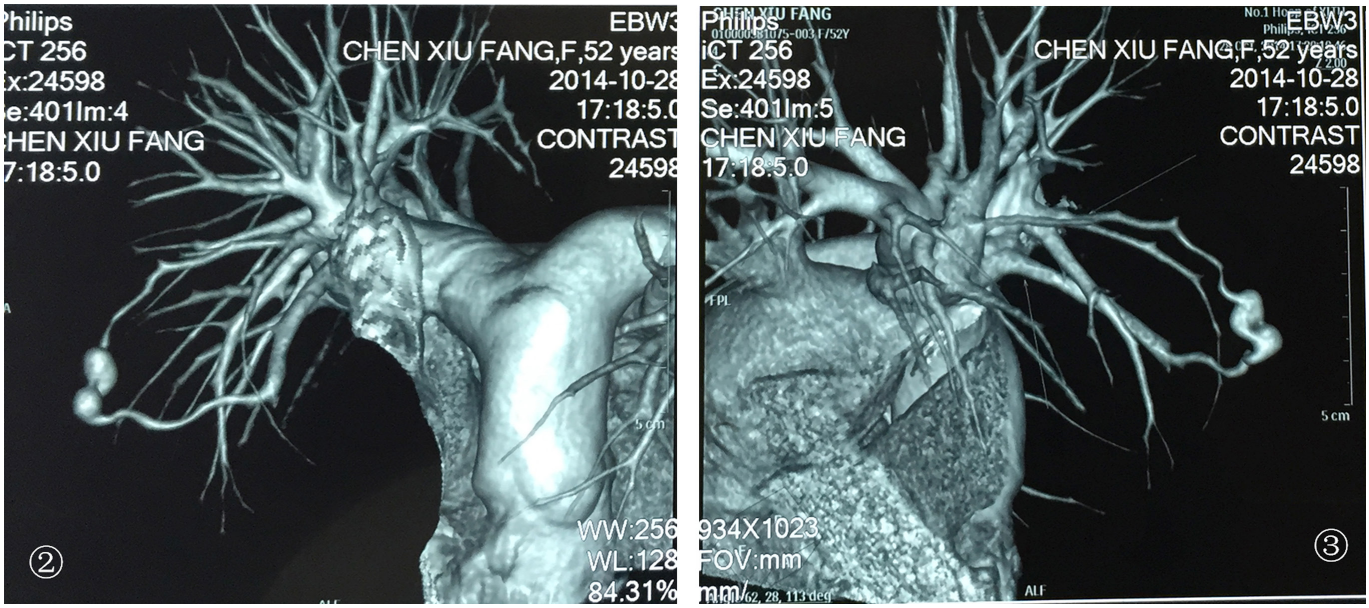

Figure 2. The bead-like cystic neoplasm (approx. 15 mm × 7.8 mm) visualized in the anterior segment of the superior lobe of right lung, to which the artery in the anterior segment and the anterior branch of the vein in the superior lobe of right lung are connected.

was suspended for additional evaluation, and afterwards the pulmonary arteriovenous malformation (PAVM) was indicated in the anterior segment of the superior lobe of the right lung through CT pulmonary angiography (CTPA) (Figure 2).

For prevention from the recurrence of cerebral infarction in this patient, the percutaneous closure of PAVF was performed by implantation of one 10/12 mm patent ductus arteriosus (PDA) occluder (Figure 3). Furthermore, the findings in selective right pulmonary arteriography included the disappearance of original PAVF, and the non-affected pulmonary artery and its branches; a tiny amount of RLS was indicated in the contrast echocardiography of the right pulmonary artery, after which the occluder was released. Then aspirin 100 mg/d and clopidogrel 75 mg/d were administrated orally after the procedure for 6 months. In the follow-up for 2 years, the patient did not complained of any discomfort.

DISCUSSION

The PAVF refers to direct connection of one or more pulmonary arteries with pulmonary veins, forming a fistula or tumor-like lesion and bypassing blood capillaries, and in rare cases direct connection of systemic arteries with pulmonary arteries and/or pulmonary veins with the left atrium. As an uncommon kind of vascular malformation, the disease is congenital in most cases with the incidence of 2-3 per 100,000. With varied clinical symptoms, the disease is identified as single, multiple, or diffuse lung placeholders, and hypoxia caused by the shunt; 47-80% of PAVF patients have hereditary hemorrhagic telangiectasia (HHT), showing the expanding and bleeding capillaries in the skin surface and mucosa and vascular malformations in the lung, brain, and liver. Multiple complications in the central nervous system are common and probably initially manifested in PAVF, including brain abscess, transient ischemic attack (TIA), migraine, and cerebral infarction with the incidence of 10-19% [Cottin 2007].

Paradoxical embolism refers to the embolism caused by a variety of emboli entering from the low-pressure venous system or the right heart into the systemic circulation through intracardiac or abnormal arteriovenous communicating branches, eventually detained in the arterial system of the brain or other organ, provided that the RLS and paradoxical emboli exist. In this patient, paradoxical embolism was mainly responsible for her cerebral infarction, where the shunt pathway for paradoxical embolism was created by blood flowing from pulmonary arteries directly to pulmonary veins without capillary filtration. The thrombosis in the lower limbs or pelvic vein, and even the non-detected thrombosis in the hemorrhoidal vein may be the main source of the emboli. This patient probably had the emboli from the smaller veins of the lower limbs, due to her history of the varicose great saphenous. Unfortunately, no direct evidence for this presumption was obtained in clinical examinations, possibly because of limited testing methods or the dropped emboli. As reported in previous rare cases, the embolism might be caused by local blood stasis in PAVF as well as the formed and dropped primary thrombi [Cohen 2006]; the polycythemia secondary to chronic hypoxia may also facilitate the thrombosis [Cottin 2007].

Currently, PFO is widely recognized as an intracardial shunt pathway. But the extracardial pathway in PAVF, rarely reported, has not been clearly known. Based on positive results in both cTTE and cTCD, PFO was determined as the cause for recurrent cerebral infarction of this patient, which was verified by TEE. From this point, the reasons for missed diagnosis and preventive measures are summarized as following: 1) The characteristics of embolism shown in brain imaging should be primarily investigated, in combination with the patient's medical history, family history, and physical

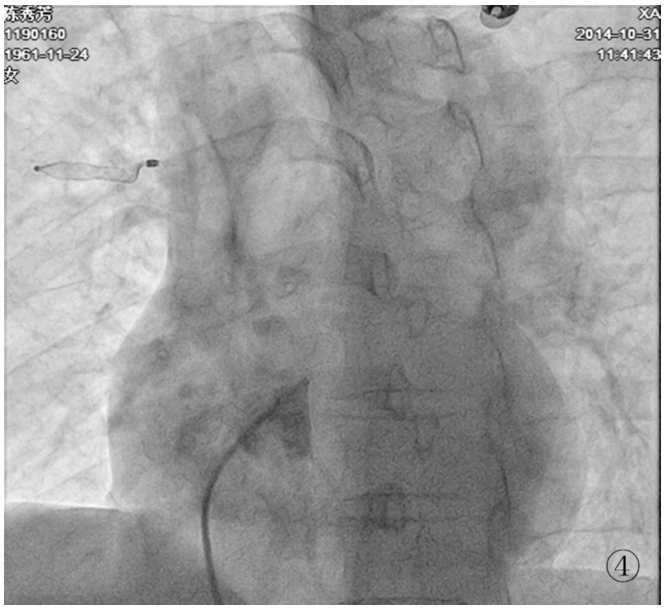

Figure 3. Insertion of 10/12 mm patent ductus arteriosus occluder in the right upper pulmonary arteriovenous fistula.

examination for the signs (including bleeding and hypoxia) in lung, skin surface, and mucosa. There is a high possibility of missed diagnosis in the case that cerebral infarction is the initial and only clinical manifestation, particularly in patients at an older age of onset with conventional risk factors for cerebral infarction. 2) Routine screening of RLS is required in patients with cryptogenic stroke, and PFO is not the only cause for RLS. RLS is indicated by positive results in cTCD, including intracardial and extracardial pathways. Intracardial pathway is mostly common in PFO, also found in atrial septal defect (ASD); extracardial pathway occurs in intrapulmonary shunt (e.g., PAVF) and rarely in other channels (e.g., azygos vein to pulmonary vein fistula [Huang 2010]). For this patient, the four findings were observed in cTCD and cTTE: continuous RLS (even in resting state), early RLS (upon injection), a large amount of shunt (curtain microembolic signals), and a minor effect of VM on RLS. The identification between PFO and PAVF could not be achieved according to a single one of the findings, which was nevertheless suggested by comprehensive analysis of all the findings. This is because the shunt pathway in PFO mainly depends on the size of PFO and the pressure gradient, and RLS occurs only when the pressure in the right atrium is higher than that in the left atrium. A tiny amount of shunt or no shunt was found in PFO with a small size in the resting state, and a larger amount of shunt occured after VM; the substantial, continuous shunt may be observed in the large-sized PFO in case of right atrial enlargement and pulmonary hypertension. If no shunt or a minor shunt occurs in resting state and a larger increased amount of shunt exists after VM, PAVF is unlikely developed; a tiny amount of shunt in resting state indicates the exclusion of PAVF that causes a large amount of shunt and clinically suggests a risk of embolism, but no exclusion of PAVF. Though theoretically, it is supposed that the shunt through pulmonary circulation in PAVF occurs later than that in PFO. The shunt was observed within 10 seconds in this patient; also as reported in some literature the shunt in PFO could occur up to 40 seconds [Jauss 2000]. Therefore, PFO and PAVF cannot be reliably identified merely based on the occurrence time of shunt.

This patient had no symptoms before and after the onset of stroke, which suggests that cerebral infarction was the early clinical manifestation in PAVF. From this point, PAVF may occur in patients with cryptogenic stroke. CTPA, a highly sensitive diagnostic method for PAVF, has obvious advantages over pulmonary arteriography in the diagnosis of PAVF and the visualization of anatomy, which is expected in substitution for arteriography. Considering the patients have a significantly increased risk of embolism, with the feeding artery of a diameter over 3 mm [Sun 2012], active treatments should be given for them upon diagnosis of PAVF, including surgical therapy taken as the primary treatment option for PAVF.

Conventional treatment for PAVF by surgical resection of the affected lobe of lung has some disadvantages, including large wounds, more complications, slower recovery, and partially lost pulmonary functions; surgical procedures are contraindicated in those with multiple PAVF. Transcatheter interventional closure of PAVF, an alternative for surgical procedure, is preferred in the treatment of PAVF, with multiple advantages over surgical approach including simple operation, small wounds, and safe and effective treatment [Joseph 2013]. As reported in the literature, the PAVF occluder is mostly composed of easy-to-shift spring coils [Abushaban 2004], which probably causes systemic embolism by transportation, residual shunt, and a high recanalization rate [Mager 2004]. The occluder device made in China was chosen for this patient with PAVF through the access vessel with a larger diameter (5 mm) because a number of coils are costly if the device is purchased overseas. Similar to the Amplatzer Plug, the made-in-China PDA occluder device has some desirable features, including easy-to-use and one-step operation for complete closure; this recyclable and relocatable device has been successfully applied in interventional treatments of PAVF in patients with the large-sized feeding artery. The occluder device should be placed on the distal section of the feeding artery, with a diameter 2-4 mm larger than the artery, for prevention from its falling into the cystic neoplasm and the closure of normal proximal pulmonary arteries, thus contributing to a far lower incidence of the occluder shifting and falling-out and a lower possibility of postoperative recanalization compared with the device containing coils.

In conclusion, transcatheter closure of PAVF is a good interventional treatment ensuring small wounds, high safety, and exact clinical effects, provided that the indications are strictly followed. The spring coils could be used for PAVF closure in the access vessel with a diameter smaller than 5 mm, and Amplatzer Plug or PDA occluder is the preferred choice for the access vessel with a diameter larger than 5 mm.

Unfortunately, no direct evidence of thrombi in the overriding pulmonary arteries and veins was found in this patient, which only presumptively suggests her recurrent cerebral infarction induced by paradoxical embolism. Although this patient had good recovery in the 2-year follow up, the safety

and effectiveness of the closure procedure remains to be determined in the long-term follow up. Using this method of clinical diagnosis and treatment, clinicians have profound insights in the recognition of paradoxical embolism, knowing that PAVF-paradoxical embolism can induce cryptogenic stroke, in addition to the correlation of the embolism with intracardiac abnormal channels. This provides a new basis for the screening of etiological factors in the patient population with cryptogenic stroke.

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