Pulmonary Arteriovenous Malformation as a Cause of Cerebrovascular Accident

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Abstract

Although pulmonary arteriovenous malformation is not a common condition, it is one of the causes of paradoxical embolism presenting with neurological manifestation. Therefore, pulmonary arteriovenous malformation should be considered in any patient with an arterial embolism, especially patients with cerebrovascular accident from an unidentified source. We report a case of paradoxical embolism of the brain due to an isolated pulmonary arteriovenous malformation (Iranian Heart Journal 2013; 13 (1):57-60).

Pulmonary arteriovenous malformation (PAVM) is a rare condition in which there is an abnormal pulmonary artery to vein communication. PAVM is often congenital but it can be found in a variety of clinical situations such as liver disease or after congenital heart disease repair or in the context of systemic disorders like hereditary hemorrhagic telangiectasia (HHT).¹ Approximately, 70% of PAVMs occur in patients with HHT but they can be isolated as well.² PAVMs occur twice in women and nearly 10% of PAVMs are diagnosed in infancy or childhood, followed by a slight rise in incidence through the fifth and sixth decades.³ PAVM is one of the rare but well-recognized causes of paradoxical brain embolism due to a continuous right-to-left shunt⁴⁻⁷ and as such should be suspected in any patient with an arterial embolism from an unknown origin. Patients with paradoxical embolism present with neurological manifestations attributed to a stroke or transient ischemic attack or clinical features suggesting carotid, renal, splenic, mesenteric, or peripheral arterial embolism. Since classic clinical manifestations or radiologic findings may be absent in some patients with PAVM, which renders the diagnosis more difficult, any patient with cryptogenic stroke should be probed for PAVM, not least in the absence of patent foramen ovale (PFO).

Case Report

A 48-year-old woman was admitted to our hospital due to vertigo, nausea and vomiting of two days’ duration, and numbness of the left side of the face. History and physical examination revealed ataxia and some degree of the involvement of the maxillary and mandibular branches of the left trigeminal nerve. Right plantar reflex was positive, and deep tendon reflex in the upper extremities was hyperactive. Other examinations such as heart, lung, abdomen, and skin were normal. Brain MRI showed right cerebellar hemisphere infarction and ischemic disease of the small vessels. Electrocardiography (ECG), chest X-ray, and laboratory finding were normal. Doppler sonography of the cervical vessels showed no vascular pathology. Normal antiphospholipids and anticardiolipins excluded collagen-vascular disease as well.

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Transthoracic echocardiography did not reveal any source of emboli, including valvular disease, dilated cardiomyopathy, akinetic left segments, intracardiac thrombus, and myxoma. Transesophageal echocardiography (TEE) was performed, which demonstrated that the left atrium and left atrial appendage had no thrombus and the left ventricle and aorta were normal. In contrast echocardiography after the injection of contrast and opacification of the right atrium, the left atrium was filled by contrast too but the intra-atrial septum was normal with no atrial septal defect, PFO, or aneurysm. The contrast study was repeated for an evaluation of the left pulmonary vein. Just after the opacification of the right atrium, a large amount of contrast bubble entered the left atrium via the left upper pulmonary vein, revealing the presence of a pulmonary arteriovenous fistula at the level of the left upper pulmonary vein.
Discussion

There is a strong association between PAVM and various neurological problems. Cerebral cortical infarction was documented in 14% of patients with single and higher multiple malformations. The incidence of stroke in the White et al. study was reported 18% and in other studies was also reported from 2.6% to 25% with an average of 8.5%. However, only a few cases of paradoxical embolism of the brain due to an isolated PAVM have thus far been reported. In the patient presented herein, the other causes of stroke such as atherosclerotic and non-atherosclerotic arteriopathies, hematological and systemic disorders, and coagulopathies were ruled out and no embolic sources were detected. It has been previously demonstrated that deep vein thrombosis (DVT) may be occult in more than 50% of patients with paradoxical embolism. We failed to find any DVT in our patient. We also ruled out PFO, atrial septal defect, and left atrial and atrial appendage clot as a source of emboli by TEE. The patient had a single PAVM in the left upper pulmonary lobe, which led to right cerebellar hemisphere infarction, while the most common site of PAVM is the lower lobe often next to the pleura.

The present study confirmed that although the PFO is the most important cause of young adult stroke after arrhythmias (similar to atrial fibrillation and valvular and vascular diseases), it is prudent that PAVM be taken into consideration as well. It should also be noted that PAVM may be sometimes misdiagnosed with the PFO in contrast echocardiography. Therefore, there should be a high clinical suspicion when making the diagnosis.

References


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