Journal of Community Hospital Internal Medicine Perspectives

co-action PUBLISHING

CASE REPORT

Intrapulmonary arteriovenous malformation causing recurrent strokes

Kareem Abed, MD, MBA*, Lalith Premachandra, MD, Viren Vankawala, MD, FACC and Qi Sun, MD

AtlantiCare Regional Medical Center, Atlantic City, NJ, USA

This case reveals a left pulmonary arteriovenous malformation (PAVM) as a cause of recurrent cerebral and cerebellar emboli. Extensive workup excluded other etiologies of emboli formation, and the patient was transferred to a tertiary care center for percutaneous embolotherapy. In the absence of a clear etiology, PAVM should be considered as a potential cause of recurrent cerebral emboli, especially in the absence of carotid disease, intracardiac thrombus, atrial septal defect, and patent foramen ovale. Diagnostic work-up for the PAVM can be cost effective and expedited by utilization of agitated saline contrast echocardiography, as noted in our case.

Keywords: *PAVM*; *embolotherapy*; *contrast echocardiography*; *intrapulmonary arteriovenous malformation*

*Correspondence to: Kareem Abed, 2029 Cedar Lane Ext., Bordentown, NJ 08505, USA, Email: abedlkareem@gmail.com

Received: 12 June 2015; Revised: 17 August 2015; Accepted: 21 August 2015; Published: 19 October 2015

Ischemic cerebral vascular accidents, especially wake-up strokes with unclear etiology, have been difficult to manage. In the absence of carotid disease, atrial fibrillation, or atrial septal defects such as patent foramen ovale, many patients are discharged home without a clear source of their cerebral infarct (1). This case report stresses the impact of intrapulmonary arteriovenous malformation and the methods of establishing a diagnosis when the cause of the cerebral infarct is unclear.

Introduction

Intrapulmonary arteriovenous malformations were first diagnosed in 1897 and are generally rare, occurring at an annual rate of 2–3 cases per 100,000 (2). Pulmonary arteriovenous malformation (PAVM) is an arteriovenous shunt, which can be the source of paradoxical cerebral emboli. PAVMs are more common in females, and most patients are relatively asymptomatic during the course of their lifetime (3). Nearly 60% of PAVMs are asymptomatic, and one third of patients have neurologic findings (4). A direct connection between the high-flow pulmonary artery and low-resistant veins, which bypasses the capillary bed within the pulmonary vasculature creating a right to left shunt, can lead to dyspnea, hemoptysis, hypoxia, stroke, and brain abscess.

Case report

A 63-year-old Caucasian male with a past history of supraventricular tachycardia and recurrent deep vein thrombosis, who had been non-compliant with his daily Rivaroxaban® 20 mg, presented with pleuritic chest pain and was found to have a right lower lobe pulmonary embolus. The patient was subsequently restarted on Rivaroxaban® and discharged. He presented again 5 days later with left arm weakness and was found to have multiple areas of punctuate infarctions of both cerebral and cerebellar hemispheres, more confluent within the right frontal lobe on magnetic resonance imaging (MRI) of the brain (Figs. 1 and 2). He has been compliant with his Rivaroxaban® since the previous hospital discharge 5 days prior. The patient was admitted and subsequently started on unfractionated heparin. On admission, his vital signs were blood pressure 138/93, heart rate 65 beats per minute, respiratory rate 16, and oxygen saturation 97% on room air. Cardiopulmonary examination revealed regular rate and rhythm, without murmurs, rubs, or gallops and clear lungs on auscultation. Neurologic examination revealed intact cranial nerves, preserved strength in all extremities with an exception of mild dysmetria in the left upper extremity and an NIH score of 1. Electrocardiogram revealed normal sinus rhythm, non-specific ST changes, and a premature ventricular contraction. Transthoracic echocardiogram was notable for normal left ventricular function with mild tricuspid regurgitation and stage 1 diastolic dysfunction. Transesophageal echocardiogram was negative for intracardiac thrombus. The echocardiography with agitated saline contrast revealed microbubbles of moderate opacity entering the left atrium

Citation: Journal of Community Hospital Internal Medicine Perspectives 2015, **5**: 28843 - http://dx.doi.org/10.3402/jchimp.v5.28843

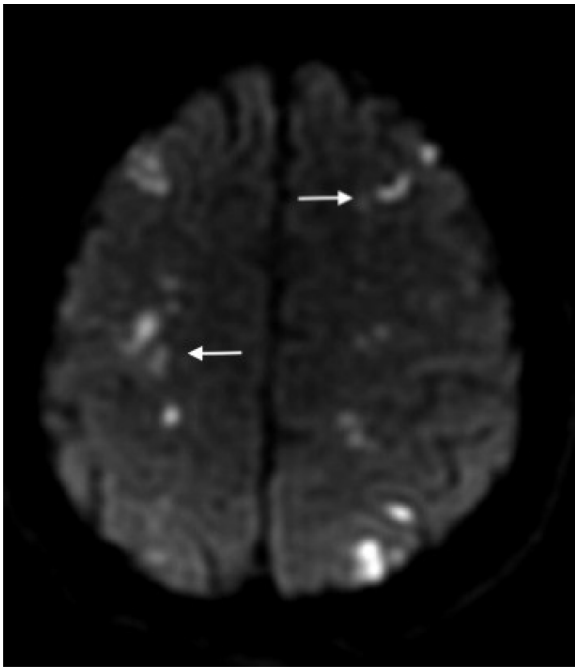

Fig. 1. Diffusion MRI brain without contrast showing extensive infarctions within both cerebral hemispheres (see arrows).

from the left lower pulmonary vein indicative of a grade 2 intra-pulmonary arteriovenous shunt (Fig. 3). There was no atrial septal defect or patent foramen ovale. Unfractionated heparin was continued, and the patient was transferred to a tertiary care center for embolization of the

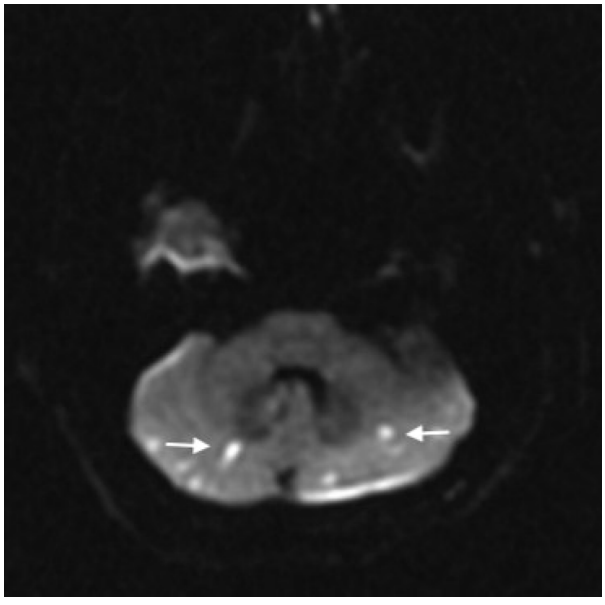

Fig. 2. Diffusion MRI brain without contrast showing multiple infarctions within the cerebellar hemispheres (see arrows).

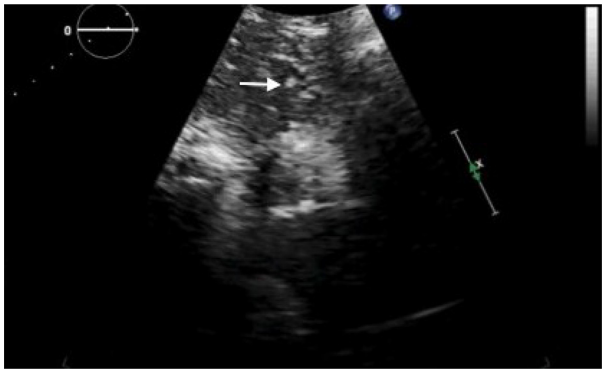

Fig. 3. Echocardiography with agitated saline contrast noting microbubbles entering the left atrium from the left lower pulmonary vein.

shunt. There, he developed recurrent cerebral infarctions while anticoagulation was temporarily held. Computed tomography of the abdomen and pelvis was done for persistent abdominal pain, and noted an omental mass, and subsequent exploratory laparotomy revealed an adenocarcinoma within the omentum with nodular peritoneal implants. The patient was placed on weight-based Enoxaparin® and discharged home with scheduled outpatient chemotherapy. Embolotherapy of the intrapulmonary shunt was deferred given this new diagnosis of malignancy.

Discussion

This case illustrates the embolic outcome of intrapulmonary arteriovenous (AV) shunt and the characteristic presentation of those with paradoxical emboli. Classic symptoms of dyspnea, clubbing, and cyanosis are only seen in a small population of patients. These may be due to hereditary or idiopathic phenomena. Most cases are present at birth and are generally benign until the fifth or sixth decade of life when increased pulmonary vascular pressure is more prominent and patients become symptomatic, leading to higher rates of mortality if left untreated (5). Intrapulmonary AV malformations have a 60–90% association with Hereditary Hemorrhagic Telangiectasia, an autosomal dominant syndrome, also referred to as Osler–Weber–Rendu syndrome, suggesting a primary cause of PAVM (5). Patients have unilateral segments of AVM present in the lower lobe of the lung in more than 50% of cases, and one third of the patients have multiple lesions, which are typically 1–10 cm.

Use of agitated saline contrast echocardiography is noninvasive and readily provides diagnosis of intrapulmonary AV shunt. After contrast is injected with agitated saline, significant amount of bubbles entering the left atrium from the left pulmonary vein is diagnostic, as noted in our case. Most shunts drain into the left atrium,

Citation: Journal of Community Hospital Internal Medicine Perspectives 2015, **5**: 28843 - http://dx.doi.org/10.3402/jchimp.v5.28843

as noted in this patient, however some may drain into the inferior vena cava or even into the innominate vein (6). Other modalities for diagnosis of intrapulmonary shunt include: nuclear scintigraphy with injection of technetium Tc99m labeled macroaggregated albumin, computed tomography of the chest with contrast, and pulmonary angiography. Although computed tomography with contrast is used as the modality of choice, agitated saline contrast echocardiography is certainly more feasible and cost-effective, and provides a favorable method to obtain this diagnosis. Percutaneous embolotherapy of the shunt is the preferred treatment; however, surgical intervention may still be an option in select individuals (7).

Conflict of interest and funding

The authors have not received any funding or benefits from industry or elsewhere to conduct this study.

References

1. Papagiannis J, Apostologpoulou S, Sarris GE, Rammos S. Diagnosis and management of pulmonary arteriovenous malformations. Paediatr Cardiol 2002; 4(1): 33–49.
2. Khurshid I, Downie G. Pulmonary arteriovenous malformation. Postgrad Med J 2002; 78: 191–7.
3. Coley S, Jackson J. Pulmonary arteriovenous malformations. Clin Radiol 1998; 53: 396–404.
4. Gossage J. Pulmonary arteriovenous malformations. Am J Respir Crit Care Med 1998; 158: 643–61.
5. Meek M, Meek J, Beheshti M. Management of pulmonary arteriovenous malformations. Semin Intervent Radiol 2011; 28: 24–31.
6. Chang RK, Alejos JC, Atkinson D, Jensen R, Drant S, Galindo A, et al. Bubble contrast echocardiography in detecting pulmonary arteriovenous shunting in children with univentricular heart after cavopulmonary anastomosis. Pediatr Cardiol 1999; 33(7): 2052–8.
7. Shovlin C. Pulmonary arteriovenous malformations. Am J Respir Crit Care Med 2014; 190: 1217–28.