

Unusual Cause of Stroke in a Middle-Aged Woman

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INTRODUCTION

Many ischemic strokes are considered cryptogenic as they occur without a well-defined etiology. However, uncovering the pathophysiology affects prognosis, outcome, and management. The Trial of Org 10172 in Acute Stroke Treatment (TOAST) began classifying ischemic strokes as large-artery atherosclerosis, cardioembolic, small-vessel occlusion, stroke of other determined etiology, and stroke of undetermined etiology.¹ Twenty-five to forty percent of ischemic strokes are considered cryptogenic strokes, which are diagnosed through exclusion. The Northern Manhattan Study showed that the prevalence of cryptogenic strokes was higher in African Americans and Hispanics than in Caucasians.² No clear risk association has been found for age and gender.^{3,4}

Multiple mechanisms have been proposed for cryptogenic strokes such as cardiac embolism secondary to atrial fibrillation, paradoxical embolism through a cardiac septal defect, undefined thrombophilia, and sub-stenotic cerebrovascular disease.⁵ However, there are other important and unidentified mechanisms to uncover.

This case study describes a middle-aged woman who presented for an ischemic stroke and was found to have a stenosis of the brachiocephalic vein, likely secondary to chronic pleurisy or congenital malformation.

CASE REPORT

A 44-year-old white female with no significant past medical history except for recurrent left-sided pneumonia presented to the emergency department with left leg numbness and dysarthria. Social history was noncontributory. She was found to have a right middle cerebral artery territories ischemic stroke on magnetic resonance imaging (MRI). Computerized tomography (CT) of the head and CT angiography of the neck were normal. Tissue plasminogen activator was not administered as she was outside the time window. A 2D echocardiogram with bubble study was performed with an injection of the saline contrast through the left antecubital vein. The bubbles appeared initially in the left-sided cardiac chambers before the right-side chambers, which is extremely unusual (Figure 1).

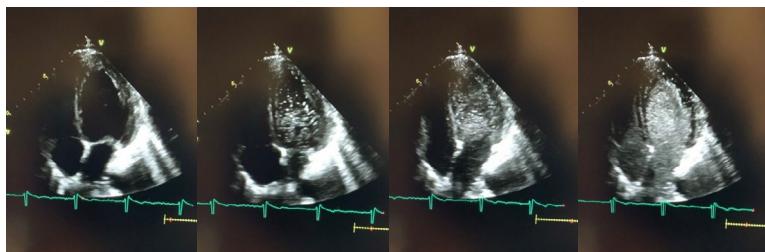


Figure 1. Bubble study showing bubbles in the left-sided chambers before the right.

A CT angiogram of the chest showed a persistent left superior vena cava (SVC) with anomalous venous vasculature consistent with severe stenosis of the brachiocephalic vein with significant collateralization (Figure 2). A transesophageal echocardiography showed a small patent foramen ovale with small densities on the mitral valve (Figure 3).

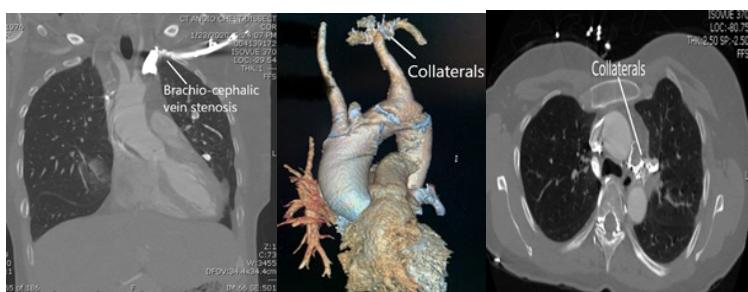


Figure 2. 2D and 3D CT angiogram of the chest showing the brachiocephalic vein stenosis with extensive collateralization.

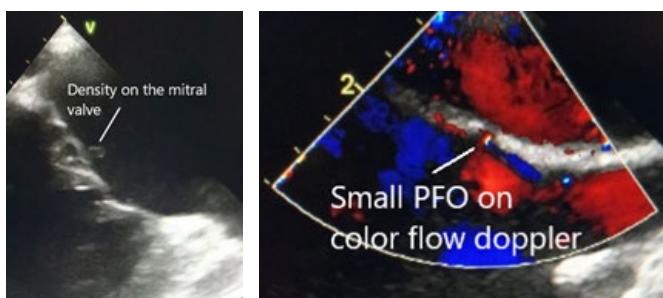


Figure 3. On the left: Small density suggestive of a vegetation on the mitral valve. On the right: Small PFO seen on color flow doppler.

Upper and lower extremities venous Doppler ultrasounds were negative for thrombus. The patient was started on warfarin for anticoagulation and antibiotics for possible endocarditis. She was referred to a congenital heart disease program in a larger community.

DISCUSSION

The patient was found likely to have an embolic stroke on MRI. The carotid arteries were normal, and she had no cardiovascular risk factors. No significant arrhythmias were found on electrocardiogram or telemetry. A suspicion for a cardiac source of embolization was raised. A 2D echocardiogram with bubble study was ordered to rule out a cardiac mass or a patent foramen ovale or atrial septal defect. The bubbles were injected in the left anti-cubital vein and unexpectedly appeared in the left-side chambers initially, instead of the right. This raised the suspicion for anomalous venous return from the left upper extremity to lead the bubbles straight to the left atrium. A persistent left SVC typically would drain the bubbles into the coronary sinus then the right atrium (90% of the cases); 10% of patients with persistent left SVC have a direct connection to the left atrium.² A CT angiogram of

the chest was ordered to evaluate for the left SVC, uncovering severe stenosis of the brachiocephalic vein leading to large collaterals, which in turn anastomosed with the pulmonary veins. Congenital anomalies of the brachiocephalic vein also were a possibility.

On further interrogation, the patient admitted to a history of recurrent pneumonia on the left side which may have caused pleurisy and contributed to the stenosis of the brachiocephalic vein. The densities on the mitral valve may have been new or healed vegetations and could be due to bacterial showering of the left-sided valves from the left upper extremity venous drainage. The patient was treated for possible infectious endocarditis as well as thromboembolic event from the venous malformations. The medical team was fortunate that the IV was placed on the left side, otherwise her venous malformation may have never been discovered. The patent foramen ovale was small and unlikely to have contributed to the patient's presentation.

CONCLUSIONS

The pathophysiology of cryptogenic stroke likely is heterogeneous. This case report proposed a new and unusual mechanism of an ischemic stroke in a young, middle-aged woman. The acute management was similar to other ischemic stroke subtypes and the prognosis tended to be better with a lower long-term risk of recurrence.^{6,7} However, in this case, a cardiothoracic surgeon was consulted to evaluate for possible lobectomy and the interventional radiology service was consulted for stenting of the brachiocephalic vein. After review of the case, it was recommended to transfer the patient to a congenital heart disease team for further evaluation and treatment of this complex vascular patient. Thus, addressing the vascular pathology likely will resolve the cause of her strokes and reduce the risk of recurrence.

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Keywords: persistent left superior vena cava, pulmonary vein stenosis, cryptogenic stroke, incidental findings, case study