Membranous Ventricular Septal Aneurysm Leading to Embolic Stroke

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INTRODUCTION

Echocardiography is often ordered to assess for a cardiac source of embolism after an acute neurologic event. Common cardiac culprits for thromboembolism include cardiac masses, thrombi, vegetations, mobile atheromas, or a cryptogenic embolism in the setting of patent foramen ovale. Membranous ventricular septum aneurysm (MVSA) is a rare abnormality that may be congenital or acquired. Blood stasis within these aneurysms increases the risk of thrombosis formation.

CASE PRESENTATION

A 51-year-old woman with a medical history of diabetes, hypertension, and cognitive impairment presented with aphasia and right-sided hemiplegia. She had no history of congenital heart disease. Her vital signs were within normal limits, and the cardiac examination was unremarkable. Pertinent medication included a daily oral contraceptive. An emergent computed tomography angiogram of the head and neck revealed an acute left middle cerebral infarct, and subsequent cerebral angiogram showed a nonocclusive thrombus within two middle cerebral artery branches consistent with embolic etiology.

A transthoracic echocardiogram (TTE) was ordered to assess for a cardiac source of emboli. The TTE showed a thin-walled outpouching of the basal segment of the MVSA protruding into the right ventricular outflow tract (RVOT), consistent with a MVSA (Figures 1-3, Videos 1-5). Flow was seen into the MVSA by color Doppler (Video 1) with no evidence of a ventricular septal defect (VSD). There was no evidence of RVOT obstruction by pulsed-wave Doppler (Figure 2). Administration of an ultrasound-enhancing agent (UEA) revealed a discrete echo density within the aneurysm suggestive of thrombus (Figure 3B, Video 4). For further assessment of an echodensity, an electrocardiogram-gated cardiac computed tomography (CCT) was ordered that confirmed the MVSA and a 0.8 × 1.2 cm thrombus within the aneurysmal space (Figure 4). No thrombus was seen in the left atrial appendage.

The patient was seen by cardiothoracic surgery, and medical management was recommended. The patient’s oral contraceptive was discontinued, and oral anticoagulation was initiated with close outpatient cardiology follow-up.

DISCUSSION

The membranous septum is located in the midportion of the ventricular septum just inferior to the aortic valve annulus. Aneurysm formation of this segment is an uncommon occurrence and may be related to a spontaneous partial or complete closure of a congenital VSD by approximation of the septal leaflet of the tricuspid valve across the defect. These may also be idiopathic or develop after previous infection or trauma. Transthoracic echocardiogram is the first-line imaging modality; however, supplementary CCT may be helpful when the anatomy remains unclear. Cardiac computed tomography provides improved spatial resolution and is devoid of acoustic shadowing. Given the proximity of the membranous septum to the aortic valve, a sinus of Valsalva aneurysm (SoVA) must be included in the differential diagnosis. Sinus of Valsalva aneurysm is associated with a more aggressive course and a higher likelihood of rupture. Cardiac computed tomography can help distinguish a SoVA from an MVSA as it provides sharper delineation of the cardiac structures. A SoVA shows saccular dilatation of the aortic sinus, compared with an MVSA, which shows an outpouching of the ventricular septum below the aortic valve.

Membranous ventricular septum aneurysms are often clinically silent; however, if present they can be associated with significant complications including thromboembolism, aortic valve leaflet prolapse, tricuspid regurgitation, RVOT obstruction, conduction abnormalities, or acute left-to-right shunting secondary to aneurysmal rupture. The aneurysmal sac allows for stasis of blood flow and the potential for thrombus formation. Distal embolization due to thrombus formation within the aneurysmal space is rare, with few documented case reports. Due to the anatomical location of the membranous septum, interference with the aortic valve can lead to aorto valve leaflet prolapse and subsequent aortic insufficiency. The aneurysm may impinge on the tricuspid valve apparatus, leading to tricuspid regurgitation. If large, the MVSA can protrude into the RVOT and cause obstruction. Despite the large size of the aneurysm in our case, there was no RVOT obstruction or significant aortic or tricuspid regurgitation (Figure 2, Video 6).

Valvular calcification, echocardiographic artifacts, slow flow into the chambers, and poor imaging windows may add technical challenges in evaluating an aneurysm for thrombus. Opacifying aneurysm chambers with an UEA is helpful to delineate chamber borders and evaluate for filling defects that are seen with masses.
Ultrasound-enhancing agent hyperenhancement of visualized masses suggests vascularity and thus can help distinguish neoplasm from thrombus. It is important to note that sweeping through a large aneurysm or using biplane imaging is helpful and often necessary to fully assess the entire aneurysm for the presence or absence of a filling defect. A small thrombus within the aneurysm may be missed if only a single location is imaged.

Cardiovascular magnetic resonance imaging provides a higher sensitivity for identifying left ventricular thrombus compared with TTE. Cardiovascular magnetic resonance imaging offers tissue characterization that is highly effective in differentiating thrombus from neoplasm. Thrombi, as opposed to neoplasms, lack a vascular supply and, using the late gadolinium enhancement technique, do not enhance.

There are no established management guidelines for the treatment of an MVSA in the absence of significant related complications or concomitant cardiac surgery. If a thrombus is identified within the aneurysm, initial management is focused on treatment of the thrombus with anticoagulation and the consideration of surgical intervention. Percutaneous interventions with coiling and/or placement of occluder devices to exclude left ventricular aneurysms in other locations of the left ventricle have been reported. The location of this patient’s aneurysm would make percutaneous closure challenging due to proximity to the aortic valve and the wide base of the aneurysm.

Video 1: Two-dimensional TTE, modified parasternal long-axis view showing the MVSA with flow seen by color Doppler into the aneurysm. There was no evidence of a VSD by color Doppler.

Video 2: Two-dimensional TTE, modified parasternal short-axis view at the level of the aorta demonstrating the aneurysm adjacent to the aortic valve and protruding into the right ventricle.

Video 3: Two-dimensional TTE, apical five-chamber view of the aneurysm seen at the MVSA.

Video 4: Two-dimensional TTE conventional apical four-chamber view with UEA. Large MVSA seen protruding into the RVOT. No thrombus is appreciated on this view.

Video 5: Two-dimensional TTE, slightly modified apical four-chamber view with UEA. Although not seen on the conventional orientation, a filling defect, suggestive of a thrombus within the aneurysm, is now clearly visualized.

Video 6: Two-dimensional TTE, apical five-chamber view, color flow Doppler showing absence of aortic regurgitation.

View the video content online at www.cvcasejournal.com.

Figure 1 Two-dimensional TTE, modified parasternal long-axis orientation in late-diastolic phase demonstrates the large MVSA (arrow). AV, Aortic valve; LA, left atrium; LV, left ventricle; RV, right ventricle.

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CONCLUSION
Membranous ventricular septum aneurysms are rare congenital anomalies that may lead to blood stasis and thrombus formation. Color Doppler echocardiography is useful in confirming or excluding the presence of concomitant VSD. When evaluating a patient for a cardiac source of embolus, thorough evaluation of ventricular aneurysms using color Doppler and an UEA is recommended to exclude or confirm the presence of a thrombus. Supplemental imaging with CCT and cardiovascular magnetic resonance imaging may also be useful in certain cases.

Figure 2 Two-dimensional basal parasternal short-axis view, pulsed-wave Doppler of the RVOT.

Figure 3 Two-dimensional TTE, apical five-chamber view, diastolic phase without (A) and with (B) an UEA demonstrates the large MVSA (*) bulging into the RV. A filling defect is seen inside the MVSA (arrow) consistent with a suspected thrombus (B). LA, Left atrium; LV, left ventricle; RV, right ventricle. The arrow indicates the thrombus.
Figure 4 Contrast-enhanced CCT scan, multiplanar reconstruction, coronal display (A), and maximal intensity projection with increased slice thickness, oblique sagittal short-axis display (B). The filling defect (thrombus) is well visualized (arrows) and can be measured at 0.8 × 1.2 cm. Ao, Ascending aorta; AV, aortic valve; LV, left ventricle; PV, pulmonic valve. The arrow indicates the thrombus.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2022.01.010.

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