Multimodal Imaging Characterization of a Congenital Multilobular Interventricular Membranous Septal Aneurysm in a Patient Presenting with Embolic Stroke

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INTRODUCTION

An interventricular membranous septal aneurysm (IVMSA) is a rare cardiac defect, with stroke being an even more rare complication of IVMSA.[1,2] This report highlights an extremely unique case of recurrent strokes presumed secondary to IVMSA. Multidisciplinary care is necessary in the management of patients with IVMSA presenting with complications.

CASE PRESENTATION

A 71-year-old woman presented as a transfer from an outside hospital with numbness and weakness in her right hand along with difficulty finding words. She was diagnosed with a distal left middle cerebral artery occlusion with resolution of symptoms following thrombolytic therapy. On examination, she was hypertensive with a blood pressure of 162/99 mm Hg but with otherwise normal vital signs. With the exception of her presenting symptoms, her physical examination was unremarkable. Her medical history included breast cancer, for which she was on maintenance tamoxifen therapy, hypertension, osteoporosis, and a remote "childhood murmur." The differential diagnosis of her stroke included atherosclerosis, small vessel occlusion, a hypercoagulable state (given her history of cancer and maintenance tamoxifen therapy), and cardioembolism.

Magnetic resonance imaging (MRI) of the head revealed acute cortical infarction and white matter disease consistent with chronic small-vessel ischemic changes. It was additionally concerning for multiple past strokes in varied vascular territories. Laboratory evaluation was remarkable for an elevated factor V activity. As a part of the stroke workup, initial transthoracic echocardiogram (TTE) with bubble study was remarkable for a mobile mass noted in the subvalvular apparatus of the tricuspid valve, with no evidence of a patent foramen ovale (PFO) or atrial shunt (Figure 1A; Videos 1-3). The left ventricular ejection fraction was normal at 50%-55%; however, moderate tricuspid regurgitation (by visual estimation) was noted.

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A repeat TTE with additional tricuspid views noted a large IVMSA associated with the septal leaflet of the tricuspid valve (Figure 1B).

Cardiovascular MRI (CMR) was notable for a complex lobular outpouching projecting into the right-sided cardiac chambers, which was thought to be an IVMSA, with no evidence of a shunt, mass, or thrombus (Figure 2A and 2B; Videos 4 and 5). Additional CMR findings included left ventricular ejection fraction, 54%; left ventricular end-diastolic volume, 104 mL; left ventricular end systolic volume, 48 mL; right ventricular ejection fraction, 59%; right ventricular end-systolic volume, 32 mL; and right ventricular end-diastolic volume, 78 mL.

In this patient's case, abnormal development of the membranous septum led to a nonfenestrated aneurysm, causing apical displacement of the anterior portion of the septal leaflet. While the tricuspid valve septal leaflet was displaced >0.8 mm/m^2, meeting the two-dimensional TTE criteria for Ebstein anomaly, this was thought to be a likely by-product of the changes associated with the congenital IVMSA (Figure 2B and Video 5). The lack of a shunt was confirmed by CMR with a Qp:Qs of 1, and there was no color Doppler consistent with a shunt. Additionally, the agitated saline study from the aforementioned echocardiogram did not show a negative contrast jet consistent with a ventricular septal defect (VSD; Video 3).

Subsequent transesophageal echocardiogram further characterized the multilobulated subaortic valvular aneurysms (Figure 3). The largest aneurysms measured 10 mm in diameter with an ostium of approximately 11 mm (Video 6).

Additional evaluation with cardiac CT (CCT) allowed for three-dimensional reconstruction and characterization of the aneurysm to allow for possible surgical planning. The CCT showed a complex lobular outpouching from the left ventricular outflow tract (LVOT) consistent with an IVMSA (Figure 4) that measured 3.0 $\times$ 1.5 cm in the coronal oblique plane and 1.4 $\times$ 1.6 cm in the axial plane. Its communication with the LVOT was noted to measure 1.4 $\times$ 1.1 cm.

Despite extensive collateral testing including blood cultures and telemetry monitoring, no other etiology of emboli was identified.

Although our patient was on maintenance tamoxifen and had elevated factor V activity, an embolic etiology of stroke was favored due to evidence of multiple strokes in varied vascular territories on brain MRI. A comprehensive multidisciplinary conversation among cardiology, cardiothoracic surgery, and adult congenital cardiology yielded a consensus that a thrombus originating in the IVMSA was the likely etiology of the patient's embolic strokes.

In our patient, imaging did not show a VSD, and the risks of percutaneous or surgical closure were deemed to outweigh the benefits. Additionally, as no thrombus was visualized, the identification of the aneurysm as the source of embolization could not be definitively proven.

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, basal short-axis view showing the relationship of the IVMSA (*arrow*) with the LVOT and right ventricular outflow tract.

Video 2: Two-dimensional TTE superior sweep from an apical four-chamber to an apical five-chamber showing the relationship of the IVMSA (*arrow*) with the LVOT and right ventricular outflow tract.

Video 3: Two-dimensional TTE, zoomed apical five-chamber view with color Doppler highlighting the absence of a shunt or valve regurgitation.

Video 4: CMR basal SAX SSFP cine image highlighting the multiple lobes of the IVMSA (*arrow*).

Video 5: CMR LAX SSFP cine images, four- and five-chamber views, highlighting the multiple lobes of the IVMSA (*arrow*).

Video 6: Two-dimensional transesophageal echocardiogram, midesophageal 127° view showing the IVMSA (*arrow*) at the LVOT.

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

Based on the available information, the interdisciplinary team decided on an anticoagulation strategy with warfarin over surgical intervention. The patient was discharged home with an international normalized ratio goal of 2-3. An outpatient implantable loop recorder showed no evidence of atrial fibrillation or other arrhythmias. Later, given continuing difficulty with international normalized ratio maintenance despite medication and diet compliance, the patient was transitioned to apixaban. The patient underwent a repeat TTE 6 months later, which was consistent with previous imaging, with plans to continue serial monitoring every 9-12 months. Since then, the patient has had no recurrent events.

DISCUSSION

The interventricular septum of the heart can be divided into the following portions: membranous, inlet, infundibular, and muscular. The midseptum is termed the membranous septum and comprises both the upper and posterior portion of the septum, thereby separating the aortic passage from the lower right atrium and upper right ventricle. This membranous portion notably lacks myocardium, which, in turn, predisposes the structure to the development of IVMSA upon exposure to a high-pressure gradient.[1]

While the exact incidence of IVMSA is unclear, reports suggest that it occurs in approximately 0.3% of patients with congenital heart disease.[1,3] IVMSA is significantly less common than an atrial septal aneurysm, which has been well documented in the literature to be independently associated with recurrent stroke, especially in patients with a PFO.[4,5] Potential etiologies of IVMSA include spontaneous formation, closure of a VSD, and previous infections or trauma. It is most likely that this patient had a membranous VSD as a child (given the clinical history of a murmur) and that this spontaneously closed via an aneurysmal pouch from the tricuspid valve tissue, which, although rarely this complex, is not an unusual finding. Complications of IVMSA include aortic valve incompetence, subpulmonic stenosis, arrhythmias, endocarditis, and thromboembolism, likely due to abnormal ventricular wall motion causing turbulent flow.[1,3]

Of note, IVMSA is just one of many sources of cardioembolic stroke, the differential diagnosis of which is incredibly broad. Between 14% and 30% of strokes can be attributed to a cardiac source, with cardioembolic strokes being more common as a patient's age increases.[6] Common causes include arrhythmias (such as atrial fibrillation and atrial flutter), mechanical valves, dilated cardiomyopathy, infective endocarditis, thrombus (commonly either a left ventricular thrombus or thrombu associated with a PFO), dilated cardiomyopathy, myxoma, and rheumatic cardiac disease.[7] Cardioembolic strokes are associated with a sudden onset of symptoms (typically less than 5 minutes), altered consciousness, Wernicke's aphasia, and occurrence of both systemic and cerebrovascular emboli.[6,7] It is crucial to consider a wide variety of differential diagnoses as, of the many stroke subtypes, cardioembolic sources of stroke are associated with the highest in-hospital mortality.[6]

For patients presenting with a stroke with concern for a thrombus within an IVMSA, serial echocardiograms have been recommended to monitor for growth of the aneurysm and development of a thrombus. Surgical intervention is generally only indicated when concurrent cardiac disease, hemodynamic changes, or other complications are noted. Specifically, for patients without evidence of a thrombus on echocardiography, surgical correction is only warranted

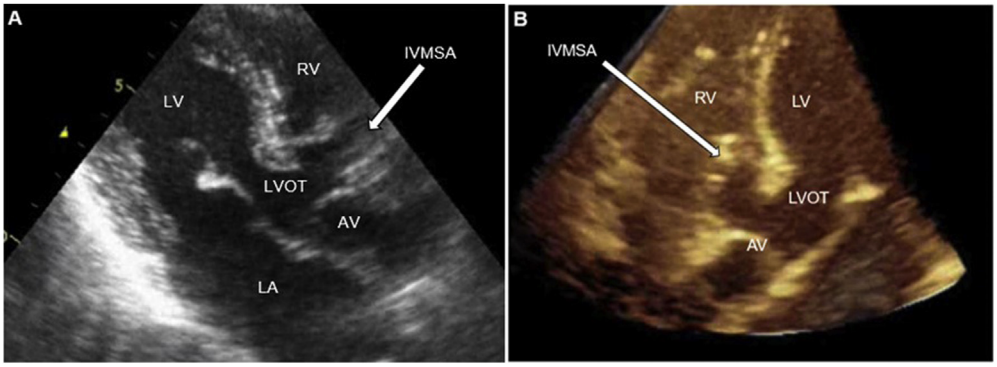

Figure 1 **(A)** Two-dimensional TTE, apical three-chamber view in diastole. **(B)** Three-dimensional TTE, apical five-chamber view in diastole. The *arrow* points to the pathologic outpouching from the LVOT later demonstrated to be an IVMSA. *AV*, Aortic valve; *LA*, left atrium; *LV*, left ventricle; *RV*, right ventricle.

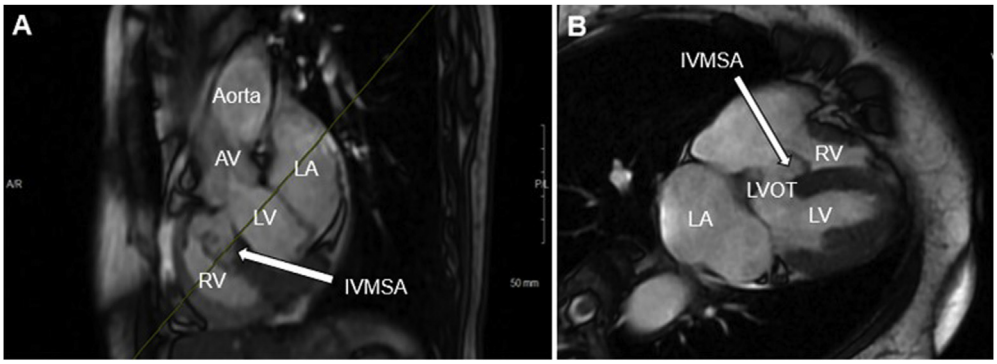

Figure 2 CMR SSFP cine images. **(A)** Basal, sagittal systolic display showing the IVMSA (*arrow*). The *green line* indicates the cut-plane display shown in panel B. **(B)** Oblique axial, systolic display approximating a four-chamber view and highlighting the IVMSA (*arrow*). *AV*, Aortic valve; *LA*, left atrium; *LV*, left ventricle; *RV*, right ventricle.

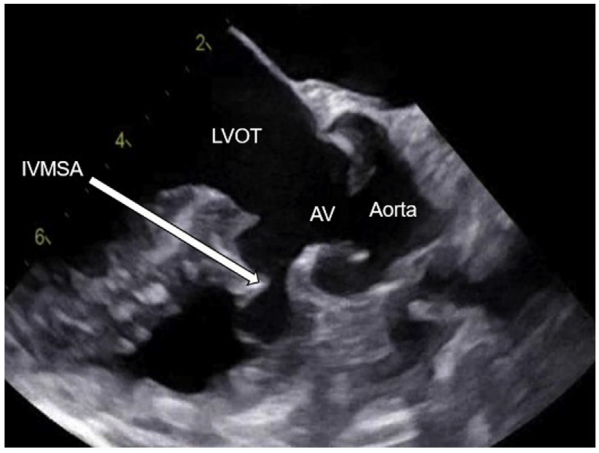

Figure 3 Two-dimensional transesophageal echocardiogram, midesophageal, long-axis view in systole highlighting the two connected saccular structures (~10 mm in maximal diameter) consistent with the multilobular IVMSA, demonstrated by the *arrow*. *AV*, Aortic valve.

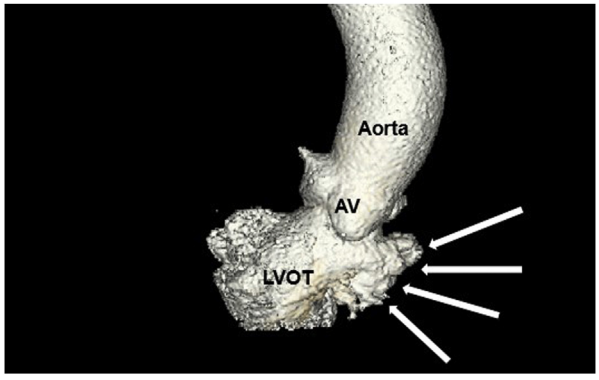

Figure 4 Contrast-enhanced CCT three-dimensional volume-rendered display of the LVOT and aorta demonstrating the multilobular IVMSA (*arrows*). *AV*, Aortic valve.

in those with cerebrovascular emboli despite initiation of anticoagulation. Risks specific to surgical intervention of an IVMSA include residual communication and recurrence, along with distortion of the tricuspid leaflets.[1,3] It is important to note that comprehensive imaging studies should be used to delineate whether the patient has an IVMSA or an aneurysm of the sinus of Valsalva as the surgical indications and further management differ. In our patient, other differential diagnoses that were initially considered included other cardiac aneurysm, diverticula, cyst, or crypt (if in the muscular portion of the ventricle).

It is worth noting that our patient presented with only one symptomatic stroke but had multiple infarcts on MRI. Per the discussions at our institution, there may be a utility for screening MRIs in similar patients to assess for additional cerebral infarcts while on anticoagulation, which would then assuredly qualify a patient for surgical intervention.

CONCLUSION

A heightened awareness of IVMSA among the cardiovascular community can prevent underdiagnosis of this entity and thereby prevent associated complications such as strokes. Multidisciplinary team care is essential in the management of these patients to minimize complications and develop an individualized treatment strategy. Due to there being only a few reported cases, there is no consensus on treatment for recurrent strokes in a patient with underlying IVMSA, and both surgical repair and anticoagulation can be considered. Routine cardiac surveillance for expansion and thrombus formation as well as periodic neurologic imaging in asymptomatic patients may help determine anticoagulation failure. Further research may help delineate appropriate management and anticoagulation regimens for patients with strokes secondary to IVMSA.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2022.01.005.

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