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CASE REPORT
Cardiovascular imaging

A case report of successful transaneurysmal repair of a giant isolated membranous ventricular septal aneurysm

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Background Isolated membranous ventricular septal aneurysms are infrequent in clinical practice. Furthermore, current guidelines do not dictate how to diagnose or manage such lesions.

Case summary A 54-year-old male patient with a history of essential hypertension and tobacco use presented with chest pain associated with dyspnoea and nausea. Electrocardiogram was unrevealing. Physical exam was significant for a diastolic murmur heard best in the apex. Computed tomography angiography of the chest revealed an aneurysm measuring 5 cm in diameter along the ascending aorta. Transoesophageal echocardiography showed that the aneurysm originated from the membranous ventricular septum, coursed along the ascending aorta, and ended anteriorly to the surface of the right ventricle and ascending aorta. Cardiac magnetic resonance imaging confirmed these findings and demonstrated that the aneurysm comprised of two loculations. Given the size of the aneurysm and its proximal location to major cardiovascular structures, percutaneous repair was considered unsafe. Following a multidisciplinary meeting, the lesion was successfully resected via a transaneurysmal approach.

Discussion Isolated membranous ventricular septal aneurysms are best imaged via a combination of transoesophageal echocardiogram and cardiac magnetic resonance imaging and best managed via a multidisciplinary approach for optimal outcomes.

Keywords Cardiac magnetic resonance imaging • Case report • Membranous ventricular septal aneurysm • Multidisciplinary approach • Multimodality imaging • Transoesophageal echocardiogram

ESC curriculum 2.1 Imaging modalities • 2.3 Cardiac magnetic resonance • 2.2 Echocardiography • 7.5 Cardiac surgery

Learning Points

- Isolated membranous ventricular septal aneurysms are uncommon in clinical practice.
- A multimodality imaging approach using transoesophageal echocardiogram and cardiac magnetic resonance imaging is key for an accurate diagnosis of an isolated membranous ventricular septal aneurysm.
- Once a diagnosis of an isolated membranous ventricular septal aneurysm is made, a multidisciplinary approach should be pursued for optimal outcomes.

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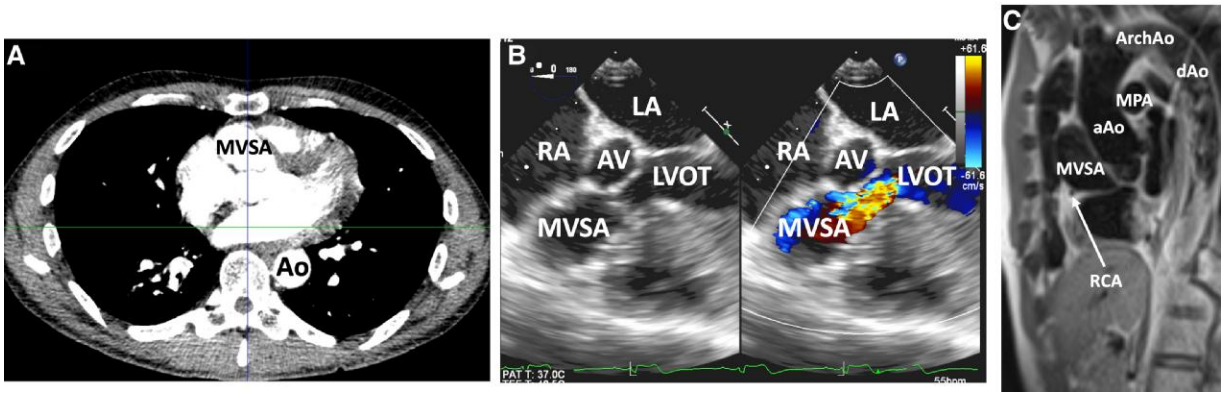
Introduction

Membranous ventricular septal aneurysms (MVSAs) often arise secondary to congenital heart disease, such as ventricular septal defect (VSD) or transposition of the great arteries, but they can also occur following endocarditis or myocardial infarction.[1–3] Isolated MVSAs are an even rarer phenomenon. In general, MVSAs are asymptomatic but have shown to cause cerebral embolism, cardiac arrhythmias, and right ventricular outflow tract obstruction. They are often hard to diagnose, so multimodality imaging and a multidisciplinary approach are frequently required for optimal outcomes. Here, we report an adult patient with a symptomatic isolated MVSA, which was successfully surgically repaired.

Summary figure

Time point	Event
Day 0	Patient presented with chest pain associated with dyspnoea and nausea.
Day 1	Computed tomography angiography of the chest showed a 5.5 cm aneurysm alongside the ascending aorta.
Day 3	Transoesophageal echocardiogram showed that the aneurysm originated from the membranous ventricular septum, coursed along the ascending aorta, and ended anteriorly to the surface of the right ventricle and ascending aorta.
Day 6	Bilateral heart catheterization showed normal haemodynamics and nonobstructive coronary arteries.
Day 8	Cardiac magnetic resonance imaging confirmed the prior findings and showed that the aneurysm had two loculations.
Day 9	Patient underwent successful transaneurysmal repair of the isolated membranous ventricular septal aneurysm.
Day 13	Patient was safely discharged to home.



Case presentation

A 54-year-old male with a history of essential hypertension and tobacco use presented with a chief complaint of midsternal, non-radiating, pressure-like, and intermittent chest pain associated with dyspnoea and nausea. These symptoms started several hours prior to presentation, and each episode lasted a few seconds.

Physical exam was significant for a diastolic murmur heard best in the apex. Labs were significant for a troponin I of <0.02 ng/mL and B-type natriuretic peptide of 45 pg/mL. Electrocardiogram revealed a normal sinus rhythm.

Due to a concern for aortic dissection, computed tomography angiography of the chest was ordered, and it showed a contrast-filled and saccular-shaped aneurysm measuring 2.8 cm × 2.5 cm × 5 cm located anterior to the aortic root and superior to the junction of the right atrium and right ventricle (*Figure 1*). Similarly, due to a concern for acute coronary syndrome, transthoracic echocardiogram was ordered, and it showed a possible ascending aortic root aneurysm measuring 5.5 cm in diameter. In order to better characterize the aneurysm, a transoesophageal echocardiogram (TEE) was ordered, and it showed a large unruptured aneurysm measuring 3 cm × 3.5 cm × 5 cm originating from the membranous ventricular septum below the junction of the right coronary and non-coronary cusps, coursing to the right of the ascending aorta in the plane between the aorta and right ventricle and right atrium, and ending anteriorly to the surface of the right ventricle and ascending aorta (*Figure 2*). Furthermore, the TEE revealed that the aneurysm filled with blood for a brief period during systole and emptied into the left ventricular outflow tract during diastole. Lastly, the TEE demonstrated that the aneurysm comprised of two loculations, with the smaller one dissecting into the muscular ventricular septum and communicating with the larger one through a narrow orifice. Cardiac magnetic resonance imaging (MRI) confirmed these TEE findings (*Figure 3*). Of note, coronary angiography was performed to evaluate for coronary artery disease, and it showed non-obstructive coronary arteries.

Given the large size of the aneurysm and its proximal location to many essential cardiovascular structures, a multidisciplinary approach involving cardiothoracic surgery and cardiology was undertaken to determine the optimal solution. Based on the complex anatomy, percutaneous repair was considered unsafe, and patient was taken to the operating room for surgical repair under coronary bypass. Briefly, patient underwent median sternotomy, pericardiectomy, and aneurysmal repair via a transaneurysmal approach. The operator noticed extensive adhesions around the entire heart and confirmed that the aneurysm originated from the membranous ventricular septum at the site of the aortic root and extended into the right ventricular outflow tract. In addition, it was observed that the aneurysm pulsated with the right

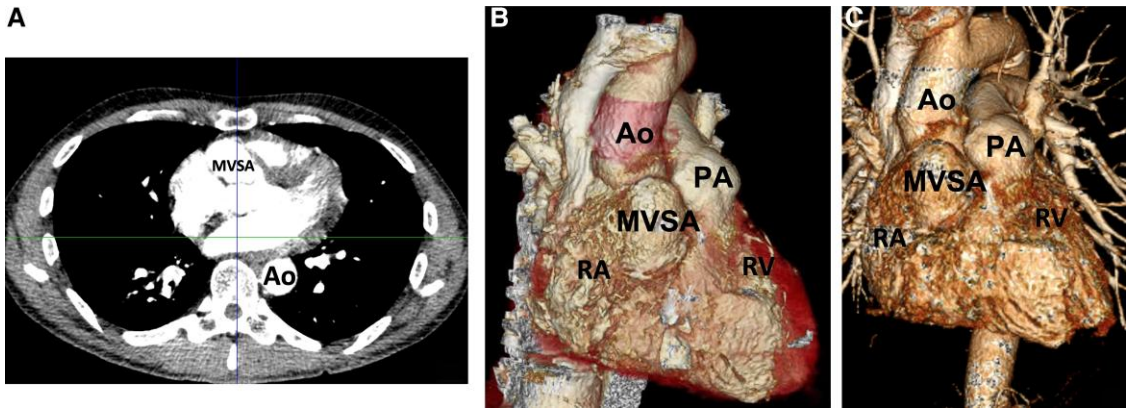


Figure 1 (*A*) Computed tomography angiography of the chest (*B*, *C*) with 3D reconstructions showing a saccular contrast-filled structure measuring 2.8 × 2.5 × 5 cm located just anterior to the root of the aorta and superior to the junction of the right atrium and right ventricle. Ao, aorta; MVSA, membranous ventricular septal aneurysm; PA, pulmonary artery; RA, right atrium; RV, right ventricle.

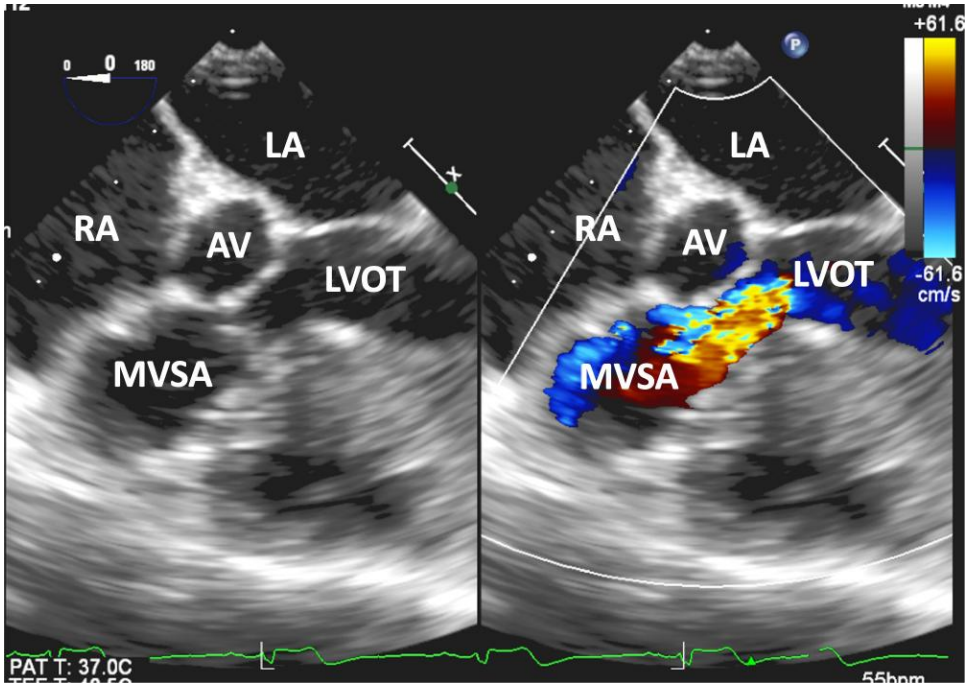


Figure 2 Transoesophageal echocardiogram revealing a giant and unruptured membranous ventricular septal aneurysm with evidence of blood flow into the aneurysm during systole that empties into the left ventricular outflow tract during diastole. AV, aortic valve; LA, left atrium; LVOT, left ventricular outflow tract; MVSA, membranous ventricular septal aneurysm; RA, right atrium.

ventricle. The operation was successful, and it took ~4 h to complete. Patient was transferred to the intensive care unit for overnight monitoring and subsequently discharged to home a few days after.

Discussion

This is, to the best of our knowledge, the first case report to demonstrate successful transaneurysmal repair of a symptomatic isolated MVSA in an adult patient. In addition, it highlights the importance of multimodality imaging and multidisciplinary approach for optimal outcomes in patients with challenging diagnoses.

The literature consists of an ample amount of data on MVSA secondary to VSD, but there are very few publications related to isolated MVSA and none regarding management of such lesions. Assaf *et al.*[4] reported a case of a 42-year-old patient with a 1.3 cm × 1.7 cm isolated MVSA associated with no gross complications who did well with serial echocardiography monitoring. Colangelo *et al.*[5] reported a case of a

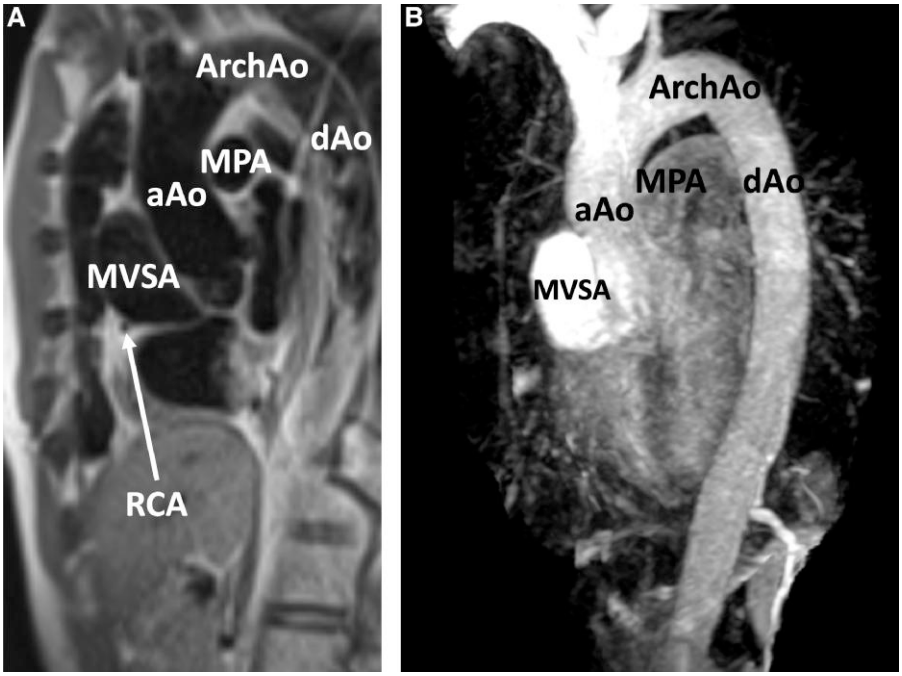


Figure 3 Cardiac magnetic resonance imaging with (*A*) black blood contrast sequencing and (*B*) contrast sequencing in the sagittal view revealing a 3.0 × 3.5 × 6.0 cm unruptured membranous ventricular septal aneurysm located just below the junction between the right and non-coronary cusps and coursing to the right of ascending aorta, initially below the right coronary artery and subsequently above the right coronary artery, and eventually in front of the right ventricle and ascending aorta. aAo, ascending aorta; ArchAo, aortic arch; dAo, descending aorta; RCA, right coronary artery; MPA, main pulmonary artery; MVSA, membranous ventricular septal aneurysm.

51-year-old patient who developed an acute left middle cerebral infarct likely secondary to a thrombus within an isolated MVSA that was ultimately medically managed with initiation of oral anticoagulation and discontinuation of oral contraceptives. Hawa *et al.*[6] reported a case of a 72-year-old patient with a 1.2 × 1.0 cm isolated MVSA who was able to undergo successful transcatheter aortic valve replacement. Abdul Jabbar *et al.*[7] reported three adult patients with isolated MVSA that were conservatively managed. In all of these four instances, the MVSA was managed non-surgically. The MVSA in our patient required treatment given it was causing symptoms likely secondary to the large size obstructing the right ventricular outflow tract. One can appreciate the size of the MVSA in this patient by noting that a retrospective review of cardiac computed tomography scans from 3402 patients revealed eight MVSAs, and the MVSA diameter ranged between 1.0 and 2.2 cm, which is less than half the value of the aneurysm described in this case.[1]

Current guidelines do not clearly discuss the diagnosis and management of isolated MVSA, highlighting the rarity of the condition and lack of expert consensus[8] and need for a multimodality imaging and multidisciplinary approach. Transoesophageal echocardiogram can yield important information about the tortuous course of a given MVSA, whereas cardiac MRI can give important information about the structure of the MVSA. Similarly, involvement of individuals from different specialists can help decide between repair type (percutaneous vs. surgical) and specifics of the repair of choice. In our patient, a transverse aortotomy approach was initially pursued as a means of repairing the MVSA, but it did not yield a clear visualization of the defect, so a transaneurysmal approach was undertaken. Furthermore, after making the decision to undertake a transaneurysmal approach, the question of whether to repair the MVSA from the inside or outside arose, and following consultation with colleagues, the surgeon opted to perform the repair via an outside-to-inside approach due to concerns that an inside-to-outside approach would place the right coronary cusp, left coronary cusp, atrioventricular node, and conduction tissue at risk for damage.

Conclusion

Isolated MVSAs are rare in clinical practice. Accordingly, if initial workup raises suspicion for such, multimodality imaging involving a combination of TEE and cardiac MRI can be utilized to clarify the findings. Once a definitive diagnosis is made, a multidisciplinary approach should be undertaken for optimal outcomes.

Lead author biography

Preetham Kumar is currently working as a Clinical Instructor of Medicine at the University of California, Riverside. He earned his Bachelor of Science in Biomedical Engineering: Premedical from the University of California, Irvine, and his Doctor of Medicine from Virginia Commonwealth University. Upon completion of Internal Medicine Residency at Huntington Hospital, he worked as a clinical research fellow under interventional cardiologist Dr Jonathan Tobis at the University of California, Los Angeles, followed by as a cardiac hospitalist at the University of Washington.

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Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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Data availability

The data underlying this article will be shared upon reasonable request to the corresponding author.

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