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Isolated Ventricular Septal Aneurysm: A Differential Diagnosis for a Right Sinus of Valsalva Aneurysm

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

EF 1 **Andrew Assaf**
EF 2 **Ryan Berry**
CDEF 3 **Yogamaya Mantha**
ABCDEF 1 **Marcel Zughaib**
ABCDEF 1 **Souheil Saba**

1 Department of Cardiology, Ascension Providence Hospital, Southfield, MI, U.S.A.
2 School of Medicine, Michigan State University, Lansing, MI, U.S.A.
3 Department of Internal Medicine, Texas Health Presbyterian Hospital of Dallas, Dallas, TX, U.S.A.

Corresponding Author: Andrew D. Assaf, e-mail: andrewdavis.assaf@gmail.com

Conflict of interest: None declared

Patient: Male, 42-year-old
Final Diagnosis: Ventricular aneurysm
Symptoms: Palpitations
Medication: —
Clinical Procedure: —
Specialty: Cardiology

Objective: Congenital defects/diseases

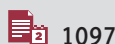
Background: A ventricular septal aneurysm (VSA) is rare and almost always an incidental finding on cardiac imaging. It is rarely an isolated phenomenon and is more commonly associated with other forms of congenital heart disease such as a ventricular septal defect (VSD). Differentiating a ventricular septal aneurysm from an aneurysm of the right sinus of Valsalva is crucial as the latter usually has a more aggressive course and may require surgical intervention. Cardiac computed tomography (cardiac CT) or cardiac magnetic resonance imaging (CMR) may help confirm the diagnosis.

Case Report: We report a case of a 42-year-old obese Japanese man with a past medical history of hyperlipidemia who described occasional effort-related palpitations when climbing stairs over the past few months but no anginal symptoms. Echocardiogram revealed normal left ventricular systolic with a presumed right sinus of Valsalva aneurysm measuring around 1.5 cm. A coronary CTA was obtained to further delineate the aneurysm and revealed normal CT angiographic appearance of a right dominant coronary artery circulation with a small aneurysmal outpouching of the membranous ventricular septum measuring 13×17 mm without any evidence of shunting, along with focal calcification of the medial aspect of the tricuspid annulus. The right sinus of Valsalva appeared normal on coronary CTA.

Conclusions: Membranous ventricular septal aneurysm is a rare condition that is almost always an incidental finding on echocardiography and can be mistaken for an aneurysm of the right sinus of Valsalva. Multimodality imaging and high degree of clinical suspicion are needed to accurately diagnose a ventricular septal aneurysm and to achieve favorable outcomes. A VSA usually has a benign course and is rarely a cause of arrhythmia, right ventricular outflow obstruction, or valvular insufficiency.

Keywords: Heart Septal Defects, Ventricular • Sinus of Valsalva • Ventricular Septum

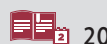
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Background

The interventricular septum (IVS) is a robust wall that separates the right and left ventricle and consists of multiple parts, including the infundibular or outlet part, membranous part, inlet or atrioventricular canal part, and the muscular or trabecular part. A defect or an aneurysm in any of these parts can occur and is termed a ventricular septal defect (VSD) or a ventricular septal aneurysm (VSA), respectively. A ventricular septal defect is the most common congenital cardiac anomaly; it occurs in about 4 of every 1000 births and is the second most common in adults, the first being bicuspid aortic valve in this age group [1-3]. In contrast, a ventricular septal aneurysm, which is an outpouching of the ventricular septum into the right ventricle because of the pressure difference between the right and left ventricles, is very rare and accounts for approximately 0.3% of patients with congenital heart disease, which is roughly 1% of the population [4].

A VSA is rarely isolated and is more commonly associated with other congenital heart defects such as a VSD, but most commonly transposition of the great arteries [5]. The clinical presentation of a VSA is often asymptomatic unless there is another congenital heart defect or a complication. Occasionally, the VSA causes subpulmonic stenosis, which can give rise to right atrial enlargement and right ventricular hypertrophy [6-8].

A VSA is often mistaken for an aneurysm of the right sinus of Valsalva on echocardiography and is often detected incidentally prior to surgery for aortic valve disease. It is best visualized on coronary computed tomography angiography (CCTA) or cardiac magnetic resonance imaging (CMR) [9]. Complications of VSA include, but are not limited to, acquired aortic valve insufficiency due to impingement with subsequent leaflet prolapse, complete atrioventricular (AV) and bundle branch block because of close proximity to the conduction system of the heart, arrhythmias (eg, ventricular tachycardia) and subpulmonic stenosis [10,11]. Thromboembolism and bacterial endocarditis have been reported in patients with VSA [12,13].

Surgical repair is rarely indicated since most patients who have an isolated VSA are asymptomatic, but treatment may be required in a patient with other concomitant congenital heart disease or with a complication arising from a VSA [13,14].

Case Report

We report the case of a 42-year-old obese Japanese man with a past medical history of hyperlipidemia who described occasional palpitations when climbing stairs over the past few months but no anginal symptoms, who was referred to our cardiology clinic for further evaluation after noting a left anterior

hemiblock on his electrocardiogram (ECG). The patient had no other symptoms. He worked as an engineer and smoked 5 cigarettes per day. He had no family history of premature coronary artery disease. Physical exam and laboratory workup were unremarkable. The patient was able to exercise 8 minutes 7 seconds on a standard Bruce protocol and achieved target heart rate without any symptoms or ST-T changes on ECG. Echocardiogram (Figures 1-3) revealed normal left ventricular systolic and diastolic function, normal right ventricular systolic function, mild mitral valve and tricuspid valve regurgitation, and a presumed right sinus of Valsalva aneurysm measuring around 1.5 cm. A coronary CTA (Figures 4-6) was obtained to further delineate the aneurysm and revealed normal CT angiographic appearance of a right dominant coronary artery circulation with a small aneurysmal outpouching of the membranous ventricular septum measuring 13×17 mm without any evidence of shunting, along with focal calcification of the medial aspect of the tricuspid annulus. The right sinus of Valsalva appeared normal on coronary CTA.

Discussion

A membranous ventricular septal aneurysm involving the membranous septum is rarely an isolated phenomenon and is more commonly associated with other forms of congenital heart disease such as a ventricular septal defect [4]. However, a VSA involving the muscular portion of the interventricular septum is likely the result of a myocardial infarction [15]. There are insufficient data in the literature describing the natural history of a membranous ventricular septal aneurysm and potential progression, although, thus far, a membranous VSA is considered to be at a low risk of rupture and is almost always an incidental finding unless associated with other pathologies [15]. It has been suggested that the development of a membranous VSA is probably related to delayed partial or complete closure of a VSD [16]. A membranous ventricular septal aneurysm is presumed to be a true aneurysm involving all layers of the myocardium [20].

A ventricular septal aneurysm is frequently found incidentally on transthoracic or transesophageal echocardiography. Echocardiography will also help in determining the presence of other pathologies that could be associated with a VSA (eg, ventricular septal defect associated with an aneurysm, and right ventricular outflow obstruction with subsequent right atrial and right ventricular enlargement) [17]. In the setting of suboptimal echocardiographic images or significant artifact, cardiac computed tomography or cardiac magnetic resonance imaging can provide better spatial resolution and subsequently confirm the diagnosis of a ventricular septal aneurysm [17].



Figure 1. Parasternal short-axis view at the level of the aortic valve and right ventricular outflow tract (zoomed in view). A trileaflet aortic valve is noted. The arrow indicates the presumed sinus of Valsalva aneurysm involving the right coronary artery, measuring around 1.5 cm, which was confirmed to be a membranous ventricular septal aneurysm, as noted on CTA of the coronary arteries.

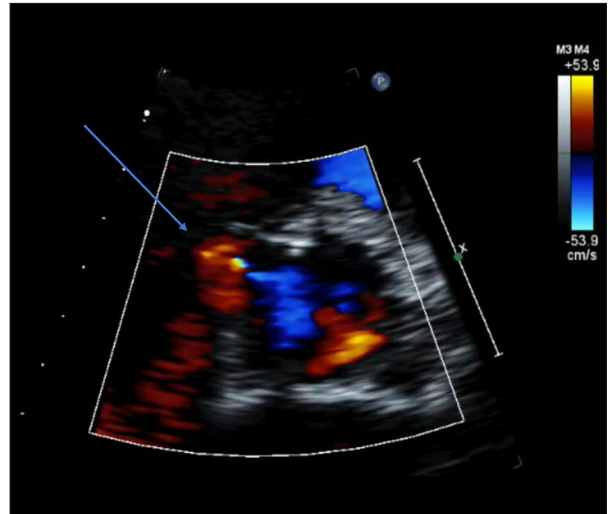


Figure 2. Parasternal short-axis view at the level of the aortic valve and right ventricular outflow tract (zoomed in view). Color Doppler overlying the aortic valve revealing no evidence of shunting. The arrow indicates a presumed sinus of Valsalva aneurysm involving the right coronary artery, measuring around 1.5 cm, which was confirmed to be a membranous ventricular septal aneurysm, as noted on CTA of the coronary arteries.

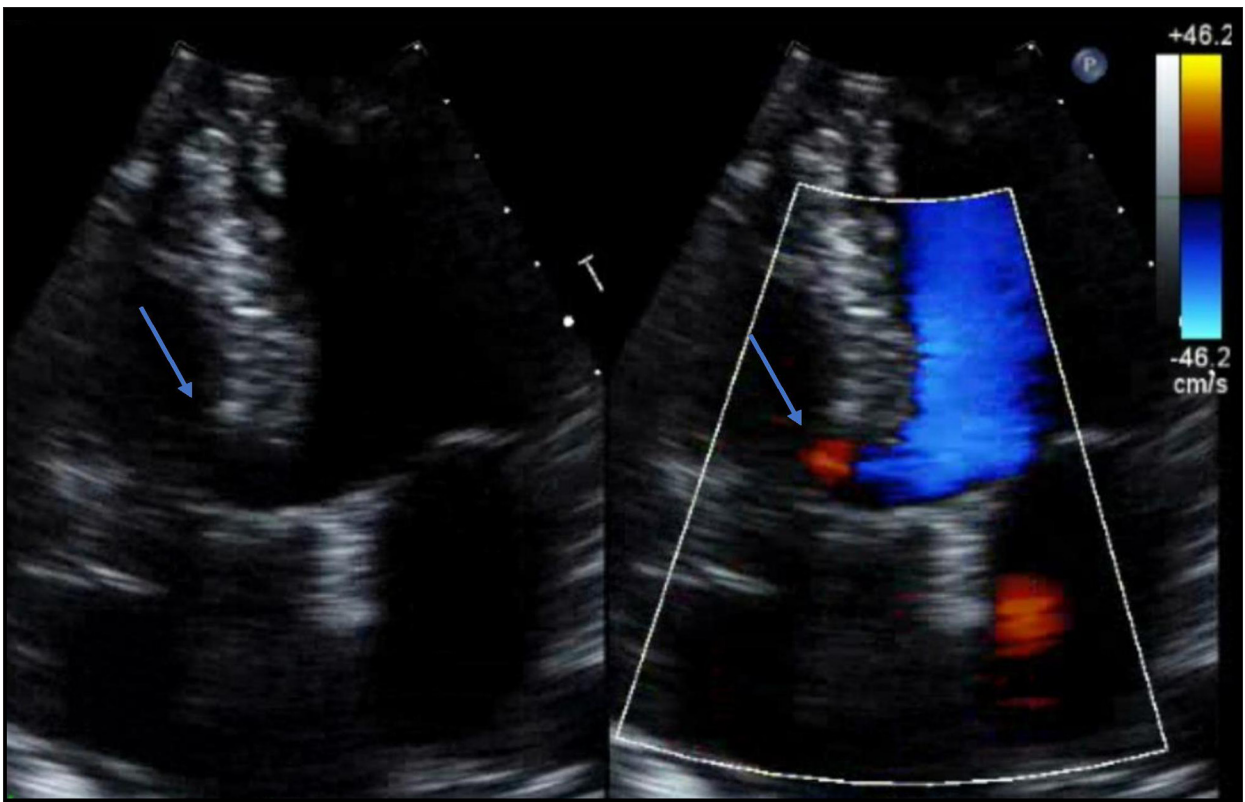


Figure 3. Apical 4-chamber view. Color Doppler overlying the presumed sinus of Valsalva aneurysm revealed no evidence of shunting. The arrow indicates the presumed sinus of Valsalva aneurysm involving the right coronary artery, measuring around 1.5 cm, which was confirmed to be a membranous ventricular septal aneurysm, as noted on CTA of the coronary arteries.

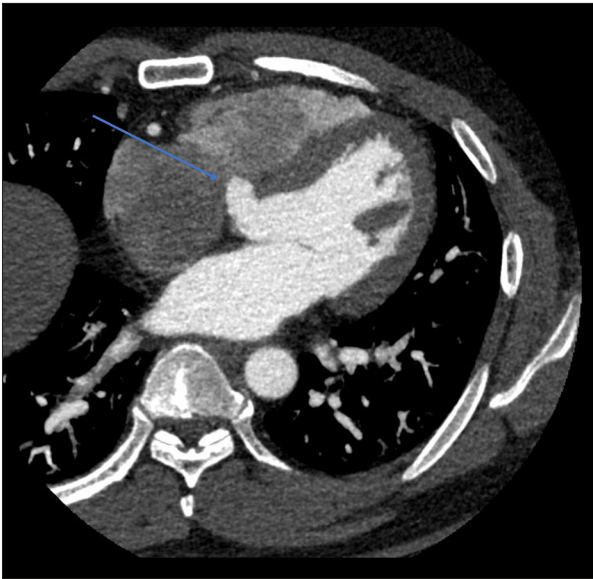


Figure 4. Axial view. 180° left anterior oblique and 90° caudal view. The arrow indicates a membranous ventricular septal aneurysm measuring 13×17 mm, without no evidence of shunting.

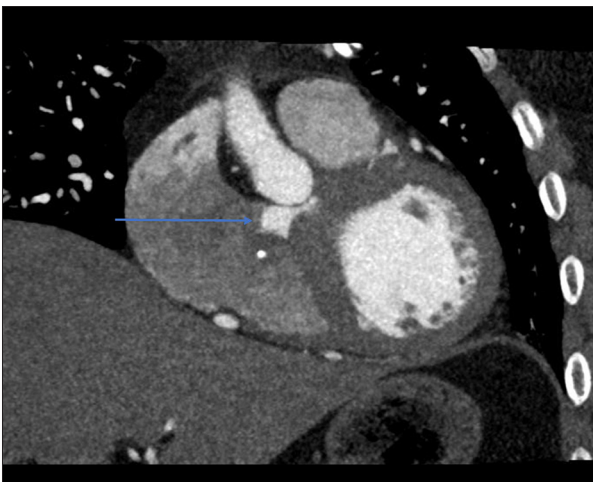


Figure 5. Anterior view. 0° left anterior oblique and 0° caudal view. The arrow indicates a membranous ventricular septal aneurysm measuring 13×17 mm, without no evidence of shunting.

A crucial aspect of cardiac imaging is differentiating a ventricular septal aneurysm from an aneurysm of the sinus of Valsalva because of the more aggressive course of an aneurysm of the sinus of Valsalva, which could potentially require surgical intervention. Adults with a small residual VSD with no other coexisting pathology should be followed up in 3 to 5 years based on the ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease; however, no statement exists with regards to treatment and follow-up of a membranous ventricular septal aneurysm [18].

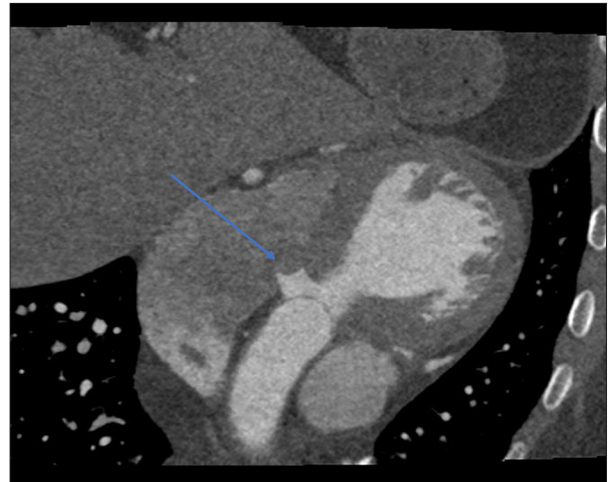


Figure 6. Posterior view. 180° left anterior oblique and 0° caudal view. The arrow indicates a membranous ventricular septal aneurysm measuring 13×17 mm, without no evidence of shunting.

In the rare occasion that a ventricular septal aneurysm requires surgical intervention, it is worth noting that surgical treatment can result in tricuspid regurgitation or even complete heart block because of the proximity of the septal leaflet of the tricuspid valve and the atrioventricular node to the membranous ventricular septum.

Our patient was found to have a 13×17 mm VSA outpouching into the right ventricle without any evidence of shunting, vegetation, subpulmonic stenosis, or associated congenital heart disease. Coronary computed tomography did not reveal any evidence of obstructive epicardial coronary artery disease. Therefore, no intervention was offered, and the patient will be followed with serial echocardiography every 3 to 5 years if he remains asymptomatic.

Conclusions

Membranous ventricular septal aneurysm is a rare condition that is almost always an incidental finding on echocardiography and can be mistaken for an aneurysm of the right sinus of Valsalva. However, in certain circumstances, it can cause arrhythmias or hemodynamically significant complications such as obstruction of the RVOT, VSD, and valvular insufficiency. Multimodality imaging and high degree of clinical suspicion are needed to accurately diagnose VSA and to achieve favorable outcomes.

Conflict of Interest

None.

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