Ruptured Sinus of Valsalva Aneurysm: An Unusual Cause of Heart Failure in a Young Woman

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INTRODUCTION

Sinus of Valsalva aneurysm (SOVA) is an abnormal dilatation of the aortic root between the valve annulus and the sinotubular junction occurring at the level of the elastic lamina.[1] It is a rare cardiac anomaly with an estimated prevalence of 0.09% and accounts for up to 3.5% of congenital cardiac defects.[2] Aneurysm enlargement can lead to disturbance of contiguous structures, and symptoms tend to relate to the chambers involved and severity of valvular regurgitation. Multimodality cardiac imaging is effective for diagnosis and for guiding intervention. Procedural correction is recommended, and long-term outcomes remain favorable.

CASE PRESENTATION

A 22-year-old woman with a medical history of a nonspecific autoimmune arthritis presented with 2 weeks of progressive shortness of breath, dry cough, and abdominal distention. Three months prior to admission, during the naissance of the COVID-19 pandemic in Europe, she returned to New York from a trip to London. She subsequently developed cough and shortness of breath, which persisted for 1 week despite daily use of a fluticasone inhaler prescribed by her primary care physician. The patient tested negative for COVID-19 by nasal swab polymerase chain reaction, but her chest x-ray was consistent with a viral pneumonia. She was diagnosed with presumed COVID-19 infection and treated with supportive measures. The patient's symptoms gradually improved until she developed progressive lower extremity edema, abdominal distention, and exertional dyspnea 1 month later. She presented to her primary care physician, who noted a new murmur and referred her to the emergency department for further evaluation.

In the emergency department, the patient was tachycardic to 113 beats per minute and normotensive and had an oxygen saturation of 97% on ambient air. Her cardiac exam was notable for a jugular venous pressure of 11 cm H_2O, a normal first heart sound and pronounced second sound, a parasternal heave, and a III/VI continuous murmur at the left and right sternal borders and through the apex of the heart. She additionally had decreased bibasilar breath sounds, a distended abdomen, and trace pitting lower extremity edema. Initial laboratory data were remarkable for elevated transaminases, including an aspartate aminotransferase of 46 U/L and alanine aminotransferase of 74 U/L, an elevated B-type natriuretic peptide of 686 pg/mL, and a positive D-dimer of 900 ng/mL. Chest computed tomography (CT) with intravenous contrast demonstrated bilateral pleural effusions without pulmonary emboli. Abdominopelvic CT revealed hepatomegaly with moderate ascites. She was admitted to the general medicine service for further evaluation and cardiology consultation.

Given the concern for new-onset heart failure in a young patient, a broad differential was entertained, including postviral/post-COVID myocarditis, endocarditis, chronic thromboembolic pulmonary hypertension, or a cardiomyopathy associated with her undefined autoimmune condition. Since her cardiac murmur was new, acute valvular pathology or intracardiac shunt was also considered.

Continuous murmurs are generated by flow occurring throughout the cardiac cycle. The differential diagnosis is broad and includes both physiological and pathological etiologies that can be grouped according to their mechanism, namely, rapid blood flow, high- to low-pressure shunt, or localized arterial stenosis.[1] Rapid blood flow due to a venous hum, mammary souffle, and hyperthyroidism can each produce a continuous murmur. Next, any shunting of blood from a higher to lower resistance circulation will also result in a continuous murmur. Systemic artery to pulmonary artery (e.g., patent ductus arteriosus, anomalous left coronary artery, etc.), systemic artery to right heart (e.g., ruptured SOVA, coronary artery fistula), left to right atrial (e.g., atrial septal defect with or without the presence of mitral atresia or stenosis), venovenous (e.g., anomalous pulmonary veins), or arteriovenous shunts will also produce continuous murmurs. Lastly, localized arterial stenosis may lead to similar auscultative findings; flow through large collateral vessels in the presence of aortic coarctation and branch pulmonary stenosis are two examples.[2] The differential diagnosis in this case could more smoothly be narrowed because the murmur was most pronounced at the precordium. The patient's new murmur raised the concern for an acute issue such as ventricular septal defect, atrial septal defect with elevated left-sided pressures, ruptured SOVA, or development of symptoms associated with congenital shunts such as coronary arteriovenous fistula or anomalous left coronary artery.

Transthoracic echocardiography (TTE) revealed mildly reduced global left ventricular systolic function with biplane ejection fraction of 51%, a normal-size inferior vena cava without respirophasic variation, and a small mobile echo density on the septal leaflet of the tricuspid valve best visualized as discontinuity at the noncoronary sinus in the parasternal short-axis view at the level of the aortic valve and right ventricular outflow tract (Figure 1A, Video 1). Color Doppler over the discontinuity revealed flow through the evident tissue defect

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VIDEO HIGHLIGHTS

Video 1: TTE in the parasternal short-axis view at the level of the aortic valve and right ventricular outflow tract demonstrating discontinuity at the noncoronary sinus.

Video 2: TTE in the parasternal short-axis view at the level of the aortic valve and right ventricular outflow tract with color Doppler demonstrating discontinuity at the noncoronary sinus.

Video 3: TEE in the midesophageal 30° short-axis view of the aortic valve demonstrating large noncoronary SOVA protruding into the RA.

Video 4: TEE in the midesophageal 30° short-axis view of the aortic valve with color Doppler demonstrating rupture and resultant left-to-right shunt.

Video 5: TEE in the midesophageal four-chamber view showing the SOVA abutting the tricuspid valve.

Video 6: TEE in the midesophageal four-chamber view with color Doppler showing the SOVA abutting the tricuspid valve.

Video 7: Aortic angiogram revealing the ruptured SOVA and significant left-to-right shunting.

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(Figure 1B, Video 2). To further elucidate the patient's anatomy, transesophageal echocardiography (TEE) was pursued. It was notable for a large noncoronary SOVA protruding into the right atrium (RA; Figure 2, Video 3) with color Doppler demonstrating rupture and resultant left-to-right shunt (Figure 3, Video 4). The aneurysm was seen abutting the tricuspid valve, although the leaflets themselves were normal (Videos 5 and 6). Diastolic flow reversal in the aorta suggested the degree of shunting to be severe. Cardiovascular magnetic resonance imaging (CMR) was also performed, confirming the presence of a high-velocity left-to-right shunt near the aortic valve with flow into the RA and excluding any concomitant congenital heart lesions. The cardiothoracic surgery team was consulted, preoperative left heart catheterization was sought to exclude for the presence of coronary disease, and the ruptured aneurysm and significant left-to-right shunting were reimaged on aortic angiography (Figure 4, Video 7).

The following day, the patient underwent urgent repair of the ruptured noncoronary SOVA with a woven Dacron patch and aortic valve repair. Postoperative TEE demonstrated no residual shunt. The patient was extubated on postoperative day 1 and discharged a few days thereafter. A TTE 2 months later revealed normal left ventricular systolic function, and CMR 3 months postoperatively confirmed no evidence of residual shunt; alternate views not shown revealed normal right ventricle size and near resolution of the pleural effusions.

Of note, her family history was pertinent for a maternal grandmother with ascending aorta dilatation. Given this family history, the patient underwent genetic testing for genes associated with thoracic aortic aneurysms and/or dissections, which did not demonstrate any pathologic variants in the genes evaluated.

DISCUSSION

The normal function of the sinuses of Valsalva is to prevent coronary ostia occlusion as the aortic valve opens during systole. Congenital and acquired etiologies for SOVA include connective tissue diseases (e.g., Marfan syndrome, Ehlers-Danlos syndrome), infections (e.g., syphilis, endocarditis, tuberculosis), trauma, chronic atherosclerosis, and cystic medial necrosis.[3] Coexisting cardiac lesions, such as ventricular septal defects, or aortic regurgitation may be present in up to 30%-40% of cases.[4,5] The right coronary sinus is most commonly involved, followed by the noncoronary sinus and the left coronary sinus. Aneurysm enlargement can lead to disturbance of contiguous structures and result in complications such as arrhythmias or heart block, myocardial ischemia, or aortic regurgitation.[3,6] Although most SOVAs remain intact, rupture occurs in about 35% of cases and commonly appears between 20 and 40 years of age.[6] The anatomic positioning of each sinus determines the clinical outcome in the case of SOVA rupture.[7] Rupture of the right and noncoronary sinuses typically results in communication between the aorta and the right-sided chambers, potentially leading to right ventricular overload and failure, as was seen in our patient. Left sinus rupture is usually less significant and allows blood to recirculate into the left atrium or the left ventricular outflow tract.[8-10] Symptoms depend on the acuity and severity of rupture and can include chest pain, dyspnea, palpitations, or hemodynamic collapse. Diagnosis can be made with multimodality imaging. Traditionally, TTE and TEE have been the first-line imaging techniques, but cardiac CT (CCT) and cine CMR have gained recent popularity.[3,7,11,12] Although both CCT and CMR can visualize cardiac anatomy with precision, CMR has the advantage of sparing radiation and providing enhanced contrast between tissue and vasculature. Cardiac catheterization is performed to evaluate coronary anatomy prior to surgery, although this is not without risk of injuring the already abnormal aortic root. For this reason, CCT may be an appropriate alternative in the absence of hemodynamic instability, large body

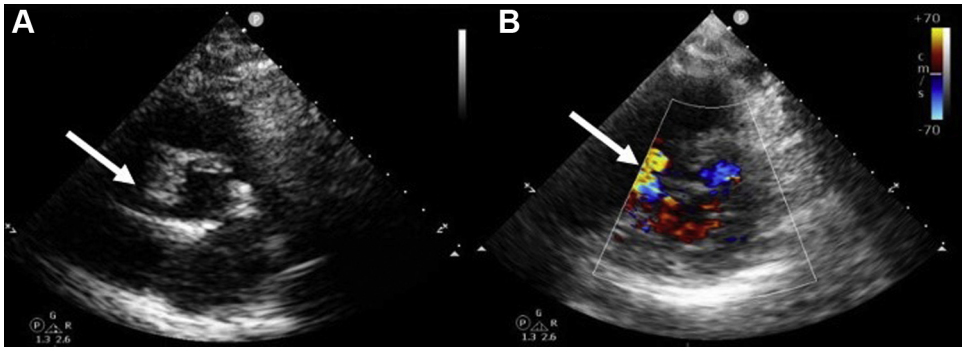

Figure 1 **(A)** TTE in the parasternal short-axis view at the level of the aortic valve and right ventricular outflow tract demonstrating discontinuity at the noncoronary sinus (*arrow*) with **(B)** color Doppler over the defect revealing shunt.

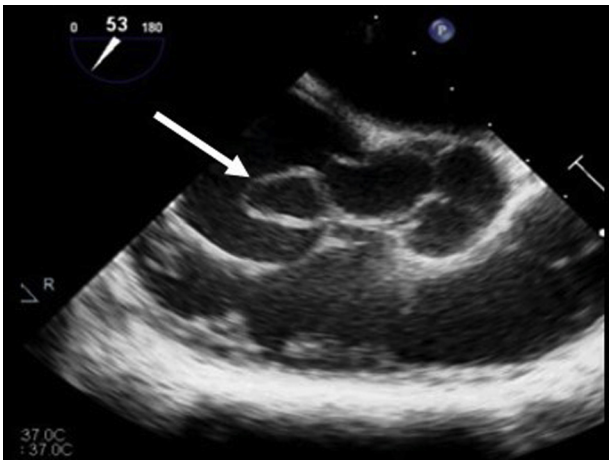

Figure 2 TEE in the midesophageal 30° short-axis view of the aortic valve demonstrating "windsock" deformity of the noncoronary SOVA (*arrow*).

habitus, decompensated heart failure, or inability to tolerate heart rate-slowing medications. Mortality of untreated ruptured SOVA is high, with mean survival of only 4 years without surgery.[13] Surgical management remains the standard of care, with primary closure reserved for small SOVAs and patch closure preferred for larger SOVAs.[7] Outcomes following surgery are typically favorable, as 15-year survival rates may exceed 95%. Alternate therapeutic options include transcatheter closure with septal or ductal occluders. Multiple case series have demonstrated favorable procedural outcomes in these patients as far out as 60 months, although there are no randomized clinical trials that have addressed this.[14,15] In these cases, echocardiography is key in understanding anatomy and recognizing contraindications to percutaneous closure, such as defect size or concomitant structural heart disorders.

CONCLUSION

Clinical presentation with a right heart failure syndrome in the presence of a continuous loud murmur may be a consequence of a ruptured SOVA. Transthoracic echocardiogram, TEE, and CMR are useful for diagnosis. Given the poor prognosis if left untreated, prompt surgical referral is necessary. Outcomes are favorable with surgery, although transcatheter interventions remain a growing alternative option in the appropriate population.

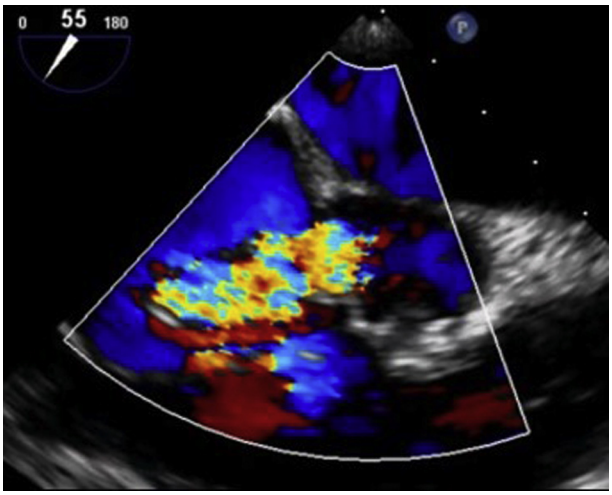

Figure 3 TEE again in the midesophageal 30° short-axis view of the aortic valve color Doppler showing flow between SOVA rupture and RA.

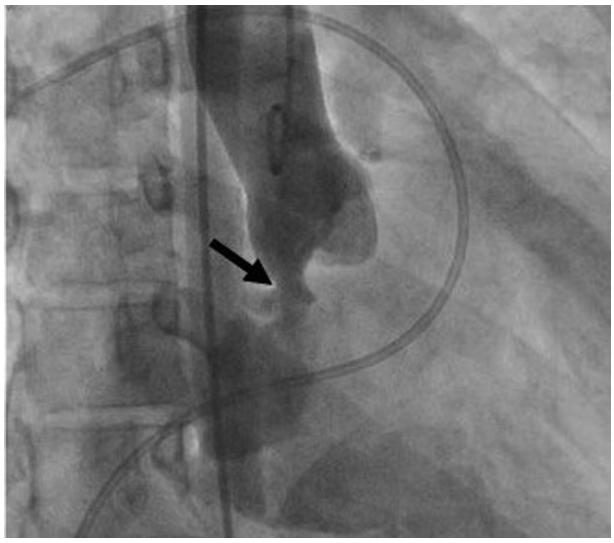

Figure 4 Aortogram showing connection between the noncoronary sinus and RA (*arrow*).

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2021.10.001.

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