

Case in Point

Three Anomalies and a Complication: Ruptured Noncoronary Sinus of Valsalva Aneurysm, Atrial Septal Aneurysm, and Patent Foramen Ovale

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The confluence of atrial septal aneurysm and patent foramen ovale in noncoronary sinus of Valsalva has not been previously documented in the literature.

A 53 year-old white male with a past medical history of hypertension, hyperlipidemia, and former tobacco use was referred to the Dayton VAMC in Ohio for symptoms that included shortness of breath and a recent abnormal stress test. The patient reported no history of known coronary artery disease (CAD), congestive heart failure, or other cardiovascular diseases. The patient also reported no recent fever, bacterial blood infection, syphilis infection, recreational drug use, or chest trauma.

A physical examination was remarkable for grade 3/6 continuous murmur at the 5th interspace to the left of the sternum and a loud “pistol shot” sound heard over the femoral artery. The patient had jugular venous distension and 2+ leg edema bilaterally. His vital signs were normal, and laboratory blood tests showed

normal hemoglobin level and kidney function.

An electrocardiogram showed nonspecific ST segment changes and a transthoracic echocardiogram (TTE) revealed a high-velocity jet in the right atrium (RA) above the tricuspid valve concerning for sinus of Valsalva aneurysm (SVA). A transesophageal echocardiogram (TEE) showed a “windsock” appearance of the noncoronary SVA with possible rupture into the RA (Figure 1) and atrial septal aneurysm (ASA) with more than 2-cm displacement beyond the plane of the atrial septum and a 2-mm patent foramen ovale (PFO) (Figure 2).

Right heart catheterization revealed elevated RA pressures with positive shunt study showing oxygen saturation step-up in the RA (Figure 3). Left heart hemodynamic measurement from an aortic approach to the distal part of the noncoronary cusp SVA revealed an RA pressure-tracing pattern consistent with rupture of the noncoronary SVA into the RA (Figure 4). Coronary angiography re-

vealed single vessel CAD involving the proximal right coronary artery.

The primary diagnosis was of acute heart failure secondary to ruptured aneurysm of the noncoronary SVA into RA. The patient also received a secondary diagnosis of atrial septal aneurysm and PFO.

TREATMENT & OUTCOME

The patient was treated with aggressive diuresis and responded well to therapy. Considering the high mortality rate associated with a ruptured SVA, the patient was referred to a tertiary care center for surgical evaluation. He underwent repair of aorto-right atrial communication with a Cormatrix patch (Roswell, GA) from the aortic side and with primary closure from the right atrial side with resection of the windsock tract; coronary artery bypass graft x1 with right internal mammary artery to the right coronary artery; closure of the PFO with the Cormatrix patch.

The postoperative TEE confirmed preserved LV and RV function, no shunts, no aortic or tricuspid

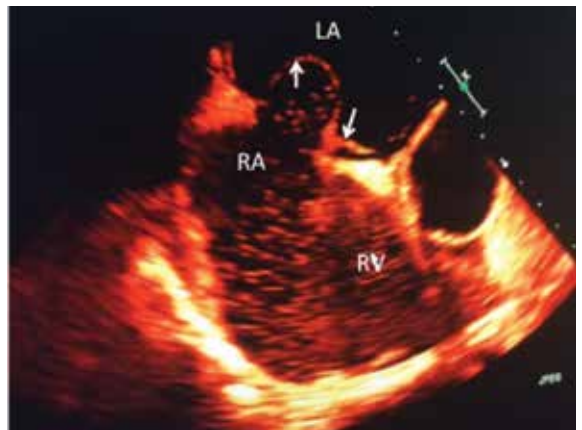
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Figure 1. Transesophageal Echocardiography



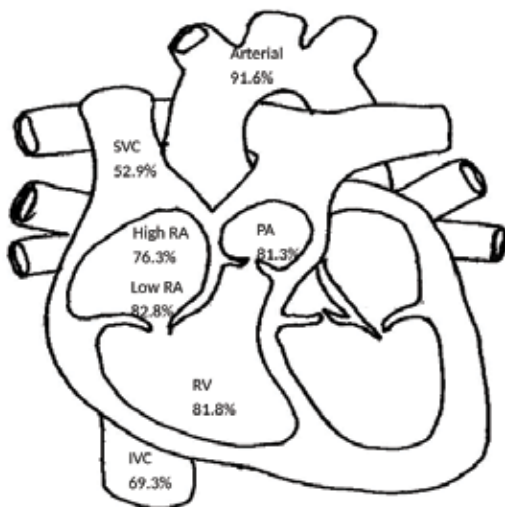
Transesophageal echocardiography demonstrating “windsock” appearance of the noncoronary SVA (arrow) with possible rupture into the right atrium.
Abbreviations: LA, left atrium; RA, right atrium; RV, right ventricle; SVA, sinus of Valsalva aneurysm.

Figure 2. Atrial Septal Aneurysm



Transesophageal echocardiography demonstrating atrial septal aneurysm (upward arrow) protruding into the left atrium; patent foramen ovale also is seen.
Abbreviations: LA, left atrium; RA, right atrium; RV, right ventricle.

Figure 3. Oxygen Saturation Values in Different Intracardiac Locations



Oxygen saturation step-up at the levels of high right atrium and low right atrium confirms left to right shunt.
Abbreviations: IVC, inferior vena cava; PA, pulmonary artery; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

insufficiency. Biopsy of the tissue resected showed intimal fibroplasia. A TTE completed 1 year after surgery showed normal valvular function and without any structural abnormalities. The patient had improvement

in symptoms and an uneventful year after surgical intervention followed by 24 session of cardiac rehabilitation.

DISCUSSION

Sinus of Valsalva aneurysm is a dilation of the aortic wall between the aortic valve and the sinotubular junction that is caused by the lack of continuity between the middle layer of the aortic wall and the aortic valve.¹ Cases of SVA are rare cardiac anomalies with prevalence of 1% in patients undergoing open-heart surgery.² Between 65% and 85% of SVA cases originate from the right coronary sinus, 10% to 20% from the noncoronary sinus, and < 5% from the left coronary sinus.³

Sinus of Valsalva aneurysm is usually congenital, although cases associated with syphilis, bacterial

endocarditis, trauma, Behçet disease, and aortic dissection have been reported. Structural defects associated with congenital SVAs include ventricular septal defect, bicuspid aortic valve, and aortic regurgitation. It is less commonly associated with pulmonary stenosis, coarctation of the aorta, patent ductus arteriosus, tricuspid regurgitation, and atrial septal defects.

The most common complication of the SVA is rupture into another cardiac chamber, frequently the right ventricle (60%) or RA (29%) and less frequently into left atrium (6%), left ventricle (4%), or pericardium (1%).¹ Patients with ruptured SVA mainly develop dyspnea and chest pain, but cough, fatigue, peripheral edema, and continuous murmur have been reported.¹

Atrial septal aneurysm is an uncommon finding in adults, with an incidence of 2.2 % in the general population, and it is often associated with atrial septal defect and PFO.^{1,4} Although ASA formation can be secondary to interatrial differences in pressures, it can be a primary

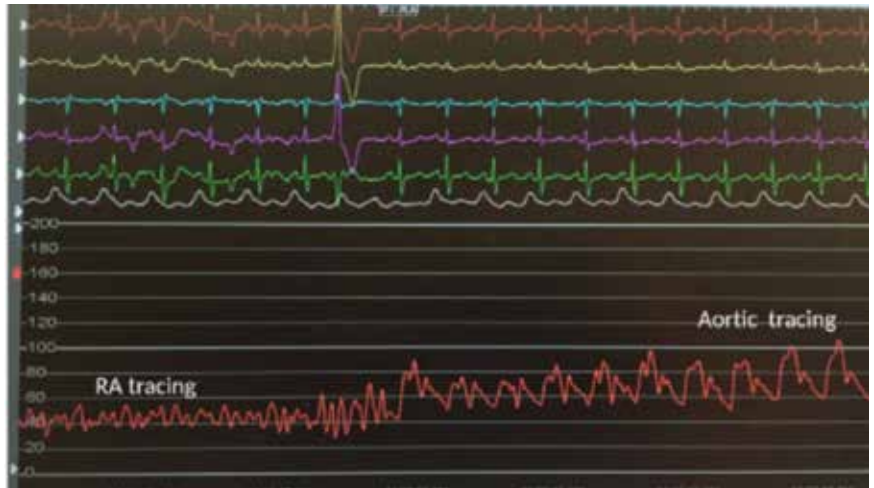
malformation involving the region of the fossa ovalis or the entire atrial septum.⁴ Atrial septal aneurysm may be an isolated anomaly, but often is found in association with other structural cardiac anomalies, including SVA and PFO.^{4,5}

CONCLUSION

Although coexistence of SVA and ASA has been reported previously, the case reported here, a ruptured noncoronary SVA that was associated with a large ASA and a PFO, has not been previously documented in the English literature. This patient's anomalies are most likely congenital in origin. Progressive dyspnea and chest pain in the presence of a continuous loud murmur should raise the suspicion of ruptured sinus of Valsalva. Although no significant aortic regurgitation was noted on echocardiography, the pistol shot sound heard over the femoral artery was believed to be due to the rapid diastolic runoff into the RA through the ruptured SVA.

The significant increase in the RA pressure made the ASA and PFO more prominent. A TEE, left and right heart catheterizations with shunt study are vital for the diagnosis of SVA. If left untreated, SVA has an ominous prognosis. Surgical repair of ruptured SVA has an accepted risk and good prognosis with 10-year survival rate of 90%, whereas the mean survival of untreated ruptured SVA is about 4 years.^{6,7} Hence, the patient in this study was referred to a tertiary care center for surgical intervention. ●

Figure 4. Cardiac Pressure Tracing Measured by Cardiac Catheterization



Pullback from the distal noncoronary sinus of Valsalva to aorta: RA tracing pattern at the distal noncoronary sinus of Valsalva confirmed a shunt between and the sinus and the RA. Abbreviation: RA, right atrium.

Author disclosures

The authors report no actual or potential conflicts of interest with regard to this article.

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