CASE REPORTS

Rare cause of right heart failure: contained rupture of a sinus of Valsalva aneurysm associated intraventricular septal aneurysm

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Abstract Sinus of Valsalva aneurysm (SVA) is a rare congenital lesion described first in 1840 by John Thurnam (Cited by Boutefou JM, Moret PR, Hahn C, Hanf E. Aneurysms of the sinus of Valsalva: report of seven cases and review of the literature. Am J Med 1978;65:18–24). In most cases unruptured Sinus of Valsalva aneurysm (SVA) is clinically silent; however, if it progressively enlarges it may cause coronary artery compression, complete heart block, or right ventricular outflow tract obstruction (Meier JH, Seward JD, Miller FA, Oh J, Sarano ME. Aneurysms in the left ventricular outflow tract: clinical presentation, causes, and echocardiographic features. J Am Soc Echocardiogr 1998;11:729–45; D'Silva SA, Dalve BV, Lokhandwala YY, Kale PA, Tendolkar AG. Unruptured congenital aneurysm of the left sinus of Valsalva presenting as acute right heart failure. Chest 1992;101:578–79) or is a potential source of cerebrovascular emboli. (Shahrabani RM, Jairaj PS. Unruptured aneurysm of the sinus of Valsalva: a potential source of cerebrovascular embolism. Br Heart J 1993;69:266–67). In this report, we describe a case of right coronary sinus of Valsalva aneurysm with a contained rupture. The containing rupture is in intraventricular septal aneurysm; the patient presents with right-sided heart failure.

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KEYWORDS
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Background

In the United States, SVA accounts for only 0.43% of all cardiopulmonary bypass procedures.\(^1,5\)

Rupture of the aneurysm may be a spontaneous occurrence or secondary to trauma, exertion, or iatrogenesis during cardiac catheterization. The rupture may be clinically silent or present with heart block, right ventricular outflow obstruction, acute right heart failure or stroke.\(^2-4\) In addition, it may present as symptoms of dyspnea, chest pain, or a new continuous to-and-fro murmur. Rupture into the right ventricle is most common (60\(\text{--}\)90%) followed by rupture into the right atrium.

Figure 1  A basal short axis view on transesophageal echocardiogram of the aortic sinuses. The right sinus of Valsalva (RSV) is dilated with aneurysm formation. The aortic valve (BoV), left sinus of Valsalva (LSV) and noncoronary sinus are essentially normal. The right ventricular outflow tract (RVOT) is obstructed.

Figure 2  The basal short axis view of the aortic valve (AoV) at the level of the sinuses of Valsalva. Color flow Doppler shows a right ventricular outflow tract (RVOT) obstruction. Trivial aortic regurgitation is noted.
(10%) and left atrium (2–3%) and rarely into the pericardium. If left untreated after symptomatic presentation, most patients die within 1 year. It is thought that the primary defect involves a separation of the media in the sinus of Valsalva from the media adjacent to the aortic annulus. Endocarditis, syphilitic aortitis, cystic medial necrosis (Monckeberg’s disease) or traumas are less common etiologies.

Transthoracic echocardiography usually clinches the diagnosis, transesophageal echocardiography or cardiac magnetic resonance imaging may be necessary on occasion.

Case presentation

A 76-year-old Caucasian male with a 17-year history of hypertension presents with exertional dyspnea, ankle swelling, two-pillow orthopnea, and presyncope when bending forward. No other known cardiac history, two dimensional echocardiogram done 10 years earlier showed mild dilatation of the RSV. He was treated with furosemide and felt symptomatically better and returned four weeks later for cardiac assessment.

Physical examination revealed an elderly gentleman, not distressed, with a heart rate of 78 beats a minute, and blood pressure 180/80, both left and right arms; neck veins were not distended. Heart sounds were normal with an intermittent ejection systolic murmur at the base, graded I/VI. No obvious diastolic murmur was noted. No hepatomegaly. Extremities showed peripheral edema. EKG showed sinus bradycardia with suspicion of an old inferior wall myocardial infarction. Laboratory data showed a BUN of 17, creatinine of 1.7, and normal electrolytes. Two dimensional echocardiogram showed a large 57 mm sinus of Valsalva aneurysm involving the right coronary sinus in the parasternal long axis window with suggestions of right ventricular outflow tract obstruction. The ascending aorta was dilated and the right ventricular systolic pressure recorded at 54 mm of mercury. Mild concentric left ventricular hypertrophy was noted with normal left ventricular systolic function and no regional wall motion abnormalities. No evidence of intracardiac shunt was noted. The patient underwent transesophageal echocardiography that confirmed a large sinus of Valsalva aneurysm and right ventricular outflow tract obstruction (Figs. 1–3) and moderate aortic regurgitation graded 2/4 as a result of annular-aorta dilatation. The proximal and mid-ascending aorta was dilated as well. No ventricular septal defect was noted.

The patient underwent elective aortic valve repair, repair of the sinus of Valsalva and resection of the ventricular septal aneurysm, which was described as “tissue paper thin” and at risk for impending rupture, but no evidence of communication between the ventricles. The patient
came off bypass easily with excellent hemodynamics and transesophageal echo showed only trivial residual aortic valve regurgitation. There were no perioperative complications. Six years postoperatively, the patient was seen in follow-up and doing well.

References