Case Reports

Contained Rupture of Sinus of Valsalva Aneurysm in a 64-Year-Old Man

Anjali Garg, MD
Nivas Balasubramaniyam, MD
Rocco Lafaro, MD
Robert Timmermans, MD
Wilbert S. Aronow, MD
Howard A. Cooper, MD
Julio A. Panza, MD

We report a contained rupture of a right coronary sinus of Valsalva aneurysm, in which repair resulted in symptomatic improvement. Patients often present with symptoms secondary to rupture of the sinus of Valsalva aneurysm into one of the cardiac chambers, or secondary to the compression of adjacent structures. Whereas sinus of Valsalva aneurysms and their rupture are well reported in the literature, contained ruptures have been described only rarely. In those cases, symptoms often arose from compression of adjacent structures.

Although transesophageal echocardiography is considered to be the diagnostic method of choice, cardiac magnetic resonance imaging and computed tomography can be equally helpful in establishing the diagnosis and delineating the lesion. Diagnosis and prompt repair in our 64-year-old patient resulted in the rapid resolution of his symptoms. ***(Tex Heart Inst J 2016;43(5):433-6)***

Key words: *Aortic aneurysm/pathology; aortic valve insufficiency; cardiac magnetic resonance imaging; echocardiography, transesophageal; sinus of Valsalva aneurysm/contained rupture*

From: *Departments of Internal Medicine (Dr. Garg), Cardiology (Drs. Aronow, Balasubramaniyam, Cooper, Panza, and Timmermans), and Cardiothoracic Surgery (Dr. Lafaro), Westchester Medical Center/New York Medical College, Valhalla, New York 10595*

Address for reprints: *Nivas Balasubramaniyam, MD, Department of Cardiology, Westchester Medical Center, 100 Woods Rd., Valhalla, NY 10595*

E-mail: *nivaseras@gmail.com*

Sinus of Valsalva aneurysms (SVA) are rare. They can be congenital or acquired. Congenital SVAs are more prevalent and are caused by weakness at the junction of the aortic media and fibrotic annulus.[1] Diseases affecting the aortic wall—such as infections, trauma, and degenerative diseases—can cause acquired aneurysms.[2] Sinus of Valsalva aneurysms typically occur in the right coronary sinus (in 65%–85% of cases), followed by the noncoronary sinus (10%–30% of cases) and, rarely, by the left coronary sinus (1%–5% of cases).[3] They usually remain asymptomatic[4] and are discovered incidentally upon diagnostic study. Infrequently, they can act as a space-occupying lesion—obstructing the right ventricular outflow tract (RVOT) or the left ventricular outflow tract (LVOT), interfering with aortic valve function, distorting the coronary ostia and thereby leading to ischemia, and even causing conduction disturbances by compressing the conduction system.[5] Whereas SVAs and their rupture are well reported in the literature, contained ruptures are described only rarely.[6,7] Although transesophageal echocardiography (TEE) is considered the SVA diagnostic method of choice,[8] cardiac magnetic resonance and computed tomography (CT) can be equally helpful in establishing the diagnosis and delineating the lesion. Patients with contained rupture present with symptoms arising from the compression of adjacent structures. We describe the case of a patient who most likely had symptoms from LVOT obstruction, aortic insufficiency (AI), or both.

Case Report

In January 2014, a 64-year-old man with a history of hypertension and sarcoidosis presented with increasing dyspnea on exertion. He was taking prednisone and azathioprine for sarcoidosis. He was a former smoker (20 pack-years), but had quit 9 years earlier. He reported no chest discomfort, paroxysmal nocturnal dyspnea, orthopnea, edema, fatigue, dizziness, or palpitations. On physical examination, he was afebrile with a blood pressure of 120/72 mmHg, a heart rate of 90 beats/min, a respiratory rate of 16 breaths/min, and an oxygen saturation of 98% on room air. His body mass index was 29 kg/m^2. He had no jugular venous distention. His lungs were clear to auscultation. His point of maximal impulse, S_1, and S_2 were normal. No heart murmurs were present.

He reported his dyspnea to his physician. A chest radiograph showed diffuse patchy airspace opacities (most likely indicative of infiltrates or pulmonary edema), and me-

diastinal adenopathy accompanied by a questionable subaortic mass. His daily steroid dosage was increased from 10 mg to 20 mg, but his dyspnea persisted. His electrocardiogram revealed sinus rhythm at 70 beats/min, in addition to an old anterolateral myocardial infarction. Routine laboratory test results were within normal limits. A transthoracic echocardiogram (TTE) showed a 24-mm-long × 21-mm-wide, basal anteroseptal mass—heavily calcified, spherical, cystic, and fixed—positioned anterior to and somewhat to the left of the LVOT, and adjacent to the RVOT. The TTE also identified moderate AI and normal left ventricular function.

Cardiac magnetic resonance with and without contrast medium showed a 4.4 × 3.6 × 3.9-cm mass extending into the LVOT with central flow, best seen on magnetic resonance angiography. The mass, immediately adjacent to the left main coronary artery and the left atrial appendage, was suspected to arise from the left sinus of Valsalva (Fig. 1). This partially thrombosed paravalvular aneurysm was associated with mild AI.

Further evaluation of the cardiac mass by CT with contrast solution (Fig. 2) revealed it to be peripherally calcified and nearly completely thrombosed, and to arise from the right coronary cusp. It extended posteriorly, abutted the left coronary cusp, and extended into the LVOT (there was no communication between the outflow tract and the mass). The patient was diagnosed with contained rupture of the right SVA (RSVA), and with AI.

Intraoperatively, the patient was noted to have a perforation of approximately 45 mm in the right sinus, which was probed and found to enter a pseudo-sac. He underwent patch closure of the perforation, together with aortic valve repair by augmentation of the right leaflet with bovine pericardium. An intraoperative TEE revealed mild residual AI. He had an uneventful postoperative course and was discharged from the hospital. His symptoms resolved during the postoperative follow-up evaluation.

Discussion

Sinus of Valsalva aneurysms are relatively rare, with a reported incidence of 0.14% to 0.96% in patients undergoing open-heart surgery.[9] These lesions result from separation or lack of fusion between the media of the aorta and the annulus of the aortic valve. Rupture usually occurs in the 3rd or 4th decade of life. Most often, the aortic–cardiac fistula is between the right coronary cusp and the right ventricle, but occasionally, when the noncoronary cusp is involved, the fistula drains into the right atrium. A left SVA can rupture into the left ventricle or left atrium, and infrequently into the pulmonary trunk or pericardium.[10] Acquired SVA can be associated with endocarditis, atherosclerosis, syphilis, or aortic dissection.[11] Degenerating conditions like cystic medial necrosis and injury from deceleration trauma are also associated with acquired SVA.[12]

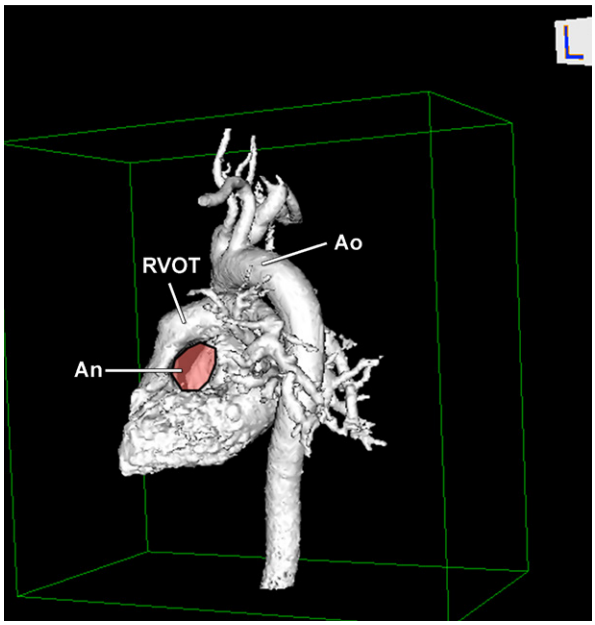

Fig. 1 *Cardiac magnetic resonance image (3-dimensional reconstruction) shows the location of the thrombosed aneurysm (An) at the aortic root, extending to the right ventricular outflow tract (RVOT).*

Ao = aorta

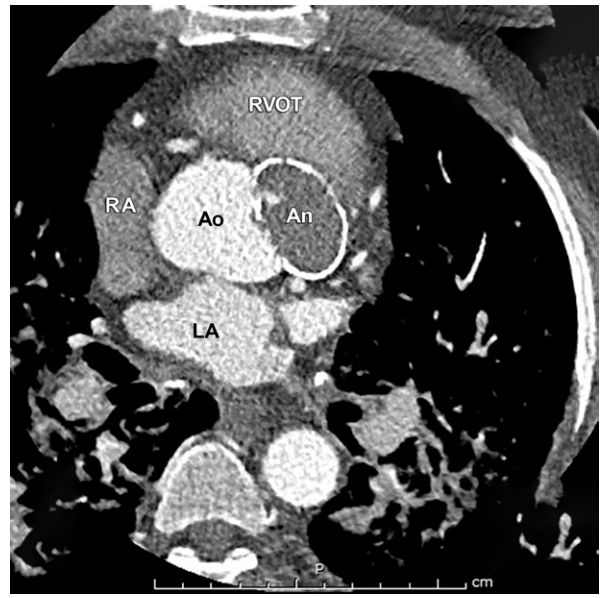

Fig. 2 *Computed tomogram of the heart with contrast medium shows that the 4.5 × 3 × 4.7-cm mass arises from the right coronary cusp, is partially thrombosed and calcified, and is probably a thrombosed pseudoaneurysm of the right coronary cusp.*

An = thrombosed aneurysm; Ao = aorta; LA = left atrium; RA = right atrium; RVOT = right ventricular outflow tract

TABLE I. Classification Systems for Ruptured Sinus of Valsalva Aneurysm[15,16]

Type	Sakakibara Classification	Modified Sakakibara Classification
I	Originating from left part of right coronary sinus (CS): protruding into conus of right ventricle (RV), just beneath commissure of right and left pulmonary valves	Protrusion and rupture into RV just beneath pulmonary valve
II	Originating from central part of right CS; protruding into RV; penetrating crista supraventricularis	Penetration and rupture into or just beneath crista supraventricularis of RV
IIIv	Originating from posterior part of right CS; protruding into RV, just beneath septal leaflet of tricuspid valve; penetrating membranous septum	Penetration and rupture into RV adjacent to or at tricuspid annulus
IIIa	Originating from posterior part of right CS; protruding into right atrium (RA), near commissure of septal and anterior leaflets of tricuspid valve	Penetration and rupture into RA adjacent to or at tricuspid annulus
IV	Originating from right part of noncoronary sinus; protruding into RA, near septal leaflets of tricuspid valve	Protrusion and rupture into RA
V	———	Other rare conditions (for example, rupture into left atrium, pulmonary artery, left ventricle, or other structures)

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Abrupt SVA rupture causes chest pain, a bounding pulse, a continuous murmur in the left sternal border accentuated in diastole, and volume overload of the heart. Although symptoms from an enlarging aneurysm occur most commonly between the ages of 20 and 40 years, presentations in infancy and in the 7th decade of life have been described.[13] Typically, unruptured SVAs are silent and are diagnosed incidentally. Rarely, an unruptured aneurysm causes myocardial ischemia, AI, and conduction disturbances (including complete heart block); and it can be a source of emboli.[14]

In 1962, Sakakibara and Konno[15] proposed the first formal classification system for SVA, according to the sites of origin and rupture (Table I). This anatomic classification system described only 4 types of aneurysms, arising from either the right coronary sinus or the noncoronary sinus, and did not account for all possible chambers of penetration.

In 2013, a modified Sakakibara classification system was proposed,[16] wherein the origin of the lesion is no longer emphasized. Instead, the RSVA is classified in accordance with the anatomic location of the protrusion site. Because most RSVA lesions rupture into the right ventricle or right atrium, a detailed classification was established for RSVAs involving these 2 heart chambers.

The diagnosis of SVA is confirmed by means of 2-dimensional and color-flow Doppler echocardiographic studies. In 75% of patients, TTE in Doppler mode correctly but often suboptimally establishes the diagnosis of ruptured aneurysm, thereby imprecisely delineating the anatomy of the aneurysm and its relationships with associated lesions. To date, TEE has become the gold standard for the diagnosis of this lesion.[8] In color-flow Doppler mode, TEE precisely defines the location, morphology, size, associated lesions, and complications of the defect. Cardiac catheterization quantitates the left-to-right shunt, and thoracic aortography portrays the fistula. Cardiac magnetic resonance has the advantage of providing better soft-tissue contrast, thereby enabling better delineation of components of the mass.[17]

Medical management is directed at managing cardiac failure, arrhythmias, or endocarditis. At surgery, the aneurysm is closed and excised, and the aortic wall is reunited with the heart, either by direct suture with pledgets or with a pericardial (or Dacron) patch or prosthesis. Patients who underwent surgical repair of SVA had an 95% overall survival rate of 20 years,[18] whereas the mean survival rate in patients with untreated RSVA is about 4 years.[19]

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