

Rupture of right coronary sinus of Valsalva aneurysm into right ventricle

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A sinus of Valsalva aneurysm is a rare cardiac anomaly that may be congenital or acquired; a coexisting cardiac lesion might be present. If the aneurysm ruptures, it causes acute symptoms of dyspnoea. Echocardiography and cardiac magnetic resonance imaging are useful for diagnosis. The treatment of choice is surgery. We present a case of a patient with acute onset of symptoms due to a ruptured sinus of Valsalva aneurysm. (Neth Heart J 2010;18:209-11.)

Keywords: Aortic Aneurysm; Aortic Rupture; Echocardiography; Magnetic Resonance Imaging; Sinus of Valsalva; Case Reports

A ruptured sinus of Valsalva aneurysm (SVA) is a rare cardiac abnormality, and is associated with a severe left-to-right shunt if communicating with the right-sided heart chambers. Uncorrected, the rupture almost invariably causes deterioration in heart function. Early surgical intervention is the treatment of choice.

Case report

A 55-year-old man was referred to our hospital because of sudden onset of severe dyspnoea, palpitations, and deterioration in exercise capacity. On

admission, his functional class was New York Heart Association class III. A loud continuous murmur and a sinus tachycardia were present. Transthoracic echocardiography was performed and showed a slightly dilated right ventricle in the presence of a left-to-right shunt from the aorta into the right ventricle. Transoesophageal echocardiography (TEE) revealed a ruptured aneurysm of the right coronary sinus of Valsalva causing a large left-to-right shunt into the right ventricle, diagnosed by colour Doppler. The ventricular septum showed no defect and mild aortic regurgitation was present (figure 1). The rupture into the right ventricular outflow tract was confirmed using cardiac magnetic resonance imaging (figure 2). Cardiac catheterisation was performed and did not reveal any coronary artery disease; the mean pulmonary artery pressure was 32 mmHg and the Qp/Qs was 1.55. Urgent cardiac surgery was performed. Cardiopulmonary bypass was instituted after a median sternotomy and coronary ostial cardioplegia infusion was used. Aortotomy and right atriotomy were performed. The right coronary sinus showed an aneurysm with an opening of 10 mm leading into the right ventricle outflow tract (figure 3). The aneurysmatic tissue was excised and a bovine pericardial patch was used to close the hole entering into the right ventricle. The aortic valve was excised and a mechanical aortic valve (Carbo-medics® 25 mm) was successfully implanted. After the procedure a TEE was performed and showed no residual shunt.

Discussion

SVA is a rare cardiac anomaly, and arises mainly from a congenital defect of the aortic media or may follow bacterial endocarditis. It occurs in between 0.09 and 0.15% of cases, and comprises up to 3.5% of all congenital cardiac anomalies.¹ Coexisting cardiac lesions, especially a ventricular septal defect or aortic valve regurgitation, may both be present in about 30 to 40% of patients.^{2,3} The aneurysms originate predominantly from the right coronary sinus (70%), and are more prevalent in males and people

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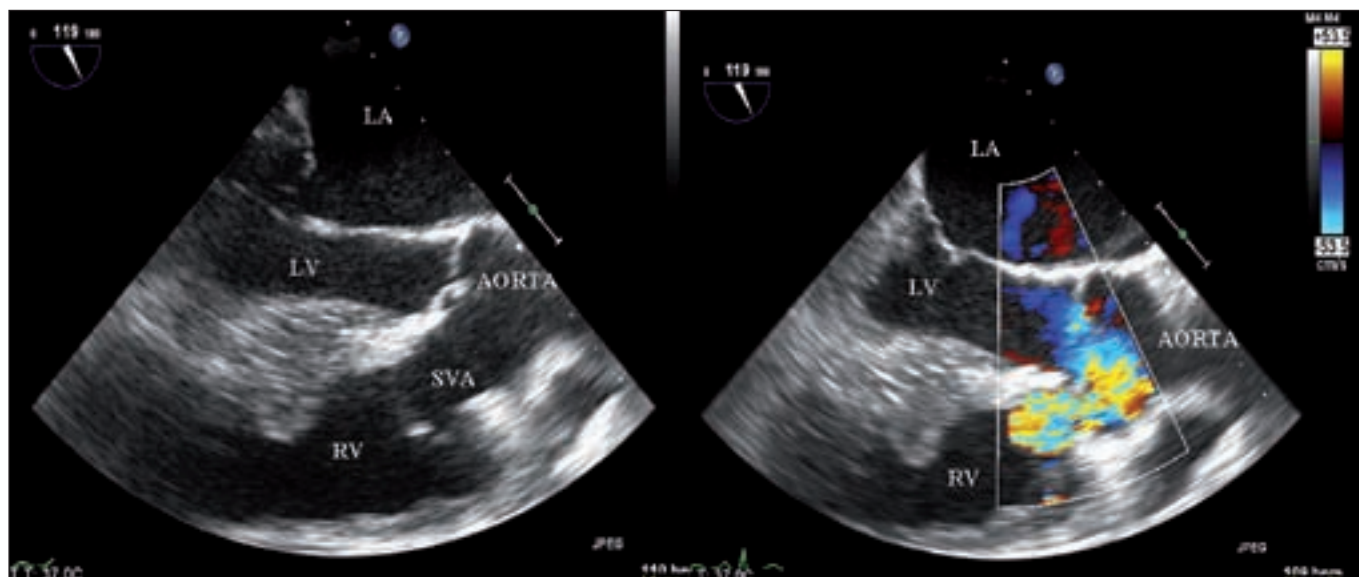


Figure 1. Transoesophageal echocardiography shows a ruptured aneurysm of the right coronary sinus with an obligate left-to-right shunt into the right ventricle, using colour Doppler technique (left). LV=left ventricle, LA=left atrium, RV=right ventricle, SVA=sinus of Valsalva aneurysm.

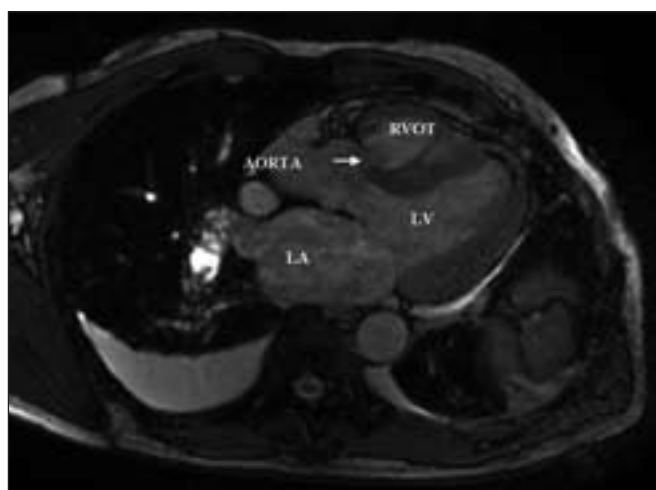


Figure 2. Cardiac magnetic resonance imaging shows a ruptured right coronary sinus of Valsalva into the right ventricle. Arrow shows the ruptured sinus of Valsalva. RVOT=right ventricular outflow tract, LV=left ventricle, LA=left atrium.

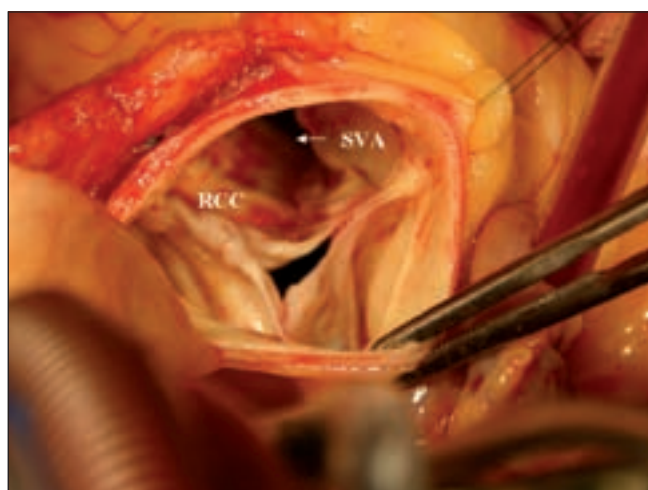


Figure 3. Surgeon's view from the opened aorta into the aneurysm of the right coronary sinus of Valsalva. RCC=right coronary cusp, SVA=sinus of Valsalva aneurysm.

from Asian descent.² The presence of an aneurysm may lead to a compression of an adjacent chamber, a coronary artery or the conduction system, leading to myocardial ischaemia and/or conduction disturbances. Symptoms occur in 80% of patients, most commonly between 30 and 45 years of age.² An aneurysm of the sinus of Valsalva ruptures in 35%, leading to acute symptoms in one fourth of the patients.⁴ Symptoms associated with rupture are shortness of breath, chest pain, and fatigue.^{5,6} The severity of the shunt, the presence of associated lesions, and age at presentation determine the severity of symptoms.²

Most aneurysms rupture into the right-sided heart chambers, and rarely into the left-sided heart chambers, pericardium, and pulmonary artery or superior caval vein.^{4,6} Echocardiography, especially transoesophageal echocardiography, is important for diagnosing ruptured and non-ruptured aneurysms. It allows accurate evaluation of the involved sinus, the presence of a left-to-right shunt or associated cardiac lesions. Magnetic resonance imaging might be useful in diagnosing the coexisting cardiac lesions more precisely. Cardiac catheterisation is currently only performed to evaluate coronary anatomy prior to surgery.

The optimal management for a ruptured SVA is surgical repair. However, the best therapy for non-symptomatic non-ruptured SVA is currently unknown. The mean survival in patients with an untreated ruptured SVA is about four years.⁴ The most used surgical technique is the 'dual exposure technique' where both the aorta and chamber of termination are explored. The aneurysmal sac is excised and the resultant defect will be repaired either by direct suturing or patch closure. Coexisting lesions will be repaired within the same surgical procedure. Today, the ten-year survival rate after surgical repair of a ruptured SVA is 90%.³ However, aortic regurgitation might occur after surgical repair.^{1,4}

In conclusion, sudden onset of acute dyspnoea in the presence of a continuous loud murmur might be due to a ruptured sinus of Valsalva. Echocardiography and magnetic resonance imaging are useful for diagnosis. Left untreated it carries a dire prognosis. The treatment of choice is surgery, and

associated cardiac lesions must be repaired within the same procedure. ■

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