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Aneurysm of sinus of valsalva

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CASE REPORT

Aneurysm of Sinus of Valsalva

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ABSTRACT

Aneurysm of sinus of Valsalva is a rare cardiac abnormality with congenital origin in most of the cases. If it is located in the right coronary sinus, it usually ruptures into a right heart chamber and frequently a ventricular septal defect (VSD) co-exists with this condition. Early diagnosis and immediate surgical treatment can save the patient's life in most cases. All the 3 cases reported in this series had aneurysm of right sinus of Valsalva with associated VSD and mild degree of aortic regurgitation (AR). Two of the cases ruptured aneurysm into the right ventricle. Trans-esophageal echocardiography was used to confirm the diagnosis and all three showed good results with surgery.

Key words: *Sinus of Valsalva aneurysm. Ruptured sinus of Valsalva. Ventricular septal defect. Complication. Aortic regurgitation. Trans-esophageal echocardiography.*

INTRODUCTION

Aneurysm of the sinus of Valsalva is a relatively rare condition. It comprises approximately 0.1-3.5% of all congenital cardiac anomalies. Discovery in the pediatric age group is unusual. There is an association with ventricular septal defect (VSD). In a large series of 129 patients, associated VSD was found in 11.6% patients with sinus of Valsalva aneurysm.[1] Here we are reporting 3 cases of sinus of Valsalva aneurysm with associated VSD.

CASE REPORT

Case 1: A 24 years old female medical student presented with sustained ventricular tachycardia. She was a known case of supra-cristal type of VSD and was having intermittent palpitation and breathlessness for one and a half months. Recent echocardiogram revealed dilated right coronary sinus with a small VSD and mild aortic regurgitation (AR). On presentation she required a brief cardiopulmonary resuscitation and intravenous amiodarone infusion. After resuscitation her examination revealed a blood pressure of 115/40 mmHg, a collapsing pulse with a rate of 100/minute, left parasternal thrill along with a continuous murmur. Her trans-esophageal echocardiogram (TEE) revealed ruptured right sinus of Valsalva aneurysm into right ventricle (Figure 1) along with small perimembranous VSD. A successful surgical repair of ruptured sinus of Valsalva aneurysm and VSD was done on an urgent basis. After surgery while in the hospital she had non-sustained polymorphic ventricular tachycardia, which was treated with beta blockers along with amiodarone. She was found to have long QT interval and developed a bradycardiac arrest requiring pacemaker implantation. Her subsequent course was uneventful.

Case 2: A 48 years old woman, known to have VSD since childhood, presented with 4 months history of intermittent episodes of breathlessness and palpitation. She had an episode of near syncope during this period. Twenty four-hours Holter monitoring showed non-sustained ventricular tachycardia. Clinical examination on presentation revealed a blood pressure of 100/60 mmHg, pulse of 80/minute, a 4/6 pansystolic murmur over the left sternal border and a continuous to and fro murmur all over the precordium.

ECG showed non-specific T-wave changes. Her echo-cardiogram (TEE) revealed a small perimembranous VSD, a large un-ruptured aneurysm of right sinus of Valsalva (Figure 2) and mild AR. Normal coronaries were found on pre-operative coronary angiography. She underwent a successful repair of both VSD and aneurysm of sinus of Valsalva. Operative findings revealed a large aneurysm (4 cm in size) which was protruding through the right ventricular outflow tract (RVOT) into the right ventricle causing RVOT obstruction. Her peri and postoperative course was uneventful.

Case 3: A 28 years old man with no known prior cardiac problem, presented with one week history of fever and vomiting. Clinical examination revealed marfanoid features, a blood pressure of 110/40, pulse of 96/minute which was collapsing in nature, a thrill with a continuous murmur at left sternal border. ECG showed biventricular hypertrophy and left atrial enlargement. Echocardio-gram revealed mildly dilated cardiac chambers, a small perimembranous VSD, mild AR, ruptured aneurysm of

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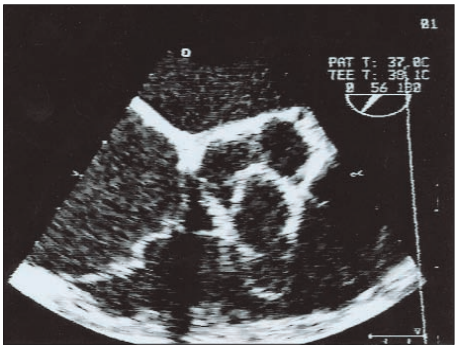

Figure 1: Transesophageal echo view showing ruptured right sinus of Valsalva aneurysm.

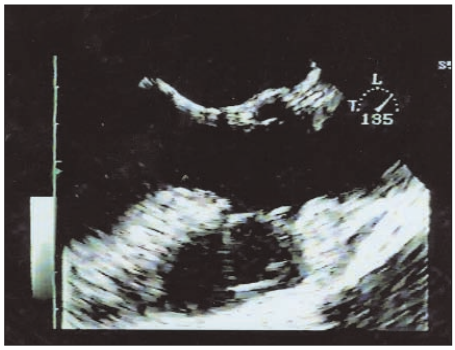

Figure 2: Transesophageal echo view showing small perimembranous VSD and large un-ruptured aneurysm of right sinus of Valsalva .

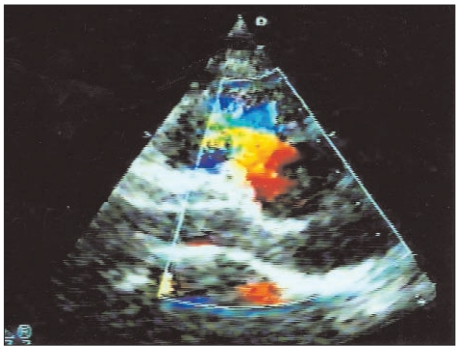

Figure 3: Transthoracic echo view showing color flow through the ruptured aneurysm.

right sinus of Valsalva into right ventricle (Figure 3) and a vegetation on ruptured margin. Parenteral antibiotics were started and he was operated urgently. His blood cultures grew corynebacterium species. Postoperatively he completed 6-8 weeks course of appropriate antibiotics and subsequently remained well.

DISCUSSION

Aneurysm of the sinus of Valsalva is a dilatation of one or more sinuses of Valsalva. It may be congenital or acquired, arising due to dehiscence in attachment of media of the aorta to the annulus fibrosus of the aortic valve ring leading to progressive dilation of the intervening wall and/or formation of fistulous track. Approximately two thirds of surgically resected cases are congenital. Acquired ones are caused either by endocarditis, syphilis or atherosclerosis.[2] Although the condition is more common in males,[3] with a male to female ratio of 4:1, however, two of the patients presented here were females.

All 3 cases had aneurysm of right sinus of Valsalva which is also the most common site of involvement. As per literature the sites of involvement are the right sinus of Valsalva in 76.8%, non-coronary sinus 20.2% and left sinus in less than 3% of cases.[4]

The most common co-existing congenital heart diseases are VSD, usually sub-aortic (25-55%) and regurgitation of the aortic valve and rarely pulmonary stenosis, patent ductus arteriosus, atrial septal defect, subaortic stenosis and tetralogy of Fallot.[5] All three of our patients had associated VSD with mild degree of AR.

Unruptured aneurysms usually produce no symptoms and are often incidentally found during cardiac catheterization or echocardiographic examination, or at autopsy. However, unruptured aneurysms may present with aortic regurgitation, right ventricular outflow obstruction causing cogestive heart failure, complete hearty block, coronary artery compression, resistant ventricular tachycardia and left/right ventricular inflow obstruction.

Aneurysms of sinus of Valsalva can remain silent for several years and usually rupture during the 3rd or 4th decade of life. Rupture is generally believed to be spontaneous, partly depending on the size of aneurysm. Two of our cases had ruptured aneurysm. Bacterial endocarditis occurring in an aneurysm may also lead to its rupture and this was probably the cause in case 3. Rupture results in sudden onset of chest pain, shortness of breath, congestive heart failure, wide pulse pressure, murmur and a thrill.

Aneurysms of sinus of Valsalva may rupture into the heart chambers (usually the right chambers), into the interventricular septum or in the pericardial cavity.[5,6] The former condition was seen in two of these patients.

Trans-thoracic echocardiography is the main diagnostic tool for the identification of the aneurysms of sinus of Valsalva,[7] while multiplane trans-esophageal imaging may further help in the diagnosis and peroperative surgical correction of the lesion, especially when VSD is also present.[8] We used trans-esophageal echocardiography (TEE) for confirmation of diagnosis in all of our patients. In one of the patient TEE was also used intra operatively to detect any residual defect post repair.

Hemodynamic and angiographic study, although limited by the echocardiographic examination, is used to estimate the patency of the coronary vessels, particularly in elderly patients. Finally cardiac magnetic resonance imaging may be used to confirm the diagnosis.[9]

It is generally believed that any aneurysm, ruptured or un-ruptured should be operated upon because of potential morbidity and mortality associated with it. Surgical correction involves removal of aneurysm, direct or patch closure of aortic defect, while protecting the aortic valve and origin of coronary arteries, and closure of VSD if present.[2] Surgical treatment presents low operative risk and has a high long-term survival rate. Lukacs *et al.* reported zero peri-operative mortality in a total of 30 patients with ruptured aneurysm of sinus of Valsalva and advised early intervention.[10] These cases also showed good results with surgery.

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