

Case Report



Ruptured Sinus of Valsalva Aneurysm

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Abstract

Sinus of Valsalva Aneurysms are extremely rare cardiac anomalies. The sinus of Valsalva aneurysm (SVA) is a small dilatation caused by a separation between the aortic media and annulus fibrosus. Its may be either acquired or congenital. The right coronary sinus is most frequently affected. The most common complication being rupture. We present the case of a 12-year old girl with ruptured non-coronary sinus of valsalva aneurysm associated with ventricular septal defect , mitral regurgitation grade II and severe pulmonary hypertension. Echocardiography is sufficient to diagnose SVA, its complications, repercussions, and surgical options. SVA, even if asymptomatic, has potential risks of expansion, rupture, cardiac failure, endocarditis, embolism and sudden death. This justifies surgical correction, with a low mortality rate in both the short- and long-term.

Keywords: Sinus of Valsalva, Aneurysm rupture.

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Introduction

The SVA is defined as a significant dilatation of the aortic wall between the aortic valve and the sinotubular junction.¹ Sinus of Valsalva aneurysms are extremely rare cardiac anomalies that may be acquired or congenital with an incidence ranging from 0.1% to 3.5% of all congenital heart defects.² Most sinus of Valsalva aneurysms arise from the right or the non-coronary sinuses, and they commonly protrude and rupture into the right ventricle or right atrium. Aneurysms may also rupture into the left ventricle, pulmonary artery, left atrium, or pericardial cavity, but these entry sites are exceptional.^{3,4} Aneurysms usually remain asymptomatic unless they are complicated by rupture. Ruptured sinus of Valsalva aneurysms are frequently associated with ventricular septal defects (VSD).⁵ We report

herein the successful treatment of a patient who had a rare combination of a ruptured aneurysm of the non-coronary sinus of Valsalva, a VSD.

Case Report

We present the case of a 12-year-old girl with a history of exertional dyspnoea , palpitation, repeated respiratory tract infection and occasional haemoptysis. He presented clinically with continuous murmur, bilateral basal crepitations in lungs and edema of lower extremities. Chest x-ray showed cardiomegaly with bilateral pulmonary hypertension and oedema. We reported an EKG with sinus tachycardia, right axis deviation and biventricular hypertrophy. Transthoracic

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echocardiogram (TTE) showed ventricular septal defect (perimembranous), ruptured non coronary sinus of valsalva aneurysm into right atrium (Figure 1), left to right shunt (Figure 2), mitral regurgitation (MR) Gr II, pulmonary hypertension, pericardial effusion and normal biventricular systolic function. Corrective surgery was performed. Peroperative findings :Right atrium and both ventricles were dilated. Aortic valve was found competent with noncoronary SVA of 3 cm in length, ruptured towards the RA, and adhered to the tricuspid valve without affecting its functionality (Figure 3). A 10mm×10mm ventricular septal defect (VSD) was found at membranous septa restricted by chordae of tricuspid valve. The repair was performed with the assistance of a cardiopulmonary bypass (CPB) and moderate hypothermia (32°C). The approach was made through aortotomy and right atriotomy. The aneurysm was exposed and resected from the RA (Figure 4) and the defect was closed with a gluteryldehyde treated pericardial patch with a prolene suture 4-00 (Figure 5). VSD was separately closed with a gluteryldehyde treated pericardial patch with a prolene suture 4-00. The patient's postoperative evolution was uncomplicated. Follow up transthoracic echocardiogram (TTE) showed successful repair of ruptured sinus of valsalva and VSD with competent aortic valve and mild MR.

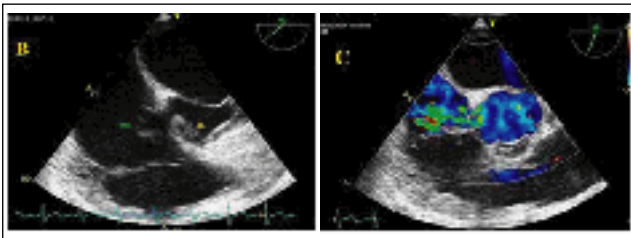


Figure 1 and 2: Ruptured non coronary sinus of valsalva aneurysm into right atrium & left to right shunt.



Figure 3 and 4: Rupture of noncoronary sinus of Valsalva aneurysm (SVA) into right atrium and Resection of aneurysmatic sac of noncoronary SV.

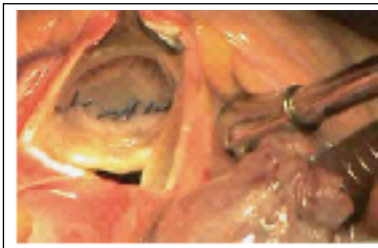


Figure 5: Closure of the defect with gluteryldehyde treated pericardial patch.

Discussion

Sinus of Valsalva aneurysms (SVA) are very uncommon, have a marked male preponderance (4:1), and their incidence is highest in Asian populations. SVA can be congenital or have an acquired origin. The acquired aneurysm is caused by conditions affecting the aortic wall, such as infections (syphilis, bacterial endocarditis, and tuberculosis), trauma, and degenerative disease (connective tissue disorders, Marfan syndrome, and cystic medial necrosis).⁶ The congenital aneurysm is more common and is most often caused by weakness at the juncture of the aortic media and the annulus fibrosus.⁷ They are associated with other heart defects, VSD in 30-60% of cases, aortic valve abnormalities such as aortic insufficiency in 20-30% of cases, bicuspid valve (10%), aortic stenosis (6.5%). They may also be associated with pulmonary stenosis (9.7%), coarctation of the aorta (6.5%), persistence of the conductus arteriosus (3.2%), tricuspid insufficiency (3.2%) and interatrial defect.^{1,8-14}

Aneurysms may originate in the right coronary sinus (65%-85%), the noncoronary sinus (10%-30%), and, rarely, the left coronary sinus (1%-5%).¹⁵ The frequency of ruptured SVA varies according to the location: 60% in the right sinus, 42% in the noncoronary sinus and only 10% in the left sinus. Rupture may be spontaneous, after trauma, extreme physical exercise or due to endocarditis. Rupture of a SVA occurs principally at the RV (60%) or at the RA (29%), the LA (6%), LV (4%) or at the pericardium (1%). Extracardiac ruptures are rare, usually fatal, and occur towards the pericardium or the pleural space and are more common when the SVA is of acquired origin.^{1,8,11,12,16,17,19} Physiopathological consequences of SVA rupture depend on the volume of flow through communication, velocity of establishment of the rupture and cardiac chamber with which it communicates. The perforation of acutely high flow does not allow hemodynamic compensation, developing a sudden CI.^{14,16,19,20} A small, gradual and progressive perforation can be tolerated temporarily, occurring in 25% of cases.¹¹ The intact SVA may be asymptomatic, produce thrombus or distortion and compression of the coronary arteries, leading to ischemia,^{10,16,20} arrhythmias, tricuspid insufficiency, aortic regurgitation or, very rarely, a right ventricular outflow tract obstruction. Rupture of sinus of valsalva aneurysm (RSOV) is associated with dramatic onset of symptoms and deterioration to a state of biventricular cardiac failure due to sudden volume overload.¹ SVA rupture causes thoracic pain, dyspnea, cough, fatigue, tachycardia, or peripheral edema, as well as a continuous murmur. Clinical presentation is less severe in case of preexisting ventricular septal defect (VSD). Diagnosis is an indication for early surgical intervention, as the mean survival period for untreated RSOV patients is 1-3.9 years. Unruptured SVA that produces malignant arrhythmias, infection, blockage of coronary arteries or ventricular outflow tracts indicate the need for surgery.^{1,8,11,17}

Conclusion

Clinical suspicion followed by echocardiographic evaluation is the key for diagnosing this condition. The traditional treatment modality is a surgical closure of a ruptured coronary sinus. There is less experience with transcatheter closure using Amplatz occluder. Surgical procedure in this case is connected with a low risk of postoperative mortality (1.9 -3.6%) and with good long-term results.^{21,22}

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