Percutaneous device closure of ruptured sinus of Valsalva aneurysm in an adult male

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Abstract:
Forty seven years old man, known diabetic, hypertensive and non smoker presented with gradually progressive shortness of breath NYHA class II for 2 years. He was found to have a grade 56 superficial, loud continuous murmur with harsh sawing character. Transthoracic echocardiography revealed a ruptured aneurysm of the right aortic sinus with continuous flow into the right ventricular outflow tract. Catheterization study revealed an 8 mm defect in the right coronary sinus along with trivial aortic regurgitation. Transcatheter device closure of the ruptured sinus of Valsalva (RSOV) aneurysm was done using a 1210 Lifetech PDA device. The post procedure period was uneventful and he was discharged within 24 hours.

Keyword: Sinus of Valsalva aneurysm, heart failure, device closure, transthoracic echocardiography, right ventricular outflow tract

Introduction and objectives:
Aneurysms of sinuses of Valsalva (ASOV) are thin-walled saccular or tubular outpouchings of the aortic sinuses, which can be either congenital or acquired. They can rupture into heart chambers, the pulmonary artery or the pericardial space. Congenital sinuses of Valsalva aneurysms commonly present as unruptured aneurysm with no symptoms which are detected incidentally. Rupture may follow severe exertion or chest trauma. If a huge shunt develops rapidly, the symptoms of congestive heart failure appear almost immediately. Some small insidious perforations progress gradually and initially go unnoticed. Mild dyspnea without pain can precede congestive heart failure by months or years. We hereby discuss an adult male who presented with gradual onset dyspnea of 2 years duration.
Case Description:
Forty seven year old man, known diabetic and hypertensive, non-smoker presented with gradual onset dyspnea NYHA class II for 2 years. There was no associated chest pain on exertion or any sudden severe chest pain at the onset. There was no history of prolonged fever or chest trauma. There was no past history of acute coronary syndrome. On examination, there was no pallor, clubbing, cyanosis or pitting pedal edema. Jugular venous pressure was not elevated and had normal waveforms. Pulse was 84 per minute, regular, normal volume and character and was bilaterally equal and symmetrical. The blood pressures were 130/70 mm Hg in both upper limbs and 140/80 mm Hg in the right lower limb. The apex beat was localized in the left 5th intercostal space in the mid-clavicular line and was of normal character. There was a palpable continuous thrill in the left third intercostal space. The first and second heart sounds were normal. There were no additional sounds. There was a grade 5/6 loud, superficial
continuous murmur with a harsh sawing character which was best heard in the left 3rd intercostal space. The murmur was loudest during systole and less loud during diastole. There were no other coexisting murmurs. Examination of other systems revealed no abnormalities. Routine blood investigations revealed no abnormality. His electrocardiogram showed sinus rhythm and left ventricular hypertrophy by voltage criteria. The chest radiograph revealed normal cardiothoracic ratio with clear lung fields. Transthoracic echocardiogram revealed a 4 mm defect in the right coronary sinus of Valsalva rupturing into the right ventricular outflow tract with continuous left to right flow. There was no associated ventricular or atrial septal defect, aortic or mitral regurgitation, bicuspid aortic valve, patent ductus arteriosus or coarctation of aorta. The left ventricular ejection fraction was normal. His coronary angiography revealed normal epicardial coronaries. Aortic root angiogram showed an 8 mm defect in the right coronary sinus. There was trivial aortic regurgitation. Left ventriculogram did not show ventricular septal defect or mitral regurgitation. The ruptured sinus of Valsalva (RSOV) was crossed from the aorta into the right atrium using a 6F Judkins Right diagnostic catheter and a 0.035" (300 cm exchange length) Terumo glide wire. The glide wire was passed up into the inferior vena cava across the RSOV. The glide wire was snared out from the right femoral venous approach using a 6F 20 cm loop goose neck snare and exteriorized. An 8F 55 cm sheath was tracked over this wire with its distal end in the ascending aorta. A 12/10 Lifetech PDA device was back loaded into 7 F short sheath and passed onto the 55 cm long sheath. The device was positioned in the right coronary sinus of Valsalva aneurysm under fluoroscopy and rechecked by transthoracic echocardiogram. Aortogram confirmed adequate position and the device was deployed. Post deployment angiogram revealed no flow through the device. On follow up, the patient’s functional capacity had improved to normal effort tolerance and transthoracic echocardiogram revealed no flow across the device.

Discussion:
A localized weakness of a sinus of Valsalva is a relatively rare lesion. This was first reported in 1839 by Hope. (1) The localized aneurysms are usually congenital but can follow infective endocarditis. Rupture of the aneurysm can follow acute chest trauma or severe exertion. Ninety to 95% originate in the right or non coronary sinus and project into the right ventricle or right atrium leaving <5% originating in the left coronary sinus. (2) Those arising in the non coronary sinus almost all rupture into the right atrium. Those arising from the right coronary sinus rupture mostly into the right ventricular, occasionally into the right atrium and rarely rupture into the pulmonary artery, left ventricle, left atrium or the pericardial sac. The physiological consequences or rupture depends on (2)

1 Amount of blood flowing into the rupture

2 The rapidity with which the rupture develops

3 The chamber that receives the rupture
The various associations of these aneurysms are with ventricular septal defects (especially defects of the outlet septum), atrial septal defect, patent ductus arteriosus, coarctation of aorta and tetralogy of Fallot. Ruptured congenital sinus of Valsalva aneurysm occur chiefly in males with a sex ration of 4:1.
It is more common in young men after puberty but before the age of 30 years. Rupture is usually accompanied by a tearing pain in the chest or upper abdomen. If a huge shunt develops rapidly, symptoms of congestive heart failure develop acutely. However, with small fistulas the symptoms may take several months to develop. About 20% of cases are asymptomatic. These aneurysms can symptoms without rupture by obstructing the right ventricular outflow tract, distorting the aortic valve causing aortic regurgitation, compressing the left coronary artery causing myocardial ischemia or causing conduction disturbances including complete heart block by compressing the conduction system.(2) With a small fistula, there may only be a continuous murmur resembling that of patent ductus arteriosus, but with its maximal intensity at the 3rd or 4th intercostal space near the sternal edge. If the aneurysm ruptures into the right atrium, the murmur may be maximal at the right of the sternum. A large fistula entering into the left ventricle may simulate aortic regurgitation. If a ventricular septal defect is present along with infundibular narrowing, the combined murmurs can be confusing. With larger fistulas, there will be a wide pulse pressure, a collapsing pulse and left ventricular hyperactivity. The electrocardiogram shows enlargement of the appropriate chambers depending on the site of rupture. Occasionally, signs of myocardial ischemia or conduction defects may be present. The chest radiograph will show enlargement of the appropriate chambers along with pulmonary overcirculation if there is a large left to right shunt. Evidence of congestive heart failure may be seen. Rarely an aneurysm of the left coronary sinus presents as a localized convex radiologic prominence immediately below the pulmonary trunk, or large saccular aneurysm of the right coronary sinus presents as a prominent right paracardiac density. Echo-cardiography with color flow imaging and Doppler establishes the diagnosis of ruptured or unruptured sinus of Valsalva aneurysm along with associated abnormalities. Two dimensional echocardiogram identifies the aneurysmal sac along the sinus of origin. Doppler studies define the flow pattern in the ruptured aneurysm. Transesophageal echocardiography and 3D echocardiography gives information not obtainable by routine transthoracic echocardiogram.(3) Further non invasive imaging with computed tomography or magnetic resonance imaging also provides excellent definition of the aneurysm and the tissue planes involved. The current definitive therapy is surgical anatomic correction. However, percutaneously delivered devices have been used to occlude the ruptured aneurysm, but caution should be advised so as to not cause future aortic regurgitation by the device. The transcatheter technique for RSOV closure was first reported by Cullen et al, with a Rashkind umbrella device in 1994.(4) Since then, Gianturco coils, Amplatzer duct occluders, and Amplatzer septal occluders have been used for device closures of RSOV.(5)

Summary:
Ruptured sinus of Valsalva aneurysm usually presents acutely and the symptoms of congestive heart failure appear almost immediately. However, sub acute presentation with dyspnea is occasionally present. The presence of the characteristic continuous murmur suggested the diagnosis, which was confirmed by transthoracic echocardiogram. Transcatheter technique of device closure was successful, with less morbidity and a shorter hospital stay compared to surgical correction.
Conclusion:
The transcatheter technique provides good efficacy in the treatment of ruptured sinus of Valsalva aneurysm and avoids the risks of open heart surgery and cardiopulmonary bypass.

Bibliography


