Ruptured sinus of valsalva

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ABSTRACT

This case report is about a 24-year-old lady, labeled case of ventricular septal defect since birth, who presented with sudden onset of chest pain and palpitation followed by acute pulmonary edema. She was found to have clinical cardiomegaly, biventricular impulse, pansystolic murmur of ventricular septal defect with continuous murmur at the right lower sternal border. Electrocardiogram revealed frequent ventricular premature beat and chest skiagram revealed cardiomegaly with pulmonary venous congestion. Transthoracic color doppler echocardiography revealed ruptured non coronary sinus of valsalva to right ventricle with large left to right shunt, perimembranous sub aortic ventricular septal defect with left to right shunt and prolapse of right coronary cusp with mild aortic regurgitation and good left ventricular systolic function. She underwent corrective surgery of ruptured sinus of valsalva and ventricular septal defect and aortic valve replacement

Keywords: Ruptured sinus of valsalva, non coronary sinus, perimembranous sub aortic ventricular septal defect, aortic regurgitation.

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Congenital aneurysms of sinus of valsalva is an uncommon condition and usually presents after its rupture. In 1839 Hope1 described "a case of aneurysmal pouch of aorta bursting into the right ventricle" which, was the first published account of a ruptured congenital aneurysm of sinus of valsalva. The most commonly involved sinuses are the right and less commonly noncoronary sinus.2 The involvement of the left coronary sinus is rare.3 Aneurysms arising from the right sinus generally communicate with the right ventricle and occasionally to right atrium, however aneurysms originating in the noncoronary sinus almost always ruptures to right atrium.2 It is to be noted that about 20% of the congenital sinus of valsalva aneurysm are unruptured and were chance findings at autopsy or cardiac surgery4 but now readily identified by color doppler echocardiography.5,9 We are presenting a case of rupture of aneurysm of non coronary sinus of valsalva to right ventricle diagnosed by color doppler echocardiography.

Case Report. A 26-year-old unmarried lady labeled to have ventricular septal defect (VSD) since childhood was admitted with history of severe bilateral chest pain for 2 hours followed by palpitations and shortness of breath for 3 days prior to admission. She was asymptomatic prior to this illness and denied history of prolonged fever, undue physical stress or blunt trauma to chest. On admission she was orthopneic, pulse was 110/min collapsing in nature, blood pressure 150/60. Mean pressure of JVP was not elevated. Cardiovascular examination revealed clinical cardiomegaly with biventricular hyperdynamic impulse, prominent systolic thrill along the left sternal border and pulmonary artery pulsations. First and 2nd heart sounds were normal.
sounds were audible and she had grade 4/6 systolic murmur on the left sternal border, prominent continuous murmur with diastolic augmentation at the right lower sternal border and grade 3/6 ejection systolic murmur at the pulmonary area. She had bibasilar rales upto the infrascapular regions of chest. Electrocardiogram revealed normal PR interval with QRS axis + 60 and frequent ventricular premature beats. Chest skiagram showed cardiomegaly with cardiothoracic ratio 60% with pulmonary venous congestion and increased pulmonary blood flow. Echo colour doppler study revealed left ventricular internal diameter (LVID) diastole 4.6 cm, LVID systole 3.1 cm, left atrium 3.96 cm, Aorta 3.6 cm left ventricular ejection fraction 57%. Echocardiography revealed ruptured aneurysm of non coronary cusp to right ventricle measuring 7 mm (Figure 1), sub aortic perimembranous ventricular septal defect (VSD) 4 mm with trileaflet aortic valve with good opening motion with thickened tips, mild prolapse of right coronary cusp. Mitral valve was normal and so was the pulmonary and tricuspid valves and no vegetations were noticed. Color flow imaging revealed left to right shunt through non coronary sinus and also through the ventricular septal defect (Figure 2). She also had mild aortic regurgitation (Figure 3). Continuous wave doppler interrogation of ruptured sinus revealed continuous flow in right ventricle and aortic regurgitation with pressure half time of 540 milli seconds. There was no evidence of mitral or tricuspid regurgitation. Peak instantaneous gradient across pulmonary valve was 26 mm Hg. Interatrial septum was intact and the systemic and pulmonary venous drainage was normal. Her renal functions were normal and blood cultures did not yield any growth.

**Discussion.** Rupture of sinus of valsalva is an uncommon disorder and results due to aortic medial separation from the anulus fibrosus resulting in aneurysmal dilatation of one or more of the aortic sinus and rupture to form an aortocardiac fistula. Diffuse dilatation of one or more aortic sinuses, with rare presentation of rupture are also reported in Marfans Syndrome. Though rupture of sinus of Valsalva is more commonly observed in adults, cases have also been reported in neonates and in pediatric age group. Depending on the anatomical location of the aneurysmal bulge a number of complications may ensue. These frequently rupture into the right ventricle, right atrium, left ventricle, and pulmonary artery but rarely into pericardium. Dissection into the interventricular septum is a very rare complication of sinus of valsalva aneurysm which originates from the right coronary sinus. Associated congenital abnormalities with ruptured sinus of valsalva included supracristal VSD, Gerbode defect, aortic valve regurgitation, bicuspid aortic valve.
atrium. However, this patient had rupture of non coronary cusp right atrium, as also reported by Lim et al., rupture of non coronary cusp aneurysm to pericardium, and rupture of right coronary cusp to right ventricle simultaneously had been reported by Lim et al., rupture of right coronary cusp aneurysm to left ventricle, and rupture of left coronary cusp right atrium, has also been reported by various authors. This patient had acute development of large perforation resulting in sudden onset of dyspnea with large left to right shunt with clinical and radiological evidence of cardiac failure. No atrio ventricular conduction abnormalities were observed. Two dimensional echocardiography plays an important role in diagnosing ruptured or unruptured sinus of valsalva aneurysm, and with use of contrast echocardiography, and pulsed doppler, and color doppler imaging help to confirm the defect. Use of transesophageal echocardiography, and magnetic resonance imaging are complimentary and helps to confirm the anatomical details. Two dimensional color doppler and pulsed doppler echocardiography clearly identified the rupture of non coronary cusp aneurysm of sinus of valsalva to right ventricle with association of perimembranous sub aortic VSD and mild aortic valvular regurgitation. She improved on decongestive therapy and subsequently she underwent corrective surgery with patch closure of the VSD and ruptured aneurysm with replacement of aortic valve.

In conclusion, congenital ruptured sinus of valsalva is an uncommon disorder but is to be suspected in patients with long history of VSD with sudden onset of cardiac failure with continuous murmur with wide pulse pressure. Of the various aneurysms of sinus of valsalva the occurrence of rupture of non coronary cusp aneurysm to right ventricle is rare. The presence of perimembranous sub aortic VSD is also not a common association as sub arterial VSD is more often reported with aneurysm of sinus of valsalva. Two dimensional colors doppler and pulsed doppler echocardiography are extremely useful tools for accurate diagnosis and for documenting the communication with various cardiac chambers. These patients require patch closure of the aneurysm and closure of VSD with plication of aortic valve or aortic valve replacement.

References

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