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Unusual case of ruptured sinus of Valsalva: rupture into left ventricle cavity along with distortion of mitral valve requiring double valve replacement

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Abstract

We report a rare case of sinus of Valsalva aneurysm of left aortic sinus, which was bulging into left atrium (LA) distorting mitral valve and rupture into the left ventricle with severe aortic regurgitation in a 25 years old male patient. Due to the rupture, both aortic and mitral valves were irreparable and required a double valve replacement along with the closure of the neck of the aneurysmal cavity occupying the left atrial roof.

MeSH: Sinus of Valsalva, Echocardiography, Aortic valve, Mitral valve, Heart defects, congenital

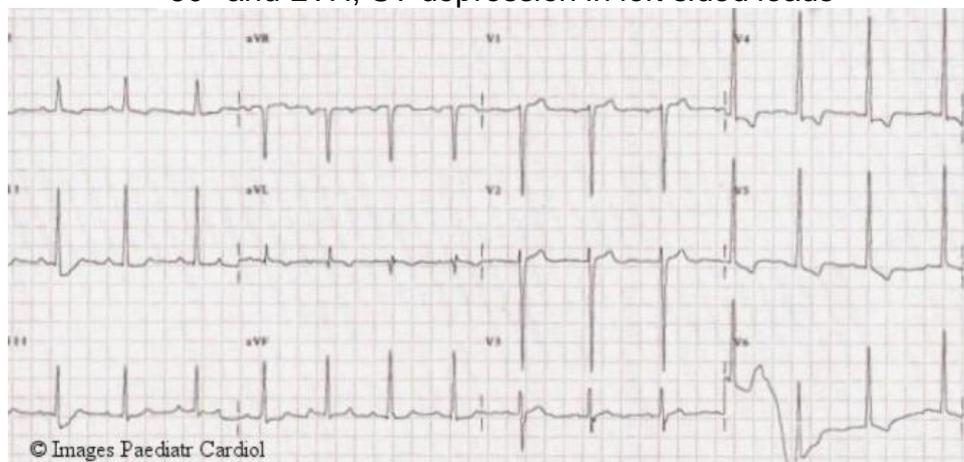
Introduction

Though well known, ruptured sinus of Valsalva (RSOV) aneurysms are rare comprising 0.3-3.56 % of all congenital heart diseases.¹ They are thin walled outpouchings of the sinus of Valsalva and may be tubular or saccular in shape. The right sinus of Valsalva is most commonly involved and usually ruptures into right heart chambers. Uncommonly they rupture into left heart chambers (left atrium, left ventricle), pulmonary artery, interventricular septum or the pericardial cavity.^{2,3} Rupture into the left ventricle (LV) is very rare.^{4,5} We report a young male patient who was diagnosed to have a large aneurysm of left sinus of Valsalva with rupture into LV along with distortion of mitral valve requiring replacement of both aortic and mitral valve. To our knowledge, an aneurysm of the left Valsalva sinus bulging into the left atrial (LA) roof and damaging the mitral valve along with concomitant rupture into the left ventricular cavity leading to severe aortic regurgitation (AR) and thus requiring a double valve replacement (DVR) has not been reported.

Patient

PK, a 25 year old male patient was referred to our hospital with chest pain and dyspnea on exertion for the previous two months. Chest pain used to increase in severity on lying down. There was no history of prolonged fever or trauma. On examination, he was of an average built. There was pallor, tachycardia (HR-110/minute), tachypnoea (respiratory rate-24/minute) and wide pulse pressure (BP 124/40 mmHg in right upper limb). Cardiac examination revealed cardiomegaly, a hyperdynamic precordial impulse, an ejection systolic murmur grade 2/6 and a pandsystolic murmur at the left parasternal border. Chest roentgenogram (postero-anterior view) showed gross cardiomegaly. Twelve lead electrocardiogram showed sinus tachycardia, QRS axis +60° and LV hypertrophy, and ST depression in the left sided leads (figure 1). Transthoracic echocardiography done on Phillip IE33 showed a large aneurysm of the left sinus of Valsalva with rupture of the aneurysm into the LV (figures 2-6).

Figure 1 12 lead ECG showing sinus tachycardia(HR-90/minute), QRS axis +60° and LVH, ST depression in left sided leads



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Figure 2 Apical 4 chamber view with anterior tilt (5 chamber view) showing aneurysm from left sinus bulging into left atrium and impinging to mitral valve. Left ventricle is dilated.



Figure 3 Parasternal short axis view at the level of great vessels showing large aneurysm arising from left sinus of Valsalva

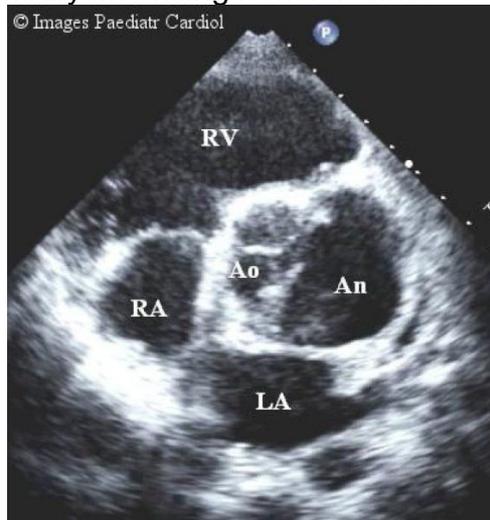
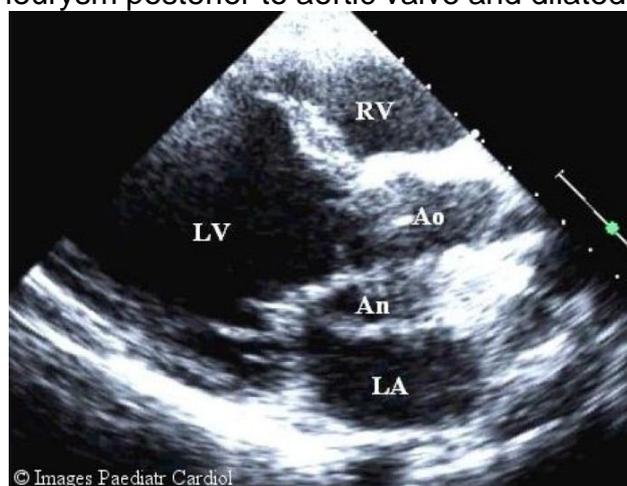


Figure 4 Standard Parasternal long axis view showing aortic valve and small portion of aneurysm posterior to aortic valve and dilated left ventricle.



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Figure 5 Anterior tilt from figure 4 opened up whole aneurysm extending anteriorly to right ventricle outflow and posteriorly bulging into left atrium.

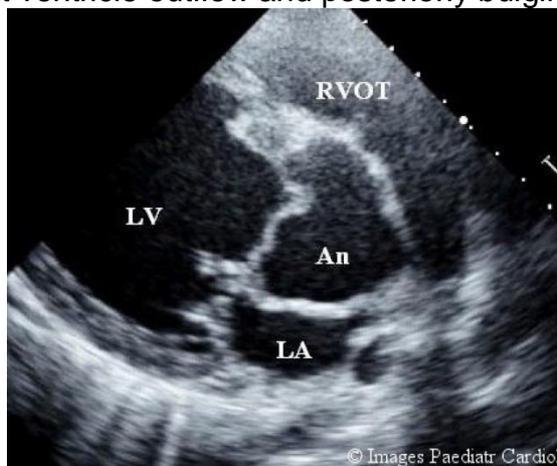
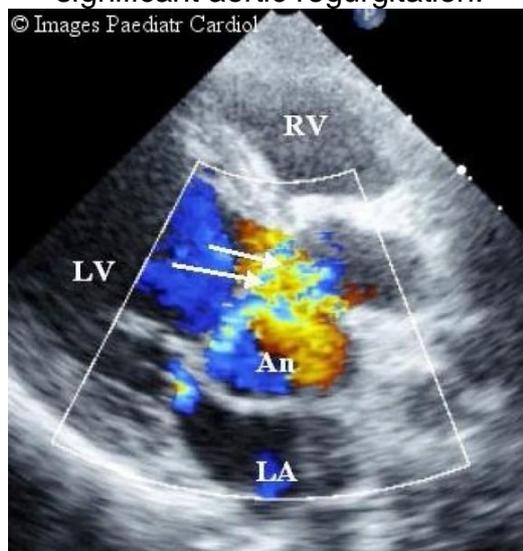


Figure 6 Parasternal long axis view with color flow mapping showing significant aortic regurgitation.



LA-left atrium, RA-right atrium, LV-left ventricle, RV-right ventricle, MV-mitral valve, Ao-aorta, An-aneurysm, RVOT-right ventricle outflow tract

The aneurysm bulged into the mitral inflow distorting the mitral valve (figures 2,3). Color flow mapping showed 3 jets of aortic regurgitation, one central jet and two other from the ruptured aneurysm (figure 6). It also revealed turbulent flow across mitral inflow and moderate mitral regurgitation (MR). Continuous wave Doppler interrogation of the mitral inflow revealed a transmitral mean gradient of 7 mmHg. As transthoracic windows were adequate, transesophageal echocardiography was not done.

Early surgical intervention was advised and was performed through midsternotomy under routine cardiopulmonary bypass. At surgery, a large aneurysm of the left sinus of Valsalva measuring 5 by 6 cm was found originating just distal to the orifice of the left coronary artery and protruding into the LV cavity. The aneurysm had ruptured into the LV cavity at its distal extent. The left coronary cusp was distorted and stretched resulting in severe aortic regurgitation. Posteriorly, the aneurysm had burrowed into the roof of

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the left atrium causing displacement of the anterior mitral leaflet, resulting in significant MR.

It was apparent that both aortic and mitral valves needed to be replaced. The aortic valve and most of the aneurismal sac were excised through the aortotomy. The mouth of the aneurysm was closed with a Gore-Tex (expanded PTFE) patch and the aortic valve was replaced with a 19 mm St. Jude aortic valve prosthesis. The left sinus portion of the valve was anchored to the Gore-Tex patch. The mitral valve was inspected through a left atriotomy to assess feasibility of repair. There was marked distortion of the mitral annulus by the aneurysm and the valve cusps were considerably thickened. Feasibility of durable repair was ruled out and a valve replacement with 25 St Jude Mitral valve was performed. The patient had fairly uneventful postoperative recovery.

Discussion

The earliest descriptions of sinus of Valsalva were in the reports of Hope and Thurman.^{6,7} Later, Abbott described this malformation as congenital and not acquired in origin.⁸ Most aneurysms are congenital in origin but they may be seen after bacterial endocarditis, atherosclerosis or chest trauma.^{2,9} Aneurysms of sinus of Valsalva are thought to result from the absence of normal elastic and muscular tissue, which leads to thinning of the wall of the aortic sinus.⁴⁻⁷ In a study of 332 patients collected from Western and Oriental literature by Chu et al, pathological rupture of a sinus of Valsalva most frequently involved the right sinus (76.8%), followed by the noncoronary (20.2%), and least commonly, the left sinus of Valsalva (3%).¹ On reviewing the English literature we could find only 26 cases of RSOV ruptured into the left ventricle and out of them only 3 arose from a left sinus of Valsalva aneurysm.⁵ In all of them, the aneurysm was repaired along with aortic valve replacement.

Presentation of sinus of Valsalva aneurysm is varied.¹⁻⁵ With rupture, the presentation is acute with chest pain and dyspnea. Chest pain is due to acute AR from the ruptured sinus of Valsalva, frank or dynamic coronary artery compression, or dissection into a coronary artery. Unruptured sinus of Valsalva aneurysms are usually asymptomatic but may sometimes present as right ventricular outflow tract obstruction when they bulge into right ventricular outflow tract, or subaortic stenosis when they bulge into the left ventricular outflow tract. Rarely, complete heart block occurs when it erodes into the interventricular septum.¹²

Associated heart defects that have been reported are ventricular septal defect (in about 50% of cases), pulmonary stenosis, tetralogy of Fallot, coarctation of aorta, bicuspid aortic valve, valvular and supra-valvular aortic stenosis, left superior vena cava and one case report of anomalous origin of left coronary artery from pulmonary artery.²

Initially, angiography was the gold standard for the diagnosis but now, noninvasive modes of diagnosis such as echocardiography (transthoracic, transesophageal), spiral computed tomography and cardiac magnetic resonance imaging have taken over as preferred investigations. In our case we were able to delineate the detailed anatomy by transthoracic echocardiography and the patient underwent surgery requiring replacement of both aortic and mitral valve.

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