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Case Report

Unusual presentation of ruptured non coronary sinus of valsalva with new onset heart failure

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ABSTRACT

Rupture of sinus of Valsalva is a rare cardiac condition which is associated with severe left to right shunting. Symptoms may include breathlessness, chest pain and fatigue or even cardiogenic shock and when untreated, this condition carries a grave prognosis. We report a case of a 57-year-old gentleman without any past comorbidities who presented to our hospital with features of acute heart failure due to rupture of sinus of Valsalva which was diagnosed by echocardiogram and further confirmed by cardiac computed tomography scan. Patient underwent successful patch closure of the defect and made a good recovery. His hospital stay was uneventful and is under regular follow up. This highlights the importance of multimodality imaging and timely treatment for cases of rupture of sinus of Valsalva.

Keywords: Echocardiogram; ruptured sinus of Valsalva; sinus of Valsalva aneurysm.

INTRODUCTION

The sinuses of Valsalva are three focal expansions that form the walls of the aortic root.[1] The anatomic position of the sinus within the heart determines the clinical outcome in case of aneurysm rupture.[2] Although the true prevalence of Sinus of Valsalva Aneurysm (SOVA) is unknown, the estimated rate is approximately 0.09% of the general population.[3] SOVAs usually affect the right coronary sinus (in 70% of cases), followed by the noncoronary sinus (in 25% cases) and rarely the left coronary sinus.[4] Typically men are more affected (4:1) and there is a higher reported incidence in Asian groups.[5] In this report, we describe case of a middle aged gentleman who presented with rupture of non coronary sinus of Valsalva presenting as acute heart failure.

CASE REPORT

A 57 year old gentleman presented to another hospital for complaints of sudden onset shortness of breath which was rapidly progressive in nature. There was no associated chest pain, cough or palpitations or any history of fever or any skin rash. There was no any history of prior congenital heart disease or any family history of cardiac illnesses. There was no history of TB in the past. He had received the initial symptomatic treatment at other centre for his symptoms but which didn't subside, and so was referred to our centre.

On clinical examination, he was found to be in respiratory distress and hypotensive with blood pressure of 84/66 mmHg, pulse of 110 beats per minute. Physical examination revealed fine basal crepitations at both the lung bases and a loud continuous murmur of Grade IV with thrill along the left sternal edge. Jugular venous distention was present and there was bilateral pitting pedal edema.

A 12 lead ECG done showed sinus tachycardia while his chest X-ray revealed bilateral mild pleural effusion. Other lab parameters showed Hemoglobin - 14.8 g/dl, total leucocyte counts -16900, platelets count - 289000, Serum Urea - 54 mg/dl, Serum creatinine - 1.3 mg/dl, Cardiac markers - Troponin I negative , Serum BNP - 980 pg/ml. Blood culture sets from two different sites was sent which reported no growth.

Further, a Transthoracic echocardiogram was done which showed abnormal turbulent flow across aortic sinus into the RA- RV junction with peak gradient of 111 mmHg

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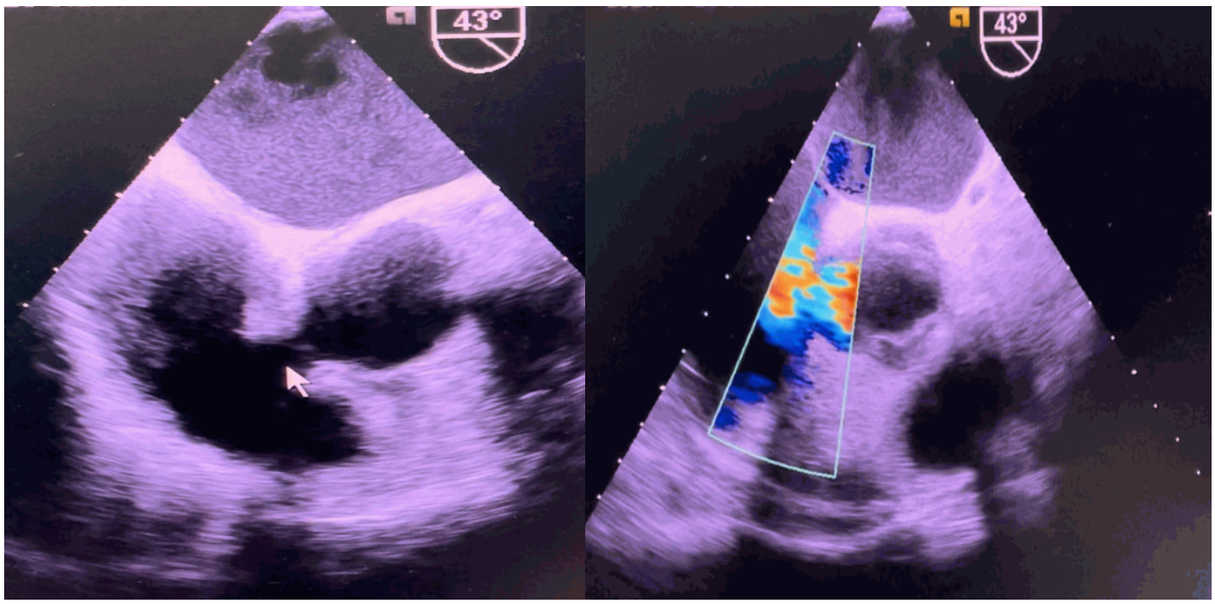

Figure 1. TEE at midesophageal level at 43 degrees showing ruptured noncoronary sinus of valsalva (white arrow). Color doppler showing left to right shunting from ruptured sinus of valsalva to right atrium.

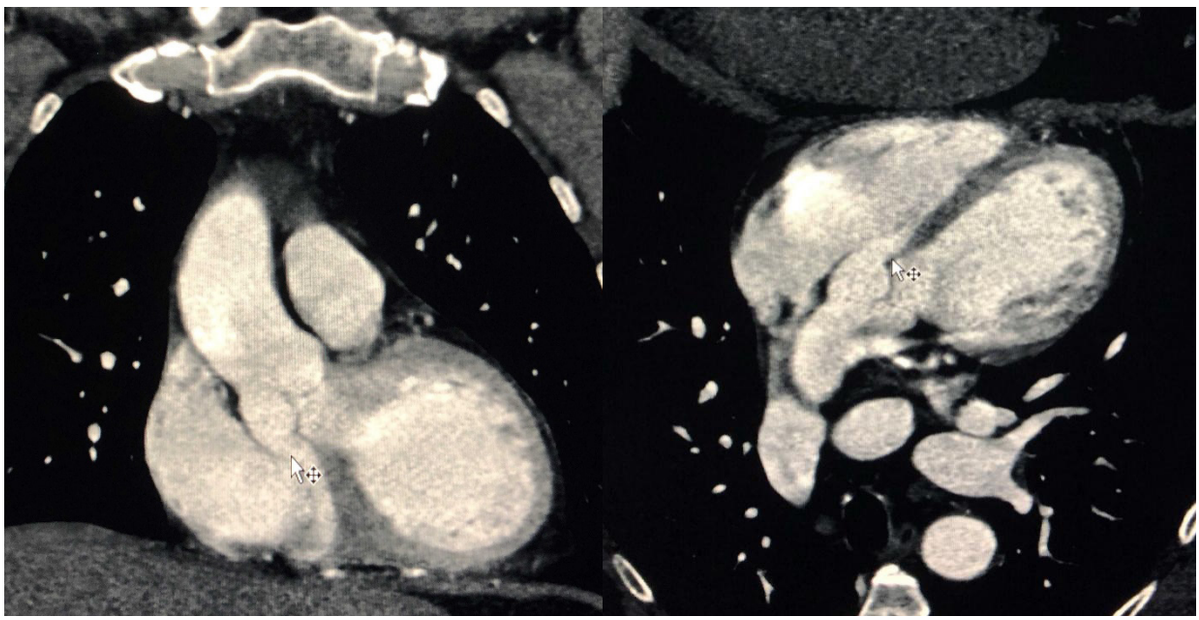

Figure 2. CT aortogram showing the ruptured sinus of valsalva with communication into right atrium (white arrow).

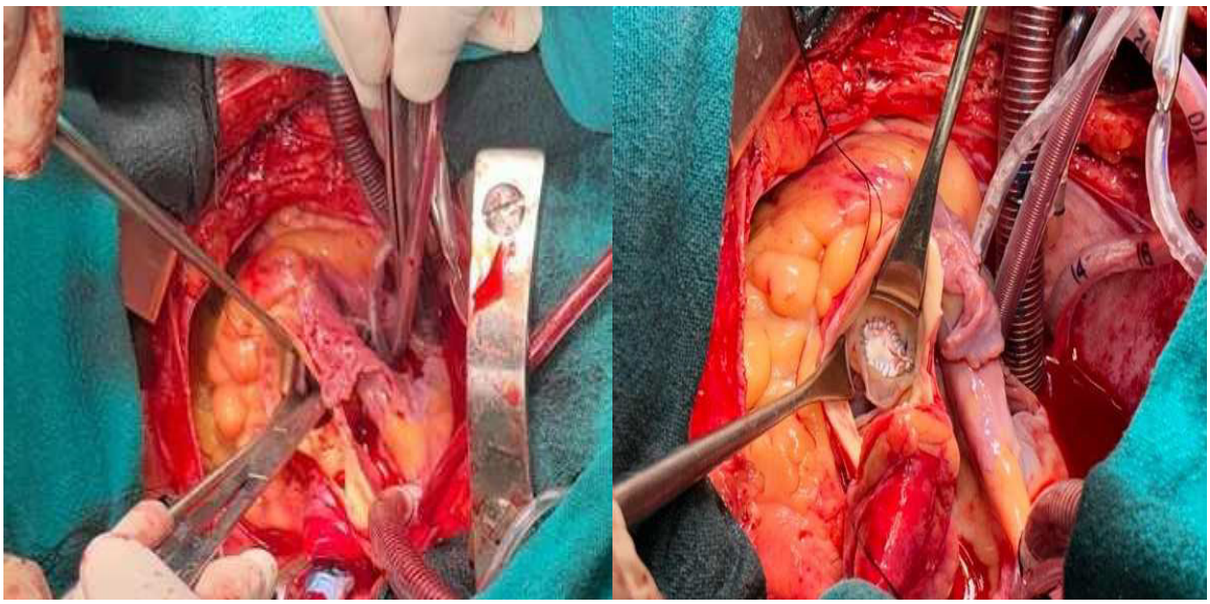

Figure 3. An artery forceps (hemostat) passed showing the communication between sinus of valsalva and right atrium; same defect closed with PTFE patch.

and left to right shunt and mild tricuspid regurgitation with moderate pulmonary hypertension with ejection fraction 60%. Transesophageal echocardiogram done at mid- esophageal level showed ruptured non-coronary sinus of Valsalva with left to right shunting into right atrium (figure 1). An Ultrasonogram of abdomen showed prominent hepatic veins with hepatomegaly. Cardiac CT and Aortogram showed about 7.8 mm defect with communication between right posterior non-coronary sinus with right atrium just above the atrio-ventricular valve; with normal origin of both left main coronary artery and right coronary artery. There was no evidence of aortic dissection or any vegetation (figure 2).

Patient underwent surgical fibrous tissue excision with PTFE (polytetrafluoroethylene) patch closure with direct closure of incidental PFO noted during the surgery (figure 3). The patient tolerated the procedure well and made a good recovery. He is doing well and is under regular followup with no clinical symptoms.

DISCUSSION

SOVAs can either be congenital or acquired. A congenital SOVA is frequently associated with Marfan syndrome, Ehlers-Danlos syndrome or other connective tissue disorders.[2,6] Congenital SOVAs are usually associated with other cardiac anomalies like ventricular septal defect, aortic valve dysfunction, bicuspid aortic valve and coarctation of aorta.[7] Acquired aneurysms have been associated with trauma, atherosclerosis, infective endocarditis, iatrogenic injury during aortic valve replacement, syphilis and collagen vascular disorders.[2,6]

Rupture of sinus of valsalva (RSOV) may have a diverse presentation ranging from asymptomatic murmur to acute heart failure and cardiogenic shock.[8] Symptoms associated with rupture are shortness of breath, chest pain, and fatigue. Our index case presented with an unusual presentation of acute new onset heart failure in a very rare case of rupture of non coronary sinus of Valsalva. The pathology is due to failure of fusion between aortic media and the heart at the level of the annulus fibrosus of the aortic valve, with subsequent aneurysmal enlargement at this weak point due to the high head of pressure at the root of the aorta.[9]

Traditionally, transthoracic and transesophageal echocardiogram have been the first-line imaging techniques; which can visualize the aneurysm walls and the disturbed blood flow at the site of perforation.[10] Echo features include aneurysmal dilatation of sinus of valsalva and on colour flow imaging, there is a unidirectional continuous mosaic high velocity jet from the aorta to the right heart chamber. Transoesophageal echocardiography is also helpful for better anatomic definition during preoperative evaluation. Other imaging tools including Magnetic resonance imaging, contrast aortography and CT aortogram have been used as supplemental or confirmatory tests.

Ruptured SOVAs require early surgical intervention since median survival is 3.9 years if untreated.[2] Death is usually due to congestive heart failure. Surgical treatment procedures include primary closure, patch repair or aortic root replacement with or without valve replacement. Primary closure is used for the repair of small SOVAs. Patch closure is preferred for repair of larger SOVAs. The operative mortality rate is 1.9% - 3.6% with 90% survival at 15 years. An alternative to open-heart surgery is percutaneous transcatheter closure of RSOV using different occlusive devices.

CONCLUSIONS

Sudden onset of acute breathlessness in the presence of a continuous murmur should alarm the clinician to suspect a case of a ruptured sinus of Valsalva aneursysm. The fistulous tract will cause left to right shunting and lead to congestive heart failure if not managed timely. Noninvasive imaging like echocardiogram, CT Aortogram and Magnetic resonance imaging can identify the aneurysm and fistulous tract. If left untreated it carries a dire prognosis.

CONFLICT OF INTEREST:

None

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