A Case of Ruptured Sinus of Valsalva Aneurysm

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Abstract

Introduction: Sinus of Valsalva aneurysm (SVA) is an abnormal dilation of one or more aortic sinuses located between the aortic valve annulus and the sinotubular junction. SVA rupture can cause shunting into cardiac chambers creating hemodynamic compromise and has a high morbidity and mortality requiring prompt recognition and treatment.

Case presentation: A 62-year-old male with past medical history of heart failure, COPD, hypertension, hyperlipidemia, schizophrenia, and dementia, presented with a two-day onset of shortness of breath. BNP was elevated and chest X-ray showed bilateral pleural effusions and volume overload. The patient had a TTE (transthoracic echocardiogram) performed on May 2023 which showed normal ejection fraction, diastolic flattening of ventricular septum consistent with RV volume and pressure overload; atrial shunt, moderate TR, moderate free-flowing pericardial effusion; systolic pulmonary artery pressure of 77 mmHg. TEE (transesophageal echocardiogram) was performed and showed a small right-to-left atrial level shunt, a ruptured non-coronary sinus of Valsalva aneurysm into the right atrium with continuous systolic and diastolic left to right shunt on spectral and color Doppler interrogation. During the hospitalization TTE was obtained which re-demonstrated the aneurysmal dilation of the noncoronary sinus of Valsalva with protrusion into the right atrium, associated with rupture and shunting into the right atrium. Patient underwent repair of sinus of Valsalva aneurysm with fistula to right atrium using bovine pericardium patch, repair of atrial septal defect as well as coronary artery bypass grafting and was ultimately discharged.

Conclusion: A ruptured sinus of Valsalva aneurysm is a rare condition which can cause heart failure with volume overload as well hemodynamic compromise. This condition has high morbidity and mortality, therefore must be promptly identified, and treated surgically.

Keywords: Sinus of Valsalva aneurysm, Rupture, Atrial shunt, Echocardiography

Abbreviations: SVA: Sinus of Valsalva Aneurysm; TTE: Transthoracic Echocardiogram; TEE: Transesophageal Echocardiogram

Introduction

A sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly, accounting for only 1% of congenital cardiac anomalies [1]. The condition is described as an abnormal dilation of one or more of the aortic sinuses located between the aortic valve annulus and the sinotubular junction that ensues from the dysplasia of the vascular media [2]. It can mainly arise from a congenital defect of the aortic media or may be acquired following trauma, connective tissue disorders, endocarditis, atherosclerosis, or syphilis [3]. Unruptured sinus of Valsalva aneurysms are usually clinically silent, but greater availability of diagnostic imaging in recent years has led to an increased number being diagnosed incidentally on echocardiography or cardiac MRIs (magnetic resonance imaging) [4]. A SVA can rupture into adjacent cardiac chambers presenting with...
sudden hemodynamic collapse and has a very poor prognosis with high morbidity and mortality and should prompt urgent cardiothoracic evaluation for consideration of repair [5].

We present a case of a patient with acute onset of symptoms due to a ruptured sinus of Valsalva aneurysm.

**Case Presentation**

**History of presentation**

A 62-year-old male with a past medical history of heart failure, COPD, hypertension, hyperlipidemia, schizophrenia and dementia, presented with two-day onset of shortness of breath. The patient reported that the shortness of breath started suddenly, and he felt as if he could not catch his breath. He also endorsed orthopnea, paroxysmal nocturnal dyspnea, and leg swelling. He denied chest pain. It was unclear whether the patient was compliant with his home medications, one of which was Furosemide.

**Investigations**

On physical examination the patient was volume overloaded, with crackles over bilateral lung fields and 1+ pitting edema on bilateral lower extremities, there was a pan systolic murmur over the aortic, tricuspid, and mitral valves.

Laboratory investigations were significant for B-type natriuretic peptide (BNP) of 502 pg/mL (1-100 pg/mL), high sensitivity troponins were elevated and peaked at 277 pg/mL (0-20 pg/mL).

Chest X-ray showed bilateral pleural effusions and volume overload (Figure 1).

Patient had a TTE (transthoracic echocardiogram) performed on May 2023 which showed normal ejection fraction, right ventricle (RV) mildly increased size and normal function, diastolic flattening of ventricular septum consistent with RV volume and pressure overload; atrial shunt, moderate TR with tricuspid regurgitant peak velocity of 4.1 m/sec, moderate free-flowing pericardial effusion; systolic pulmonary artery pressure 77 mmHg (Figure 2). He was scheduled for an outpatient TEE (transesophageal echocardiogram) which was performed in December 2023. It demonstrated a small right-to-left atrial level shunt, a ruptured non-coronary sinus of Valsalva aneurysm into the right atrium with continuous systolic and diastolic left to right shunt on spectral and color Doppler interrogation (Figure 3). Patient was scheduled to follow up in cardiology and cardiothoracic surgery clinic, however he did present to his appointments.

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**Figure 1.** Chest X-ray demonstrating bilateral pleural effusions and volume overload.

**Figure 2.** TEE (midesophageal long axis aortic valve view) demonstrating ruptured non-coronary sinus of Valsalva. NCC: Non-Coronary Cusp; LCC: Left Coronary Cusp; RCC: Right Coronary Cusp.

**Figure 3.** TEE (midesophageal long axis aortic valve view) doppler demonstrating shunting into right atrium.
During the hospitalization a repeat TTE was obtained which re-demonstrated the aneurysmal dilation of the noncoronary sinus of Valsalva with protrusion into the right atrium, associated with rupture and shunting into the right atrium.

Patient underwent left and right heart catheterizations, which showed obstructive CAD (coronary artery disease) with a 40% pLAD (proximal left anterior descending artery), 70% pRCA (proximal right coronary artery) and a diffuse 70-80% mRCA (mid right coronary artery). The LVEDP (left ventricular end diastolic pressure) was 21 mmHg with a mean pulmonary artery pressure of 29 mmHg. A shunt study demonstrated an abnormal 12% step-up increase in oxygen saturation from the inferior vena cava to the right atrium indicating a left to right atrial shunt. Aortogram demonstrated coronary sinus aneurysm with a shunt from the aorta into the right atrium.

Management

Cardiothoracic surgery was consulted, and the patient was prepped and underwent repair of sinus of Valsalva aneurysm with fistula to right atrium using bovine pericardium patch, repair of atrial septal defect as well as coronary artery bypass grafting of the right posterior descending artery using reverse saphenous vein graft. During the surgery chest tubes as well as a temporary pacing wires were placed. Patient was extubated on post operative day 1 and weaned off Dopamine drip. Chest tubes were removed on post operative day 2 and 3 and patient was downgraded from surgical ICU to medical step down. Hospital course was complicated by post operative atrial fibrillation/flutter managed with amiodarone drip.

Repeat TTE showed EF 55-60%, mild mitral regurgitation with small residual flow through the surgical site from the aorta to the right atrium as well as mild regurgitation of the aortic valve and small pericardial effusion.

Outcome

Patient was discharged to a skilled nursing facility with outpatient follow up with cardiology and cardiothoracic surgery.

Discussion

When a right or noncoronary sinus ruptures, a fistula forms between the aorta and either the right atrium or the right ventricular outflow tract, creating a left to right shunt, which can lead to right ventricular overload and right-sided heart failure. Left sinus of Valsalva aneurysm ruptures are clinically less significant, causing communication to the left atrium or left ventricular outflow tract [1-3]. The diagnosis depends on imaging tools such as echocardiography, computed tomographic angiography (CTA), magnetic resonance imaging (MRI), and coronary angiography (CAG). Both TTE and TEE are initial diagnostic tools as they are fast, noninvasive, inexpensive, and real-time ways to assess the characteristics of aneurysmal dilation, cardiac chamber involvement, and cardiac function. Spectral color Doppler imaging is also crucial in aiding the diagnosis as it helps to reveal the persistent turbulent flow between the aneurysm and the receiving chamber [4]. As seen in our case, initial TTE showed an atrial shunt and a TEE with color Doppler visualized the ruptured noncoronary sinus of Valsalva aneurysm into the right atrium with systolic and diastolic flows. Our patient also underwent catheterization which is also commonly conducted, and considered gold standard for the diagnosis of SVA, as it assesses coronary anatomy before surgery and demonstrates elevated PAO2 saturations in the right side of the heart, solidifying the presence of a left to right shunt. Though CTA and MRI can offer precise visualization of cardiac anatomy, their use is limited due to risks of ionizing radiation, cost [4,5].

Once presence of a left to right shunt from a ruptured aneurysm of sinus of Valsalva is diagnosed, urgent surgical evaluation is recommended as mortality rate is alarmingly high. Ruptured SVAs produce serious hemodynamic changes from continuous shunting from the aorta into the receiving chamber, and patients who are left untreated die within one year [6]. Patients presents with acute heart failure symptoms like those seen in our patient and congestive heart failure is known to be the main cause of death [7]. Tricuspid stenosis and regurgitation, as seen in our patient, has also been reported [8]. There have also been cases of right ventricular outflow obstruction, coronary artery compression with infarction, conduction disturbances, endocarditis, and thrombus within the aneurysmal cavity [9]. Based on our patient’s presentation, his sudden decompensation was likely from progression of his rupture leading to worsening shunt due to his lack of prompt follow-up. Surgical interventions have good outcomes with operative mortality being between 1.9% and 3.6%, with a 15-year survival post-procedure being around 90% [10]. In our patient, given the need for a CABG, a surgical approach (as opposed to percutaneous closure repair) was considered ideal as the likelihood of SVA recurrence is relatively uncommon with surgical intervention. Cardiac conduction anomalies can be sometimes seen with surgical intervention as compared to the percutaneous approach, which was seen in our case, as the patient developed atrial flutter post-operation. However, some studies show similar outcomes in both surgical and percutaneous interventions, with both needing for close monitoring [11,12].

Conclusion

A ruptured sinus of Valsalva aneurysm is a rare condition which can cause heart failure with volume overload as well hemodynamic compromise. This condition has high morbidity and mortality, therefore must be promptly identified, and treated surgically.

Disclosures

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Consent

Informed patient consent was obtained prior to starting the case report.

References


