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Unusual Presentation of Heart Failure Secondary to Ruptured Aneurysmal Sinus of Valsalva: A Case Report

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Abstract

Sinus of Valsalva Aneurysm (SOVA) arises from an abnormal dilation of the aortic root, leading to enlargement between the aortic annulus and the sinotubular junction. Although often presenting without symptoms, these aneurysms bear the potential for life-threatening complications, primarily from the looming risk of rupture. We present the case of a 42-year-old Malay gentleman with a history of bilateral pedal edema and dyspnea on exertion who was diagnosed with a ruptured sinus of Valsalva aneurysm. The patient underwent successful surgical repair of the aneurysm, leading to symptomatic improvement and favorable outcomes. This case highlights the importance of early diagnosis and prompt surgical intervention in managing this uncommon condition.

Keywords

Heart Failure, Echocardiogram, Sinus of Valsalva Aneurysm

1. Introduction

Sinus of Valsalva Aneurysm (SOVA) is an enlargement of the aortic root area between the aortic valve annulus and the sinotubular ridge. It occurs due to the weakening of the elastic lamina at the junction of the aortic media and the annulus fibrosis [1]. It is a congenital or acquired cardiac defect that is present in roughly 0.09% [1] of the general population and often presents as an incidental finding during cardiac imaging [2]. Ruptured SOVA is the most feared complication and occurs in 34% of patients [3]. The predominant fistula was from the right sinus of Valsalva to the right ventricle. These can lead to various clinical presentations and potentially life-threatening complications. Surgery is the treat-

ment of choice for ruptured SOVA [3]. We present a case of a patient with a ruptured sinus of Valsalva aneurysm, diagnosed by echocardiogram and successfully managed surgically.

2. Case Presentation

A 42-year-old Malay gentleman presented to the cardiothoracic clinic with a history of bilateral pedal edema and dyspnea on exertion for the past 6 months. He is an ex-smoker, neither diabetic nor hypertensive, and has no history of congenital heart disease. Vital signs were stable, and as follows, blood pressure was 91/52mmHg with a heart rate of 74 bpm. He is afebrile, with a body mass index of 25.5. A physical examination revealed a grade 3 systolic murmur in the pulmonary area.

The electrocardiogram showed a normal sinus rhythm. The chest X-ray revealed cardiomegaly with a clear lung field (Figure 1). Hemoglobin was 13.6. A Transthoracic Echocardiogram (TTE) showed a dilated left ventricle with mildly depressed left ventricular function (LVEF~47%), a dilated right atrium and right ventricle, and a ruptured SOVA of the Right Coronary Cusp (RCC) into the Right Atrium (RA) with a defect size of 0.7 - 0.8 cm. We noted a trileaflet aortic valve with no aortic regurgitation. No atrial or ventricular septal defects were present in the association. We completed all investigations within a month after his first clinic visit.

The patient underwent elective admission for surgical repair of the ruptured sinus of Valsalva the following month. The Transesophageal Echocardiogram (TEE) clearly showed an aneurysm from the RCC that ruptured into the right atrium, creating an aorto-right atrial fistula (Figure 2 and Figure 3). The surgical team resected the fistulous tract and performed PTFE patch closure of the



Figure 1. CXR showed dilated right heart border, cardiomegaly.

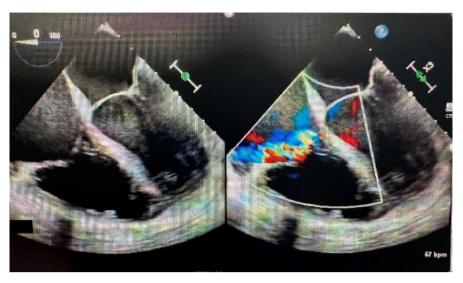


Figure 2. 2D TOE mid esophageal 4 chamber view after induction, dilated right atrium, and right ventricle with slight septal deviation to the left with abnormal jet into right atrium near tricuspid valve.



Figure 3. 2D TOE ME short axis view after induction, clear communication between the sinus of RCC to RA.

RSOV from both sides intraoperatively. The resolution of the left-to-right shunt was demonstrated by intra-operative TEE (**Figure 4**). The patient tolerated the procedure well and was discharged after 7 days of admission. During his outpatient follow-up after 3 months, he indicated the absence of a failure symptom. Repeat transthoracic echocardiography confirmed a successful repair.

3. Discussions

Sinus of Valsalva aneurysm is a rare clinical entity caused by abnormal dilatation of the aortic root located between the aortic valve annulus and the sinotubular junction [1] [2]. The estimated prevalence of SOV stands at 0.09% of the population [1],



Figure 4. 2D transesophageal echo short axis view during coming off bypass after surgical repair, closed communication of right aortic sinus to right atrium, no abnormal color flow.

with a predilection for men, particularly of Asian descent [4] [5]. SOVAs usually affect the sinus of the Right Coronary Cusp (RCC), followed by the Non-Coronary Cusp (NCC), and finally the Left Coronary Cusp (LCC).

SOVA can be either congenital or acquired. Congenital SOVA is commonly associated with Marfan's syndrome or associated with cardiac anomalies, including VSD, bicuspid aortic valve, coarctation of the aorta, patent ductal arteriosus and tricuspid regurgitation. Acquired forms have been associated with connective tissue disease, endocarditis, syphilis and tuberculosis, chest trauma, vasculitis, iatrogenic injury during surgery or secondary to atherosclerosis [2].

Non-ruptured SOVA is usually asymptomatic. However, it can also manifest as atrial fibrillation or a complete heart block. Non-ruptured SOVA can also cause coronary ostia occlusion which frequently presents with acute coronary syndrome or cause a significant aortic regurgitation in 30% to 50% [1] of cases.

Ruptured SOVA with fistula tract formation is a complication that can lead to progressive cardiac failure [6] [7]. Rupture of the right and noncoronary sinuses results in communication between the aorta and either the right atrium or the right ventricular outflow tract, which can lead to right ventricular overload and right-sided heart failure [6]. This patient presented with heart failure symptoms as his RCC SOVA ruptured into the right atrium. Considering the absence of a physical examination pointing towards a connective tissue disorder, along with a negative family history, most likely the patient's aneurysm was congenital.

Several imaging techniques can be employed for diagnosing a SOVA, with or without rupture. The initial imaging methods have been transthoracic and transesophageal echocardiograms. In the scenario of a ruptured SOVA, echocardiographic assessment utilizing color Doppler reveals continuous flow throughout both systole and diastole, given the aorta's high-pressure nature [2]. TOE pre-

cisely defines the location, size, morphology, associated lesions and complications of the defect [8]. Additional or confirming tests have included magnetic resonance imaging, contrast aortography, and Multi-Slice Computed Tomography (MSCT). Magnetic resonance imaging, particularly with multiplanar sequencing, has enabled the assessment of intracardiac shunts in ruptured SOVAs [2].

The unknown natural progression of asymptomatic, unruptured SOVA makes its optimal management unclear [9]. Ruptured SOVA requires early surgical intervention since median survival is 3.9 years if left untreated [10]. Death often results from congestive heart failure. Historically, surgical intervention has been the standard approach for dealing with ruptured SOVAs. However, numerous recent studies have highlighted favorable clinical results through the utilization of transcatheter closure devices, such as septal occluder devices, ductal occluder or Amplatzer vascular plugs offering surgical alternatives but large clinical trials with long-term follow-up are lacking [11] [12] [13]. The optimal decision regarding whether to pursue surgical or percutaneous intervention is best made by a multidisciplinary team, considering factors such as the nature and location of the defect, along with the expertise of the involved cardiac surgeon and interventional cardiologist [7]. Our patient was managed surgically and the outcome was good. This case underscores the importance of early recognition of ruptured SOVA and swift surgical management in addressing this uncommon yet impactful condition.

4. Conclusion

To sum up, while the rupture of a sinus of Valsalva aneurysm is uncommon, it should be considered when a patient presents with failure symptoms like breathlessness, chest pain, and palpitations. Echocardiography is among the imaging methods that can be employed to validate the diagnosis. It is recommended to promptly intervene in cases of ruptured SOVA before symptoms worsen and complications arise.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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