CASE REPORT

Valsalva Aneurysm of Right Sinus: A Case Report and Review of Literature

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Abstract

In this case report, we describe the diagnostic modality of sinus of Valsalva aneurysm (SOVA) in combination with congenital cardiac defect, aortic valve involvement, and conduction abnormality in a 19-year-old patient. Aim of article was to understand the importance of clinicians being cautious about SOVA presenting in young patients, despite cases being rare, and that SOVA requires a thorough SOVA diagnostic approach. We further provide a review of literature highlighting and comparing the treatment options for both unruptured and ruptured SOVAs. The patient presented for examination due to tachycardia and palpitations. A murmur was heard, and the patient was found to have an atrioventricular nodal reentry tachycardia. Echocardiographic evaluation, magnetic resonance imaging, and computed tomography angiography confirmed an aneurysmally dilated aortic root, aortic regurgitation, and ventricular septal defect. Surgical intervention was indicated; however, the patient refused to undergo surgery.

Keywords: sinus of valsalva, aneurysm, treatment, cardiac surgical procedures.

Learning Objectives

- To understand the importance of clinicians being cautious about SOVA presenting in young patients, despite cases being rare
- To understand the SOVA diagnostic approach
- To understand to that timely intervention for SOVA in combination with VSD is imperative, due to likelihood of rupture
- To understand the adequate therapeutic modality for sinus of Valsalva aneurysm coexisting with congenital cardiac defects, aortic valve involvement, or conduction abnormalities

INTRODUCTION

In this case report, we describe the diagnostic modality of sinus of Valsalva aneurysm (SOVA) in combination with congenital cardiac defect, aortic valve involvement, and conduction abnormality in a 19-year-old patient. We further provide a review of literature highlighting and comparing the treatment options for both unruptured and ruptured

SOVAs. The main take-home message from this article is to understand the importance of clinicians being cautious about SOVA presenting in young patients, despite cases being rare, and that SOVA diagnosis requires a thorough approach. Moreover, timely intervention for SOVA in combination with VSD is imperative, due to likelihood of rupture. Understanding the adequate therapeutic modality for sinus of Valsalva aneurysm coexisting with congenital cardiac defects, aortic valve involvement, or conduction abnormalities on a case-by-case basis carries prognostic implications.

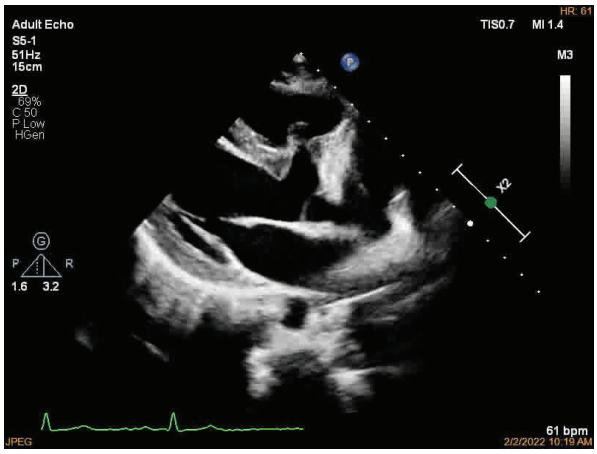

Figure 1. The parasternal long axis view (PLAX).

CASE PRESENTATION

A 19-year-old patient (body weight 70 kg, body height 175 cm) came for an examination due to tachycardia and palpitations, which occurred regardless of effort. A decrescendo diastolic murmur of intensity 2/6, according to Levine, was heard at the left upper sternal border, around the 2nd and 3rd intercostal spaces. Two attacks of atrioventricular nodal reentry tachycardia (AVNRT) with a maximum heart rate of 188 beats per minute were verified on 24-hour electrocardiogram (ECG) Holter monitoring. Per anamnesis, the father died of sudden cardiac death at the age of 52 and the brother at the age of 32. Physical exam was otherwise unremarkable.

On transthoracic echocardiogram (TTE), left and right heart cavities were with regular dimensions, along with a saccular formation that floats in the area of the distal membranous part of the interventricular septum, with an aneurysmally altered aortic root and membranous part of the septum (Figure 1).

Transesophageal echocardiography (TEE) showed aortic valve morphologically three-leaflet (with fused raphe of right and left cusp-functionally bicuspid), with an altered annulus in the form of aneurysm involving the right Valsalva's sinus (covering 30% of the circumference of the annulus), forming a triangular formation measuring 2.7x2.0 cm. On the lateral wall, there were two saccular formations of 1x0.8 cm and 1.1x0.9 cm in dimension, with the effect of filling and emptying during contractions. The entire lateral wall of this formation was of scatter structure. The mentioned formation expanded towards the membranous part of the interventricular septum and formed a channel 1.8 cm long and 0.8 cm wide (Figure 2).

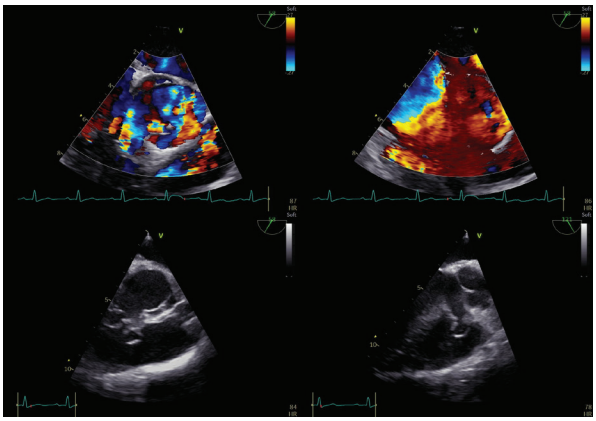

Figure 2. Altered aortic annulus in a form of aneurism, involving the right Valsalva's sinus.

The hemodynamics of the described formation was independent of the pressures in the left and right cavities. The flow from the mentioned aneurysm entered the dilated sinus of Valsalva and communicated with the left ventricle through the formed channel, along with the right ventricle (ventricular septal defect). Moderate aortic regurgitation (AR) of central type was registered, independent of the flow in the described formation, vena contracta was 0.6 cm, and aortic regurgitation/left ventricular outflow tract was 39%. Diastolic flow in descending aorta

was with peak aortic jet velocity (AV Vmax) 0.13 m/s, which corresponded to the picture of mild to moderate AR, along with an empty left atrial appendage (Figure 3). Aneurysmically altered right sinus of Valsalva is verified by aortography, as well as with cardiac magnetic resonance imaging (MRI). Computed tomography (CT) angiography of the aorta confirms the diagnosis (Figure 4). On the CT scan, an aneurysmal expansion consisting of two changes in the saccular form, measuring 9.98x8.2 mm and 10.3x9.4 mm, is described on the lateral wall of a separate compartment (area up to 3.66 cm^2), and the space, in addition to communication with the left ventricle, further communicates with the right ventricle through a tubular form with a diameter of up to 0.78 mm, a length of 18.4 mm. Aortic valve circumference is up to 110.1 mm, and total area is up to 870.1 mm^2.

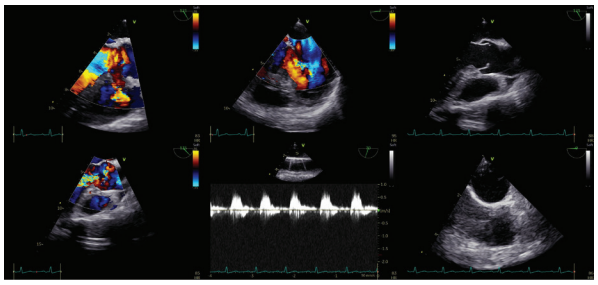

Figure 3. Moderate aortic regurgitation.

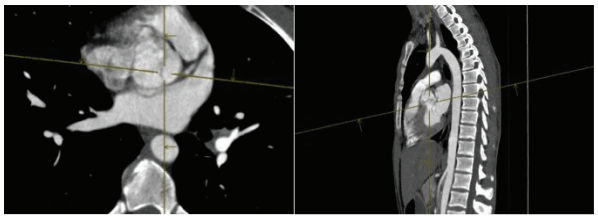

Figure 4. Computed tomography angiography of aorta.

DISCUSSION

The incidence of SOVA is 0.09% percent in the general population, 0.5% - 3% of all congenital heart defects (in 75–90% affected by RCC), occurring four times more often in men and in the Asian population (6). In the majority of cases, SOVA is discovered incidentally during cardiac imaging (2). SOVA represents dilatation of the aortic root located between the aortic valve annulus and the sinotubular junction and may be associated with the existence of connective tissue diseases (6). Our patient had no history of infections associated with acquired SOVA (syphilis, bacterial endocarditis, and tuberculosis), no Marfan syndrome or Ehlers-Danlos syndrome (or any connective tissue disorder) in the family, and the patient's rheumatological and ophthalmological findings were neat.

In 30% to 50% of cases it is associated with aortic regurgitation, VSD is associated with aneurysm in 30-60% (most often with bicuspid AV) (6). Supracrystalline VSD (frequent rupture) is more common in Asians, and perimembranous in Western populations (7). Our patient presents with an aneurysm of the right sinus of Valsalva with perimembranous VSD, functionally bicuspid AV, and moderate AR, which is an uncommon combination of congenital cardiac abnormalities, whereby the role of timing of surgery is of utmost importance.

VSD is associated with aneurysm in 30-60% (most often with bicuspid AV), and the association with VSD brings a higher risk of rupture (4). Rupture can occur spontaneously due to trauma or endocarditis (in 60% of RV); most often sudden, in previously undiagnosed cases, before the age of 40 (4,6). Also, the occurrence of rhythm disorders is something that should be taken into account. Our patient also has verified AVNRT, which represents an additional problem in understanding the therapeutic modality.

An important consideration is that it may be challenging to differentiate the sinus of Valsalva aneurysm associated with perimembranous VSD from membranous septal aneurysm, owing to the close anatomical location.

Surgical treatment of SOVA is widely acknowledged and associated with low mortality, the choice of technique depending on the presence of VSD (8). Operative treatment goes in two directions. The first option is transcatheter closure (Rashkind umbrella, septal occluder device, ductal occluder,

Table 1: Case reports of SOVA originating from right coronary sinus with associated perimembranous VSD and aortic valve disease in young adults (age group 18-25 years old)

Reference	Age, sex	Presentation	SOVA	Associated conditions	Mode of diagnosis	Changes on ECHO	Treatment	Outcome
Udora et al. (2023) (1)	24-year-old, female	Chest pain, SOB, generalized body swelling, weight loss, cough, paroxysmal nocturnal dyspnea, orthopnea, palpitation, dizziness; grade 4/6 pansystolic murmur at the left mid-sternal border, loud, nonsplitting pulmonary component of S2	Unruptured	Grade 2 AR, bidirectional perimembranous VSD 1.1 cm	TTE	N/A	N/A	N/A
Mhanna et al. (2022) (2)	23-year-old, female	Palpitations, exertional dyspnea; continuous murmur; HR 110 bpm	Ruptured	AR	TEE	Ruptured 1.8 cm SOVA of the non-coronary cusp to the right ventricle, with significant left-to-right shunt and pulmonary hypertension	Surgical correction	Significant resolution of the shunt, pulmonary pressure was normalized
Kumar et al. (2016) (3)	24-year-old, male	Gradually progressive SOB and palpitations on exertion; grade 3/4 early diastolic murmur over the aortic area, wide pulse pressure with presence of peripheral aortic run off; two previous syncope episodes	Unruptured, dissecting into IVS	Severe AR, perimembranous VSD	TTE TEE	LVEF 40%, LVEDD of 5.4 cm, LVESD of 4.0 cm; aortic annulus dilated 3 cm; severe AR without evidence of leak into any of the chambers	Repair and closure of the right SOVA with Dacron patch	Completely healed and thrombosed aneurysmal sac within the IVS
Hyung Rae Kim et al. (2015) (4)	20-year-old, male	Chest pain, HF symptoms, history of VSD	Ruptured	Perimembranous VSD 1 cm	TTE Exercise treadmill test TEE	LVEDD of 52 mm, LVESD of 31 mm, Qp/Qs 1.2; RV enlargement with depressed function, preserved LV function; elongated right SOVA with ruptured tip	Open heart surgery - ruptured aneurysm and perimembranous VSD were closed with Dacron patch	Uneventful postop period, without residual shunt
Kumar et al. (2015) (5)	21-year-old, male	Progressively worsening effort dyspnea, long early diastolic murmur heard best over the third left intercostal space, prolonged PR interval	Ruptured, through the IVS into LV cavity	AR, cardiac conduction abnormality	TTE Right heart catheterization Aortic root angiography	Ruptured aneurysm with flow arising from the right sinus of Valsalva, traversing the IVS into the LV; LV dilatation and global hypokinesia	Right aortic sinus defect was successfully repaired with a Dacron patch	Uneventful postop period, symptomatically improved

AR: aortic regurgitation; HR: heart rate; HF: heart failure; IVS: interventricular septum; LV: left ventricle; LVEF: left ventricular ejection fraction; LVEDD: left ventricular end-diastolic dimension; LVESD: left ventricular end-systolic dimension; N/A: not available; RV: right ventricle; SOB: shortness of breath; SOVA: sinus of Valsalva aneurysm; TEE: transesophageal echocardiography; TTE: transthoracic echocardiography; VSD: ventricular septal defect

Amplatzer vascular plug (AVP II)), which has emerged as an effective alternative in carefully selected patients, revealing promising results (9). However, it is worth noting that, due to the thickness of the non-coronary cusp (NCC) and its proximity to the AV conduction system, occurrence of complete heart block after transcatheter closure of perimembranous VSD has been reported as a perturbing complication; however, a good response to high-dose steroids has been observed in such cases (9). Another option is surgical intervention, in case of rupture or without rupture, if there is a ventricular septal defect or significant aortic valve regurgitation (6). Options for surgical treatment

are the Bentall procedure, or preserving the valve with an aortic valve suture annuloplasty technique involving external stabilization of the annulus following the remodeling of the aortic root, although there is no clear evidence that preserving the valve should be an option before the classic replacement of both the aortic root and valve (10). In current practice, for specific isolated ruptured sinus of Valsalva aneurysm cases or those including a combination with perimembranous VSD, percutaneous closure employing modified double-disk occluders (which are easily retrievable, repositioned, flexible, non-space occupying, without affecting aortic valve function) is a compelling alternative to surgery; in fact, percutaneous closure has been found to be more therapeutically advantageous than surgical closure due to the avoidance of extracorporeal circulation, intensive care unit stay, and blood transfusion, as well as a shorter hospital stay and minimal invasiveness (10). In patients with mechanical aortic valve, transcatheter closure of ruptured sinus of Valsalva aneurysm with double-disc (shorter in length) perimembranous VSD occlude is a viable method for successfully repairing ruptured sinus of Valsalva aneurysms; compared with the Amplatzer Duct Occluder, thus avoiding iatrogenic right ventricular outflow tract (RVOT) obstruction (8,9). On the whole, ruptured SOVA has a poor prognosis with high mortality (3.6%) unless timely intervention is undertaken (6).

CONCLUSION

Clinicians ought to remain cautious about sinus of Valsalva aneurysm in young patients presenting with signs and symptoms of cardiac conduction abnormalities or myocardial ischemia and heart failure. The most conspicuous physical finding is a new continuous murmur, prompting urgent echocardiography to enable timely diagnosis and treatment of the aneurysm. Unruptured aneurysm of the right coronary sinus of Valsalva cases in young patients are rare and detected incidentally. Surgery is indicated when SOVA is in conjunction with a VSD and aortic valve involvement, due to likelihood of rupture. In the event of a ruptured SOVA with perimembranous VSD, treatment modalities include not only surgical repair, but also transcatheter closure in carefully selected patients. Yet, the prognosis is poor if no prompt diagnosis is made.

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