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Research Article

Ruptured Sinus of Valsalva Aneurysm from Left Coronary Sinus with Different Causes

Kang R[1#], Han M[2#] and Zhang C[1*]

[1]Department of Ultrasound, The Second Affiliated Hospital of Nanchang University, Nanchang, Jiangxi, China

[2]Queen Mary School, Nanchang University, Nanchang, Jiangxi, China

[#]Contributed Equally to this Work and regarded as Co-First Authors

***Corresponding author:** Chunquan Zhang, Department of Ultrasound, The Second Affiliated Hospital of Nanchang University, No.1 Minde Road, Nanchang, Jiangxi, 330006, China

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Abstract

Objective: Analysis of three cases in which the clinical diagnosis was ruptured sinus of Valsalva aneurysm (RSVA) from left coronary sinus, combined with similar cases in the literature, to summarize the etiology of RSVA.

Methods: Retrospective analysis of three cases of RSVA diagnosed in our hospital from 2009 to 2021. Their general condition, examination and treatment were analyzed, and the etiology of the sinus of Valsalva aneurysm (SVA) was summarized in relation to similar cases reported in the literature.

Results: All three cases were diagnosed as RSVA from left coronary sinus with a clear etiology of congenital SVA, Behcet's disease and infective endocarditis, respectively. They all underwent successful surgical treatment. In the literature, there were 12 cases of SVA from left coronary sinus of definite etiology, 10 of which ruptured.

Conclusions: SVA from left coronary sinus is a rare disease with a complex etiology, and RSVA is one of the most serious complications of SVA. Exploring the etiology of SVA will help to prevent the serious complications of RSVA by targeting the etiology for treatment and timely surgical intervention.

Keywords: Sinus of Valsalva aneurysm; Left sinus; Ruptured; Etiology; Behcet's disease; Infective endocarditis

Abbreviations

RSVA: Ruptured Sinus of Valsalva Aneurysm; SVA: Sinus of Valsalva Aneurysm; IE: Infective Endocarditis; BD: Behcet's Disease; BP: Blood Pressure; HR: Heart Rate; RR: Respiratory Rate; ESR: Erythrocyte Sedimentation Rate; B-BNP: B-Type Brain Natriuretic Peptide; ANA: Anti Nuclear Antibody; anti-dsDNA: Antibody, Anti Double Strand DNA Antibody; TTE: Transthoracic Echocardiography; LV: Left Ventricle; LVOT: Left Ventricular Outflow Tract; LA: Left Atrium; CTA: Computed Tomography Angiography; AR: Aortic Regurgitation

Introduction

Sinus of Valsalva Aneurysm (SVA) is an unusual aortic root defect that can be dangerous due to its serious complications; it is defined as dilatation of one or more of the aortic valve sinuses [1]. According to Meier JH et al., SVA commonly arise from the right sinus, (65%~85%), less commonly from noncoronary sinus (10%~30%), and rarely (<5%) from the left sinus [2]. It can be either a congenital or acquired cardiac anomaly [3], and is relatively common in oriental patients4.Congenital SVA is owing to a dilatation generally of a single sinus of Valsalva caused by a separation between the aortic media and the annulus fibrosus, and often with a deficiency of the normal elastic tissue and abnormal development of the bulbus cordis [5]. Congenital SVA is usually seen in patients with Marfan or Ehlers-Danlos syndrome [6]. Acquired SVA is mainly the results of infective endocarditis (IE), syphilis, trauma, Behcet's disease (BD), atherosclerosis, cystic medionecrosis [7,8].

Although surgical repairs for ruptured sinus of valsalva aneurysm (RSVA) are gradually matured, RSVA is still considered as a special and fetal complication of SVA with severe mortality and morbidit [5]. So it is more revelatory that we focus on the etiology of RSVA from left coronary sinus, for this rare situation is easily neglected and usually fetal. However, there are few cases of RSVA from left coronary sinus and fewer studies on the etiology of SVA at present. Therefore, this paper retrospectively analyzed and reported 3 cases of RSVA from left coronary sinus that occurred in our hospital from 2009 to 2021. 12 cases of SVA from left coronary sinus with a clear cause were also obtained from the literature. And the etiology of SVA was summarized in relation to similar cases reported in the literature.

Our cases reported in this article are remarkable for several reasons: Etiology of SVA, site of rupture, precise preoperative diagnosis and the successful surgical management. By exploring the etiology of SVA will help to prevent the serious complications of RSVA by targeting the etiology for treatment and timely surgical intervention.

Materials and Methods

We retrospectively analyzed three patients with RSVA from 2009 to 2021 IN our hospital and obtained their preoperative, intraoperative, and postoperative conditions. Also, by reviewing the literature, we obtained 12 cases of SVA from left coronary sinus with clear causes [9-20]. We also retrospectively analyzed the etiology of these 12 patients.

Results

See Table 1 and 2.

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Table 1: We got the preoperative, intraoperative and postoperative conditions of these 3 cases.

	Case 1	Case 2	Case 3
Gender	Male	Female	Male
Age	25	53	53
Reasons for admission	Persistent chest and back pain for 2 years	Sudden chest tightness and palpitation for one week	Chest tightness and shortness of breath with cough for half a month
Past medical history	No special	Recurrent oral and vulvar ulcers.	Unexplained fever
BP/HR/RR	90/30 mmHg	160/110 mmHg	135/95 mmHg
	98 beats/min	100 beats/min	100 beats/min
	22 beats/min	20 beats/min	21 beats/min
Chest physical examination	A 2/6 grade systolic murmur and moderate diastolic murmur.	Femoral artery pistol shot sound and corrigan's pulse.	A grade 2/6 systolic blowing murmur.
Abnormal laboratory examination	ESR was 53mm/h.	ANA, anti dsDNA were negative.	B-BNP was 604 pg/ml.
Electrocardiogram	Sinus heart rate	Sinus heart rate	Prolonged P-R interval.
Preoperative TTE	SVA from left coronary sinus ruptured into LVOT	SVA from left coronary sinus ruptured into LV.	A neoplasm on the aortic valve. SVA from left coronary ruptured into LA.
Preoperative CTA	An aneurysmal expansion of the left coronary sinus.	An aneurysm of the left coronary sinus at the initial segment of the ascending aorta.	This examination was not done.
Surgical approach	Ruptured aortic sinus aneurysm repair	Congenital aortic sinus aneurysm repair and aortic valvuloplasty	Aortic coronary sinus aneurysm repair and aortic valve replacement
Surgery detection	A breaking of approximately 2.0x1.0 cm in size below the left coronary artery.	Significant thickening of the aortic wall, showing changes of aortitis	A rupture of the left coronary valve of the aorta was seen, and a neoplasm was seen on it.
Pathological results/ Culture results	A pseudoaneurysm in LVOT	Arterial vascular wall fibrosis and vitreous changes.	Bacterial cultures of both blood and valve resulted in Staphylococcus aureus.
Postoperative TTE	The saccular structure had disappeared.	The saccular structure had disappeared.	The saccular structure had disappeared. Enlargement of LA and LV.
Location of rupture into the heart cavity	LVOT	LV	LA
Cause of disease	Congenital SVA	Behcet's disease	Infective Endocarditis

Table 2: A review of the literature revealed 12 cases of left Valsalva sinus aneurysm with a definite cause. The order in the references corresponds to 9-20.

Author	Age	Gender	Causes of Disease	Location of Rupture into the Heart Cavity
Killen DA, et al. [9]	62	Male	Congenital SVA	Transverse pericardial sinus
Ryan T, et al. [10]	33	Male	Sepsis with *Staphylococcus aureus* after cadaveric renal transplantation	LA
Rothbart RM, Chahine RA [11]	42	Male	Infective Endocarditis	LVOT
Greiss I, et al. [12]	56	Female	Head-on car accident	The pericardial space near LA
Saito T, et al. [13]	59	Male	Infective endocarditis	LV
Fazio G, et al. [14]	30	Male	HIV	Pulmonary artery
Ryomoto M, et al. [15]	47	Male	Congenital SVA	LV
Kawamura A, et al. [16]	29	Female	Takayasu arteritis	LV
Bakan S, et al. [17]	30	Male	Behcet's disease	Unruptured
Bae K, et al. [18]	44	Male	Infective endocarditis	LV
Chamsi-Pasha MA, Lawrie GM [19]	42	Female	Marfan syndrome	Unruptured
Talwar S, et al. [20]	35	Male	Congenital SVA	LVOT

BP: Blood Pressure; HR: Heart Rate; RR: Respiratory Rate; ESR: Erythrocyte Sedimentation Rate; B-BNP: B-Type Brain Natriuretic Peptide; ANA: Anti Nuclear Antibody; anti-dsDNA: Antibody, Anti Double Strand DNA antibody; TTE: Transthoracic Echocardiography; LV: Left Ventricle; LVOT: Left Ventricular Outflow Tract; LA: Left Atrium; CTA: Computed Tomography Angiography.

Discussion

SVA is usually thought to be an enlargement or a dilation of one sinus of the aortic root between the aortic valve annulus and the sinotubular ridge [5]. It is an unusual cardiac event with no noticeable symptoms, which poses a challenge for the early diagnosis and treatment of SVA. In the overwhelming majority of SVA cases (90%), the sinus ruptures into the right-sided chambers, and in the other 10%, it ruptures into the left atrium, left ventricle, pulmonary artery, or pericardial cavity [21]. Early diagnosis of SVA is important because surgical treatment is easier to perform at an early stage and facilitates a good prognosis [22]. RSVA is a hazardous complication that can cause AR, left to right shunt following the aneurysm rupture into a cardiac chamber, and acute progressive heart failure [23].

The natural history of SVA is undiscovered; it can be either a congenital or acquired cardiac anomaly [3]. Congenital SVA is usually thought to be the result of incomplete fusion of the two halves of the

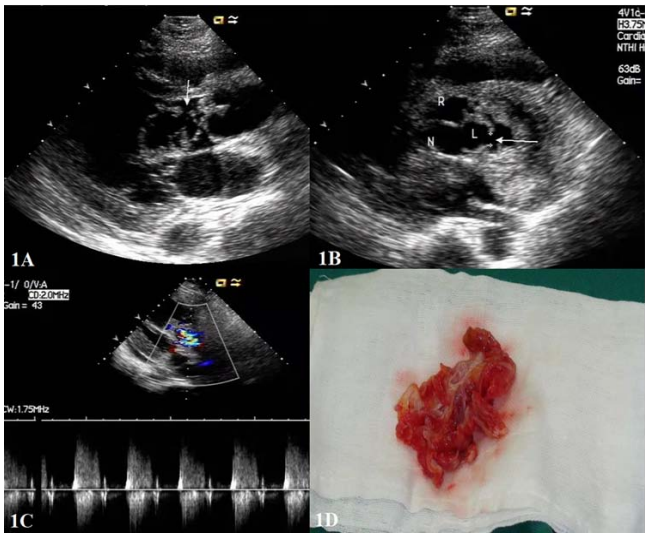

Figure 1: A: left ventricular long-axis view showed the saccular structure (SVA) in LVOT and the breach on the side of the ventricular septum; B: short-axis view of the aorta showed the entrance diameter of the saccular structure (SVA) is about 6mm; C: the multicolored blood flow signal flows from the saccular structure to LVOT, and its blood flow velocity was about 4m/s as measured by spectral Doppler; D: ruptured aortic sinus removed during surgery.

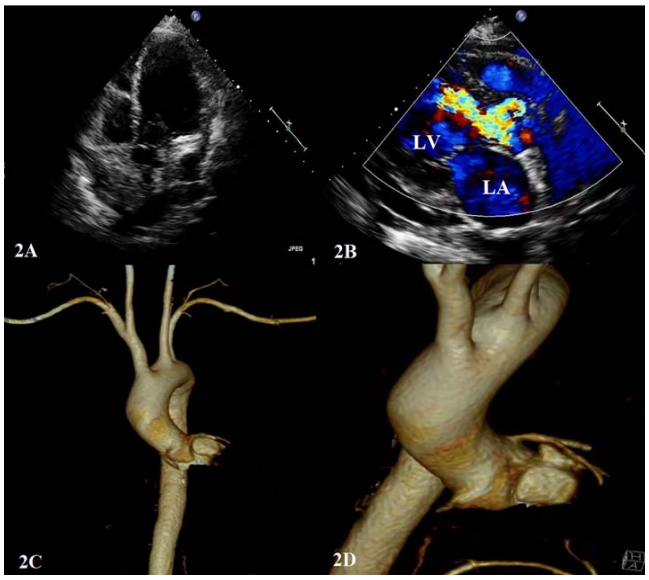

Figure 2: A: SVA from left coronary sinus rupturing into LV (arrow); B: blood flow signal from left coronary sinus to LV; C and D: CTA images of patients.

distal bulbar septum [24-26]. This is the structure that separates the aortic and pulmonic halves of the bulbus cordis, the primary exit tube of the fetal heart. Congenital weakness secondary to an incompletely fused septum predisposes the high-pressure area of the sinuses to aneurysm formation. The left coronary sinus does not arise from the distal bulbar septum, as do the right and noncoronary sinuses [24-26]. This interprets the infrequency of congenital SVA from the left sinus [26]. Case 1 is considered as a congenital SVA from the left sinus that ruptured into the LVOT. Rupture of a congenital SVA into an adjacent chamber may occur spontaneously, after trauma, after

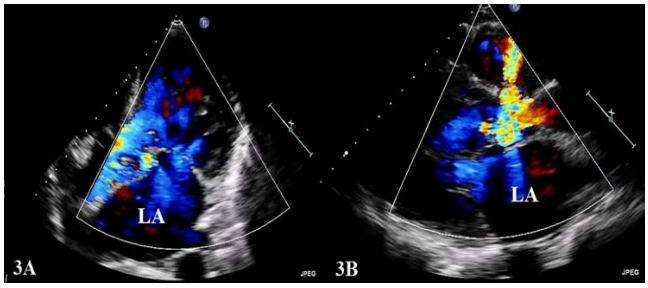

Figure 3: A and B: CDFI shows the blood flow signal from left coronary sinus to LA.

strenuous physical exertion, or from acute bacterial endocarditis.

Acquired SVA are mainly the results of infective endocarditis (IE), syphilis, trauma, Behcet's disease (BD), atherosclerosis, cystic medionecrosis [7,8], or degenerative diseases, abuse of drugs or alcoholism [27]. In case 2, the aortic wall was seen to be significantly thickened with aortitis changes under surgery. And the patient had a 5-year history of oral and vulvar ulcers, and clinical examination revealed aphthous stomatitis, genital ulceration, and erythema nodosum bilaterally, which could provide clues to differentiate diagnosis. BD is a systemic disorder of recurrent acute inflammation, characterized by major symptoms of oral aphthous ulcers, uveitis, skin lesions and genital ulcers [28]. Involvement of the heart is called Cardio-BD, cardiac manifestations include pericarditis, myocarditis, endocarditis, endomyocardial fibrosis, atrial fibrillation, ventricular arrhythmias, coronary arteritis, acute myocardial infarction, and dilated cardiomyopathy [29]. James et al. proposed that SVA was the main causes of death in patients with BD [30,31]. There is no doubt that surgical treatment is necessary, otherwise it may incur fetal complication. However, treatments that aim directly at the etiology of SVA are the most important.

In case 3, TTE suggested a rupture into LA. Under surgery, a neoplasm was seen on the left coronary valve of the aorta, and the blood and valve culture results were positive for *staphylococcus aureus*, suggesting that IE might be the cause of this patient. IE appears worldwide, and is defined by infection of a native or prosthetic heart valve, the endocardial surface, or an indwelling cardiac device [32]. *Staphylococcus aureus* is the most commonly isolated microorganism associated with infective endocarditis in high-income countries and is reported in up to 30% of cases [33,34].

Regardless of manifestation, patients with a persistent unexplained bacteremia should be investigated for infective endocarditis, especially patients with *Staphylococcus aureus* infection, should be examined with echocardiography [32]. Clinical examination presents alterable signs of disease, with fever (present in about 90% of cases) and a cardiac murmur (in about 85%) being most common. If pathology specimens are accessible (from surgery or autopsy), the diagnosis of IE can be made by histology or positive culture of vegetation or abscess tissue [35].

In summary, discussing the etiology of SVA from the left sinus will help early diagnosis of this rare cardiac anomaly and early treatment of the cause, so as to effectively prevent the serious complication of RSVA.

Conclusion

SVA from left coronary sinus is a very rare disease with multiple etiologies. It can be dangerous due to its serious complications like RSVA. Although surgical repair is gradually matured, it is still considered as a special and fetal complication of SVA. In addition to timely surgical treatment, early treatment for the etiology is also critical. The exploration of etiology of SVA can help early detection of SVA, so that timely surgical treatment can be carried out to prevent serious complication of RSVA.

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