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CASE REPORT

Ruptured Aneurysm of Sinus of the Valsalva: Case Series

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ABSTRACT

Introduction: We present three cases of ruptured sinus of Valsalva aneurysm concomitant with a cardiac lesion in 2 cases of ventricular septal defect and 1 case of bicuspid aortic valve with mild aortic insufficiency. **Case presentation:** In 2/3 cases, the site of ruptured sinus of Valsalva aneurysm originated from right coronary cusps and one from non-coronary cusps. Both ruptured sinus of Valsalva aneurysm from right coronary cusps communicated to right ventricle, and one that originated from non-coronary cusps ruptured to right atrium. All cases were diagnosed by transthoracic echocardiography and confirmed by transesophageal echocardiography. Two underwent the surgery, while one did not want to continue treatment. **Conclusion:** ruptured sinus of Valsalva aneurysm is a rare cardiac anomaly. In the Eastern countries, the most common associated anomalies with ruptured sinus of Valsalva aneurysm are ventricular septal defect and aortic regurgitation. Ruptured sinus of Valsalva aneurysm mainly originates from right coronary cusps and is ruptured into right ventricle. Transthoracic echocardiography and transesophageal echocardiography are employed to confirm the diagnosis. In this cohort, the coexistences of ruptured sinus of Valsalva aneurysm with ventricular septal defect and aortic regurgitation (case 1), ventricular septal defect (case 2), aortic regurgitation and bicuspid aortic valve (case 3) are observed.

INTRODUCTION

Sinus of Valsalva is a rare acquired or congenital cardiac anomaly with an incidence of 0.1–3.5% in all congenital heart defects [1]. The acquired aneurysm is caused by conditions affecting the aortic wall, such as infections, trauma, and degenerative disease. The congenital aneurysm is common and is often caused by weakness at the junction of aortic media and annulus fibrosis. An aneurysm may be non-coronary sinus (10–30%) or may have originated in the right coronary sinus (65–85%); it is rarely observed in left coronary sinus (1–5%) (2).

In the current study, the non-coronary sinus is observed in case 3, whereas the right coronary sinus is presented in 2 cases. Usually, the aneurysms remain asymptomatic unless complicated by rupture [2]. Reportedly, the aneurysms of the sinus of Valsalva rupture suddenly in 40% of patients presented with palpitation, fatigue, chest pain, dyspnea, and cardiac insufficiency.

In the current case series, we present three cases of ruptured sinus of Valsalva aneurysm (RSVA) concomitant with a cardiac lesion, i.e., 2 cases of ventricular septal defect (VSD) and 1 case of bicuspid aortic valve (BAV) with mild aortic insufficiency (AI).

CASE PRESENTATION

CASE 1

A 31-year-old male was admitted to our hospital because of slight fatigue and dyspnea since childhood. He had a history of VSD that was diagnosed by cardiac angiography at the age of 16 years but had not been followed up. At the time of admission, a grade 4/6 systole-diastolic murmur was audible near the left sternal border. The electrocardiogram (ECG) showed a normal rhythm. Transthoracic echocardiography (TTE) showed 60% left ventricular ejection fraction (LVEF), mild dilatation of the left ventricle (LV) with normal systolic function, and a systole-diastolic flow between aorta and right ventricle (RV) without diastolic

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accentuation. In addition, the patient had dilated RV, non-dilated coronary arteries, tricuspid aortic valve, and dilatation and defect in the aortic root. Moreover, transesophageal echocardiography (TEE) showed aneurysm of sinus Valsalva (size 42 mm). The site of rupture was in the right coronary cusps (RCC) below the origin of right coronary artery (RCA) that communicated into the RV, resulting in moderate aortic regurgitation. Finally, surgical repair was carried out. Subsequently, the RSVA and aortic valve were repaired. The VSD (perimembranous type) was also repaired, following which the patient made an uneventful recovery.

CASE 2

A 33-year-old woman was presented to the outpatient clinic with a complaint of gradually developed dyspnea of exertional function class II-III. A history of palpitation and dyspnea were noted since childhood that was diagnosed as VSD and had not been followed up. Physical examination detected a continuous murmur in the left sternal border, and a precordial thrill was palpable. Initially, the diagnosis of fistula between RCA and RV was considered. However, she was referred to our center (Tehran Heart Center), Tehram, Iran for further evaluation. TTE revealed that left atrium (LA) and LV were mildly dilated, mild right atrial and right ventricular dilatation exhibited mild tricuspid regurgitation (TR), and pulmonary artery was significantly dilated (47 mm). The sinus of Valsalva was dilated (41 mm), while the origin of the left main coronary artery and RCA were found normal. No diastolic accentuation was detected in the turbulent flow that was opposite to the diagnosis of fistula, and the rupture was seen in the sinus of Valsalva below the origin of RCA from the RCC (Figure 1) (Qp/Qs=1.9). The LVEF was about 55–60%, and RV systolic function was adequate. The diagnosis was confirmed by TEE. She underwent cardiac catheterization, which confirmed the echo findings. The patient was operated electively. The surgery consisted of a patch repair of ruptured aneurysm and VSD. Subsequently, the patient's blood pressure, heart rate, and rhythms were stable, but after 2 days, the general condition of the patient deteriorated and she died after 2 weeks.

CASE 3

A 24-year-old woman exhibited gradually increasing dyspnea on exertion function class III and palpitation for the last 2 years. VSD was detected during her second month of pregnancy. No history of angina, syncope, or rheumatic fever was observed. At auscultation, a continuous murmur was heard at the left lower sternal border, and ECG showed normal sinus rhythm. TTE showed a mildly dilated LV, and the global systolic function was normal. LA was normal in size, and RV was mildly dilated. The LVEF was 55%, the aortic valve was bicuspid with mild regurgitation, and rupture of sinus of Valsalva from non-coronary cusp (NCC) into the right atrium with continuous flow was detected. Moderate TR and pulmonary artery pressure of about 50 mmHg were observed. Although cardiac catheterization and surgery were recommended, the patient did not want to continue treatment.

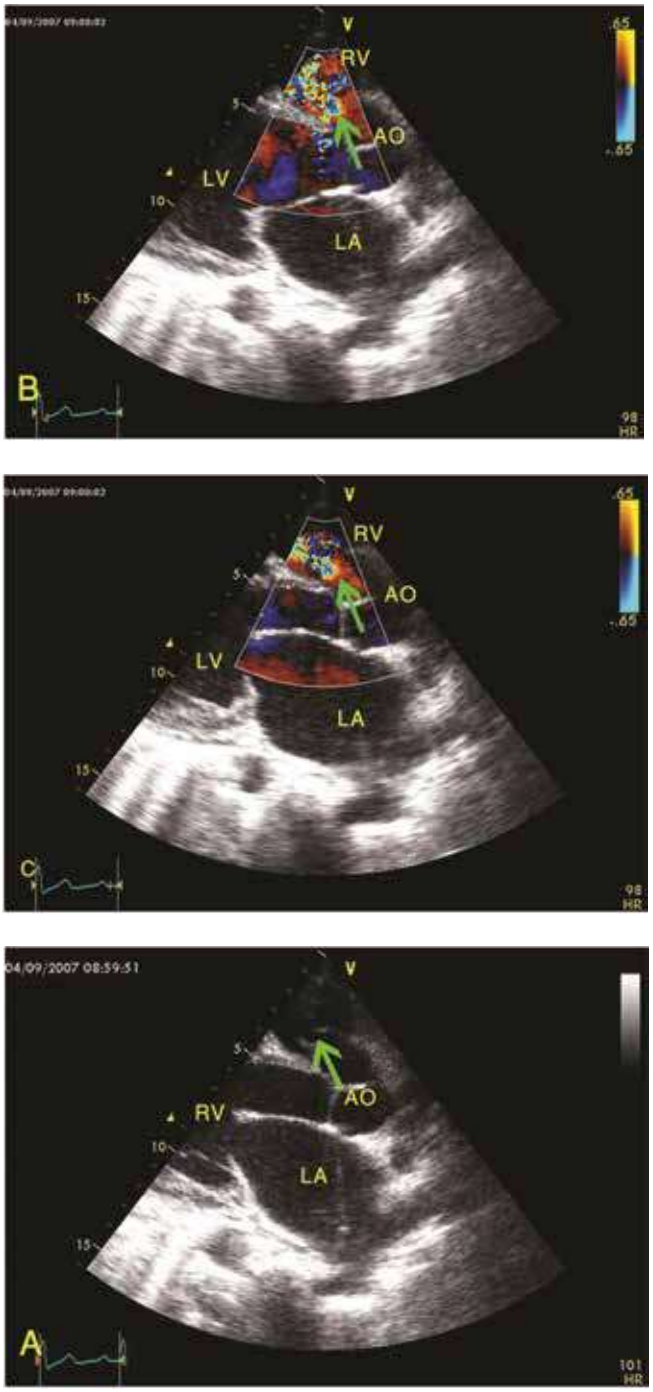

Figure 1. TTE shows RSVA (A), there is systolic (B) and diastolic (C) turbulent flow from aorta towards RV through the ruptured aneurysm.

Abbreviations; TTE: Transthoracic echocardiography, RSVA: ruptured sinus of Valsalva aneurysm, RV: right ventricle

DISCUSSION

In the current study, all the patients with RSVA had gradual developing symptoms, and their diagnosis was mainly based on TTE confirmed by TEE. The incidence of RSVA was much higher in the Asian population (1.2–4.94%) than in the Western population (0.5–1.5%) [3]. The rupture of sinus of Valsalva is frequently associated with other congenital defects, especially VSD, aortic valve regurgitation, and BAV [4]. The associated VSD is common in the Asian population, BAV is frequently seen in the Western population, and the frequency of AI does not differ significantly between the two groups (Asian vs. Western population; VSD: 52.4% vs. 37.5%, aortic regurgitation (AR): 33.6% vs. 32.7%, BAV: 0.6% vs. 7.8%). In this report, two patients had VSD, one had BAV, and 2 showed AI. RSVA originated from any of the three cusps. In Eastern countries, the origin of RSVA from RCC is high, and 72.5% ruptured into RV [3]; in the current study, two patients presented aneurysm in RCC that ruptured in RV. Occasionally, the ruptured aneurysm is from NCC [5]. The physiological consequences of a rupture depend mainly on the rapidity of the rupture [6]. The principal VSD associated with RSVA is the supra crystal type [2], but in two of the current cases, perimembranous VSD was recognized at the time of surgery. The aortic valve abnormalities and incompetence are common in patient with RSVA with influence on prognosis. VSD is a predisposing factor for AR. The coexistence of subarterial VSD with prolapsed aortic cusps could cause AR with a lack of supporting tissue. A second mechanism is the Bernoulli Effect during systole, which is related to pulling the related cusp into VSD; similar to this mechanism, the shunt flow through the ruptured site of sinus Valsalva pulled the cusp of the aortic valve in the diastole [7]. If the associated AR is mild, no surgical intervention may be necessary. However, most patients with moderate to severe AR required surgical intervention. AR resulting from cusp prolapse associated with VSD can be repaired by correcting the rupture of sinus Valsalva and VSD or valvuloplasty. TTE and TEE and spectral Doppler interrogation frequently established the diagnosis [4]. The advances in echocardiography have reduced the need for invasive angiography when evaluating Sinus of Valsalva Aneurysm . These tools allow accurate determination of the following: size of sinus, sinuses of origin, communicating chamber, the degree of valve regurgitation, and associated anomalies [1]. In our patients, the diagnosis was made by TTE and confirmed by TEE.

Catheterization is currently reserved to evaluate coronary anatomy but is being established for the diagnosis and evaluation of the hemodynamic effects of ruptured aneurysms. Early aggressive intervention is recommended to prevent endocarditis or enlargement of the ruptured aneurysms [4].

RSVA or unruptured sinus Valsalva, concomitant with VSD or aortic valve regurgitation or both, a huge sinus Valsalva causing mitral valve incompetence, right ventricular outflow obstruction, and myocardial ischemia should be surgically repaired as soon as the diagnosis is confirmed [3]. The prognosis after the surgical repair of SVA is satisfactory. Early diagnosis and surgical intervention are the optimal ways to improve survival [7]. Preoperative mortality was 7% and prolonged survival was 63% in one large series [4]. One of our cases is surviving after surgery, one did not undergo operation, and one died 2 days after the operation.

CONCLUSION

RSVA is a rare cardiac anomaly. In the eastern countries, the most common associated anomalies with RSVA are VSD and AR. RSVA mainly originates from RCC, and are ruptured into RV. TTE and TEE are employed to confirm the diagnosis. In this cohort, the coexistences of RSVA with VSD and AR (case 1), VSD (case 2), AR and BAV (case 3) are observed.

PATIENTS PROSPECTIVE

All the patients with RSVA may have gradual developing symptoms, and their diagnosis can be mainly based on TTE, which is confirmed by TEE.

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AUTHOR CONTRIBUTIONS

HS and AS interpreted and managed patient data, wrote the manuscript, and structured the figures. All authors read and approved the final manuscript.

CONFLICT OF INTERESTS

The authors declare that they have no competing interests

ETHICAL STANDARDS

Written informed consent was obtained from all three patients for publication of the case reports and accompanying images.

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