

Sinus of Valsalva aneurysm dissecting the interventricular septum with rupture into the right ventricle (RCD code: I-1B.O)

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Abstract

Sinus of Valsalva aneurysm (SVA) is a rare congenital heart disease. A possible complication of SVA is rupture into a heart chamber, causing volume overload, which could lead to heart failure and the requirement for surgical correction. Transthoracic echocardiography is a readily available diagnostic tool for assessment, while other methods of cardiac imaging (cardiac CT and MRI) confirm the diagnosis and are necessary for preoperative assessment of the pathology. We present a very rare case of right sinus of Valsalva aneurysm dissecting the interventricular septum which ruptured into the right ventricle, causing volume overload of the right heart chambers and progressive heart failure. The diagnosis was established by transthoracic echocardiography and confirmed by ECG-gated CT. Successful surgical correction resulted in regression of dilatation and dysfunction of heart chambers and to functional recovery of the patient. JRC D 2019; 4 (3): 51–54

Key words: rare disease, trauma, echocardiography, cardiac computed tomography, cardiac magnetic resonance, surgery

Case presentation

A 65-year-old man was admitted to the hospital due to dyspnoea, fatigue, and oedema of both legs. Heart failure symptoms worsened during the previous few weeks. Electrocardiogram (ECG) revealed a left bundle branch block. Since precordial auscultation revealed a loud systolic murmur along the left sternal edge, myocardial infarction with interventricular septum rupture was suspected. Laboratory findings were unremarkable. B-type natriuretic peptide (BNP) was not available. Previous medical history included arterial hypertension.

Transthoracic echocardiography (TTE) showed normal left ventricular systolic function (ejection fraction 65%), left ventricular hypertrophy (interventricular septum [IVS] 1.5 cm, posterior wall 1.5 cm), left atrial dilatation (4.8 cm), right ventricular dilatation (3.2 cm), normal left ventricular end-diastolic diameter (5.2 cm), and a cystic-like structure (aneurysm) in the right ventricle adjacent to the IVS (Fig. 1), located from the base of the septum to its mid-ventricular portion. In colour Doppler imaging, turbulent flow

was seen from the aorta into the aneurysmal sac and from its cavity into the right ventricle (RV) (Figures 1, 2). Continuous-wave Doppler showed continuous high-velocity flow from the aorta into the aneurysm and from its cavity into the RV during both phases of the cardiac cycle (systole and diastole) (Figure 3). The estimated pulmonary-to-systemic flow ratio (Qp/Qs) was 1.54. Systolic pulmonary artery pressure was 50 mm Hg.

Rupture of sinus Valsalva aneurysm (SVA) dissecting the IVS was suspected. A more detailed medical history revealed that 2 years prior, the patient suffered a fall from height with chest trauma, which could be the cause for the acquired SVA complicated by rupture.

Multi-slice cardiac computed tomography (CT) with contrast confirmed the diagnosis, showing right SVA dissecting the IVS with rupture into the RV cavity (Figures 4, 5). The perforation site between the IVS dissection aneurysm and the RV cavity was identified by cardiac CT (Fig.5).

Please cite this article: Oryshchyn N, Ivaniv Y, Kulyk L et al. Sinus of Valsalva aneurysm dissecting the interventricular septum with rupture into the right ventricle (RCD code: I-1B.O). J Rare Cardiovasc Dis. 2019; 4 (3): 51–54; doi: <https://doi.org/10.20418/jrcd.vol4no3.377>

Conflict of interest: none declared. Submitted: July 6, 2018. Accepted: November 27, 2019.

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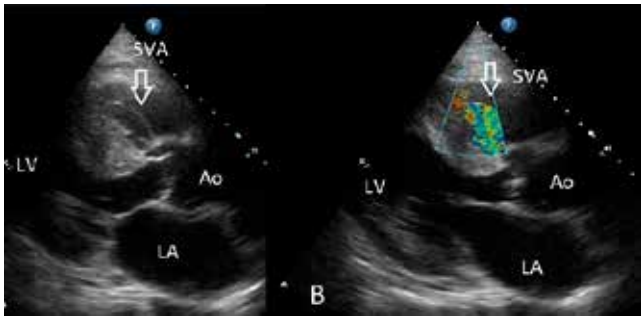


Figure 1. TTE. Parasternal long-axis view. SVA with IVS dissection seen as echo-free cavity adjacent to the right side of the IVS (A, B, arrow), turbulent flow within the cavity in colour Doppler is seen (B)



Figure 2. TTE with colour Doppler. Parasternal short axis view at the level of the aortic valve. The site of origination of SVA – arrow, SVA – arrowheads

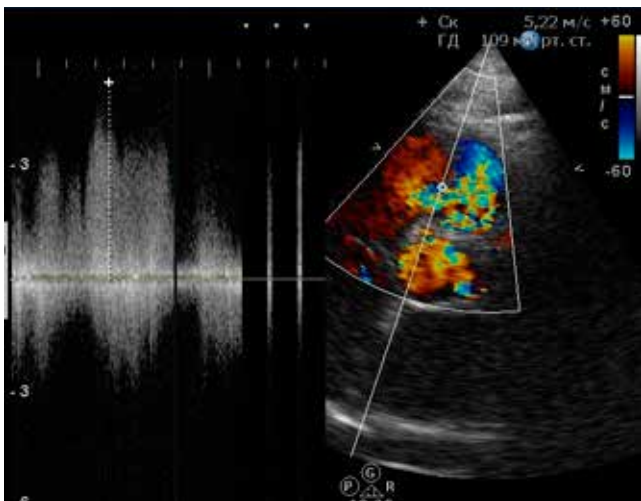


Figure 3. TTE. Parasternal short axis view at the level of the aortic valve. Continuous-wave Doppler demonstrates systolic and diastolic flow from the aorta to SVA cavity and to the right ventricle

Review of literature

SVA is a rare cardiac anomaly, caused by a defect in the aortic wall structure and can be congenital or acquired. SVA is a consequence of elastic lamina weakness at the junction of the aortic media and the annulus fibrosa. Acquired SVA are associated with connective tissue pathology, bacterial endocarditis, syphilis, atherosclerosis, and vasculitis. Chest trauma and injury during aortic valve replacement have been reported as causes of acquired SVA.

The main complication of SVA is rupture into the cardiac chamber, which causes volume overload and leads to acute or progressive chronic heart failure [1–5]. Acute onset of symptoms in SVA rupture is reported in 53% of patients, and in 42% of patients, symptoms of heart failure develop gradually [1]. The main complaints are dyspnoea (in 79% of patients), tachycardia (in 55% of patients), chest pain (in 52% of patients) [2]. During auscultation, loud murmurs from different areas are heard.

The most common site of SVA is the right coronary sinus (77 – 90%), followed by the non-coronary sinus (10 – 22%) and left coronary sinus [2, 3]. In one of the largest studies on SVA, Yan F. et al. (2014) analysed 160 cases of SVA in one centre and observed that the right coronary sinus origin was the site of SVA in 67.5% of cases, the non-coronary sinus in 31.9% of cases, and the left coronary sinus in only 0.6% of cases [6]. The rupture of SVA most commonly occurs into the right ventricle (in 55.6% of cases as reported by Yan F.), into the right atrium in 38.1% of cases, and into the left ventricle in 1.3% of cases. Unruptured SVA is rarely identified (in 5% of cases by Yan F.). Complications of SVA rupture include subaortic ventricular septal defect and aortic regurgitation, which occur in 22.5%–46.3% and 28 – 55.9% of cases respectively [3, 6, 7].

SVA rupture with IVS dissection occurs extremely rarely [8 – 13], as does biventricular rupture [14] or extracardiac rupture with cardiac tamponade [15]. Dissection of the left side of the IVS was common, together with left ventricular outflow tract obstruction. [8, 11, 12, 13]. A study including the largest cohort of patients with IVS dissection (Gu X. et al., 2017) reported 13 cases of IVS dissection, with right SVA being the most frequent cause of the dissection (6 cases, 46%) [13]. In this group, 3 patients had IVS dissection which was complicated by rupture: 2 cases ruptured into the left ventricular cavity, while 1 case ruptured into both ventricles. Thus, rupture into the RV caused by SVA with IVS dissection is an extremely rare finding. However, the exact mechanism of IVS dissection in SVA remains unclear. In the presented case, it could have been triggered by the patient's previous fall from height.

Diagnostics in SVA. For many years, cardiac catheterization and angiography have been the standard for diagnostics in SVA. Currently, TTE completely replaces the need for angiography, which is invasive and more costly. TTE became the first-line imaging technique in SVA diagnosis, while transoesophageal echocardiography is required in more complicated cases [16, 23]. TTE with colour Doppler is a sensitive diagnostic tool in the detection of intact as well as ruptured SVA and in the assessment of volume overload of the heart chambers [16]. Results of echocardiography were confirmed by angiography [1] or cardiac magnetic resonance imaging (MRI) [14]. TTE with colour Doppler can identify the site of SVA rupture and reveals continuous flow from the aorta through the de-

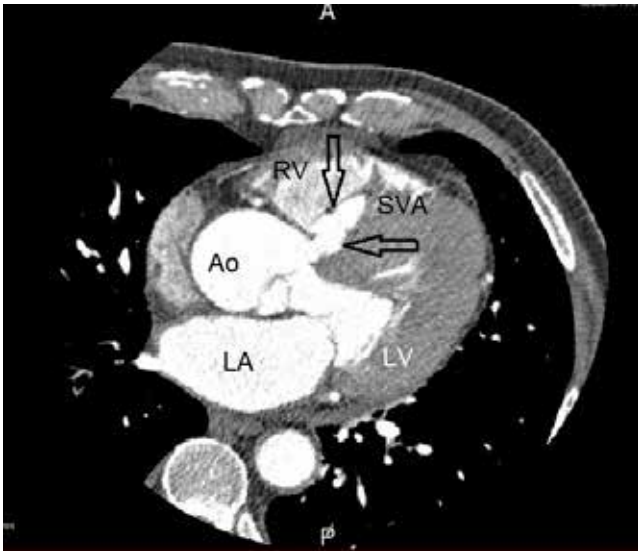


Figure 4. ECG-gated cardiac CT. SVA with IVS dissection (arrows)

fect in systole and diastole. Cheng and co-authors reported the sensitivity and specificity of TTE in SVA diagnostics to be 93.9% and 99.9% respectively [16]. Furthermore, diagnostic accuracy of aneurysm origin site identification, termination, and rupture was 99%, 99%, and 97.5%, respectively, as confirmed by surgical findings [16]. Transoesophageal echocardiography was required for confirmation of the diagnosis in a few cases [16]. TTE is the imaging modality of choice for detection of SVA rupture with IVS dissection [13]. The cavity of SVA with IVS dissection is pulsatile and is dependent on the cardiac cycle, expanding during diastole and decreasing during systole. However, the reported sensitivity of TTE in diagnostics of lesions associated with SVA was lower at 89.2%. [16].

ECG-gated cardiac CT with contrast and cardiac MRI are the preferred imaging methods prior to surgical repair of SVA, as they are able to specify anatomical features of SVA or coexisting malformations of the heart. ECG-gated cardiac CT is an accurate imaging tool for visualisation of the ascending aorta, and allows for precise assessment of the ascending aorta, including the sinuses of Valsalva [17]. Cardiac CT and MRI can identify the exact location of SVA origin and the site of its rupture, as well as any associated lesions [9, 17, 18, 19].

The presented case demonstrates the accuracy of echocardiography in the diagnostic process of SVA with IVS dissection and rupture. Cardiac CT was necessary to confirm the diagnosis and for the identification of anatomical features or associated malformations before planning surgical treatment. Cardiac CT identified the exact location of the additional cystic structure (aneurysm and dissection), its site of origin, and site of rupture.

Surgical resection is recommended as optimal care for ASV rupture and presently carries with it a low mortality rate, estimated to be 1.9–3.6% [3, 6, 7, 20, 21]. Successful percutaneous closure of SVA has been reported recently with the use of various occluders (ductal occluder, septal occluder) [4, 22, 23], with transoesophageal echocardiography being required during the procedure [4]. Surgical correction must be performed as soon as possible after diagnosis of SVA rupture has been established. The reported long-term

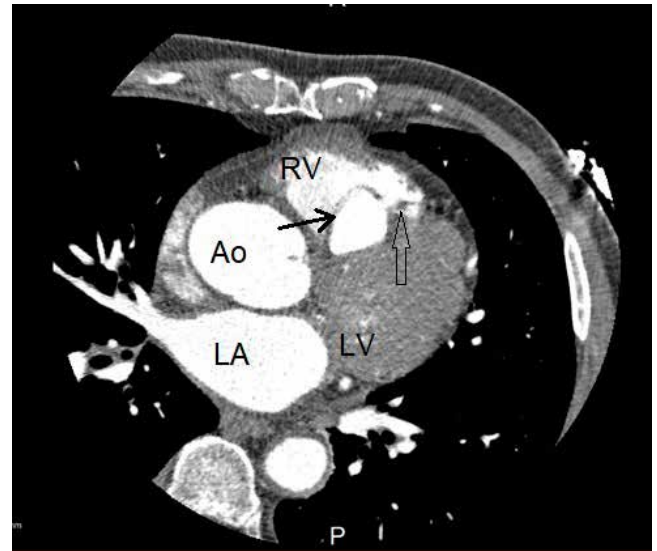


Figure 2. ECG-gated cardiac CT. Thin black arrow – SVA and IVS dissection cavity, outlined arrow – site of rupture

survival after surgical SVA resection is good. In a study involving the largest cohort (Yan), only 3 hospital deaths were reported, with a 10-year-survival rate of 94% and a 20-year-survival rate of 88% [6]. Sarikaja et al. reported survival rates of $93.4 \pm 3.7\%$ at 10 years and $87.1 \pm 5.6\%$ at 15 years [20].

Patient management

The patient was referred to a cardiac surgery unit where surgical repair of the ruptured aneurysm was performed successfully. All findings acquired by echocardiography and cardiac CT were confirmed during surgery. Three years post-operation the patient is in good physical health. His arterial hypertension is managed with standard therapy and he is classified as New York Heart Association (NYHA) functional class I-II.

Conclusion

SVA rupture dissecting the IVS is an extremely rare cardiac pathology with a potentially poor prognosis. TTE is the most readily available sensitive imaging tool for the diagnosis of SVA. Cardiac CT and MRI can more precisely identify anatomic features of the lesion prior to surgery. Surgery is recommended for SVA rupture closure. Prognosis of patients after surgical repair is good.

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