CASE REPORT

Congenital sinus of Valsalva aneurysm: insidious and unusual clinical presentation followed by an echocardiographic diagnosis

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ABSTRACT

The congenital sinus of Valsalva aneurysms (CSVAs) are infrequent and most often asymptomatic congenital heart defects. Usually, diagnosis is made by the appearance of severe complications. I described two complicated cases of CSVAs, both with insidious and unusual clinical presentation. First, with the CSVA ruptured into the RV and second in which CSVA caused aortic valve insufficiency.

INTRODUCTION

Congenital sinus of valsalva aneurysms (CSVAs) represent 0.1–3.5% of all congenital heart diseases (CHD). They are more frequent in males (4:1) and among Asian population (1.2-4.9%). They are associated with ventricular septal defects (35–59%), pulmonary stenosis, bicuspid aortic valve, tetralogy of Fallot, patent ductus arteriosus (PDA), aortic coarctation, subaortic stenosis, and single coronary artery. They may also be secondary to connective tissue diseases, endocarditis, rheumatic fever, syphilis or trauma (1–3).

In 69% of cases CSVAs are formed in the right coronary sinus (RCSV), in 26% in the non-coronary sinus (NCVS) and in less than 5% of cases in the left coronary sinus (LCSV) (3). Hong-Wei Guo et al. reported incidence of CSVAs for 82.10%, 17.51%, and 0.39% respectively (4).

The main complication of this rare congenital cardiac anomaly is the rupture, so called aortic–heart fistula (AHF). If it occurs in the acute form, it constitutes a surgical emergency. Frequency of individual AHFs is as follows: to the right ventricle (RV) – 60%, to the right atrium (RA) – 29%, to the left atrium (LA) – 6%, to the left ventricle (LV) – 4% and to the pericardium – 1% (5). Very rarely AHF may form to superior vena cava, pleura or pulmonary artery. Infrequently cases of rupture remain asymptomatic. They may have insidious clinical course and become diagnosed during echocardiogram performed for other reasons (6). If left untreated, they lead to heart failure (HF) and pulmonary hypertension (PH). Other complications of untreated AHF include endocarditis, aortic regurgitation (AR) (30 to 50%), heart block, ventricles inflow or outflow obstructions and systemic embolism (7).

CASE 1

A 6-year-old male patient with history of dyspnea, without previous hospitalizations, syncope, chest pain or trauma (for at least one year preceding hospitalization) and with no family history of CHD was presented to our clinic. He was referred to the Cardiology Department for cardiac murmur work up. The patient was in good general condition. On physical examination he had normal heart rhythm, normal first heart sound, high intensity second heart sound, normodynamic precordial activity, systolic and diastolic crescendo-decrescendo stenotic murmur in aortic focus with horizontal irradiation and high amplitude peripheral pulses, without hepatomegaly and peripheral edema. Electrocardiogram showed normal sinus rhythm and incomplete right bundle branch block (IRBBB). Chest X-ray revealed cardiomegaly with normal pulmonary flow. On transthoracic echocardiogram we ascertained an NCSV aneurysm rupturing to the RV with dilatation of the left heart cavities, with no other associated cardiac defects (Figure 1A and 1B). He was put on oral diuretic and referred for surgical repair.

CASE 2

A 7-year-old female patient, with no significant personal or familiar past medical history, was referred to Cardiology Department for cardiac murmur work up. Physical examination revealed normal weight and height, diastolic murmur grade II/IV in aortic focus irradiating to the apex, without fremitus, high amplitude peripheral pulses, and normal precordial activity. Electrocardiogram and chest X-ray were normal. Transthoracic echocardiogram showed RCSV aneurysm prolapsing into the RV outflow tract, without

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signs of obstruction or rupture. It was associated with moderate AR (Figure 3). The patient was also referred for surgical repair.

DISCUSSION

The first case of SVA was described by James Hope in 1839, one year later in 1840 Turnam reported the first case of rupture of the sinus of Valsalva (8). The physiopathological consequences of CSVA rupture depend on the volume of flow through communication, velocity of establishment of the rupture and cardiac chamber with which it communicates. When the perforation is acutely and big does not allow hemodynamic compensation, developing a sudden HF. The patient can be feel intense retrosternal and epigastric pain, which was unrelated to the effort associated with dyspnea. After an asymptomatic period, HF symptoms progress until patients’ death. In children, retrosternal pain is not a typical SVA rupture presentation. Usually, we see dyspnea, wheezing, tachycardia and gallop rhythm. Often an episode of endocarditis or trauma precedes the aneurysm rupture (9–11).

SVA rupture presents with high amplitude peripheral pulses, hyperdynamic paraesternal activity and LV apex displacement. On the chest palpation can be found systolic-diastolic thrill and during auscultation the second cardiac tone is strong (pulmonary component) and can be exist third and fourth cardiac tones (12). The first presented case corresponds to the above clinical description, though dyspnea was the only reported clinical symptom. The presence of high amplitude pulses and continuous
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Murmur raised suspicion of potential PDA and led to the echocardiogram; however the unusual location of the murmur was suggestive of ruptured SVA. Typically, the murmur of ruptured aneurysm is systolic-diastolic and stronger during systole (Figure 4), in comparison to the murmur of PDA, which is stronger during diastole (13). In the 2nd case, the presence of diastolic murmur in the aortic focus and high amplitude pulses suggested AR. The echocardiogram confirmed the clinical diagnosis and showed the presence of RCSVA (14, 15). AR was secondary to the right coronary cusp prolapse. The conclusion was congenital RCSVA, complicated with AR, but with insidious clinical presentation since the patient was asymptomatic.

Electrocardiogram is commonly normal. Although, even in cases of small ruptures, atrioventricular conduction alterations (IRBBB, atrioventricular block) can be found (16). They are more frequent when NCSV is affected, as it was in the first of our two cases.

The surgical repair remains the treatment of choice. It is indicated in all ruptured CSVA cases and when there is ventricle outflow tract obstruction. When CSVAs are not complicated or asymptomatic surgery is controversial and the only indication is AR or atrioventricular conduction alterations coexistence. Taking that into account, both of our patients were referred for surgery. Once CSVA is repaired, patients’ 15 years survival is greater than 87% (17).

Figure 3  A: Short axis view. Aortic valve is open and right coronary sinus of Valsalva aneurysm (RCSVA) prolapsing to the RV outflow tract (blue arrow) RV: right ventricle, Ao: ascending aorta, LA: left atrium, LV: left ventricle. B: Apical left ventricle outflow view. Aortic valve is closed and diastolic flow secondary aortic regurgitation. The blue arrow shows to aneurysm right coronary sinus of valsalva (RCSVA) prolapsed outlet RV Ao: Root aortic.

Figure 4  Short axis view. Doppler signal shows continuous flow secondary to aneurysm of non-coronary sinus of valsalva (NCSVA) ruptured to the RV. Typically the flow of ruptured aneurysm is more strong during systole RV: Right Ventricle.
CONCLUSIONS
The CSVAs are infrequent and most often asymptomatic congenital heart defects. Presented cases showed examples of CSVAs with insidious and unusual clinical presentation diagnosed with transthoracic echocardiography.

REFERENCES