Ruptured Right Sinus of Valsalva Aneurysm with Right Ventricular Fistula

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Introduction

Valsalva sinus aneurysm is an uncommon cardiac disorder associated with the separation or lack of fusion between the middle aortic layer and the fibrous aortic valve annulus. It is, in general, a congenital defect, but can be acquired by trauma, infections, degenerative diseases, systemic inflammatory diseases and connective tissue disorders. Its occurrence is three times more frequent in males. Its recognition is late, generally occurring in the third or fourth decade of life, when there is rupture to one chamber.

The most common aneurysm is the right coronary sinus aneurysm. The right ventricle is the most affected chamber. However, when the non-coronary valve is affected (5 to 15% of the cases), the fistula usually drains into the right atrium.

The left coronary sinus being affected with fistula to the left atrium is extremely rare. Associated anomalies are ventricular septal defect, bicuspid aortic valve and coarctation of the aorta. Unruptured aneurysms rarely cause hemodynamic repercussions. Fistula results in symptoms of acute heart failure due to volume overload. Aneurysm can also cause compression of the left coronary artery with ischemia and even acute coronary syndrome.1,2

Case Report

Male black 34-years-old individual with history of heart murmur from childhood with no regular medical follow-up. He was admitted at our institution reporting that, after performing a dental procedure, he evolved with progressive dyspnea 3 months prior, currently at small exertion, associated with lower limb edema. He was initially evaluated by an outpatient physician with initiation of diuretics and angiotensin converting enzyme inhibitor, with partial improvement of the condition. He also reported weight loss of about 18 kg in 2 months associated with night sweats. The patient had fever in the period.

Physical examination evidenced 3+/6+ diastolic murmur in the aorta. Laboratory tests with no relevant abnormalities, including negative serological tests. Sinus rhythm electrocardiogram revealed left chamber overload. Chest x-ray revealing cardiomegaly and pulmonary congestion. The patient underwent transthoracic echocardiography and, subsequently, transesophageal echocardiography. The echocardiogram showed right coronary sinus aneurysm (Figure 1; Video 1), with multiple fenestrations and systolic and diastolic flow communicating the aorta with the right ventricular outflow tract (Figures 2 to 4; Video 2), and a 12-mm collum (Figure 5). We also found a discrete increase in left chambers, preserved biventricular systolic function and pulmonary hypertension. Patent foramen ovale (PFO) (Figure 6) was evidenced.

Transesophageal echocardiogram did not show images suggestive of vegetation or abscess. Positron-emission tomography-computed tomography (PET-CT) did not reveal any signs of an inflammatory/infectious process in cardiac and large vessel topography either. The patient underwent surgical correction of the right sinus of Valsalva and closure of the fistula with bovine patch, in addition to FOP raphy. He was discharged with clinical improvement, and postoperative echocardiography only revealed a small residual aneurysm in the right Valsalva sinus, with a patch in the subaortic region, indicating subaortic interventricular communication, in addition to a flow compatible with residual right aorta-ventricle fistula.

Discussion

The Valsava sinus aneurysm is a rare condition that usually only causes symptoms after its rupture.3-5

It was first described in 1839, but a successful treatment for aneurysm rupture was only performed in mid-1950s, when Morrow et al. and Bigelow et al., independently, used hypothermia and inflow occlusion with venous return interruption. In 1956, the first cardiopulmonary bypass procedures were performed for surgical repair of the lesions.3

A retrospective study analyzing 40 years of experience of a service in the treatment of Valsalva sinus aneurysms has shown that the most common symptoms are easy tiredness, dyspnea, precordial pain and palpitations or tachycardia. In the same study, in correlation with the literature, the most affected sinus was the right coronary sinus, followed by the non-coronary sinus. In the presence of fistula, the most affected chamber is the right ventricle.1

Diagnosis of Valsalva sinus aneurysm is done by means of complementary tests, of which echocardiography is the method of choice, as it is widely available, non-invasive and easy to perform, and may even be conducted at the bedside for patients with more severe conditions. Other complementary methods include computed tomography

Keywords

Heart Defects, Congenital; Aneurysm, Ruptured; Sinus of Valsalva; Coronary Sinus; Fistula; Ventricular Dysfunction, Right; Heart Failure.
and magnetic resonance imaging. In some cases, coronary catheterization is necessary.\textsuperscript{1,4}

**Conclusion**

We presented a case of Valsalva sinus aneurysm with right ventricular fistula, with hemodynamic repercussions. Echocardiography was instrumental to the diagnosis. The patient also underwent PET-CT, available at our service, which ruled out an associated infectious process.

**Authors’ contributions**

Research creation and design: Gonçalves CSR, Silva RR, Lima MF; Data acquisition: Gonçalves CSR, Silva RR, Lima MF; Data analysis and interpretation: Gonçalves CSR, Silva RR, Lima MF; Manuscript writing: Gonçalves CSR, Silva RR, Lima MF; Critical revision of the manuscript as for important intellectual content: Gonçalves CSR, Silva RR, Lima MF.

**Potential Conflicts of Interest**

There are no relevant conflicts of interest.

**Sources of Funding**

This study had no external funding sources.

**Academic Association**

This study is not associated with any graduate program.
Figure 3 – Fistula of the coronary sinus-right ventricle on transesophageal echocardiogram.

Figure 4 – Fistula flow on continuous Doppler.
Figure 5 – Aneurysm collum.

Figure 6 – Patent foramen ovale.
Case Report

Gonçalves et al.
Ruptured right sinus of valsalva aneurysm

References

