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Cardiology

Right Sinus of Valsalva Fistula to the Right Ventricle as a Cause of Severe Heart Failure

Gabriel E. Pérez Baztarrica	MD, Department of Cardiology and Physiology.Universitary Hospital. Universidad Abierta Interamericana. Faculty of Medicine. Buenos Aires, Argentine.
Ignacio Vaca	MD, Department of Cardiology and Physiology.Universitary Hospital. Universidad Abierta Interamericana. Faculty of Medicine. Buenos Aires, Argentine.
Alejandro Botbol	MD, Department of Cardiology and Physiology.Universitary Hospital. Universidad Abierta Interamericana. Faculty of Medicine. Buenos Aires, Argentine.
Norberto Blanco	MD, Department of Cardiology and Physiology.Universitary Hospital. Universidad Abierta Interamericana. Faculty of Medicine. Buenos Aires, Argentine.
Rafael Porcile	MD, Department of Cardiology and Physiology.Universitary Hospital. Universidad Abierta Interamericana. Faculty of Medicine. Buenos Aires, Argentine.

ABSTRACT Fistula arising from ruptured sinus of Valsalva aneurysm is an uncommon cause of congestive heart failure, and it is even rarer in the absence of aneurysm.

We present the case of a female patient with acute idiopathic rupture of the right SV to the right ventricle and atrium in the absence of typical aneurysm. This anomaly resulted in a left-to-right shunt leading to rapidly progressive heart failure.

KEYWORDS : sinus of Valsalva; fistula; rupture; heart failure.

Introduction:

Fistula arising from ruptured sinus of Valsalva aneurysm is an uncommon cause of congestive heart failure, and it is even rarer in the absence of aneurysm.

Sinus of Valsalva fistulas (SVF) are an uncommon congenital or acquired cardiac anomaly.[1] Sinus of Valsalva (SV) anomalies were first described in1840 in the case of a 33-year-old man with a sudden onset of a "breaking" feeling in the heart, accompanied by syncope, palpitations, dyspnea and death within eleven weeks.[2]This condition is more common in men from Asia and Eastern Europe.[3]We present the case of a female patient with acute idiopathic rupture of the right SV to the right ventricle and atrium in the absence of typical aneurysm. This anomaly resulted in a left-to-right shunt leading to rapidly progressive heart failure.

Case report:

We present the case of a 35-year-old female patient with a history of pathological hypothyroid is maunder endocrinology follow-up and dyspnea CF II the long time. Two weak before hospitalization, the patient claimed to have sudden onset of NHYA functional class III-IV dyspnea, angina pectoris, and palpitations that a few days later progressed to lower limb edema. At the time of hospitalization, she showed signs of right congestive heart failure with 2/3 jugular ingurgitation, painful hepatomegaly, and 3/6 lower limb edema. Vital signs showed a heart rate of 115 bpm and a blood pressure of 100/70 mm Hg. Heart auscultation evidenced increased second heart sound, right R3, and continuous grade 4/6 murmur audible in the left parasternal region, third intercostal space and mesocardium, with no preferential irradiation. The electrocardiogram only showed sinus tachycardia. Laboratory tests were within normal ranges. Chest radiography revealed a normal cardiothoracic ratio and increased prominence of the main pulmonary artery and mildly increased pulmonary vascularity. The transthoracic echocardiogram showed a high-speed left-to-right shunt in the upper part of the interventricular septum, under the aortic valve. Measurement between pulmonary and systemic flows (QP/QS) was 2.4/1, with normal right cavities and preserved left ventricle systolic function. As a result of these findings, the assessment was completed with a transesophageal echocardiography (TEE), which revealed a left-to-right shunt between the right SV and the under surface of the right ventricle, with an 8-mm diameter break in continuity into the implantation site of the tricuspid septal valve **(Fig. 1A)**.

Cardiac catheterization evidenced contrast passage from the aorta to the right ventricle with a QP/QS of 2.2/1**(Fig.1B)**.

After being diagnosed with SVF to the right ventricle, the patient had surgery. Surgery showed an 8-mm hole in the right coronary sinus leading to the right cavities (the fistulous tract straddled the line between the right ventricle and atrium) **(Fig 2 A-B)**. Simple closure was performed on the hole (intraoperative TEE without shunt), with uncomplicated progress at the time of discharge.

Discussion:

SVF is a very rare condition, with an incidence of about 0.1 to 3.5% of all congenital heart malformations.[4] Anatomically, SV anomalies are classified as: 1) aneurysm; 2) aneurysm with fistula (rupture), and 3) fistula.[5] Most congenital SV anomalies affects the right SV and the non-coronary sinus; those in the left SV are usually acquired[5] and can result from trauma, endocarditis, syphilis, Behecet disease, Marfan syndrome, and senile dilation[6]; 76% of SVF are caused by sinus of Valsalva aneurysm, and few cases of SVF without aneurysm have been reported, as in our case.

Etiopathogenesis is simple in the absence of other SV disorders.[7] In contrast, SV as are related to a cardiac loop formation defect during embryonic development associated with long-lasting high pressure periods typical of the aorta.[8] The left coronary cusp is not derived from bulbar septum, and therefore, congenital left SV aneurysms do not occur.[9] SV are 3 small dilations on the wall of the aorta just above the aortic valves. The anatomical arrangement of the aortic sinus of Valsalva with respect to heart chambers may explains the hemodynamic changes arising from a ruptured sinus.[10]

The gold standard in imaging diagnosis is angiography, even though in 90% of cases initial diagnosis is provided by the TEE; cardiac magnetic resonance and multislice computed tomography can also be useful.[3]

SV rupture needs to be surgically corrected.[11] Survival in patients with non-surgical ruptured sinus of Valsalva is 3.9 years and the

postoperative survival rate after 10 and 20 years is of about 90%. Therefore, surgery is mandatory in these cases.[12] Surgical options include aortotomy, opening of heart cavities or a combination, as well as simple closure of the fistulous hole or use of patches depending on the size of the defect. Defects larger than 0.8 cm should be closed with patches.[9] In our case, the patient had an 8-mm defect; thus, simple closure was performed, with uncomplicated progress and no immediate or remote postoperative complications.

Conclusion:

In summary, we present an uncommon case of SV fistula to the right ventricle with signs and symptoms of heart failure. This is a complex condition that should be timely suspected (based on clinical presentation and physical examination), diagnosed, and treated. Otherwise, there is poor prognosis.

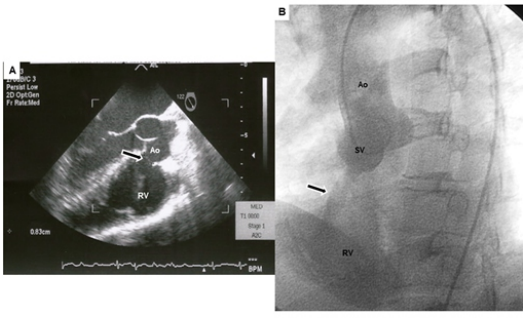

Fig.1: A) Transesophageal echocardiography (TEE) revealing left-to-right shunt between the right SV and the undersurface of the right ventricle (arrow). **B**) Cardiac catheterization evidenced the contrast passage from the aorta to the right ventricle (arrow). **Ao:** aorta; **RV**: right ventricle; **SV:** sinus of Valsalva.

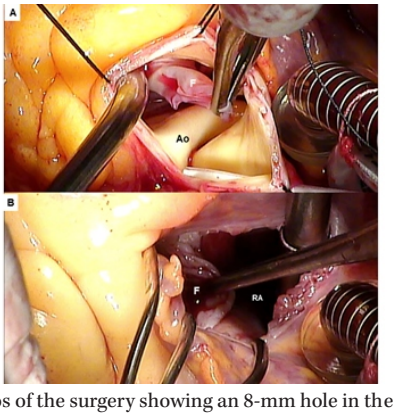

Fig. 2: Photos of the surgery showing an 8-mm hole in the coronary sinus leading to the right cavities. **A**) View from the aorta, and **B)** View from the right cavities. **Ao**: aorta; **RA**: right atrium; **F**: fistula.

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