Rupture sinus of Valsalva in a patient with dextrocardia. Case report.

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Abstract
Ruptured sinus of Valsalva (RSOV) is a rare disease with a wide range of clinical manifestations, from asymptomatic murmur to cardiogenic shock and death. We present the case of a young woman known with dextrocardia and pulmonary agenesis; she was diagnosed with RSOV and during follow-up, with endocarditis. Her evolution was good despite the association of severe heart conditions and the refusal of surgical intervention.

Keywords: sinus of Valsalva, rupture, dextrocardia, endocarditis

Introduction
Ruptured sinus of Valsalva (RSOV) is an uncommon condition with uncharacteristic manifestations, from an asymptomatic murmur to cardiogenic shock and death. As clinical examination is not entirely reliable and cardiac catheterization is not always available, echocardiography has become the definitive investigative tool not only to define and diagnose the lesion but also to quantify its severity [1].

Case report
A 42 years old female, known with dextrocardia and right pulmonary agenesis was sent for supplementary evaluation in the cardiology department from an internal medicine department. She complained of breathlessness with a sudden onset, started two months prior to admission, and progressively aggravated; physical examination revealed a systolo-diastolic murmur with maximum intensity on the right axial and posterior laterovertebral region and turgescent jugular veins. Rest electrocardiogram was concordant with dextrocardia, with normal sinus rhythm and right axis deviation. Chest X ray showed typical presentation for dextrocardia (fig 1). Lab results were within normal ranges.

Transthoracic echocardiography revealed a turbulent jet from the aortic cusp to the right atrium, tapering the tricuspid valve and important right chamber dilation, consistent with a RSOV (fig 2). Aortic insufficiency grade 2 color Doppler, mitral insufficiency grade 2 color Doppler and tricuspid insufficiency grade 2, with slightly elevated pulmonary pressure was also found. Ejection fraction was 0.48. The CT scan with contrast confirmed the diagnosis (fig 3).


The patient refused further investigation (transesophageal echocardiography, catheterization) and surgical intervention. She was discharged with indication of diuretics and beta-blocker treatment.

After discharge she continued to be symptomatic and had repeated admissions for heart failure symptoms such as dyspnea, calf edema and fatigue.
Fig 1. Chest X-Ray with dextro-position of the heart, right pulmonary agenesis, and hyperinflated left lung. Trachea is shifted to the right.

Fig 2. A non-typical view of cardiac ultrasound showing dilated sinus of Valsalva with a turbulent color Doppler jet from the aorta to the right atrium (white arrow showing the communication).

Fig 3. Contrast enhanced CT of thorax and heart showing the sinus Valsalva rupture (black arrow).

Fig 4. A non-typical view of cardiac ultrasound showing large vegetation attached to non coronary aortic cusp, floating into the right atrium.

One year later she presented for a check-up; she has done recently dental extractions, under antibiotic prophylaxis. She had no fever or supplementary signs of heart failure. On echocardiography, large vegetation (2/2.7cm) attached to the aortic cusp and floating inside the right atrium was found (fig 4). Blood cultures were positive for stafillococcus epidermidis and infective endocarditis of aortic valve was diagnosed. Treatment was given according to antibiogram. After a 6 weeks treatment with cefalosporines and oxacilin her state was improved, she had no signs of heart failure, no fever, or other general symptoms.

Discussions

ROSV is a rare entity and before echocardiography it was elusive to reliable diagnosis without non-invasive investigations. The unruptured aneurysm is usually silent and generally undiagnosed but may cause symptoms due to right ventricular outflow tract obstruction [1]. The rupture may occur into any cardiac chamber, frequently in the right ventricle or right atrium, but it may also rupture into the left ventricle, the inter-ventricular septum, or the pericardial space [2].

The aneurysm of the Valsalva sinus seems to be done...
by the failure of fusion between the aortic media and the heart at the level of the annulus fibrosus of the aortic valve, with subsequent aneurismal enlargement at this weak point due to the high head of pressure at the root of the aorta [3]. Also, it may be related to diseases of connective tissue such as Marfan’s syndrome and Behcet’s disease [4]. Males are more affected than females (3:1 ratio), and the prevalence is higher in the Eastern than in the Western population. Patients are usually diagnosed in the 3rd or 4th decade of life, when rupture occurs and symptoms develop [5].

Rupture can also occur with endocarditis affecting the aortic valve or a ventricular septal defect VSD [6], reflecting the weakening of the aortic wall due to the inflammatory process and occasionally by severe chest trauma [7]. Although the first reported case of RSOV diagnosed by echocardiography was in 1974 [8], the gold standard for diagnosis of this lesion has traditionally been cardiac catheterization and aortography. With the advent of newer generation ultrasound machines, transthoracic echocardiography and transesophageal echocardiography [9] have taken centre stage for diagnostic confirmation.

The 2-D echo features of RSOV include the aneurismal dilatation of the sinus of Valsalva where a “windsock” appearance may be identified. Doppler features include a continuous high velocity unidirectional flow through the lesion. The diagnosis is usually confirmed on color flow imaging, which reveals a unidirectional continuous mosaic jet from the aorta to the right heart chamber. These features may be more clearly seen on transesophageal echocardiography [8].

The rupture of the aneurysm of the right coronary cusp sinus of Valsalva into the right ventricle is by far the commonest type of RSOV. A few pitfalls must be kept in mind: 1) isolated VSD, if the VSD is subarterial and very close to the aortic cusps; 2) coronary arteriovenous fistula; and 3) patent ductus arteriosus present with continuous murmurs and the pattern of color flow in the right ventricular outflow tract and pulmonary artery must be scrutinized carefully to avoid misdiagnosis [8,10].

Our case was particular as the condition appeared in a patient with dextrocardia and was complicated with infective endocarditis after dental extractions. Also, despite the refusal of any surgical intervention, the evolution was good, with no signs of heart failure or systemic infection associated with a persistent image of vegetation.

The natural history of asymptomatic aneurysm of an aortic sinus is unclear, and variant cases - with rapid clinical deterioration or many years of stabilization - have been described [5,7,8]. However, once symptoms develop or rupture occurs, urgent intervention is recommended.

In conclusion, RSOV is a rare condition with a varied presentation that sometimes can be fatal if not diagnosed quickly. Right coronary sinus of Valsalva to right ventricle ruptures are the commonest type of RSOV and there is an association with VSD, aortic regurgitation and infective endocarditis. Echocardiography is an accurate and reliable noninvasive tool to diagnose the lesion and obviates the need for cardiac catheterization in most cases.

References