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Case Report

An Unusual Cause of Heart Failure. Rupture of Right Sinus of Valsalva Aneurysm into Right Ventricle and Above Pulmonary Valve

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Abstract

The rupture of sinus of Valsalva aneurysm is a rare anomaly. Its prevalence is approximately 0.09% of the general population. The rupture into pulmonary artery is extremely rare (0.2%). Here we describe a case of a 31-year-old man with symptoms of heart failure. A rupture of right sinus of Valsalva aneurysm with formation of two fistulas into right ventricle and just above the level of the pulmonary valve was diagnosed by echocardiography and Computed tomography.

The patient underwent surgical treatment with resection of the aneurysmal region of the right sinus of Valsalva and it was closed with a patch of bovine pericardium. Unfortunately, the patient had a cardiac arrest after the procedure and died.

Keywords: Rupture of sinus of Valsalva aneurysm; Heart failure; Echocardiography; Computed tomography

Introduction

The rupture of sinus of Valsalva aneurysm is a rare cardiac anomaly. Its prevalence is approximately 0.09% of the general population [1]. The most common origin is from right coronary sinus (61%), followed by non-coronary sinus (26%) and left coronary sinus (12%) [2]. When it ruptures, the most common site is into the right ventricle (66%), followed by the right atrium (30.3%), left chambers (3.1%), right ventricular outflow tract (8.5%), interventricular septum (1.6%), pericardium (0.6%) and into the pulmonary artery (0.2%) [3], it has not been reported any case of rupture of sinus of Valsalva aneurysm with formation of two fistulas. Here we describe a unique case of rupture of sinus of Valsalva aneurysm that originated two fistulas to the right ventricle and above the pulmonary valve.

Case presentation

A 31-year-old obese man with a history of systemic hypertension. He came to emergency room with progressive exertional dyspnea, nocturnal paroxysmal dyspnea, orthopnea and an increase in abdominal circumference. At his arrival, he had systolic hypertension, tachycardia, tachypnea and was afebrile. On palpation, thrill in precordial area was detected and cardiac apex was in sixth intercostal space, anterior axillar line; on auscultation, continuous murmur in accessory aortic point was heard. The N-terminal proBrain Natriuretic peptide was of 1415 pg/dl. Electrocardiogram showed sinus tachycardia with biventricular enlargement and left atrial dilation, Figure 1. Chest



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X-ray showed cardiomegaly, prominent bilateral pulmonary branches and venocapillary hypertension, Figure 2. Intravenous

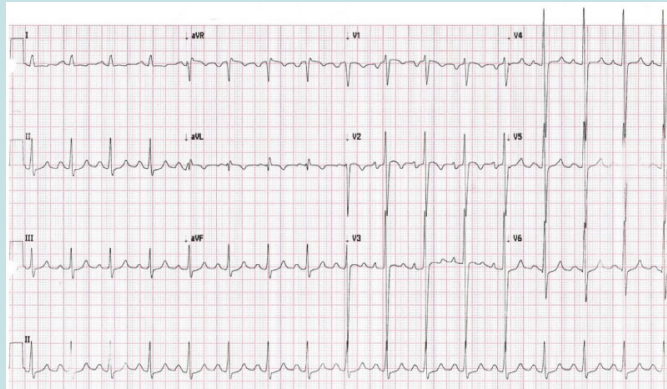


Figure 1: Electrocardiogram in sinus rhythm, with heart rate of 100 beats per minute, biventricular enlargement and left atrial dilation.

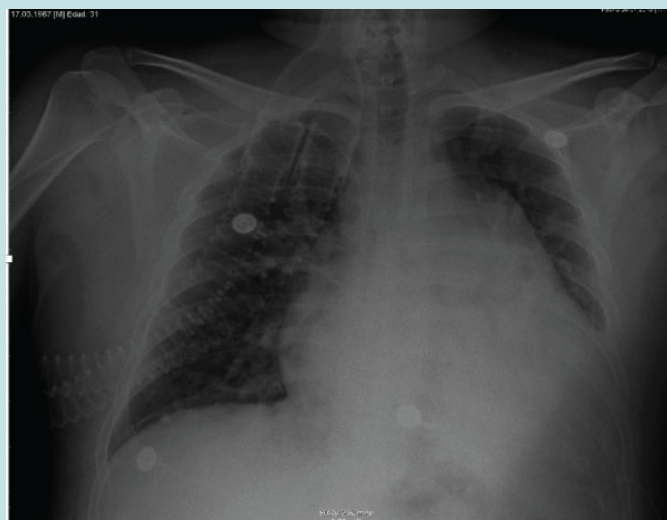


Figure 2: Chest X-ray with cardiomegaly, prominent bilateral pulmonary branches and venocapillary hypertension.

furosemide, nitroglycerin and noninvasive mechanical ventilation were initiated.

Two and three-dimensional echocardiograms showed dilation of 4 cavities, left ventricular eccentric hypertrophy, left ventricular ejection fraction of 58%, tricuspid aortic valve with mild aortic regurgitation, dilation and rupture of right sinus of Valsalva into right ventricle and above the pulmonary valve, Figure 3 and clips 1, 2, 3, 4. Cardiovascular computed tomography (CT) was performed to better characterize cardiac anatomy and two entries were found, one of 14 mm into the right ventricle and the other one of 10 mm above the pulmonary valve and biventricular enlargement, but with normal function (Left ventricular ejection fraction-61% and Right ventricular ejection fraction-67%). The coronary arteries were normal. Dilation of pulmonary trunk and branches was observed, Figure 3, Clip 5. The patient underwent surgical treatment with resection of the aneurysmal region of the right sinus of Valsalva and it was closed with a patch of bovine

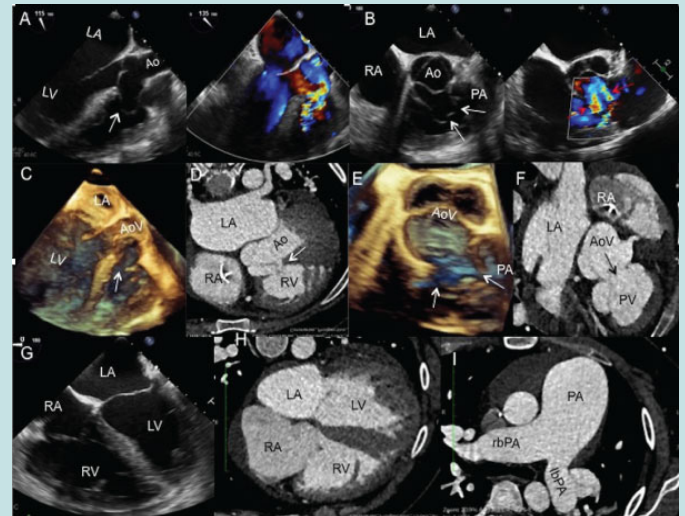


Figure 3: A-Transesophageal two-dimensional and color-flow echocardiogram, showing the rupture of the right coronary sinus aneurysm into the right ventricle (white arrow) B- above the pulmonary valve (white arrow). A mild pulmonary regurgitation is also observed. C- three-dimensional transesophageal echocardiography at 135° and computed tomography (D), pointing the rupture into the right ventricle (white arrow). E- three-dimensional transesophageal echocardiogram at 50° with rupture of the right coronary sinus into the right ventricle (white arrow) and above pulmonary valve (white arrow) and F- into the pulmonary valve visualized in the computed tomography (black arrow). G- two-dimensional transesophageal echocardiogram at 0°, and H, I- computed tomography, showing dilation of the right cavities and of the trunk and branches of the pulmonary artery.

Abbreviations: RA-right atrium; RV-right ventricle; LA-left atrium; LV-left ventricle; Ao-aorta; PA-pulmonary artery; rbPA-right branch of pulmonary artery; lbPA-left branch pulmonary artery.

pericardium. Unfortunately, the patient had a cardiac arrest after the procedure and died.

Discussion

Sinus of Valsalva aneurysms are a form of acquired or congenital aortic root aneurysm, congenital cases represent 3.5% of all congenital heart defects and often present association with ventricular septal defects, aortic insufficiency, bicuspid aortic valve and coronary anomalies. The acquired form is related to infections such as bacterial endocarditis, syphilis, and tuberculosis, or degenerative conditions such as atherosclerosis and cystic medial necrosis, injury from deceleration trauma, or iatrogenic due to aortic valve interventions [3].

Often these patients are asymptomatic, but when symptoms present, they usually are related to a mass effect on the cardiac adjacent structures or rupture of sinus of Valsalva aneurysm. The symptoms of rupture of sinus of Valsalva aneurysm include severe dyspnea, chest pain, progressive heart failure with fatigue, volume overload and hemodynamic compromise. In young patients with new-onset heart failure not otherwise explained, a rupture of sinus of Valsalva aneurysm must be suspected. On physical examination the most common finding is a systolic and diastolic murmur that intensifies during systole, which results

from the fistula formed between the aorta and a low-pressure chamber; in this case between the aorta and the right ventricle and above the pulmonary valve [2].

Diagnostic approach of rupture of sinus of Valsalva aneurysm must include an echocardiogram, CT or cardiac magnetic resonance imaging (MRI). The findings include an origin above the aortic annulus, saccular shape and normal dimensions of the adjacent aortic root and ascending aorta. The role of CT in evaluation of these cases is to define cardiac anatomy, the origin and path of the coronary arteries and associated anomalies in order to choose the best surgical technique [3].

Urgent surgical treatment is mandatory in these patients. There are two approaches: the first is a repair with patch placement during open surgery, which has been shown to be more effective than primary repair, this procedure was applied in our patient, the second is the percutaneous transcatheter closure of rupture of sinus of Valsalva aneurysm with an occluder device. The prognosis of these patients is good, when they are treated early [3].

Conclusions

The rupture of sinus of Valsalva aneurysm is a rare cardiac condition that can occur with a left to right shunt that leads to subacute or acute heart failure in young adults, it can be congenital or acquired [4]. In our patient it was congenital. Most commonly it ruptures into right ventricle and it is extremely rare above the pulmonary artery. Our case is of great interest because two entries were detected, one to the right ventricle and the other to the pulmonary valve

The diagnosis is established by image protocols that include echocardiogram, CT or cardiac MRI. The role of the CT is to further evaluate the anatomy of sinus of Valsalva aneurysm, the coronary arteries and the associated anomalies to guide the best treatment options.

Highlights

- Rupture of sinus of Valsalva aneurysm is a rare condition and is extremely uncommon with two entries.
- Echocardiography is the first step to make the diagnosis of rupture of the sinus of Valsalva aneurysm, but CT or cardiac MRI is needed to evaluate the anatomy and related cardiac anomalies and to determine proper treatment.
- Surgical treatment is mandatory to avoid complications.

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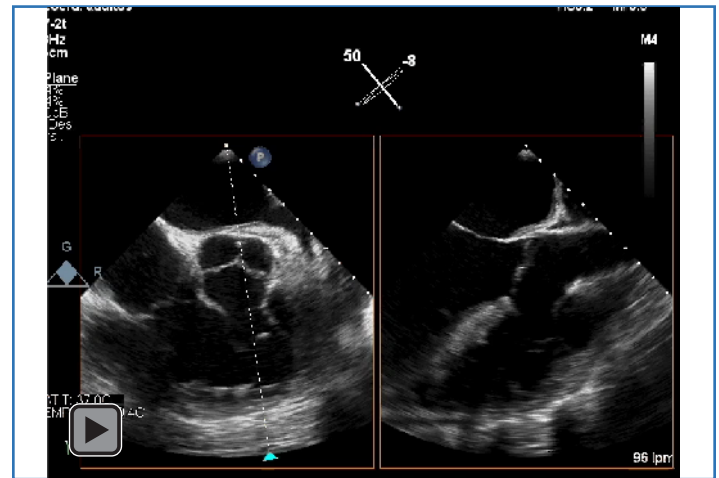
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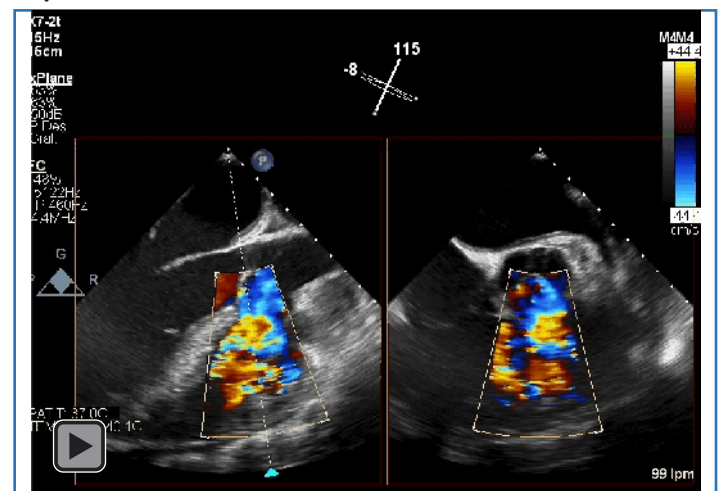
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Video Clips

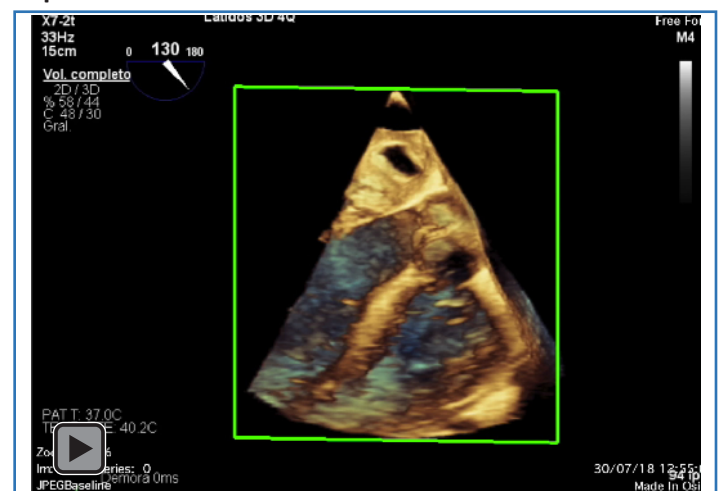
Clip 1



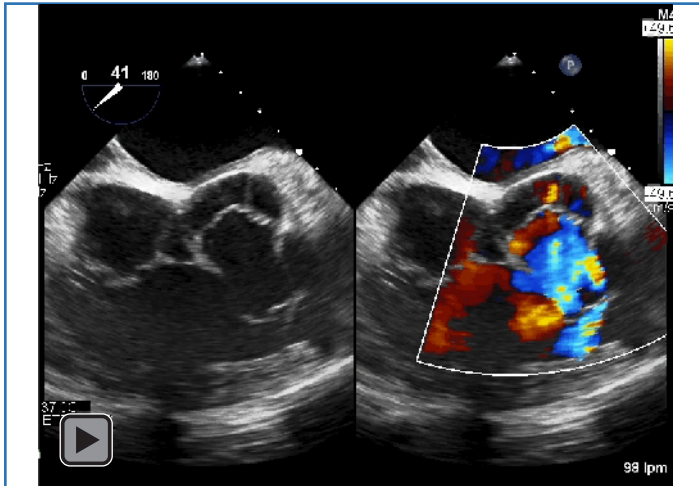
Clip 2



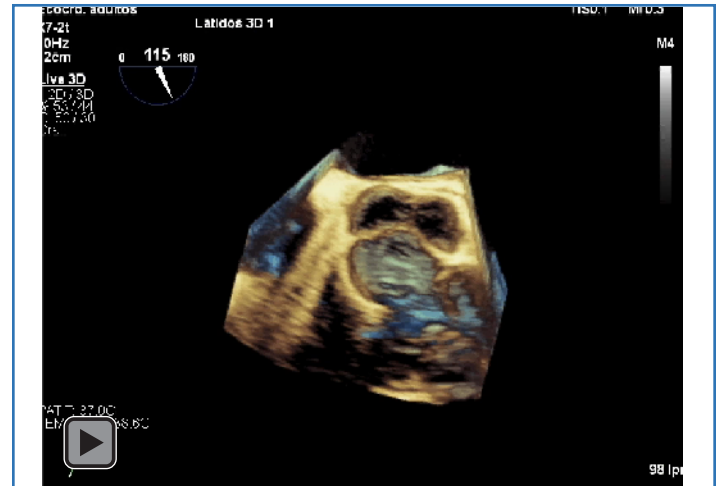
Clip 3A



Clip 3B



Clip 4



Clip 5



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