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Case Report

Multiple Unruptured Sinus of Valsalva Aneurysms Involving Right and Left Aortic Sinuses and Presenting as Severe Aortic Regurgitation

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Received: October 05, 2020; **Published:** November 25, 2020

Abstract

Sinus of Valsalva aneurysms (SVAs) is a rare but well-known defect of the aortic root. An even greater rarity is the presence of multiple unruptured aneurysms in the same patient. Unruptured SVAs is asymptomatic usually and often found incidentally. However, the condition can manifest by distortion or compression of adjacent structures as right ventricular outflow tract obstruction, aortic insufficiency, tricuspid stenosis and regurgitation, myocardial ischemia or infarction, conduction disturbances, mediastinal mass, thromboembolism and rupture. The diagnostic modalities include transthoracic and transesophageal echocardiography, multidetector computed tomography, magnetic resonance imaging angiography and cardiac catheterization. Surgical repair of unruptured SVAs should be advised in symptomatic cases.

The author describe a case of multiple unruptured SVAs involving right and left aortic sinuses presenting with signs and symptoms of congestive heart failure, due to a severe aortic regurgitation in a 25-year-old female who underwent successful surgical repair.

Keywords: *Sinus of Valsalva Aneurysm; Echocardiography; Computed Tomography; Aortic Regurgitation*

Introduction

Sinus of Valsalva aneurysms (SVAs) are a rare but well-known defect of the aortic root [1]. An even greater rarity is the presence of multiple unruptured aneurysms in the same patient [2-4]. We describe a case of multiple unruptured SVAs involving right and left aortic sinuses presenting with signs and symptoms of congestive heart failure, due to a severe aortic regurgitation in a 25-year-old female who underwent successful surgical repair.

Case Report

A 25-year-old woman presented at our hospital with symptoms of tiredness and progressively increasing dyspnea, orthopnea and lower limb edema lasting one month. The clinical examination revealed a normal Corrigan pulse of 105 minutes, blood pressure of 140/40 mmHg. A loud holodiastolic murmur over the third left intercostal space and some basal pulmonary rales were present. The abdominal palpation revealed hepatomegaly 8 cm below the right costal edge. Edema in the distal third of the lower limbs was also observed.

12-lead electrocardiogram showed normal sinus rhythm. Chest X-ray revealed cardiomegaly with features suggestive of pulmonary venous hypertension. Blood biochemical and hematological parameters were within normal range.

Two-dimensional transthoracic echocardiography (TTE) (Figure 1A) and transesophageal echocardiography (TEE) (Figure 1B) revealed two unruptured SVAs. One originating from the right aortic sinus contained a large layered thrombus, another aneurysms was

arising from the dilated and deformed left aortic sinus. In addition, there was severe aortic regurgitation (Figure 1C) and moderate left ventricular dysfunction. Contrast-enhanced multidetector computed tomography (MDCT) scan of the thorax confirmed echocardiographic findings and excluded other pathologies of the aorta (Figure 2).

The patient was underwent Bentall-de Bono procedure, postoperative course was uneventful. At twelve months follow-up the patient remains asymptomatic and doing well. Intra-operative findings included normal aortic valve leaflets and two unruptured SVAs (Figure 1D). One was arising from the right aortic sinus; another was arising from the left aortic sinus confirming the diagnosis of unruptured aneurysm of both right and left sinus of Valsalva.

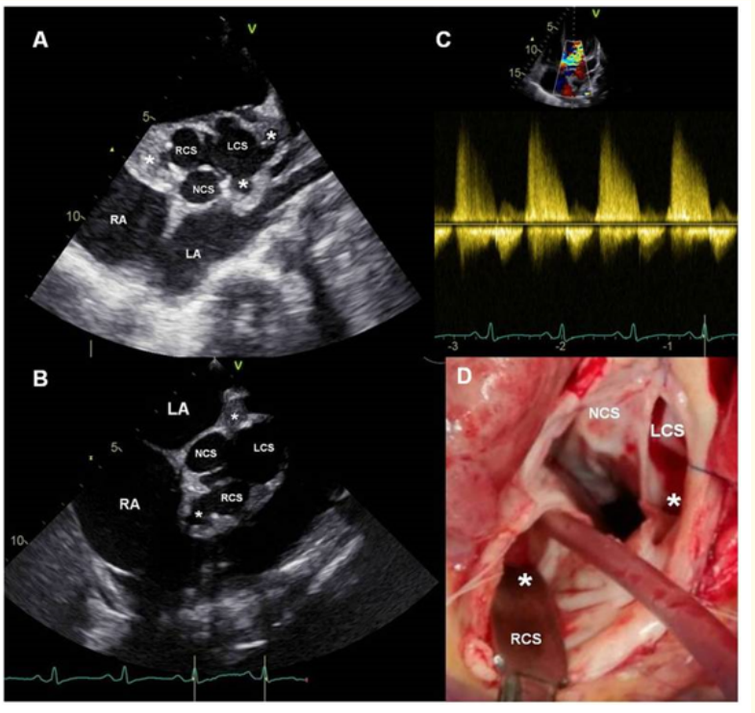

Figure 1: *A- Transthoracic short axis at level of great vessels, and B- Transesophageal echocardiography midesophageal short axis at 45º showed two unruptured ASV (asterisk) one was arising from right aortic sinus, another was arising from left aortic sinus; C- Doppler study showing severe aortic regurgitation; D- Intra-operative findings included normal aortic valve leaflets and two unruptured ASV. ASV: Aneurysm of Sinus of Valsalva; LCS: Left Coronary Sinus; NCS: Non Coronary Sinus; RCS: Right Coronary Sinus; LA: Left Atrium; RA: Right Atrium.*

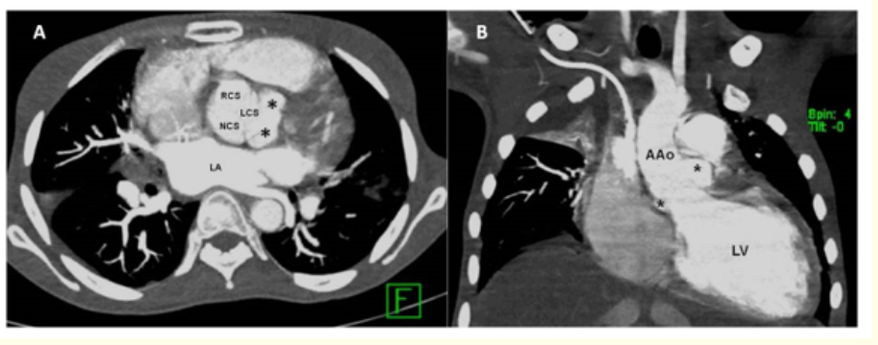

Figure 2: *Contrast-enhanced thoracic CT: A- axial view, and B- frontal view showed two unruptured ASV (asterisk). one was arising from right aortic sinus, another was arising from left aortic sinus; ASV: Aneurysm of Sinus of Valsalva; CT: Computed Tomography; LCS: Left Coronary Sinus; NCS: Non Coronary Sinus; RCS: Right Coronary Sinus; AAO: Ascending Aorta; LA: Left Atrium; LV: Left Ventricle.*

Discussion

SVAs are a rare defect of the aortic root and accounts for 0.14 - 1.5% of the cardiac surgical load. Of these aneurysms 75 - 90% arises from the right sinus of Valsalva, 10 - 25% from the noncoronary sinus and with the reminder occurring in the left coronary sinus [5]. The presence of multiple unruptured aneurysms in the same patient as the case presented herein is extremely rare [2-4].

These aneurysms can be congenital, inherited, or acquired. The congenital SVAs are consequences of incomplete fusion of the distal bulbar septum and truncal ridges resulting in fragility at the junction of the aortic annulus, the right aortic sinus media, and the right portion of the noncoronary sinus. Inherited forms occur in persons with deficiency of healthy elastic tissue resulting in annul aortic ectasia with dilatation of all three SVs and thereby progressive effacement of the sinotubular junction as seen in Marfan, Ehlers-Danlos, and Loeys-Dietz syndromes. Acquired aneurysms can result from atherosclerosis, infective endocarditis, tuberculosis, syphilis, dissecting aortic aneurysms, cystic medial necrosis, Behcet's disease, and traumatic injury involving the aortic root [6]. In our patient we ruled out all the acquired causes of the SVAs by doing the necessary investigations. In the present case the aneurysm are congenital in origin.

Most unruptured SVAs are asymptomatic and detected as incidental findings on imaging studies. However these aneurysms can compress the adjacent cardiac structures resulting in right ventricular outflow obstruction [7], aortic insufficiency and mitral insufficiency [8], tricuspid stenosis, tricuspid insufficiency [9], acute coronary syndrome [10], thromboembolism [11], mass effect [1], and conduction disturbance [1,2,12].

Our patient presented with signs and symptoms of congestive heart failure due to severe aortic regurgitation.

TTE is the first screening modality of choice given its excellent sensitivity, availability, and portability. TEE, due to its better acoustic window and higher resolution, provides a more precise characterization of the aneurysm, Doppler imaging is an advantage of both TTE and TEE imaging, helping to discriminate associated cardiac structural abnormalities and, diagnosing potential complications of SVAs, such as rupture, functional aortic regurgitation, and compressive effects on cardiac structures [13,14].

Tomographic imaging with MDCT and magnetic resonance imaging (MRI) is helpful for accurate assessment of the entire thoracoabdominal aorta. MDCT offers excellent assessment of extracardiac anatomy, at the cost of radiation exposure. Cardiac MRI offers excellent functional assessment of valvular regurgitation, as well as the extent of intracardiac shunting, although scan acquisition is more time-consuming and access to this modality is relatively more limited [13,14], Moreover, it may be unsuitable in the setting of acute SVA rupture due to its prolonged acquisition time.

In our patient the joint use of echocardiography and contrast-enhanced MDCT of the thorax allowed a more precise diagnosis and a more adequate planning of surgical treatment.

Invasive angiography has been considered the gold standard and has the advantage of both diagnostic and therapeutic potentials. However, noninvasive cross-sectional imaging modalities, consisting of color Doppler echocardiography, cardiac MDCT, and cardiac MRI, have mostly replaced it [14]. Invasive tests must be reserved for patients in whom the percutaneous closure of the fistula is possible, or the suspicion of coronary disease is present [14,15].

The management of symptomatic unruptured SVAs generally involves surgical repair. These aneurysms must be repaired by exclusion of aneurysm sac and closure of sinus defect using expanded polytetrafluoroethylene patch or pericardial patch. Surgical repair may involve adjuvant procedures like aortic valve sparing procedures or aortic valve or aortic root replacement in cases of associated aortic regurgitation or aortic root distortion [4]. In the present case, the patient underwent successfully Bentall-de Bono procedure.

Conclusion

Unruptured multiple SVAs are extremely rare cardiac anomalies with a varied clinical spectrum, including severe aortic regurgitation. Echocardiography is the first-line method in the diagnosis of SVAs. However, the combination of echocardiography with other tomographic imaging techniques (MDCT or MRI) is recommended for an accurate assessment of SVAs and a precise preoperative diagnosis. In symptomatic patients, early surgical repair is recommended.

Acknowledgments

The author thanks Dr Albino Pedro for the intra-operative images provided.

Conflict of Interests

None to declare.

Financial Support

None to declare.

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Volume 7 Issue 12 December 2020