The role of echocardiography in the diagnosis and management of a giant unruptured sinus of Valsalva aneurysm

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Introduction

Sinus of Valsalva aneurysm (SVA) is an aortic root anomaly, consisting of a lack of continuity between the aortic media and the aortic annulus, caused by a structural deficiency of muscular and elastic tissue. We present the case of a 49-year-old man with atypical chest pain. Echocardiographic imaging described a giant unruptured aneurysm of the right sinus of Valsalva which was confirmed by cardiac computed tomography and coronary angiography. The obstruction of the right coronary artery without intravascular thrombosis and the compression of the right ventricular outflow tract with dynamic obstruction gradient represent the particularities of our case.

Keywords: sinus of Valsalva; aneurysm; aorta

Abstract

Sinus of Valsalva Aneurysm (SVA) is an aortic root anomaly, consisting of a lack of continuity between the aortic media and the aortic annulus, caused by a structural deficiency of muscular and elastic tissue. We present the case of a 49-year-old man with atypical chest pain. Echocardiographic imaging described a giant unruptured aneurysm of the right sinus of Valsalva which was confirmed by cardiac computed tomography and coronary angiography. The obstruction of the right coronary artery without intravascular thrombosis and the compression of the right ventricular outflow tract with dynamic obstruction gradient represent the particularities of our case.

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Introduction

Sinus of Valsalva aneurysm (SVA) is defined as a significant enlargement of one or more of the aortic sinuses located between the aortic valve annulus and the sinotubular junction. The estimated incidence is approximately 0.09% in general population. Congenital aneurysms are more prevalent than acquired ones [1]. Usually the right coronary sinus is affected (70-90%). Unruptured SVA is often asymptomatic but occasionally can cause severe complications due either to the compression of adjacent cardiac structures or to its rupture [1,2].

Transthoracic echocardiography (TTE) is the modality of choice for SVA screening, with a high sensitivity and more than 90% diagnostic accuracy. Transesophageal echocardiography (TEE) due to its better acoustic window is strongly recommended and can improve the image quality, revealing fine structures (e.g. thrombus, vegetations, small aneurysmal defects) [2].

Case report

A 49-year-old man was admitted with atypical chest pain unrelated to physical activity and radiating into the right shoulder. There was no history of fever, collagen or vascular diseases. The physical examination was unremarkable. Electrocardiogram showed no signs of ischaemia and the chest radiography described cardiomegaly with right atrium enlargement.

Routine TTE, performed with a GE Vivid S6 echo system using a 1.5-3.6 MHz probe, discovered severe aortic root dilatation at the level of the sinus of Valsalva with a large saccular structure arising from the right sinus; the aortic valve was tricuspid with trivial aortic regurgitation, ascending and descending aorta were within normal limits (fig 1a,b). The colour Doppler mode detected turbulence in the right ventricular outflow tract (RVOT) with a dynamic gradient of 23 mmHg and a peak velocity of 2.5 m/s measured by continuous wave Doppler mode.

A TEE was performed (GE Vivid 6 echo system, 2.9-8 MHz probe). The bi-dimensional images at mid-esoph-
ageal level confirmed the presence of a right coronary sinus aneurysm, with maximum dimensions of 53x51 mm, with intrasaccular vortex, with layered thrombus along its walls. We followed the continuity of the SVAs walls and no sites of rupture were detected. The aneurysm was protruding into the right ventricle, with distortion of RVOT. Congenital defects such as ventricular septal defects, aortic regurgitation and bicuspid aortic valve were excluded (fig 1c,d).

For precisely delineating the anatomic relationships of the aneurysm and for evaluation of the coronary anatomy, a contrast enhanced cardiac computed tomography (CT) was performed. The aneurysm was not homogeneously opacified by the contrast and was protruding into the right cavities (fig 2). The right coronary artery (RCA) was described as thin, filiform.

Coronary angiography was considered necessary to distinguish the sinus aneurysm from other coronary

Fig 1. Transthoracic echocardiography: a) apical 5-chambers view: right sinus Valsalva aneurysm (arrow); b) parasternal short axis view at great vessels level: aneurysm measuring 55x70 mm. Transesophageal echocardiography – mid-esophageal level: c) long axis view of the aortic valve and ascending aorta: large right sinus of Valsalva aneurysm (arrow), with vortex; d) short axis view of the aortic valve, colour Doppler: aneurysm of the right sinus Valsalva (yellow arrow) and a tricuspid aortic valve (white arrow).

Fig 2. Contrast-enhanced cardiac computed tomography and 3D reconstruction: a) axial, b) coronal and c) sagittal reconstructed images: right sinus of Valsalva aneurysm (arrow) with nonhomogeneous opacification of the contrast; d) 3D reconstructed CT angiography: giant aneurysm of sinus Valsalva is clearly seen (arrow).

Fig 3. Aneurysm of the right sinus of Valsalva (arrows): a) coronary angiography; b) intraoperative image

Fig 4. Postoperative images. Transesophageal echocardiography: a) mid-esophageal level, long axis view of the aortic valve and ascending aorta: aortic root with neosinus reconstruction (arrow); b) mid-esophageal level, short axis view of the aortic valve: normal aortic valve; c) and d) contrast-enhanced cardiac computed tomography and 3D reconstruction: tubular Dacron prosthesis between the aortic root and ascending aorta.
anomalies (e.g. a coronary aneurysm). A nonselective injection in the right sinus showed absence of flow in the RCA. The selective injection in the left main showed normal left anterior descendant and normal left circumflex coronary artery with well developed collaterals completely filling the right coronary artery until its proximal segment (fig 3a).

The patient was referred for cardiac surgery. An aneurysmectomy with interposition of a tubular Dacron prosthesis (30 mm) between the aortic root and the ascending aorta was performed; the proximal section of the tubular graft was tailored to recreate the excised right coronary sinus. The reimplantation of the native right coronary artery was not technically possible and RCA was bypassed with a saphenous graft (fig 3b). Cross clamp time was 116 minutes, cardiopulmonary bypass time was 147 minutes. Histopathologic examination of the resected aneurysmal tissue confirmed a congenital etiology, with a deficiency of elastic fibers and mucoid deposits.

The postoperative course was uneventful. The patient was discharged on the 11th postoperative day. At the one-month follow-up the patient was asymptomatic; TTE, TEE and CT showed successful repair: an aortic root and ascending aorta with normal dimensions and a normally functioning aortic valve (fig 4).

Discussions

Published data about cases with coronary insufficiency secondary to SVA is scarce. An aggressive surgical approach is recommended to prevent acute coronary obstruction. In our patient the ostium of the RCA was obstructed, therefore the antegrade flow in the RCA was absent, but the vessel had a good opacification via contralateral collaterals developed as a consequence of a long-term compression [3,4].

The natural history of SVAs is difficult to be determined because of their rarity. Without surgery, many complications can be expected: rupture with sudden cardiac death, malignant arrhythmias, aortic regurgitation and infection. As in our case, unruptured SVAs are typically asymptomatic and there are no guidelines regarding the timing of surgical intervention; we decided to operate the aneurysm because its enlargement exceeded 50% of the normal sinus size [5,6].

The non-invasive diagnosis of SVA should include a complete echocardiographic examination (TTE, TEE) with the description of its origin, continuity of the aneurysmal wall, compression or obstruction caused by the SVA, severity of valvular insufficiency (aortic and tricuspid valves). The recommended key views are: parasternal long-axis view, short axis view of the aortic root, long axis view of RVOT, apical 5-chamber view. TEE, due to the probe’s proximity to the aortic root, is more sensitive than TTE in describing morphological details, such as presence of thrombus, vegetations, rupture and is more suitable for differential diagnosis (e.g. with pseudoaneurysm secondary to infective endocarditis, coronary artery aneurysm) [3,7,8].

In some cases, echocardiography may be extremely challenging and difficult because of a variable aneurysmal course with multiple possibilities of extension into several positions; therefore, it is recommended to complete the imagistic evaluation with 3D-echocardiography, cardiac CT or magnetic resonance imaging (MRI). In our case, cardiac CT demonstrated that RCA was not obstructed by thrombus but compressed by the SVA. This case is one of the very few where the main mechanism of reduced RCA flow was proven to be compression exercised by the SVA [3,4,9].

In conclusion, the combination of echocardiography (TTE, TEE, 3D-TEE) with other imaging techniques (CT, MRI, cardiac catheterisation) is recommended for an accurate assessment of SVA and a precise preoperative diagnosis.

References