

# Ruptured aortic sinus aneurysm with left coronary artery aneurysm-right atrial fistula

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May 18, 2022

## Abstract

A 50-year-old female presented with a history of fatigue after activities. Transthoracic echocardiography showed a 2.8 cm dilatation at the ostium of the left coronary artery and a 7.1×7.4 cm cyst-like aneurysm at the upper back of the left atrium, interlinking an anomalous fistula from the left coronary artery to the right atrial appendage, accompanied by moderate mitral and tricuspid regurgitation. Cardiac 3D-computed tomography identified the diagnosis of ruptured aortic sinus aneurysm (RASA) and a giant left coronary artery aneurysm fistula to the right atrium. Transesophageal echocardiogram and surgical vision confirmed these findings. Surgeries including RASA repair, coronary artery bypass grafting, and mitral and tricuspid valvuloplasty were then performed. Procedures proceeded successfully, the patient was fully recovered and discharged home.

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**KEYWORDS:** Rupture of aortic sinus aneurysm, Coronary artery aneurysm, Right atrium fistula

## 1 – INTRODUCTION

Coronary artery anomaly (CAA) is a common cause of sudden cardiac death, a congenital disease of abnormal coronary origin, course, and termination <sup>1</sup>. Coronary artery fistula (CAF) is an abnormally terminated CAA with a low prevalence (0.2-0.6%) and is usually detected incidentally during the diagnostic examination for ischemic heart diseases <sup>2</sup>. Ischemic symptoms of CAF are caused by coronary steal where coronary blood

flows into low-pressure areas of the fistula (eg, right ventricle, right atrium, pulmonary artery, or coronary sinus), resulting in distal myocardium ischemia. The latest data show that CAF accounts for 0.32% of total coronary anomalies in Southwest China as determined by angiography<sup>3</sup>. Especially, a giant left coronary artery aneurysm that fistula to the right atrium and accompanied by ruptured aortic sinus aneurysm (RASA) is rarely reported. Notably, even asymptomatic coronary fistulas can lead to severe complications, such as endocarditis, coronary aneurysms, myocardial infarction, or cardiac arrest; therefore, catheter-based closure or surgical therapy is often required.

## 2 – CASE PRESENTATION

A 50-year-old female presented to a community clinic due to fatigue after activities and was primarily diagnosed with giant left coronary artery aneurysm. On admission, she had not received surgery or special medication previously. Physical examination and electrocardiogram revealed left-inferior enlargement of the cardiac border, left ventricular high voltage, and a grade 3/6 systolic murmur could be heard in the auscultatory mitral area. Chest X-ray showed left atrioventricular enlargement and right coronary calcification. Transthoracic echocardiography (TTE) indicated a 7.1x7.4 cm cyst-like aneurysm at the upper back of the left atrium, which originating from the left coronary artery, detouring to the left posteriorly, and breaking into the right atrial appendage; accompanied by moderate mitral and tricuspid regurgitation (FIGURE 1). Concurrently, the left coronary artery ostium dilated up to 2.8 cm, and the possibility of RASA should be further precluded. Then cardiac three-dimensional computed tomography (3D-CT) was recommended to reconstruct the aneurysm. Cardiac 3D-CT revealed a RASA of the left coronal sinus and an aneurysm-like fistula connecting the left main coronary artery with the right atrium, squeezing the right pulmonary vein and left atrium (FIGURE 2). During operation, transesophageal echocardiogram (TEE) and surgical vision confirmed these findings (FIGURE 3, VIDEO 1-3). Under cardiopulmonary bypass, the patient underwent RASA repair, coronary artery bypass grafting (great saphenous vein graft to left anterior descending and circumflex branches), mitral and tricuspid valvuloplasty, and temporary pacemaker implantation. Following operation, TEE confirmed that there was no flow in the right atrial fistula, and the aneurysm in the posterior wall of the left atrial was collapsed. The patient was fully recovered and discharged home at 14th day after operation.

## 3 – DISCUSSION

Of all types of CAF, giant left coronary aneurysm with right atrium fistula is uncommon<sup>4</sup>. It has been reported that 90% of CAF originated from the right coronary artery, of which only 0.2% were giant coronary aneurysms (> 4 times of the normal diameter)<sup>5</sup>. CAF often has no specific clinical manifestations, with or without angina pectoris and cardiac murmur, which is closely affected by the location and size of the fistula. The optimal treatment for CAF remains controversial, the most common approaches are transcatheter closure of the fistula, surgical resection of the aneurysm, and bypass grafting of the coronary artery. Multimodal imaging and functional testing play vital roles in the diagnosis of CAF. Specially, coronary angiography and cardiac CT are universal methods for diagnosing and tracking the anatomical abnormalities of coronary. However, coronary angiography is invasive, and CT often misses tiny breaks. In this study, we proved that TTE and TEE are necessary and sufficient to determine the complex anatomy of coronary fistula and its relationship to adjacent structures, and can be used for preoperative, intraoperative, and postoperative assessments. Echocardiography has the advantages of non-invasive, real-time, and inexpensive, especially in detection of the rupture and location of fistula by color Doppler. It is worth noting that the echocardiographic images in this case need to be differentiated from the three-atrial heart, and the apical four-chamber view of TTE may easily mistake the aneurysm wall for the right atrial septum and the fistula for the septal hole.

## CONFLICT OF INTEREST

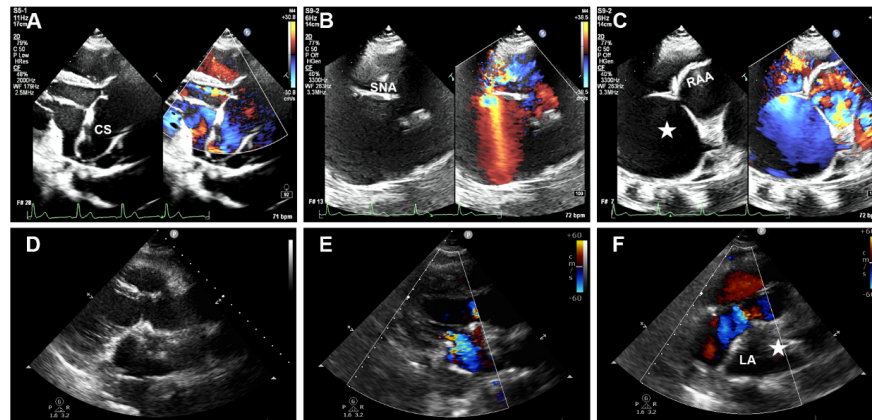
No conflict of interest.

## ORCID

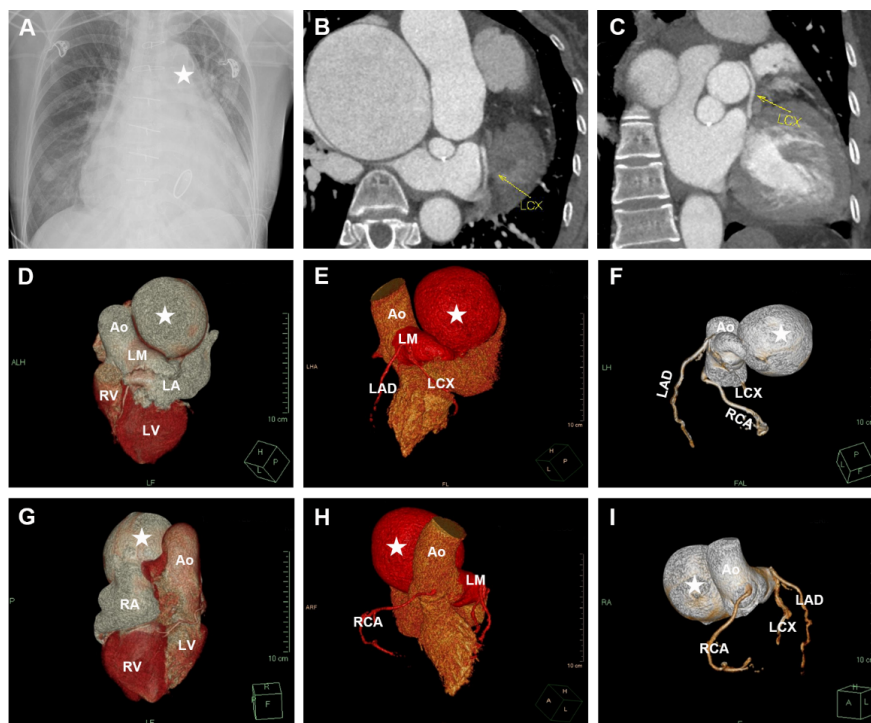
*Wei Xiong MD* <https://orcid.org/0000-0002-4899-6332>.

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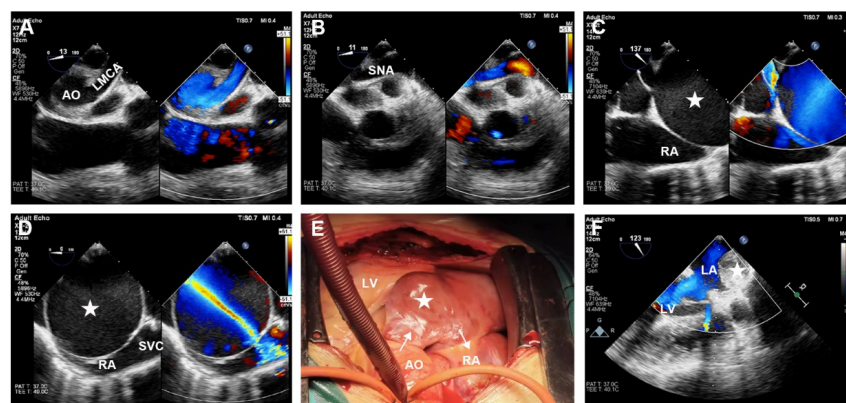
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**FIGURE 1** Pre- and postoperative TTE images. (A) Pre-TTE parasternal long-axis view reveals dilated left coronary sinus (CS), (B) Color Doppler imaging with systolic aliased flow in the sinus node artery (SNA). (C) Apical four-chamber view shows a 7.1×7.4 cm giant aneurysm (white star) fistula into the right atrial appendage (RAA). (D-F) Post-TTE shows a hypoechogenic mass of collapsed aneurysm (white star) in the posterior of the left atrium (LA).



**FIGURE 2** Multimodality image examination. (A) Chest X-ray shows a mass (white star) abutting the left atrium border. (B-C) Cardiac CT reveals a giant aneurysm in the distal of left coronary. (D-I) Cardiac 3D-CT indicated the origin and course of coronary arteries were normal. The left coronary sinus sent out a thick blood vessel branch, connecting a giant left main (LM) coronary artery aneurysm (white star) fistula to the right atrium. AO, aorta; LAD, left anterior descending; LCX, left circumflex; RCA, right coronary artery; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.



**FIGURE 3** Intraoperative TEE and surgical images. (A) Pre-TEE shows an aneurysmal dilation of left main coronary artery (LMCA) origin. (B) Color Doppler imaging with systolic aliased flow in the sinus node artery (SNA). (C-D) Pre-TEE bicaval view reveals an aneurysm-like fistula from left main coronary to right atrium (RA), which adjacent to superior vena cava (SVC). (E) A collapsed left coronary aneurysm under cardiopulmonary bypass. (F) Post-TEE long-axis view reveals a collapsed aneurysm in the posterior of the left atrium. White star represents aneurysm.

**VIDEO 1** Mid-esophageal four chamber view demonstrating an aneurysmal dilation of left coronary sinus of aorta with moderate mitral and tricuspid regurgitation.

**VIDEO 2** Mid-esophageal bicaval view showing an aneurysmal fistula drain into right atrium.

**VIDEO 3** Mid-esophageal long-axis view revealing a collapsed aneurysm in the posterior of the left atrium.

