

Right coronary artery fistula to the coronary sinus and right atrium associated with giant right coronary enlargement detected by transthoracic echocardiography

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We present the case of a 54-year-old female with a previous history of lung fibrosis secondary to methotrexate used for rheumatoid arthritis who was referred to cardiology evaluation due to precordial pain. Echocardiography showed biatrial enlargement with an enlarged coronary sinus and tubular image posterior to the heart. On the coronary angiogram, the right coronary artery was enlarged, and a distal fistula was identified. The patient underwent a contrast enhanced cardiac computed tomography which demonstrated an aneurysmatic right coronary artery with a distal fistula to the right atrium and coronary sinus. As the chest pain did not recur and there was a high risk of the intervention to correct coronary fistula, the patient remained on conservative treatment.

Keywords

Coronary fistula • Coronary anomaly • Cardiac imaging • Cardiac computed tomography • Coronary angiography

A 54-year-old female came to the outpatient clinic complaining of recurrent chest pain. The patient had a previous medical history of lung fibrosis secondary to methotrexate which she used for rheumatoid arthritis. No other risks factors were present. The physical examination was normal and the electrocardiogram was unremarkable. Echocardiography showed biatrial enlargement and a mildly aneurysmatic coronary sinus. No atrial septal defect was seen. However, colour-coded echocardiography demonstrated a vascular structure posterior to the heart (Figure 1). To exclude significant coronary artery disease, an invasive coronary angiography was performed. The left coronary artery system was normal. The right coronary artery was impressively enlarged, and a distal fistula was identified (Figure 2). Owing to its complex anatomy, invasive angiography did not clearly define the distal course and drainage of the coronary fistula. Therefore, the patient underwent contrast-enhanced computed tomography (CT) using a dualsource CT system (Definition Flash, Siemens Healthcare, Forchheim, Germany). CT coronary angiography demonstrated a significant enlarged and calcified right coronary artery with a

distal fistula connected to the right atrium and the coronary sinus (*Figure 3*).

Discussion

Coronary artery anomalies are present in invasive coronary angiograms with a prevalence of 1.3%.¹ Among them, coronary fistulas are a much rarer presentation, about 0.2%.² Most of them are accidental findings on angiography. The initial documentation of the fistula on echocardiography, as in this patient, is extremely rare. Angiography is usually useful to evaluate the fistula's clinical significance. The cardiac CT, on the other hand, is being increasingly used to evaluate those patients, as it is able better illustrate the fistula's three-dimensional structure and provide a better understanding of it, as demonstrated in this patient.

Most coronary fistulas originate from the right coronary artery and drain into the right heart, preferably in the right ventricle.³ Up to 24% of the patients demonstrate drainage into the right atrium or central veins,³ like the patient presented here.

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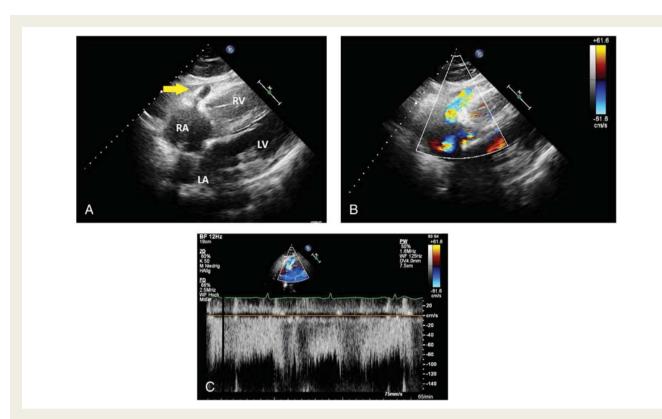


Figure I (A) Subcostal two-dimensional view demonstrated the fistula (arrow) draining into the right atrium. (B) Colour-Doppler of the same image shows the blood flow in the fistula. (C) A continuous Doppler curve of the fistula. RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle.

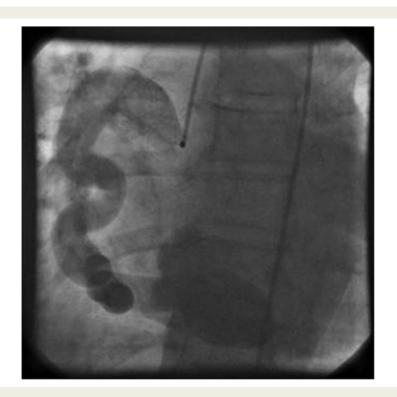


Figure 2 Right coronary artery angiogram shows an extremely enlarged coronary artery. A reliable anatomic definition of the distal course and the drainage of the fistula is not possible.

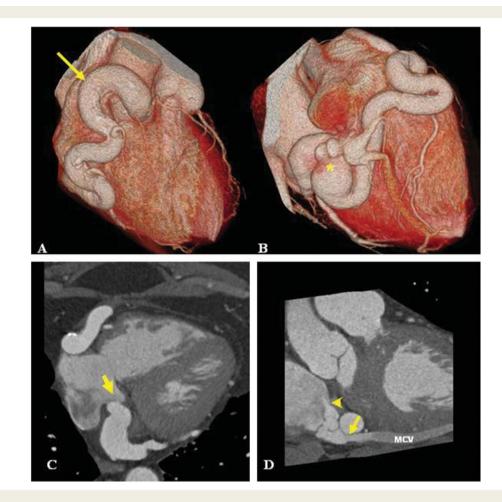


Figure 3 (A) Three-dimensional reconstruction (volume rendering technique, VRT); anterior view. The significantly enlarged giant right coronary artery (arrow) is demonstrated. (B) Three-dimensional reconstruction (VRT); posterior view. The fistula drains into the coronary sinus (asterisk at the end of the fistula into the coronary sinus). (C) Maximum intensity projection image (slice thickness, 5 mm) demonstrates the drainage of the fistula into the right atrium (arrow). (D) Multiplanar reconstruction showing a connection between the fistula and the middle cardiac vein (arrow). Finally, the fistula drains into the right atrium (arrowhead). MCV, middle cardiac vein.

The majority of the patients with coronary fistula remain asymptomatic. When symptoms occur, they are usually related to the volume overload from large left to right shunts. Although some authors suggest surgical closure or coil embolization even in asymptomatic patients, ⁴ this is not a widely recommended strategy, as most patients can adequately be managed conservative unless a significant volume overload is present. In the demonstrated patient, no surgical or interventional therapy has been recommended, not only because the intervention was considered high risk, but also because the patient remained asymptomatic after the evaluation. The patient will be followed by routine clinical and echocardiographic evaluation.

Conflict of interest: none declared.

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