Giant Aneurysm of the Left Circumflex Coronary Artery with Fistulous Communication to the Right Atrium: Multimodal Cardiac Imaging

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A healthy 36-year-old man visited his family doctor with abdominal discomfort. Electrocardiogram revealed right-axis deviation and a continuous murmur was heard at the third intercostal space left sternal border. Transthoracic echocardiogram demonstrated a continuous color jet draining into the enlarged right atrium (RA). Subsequent cardiac computed tomographic images and coronary angiogram revealed a giant left main trunk and left circumflex coronary artery (LCx) with intraluminal thrombi and fistulous communication to the RA. Cardiac magnetic resonance imaging measured a Qp/Qs of 1.24. A multimodal imaging approach helped diagnose this rare cameral fistula (LCx-to-RA) complicated by giant coronary aneurysm and intraluminal thrombi.

Keywords: Coronary aneurysm; Fistula; Multimodal imaging

INTRODUCTION

Communications between the coronary arteries and the cardiac chambers (coronary–cameral fistulae) are rare forms of congenital heart disease. The sites of origin are the right coronary artery (55%), the left coronary artery (35%), or both coronary arteries (5%); the major termination sites are the right ventricle (40%), right atrium (RA) (26%), or pulmonary arteries (17%) [1]. Symptoms of angina or congestive heart failure may occur at the time of diagnosis but most present with subclinical or atypical symptoms. Therefore, the diagnosis is often delayed or incomplete unless multimodal imaging is used. We present an asymptomatic case of an unusual giant left circumflex coronary artery (LCx) aneurysm with fistulous communication towards the RA complicated with intraluminal thrombosis, all of which was fully delineated by multimodal cardiac imaging.

CASE

A 36-year-old man came to the family medicine clinic complaining of abdominal discomfort. After 3 days of gastrointestinal medications, the abdominal discomfort has gone, but there was a continuous murmur at the 3rd intercostal space left sternal border, which brought a suggestion of cardiac diseases such as patent ductus arteriosus, pulmonary valvular stenosis-regurgitation, or coronary arteriovenous fistulae. His
blood pressure was 115/70 mmHg with a heart rate of 88 beats/min. His electrocardiogram showed a normal sinus rhythm with right-axis deviation without evidence of acute coronary syndrome. His simple chest X-ray showed a prominent pulmonary conus but was otherwise within normal limits. Laboratory tests were as follow: plasma brain natriuretic peptide, 16.4 pg/mL; total cholesterol, 273 mg/dL; low-density lipoprotein cholesterol, 208 mg/dL; high-density lipoprotein cholesterol, 43.0 mg/dL; and triglyceride, 203 mg/dL. He was an ex-smoker without any significant past medical history.

Figure 1. Transthoracic 2D echocardiography. (A) Parasternal short-axis view of the aortic valve level reveals a markedly dilated left main coronary artery ostium (LM). The right coronary artery ostium (RCA) appears normal (Supplementary Video 1). (B) Apical four-chamber view shows dilated right atrium (RA) and prominent left circumflex coronary artery (asterisk). (C) Apical two-chamber view shows parts of the dilated left circumflex coronary artery (asterisks). RVOT, right ventricular outflow tract; RC, right coronary cusp; NC, non-coronary cusp; LC, left coronary cusp of the aortic valve; LA, left atrium; LV, left ventricle; RV, right ventricle.

Figure 2. Transthoracic Doppler echocardiography. (A) RV-modified apical four-chamber view reveals a color-Doppler jet (arrow) draining into the right atrium (RA) from the dilated left circumflex artery (LCx) (Supplementary Video 2). (B) Continuous wave-Doppler demonstrates a continuous flow jet to the RA, measuring a maximum velocity of 4.1 m/s and a mean pressure gradient of 55.6 mmHg. RV, right ventricle; LV, left ventricle.
regular medications and no hypertension or diabetes mellitus.

Referred to the cardiology clinic, non-invasive transthoracic echocardiography was performed to figure out the cause of the continuous murmur. There was no regional wall motion abnormality with normal left ventricular dimension and function (ejection fraction of 68%, normal diastology). The left main coronary artery ostium was markedly dilated, and the prominent LCx was noted in the routine 2D echo views (Fig. 1).

In color-Doppler echo imaging, there was a continuous jet flow draining into the enlarged RA, presumptively from the dilated LCx, with a mean pressure gradient of 55.6 mmHg (Fig. 2). Subsequent coronary computed tomography angiography (CCTA) revealed a coronary artery anomaly: an LCx to RA fistula (diameter of 2.5 mm) and a coronary aneurysm (diameter of 20 mm) with calcified walls and intraluminal thrombi (Fig. 3). There was no stenosis in the right and left anterior

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**Figure 3.** Coronary computed tomography angiography (CCTA). (A, B) CCT demonstrates a markedly dilated left main (LM) and left circumflex coronary artery (LCx) with calcified walls and intraluminal thrombi (arrows) and a small fistula (diameter of 2.5 mm) with contrast leakage (asterisk) to the right atrium (RA). (C, D) Three-dimensional volume-rendered CCTA images reveal a coronary aneurysm (from LM to LCx). Other coronary arteries are within normal range. RV, right ventricle; LV, left ventricle; AV, aortic valve; RCA, right coronary artery; LAD, left anterior descending coronary artery.
Figure 4. Contrast-enhanced cardiac magnetic resonance (CMR) imaging. (A) Reformatted CMR angiographic imaging demonstrates a markedly dilated left main (LM) and left circumflex coronary artery (LCx) with intraluminal thrombi (arrows) and a contrasting small jet (asterisk) into the right atrium (RA) (Supplementary Video 3). (B) Delayed contrast-enhanced CMR imaging demonstrates no delayed hyper-enhancement in the left ventricle (LV) and right ventricle (RV) myocardium. RCA, right coronary artery; AV, aortic valve.

Figure 5. Coronary angiography. (A) Coronary angiography shows a markedly enlarged left main (LM) and left circumflex coronary artery (LCx) with the formation of a giant aneurysm at the distal LCx with a diameter of 20 mm. The left anterior descending coronary artery (LAD) appears normal without stenosis. The huge LCx has branches of obtuse marginal arteries (OM). The contrast dye drains into the right atrium continuously, and some dye stasis in the distal LCx with a filling defect (arrows) suggests thrombi (Supplementary Video 4). (B) On aortic root aortography, a coronary aneurysm with direct drainage into the right atrium through the small distal orifice (asterisk) is well visualized (Supplementary Video 5) with a normal right coronary artery (RCA).
descending coronary arteries. Cardiac magnetic resonance imaging (CMR) revealed no delayed hyper-enhancement of the left and right ventricular myocardium, and a Qp/Qs of 1.24 (Fig. 4). Finally, a coronary angiogram confirmed a giant coronary aneurysm from the left main trunk to the LCx with direct drainage to the RA through the distal small orifice (1.0–2.0 mm) (Fig. 5).

The 5-year follow-up was uneventful under aspirin and high-dose statins, with a resolution of the intraluminal thrombi in the LCx but increased diameters of the coronary artery aneurysm (maximal diameter of 20 mm increasing to 24 mm) on the CCTA. He’s not complaining of any ischemic or heart-related symptoms. Follow-up echocardiography showed no significant interval changes in the enlarged RA and estimated pulmonary arterial systolic pressure by trivial tricuspid regurgitation being 23 mmHg.

DISCUSSION

We described a case of a small coronary-cameral fistula (from the LCx to the RA) combined with a giant coronary aneurysm and intraluminal thrombi. Although most patients with coronary fistulae are asymptomatic, a hemodynamically significant left-to-right shunt may result in potential myocardial ischemia due to a coronary-steal phenomenon, possibly resulting in heart failure or pulmonary hypertension due to an excessive volume load on cardiac chambers [2]. Early anatomic correction may be considered when the shunt amount is significant (Qp/Qs $>1.5$) to prevent late symptoms and complications [3]. In this case, an asymptomatic patient with a small–size cameral fistula having Qp/Qs of 1.24 by CMR, normal ventricular size and function without resting pulmonary hypertension warranted a medical follow-up.

Approximately 20–25% of coronary artery fistulae are associated with aneurysmal dilatation; the prevalence of a giant aneurysm (defined by a diameter of ≥ 2 cm) is only 0.02%, and spontaneous rupture is extremely rare [4,5]. This case showed an unusual giant aneurysm combined with intraluminal thrombi, resolved with antiplatelet medication. Because coronary artery aneurysms more than 3 cm in diameter are at risk of rupture, close CCTA monitoring of growth is essential [6]. Ultimately, elimination of a coronary artery fistula (either surgical or transcatheter) is recommended by the American Heart Association/American College of Cardiology when the fistula size is large or when small to moderate but with complications (myocardial ischemia, arrhythmias, ventricular dysfunction of unexplained etiology) [7].

For diagnosis, in this case, the auscultation of a loud continuous murmur played a key role in suspicion, but relevant multimodal cardiac images identified the coronary fistula and associated anomalies both anatomically and functionally. Transthoracic echocardiography is considered a proper initial imaging technique that can reveal enlarged coronary arteries or abnormal color–Doppler flow at the termination site of the shunt. Although it is difficult to delineate the entire course of the coronary anomaly via echo, echocardiography gave useful information regarding whether there were other associated cardiac lesions (for example, tetralogy of Fallot, patent ductus arteriosus, or an atrial septal defect) or corresponding structural or functional changes (chamber enlargement, myocardial dysfunction, or elevated intracardiac pressure) [2,8]. In this case, there was no other congenital anomaly and the terminal part of the shunt chamber (RA) was enlarged but there was no heart failure or significant resting pulmonary hypertension. The CCTA had the advantage of revealing the exact delineation of the origin and termination of the fistula through the entire course, including, in this case, an associated coronary aneurysm and intraluminal thrombi which were not defined by transthoracic echocardiography. CMR confirmed the presence of the fistula and added more information such as there are no areas of delayed hyper–enhancement suggestive of previous myocardial infarction and an estimate of the shunt amount (Qp/Qs). Ultimately the size and anatomical features of the fistula were reliably confirmed by invasive coronary angiography.

The present case highlights the importance of multimodal cardiac imaging in patients with a continuous murmur either to rule out a coronary fistula or to rule it in with exact delineation of the origin and termination as well as any complication [8].
REFERENCES


