Surgical treatment of heart failure due to giant coronary artery fistula: a case report

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Abstract

Coronary artery fistula is a rare congenital cardiac anomaly that is often found incidentally during computed tomography angiography. Coronary fistula between the left circumflex coronary artery and the coronary sinus is among the less common forms of coronary artery fistula. A 60-year-old female patient presented to our outpatient cardiology department with symptoms of severe, de novo heart failure. Echocardiogram revealed severe mitral regurgitation and a dilated duct that turbulently accelerated colour Doppler flow behind the left ventricle with significant left-to-right shunt. Cardiac magnetic resonance imaging and computed tomography angiography revealed a massively dilated fistula between the left circumflex coronary artery and the coronary sinus with a diameter of 3–4 cm. The patient underwent combined heart surgery involving mitral ring annuloplasty and fistula ligation and was discharged in stable condition on guideline-based medical therapy. At 18 months of follow-up, minimal residual shunt flow and mild-to-moderate mitral regurgitation were found. We report a rare case of congenital coronary disorder resulting in heart failure and highlight the importance of complex non-invasive cardiac diagnostic procedures before planning and performing heart surgery.

Keywords: Coronary artery fistula; Heart failure; Surgery

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Introduction

Coronary artery fistula (CAF) is a rare anomaly of the coronary system that is characterized by a connection between one or more coronary arteries and a cardiac chamber or a great vessel of the systemic or pulmonary circulation.1 CAF occurs in 0.002% of the general population and accounts for 0.2–0.4% of all congenital heart disease cases. The diagnosis of CAF is often made incidentally during coronary angiography, with a prevalence of 0.05–0.9%.2,3 The appropriate diagnosis of CAF requires detailed non-invasive cardiac imaging including 2D and 3D echocardiography and cardiac magnetic resonance imaging (MRI) or coronary computed tomography angiography (CTA), as well as invasive procedures such as coronary angiography with or without fractional flow reserve measurements. A comprehensive diagnostic workup is crucial for decision-making regarding the need for transcatheter interventions or surgical procedures.4,6

Coronary fistula between the left circumflex coronary artery (LCX) and the coronary sinus (CS) is a less common form of CAF that may be asymptomatic or associated with various symptoms depending on its size. Surgical or transcatheter intervention is only indicated for extremely large fistulae or in the presence of severe dyspnoea, myocardial ischaemia, arrhythmia, aneurysm rupture, or congestive heart failure.2–6

Case report

A 60-year-old female patient presented to our department with severe dyspnoea, orthopnoea, and tachycardia. Her past medical history was unremarkable for cardiac disease and included childhood tuberculosis, bronchial asthma, and partial thyroidectomy due to goitre. Physical examination revealed extensive rales in the lung bases, and a continuous, grade
4/6 murmur all over the pericardium and lower left sternum that radiated to the left axilla and had a maximum intensity over the mitral area. No peripheral oedema was found; functional status was New York Heart Association III–IV. Electrocardiogram showed sinus rhythm with a heart rate of 95 b.p.m., as well as incomplete left bundle branch block, and left ventricular hypertrophy with strain. Routine laboratory test results were normal; however, we found elevated N-terminal pro-brain natriuretic peptide (NT-proBNP) level (1763 pg/mL). Chest X-ray showed cardiomegaly with mild bilateral pleural effusion and congestion. Initial transthoracic echocardiography (TTE) showed reduced left ventricular ejection fraction (LVEF, 46%) and dilated left and right ventricles with end-diastolic diameters of 65 and 42 mm and end-systolic diameters of 44 and 34 mm, respectively. TTE also revealed enlargement of the left and right atria (54 × 65 mm and 42 × 63 mm, respectively), dilated mitral annulus (accordingly severe secondary mitral regurgitation (MR), effective regurgitant orifice area (EROA): 0.31 cm², right ventricle: 55.2 mL), pseudo-normal left ventricular inflow (E/A: 1.33/0.81), and moderate tricuspid regurgitation with elevated pulmonary artery systolic pressure (PASP) (47 + 8 = 55 mmHg). 2D and 3D TTE images suggested coronary aneurysm with significant left-to-right shunt (Qp/Qs: 2.159) (Figure IA and IB, Supporting Information, Video S1). Cardiac MRI was performed after the initiation of routine heart failure therapy (angiotensin-converting enzyme inhibitor, beta-blocker, and loop-diuretics), which showed a dilated, hypokinetic left ventricle, severe MR, and an abnormality in the posterior interventricular sulcus (Figure 2, Supporting Information, Video S2). High-resolution black-blood contrast-enhanced T1 weighted images verified vessel origin and blood flow in the anomaly; however, the exact origin and termination site could not be determined. CTA showed no signs of significant atherosclerotic coronary artery disease (calcium score: 21) but revealed an aneurysmal enlargement of the circumferential artery with a maximal diameter of 3–4 cm, draining most of the left coronary artery flow into the CS (Figure 1C and 1D).

**FIGURE 1** Preoperative 2D and colour Doppler echocardiographic images (parasternal long axis view) of the coronary artery fistula (CAF) and mitral regurgitation (A). 3D images of the CAF (B). 3D reconstruction images of the CAF (C), and reduced calibre of the left anterior descending artery (D).
After 1.5 months of optimized heart failure therapy (angiotensin-converting enzyme inhibitor dose titration to 50% and beta-blocker dose titration to 75% of target dose), the patient experienced a deterioration of heart failure symptoms that required intervention. Transcatheter procedures were not feasible due to the size of the CAF and the degree of tortuosity; therefore, combined heart surgery was performed involving CAF ligation and mitral ring annuloplasty due to the co-existence of mitral valve regurgitation. Cardiopulmonary bypass was applied with standard aortic and bicaval cannulation, moderate systemic hypothermia, and a single dose of anterograde cold blood cardioplegia. The mitral valve was explored through a transseptal incision extending to the roof of the left atrium. Considerable annular dilatation was detected without significant disorder of the leaflets or subvalvular apparatus. A 34 mm-sized semi-rigid annuloplasty ring was used for repair. To access the inflow of the arteriovenous shunt, the main pulmonary artery was completely transected, and the fistula was ligated just distal to the LCX orifice (Figure 3A and 3B). At the end of the first postoperative week, the patient developed atrial fibrillation with normal ventricular rate. Amiodarone therapy was started to restore and maintain sinus rhythm. Five days after successful cardioversion, the patient developed atrial flutter with highly variable conduction rates and symptoms of presyncope. Following re-admission to the coronary care unit, third-degree atrioventricular block developed. Temporary pacing was not required, and after withdrawal of beta-blocker and amiodarone, atrial fibrillation with normal ventricular rate returned. Later on, sinus bradycardia was noticed, which caused dizziness and weakness. Holter electrocardiogram confirmed tachy-brady syndrome; therefore, a DDD-R pacemaker was implanted after consultation with an electrophysiologist. The patient was discharged on guideline-recommended optimal medical therapy. Her pre-discharge echocardiogram showed mild MR and an LVEF of 47%, with a decrease in PASP (36 mmHg) and NT-proBNP (968 pg/mL).

At 18 months of follow-up, echocardiography and CTA showed an improvement of left ventricular function (end-diastolic diameter: 55 mm; end-systolic diameter: 37 mm; LVEF: 55%), mild-to-moderate mitral and mild tricuspid regurgitation, and normal right-sided chamber sizes with a tricuspid
annular plane systolic excursion (TAPSE) of 18 mm and an estimated PASP of 35 mmHg. CTA showed minimal residual shunt flow (Supporting Information, Video S3). Functional status was New York Heart Association I–II (NT-proBNP: 524 pg/mL). Consequently, catheter closure of the residual CAF was not indicated.

Discussion

A wide range of cardiac conditions can result in heart failure, the most common of which are hypertension, coronary heart disease, valvular heart diseases, and cardiomyopathies. In certain cases, rare underlying pathologies such as CAF further complicate the clinical course. CAF may be asymptomatic, but the gradual dilation of small fistulae may eventually lead to late-onset heart failure. Larger fistulae can steal blood from the coronary artery and drain blood flow into lower pressure chambers or veins, which leads to coronary artery ischaemia distal to the fistula. The resulting diastolic pressure gradient can only be compensated by the progressive enlargement of the draining vessel. The development of significant left-to-right shunt may result in further deterioration of clinical symptoms due to the reduction of coronary perfusion pressure.

The majority of coronary artery fistulae originate from the right coronary artery (40–60%) and mainly connect to the right atrium, right ventricle, or pulmonary trunk. Our case represents an uncommon CAF anatomy where the fistula originated from the LCX and connected to the CS. This form of CAF is mostly diagnosed in the elderly; however, neonatal cases have also been reported. Although TTE is suitable for the first identification of CAF, the most effective diagnostic modality is CTA, which provides better inferior temporal resolution compared with cardiac MRI and can help characterize the CAF in terms of size, feeding vessel, and termination site, which is essential for choosing between a transcatheter or surgical approach. In neonatal cases, the fistula should be treated conservatively as long as the clinical status is stable, or the child is growing. American College of Cardiology/American Heart Association guidelines recommend transcatheter or surgical closure for giant fistulae or in the presence of symptoms suggesting overt heart failure. A retrospective study of 119 CAF patients undergoing intervention showed no differences in safety and effectiveness between transcatheter and surgical closure. However, transcatheter closure was associated with a shorter length of hospital stay. A prospective study involving 29 patients with CAF did not observe severe complications after surgical ligation but highlighted the importance of thrombosis prophylaxis (i.e. anticoagulation) for at least 6 months after the procedure.

In our case, combined surgical intervention was needed due to progressive symptoms of heart failure, the size and tortuosity of CAF, and significant MR. The surgical repair of the mitral annulus and ligation of CAF resulted in the prompt amelioration of symptoms as well as in an improvement of echocardiographic parameters and heart failure biomarker levels.

The treatment of patients with CAF should be based on the severity of symptoms. If surgical management is required, CTA is the best diagnostic tool for describing the exact anatomical situation and for planning and performing precise and successful heart surgery. The appropriate diagnostic workup, interventional procedure, and perioperative care can help provide good quality of life without life expectancy limitations even for patients with severe heart failure.

**FIGURE 3** Surgery (A) resulted in normalized calibre of the left anterior descending artery (3D reconstruction image) (B).
Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. All co-authors have seen and agreed with the contents and have significantly contributed to the case report. All authors approved the submission.

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Conflict of interest

None declared.

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Supporting information

Additional supporting information may be found online in the Supporting Information section at the end of the article.

Video S1. Preoperative echocardiography images of MR.
Video S2. Preoperative MRI images of the dilated hypokinetic left ventricle and MR.
Video S3. Postoperative echocardiography images of MR.