

Severe Dilated Cardiomyopathy Resulted from a Large Single Coronary Artery Fistula Drained Into the Left Ventricle

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ABSTRACT

Background: Coronary artery fistula (CAF) draining into the left ventricle (LV) is a rare condition and dilated cardiomyopathy (DCMP) that results from single coronary artery (SCA) accompanied by CAF also is extremely rare.

Case report: We report the case of a 36-year-old man, who presented with severe DCMP that resulted from SCA with CAF draining into the LV. Transthoracic echocardiogram (TTE) showed severe diffuse hypokinesia of the LV with ejection fraction (EF) of 15-20%. Coronary angiography (CAG) revealed SCA connected between left anterior descending artery (LAD) and posterior descending artery (PDA), course of the PDA was very tortuous from apex to base of the LV, and connected to posterior lateral (PL) branch, which was drained into the LV at distal part of the PL. Coronary artery computed tomography (CACT) showed LAD ran over the apex of the LV and connected to PDA, which was drained into the mid portion of lateral wall of the LV. Cardiac magnetic resonance imaging (CMRI) showed no evidence of irreversible myocardial change in global wall of the LV. The patient underwent surgical ligation of PDA near the base of the posterior wall of the LV as close to the entry of CAF to the LV as possible without any surgery-related complications. Three months after the surgical ligation, follow-up TEE showed much improved EF of 45-50%. He has been doing well without congestive heart failure (CHF) until now.

Discussion: Symptomatic CAF with hemodynamic deterioration may need mechanical correction of CAF, including surgical ligation or percutaneous interventional occlusion. How to treat this condition in terms of methodology is a very difficult issue. The detailed methods related to surgical or interventional correction of CAF have to be determined based on anatomical characteristics of CAF, underlying comorbidities, and relevant complications risk.

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INTRODUCTION

CAF is a relatively rare disease in which the coronary artery forms a shunt to other blood vessels, ventricles, or atria. Although most of CAF are asymptomatic, it has various clinical manifestations, such as heart murmur, dyspnea, fatigue, angina, CHF, arrhythmia, pulmonary hypertension, and endocarditis based on amount and site of shunt. Although conservative management often is recommended in cases of asymptomatic CAF. Hemodynamic stress or cardiac damage resulting from a large amount of shunt may need aggressive surgical or transcatheter correction of CAF. In general, most of CAFs drain into the pulmonary artery, right atrium, or right ventricle. Only a few reports have described CAFs originating from the LAD artery and draining into the LV [Fernandes 1992; Urrutia-S 1983]. Moreover, cases where CHF resulting from CAFs draining into the LV are treated surgically are very rare [Kamiya 2002; Reul 2002]. We report a case presented with severe DCMP, due to a large amount of shunt by CAF originated from SCA and drained into the LV, and successfully treated with surgical ligation of CAF.

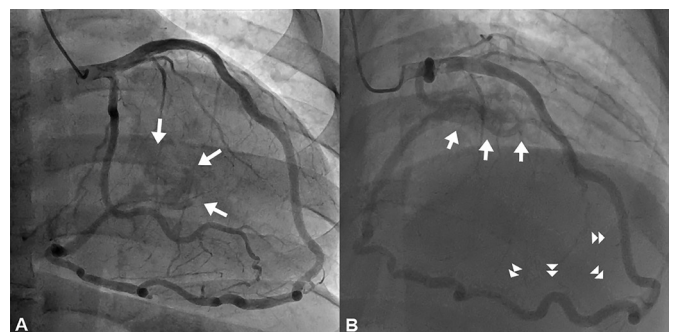


Figure 1. Selective left coronary angiography. A: Right anterior oblique view of the left coronary artery demonstrate direct drainage of PL into the LV (white arrows). B: Anterior-posterior cranial view of left coronary artery demonstrates that the PDA arising from the LAD has a very long and tortuous course (white double arrowheads) and is connected to PLA, which directly enters the LV (white arrows). LAD: left anterior descending artery, LV: left ventricle, PDA: posterior descending artery.

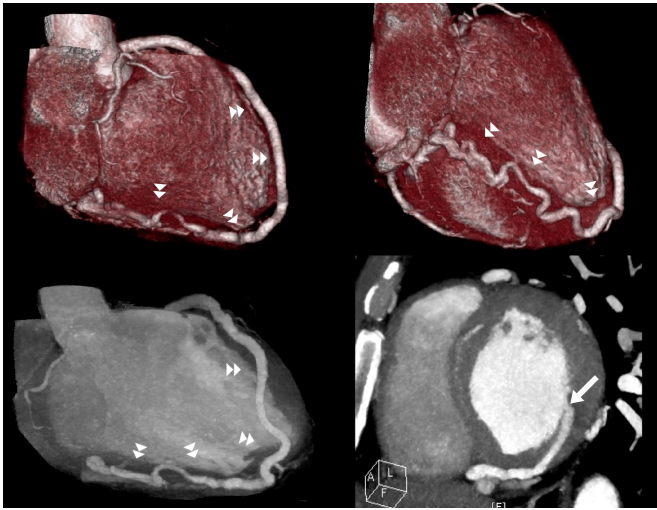


Figure 2. Coronary computed tomography shows the LAD artery extended over the apex of the LV and connected to PDA at the base of the LV (white double arrowhead) and then continues to PL, which was drained directly into the LV (white single arrow). LAD: left anterior descending artery, LV: left ventricle, PDA: posterior descending artery. PL: posterior lateral artery.

CASE REPORT

A 36-year-old man presented with dyspnea on exertion (DOE) (New York Heart Association class III/IV) that had aggravated over the span of a week. At the time of his visit to our clinic, his heart rate was 100 beats/min, blood pressure was 140/90 mmHg, and heart sounds were regular without any murmur. The patient did not have any particular medical history. He was a nonsmoker and did not heavily consume alcohol. Chest radiography revealed signs of cardiomegaly with pulmonary congestion, and NT-proBNP levels were elevated to 1949 pg/ml. Transthoracic echocardiography (TTE) revealed that the patient's EF was 15–20%, with a finding of severely diminished left ventricular global systolic function and dilated cardiac chambers. To rule out ischemic heart disease, CAG was performed, which demonstrated that SCA from LAD was connected to PDA with a very tortuous course from apex to base of the LV and then continued to the PL branch without any significant stenosis. The PL branch was drained into the LV at the distal part. The right coronary artery was hypoplastic and ended in mid-lateral wall of the right ventricle (Figure 1).

CACT showed that the LAD artery extended over the apex of the LV, was connected to the PDA, and then continued to the PL, which drained directly into the LV at the mid part of lateral wall of the LV (Figure 2).

For six months, the patient received intensive optimal medical therapy through heart failure medications, such as beta blockers, angiotensin receptor blockers and diuretics, including aldosterone antagonist. However, DCMP and relevant symptoms, such as DOE, did not improve despite optimal medical treatment. CMRI performed to investigate myocardial viability showed no evidence of myocardial necrosis or

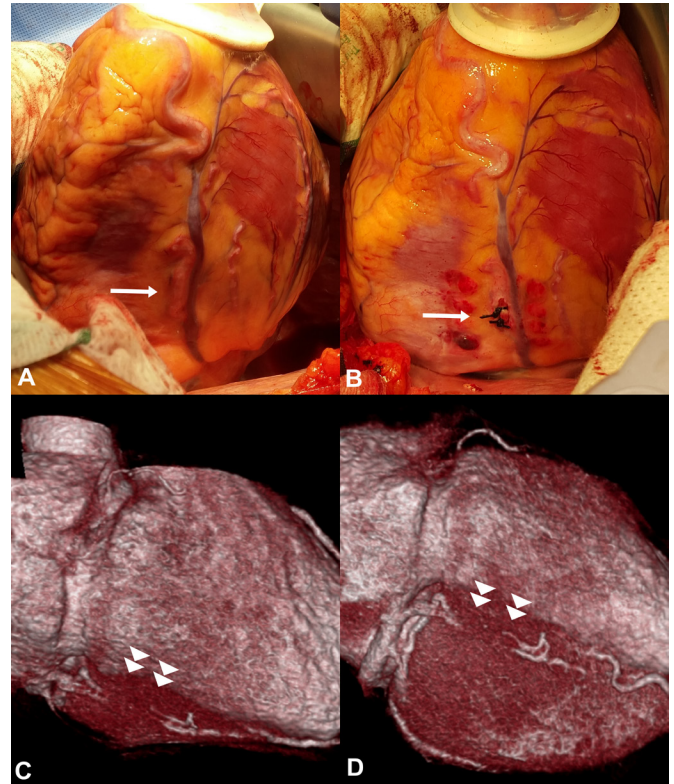


Figure 3. The intraoperative image shows the enlarged and tortuous PDA at the inferior wall of the LV (A, white arrow). PDA was dissected near the base of the LV and double ligation was done. (B, white arrow) Coronary computed tomography after double ligation surgery on postoperative day 6 shows successful ligation of the distal PDA close to the LV drainage site (white double arrowhead) and patent blood flow in the single coronary artery (C and D). LAD: left anterior descending artery, LV: left ventricle, PDA: posterior descending artery.

irreversible injury in the LV myocardium. We concluded that the patient developed DCMP, due to volume overloading and myocardial hibernation resulting from coronary flow steal. The heart team concluded that surgical correction might be a better treatment option than percutaneous closure of fistula for correction of this rare condition because of concerns about difficult access to the potential occlusion site, relevant risk of coronary artery perforation, and high risk of device embolization or thromboembolism of clots into the LV, in spite of high surgical risk due to severe LV systolic dysfunction. Therefore, we decided to perform surgical ligation of the PDA to reduce volume overloading and coronary blood flow steal. PDA was dissected near the PDA origin, and double ligation was performed at the site as close to the entry of the PL branch to the LV as possible (Figure 3).

Follow-up cardiac CT was conducted on the postoperative sixth day and revealed successful ligation of the distal PDA close to the drainage site to the LV, development of collateral vessels distal to the ligation site, and patent blood flow from the coronary ostia to the proximal part to the ligation site of the PDA and whole PL branch (Figure 3). Ten days after the surgery, the patient recovered almost from heart failure and

was discharged without any complications. Follow-up TTE after three months showed a nearly recovered left ventricular systolic function with EF 45–50%. The patient was doing well without CHF and any relevant symptoms two years after the surgery.

DISCUSSION

CAF is a rare congenital anomaly accounting for 0.4–0.5% of congenital heart disease cases [Mangukia 2012]. CAF forming a shunt between the coronary artery and the LV is observed in 2–3% of all patients with congenital CAF, while 90% of CAFs drain into the right side of heart, such as the right atrium, right ventricle, pulmonary artery, or coronary sinus, due to lower pressure [Fernandes 1992; Urrutia-S 1983; Luo 2006]. CAF drainage into the cardiac chambers is referred to as coronary cameral fistula and is very rare. DCMP presenting with a coronary cameral fistula draining into the LV also is extremely unusual.

Coronary “steal” and volume overloading of the cardiac chamber are two major pathogenic mechanisms of symptomatic CAFs. The coronary steal phenomenon in which CAF is related to shunt flow from the coronary artery runoff is presumed to lead to myocardial ischemia and hibernation, where the myocardium is supplied from the origin of the fistula [Oshiro 1990; Cheng 1982]. The internal pressure of the LV decreases during diastole, which can induce blood flow from the large coronary artery to the LV. Eventually, this flow can result in chronic LV volume overload similar to aortic regurgitation [Roberts 1986].

The optimal treatment for CAF remains a controversial issue. One reason is that many congenital CAFs are asymptomatic and incidentally detected on CAG, multi-detector computed tomography, and transthoracic echocardiography. In addition, Jebara et al and Cheung et al recommended regular screening until hemodynamic changes develop because CAFs tend to spontaneously close [Cheung 2001; Jebara 1991]. Invasive treatment of CAF is sometimes required for symptomatic patients with CAF and relevant hemodynamic deterioration.

Although the most common treatment for CAF is surgical ligation, percutaneous closure with coils or other closure devices increasingly are being performed. When percutaneous closure is selectively conducted in appropriate patients, good results can be expected. Moreover, percutaneous closure also is advantageous in that complications resulting from thoracotomy and extracorporeal circulation can be avoided [Kabbani 2008; Kassaian 2007]. However, if a CAF is large or difficult to access, technical difficulty can hinder complete percutaneous closure of the CAF. Although surgical ligation of a CAF has a high success rate, the incidence of complications, including myocardial infarction (MI), ranges from 3% to 7% [Carrel 1996; Mavroudis 1997]. Because the SCA (from LAD to the PL branch) in the present case had a very long and tortuous course and large diameter, there were several risks. These include expected risk of complications associated with percutaneous closure, risk of coronary

perforation or dissection due to difficult access to potential, risk of systemic thromboembolism due to incomplete closure, and risk of device embolization to the LV. Therefore, we concluded that the risk related to percutaneous closure outweighed the benefit from the procedure, and we decided to perform surgical ligation as close to the entry point of CAF to the LV as possible, despite high surgical risk due to severe LV systolic dysfunction. CAF has usually abundant potential collateral vessels. Therefore, when the ligation is made close to the entry site of the fistula to the LV, blood supply through the collateral vessels is possible, thereby minimizing myocardial damage. The surgical ligation was successful without any complications, including myocardial infarction.

CONCLUSION

We report that a young male presented with severe DCMP, due to a SCA with large CAF drained into the LV underwent successful surgical ligation. This patient has nearly recovered from DCMP.

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