Diffuse Coronary Artery Fistula Leading to Syncope and Treated with Transcatheter Coil Occlusion and a Defibrillator: A Case Report

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Significance of the Study

• In this report, a very rare case with extensive fistula drainage from three coronary arteries in the left ventricle is presented. Due to the development of left ventricular dysfunction and high sudden cardiac death risk, an implantable cardioverter defibrillator was implanted.

Keywords
Coronary artery fistula · Syncope · Transcatheter coil occlusion · Defibrillator

Abstract

Objectives: Coronary artery fistulas connecting coronary arteries to cardiac cavities are rare but clinically significant anomalies. Clinical Presentation and Intervention: A 47-year-old male patient presented with syncope. Left ventricular dysfunction was detected on echocardiography. Extensive coronary fistulas draining into the left ventricle were found on coronary angiography. Ventricular fibrillation was induced on electrophysiology study. Because of the induction of ventricular fibrillation, extensive fistulas, and presence of other risk factors, an implantable cardioverter defibrillator was implanted. After the detection of ischemia by nuclear scanning, microcoil occlusion of the fistula was performed. Conclusion: The present case describes extensive fistulas complicated with fatal ventricular arrhythmias due to ischemia and left ventricle dysfunction. A cardioverter defibrillator was implanted to prevent sudden cardiac death.

Introduction

Coronary artery fistulas (CAFs) are vascular connections originating from the epicardial coronary arteries and draining into the heart chambers, large vessels (vena cava, pulmonary veins, and pulmonary artery), or other vascular structures (mediastinal vessels and coronary sinus). In most cases, a solitary fistula emanates from the coronary artery and drains into the right ventricle, coronary sinus, pulmonary artery, or large veins [1–3]. In the literature, cases with fistulas draining from the coronary artery into the left ventricle have been reported as sporadic abnormalities [2]. Cases involving the presence of a fistula draining from the three coronary arteries into the left ventricle in the same patient are very rare [3].

Case Report

A 47-year-old male patient presented with a syncopal attack. Upon physical examination, his blood pressure was 130/85 mm Hg and he had a regular pulse of 72 beats per minute. His cardiac examination revealed no pathological findings, except for a short apical systolic murmur. Examination of the other systems was normal. His resting electrocardiogram showed a normal sinus rhythm,
left axis deviation, signs of left ventricular hypertrophy, and frequent unifocal premature ventricular complex. Transthoracic echocardiography revealed normal left ventricular diameters, moderate systolic dysfunction (ejection fraction, 38%), and signs of left ventricular hypertrophy at the interventricular septum. Medical treatment including metoprolol and ramipril was initiated. Coronary angiography was performed on the patient, who had non-sustained episodes of ventricular tachycardia detected on 24-hour rhythm Holter recording. No coronary atherosclerotic lesions were determined on coronary angiography. It was observed that the myocardium was intensively stained after selective injection of the left coronary artery. Fistulization into the left ventricle, especially from the left anterior descending (Fig. 1a), first diagonal (Fig. 1b), left circumflex (Fig. 1c), and the distal part of the obtuse marginal branches of the left coronary arteries, particularly from the small branches at the interventricular septum, was noted. Selective right coronary artery angiography showed that there was fistulization (Fig. 1d) involving the distal side branches of the right coronary artery with dominant flow toward the left ventricular cavity. Ventricular fibrillation was induced during the electrophysiology study of the patient due to the presence of clinical syncope, multiple fistulas, left ventricular systolic dysfunction, and a history of sudden death in his two brothers at an early age. Single chamber implantable cardioverter defibrillator (ICD) was implanted. Due to the detection of apical effort ischemia and fixed hypoperfusion on the lateral wall after myocardial perfusion scintigraphy on the lateral wall after myocardial perfusion scintigraphy performed 1 month later, transcatheter occlusion was planned. A guiding catheter was placed into the left coronary ostium. Microcoils were placed in the distal circumflex, the distal bifurcation of the left anterior descending artery, and the first diagonal artery via a microcatheter, which was transferred over the guide wire. A large number of fistulas with a diameter smaller than 0.5–1 mm were left without any intervention. No complications developed during or after the procedure. The ICD analysis interrogation at the 1-month follow-up determined that the device applied shock therapy twice due to ventricular fibrillation. Amiodarone therapy was started and control angiography was performed. During angiography, no flow was observed through the coronary fistulas where the coil occlusion was performed, but an increase in the flow from the septum towards the microfistulas (smaller than 0.5 mm) lying through the left ventricle was observed (Fig. 2a–d). On control scintigraphy of the myocardium, apical ischemia on exertion and fixed hypoperfusion on the lateral wall persisted, but the left ventricle dilatation during exercise decreased when compared to the previous myocardial perfusion scintigraphy.

**Discussion**

CAF, which provide a direct connection between the coronary arteries and heart chambers, are generally determined incidentally during coronary angiography and may cause ischemia by leading to coronary steal syndrome. CAFs are defined as direct connections between a coronary artery and a heart chamber, large vessels, or other vascular structures. Angiographic series revealed a prevalence of CAF of 0.09–0.5% [1–3]. Since fistulas between the coronary artery and the ventricles are generally asymptomatic, it is difficult to estimate the true incidence among the population. Almost 90% of CAFs flow into the
right heart chambers; thus, they act as arteriovenous fistulas. According to the draining cavities, the frequency of CAF flow is as follows: right ventricle, 41%; right atrium, 26%; pulmonary artery, 17%; and coronary sinus, 7%. There are fewer fistulas flowing into the left heart chambers: left atrium, 5%; and left ventricle, 3%. Although rare, fistulas flowing into both the ventricle and pulmonary veins have also been reported [4, 5]. Multiple CAFs are even more uncommon [3]. CAF generally originate from the right coronary artery system and usually flow into the right heart chambers. Among fistulas originating from a single coronary artery, fistulas flowing into the pulmonary artery have been reported in 17% of cases, whereas the rate of fistulas flowing into the pulmonary artery is 50% for the fistulas originating from bilateral coronary arteries [6]. Hemodynamically important fistulas may cause complaints, such as chest pain, dyspnea, and exhaustion. Symptoms of congestive heart failure mostly occur due to the volume load of the heart chambers, whereas chest pain occurs as a result of the coronary steal phenomenon or oxygen supply-demand imbalance. Pulmonary hypertension, congestive heart failure, bacterial endocarditis, rupture, coronary thrombosis, arterial aneurysm, and myocardial ischemia are potential complications [7]. Ischemic ST and T changes may occur during rest or exercise due to the coronary steal syndrome. Such ischemic changes are mostly observed if a steal syndrome has occurred on the largest branch of the left coronary artery. Lozano et al. [8] reported a patient in whom cardiac catheterization revealed multiple small fistulas from the left anterior descending coronary artery to the left ventricle, with normal systolic function, and who had an ICD implanted due to a documented ventricular fibrillation episode.

The intriguing feature of the present case is that the fistula caused left ventricular systolic dysfunction due to multiple connections, including septal microfistulas and arrhythmic syncope, as a clinical consequence. Such a condition has not been previously reported. Furthermore, the present case may have a genetic background with a potential sudden death risk, though it has not been proven through his family history.

Although complete agreement on the therapeutic approaches is lacking in fistula cases, it can be said that the asymptomatic small fistulas are benign and can be followed conservatively, whereas transcatheter or surgical treatment of the fistulas would be appropriate in symptomatic patients with fistula-related complications [1, 7]. Armsby et al. [9] performed a transcatheter closure procedure in 33 of 39 symptomatic patients having typical

**Fig. 2.** Angiographic views of the fistulas after intervention. 

- **a** Left lateral view of selective LAD injection. 
- **b** Implantation of coil via microcatheter to distal LAD (anteroposterior view, cranial angulation). 
- **c** Caudal anteroposterior view of selective Cx injection. 
- **d** Left anterior oblique view of selective LAD injection. 

LAD, left anterior descending artery; Cx, circumflex artery; RCA, right coronary artery.
murmurs and reported that all of the patients who had undergone interventional treatment showed asymptomatic and non-complicated progress at their 2.8-year follow-up. When the results of the study were compared with the present surgical case results, it was found that the transcatheter methods had the same efficacy, morbidity, and mortality as the surgical methods [10].

**Conclusion**

In the present case, as a treatment approach, we primarily performed ICD implantation because of the multiple fistulization that could not be removed completely due to the presence of systolic dysfunction, syncope, family history, induction of ventricular fibrillation on electrophysiology study, and angiographic reasons. The presence of an arrhythmia that had required shock therapy at the time of clinical follow-up showed that performing ICD implantation first was the right choice. Thereafter, due to the observed ischemia on scintigraphy, elective conservative coil occlusion was performed. After the treatment, although the major fistulas were closed, the microfistulas remained patent. There was no improvement in global left ventricle function; however, the reduction in the diameter of the left ventricle during exercise on scintigraphic imaging may be an indicator of partial efficacy. The patient is still being followed.

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