An Impressive Image of a Coronary–Cameral Fistula: A New Case Report

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ABSTRACT
A coronary artery fistula (CAF) is an abnormal connection between the heart chambers or one of the great veins. Most small fistulas have a good prognosis, while a prominent fistula or numerous small fistulas may lead to coronary steal. Herein we present the case of a patient who presented to our clinic with stable angina pectoris and who had CAFs originating from all major coronary arteries and draining into the left ventricle mimicked as left ventriculography.

Keywords: Coronary artery fistula, ventricular microfistula, coronary steal

INTRODUCTION
A coronary artery fistula (CAF) is an abnormal connection between the heart chambers (left or right ventricle) or one of the great veins (vena cava, pulmonary vein, or pulmonary artery) bypassing the myocardial distal capillary bed. Fistulas originating from the coronary arteries and draining to one of the heart chambers are also known as coronary–cameral fistulas, while those that end up in one of the veins are known as arteriovenous fistulas. The estimated prevalence of CAFs in the general population is very low (approximately 0.002-0.13%), and they are detected in 0.25% of patients undergoing coronary angiography (1-3). Most CAFs drain to the right heart chambers (90% into the right ventricle, right atrium, coronary sinus, and pulmonary trunk), while 52-60% originate from the right coronary artery (RCA), 30% from the left anterior descending artery (LAD), and 18% from the circumflex artery (Cx) (4, 5). However, a CAF originating from all three major coronary arteries at the same time and draining directly into the left ventricle is extremely rare and has an unidentified clinical importance. In this report, we present the case of a patient who presented to our clinic with stable angina pectoris and had a CAF originating from all three coronary arteries and draining into the left ventricle mimicked as ventriculography.

CASE REPORT
A 63-year-old male patient presented to our clinic with typical chest pain and dyspnea, which worsened with ongoing exercise for 2 months. He had no medical history except for hypertension that was diagnosed 10 years ago. His physical examination result was normal, except for tachycardia. His electrocardiogram showed 94 bpm with sinus rhythm. His cardiothoracic index and pulmonary vasculature were normal on the chest X-ray. An echocardiographic examination revealed an ejection fraction of 50% with modified Simpson’s method, while there was no valvular anomaly or ventricular hypertrophy. Marked ST depression was detected in his exercise stress test, and coronary angiography was planned. Coronary angiography revealed hemodynamically insignificant atherosclerotic plaques and several microfistulas originating from left and right coronary system circulation through the myocardium draining into the left ventricle and mimicking ventriculography with intense opacification of the left ventricle and atrium (Figure 1. a-d, Figure 2. a-d). Coronary angiography demonstrated that the microfistulas originated from the middle and distal portions of the LAD and Cx and the distal portion of the RCA. In addition, severe tortuosity and dilatations were present, particularly in the left coronary system (Video 1. See corresponding video/movie images at https://doi.org/10.5152/etd.2017.17054). These fistulas are thought to be the cause of the patient’s symptoms, and ST depressions in the exercise test while tachycardia is the precipitating factor. As these multiple microfistulas are not suitable for percutaneous or surgical closure, it was decided to administered medical treatment to the patient. The patient was discharged with acetylsalicylic acid, statins, and high-dose beta-blockers. At the follow-up visit after one month, the patient’s heart rate was 65/bpm and he was asymptomatic.
DISCUSSION

Most CAFs are typically asymptomatic during childhood, and most of them (18-63%) became symptomatic after 18 years of age (3, 6). While the most frequent symptom is exercise dyspnea, patients may also present with fatigue, atrial arrhythmias, pulmonary hypertension, heart failure, endocarditis, and even sudden cardiac death. On the other hand, angina generally occurs if there is a co-existing condition such as atherosclerotic heart disease, hypertrophic cardiomyopathy, or aortic stenosis (3, 7). The hemodynamic significance of coronary fistulas are determined by the flow rate across the fistula itself or the heart chambers they originate from and drain into. For example, clinical consequences of fistulas draining into the left ventricle are similar to those of aortic regurgitation (3, 8).

Most small fistulas have a good prognosis, while a prominent fistula or numerous small fistulas may lead to coronary steal. A prominent single fistula may cause infective endocarditis, superior vena cava syndrome, heart failure, stroke, or myocardial ischemia and should be treated with surgery or catheter-based procedures; however, similar to our case, there is no definitive treatment method for ischemic conditions caused by several microfistulas (9, 10).

In our patient who presented with new-onset angina pectoris and dyspnea, we detected several microfistulas originating from all three major coronary arteries, which led to severe opacification of the left ventricle during coronary angiography. These kind of CAFs are extremely rarely described in the literature. Coronary artery dilatations in our patient are thought to have occurred as a consequence of coronary steal, which was prominent in the left coronary artery. These dilatations were found to be permanent, even after the closure of CAFs, in the literature (9). As there were several microfistulas, our patient was not suitable for undergoing surgical or percutaneous closure. In the literature, there are similar cases treated with beta-blockers and nitrate therapy (6, 8). Fractional flow reserve measurement may be used to evaluate proximal and distal flow rates in these patients and as a guide to determine whether if shunts are hemodynamically significant.

CONCLUSION

We need more detailed experiences for the management of symptomatic or co-incidentally detected coronary-cameral microfistulas after performing coronary angiography.

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