CASE REPORT

Diffuse Plexus-Like Coronary Artery-Left Ventricular Fistulae Leading to Coronary Steal Syndrome: A Pattern of Anomalous Coronary Microvascularization

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Abstract

Coronary artery fistulae that drain into the left ventricle are extremely rare, and even fewer cases of fistulae involving all three of the coronary arteries have so far been reported. We herein report a 64-year-old woman with a unique pattern of coronary artery-left ventricular fistulae that involved all three of the coronary arteries. The multiple fistulae presented in a diffuse plexus-like arrangement. The fistulae resulted in a diastolic volume overload of the left ventricle (left-to-left shunt), as well as “coronary steal” with the shunting of blood away from the myocardium since the fistulae represented the path of least resistance.

Key words: coronary steal, coronary artery anomaly, fistula


Introduction

The presence of a coronary artery fistula between a coronary artery and a cardiac chamber is a rare condition, occurring in only 0.2% of patients who have undergone cardiac catheterization (1, 2). The majority of these cases result in a left-to-right shunt, where the fistulae drain into a right cardiac structure. Fistulae that drain into the left ventricle are extremely rare, and even fewer cases of fistulae involving all three of the coronary arteries have been reported. We herein report the findings of a 64-year-old woman with a unique pattern of coronary artery-left ventricular fistulae that involved all three of the coronary arteries. The multiple fistulae presented in a diffuse plexus-like arrangement.

Case Report

A 64-year-old woman who was regularly receiving hemodialysis for the treatment of chronic renal failure was admitted to our hospital with progressive exertional dyspnea and paroxysmal nocturnal dyspnea. Upon admission, her pulse rate was 65 bpm and regular, and her blood pressure was 120/75 mmHg. Her cardiac enzymes were normal and a chest X-ray revealed cardiomegaly with an increased pulmonary vasculature. A twelve-lead electrocardiogram (ECG) revealed a normal sinus rhythm and non-specific ST-T changes (Fig. 1A). Transthoracic echocardiography demonstrated a mild left ventricular dilation, a preserved left ventricular systolic function with an ejection fraction of 65% and moderate pulmonary hypertension. After admission, she presented with typical angina and dyspnea during hemodialysis. An ECG taken during angina showed a normal sinus rhythm with biphasic and inverted T waves in leads V2-V5 and T wave inversion in leads II, III and aVF (Fig. 1B), thus indicating a possible multi-vessel myocardial ischemia. Cardiac catheterization with coronary angiography was performed and disclosed patent epicardial coronary arteries with all three coronary arteries draining into the left ventricle via a diffuse plexus-like pattern of coronary fistulae (Fig. 2, Online Video 1 and 2). Coronary angiographies revealed diastolic opacification (Fig. 2A, C) and systolic emptying (Fig. 2B, D) of the left ventricle. The coronary sinus was normal in the levophase of angiography. Coronary angi-
Figure 1. (A) The twelve-lead ECG performed upon admission shows a sinus rhythm and nonspecific ST-T changes. (B) The ECG taken during hemodialysis and angina shows a sinus rhythm with biphasic and inverted T-waves in leads V2-V5, as well as T wave inversion in leads II, III and aVF.

The presence of coronary artery fistulae between a coronary artery and a cardiac chamber is noted in only 0.2% of the patients who undergo diagnostic cardiac catheterizations (1, 2). In more than 90% of these cases, the fistulae arise from the right coronary artery and drain into a right heart structure (right ventricle, right atrium or pulmonary artery), thus resulting in a left-to-right shunt. Fistulae draining into the left ventricle are extremely rare, and even fewer cases of fistulae involving all three of the coronary arteries have been reported. Our patient shows a unique pattern of multiple coronary artery-left ventricular fistulae involving all three of the coronary arteries and presenting in diffuse plexus-like pattern. These congenital fistulae are different from Thebesian vessels (also called venae cordis minima or smallest cardiac veins) that represent a persistence of the intertrabecular vascular network (2, 3). Rather, these fistulae can result in a diastolic volume overload of the left ventricle (left-to-left shunt), dilated epicardial arteries and a “coronary steal” with the shunting of blood away from the myocardium since the fistulae represented the path of least resistance for the blood flow. The clinical diagnosis of coronary artery-left ventricular fistulae is difficult because the clinical presentation and the laboratory and ECG manifestations are nonspecific. As with our patient, the presence of the abnormal structures results in typical angina or exertional dyspnea associated with patent coronary arteries (4). The main mechanism of myocardial ischemia might be related to the coronary steal phenomenon (5). The definitive treatment for coronary artery fistulae is surgery or transcatheter embolization. However, surgical treatment or embolization is not suitable for the presence of multiple coronary artery-left ventricular fistulae. The administration of beta blockers or non-dihydropyridine calcium channel blockers (e.g., diltiazem or verapamil) could improve the symptom of angina (4), possibly by improving the myocardial oxygen supply-demand mismatch. In our patient, the symptoms (angina and dyspnea during dialysis) were relatively relieved through treatment with diltiazem and no surgical treatment was warranted.

The authors state that they have no Conflict of Interest (COI).
Figure 2. Coronary angiogram of the left (A and B) and right (C and D) coronary arteries in the right anterior oblique view shows normal epicardial vessels, and the sequence demonstrates multiple fine fistulae draining to the cavity of the left ventricle. As a result, a striking pattern of filling and emptying of this chamber is visualized during diastole (A and C) and systole (B and D), respectively (arrow heads indicate the left ventricular cavity). The fistulae emerged from the proximal, mid, and distal segments of the left anterior descending and left circumflex arteries (A).

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References