

Left ventricular steal syndrome caused by multiple plexiform coronary artery fistulae: case report, literature review and treatment

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Anomalous coronary arteries are rarely seen in 0.3% to 1% of the general population [1]. Coronary artery anomalies may be classified as anomalies of origin, course and termination [2]. Hemodynamically significant anomalies may cause myocardial perfusion abnormalities, symptoms of myocardial ischemia or sudden death [3]. Coronary artery fistulae (CAF) consist of termination abnormalities of coronary arteries, in which there exists a communication between one coronary artery and other coronary arteries, pulmonary artery, coronary sinus, superior vena cava or a cardiac chamber [3]. Coronary artery fistulae are seen in approximately 0.1% to 0.2% of patients undergoing coronary angiography and more commonly involve the right coronary system than the left coronary system (60%, 40%, respectively) [4]. Coronary artery fistulae originating from both systems represent less than 5% of cases [3]. Involved coronary arteries are often dilated and tortuous [3]. In terms of morphology and drainage site, CAF are variable, with either single or multiple communications or a maze of fine vessels or plexus with intramural distributions [3]. The drainage site is of higher clinical importance as per origin. The most commonly seen drainage sites are the right ventricle (45%), right atrium (25%), pulmonary artery (15%), left ventricle or left atrium (10%) [3].

Here, we present a case of multiple CAF causing myocardial ischemia and symptoms of angina pectoris and would like to discuss the treatment approaches of this type of CAF.

A 62-year-old woman presented with symptoms of resting chest pain lasting for 3 to 5 min, which had started in the last 2 weeks. In the patient's medical history, she had experienced hypertension for 2 years and occasionally mild exertional chest pain. She had no history of known coronary artery disease, diabetes mellitus, hyperlipidemia or smoking. She was taking amlodipine 5 mg once daily for the treatment of hypertension. Her physical examination revealed 2/6 degree mild diastolic murmur at the apex. Electrocardiography showed normal sinus rhythm and no ST segment abnormality. Two-dimensional echocardiography showed no hypertrophy or other abnormality. The levels of serum creatinine kinase MB fraction and troponin T were found to be normal. The patient was diagnosed with low risk unstable angina. Exercise stress test showed 2 mm horizontal ST segment depressions at peak exercise in derivations of V3-6, DI-III, aVL and aVF. The patient, therefore, underwent cardiac catheterization. Selective coronary angiography showed no

atherosclerotic lesions but revealed flow of contrast agent from the distal endings of all three coronary arteries to the left ventricle via plexuses of intramural vessels. The left ventricle (LV) stained and LV walls and borders were seen throughout 3 cardiac cycles (Figure 1). We concluded that the pathophysiological mechanisms of myocardial ischemia in this case were reduction of blood flow to the myocardium resulting from the left ventricular steal phenomenon and the vasodilating effect of the dihydropyridine calcium channel blocker on the plexus of the CAF, which increases the shunt fraction. Therefore, amlodipine therapy was switched to bisoprolol 10 mg once daily. In the follow-up period, ivabradine was added to the therapy for effective heart rate reduction (< 70 per min). In 2 years of follow-up, the patient has had no symptoms of angina pectoris and no cardiovascular event to date.

This case presentation concerns an elderly woman with multiple plexiform CAF between all three major coronary arteries and the left ventricle causing anginal symptoms. The patient's symptoms were most likely the result of left ventricular steal of oxygen-rich blood via fistulous channels bypassing the myocardium. CA-LV fistulae originating from three major coronary arteries have been encountered rarely in the catheterization laboratory. Heper *et al.* reported an incidence of 1 per 14000 coronary angiographies, but others identified 8 cases in 7262 coronary angiographies [5].

It has been reported that CAF are generally asymptomatic in the first two decades of life, in particular when they are hemodynamically insignificant. In fact, a few may close spontaneously. Coronary artery fistulae may increase in size over time and form complex, long, tortuous and aneurysmal connections, and the largest shunts are

generally seen when there is a right-sided shunt rather than a left-sided one [6]. After this period, the frequency of symptoms and complications increases [6]. Complications of CAF consist of steal phenomenon from the adjacent myocardium causing myocardial ischemia, thrombosis and embolism, cardiac failure, rupture, endocarditis, endarteritis, atrial fibrillation and other arrhythmias [6].

In older patients, symptoms related to CAF increase and commonly include angina or dyspnea on exertion and arrhythmias. When there is a shunt to a right-sided heart chamber, the hemodynamics are similar to an extra-cardiac left-to-right shunt, but when there is a connection to a left-sided heart chamber, the hemodynamics resemble those of aortic insufficiency [3]. Patients who have CAF between coronary arteries and the left ventricle (CA-LV fistulae) may have mildly elevated pulmonary capillary wedge pressure and passive pulmonary hypertension, which is likely due to LV diastolic dysfunction associated with decreased LV compliance from increased diastolic blood flow via the multiple CAF.

Diastolic perfusion of the myocardium supplied by the coronary artery with CA-LV fistulae may be reduced, which results from a hemodynamic steal phenomenon and may lead to myocardial ischemia [3]. These pathways may become hemodynamically significant only if a pressure gradient exists between two sides of the shunt [7]. In diastole, such a pressure gradient develops between coronary arteries and the left ventricular cavity. Since the majority of coronary flow and myocardial perfusion occurs in diastole, these shunts may cause left ventricular steal syndrome and myocardial ischemia. Vasodilating agents such as nitrates and dihydropyridine group calcium channel blockers may increase the shunt volume

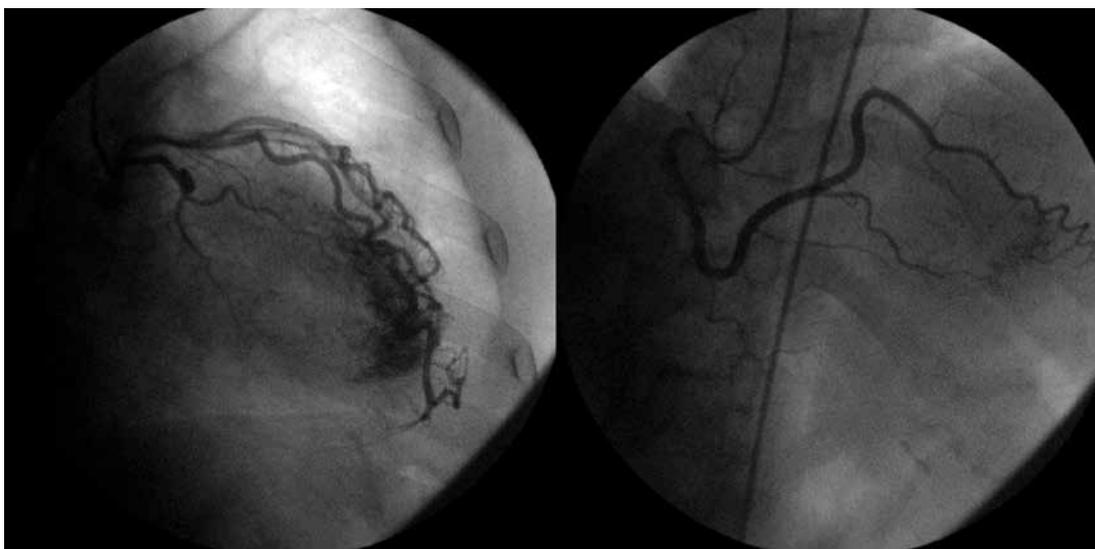


Figure 1. Multiple small cameral fistulae between coronary arteries and left ventricle

between coronary arteries and the left ventricle, and increased left ventricular steal may worsen myocardial perfusion and myocardial ischemia [5]. β -Blockers and ivabradine prolong diastole time, which determines the duration of coronary flow and myocardial perfusion time. Thus, increased duration of diastolic coronary flow and myocardial perfusion time may increase myocardial perfusion and decrease myocardial ischemia [8, 9]. According to our knowledge, this is the first report on ivabradine use in the medical treatment of coronary cameral fistulae and has the longest follow-up period.

The treatment of CA-LV cameral fistulae is essentially medical; conservative management with continued follow-up of these patients appears to be appropriate [10]. Surgical treatment is exceptional and should be considered only in severe cases refractory to medical treatment [10].

In conclusion, because the coronary artery to LV communications are small and multiple, surgical or transcatheter occlusion therapies may not be appropriate in such cases. In the light of the worsening of angina under nitrate and dihydropyridine group calcium channel blocker therapy, reducing heart rate with a β -blocker and ivabradine in patients with sinus rhythm may be effective for angina control in the treatment of CA-LV cameral fistulae.

Conflict of interest

The authors declare no conflict of interest.

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