

Multiple Coronary Artery-Left Ventricular Microfistulae in a Patient with Apical Hypertrophic Cardiomyopathy: A Demonstration by Transthoracic Color Doppler Echocardiography

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Among the congenital coronary artery fistulae, multiple coronary artery microfistulae arising from the left and right coronary artery and emptying into the left ventricle are very rare and little is known of their anatomic and clinical features, especially in apical hypertrophic cardiomyopathy. A 67-year-old woman was referred for the evaluation of chest pain at exertion, and shortness of breath. Electrocardiographic and echocardiographic findings were typical of apical hypertrophic cardiomyopathy. Coronary arteriography showed normal epicardial coronary arteries, but multiple coronary artery-left ventricular microfistulae arising from the left and right coronary arteries. Transthoracic color Doppler echocardiography, using a high frequency transducer with a low Nyquist limit, demonstrated multiple coronary artery-left ventricular microfistulae just beneath the apical impulse window.

Key Words: Apical hypertrophic cardiomyopathy, transthoracic Doppler echocardiography, coronary artery-left ventricular microfistulae

INTRODUCTION

A number of abnormal forms of congenital coronary arterial fistulae are known. Most of the fistulae arise from either the right or left coronary artery and drain into the right side of the heart.^{1,2}

Multiple coronary artery to left ventricular microfistulae is rare and the clinical and hemodynamic sequelae are still incompletely understood.^{3,4} In particular, little is known about the association between multiple coronary artery-left ventricular microfistulae and apical hypertrophy.⁵ Moreover, clinical symptoms related to multiple coronary artery-left ventricular microfistulae are highly variable. Most patients are asymptomatic, although some may present in infancy with congestive failure, or later in life with myocardial ischemia, due to a coronary steal phenomenon.⁶⁻⁸ Several cases of acute coronary syndrome have been ascribed to the presence of multiple coronary artery-left ventricular microfistulae in patients with apical hypertrophic cardiomyopathy.⁹ We report a case of multiple coronary artery to left ventricular microfistulae, presenting as chest pain and a shortness of breath in a patient with apical hypertrophic cardiomyopathy, which was diagnosed by coronary angiography and demonstrated by transthoracic color Doppler echocardiography.

CASE REPORT

A 67-year-old woman was referred to our hospital for the evaluation of chest pain and a shortness of breath that had occurred 3 days previously. She had a 2-year history of recurrent episodes of anterior chest pain, which were typi-

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cally provoked by physical exertion or emotional distress and readily relieved by sublingual nitroglycerin. She also had a 1-month history of hypertension. On physical examination, fine crackles were audible in both lower lung fields, and she had a high blood pressure (180/100 mmHg). No heart murmurs were audible; there was no ankle edema; and chest radiography showed mild cardiomegaly and pulmonary edema in both lung fields. A resting electrocardiogram (ECG) showed left ventricular hypertrophy with inverted T wave in the precordial leads (Fig. 1). The CK-MB fraction was slightly increased (12.03 ng/ml) but the cardiac troponin-T level was within the normal range. The pO_2 was 49.2 mmHg and pCO_2 was 53.5 mmHg. O_2 saturation was 82% at the initial arterial blood gas analysis (room air). Two dimensional transthoracic echocardiography showed typical of apical hypertrophic cardiomyopathy without regional wall motion abnormality and with normal left ventricular systolic function. The patient was treated with a nasal O_2 supply, aspirin, isosorbide dinitrate, beta-blocker, angiotensin converting enzyme(ACE) inhibitor, and diuretics at the intensive care unit. After an improvement in the patient's symptoms, she underwent coronary angiography, which showed no stenotic lesion of epicardial coronary arteries. However, left coronary angiography showed multiple microfistulae originating from the 1st diagonal branch of the left coronary artery and draining into the left ventricular cavity (Fig. 2). Right coronary angiography showed opacification of the left ventricle through a diffuse microfistulae network originating from the right posterior descending artery

(Fig. 3). Left ventricular angiography revealed a spade shaped left ventricle, typical of apical hypertrophic cardiomyopathy without regional wall motion abnormality (Fig. 4). Transthoracic color Doppler echocardiography just beneath the apical window, using a high frequency transducer (4-7 MHz, ATL HDI 5000, USA) with a low Nyquist limit (12 to 16 cm/s), showed the presence of multiple linear color flow signals perpendicular to the epicardial surface, draining into the left ventricular cavity, the demonstrating multiple coronary artery-left ventricular microfistulae arising from the left coronary artery (Fig. 5). She was discharged a week later with clinical improvement

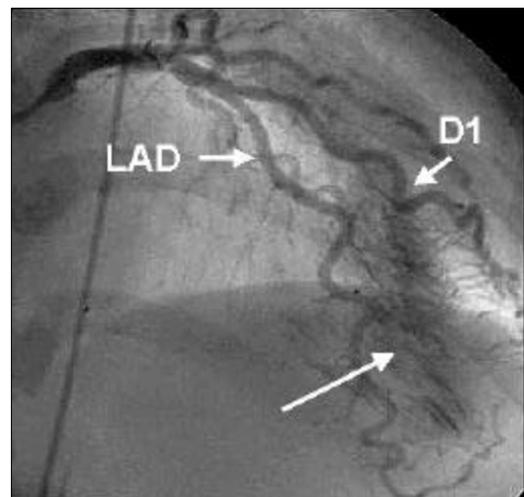


Fig. 2. Left coronary angiogram showing multiple microfistulae originating from the 1st diagonal branch (D1) of the left coronary artery and draining into the left ventricular cavity (arrow).

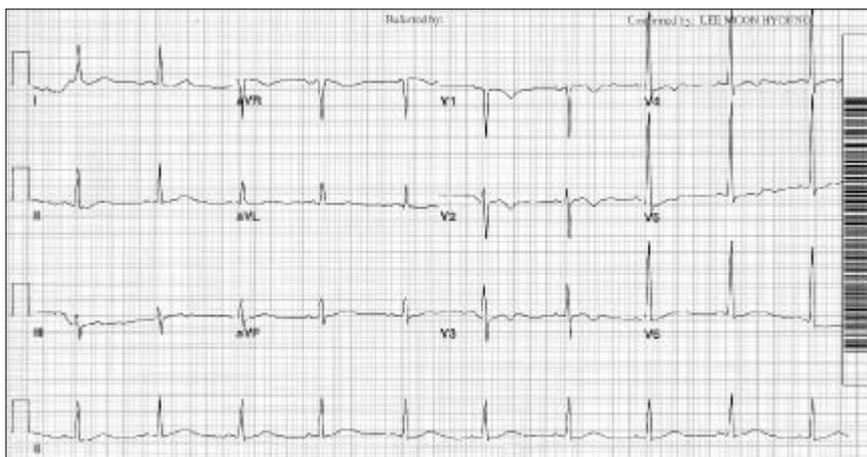


Fig. 1. A resting electrocardiogram showing left ventricular hypertrophy with an inverted T wave in the precordial leads.

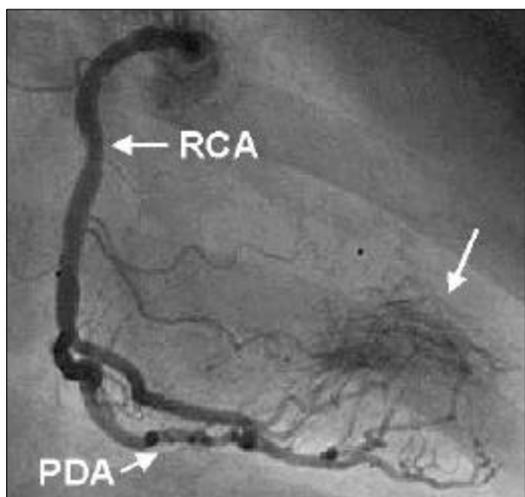


Fig. 3. Right coronary angiogram showing opacification of the left ventricle through a diffuse microfistulae network originating from the right posterior descending artery (arrow). PDA; posterior descending artery.



Fig. 4. Left ventricular angiogram showing the spade-like shape of left ventricle, which is representative of apical hypertrophic cardiomyopathy.

after medication; oral isosorbide dinitrate, beta-blocker, aspirin, and angiotensin converting enzyme inhibitor.

DISCUSSION

Congenital coronary artery fistulae between a coronary artery and a cardiac chamber can occur

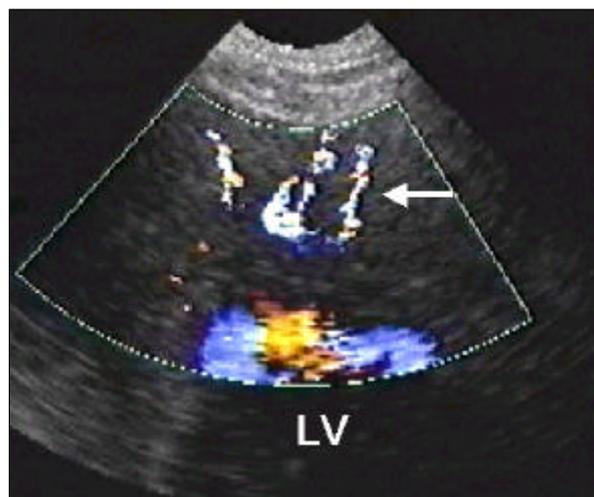


Fig. 5. Transthoracic color Doppler echocardiogram showing multiple linear color flow signals (arrow) perpendicular to the epicardial surface in the left ventricular apex, which drained into the left ventricle, thus demonstrating multiple left coronary artery-left ventricular microfistulae.

in 0.2% of diagnostic cardiac catheterizations and usually arise from the right coronary artery to drain into a right heart structure (right atrium, right ventricle or the pulmonary artery). Fistulae draining into the left ventricle are uncommon,⁶ and the pathophysiologic origin of the malformation is obscure. Morphological studies suggest the partial persistence of embryonic myocardial sinusoids that arise from endothelial protrusions into the intertrabecular spaces. Fetal regression of these structures results in the formation of the Thebesian vessels of the adult heart. Thus, interference with developmental changes might produce an abnormally prominent Thebesian system with the morphological appearance of multiple coronary microfistulae.^{7,8} Few reports exist upon coronary artery to left ventricular fistulae in apical hypertrophic cardiomyopathy, and it is not clear whether the apical hypertrophy is a reactive change to chronic volume overload of the left ventricle through the coronary artery to the left ventricular shunt or whether itself results in multiple coronary microfistulae, possible due to the disarray of myocardial cells. This association may not be coincidental as multiple microfistulae could cause abnormalities of the microcirculation and result in reactional myocardial hypertrophy.⁵

Most patients in whom this anomaly is diagnosed present with typical or atypical angina pectoris in adult life. The clinical symptoms have been attributed to a coronary steal phenomenon due to the shunting of blood via the low resistance fistulae. According to the literature, all patients experience their first anginal attack during advanced adulthood (older than 40 years) despite the assumed congenital origin of the malformation.¹⁰ Symptoms of congestive heart failure are rarely reported in this entity, and when observed, seem to be related to concomitant atrial fibrillation or severe apical hypertrophic cardiomyopathy.^{10,11} While diastolic volume overload of the left ventricle, giving the clinical picture of aortic incompetence, was found to be predominant in single coronary artery to left ventricular fistula, myocardial ischemia and the absence of a continuous murmur were found more prevalent in multiple coronary artery to left ventricular fistulae.¹² In the present case, the patient had acute pulmonary edema on chest X-ray, probably due to diastolic dysfunction and myocardial ischemia by apical hypertrophic cardiomyopathy and multiple coronary artery to left ventricular fistulae.

The clinical diagnosis of coronary artery left ventricular fistula is difficult because the clinical presentation, laboratory, and ECG manifestations are nonspecific. In most of cases, coronary artery to left ventricular fistulae is identified during coronary angiography, performed to evaluate patients with suspected coronary artery stenosis. A variety of diagnostic approaches have been proposed in order to document the presence of myocardial ischemia and the hemodynamic consequences of fistulization.¹³ To demonstrate myocardial ischemia, exercise electrocardiography and scintigraphy have been utilized to provoke ischemic changes in this condition.¹⁴⁻¹⁶ In this case, we visualized multiple coronary artery to left ventricular microfistulae in the apical window by transthoracic color Doppler echocardiography using a high frequency transducer with a low Nyquist limit,¹⁷ which showed the presence of multiple linear color flow signals, perpendicular to the epicardial surface draining into the left ventricular cavity.

The management of patients with coronary artery fistulae remains controversial. Clear indica-

tions for surgical intervention include, the presence of large shunts, other major cardiac lesions, concomitant atherosclerotic coronary artery disease, and symptoms, or complications including, progressive enlargement, bacterial endocarditis, rupture, pulmonary hypertension, and thromboembolism.¹⁴ Medical therapy with nitrates in conjunction with beta-blockers or calcium-antagonists have proven to be efficient in a few cases.

The natural history of coronary artery-left ventricular fistulae remains largely unknown. The majority of adults remain asymptomatic, without a tendency to progressive enlargement of the fistulae during periods of follow-up, which have extended up to 10 years.⁶ It remains uncertain whether there is an association with premature atherosclerosis. Moreover, large left-to-right shunts may promote the development of mild-to-moderate pulmonary hypertension,⁶ and bacterial endocarditis has been reported to be a complication independent of fistula size.¹⁸

We report a case of multiple coronary artery-left ventricular microfistulae in a patient with apical hypertrophic cardiomyopathy, which was demonstrated by transthoracic color Doppler echocardiography. This case is worthy of attention due to its rarity and the possible role of transthoracic color Doppler echocardiography using a high frequency transducer with a low Nyquist limit for demonstrating multiple coronary artery-left ventricular microfistulae in patients with apical hypertrophic cardiomyopathy.

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