Coronary artery-left ventricular microfistulae associated with apical hypertrophic cardiomyopathy

Özgül Uçar, Hülya Çiçekçioglu, Mustafa Çetin, Mehmet Ileri, Sinan Aydoğan

Department of Cardiology, Ankara Numune Education and Research Hospital, Talatpaşa Bulvari, Sihhiye, Ankara, Turkey

Abstract
A 58 year-old Caucasian man was admitted to the coronary care unit with angina pectoris. There were deep inverted T waves and ST segment depression at anterior precordial derivations. Coronary angiography revealed widespread coronary artery to left ventricular microfistulae arising from distal portions of both left and right coronary systems. Left ventriculography and transthoracic echocardiography revealed typical features of apical hypertrophic cardiomyopathy. Angina pectoris was alleviated by beta-blocker therapy. Both multiple coronary artery to left ventricular microfistulae and apical hypertrophic cardiomyopathy are rare conditions and little is known about pathophysiological and clinical aspects of this combination. Accumulating evidence will provide us this information so that the management of the patients will be enhanced. (Cardiol J 2011; 18, 3: 307–309)

Key words: coronary fistula, cardiomyopathy, angina, coronary angiography

Introduction
Congenital coronary artery fistulae are rare (with a prevalence of 0.13–0.2%) in patients undergoing coronary angiography [1, 2]. Multiple microfistulae between coronary arteries and left ventricle are even rarer. Clinical presentation depends on the site of origin, size, multiplicity and the site of drainage of the coronary fistula. Angina pectoris, heart failure, cardiac arrhythmia, syncope, myocardial infarction and endocarditis have all been reported in relation to coronary fistulae. We report a case of multiple coronary artery to left ventricle microfistulae in association with apical hypertrophic cardiomyopathy (AHC) in a patient who presented with angina.

Case report
A 58 year-old Caucasian man presented to the emergency service with angina pectoris and was admitted to the coronary care unit with suspected acute coronary syndrome. There were deep negative T waves and ST segment depression through the leads V2 to V6 on 12-lead electrocardiogram (ECG). Cardiac auscultation findings were normal. He had no coronary risk factors other than age and male gender. During his follow-up, neither cardiac enzyme nor troponin levels were elevated. A coronary angiogram revealed dilated and tortuous epicardial coronary arteries with no atherosclerotic lesions. However, a heavy stream of contrast agent entered the left ventricle via a plexus of intramural

Address for correspondence: Özgül Uçar, MD, Keklikpinari Mah, 463/7, Posta Kodu: 06450, Dikmen, Ankara, Turkey, tel: +905053491009, +903125084783, +903124769578, fax: +903124324356, e-mail: ozgul_ucar@yahoo.com
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vessels, from both the distal portions of the left and right coronary artery systems (Figs. 1A, B). On left ventriculography there was apical obliteration at end-systole and spade-like configuration at end-diastole. Two-dimensional transthoracic echocardiography showed typical features of AHC without regional wall motion abnormalities (Fig. 2). No intracavitary systolic gradient was detected. Due to the diffuse character of the microfistulae between the coronary arteries and the left ventricle, neither surgery nor transcatheter occlusion was feasible. We started 100 mg/day metoprolol and the patient experienced no angina pectoris under this therapy.

**Discussion**

Multiple coronary artery-left ventricular microfistulae originating from both left and right coronary systems are rare. This condition is believed to result from persistence of the embryological myocardial microcirculation [3]. It causes left-to-left shunting similar to aortic regurgitation, and most patients with this anomaly present with angina pectoris during advanced adulthood. Ischemia in the presence of multiple coronary artery microfistulae has been attributed to ‘coronary steal phenomenon’ which is shunting of blood via the low resistant fistulae [4]. AHC is a relatively rare morphological expression of hypertrophic cardiomyopathy (< 5% of patients) which was initially described in Japan.

The disease has two forms: in the Japanese form, hypertrophy is located at the apex and in the non-Japanese form hypertrophy extends under the level of chordae tendinea. The characteristic ECG finding is giant negative T waves at anterior precordial derivations. The reports about the coexistence of multiple coronary artery to left ventricle microfistulae with AHC are increasing [5–10]. There are numerous mechanisms postulated about this association. Microfistulae can cause reactive left ven-

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**Figure 1.** Left (A) and right (B) coronary angiography demonstrates multiple coronary artery — left ventricle microfistulae.

**Figure 2.** Two-dimensional transthoracic echocardiography shows left ventricular apical thickening in apical-four-chamber view; LV — left ventricle; LA — left atrium; RV — right ventricle; RA — right atrium.
tricular hypertrophy through induction of ischemia by means of ‘coronary steal phenomenon’ or through chronic volume overload mimicking aortic regurgitation.

Another suggestion is that myocardial disarray in AHC may induce a developmental anomaly in the Thesbian venous system; or there can be a common etiologic factor, such as a genetic mutation resulting in both anomalies. Both coronary artery fistulae and AHC can cause angina pectoris and their association may aggravate the clinical condition. Increased oxygen demand due to myocardial hypertrophy, together with decreased coronary vasodilatory capacity, are classical mechanisms of angina in the latter.

Due to the rarity of this condition, the natural course of multiple coronary artery to left ventricle microfistulae is not known. Banks et al. [11] described a ten year case history who initially presented with ischemic symptoms and later developed left heart failure with elevated left ventricular end-diastolic pressure. Ischemia in the context of multiple coronary artery to left ventricular microfistulae and AHC is usually benign. No cases of myocardial infarction have been reported. In previous reports about multiple coronary microfistulae, exercise stress test or thallium scintigraphy was performed in order to demonstrate objective evidence of ischemia. However, the utility of these tests is limited when multiple coronary microfistulae coexist with AHC. ECG changes related to AHC obscure ST segment analysis in exercise stress test or AHC itself causes reversible perfusion defects on thallium scans, even when the coronary arteries are normal [12]. Currently, symptomatic treatment with beta-blockers or non-dihydropyridine calcium channel blockers appears the optimal treatment strategy. This therapy proved successful in nearly all of the reported cases, as well as in our case.

In conclusion, our case report contributes to the growing evidence about the association of coronary artery to left ventricular microfistulae and AHC. Understanding the pathophysiological implications of both clinical conditions will enhance the management of patients.

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References