BILATERAL CORONARY ARTERY-LEFT VENTRICLE FISTULAS PRESENTING WITH STABLE ANGINA PECTORIS RESISTANT TO PHARMACOLOGICAL THERAPY: A Case Report

Erhan BABALIK[a] MD, Murat MERT[b] MD, Alev Arat-ÖZKAN[a] MD, Tevfik GÜRMEN[a] MD

[a]Departments of Cardiology, and [b]Cardiovascular Surgery,
Institute of Cardiology, Istanbul University, Istanbul, Turkey

Summary

Bilateral coronary artery fistulas, which drain into the systemic high-pressure arterial system, are rare anomalies. This article will report an unusual case of multiple, bilateral coronary artery to left ventricle fistulas causing myocardial ischaemia and angina pectoris that did not respond well to pharmacological treatment. ***(Arch Turk Soc Cardiol 2003;31:294-7)***

Key words: *Angina pectoris, coronary artery, anomaly coronary artery fistula*

Özet

İlaç Tedavisine Dirençli Kararlı Angina Pektoris ile Beliren Bilateral Korner Arter-Sol Ventrikül Fistülleri: Bir Olgu Sunumu

Özellikle yüksek basınçlı arteryel sisteme açılan bilateral koroner arter fistülleri nadir görülen anorlalilerdir. Sempomlu hastalarda, fistülün anatomik yapısına ve eşlik eden bir kalp hastalığı varlığına göre cerrahi veya perkütan girişim yöntemleri genellikle kabul görmektedir. Bu yazıda farmakolojik tedaviye iyi yanıt vermeyen miyokard iskemisi ve anginaya neden olan, sol ventrikül ile her iki koroner arasındaki multipl fistülleri olan nadir bir olguyu sunuyoruz. ***(Türk Kardiyol Dern Arş 2003;31:294-7)***

Anahtar kelimeler: *Angina pektoris, koroner arter anomalisi, koroner arter fistülleri*

Coronary artery fistulas (CAFs) represent one of the most common congenital anomalies of the coronary arteries[1]. A coronary artery with an anatomically normal origin terminates abnormally and communicates with a cardiac chamber or a great vessel. The clinical importance of such a communication depends upon the amount of blood traversing the communication, the cardiac chamber or the great vessel into which the fistula drains. Although many patients with CAFs are asymptomatic, the clinical presentation of symptomatic cases includes angina pectoris, atypical chest pain, palpitation, fatigue, bacterial endocarditis, congestive heart failure, pericardial effusion, or even sudden cardiac death[2-4]. Less than 10% of the CAFs drain into the systemic high-pressure arterial system[1]. Bilateral CAFs are rare anomalies and multiple ones are even rarer[5-7]. This article will

Address for Correspondence: Dr. Erhan Babalık. Ethemefendi cad. Sadi Yaver Ataman sok. 11/17 Erenköy, 34738 Istanbul / TURKEY
Phone: 0216 302 46 88 / Fax: 0212 511 66 46
E-mail:erhanbabalik@yahoo.com
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report an unusual case of multiple, bilateral coronary artery to left ventricle fistulas causing myocardial ischaemia and angina pectoris in a middle-aged male.

REPORT of CASE

Eight months before his current admission, a 49-year-old male was performed a coronary angiography in our institution because he had angina at effort with a positive exercise test. At first admission, blood pressure was 120/70 mmHg and pulse rate was 76 bpm otherwise the physical examination was unremarkable. Coronary angiography demonstrated multiple, bilateral coronary artery to left ventricle fistulas without atherosclerotic lesions (Fig. 1 and Fig. 2). He was prescribed 50 mg of metoprolol twice daily and 5 mg of amlodipine once daily to relieve symptoms. In fact, there was not any alternative therapy (surgical ligation, interventional procedures) to pharmacological treatment for this patient because fistulas were composed of a plexiform network of vessels. Eight months after drug initiation, the patient was admitted again to the emergency service of our institution with complaints of half an hour-lasting anginal pain and dizziness after exertion. He had angina pectoris (CCS class I-II) and fatigue on exertion, even milder, despite using drugs regularly. He had a history of 365-pack/year-cigarette smoking. On admission his arterial blood pressure was 100/60 mmHg and he had a pulse rate of 60 bpm. There was no audible murmur on the chest wall, and the rest of the physical examination was normal. Laboratory tests revealed: total cholesterol 199 mg/dL, HDL-cholesterol 45 mg/dL, LDL-cholesterol 134 mg/dl, triglyceride 109 mg/dL, glucose 78 mg/dl, creatine phosphokinase (CPK) 396 U/L, CPK-MB isoenzyme 12 U/L and cardiac troponin-T negative. The electrocardiogram showed sinus rhythm with a rate of 60 bpm, normal axis and T-wave inversion in precordial leads V_3 to V_5. On echocardiography, the left ventricular function was normal (ejection fraction was 60%), the left ventricular end-diastolic diameter was 53mm. Structures and functions of heart valves were also normal. During in-hospital follow-up, the patient did not have angina with oral triple anti-ischaemic therapy (metoprolol 50 mg once a day, isosorbide mononitrate 40 mg once a day, amlodipine 5 mg once a day) at bed rest. No signs of redistribution were found on exercise thallium-201 perfusion scintigraphy, despite the patient had chest pain during exercise, and the ECG showed 0.1mV horizontal ST-segment depression in precordial leads V_5 and V_6 (Fig. 3). We did not repeat coronary angiography, because it was performed recently, and revealed bilateral coronary artery to left ventricle fistulas without atherosclerotic lesions. We had no chance other than pharmacologic therapy to treat the patient, and we prescribed him triple anti-ischaemic drugs.

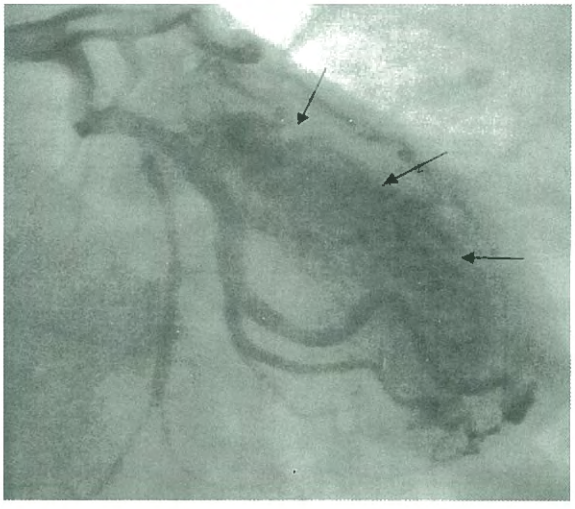

Figure 1: *Left coronary arteriograms with left caudal projection demonstrate left anterior descending coronary artery to left ventricle fistulas with multiple origins and multiple terminations (arrows)*

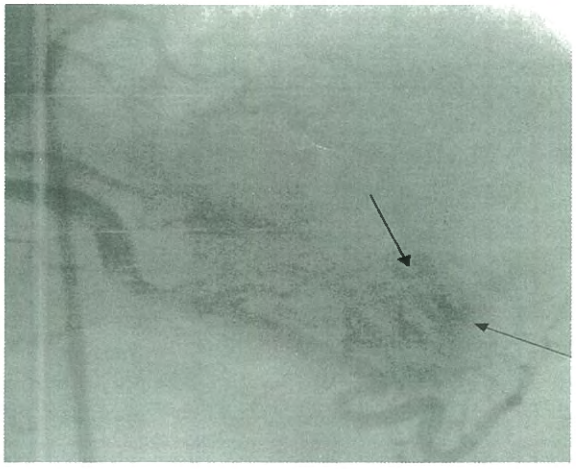

Figure 2: *Right coronary arteriogram demonstrates right posterior descending coronary artery to left ventricle fistulas with multiple origins and multiple terminations (arrows)*

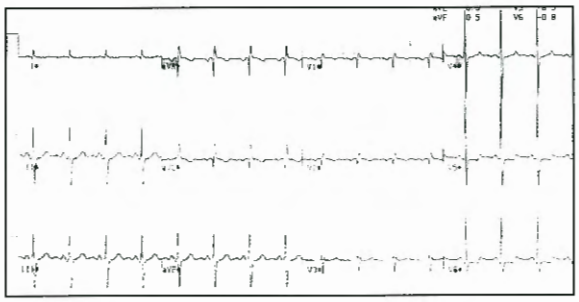

Figure 3: *While patient had angina during exercise test, the electrocardiogram showed 0.1mV horizontal ST-segment depression in precordial leads V5 and V6. But perfusion scintigraphy could not demonstrate a reversible defect*

DISCUSSION

The prevalence of CAFs has been reported as 0.1 to 0.2% in selective coronary angiography series[2-8]. The arterio-arterial fistulas (left ventricle, left atrium, pulmonary veins) are very rare. Cardiac symptoms such as precordial pain, dyspnoea and palpitation associated with a CAF do not usually appear until adult life. The presence and significance of symptoms depends upon the magnitude of shunting, whether or not myocardial blood flow is affected by the shunt. Surgery or percutaneous interventional procedures are generally accepted for patients with symptomatic CAFs, depending on the anatomy of the fistula and concomitant heart disease[3,9,10]. Pharmacological treatment is preserved for asymptomatic or symptomatic patients with "small" CAFs, or patients without favorable anatomy for surgery and interventional procedures. In the presented case, bilateral coronary artery to left ventricle fistulas resulted in coronary steal phenomenon producing myocardial ischaemia. The occurrence of coronary steal phenomenon in a coronary artery to left ventricular fistula has been previously documented[11]. It has been shown that the ECG or myocardial perfusion scan could not demonstrate ischaemia in patients with angina associated with CAFs[8,12,13]. Although the patient had angina during exercise protocol and the ECG showed 0.1 mV ST-segment depression in precordial leads V_5 and V_6, a reversible perfusion defect could not be demonstrated on myocardial scintigraphy. Selective coronary angiography is considered to be the gold standard for definite diagnosis of CAFs. In our patient, coronary arteriography demonstrated multiple, bilateral coronary artery to left ventricular fistulas without atherosclerotic lesions. The amount of contrast media traversing the fistulas was enough to fill the entire left ventricular cavity.

The presented case carries two interesting aspects. First, multiple, bilateral coronary artery to left ventricle fistulas are rare congenital anomalies. Second, the coronary steal through the fistulas resulted in ischaemia and angina pectoris resistant to anti-ischaemic therapy. Although therapy with beta-blockers has been successfully established in patients with CAFs[11], our patient still had angina and fatigue despite concomitant use of a beta-blocker and a calcium antagonist. This case showed us that new therapeutic modalities should be tested in such patients in whom surgical repair or interventional procedures seem technically impossible.

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