A Clinically Silent Case of Prominent Thebesian System: Diagonal Branch of Left Anterior Descending Coronary Artery to Left Ventricular Communication

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ABSTRACT

Fistulous communication from the coronary artery to the left ventricle (LV) is extremely rare and has received little attention in the literature. Moreover, a case of coronary artery fistula (CAF) from a diagonal branch of the left anterior descending coronary artery to the LV has been reported only once in the literature. Herein, we report a case of CAF from a diagonal branch of the left anterior descending coronary artery to the LV, along with a review of the pertinent literature regarding this disorder. (Korean Circulation J 2004;34(1):107-111)

KEY WORDS: Coronary vessels; Vascular fistula; Veins.

Introduction

Coronary artery fistula (CAF) is an unusual anomaly and a predominantly congenital communication between the coronary arterial system and the chambers or great vessels of the heart. Generally CAF presents as a single fistula terminating into the right heart or the pulmonary trunk. Fistulas to the LV are extremely rare. Although coronary artery-to-left ventricular fistula (CALVF) is considered as a possible cause of exertional angina, some cases of CALVF are clinically silent. In this report, we describe a case of CALVF (diagonal branch of left anterior descending coronary artery to LV). An accompanying review of the literature shows the broad spectrum of its clinical presentation.

Case

A 68-year-old man was referred from a local clinic for further cardiac evaluation. He was incidentally told that his ECG was abnormal at routine tests for cataract surgery. He denied chest pain and shortness of breath, but did report occasional episodes of palpitation. He smoked 20 cigarettes per day for 40 years and there was no family history of cardiac disease. Past health status was good, except for hypertension, which had not been treated with any medication. Physical examination revealed a blood pressure of 148/92 mmHg and an irregular pulse of 68/min. There was no heart murmur, gallop, or click. The neck veins were flat and the lungs clear. Cardiomegaly (a cardiothoracic ratio of 0.62), without lung congestion, was seen on a chest x-ray and routine lab tests were normal. The ECG revealed atrial fibrillation, left ventricular hypertrophy and a nonspecific abnormal ST-T change in leads II, III, AVF, and V 5-6. The treadmill exercise test showed a good exercise tolerance without chest pain, but it disclosed a horizontal

ST-segment depression from the second stage of exercise to the 2-minute recovery period. Stress technetium-99m (^{99m}Tc)-sestamibi single-photon emission computed tomography (SPECT) showed no fixed lesions or reversible myocardial ischemia. Echocardiographically, the patient had concentric left ventricular hypertrophy (LVH) and an enlarged left atrium, in which spontaneous echo contrast was visible. The global LV function was normal and there was no segmental wall motion abnormality or valvular dysfunction. Left ventriculography revealed the LV to be normal in size and contractility. Coronary angiography showed no significant stenosis. The vessels appeared somewhat dilated and elongated, and the egress of contrast from the distal quarter of the diagonal branch was observed to form a whirling smoke-like shadow, draining quickly into the left ventricular cavity (Figure 1). The coronary venous drainage was normal. The patient was discharged on verapamil to control hypertension. During an eight-month follow-up period, there was no symptom related to CALVF.
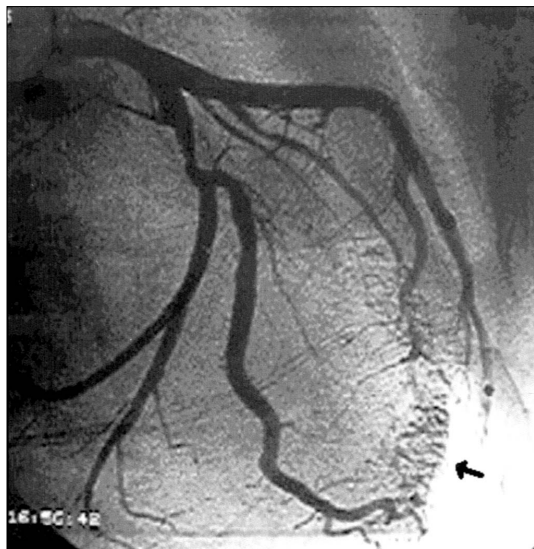

Figure 1. Selective left coronary arteriogram in right anterior oblique projection showing diagonal branch of left anterior descending coronary artery draining into left ventricle via a network of fine vessels (arrow).

Discussion

Although direct communication between the coronary arteries and the cardiac chambers was first recognized by Vieussens in 1706, the pathogenesis of CALVF is not clear.[1] It is presumably congenital in origin, even though it could be acquired due to a localized inflammatory process or local injury including stab wound, percutaneous coronary angioplasty, and endomyocardial biopsy. It is also possibly associated with myocardial infarction, thrombi or myxomas.[2-6] In case of combinedhypoplasia of the coronary sinus and some of the cardiac veins, the abnormality is also described to drain the major coronary venous blood.[7]

It has been hypothesized that CALVF most likely represents partial persistence of the network of Thebesian veins that provide blood supply to the subendocardial regions of the myocardium during embryogenesis and are usually obliterated in normal development.[1)6-13] The Thebesian veins are traditionally known to have two separate functions, a route of myocardial drainage and an alternative route of nourishment to the myocardium.[1] The Thebesian foramina were identified throughout the ventricular walls and interventricular septum, in particular at the apex of the ventricles and at the base of the papillary muscles, in human and animal hearts using the microangiographic method.[1)9] Wearn (1928) and Wearn et al. (1933) found Thebesian openings to be more numerous in the ventricles than in the atria; they also found direct connections between the arteries and Thebesian veins, which they named arterioluminal and arteiosinusoidal vessels.[1] These communications may evacuate their contents to the myocardial sinusoids, direct to the intertrabecular spaces, or to the heart cavities from the top of the trabeculae.[10] They may have a plexiform pattern or appear as a large single conduit.[5)6)11]

In this case, small vessels, consisting of a plexiform fine vascular network, were interposed between the epicardial coronary artery and the heart cavity and emptying into the heart. Because our patient was otherwise healthy and did not have any other medical or trauma history,

his anomaly can be best classified into the congenital arteriosinusoidal type of CALVF, where communication is through a myocardial sinusoidal network and also traverses capillary beds.

Congenital CAF have been reported in 0.08−0.3% of 126,595 adults undergoing coronary angiography.[14] The true incidence of CAF in the general population is not known, since the absence of symptoms, in most cases, does not promote diagnostic investigation. In a series of 101 cases, 79% of patients were found to have a single fistula, while 21% displayed multiple fistulas.[15] CAF arise from the right coronary artery or its branches in 10−50% of cases; the remaining fistulas arise from the left coronary system in 42−87%.[15-17] In only 3−5% of patients were both coronary arteries involved.[16,17] Right-sided structures such as right atrium, right ventricle, pulmonary artery, or coronary sinus were the recipient site in 76−92% of patients with CAF. Accordingly, drainage into the left atrium or left ventricle was present in only 8−24% of these patients.[15-17] Moreover, fistulas draining into the LV are extremely rare, amounting to only 3−17% of all CAF.[15] To our knowledge, this is the second report of focal CAF from a diagonal branch of the left anterior descending coronary artery to the LV in the world-wide literature.[2]

In most cases of CALVF, patients are asymptomatic and the fistulas are coincidentally found.[8,11,15,16,18,19] However, anginal symptoms have frequently been described.[5,7,13,20-25] Myocardial ischemia could result from a shunt-related coronary steal, due to the diversion of oxygen-rich blood into the LV cavity via the low-resistance fistulous channels bypassing the myocardium, or a critical drop in perfusion pressure because of rapid run-off and impaired subendocardial perfusion of the hypertrophied myocardium.[12,25] Exercise stress test, myocardial perfusion scintigraphy, Holter monitoring, and coronary sinus lactate determination during atrial pacing have all been used to demonstrate myocardial ischemia.[3,5-8,11,12,20-24]

Other patients rarely show symptoms of congestive heart failure, as a consequence of the left-to-left shunt into the LV causing diastolic volume overload, which can give the clinical feature of aortic valve insufficiency.[5,20,23] However, shunt volumes and the hemodynamic significance of CALVF are highly variable.[5,12] In our case, the normal pulse pressure and the absence of LV enlargement, as well as the absence of a continuous or diastolic murmur indirectly suggested that there was no hemodynamically important shunt and volume overload of the LV. LVH might be caused by chronic untreated hypertension.

The CALVF can also cause infective endocarditis, a huge coronary aneurysm, and rupture or thrombosis of the fistula.[5,25,26]

Clinical diagnosis of CALVF is difficult. Although CAF has been known to be associated with continuous or purely diastolic murmurs, typically there are no pathological physical findings, ECG, chest x-ray, and echocardiogram findings in CALVF, except when associated with cardiac anomalies.[5,15,20,22-24] Sometimes a fourth heart sound is audible.[8,13,20] A widened pulse pressure may also be present.[20]

Abnormal ST-T changes, LVH and left bundle branch block have been described on resting ECG.[22] Chest x-ray and two dimensional echocardiogram including color, continuous and pulsed wave Doppler are usually normal.[6,23] Diastolic flutter of the anterior mitral leaflet and concentric LVH can be found in association with diastolic blood flow into the LV and consequent volume overload.[6,20]

With the advent of high-resolution 2-dimensional and color Doppler transthoracic echocardiography, however, the incidental diagnosis of clinically silent CAF including CALVF is increasing.[17,27] Color Doppler flow mapping has proved to be a highly sensitive method of identifying small CAF.[17] Transesophageal echocardiography has also been utilized to diagnose CALVF.[28]

But, in most cases, CALVF is identified during routine coronary angiography and emergent catheterizations.[23,24] Occasionally, concomitant coronary atherosclerosis is found.

Angiographically, a plexus of small vessels that origi-

nate from the coronary artery and empty contrast medium into the LV cavity, which is described as "diastolic rain", is visualized, together with LV filling.[22][24] The epicardial coronary artery proximal to the fistula can be normal or, in large shunts, can be tortuous, elongated, or aneurysmally dilated.[5] Some cases showed that the feeding artery was totally occluded distally to the origin of the communications, and significant atherosclerosis was present in either the fistulous or the non-fistulous coronary artery.[24] Interestingly, this patient had a localized fistulization from a single coronary artery, but all three major arteries were dilated and elongated. The diffuse coronary ectasia was, we think, either another anomaly or an early consequence of atherosclerosis, known as positive remodeling.

The management of patients with CALVF remains controversial. Given the relatively benign nature of this disease, treatment is usually required only for patients with angina or symptoms of congestive heart failure.[5][15][17][29]

Reduction of angina pectoris has been usually achieved with β-blockers, nitrates, and calcium channel blockers.[3][5][6][12][20][21][23] However, it should be noted that calcium channel blockers and nitrates may also dilate fistulas and increase the magnitude of steal.[5][6] Surgical intervention for CAF is indicated and feasible only in the presence of large shunts, other major cardiac lesions, concomitant atherosclerotic coronary artery disease, symptoms, or complications including progressive enlargement, bacterial endocarditis, rupture, pulmonary hypertension, and thromboembolism.[23] Antibiotic prophylaxis against infective endocarditis seems appropriate for all patients with CALVF.[5][6][23]

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