

A case of multiple coronary microfistulas to the left ventricle and apical myocardial hypertrophy coexisting with stable angina



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A coronary artery fistula consists of a communication between a coronary artery and a cardiac chamber, a great artery or the vena cava. It is the most common congenital anomaly that can affect coronary perfusion. However, coronary fistulas to one of the cardiac chambers and coexisting apical myocardial hypertrophy are infrequent anomalies, and usually are found unexpectedly. Herein, we report a case in which all three major coronary arteries emptied into the left ventricle with apical hypertrophy, through multiple microfistulas.

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Keywords: Coronary artery fistula, Coronary steal phenomenon, Stable angina

Introduction

Coronary artery fistula is a rare anomaly occurring in 0.2–0.4% of patients in previous studies of congenital heart disease, and its incidence in overall populations is estimated to be approximately 0.002%. Blood in the coronary artery fistula is shunted into a cardiac chamber, great vessel or other structure, bypassing the myocardial capillary network. Coronary artery fistula is also the most common congenital coronary anomaly affecting hemodynamic parameters [1–4]. Approximately half of all patients with coronary artery fistula remain asymptomatic; congestive heart failure, infective endocarditis, myocardial ischemia induced by a coronary ‘steal’ phenomenon,

or rupture of an aneurysmal fistula may develop in the other half [5–6]. Multiple arterio-systemic fistula, arising from all three major coronary arteries and draining into the left ventricle, are rare, and the clinical and hemodynamic sequelae are incompletely understood. This report describes the three vessel involvement of a coronary artery fistula, and this was coexistent with apical left ventricle hypertrophy.

Case report

A 58-year-old woman presented with complaints of progressive dyspnea on effort and also chest discomfort for two months. Her medical history

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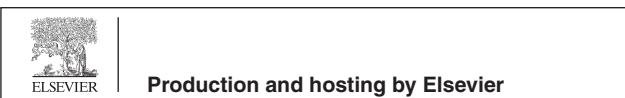


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included hypertension but unknown coronary artery disease, diabetes mellitus hyperlipidemia or smoking. There were no abnormal findings on her physical examination. The electrocardiogram displayed normal sinus rhythm; the chest X-ray showed no signs of pulmonary hyperemia or mass lesions, and the central shadow was normal (cardiothoracic ratio = 0.40).

The transthoracic echocardiography demonstrated left ventricular diastolic dysfunction and apical hypertrophic cardiomyopathy (Fig. 1). Myocardial perfusion scintigraphy showed a massive area of residual ischemia within culprit lesion of the anterior zone starting from the subapical level to the basal cross-section (Fig. 2). The patient therefore underwent cardiac catheterization. There were no atheroma and stenotic lesions on the coronary angiogram; however, a heavy stream of contrast agent entered the left ventricle via an apparent plexus of intramural vessels, from the distal third of both the left and right coronary arterial systems and it revealed coronary artery micro fistulae coexistent with stable angina. All fistulae drained into the left ventricle; the first originated from the distal septal branch of the left anterior descending artery; the other originated from circumflex artery and the latter originated from a distal right ventricular branch of the right coronary artery (Figs. 3 and 4). She was treated with medically and was discharged with metoprolol and aspirin. She told that her symptoms were regressed after the treatment at three and six months follow up.

Discussion

The pathophysiologic importance of a coronary arterial fistula is related to the amount of blood that flows through the communication to the

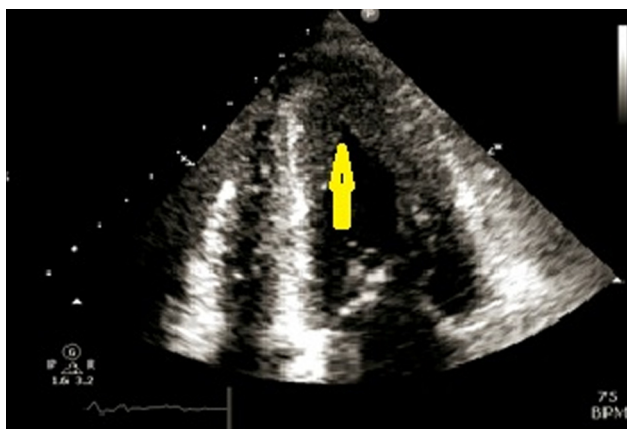


Figure 1. On 2-D echocardiography, arrow indicates hypertrophy in the apical region and apical 4-chamber view of the left ventricle.

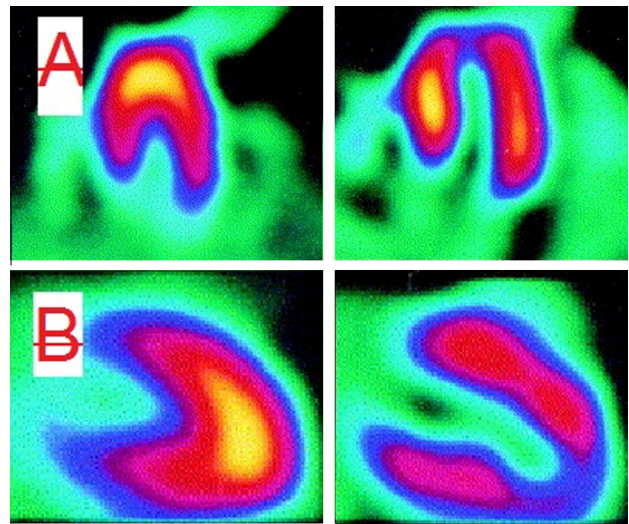


Figure 2. Rest (left) and stress (right) SPECT horizontal (A) and vertical (B) long-axis slices from a patient in the apical hypertrophic cardiomyopathy. Increased resting apical tracer uptake and stress-induced apical and inferoapical perfusion defects are noted.

chamber or vessel into which the fistula drains; myocardial ischemia is believed to be the result of this fistulous bypass. If the fistulous pathway drains into the left ventricle, the hemodynamic results are similar to those of an aortic regurgitation. In the diagnosis of coronary fistula, electrocardiography is unhelpful, although it can sometimes point out left ventricle overload and ischemic ST-segment changes. Echocardiography may help to identify large fistulae. However, exact diagnosis is possible only upon coronary angiography [7].

The main presenting symptom of our patient was chest pain which was indicative of ischemia referred by myocardial perfusion scintigraphy. We inferred that the pathophysiologic mechanism of this patient's ischemia might be myocardial



Figure 3. Fistulas of circumflex and left anterior descending arteries.

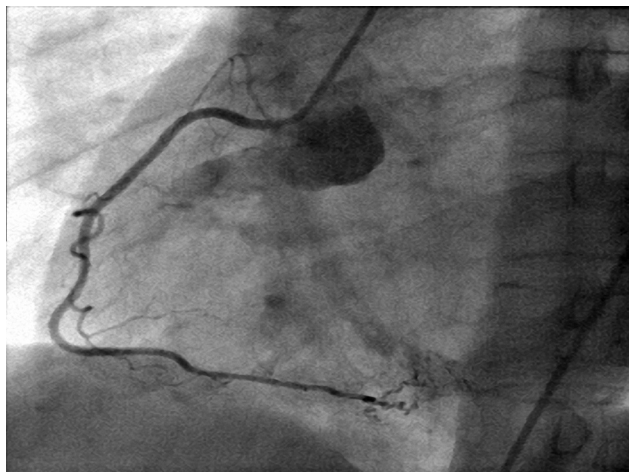


Figure 4. Fistula of right coronary artery.

steal, due to reduction of blood flow at points distal to the fistula or apical hypertrophic cardiomyopathy. The mechanism is probably related to the diastolic pressure gradient caused by blood run-off from the coronary vasculature to a low-pressure receiving cavity. If the fistula is large or multiple, as observed in our patient, we suggest that intracoronary diastolic perfusion pressure may diminish progressively. The myocardium beyond the origin of the multiple fistula is at risk of ischemia, which is most frequently evident in association with exercise or other physical activity (increased myocardial oxygen demand). We found reversible perfusion defects in multiple myocardial areas by means of stress/rest Tc 99 m sestamibi myocardial perfusion single-photon emission tomography. The correspondence between the sites of the coronary artery where the fistula originated, identified by coronary angiography, and the sites of the reversible perfusion defects, identified by myocardial perfusion scintigraphy, indicated that anomalous flow through the fistula was the likely cause of the myocardial ischemia. Because the coronary artery-cardiac chamber communications were multiple and diffuse, neither surgery nor transcatheter coil occlusion were considered in this case.

Generalized arterio-systemic fistula originating from all three major coronary vessels has been reported in very rare cases in the literature.[8] Myocardial infarction in cases of coronary artery fistula is rare. It was thought that the volume increase in the left ventricle due to coronary steal syndrome formed conditions suitable for thrombosis.

Indications for treatment include a large left-to-right shunt, myocardial ischemia, endocarditis, and congestive heart failure. The goal of treatment is to occlude the fistula while providing normal

coronary circulation. In some cases, coronary fistulae are spontaneously obstructed as a result of atherosclerosis, leading to asymptomatic patients.[9]

Surgical intervention is not without risk. Myocardial infarction may develop in 5% of patients during surgery, and then fistula may recur. Medical treatment is recommended for such patients, and the number of surgical procedures has decreased considerably due to the development of percutaneous closure methods. Such methods include the use of coils, detachable balloons, or alcohol injection.

There are some reported cases of multiple coronary artery-systemic fistula with apical hypertrophic cardiomyopathy [10]. The cardiomyopathy itself is thought to be the cause of micro fistulae formation by inducing an anomaly in the Thebesius Venous System [11]. Our patient also had hypertrophic cardiomyopathy.

In conclusion, multiple coronary-systemic fistulae are a rare cause of myocardial ischemia via the coronary steal phenomenon. The anatomical types of fistulous connections and the severity of leakage vary, and this may alter the myocardial ischemia level and clinical symptoms.

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