

A very rare cause of continuous murmur and coronary ischemia: high-flow coronary-to-pulmonary artery fistula

Devamlı üfürüm ve iskeminin çok nadir bir nedeni:
Yüksek akımlı sol ana koroner arter-pulmoner arter fistülü

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A 52-year-old woman presented with complaints of chest discomfort and angina radiating to the neck and left arm on exertion. She reported that the severity of angina was increasing after sublingual nitrate intake. Coronary angiography showed a high-flow fistula between the left main coronary artery and pulmonary artery. Left coronary arteries were normal, but there was a stenotic lesion in the right coronary artery. Coronary artery fistula was ligated successfully under bypass surgery. It was thought that the severity of myocardial ischemia caused by the high-flow fistula was aggravated by sublingual nitrate due to coronary steal phenomenon.

Key words: Coronary vessel anomalies/complications; hypertension, pulmonary/etiology; myocardial ischemia/etiology; vascular fistula/complications/surgery.

Coronary artery fistula (CAF) is a rare anomaly in which a communication exists between a coronary artery and a cardiac chamber or another vascular structure. It is observed in 0.3% to 0.8% of patients undergoing coronary angiography.^[1,2] Its frequency is higher in the right coronary artery compared to the left coronary artery and it arises from both in approximately 5% of patients.^[3] Coronary angiography is the gold standard diagnostic approach in patients with a CAF. Left main coronary artery fistula has been rarely reported in the literature.

We presented a case in which a high-flow fistula between the left main coronary artery and pulmonary artery caused severe myocardial ischemia that was complicated by sublingual nitrate use due to coronary steal phenomenon.

Elli iki yaşında bir kadın hasta, göğüste rahatsızlık ve egzersizle ortaya çıkan ve boyna ve sol kola yayılan göğüs ağrısı yakınmalarıyla kliniğimize başvurdu. Hasta, göğüs ağrısının dilaltı nitrat kullanımıyla arttığını bildirdi. Koroner anjiyografide sol ana koroner arterle pulmoner arter arasında yüksek akımlı bir fistül görüldü. Sol koroner arterler normaldi; fakat, sağ koroner arterde darlık vardı. Koroner arter fistülü baypas cerrahisi ile başarılı bir şekilde bağlandı. Yüksek akımlı fistülden kaynaklanan miyokard iskemisinin, koroner çalma fenomeninin etkisiyle dilaltı nitrat kullanımıyla arttığı düşünüldü.

Anahtar sözcükler: Koroner damar anomalisi/komplikasyon; hipertansiyon, pulmoner/etyoloji; miyokard iskemisi/etyoloji; vasküler fistül/komplikasyon/cerrahi.

CASE REPORT

A 52-year-old woman had complaints of chest discomfort and effort-related angina that spread to the neck and left arm for two-years. She had been receiving antihypertensive therapy for five years. She also gave an account of sublingual nitrate use when angina occurred on exercise, but the severity of angina worsened following nitrate use. On physical examination, blood pressure was 165/90 mmHg and heart rate was 74 bpm. Chest auscultation revealed a continuous murmur of grade 2/6 at the upper left sternal border. Electrocardiography showed T-wave inversion in leads DI, aVL, and V1-6 derivations. Echocardiography showed normal ventricle wall motion and ejection fraction was 65%. Pulmonary artery systolic pressure measured using tricuspid regurgitant jet velocity

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was 45 mmHg. As she had gonarthrosis, treadmill exercise test could not be performed. Thus, myocardial ischemia was estimated by myocardial perfusion scintigraphy, which showed severe (3+) ischemia in anterior and lateral wall segments. On coronary angiography, we did not find coronary artery stenosis in the left coronary artery system, but found a high-flow coronary artery fistula originating from the left main coronary artery and draining into the pulmonary artery (Fig. 1a, b) and coronary stenosis (60%) in the right coronary artery (Fig. 2). We recommended surgery for ligation of the fistula on cardiopulmonary bypass because coil embolization would not be safe due to the close localization of the fistula to the ostium of the left main coronary artery. Fistula repair was performed successfully, and the patient was discharged four days after the operation.

DISCUSSION

Coronary artery fistula is a rare anomaly in which a communication exists between a coronary artery and a cardiac chamber or another vascular structure.^[1,2] The site of the fistula drainage is of great significance, because it determines its flow rate: fistulas that drain into a low-pressure site would have a higher flow rate.^[4] A coronary artery fistula with a high flow reserve has significant clinical implications as in our case. Myocardial ischemia may occur on exertion in the presence of a high-flow fistula due to coronary steal. If fistula flow is high, pulmonary artery pressure increases and right heart failure may ensue. Percutaneous embolization or surgical ligation should be considered to prevent right heart failure if the flow of the fistula is high. Our case is of particular interest, in that the high-flow fistula caused myocardial ischemia which was further worsened due to coronary steal following sublingual nitrate intake. A high-flow CAF is considered when the severity of ischemia or angina increases despite nitrate therapy. In patients with a CAF draining into the left side of the heart or pulmonary artery, pulmonary hypertension may occur.^[5] In our case, fistula was draining into the pulmonary artery and systolic pulmonary pressure was measured as 45 mmHg.

In conclusion, a high-flow CAF may cause myocardial ischemia resulting in angina symptoms. In this situation, nitrate therapy may aggravate angina due to coronary steal. Percutaneous closure or ligation under cardiac bypass surgery should be considered to prevent increases in pulmonary artery pressure and/or heart failure.

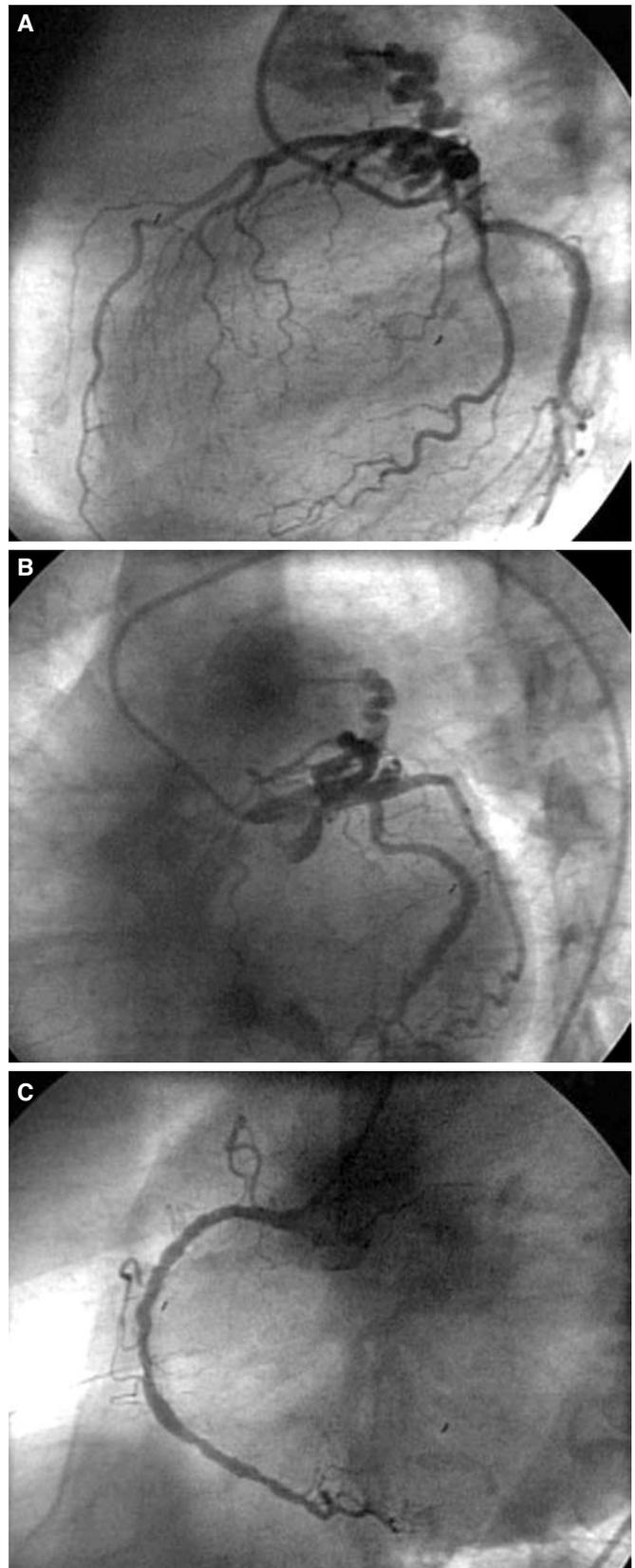


Figure 1. Left anterior (A) oblique (90°) and (B) oblique caudal (35°) views show a high-flow fistula originating from the left main coronary artery and draining to the pulmonary artery. (C) Left anterior oblique (45°) view shows diffuse stenosis in the right coronary artery with significant narrowing after the right ventricular branch.

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