Congenital coronary artery fistula as a cause of angina pectoris

Göğüs ağrısı nedeni olan doğuştan koroner arter fistülü

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Coronary arteriovenous fistula is an uncommon anomaly, representing an incidental finding in 0.1% to 0.2% of coronary angiograms. A 46-year-old man presented with a five-month history of palpitation and atypical stabbing chest pain on exertion and/or at rest. The electrocardiogram showed normal sinus rhythm, incomplete right bundle branch block with normal axis, and borderline left atrial abnormality. Echocardiography showed normal left ventricular function, mild left ventricular hypertrophy, moderate left atrial enlargement with mild mitral insufficiency, and moderate right atrial enlargement with mild-moderate tricuspid valve regurgitation. During exercise test, frequent ventricular ectopic beats were noted and the patient complained of atypical chest pain. Scintigraphy showed an ischemic defect in the inferior wall of the left ventricle. On coronary angiography, the right coronary artery and left anterior descending artery were normal, but the circumflex coronary artery was connected to the pulmonary artery through a congenital fistula. The patient refused any further intervention for fistula closure.

Key words: Angina pectoris/etiology; coronary angiography; coronary vessel anomalies; fistula; myocardial ischemia/etiology.

Koroner arteriyovenöz fistül nadir görülen, koroner anjiyografilerin %0.1-0.2'sinde tesadüfen rastlanan bir anomalidir. Kırk altı yaşında bir erkek hasta, beş aydır var olan çarpıntı ve hem istirahatte hem de egzersiz sırasında kendini gösteren atipik göğüs ağrısı yakınmalarıyla başvurdu. Elektrokardiyografide normal sinüs ritmi, normal eksenli inkomplet sağ dal bloku ve sınırda sol atriyal anormallik gözlendi. Ekokardiyografide sol ventrikül fonksiyonu normal bulunurken, hafif sol ventrikül hipertrofisi, hafif mitral yetersizlikle beraber orta derecede genişlemiş sol atriyum, hafif-orta triküspid kapak yetersizliği ile beraber orta dereceli sağ atriyum genişlemesi vardı. Egzersiz testinde sık ventrikül ektopik vuruları gözlendi ve hasta atipik göğüs ağrısından yakındı. Sintigrafide sol ventrikül inferior duvarında iskemik defekt görüldü. Koroner anjiyografide sağ koroner arter ve sol ön inen arter normal bulunurken, sirkumfleks koroner arteri pulmoner artere bağlayan doğuştan fistül izlendi. Hasta, fistülün kapatılması için herhangi bir girişimi kabul etmedi.

Anahtar sözcükler: Angina pektoris/etyoloji; koroner anjiyografi; koroner damar anomalisi; fistül; miyokard iskemisi/etyoloji.

Coronary artery fistula (CAF) is an abnormal precapillary connection between a coronary artery and neighboring structures, such as the right atrium, right ventricle, pulmonary artery, superior vena cava, or coronary sinus. It is observed in 0.1% to 0.2% of coronary angiographic studies.[1] This infrequent abnormality can be seen at any age. Although a CAF may originate from any site of the three major coronary arteries, including the left main trunk, it usually arises from the right coronary artery or left anterior descending artery. The circumflex coronary artery is rarely involved.[2]

Low-pressure structures are the most common sites of CAF drainage.[1] Over 90% of the fistulas drain into the venous circulation. It is usually accompanied by coronary artery dilatation whose extent is not always related to the shunt size.[3] Most fistulas present as single communications, but multiple fistulas have been reported.[4]

Although noninvasive coronary artery imaging may facilitate the diagnosis and identification of the origin and insertion of a CAF, cardiac catheterization and coronary angiography are necessary for the precise delineation of coronary anatomy, assessment of hemodynamic parameters, and to detect concomitant atherosclerosis and other structural anomalies. In symptomatic cases, therapeutic options include surgical correction or transcatheter embolization.

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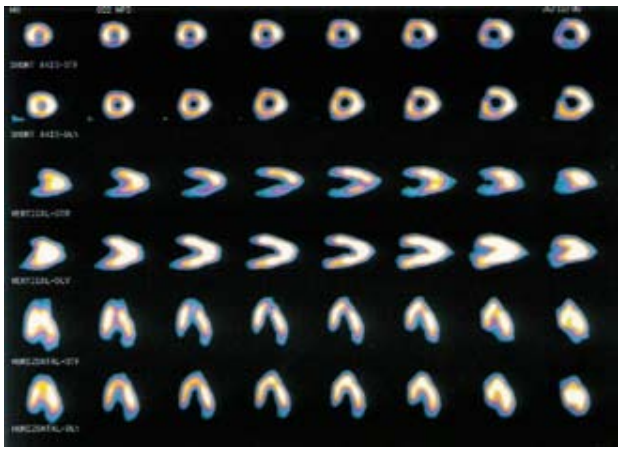

Figure 1. Scintigraphy shows an ischemic defect in the inferior wall of the left ventricle.

We report a symptomatic patient with a congenital coronary arteriovenous fistula, originating from the proximal part of the left circumflex artery and draining into the pulmonary artery.

CASE REPORT

A 46-year-old man presented with a five-month history of palpitation and atypical stabbing chest pain on exertion and/or at rest. He had no complaints of shortness of breath, paroxysmal nocturnal dyspnea, orthopnea, or peripheral edema. He was a nonsmoker, had no cardiac risk factors or a positive family history for coronary artery disease, congenital heart disease, or sudden cardiac death. In addition, there was no history of significant chest wall trauma or any invasive cardiac procedure. On physical examination, blood pressure was 140/70 mmHg and heart rate was 76 beats/sec. There was no rise in jugular venous pressure, cardiac auscultation was normal without added sounds or murmurs, and the chest was clear with adequate air entry on both sides. The electrocardiogram showed normal sinus rhythm, incomplete right bundle branch block with normal axis, and borderline left atrial abnormality. Echocardiography showed normal left ventricular function without regional wall motion abnormalities, mild left ventricular hypertrophy, moderate left atrial enlargement with mild mitral insufficiency, and moderate right atrial enlargement with mild-moderate tricuspid valve regurgitation. Pulmonary artery pressure was estimated as 36 mmHg. The cardiothoracic ratio was normal on a posteroanterior chest x-ray view. Laboratory investigations were normal. He achieved maximal exercise treadmill test with the standard Bruce protocol without significant ST-segment changes. However, during exercise test, frequent ventricular ectopic beats were noted and he complained of atypical chest pain. On 99mTc-MIBI scintigraphy, an ischemic defect was identified in the inferior wall (Fig. 1). Cardiac catheterization showed no significant step-up in oxygen saturation in the right heart and normal pulmonary artery pressure. Coronary angiography showed normal right coronary artery and left anterior descending artery, but the circumflex coronary artery was connected to the pulmonary artery through a fistula (Fig. 2). The patient was strongly recommended surgical intervention via transcatheter embolization or surgical closure under cardiopulmonary bypass for the CAF between the circumflex artery and the coronary sinus, but he refused any further intervention.

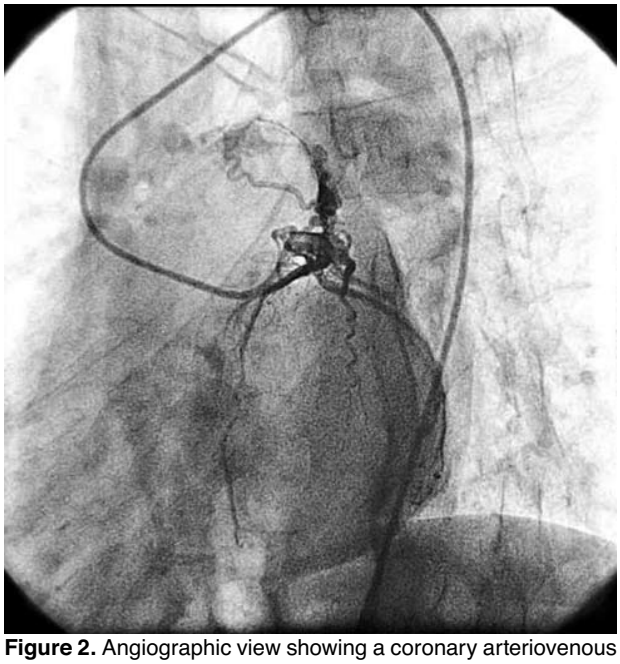

Figure 2. Angiographic view showing a coronary arteriovenous fistula originating from the proximal part of the left circumflex artery with drainage into the pulmonary artery.

DISCUSSION

Most coronary artery anomalies are found accidentally during angiographic evaluation for specific/nonspecific cardiac symptoms. They may be congenital or acquired as a result of trauma (stabbing, gunshot and projectile injuries) or iatrogenic due to invasive cardiac procedures (pacemaker electrode implantation, endomyocardial biopsy, and coronary angiography). They predominantly originate from the right coronary artery, may not be associated with other congenital heart diseases,[5] and are not gender-specific. Drainage depends on the site of origin, size of the fistula, and on the receiving chamber, while the hemodynamic effect of CAF depends on the site of drainage, and resistance within the fistula.

The clinical presentation of CAF is mainly dependent on the severity of the shunt. Although about half of the patients are asymptomatic and the vast majority of adults have good prognosis,[1] symptomatic patients may present with palpitation, chest discomfort, heart failure, bacterial endocarditis, or with an incidental continuous murmur which is characteristically heard over the left sternal border and apex.[6] Most elderly patients present with shortness of breath, right ventricular enlargement or dysfunction as a result of progressive enlargement of the fistula, and increased left-to-right shunting. Coronary artery anomalies may be complicated by myocardial ischemia or infarction due to stealing of coronary blood flow down the fistulous tract, and rarely rupture.[7,8] Transthoracic echocardiography (TTE) is an important primary noninvasive tool for identifying the anomalous origin of coronary arteries. A continuous systolic and diastolic turbulent flow pattern characterizes the shunt entry site, while flow in the distal site of a CAF may not be detected via TTE. Compared to TTE, transesophageal echocardiography (TEE) more accurately defines the origin, course, and drainage site of CAF.[1] Contrast-enhanced electron-beam tomography is a newer, more sensitive diagnostic tool with excellent spatial resolution, enabling identification of most coronary anomalies including origin, course, and insertion of CAFs. However, cost of ionizing radiation and potential contrast agent-related complications such nephrotoxic or allergenic reactions relatively restrict its use. Magnetic resonance imaging (MRI), on the other hand, is not associated with radiation- or contrast-related risks and provides excellent visualization. Of all these noninvasive modalities, coronary angiography still remains to be the gold standard imaging tool for diagnosing coronary anomalies and can be used selectively as a diagnostic and therapeutic procedure.[1]

Closure of congenital CAFs is strongly recommended in symptomatic patients. Clinical symptoms of coronary ischemia, such as exertional angina or dyspnea are the primary indications for closure of a fistula.[7] Surgical closure by vessel ligation is effective and provides long-term safety. However, catheter-based closure has become the preferred treatment option, if technically available. The most frequent complication associated with this procedure is embolization of the occlusion device, occurring in seven of 40 patients in one series.[9]

Small fistulas usually show a benign prognosis, symptom-free course, and even in some cases, tendency to spontaneous closure.[1] However, elective closure of congenital CAFs is recommended in asymptomatic patients to prevent potential complications,[9] such as congestive heart failure, infectious endocarditis, myocardial infarction, and death.[10] The incidence of these complications may be higher in patients older than 20 years than in younger patients (63% *vs* 19%).[10] Considering that fistula-related complications increase with age, and that the presenting symptom in our case was anginal pain caused by CAF, closure of congenital CAFs in asymptomatic patients seems to be a more reasonable approach.

In conclusion, despite their rare existence, CAFs may lead to specific/nonspecific cardiac symptoms and life-threatening complications; therefore, they should always be considered during diagnostic work-up.

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