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Surgical treatment of congenital coronary artery-pulmonary artery fistula: a case report

Doğuştan koroner arter-pulmoner arter fistülün cerrahi tedavisi: Olgu sunumu

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Congenital coronary artery fistula is a rare congenital heart disease. A 65-year-old female case was admitted to our outpatient clinic with the complaints of back and chest pain lasting for one year and increasing with exercise. A fistula originating from the left anterior descending coronary artery and drained into the main pulmonary artery was detected. The fistula was ligated using off-pump cardiopulmonary bypass.

Key words: Congenital coronary artery fistula; pulmonary artery; surgery.

Doğuştan koroner arter fistülü, nadir bir doğuştan kalp hastalığıdır. Altmış beş yaşında kadın olgu, bir yıl önce başlayan, egzersizle artan sırt ve göğüs ağrısı yakınması ile polikliniğimize başvurdu. Sol ön inen koroner arterden köken alan ve ana pulmoner artere drene olan bir fistül tespit edildi. Fistül kardiyopulmoner baypas yapılarak atan kalpte ligatüre edildi.

Anahtar sözcükler: Doğuştan koroner arter fistülü; pulmoner arter; cerrahi.

A congenital coronary artery fistula (CCAF) consists of communication between one of the coronary arteries and a cardiac chamber or a great artery [the superior vena cava (SVC), the inferior vena cava (IVC), or the pulmonary artery or pulmonary veins]. This type of fistula is rare with a reported incidence of 0.1 to 0.2%.[1]

A pathogenetic explanation has been given for this anomaly which states that during the embryologic life, communication between the coronary arteries and great vessels or ventricular chambers persists. Later on, this communication matures into a normally calibrated capillary network. Failure to complete this process could lead to the creation of major congenital fistulas.[1] There are a limited number of reports about CCAFs in our country.[2-6] Therfore, we present a case of successful surgical treatment for left anterior coronary artery to pulmonary artery fistula.

CASE REPORT

A 65-year-old female was admitted to our outpatient clinic with a one-year history of back and chest pain on minimal effort that had been progressive in nature.

Physical examination revealed no significant findings. The electrocardiogram (ECG) was normal with no signs of ischemia at rest and normal sinus rhythm. However, left bundle-branch block was seen. Echocardiography showed normal left ventricular function and mild left ventricular hypertrophy. On coronary angiography, the coronary arteries were normal, but the left anterior descending coronary artery (LAD) was connected to the pulmonary artery through a congenital fistula (Figure 1). The pulmonary to systemic flow ratio (Qp/Qs) was 1.7:1. In addition, left ventriculography revealed normal function in the left ventricle. The patient was operated on through a midline sternotomy.

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After opening the pericardium, a tortuous CCAF was seen to course from the proximal segment of the LAD to the entrance of the proximal part of the main pulmonary artery (MPA) (Figure 2). Because of the pulmonary artery orifice, it was decided to ligate the fistula under beating heart cardiopulmonary bypass (CPB) surgery. The extracardiac fistula was ligatured with 5/0 monofilament transfixion sutures. Afterward, the internal fistula orifice and pulmonary artery were closed using the same suture. The postoperative course was uneventful, and the patient was discharged eight days after the operation. She remains asymptomatic one year after her surgery.

DISCUSSION

Krause was the first to describe CCAFs in 1865. In 50% of the cases, they have been reported to arise more commonly from the right coronary artery (RCA). They originate less frequently from the LAD (25%), the circumflex coronary artery (CCA) (18.3 %), diagonal branch (1.9%), and the left main coronary artery (LMCA) or the circumflex-marginal branch (both 0.7%). The coronary arteriovenous (AV) fistula drainage site was the pulmonary artery in 29.8-43% of patients while the right ventricle was the location in 14-40%, the right atrium in 19-20.2%, the left ventricle in 5.8-19%, and the left atrium in 5% of patients.[1] The coronary artery fistula may be isolated in some cases (55-80%), or it may be associated with other congenital cardiac anomalies (20-45%). These associated anomalies include tetralogy of Fallot, atrial septal defect, patent ductus arteriosus, ventricular septal defect. Besides, superimposed coronary artery disease may be associated in proportion as 35%.[1,3,7]

Drainage depends on the site of origin, size of the fistula, and the receiving chamber, whereas the hemodynamic effect of CCAF depends on the drainage site and resistance within the fistula. Most patients (40-50%) are asymptomatic.[1]

There are more asymptomatic CCAF patients who are under 20 years old. Indeed, age is an important determinant for the frequency of both symptoms and fistula-related complications.[7]

Symptomatic patients may present with dyspnea on exertion, fatigue, palpitation, chest discomfort, signs of fistula-related complications, or an incidental continuous murmur which is characteristically heard over the left sternal border and apex. The fistula-related complications include angina pectoris, myocardial infarction, subacute bacterial endocarditis, arrhythmias, sudden death, congestive heart failure, pulmonary arterial hypertension, and aneurysm formation. Angina pectoris and myocardial infarction due to coronary artery steal occur in 3-7% of CCAF patients. Elderly CCAF patients present with shortness of breath and right ventricular enlargement or dysfunction (congestive heart failure) as a result of the progressive enlargement of the fistula and increased left-to-right shunt.

Congestive heart failure develops in 19% of CCAF patients while subacute bacterial endocarditis develops 20%.[8] In rare instances, sudden death, arrhythmias, and conduction defects may be the first manifestation of CCAF. Although overall morbidity and mortality from anomalously terminating coronary arteries are low, most patients who do not have surgery for this condition develop symptoms and fistula-related complications as they get older. Fistula related complications develops

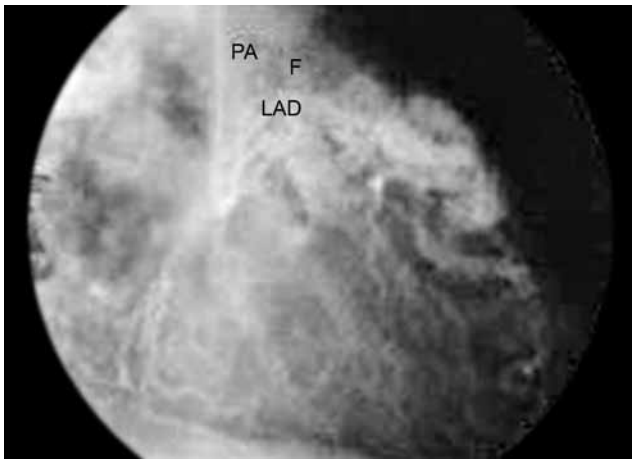

Figure 1. Coronary angiography finding of a congenital artery fistula. PA: Pulmonary artery; F: Fistula; LAD: Left anterior descending coronary artery.

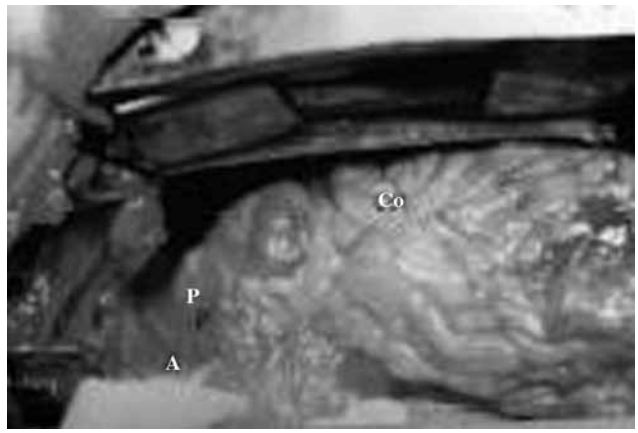

Figure 2. Intraoperative finding of tortuous fistulous communication between the proximal left anterior coronary artery and the pulmonary artery and collaterals. Co: Collaterals; P: Pulmonary artery; A: Aorta.

19% for patients younger than 20 years old, respect that 63% for patients older than 20 years old.[7] In addition, increased morbidity and mortality occurs when ligation is performed later in life. In a study by Liberthson involving 73 patients, left-to-right shunt flow averaged 1.6:1 (Qp/Qs), and there was no correlation between the clinical symptoms and the degree of shunting. However, in the patients who were over 20 years of age, there seemed to be an increased incidence of complications related to the shunt.[7]

There is general agreement that all symptomatic patients with an anomalously terminating coronary artery should undergo closure of the fistula, perhaps as soon as the diagnosis is made. Small fistulas usually show a benign prognosis, have a symptom-free course. In some cases, they even have a tendency toward spontaneous closure.[9] However, most surgeons would agree that treatment is also indicated in asymptomatic patients in order to prevent potential complications due to overload on the cardiac chambers in the case of a significant CCAF.[7,10]

Possible therapy for the management of CCAF includes prophylaxis of endocarditis, transcatheter embolization, and surgical ligation of fistula, but most surgeons also suggest that surgical ligation should be considered as the gold standard.[11]

Surgical closure by vessel ligation has been the most commonly used technique, yielding 100% survival and 100% closure rates as well as effective long-term safety.[1-10] Consensus exists concerning the surgical treatment of symptomatic patients regardless of age. For patients with asymptomatic fistulas, however, the timing and indications for surgery are controversial. Various surgical techniques to deal with CCAF have been utilized, including internal cameral closure via CPB, tangential arteriography (either with or without bypass), distal ligation, ligation, and saphenous vein/ internal mammary artery bypass.[1-10]

In general, the fistulas that are short and intracameral along with those that are near a critical coronary supply or associated with an aneurysm or congenital heart disease are closed with the use of CPB.[1-7]

Internal closure in the right heart chamber (right atrium or pulmonary artery) can be performed using CPB without cross-clamping or cardioplegia on a beating or fibrillating heart.[8] Right ventricular fistula closure has been accomplished by tangential arteriography and by distal ligation of the feeding artery on the surface of the heart, often without CPB. A congenital coronary artery fistula to the left ventricle was closed by ligation during CBP.[1]

In contrast, coronary artery fistulas that are extracameral and anatomically accessible are usually controlled with ligature without CPB.

Catheter-based closure for CCAFs, if technically possible, has become the preferred treatment option. The successful use of percutaneous transcatheter closure devices has been described, and these are increasingly being used, especially in the pediatric population.[12,13] The most frequent complication associated with this type of procedure is the embolization of the occlusion device, which occured in seven of 40 patients in one series.[10]

Our patient was admitted to our hospital with progressive chest pain and dyspnea along with CCAF with a left-to-right shunt of 1.70 surgical ligation at beating heart discovered via CPB.

In summary, CCAFs are rare congenital malformations which should be treated as soon as the diagnosis is made to prevent complications and symptoms just like developed in our patient. The surgical treatment remains the gold standard with a weak morbidity and mortality even if some cases can benefit from percutaneous closing.

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