Treatment Strategies and Outcomes of Coronary Artery Fistulas in a Turkish Adult Population

Türk Toplumunda Erişkinlerde Koroner Arter Fistüllerinin Tedavi Stratejileri ve Sonuçları

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ABSTRACT Objective: Congenital coronary artery fistulas (CAF) are uncommon abnormalities that are known as abnormal communications between a coronary artery and a cardiac chamber or a major vessel. Incidence of CAF ranges between 0.1% and 0.2% 2 in literature,however, there is no detailed data on the incidence of CAF in a Turkish population. In this article, we evaluated the clinical manifestations, symptoms, chosen treatment strategies, and long term follow-up results of CAF in Turkish adult population. **Material and Methods:** In this multicenter study carried out between 2001 and 2009, a total of 25 400 angiograms which were taken for various reasons were evaluated retrospectively. Sex, age, clinical manifestations, symptoms, number of the fistulas, chosen treatment strategies, and long term follow up results of the study population were recorded and analyzed. **Results:** CAF were found in 32 of these cases (0.13%). Eighteen of the cases were males, and 14 were females with a median age of 57.6 years. Fifteen cases had surgery, graft-stent was applied to two cases and coil embolization was preferred in two cases. Thirteen cases were followed up under medical treatment. Graft stents were occluded after 4 and 6 months following implantation in left anterior descending artery in both cases, and hence, these patients required surgery. Mean follow-up period was 48 ± 27 months. All cases were asymptomatic (at the end of the follow-up period) and were doing well. **Conclusion:** In conclusion, incidence of CAF was found as 0.13% in the study population, and surgical treatment and coil embolization were considerably successful compared to stent graft application in CAF patients.

Key Words: Coronary vessel anomalies; coronary vessels; fistula

ÖZET Amaç: Doğumsal koroner arter fistülleri (KAF) bir koroner arterle bir kalp odacığı veya bir ana damar arasındaki anormal bağlantı olarak bilinen nadir görülen anomalilerdir. Literatürde KAF insidansı %0.1 ile 0.22 arasında değişir fakat Türk toplumunda erişkinlerde KAF ile ilgili detaylı veri yoktur. Bu makalede, Türk toplumundaki erişkinlerde KAF'ın klinik görünümlerini, belirtilerini, seçilen tedavi stratejilerini ve uzun dönem izlem sonuçlarını değerlendirdik. **Gereç ve Yöntemler:** 2001 ve 2009 yılları arasında yapılan bu çok merkezli çalışmada çeşitli nedenlerle çekilen toplam 25 400 anjiografi geriye dönük olarak değerlendirildi. Çalışma grubunun cinsiyet, yaş, klinik görünüm, belirti, fistül sayısı, seçilen tedavi stratejileri ve uzun dönem izlem sonuçları kaydedildi ve değerlendirildi. **Bulgular:** Bu olguların 32'sinde (%0.13) KAF bulundu. Olguların 18'i erkek, 14'ü kadındı, ortalama yaş 57.6 yıldı. Onbeş olgu ameliyat edildi, iki olguya greft-stent uygulandı ve iki olguda sarmal embolizasyon tercih edildi. Onüç olgu tıbbi izleme alındı. Her iki olguda implantasyondan sonra 4-6 ayda sol ön inen arterdeki greft stentler tıkandı ve bu nedenle hastaların ameliyat edilmesi gerekti. Ortalama izlem süresi 48 ± 27 aydı. Tüm hastalar asemptomatikti (izlem döneminin sonunda) ve durumları iyiydi. **Sonuç:** Sonuç olarak, çalışma popülasyonunda KAF insidansı %0.13 bulundu ve KAF hastalarında cerrahi tedavi ve sarmal embolizasyon stent greft uygulamasına göre daha başarılıydı.

Anahtar Kelimeler: Koroner damar anomalileri; koroner damarlar; fistül

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Coronary artery fistulas (CAFs) are quite rare termination abnormalities due to congenital or traumatic reasons. Most of CAFs are congenital without a gender predilection. The abnormal communication in CAFs occurs between coronary arteries and cardiac chambers, large vessels or other vascular structures. Most of the CAFs originate from the right coronary artery (RCA), while left coronary artery and circumflex artery are less frequently involved. CAFs frequently terminate in the right ventricle, right atrium, pulmonary artery, and less frequently, in the coronary sinus.[1] Coronary artery fistulas are coincidentally found when physicians examine cases with continuous murmurs, or causes of cases like acute myocardial infarction, congestive heart failure or endocarditis. On the other hand, diagnostic modalities of cardiovascular practice such as echocardiography or angiography, which are being performed intensely in the recent years, have increased the frequency of diagnosis of asymptomatic fistulas that could otherwise go undetected. Incidence of CAFs in angiographic series ranges between 0.1% and 0.2%.[2] Along with the increasing number of diagnostic coronary angiograms in our country, CAFs have become noticed, however, true incidence of CAF in the Turkish population has not been shown in a large scale trial.

In this study, diagnoses, treatments and follow-up of 32 cases with coronary artery fistulas out of 25 400 cases with available diagnostic coronary angiograms have been evaluated.

MATERIAL AND METHODS

In this multicenter study carried out between 2001 and 2009, a total of 25 400 angiograms in participating clinics were evaluated retrospectively. The current study was approved by local ethical committee. CAFs were found in 32 of these cases (0.13%). Eighteen of these cases were males and 14 were females, with ages ranging between 45 and 75 years (mean age: 57.6 years).

All positive cases were evaluated carefully by an experienced author in each center for the presence of CAFs (Figure 1). There was an accompanying coronary artery disease (CAD) in 12 (41%), mitral valve disease in 4 (12%), and coronary artery disease with coarctation of aorta in one (3%) of the cases with CAF.

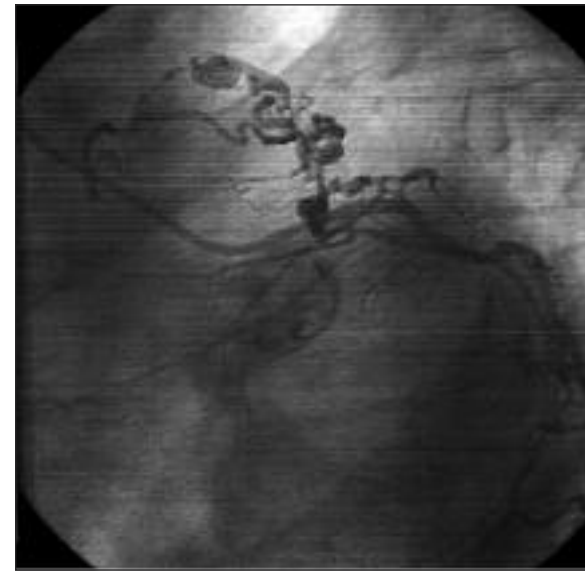

FIGURE 1: Arteriovenous fistula with aneurysmal dilatation between the left anterior descending coronary artery and pulmonary artery.

Surgery for the management of CAF was indicated due to the presence of symptoms of myocardial ischemia or cardiac insufficiency, presence of a fistula with hemodynamic significance (Qp/Qs ≥1.5).[1]

RESULTS

In our series, the mean age of the cases was 57.6 years. Twenty-one (65%) cases had angina pectoris, and six (19%) cases had shortness of breath. Congestive heart failure was present in three (9%) cases. In two (6.2%) of the cases, a systole-diastolic murmur characteristic for CAF was heard in auscultation. Twenty-eight (88%) of the cases had sinus rhythm, and four had mitral valve disease, and four (12%) had atrial fibrillation. Eight of the cases had electrocardiogram (ECG) signs of myocardial ischemia. Besides, twenty-one (65%) cases had chest pain.

Isolated CAF was found in 15 (47%) cases and accompanying pathologies together with CAF were found in 17 (53%) cases. There was one CAF in 29 cases and two fistulas in three cases. CAF was originated from left anterior descending artery

(LAD) in 15 (43%) cases, circumflex artery (Cx) in three (9%), RCA in 13 (37%), and diagonal artery in four (11%). CAFs drained into pulmonary artery in 22 (63%) cases, right ventricle in eight (23%), right atrium in four (11%), and the ascending aorta in one (3%) of the cases. Patient characteristics are presented in Table 1.

Fifteen cases had surgery, graft-stent was implanted in two cases and coil embolization was performed in two cases. Thirteen cases had medical follow-up. Surgery was performed through median sternotomy. CAF was epicardially ligated in all cases. Isolated epicardial fistula ligation was performed in two cases, both of whom had significant shunt (Figures 2 and 3). Of the other who had surgery cases coronary bypass surgery was performed in eight cases together with/ epicardial fistula ligation, cardiac valvular surgery in four and correction of coarctation of aorta together with coronary bypass surgery in one case. Cardiopulmonary bypass (CPB) was performed in 13 cases. In the three of the four with cases additional coronary artery diseases, coronary stent (two RCAs, one LAD) was implanted. No operative mortalities were observed. All of the patients were discharged without any problems. Control coronary angiography was performed in the early period in six cases; two of them had isolated CAF treated with stent grafts and the other two were treated with coil embolization, and no residual fistula was found. However, in two cases with graft stent, implanted in LAD, stents were found to be occluded in the control angiograms. These cases were referred to coronary bypass surgery. Long-term follow-

TABLE 1: Patient characteristics of the cases.

Age (year)	57.6
Gender n, (%)	
Male	18 (56)
Female	14 (44)
Symptoms/Findings n, (%)	
Hypertension	2 (6)
Chest pain	21 (65)
Shortness of Breath	6 (19)
Congestive Heart Failure	3 (9)
Origin n, (%)	
LAD	15 (43)
D	4 (11)
Cx	3 (9)
RCA	13 (37)
Termination n, (%)	
PA	22 (63)
RV	8 (23)
RA	4 (11)
Asc. Aorta	1 (3)
Accompanyingpathologies n, (%)	
CAD	12 (38)
MR	4 (12)
Coarct. of aorta	1 (3)

Cx: Circumflex coronary artery, D: Diagonal coronary artery, CAD: Coronary artery disease, LAD: Left anterior descending coronary artery, MR: Mitral regurgitation, PA: Pulmonary artery, RA: Right atrium, RCA: Right coronary artery, RV: Right ventricle.

FIGURE 2: Appearance of the arteriovenous fistula with aneurismal dilatation between the left anterior descending coronary artery and pulmonary artery.

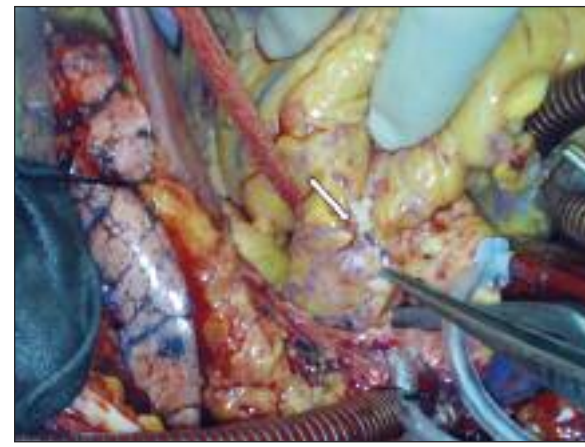

FIGURE 3: Epicardial ligation of the fistula between the left anterior descending coronary artery and pulmonary artery.

up was performed in 26 (81%) cases. Mean follow-up period was 48 ± 27 months. All cases were asymptomatic (at the end of the follow-up period) and were stable.

DISCUSSION

Coronary artery fistulas are very rare and encountered with an incidence of 0.002% in the general population. They are incidentally found during diagnostic tests, and diagnosis is not possible in most of the cases before death.[1] Advanced imaging technology enabled physicians to recognize asymptomatic fistulas which normally go undetected in the absence of cardiac surgery. The incidence of angiographically shown CAFs ranges between 0.3% and 0.8%.[3,4] CAF was diagnosed in 32 cases out of 25 400 angiography cases (0.13%) in our study. In this study, only two cases were admitted with complaints, possibly related to fistula.

Clinical course of CAF depends on many factors such as age, flow rate of the fistula, resistance at the terminal location or presence myocardial ischemia. Effort dyspnea, fatigue, angina pectoris, and atrial arrhythmias may be noted in symptomatic cases. In some cases, complications like myocardial infarction, heart failure, endocarditis, or peripheral embolism may be seen. Angina pectoris is rare in the absence of apparent coronary artery disease. In our series, half of the patients had accompanying pathologies. Classical finding of the physical examination in CAF is continuous soft murmur in crescendo-decrescendo style with a stronger systolic component, although it is heard both in systole and diastole. The prevalence of continuous murmur in confirmed cases with CAF is around 3%.[1,5] In our series, two cases with hemodynamic sequelae were diagnosed with physical examination (6.2 %). The size and flow rate of coronary fistulas tend to increase by aging, therefore early operation is recommended in those with auscultation findings.[1,6] The mean age of the cases was higher compared to that of other series in the literature.[5] Congenital CAF frequently arises from the right coronary artery and mostly drains into the right ventricle (45%), right atrium (25%), coronary sinus (7%), or pulmonary artery (15-20%).[7] While RCA is the most frequent origin with 55% in the literature, involvement of the left coronary artery is about 35%.[2,7,8] In our series, fistulas originated from the left coronary artery in 63% of cases, and from the right coronary in 37% of cases. In our series, pulmonary artery (63%) was the most frequent destination of CAF. Drainage into left atrium and left ventricle has been encountered very rarely.[7] Although coronary artery fistulas are solitary in general, multiple fistulas can also be noticed, though, very rarely.[1,2,7,8] There were multiple fistulas in 3 cases (9.3%) in our series.

There were no mortalities or morbidities associated with CAF in our series, similar to previous reports in literature.[5,8] There is no agreement in the literature regarding the surgical indication of asymptomatic CAF.[8] It has been suggested that asymptomatic cases without significant shunts should be followed up with regular control visits until symptoms appear or shunt becomes hemodynamically important.[8,9] It is suggested that surgical intervention provides an effective and rapid hemodynamic correction and protects the heart from volume overload, and it provides rather low postoperative recurrence rate. Some authors suggest that CAF should be closed particularly in younger cases, even when they are asymptomatic, in order to prevent angina, endocarditis, myocardial infarction, heart failure, development of aneurysms that might cause rupture or embolism, and development of pulmonary hypertension.[1,2,6] Thirteen (41%) of our cases had medical follow-up, surgical interventions were indicated in 19 (59%). All cases who had medical follow-up had small and nonsignificant shunts (Qp/Qs< 1.5). Surgery is justified in symptomatic cases with complications, in those with significant left-to-right shunt even if they were asymptomatic .[10]

Although CAF is generally closed with median sternotomy, minimal incisions without CPB are also used for closure. Operation technique varies according to the involved coronary artery, chamber of drainage and the localization of the connection. The basic procedure in closing CAF is the ligation of the fistula right on the point of entry to the cardiac chamber. This intervention prevents

development of myocardial ischemia. However, it ligation jeopardizes distal coronary flow, coronary bypass must be performed to ensure optimal flow.[5,10,11] In our cases, 13 of the surgical interventions were performed with CPB, two without using CPB, and coronary artery by-pass grafting (CABG) was performed in 9 cases. The postoperative course was uneventful.

CAF is closed successfully using trans-catheter methods in the last two decades.[9] These are coils, coils coated with stainless steel, separable balloons, and double-umbrella devices.[9] Selection of the occlusion method depends on the structure of the fistula. While coils are preferred for smaller fistulas, double-umbrella occlusion method is preferred for larger fistulas. This procedure protects cases from median sternotomy, CPB and from the disavantages of the surgical intervention. However, even rare, transcatheter method can cause arrhythmia, transient ischemic ECG changes, myocardial ischemia, and even death due to left coronary artery occlusion.[12-16] Ligation of CAF was preferred in cases who were referred to cardiac surgery due to accompanying pathologies. Graft-stent was implanted in two cases who refused surgery. However, graft stents were occluded after four and six months following implantation in LAD in both cases, and hence, these patients required CABG. Graft stents, which have quite high restenosis risk, are reserved only for cases with iatrogenic coronary artery perforations or giant aneurysms.[17] CAFs, closed with trans-catheter method or surgery, must be followed up for recurrences.[9]

Although medical treatment following closure is controversial, anti-aggregation treatment is recommended in a number of many articles since closure creates a chamber which can be source of thrombi.[12-15] We also recommended anti-aggregation therapy and bacterial endocarditis prophylaxis in cases who have medical follow-up.

CONCLUSION

In conclusion, while the surgical treatment and coil embolization of CAF were safe and successful, graft-stent did not seem to be an appropriate treatment choice. Ligation of CAF in patients, referred to cardiac surgery, seems safe without increasing the duration or complication risks of the surgery and might prevent hemodynamic problems driven by progression of CAF.

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