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Congenital coronary arteriovenous fistulas: an evaluation of 10 pediatric patients

Doğuştan koroner arteriyovenöz fistüller: 10 pediyatrik hastanın değerlendirilmesi

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ABSTRACT

Background: This study aims to assess the characteristics of congenital coronary arteriovenous fistulas (CAVFs) and discuss the timing of treatment.

Methods: Between January 2008 and December 2012, 10 pediatric patients with CAVF were diagnosed in our institution. Demographic characteristics, symptoms and clinical findings, electrocardiographic, echocardiographic, angiographic findings, clinical courses after treatment were evaluated retrospectively.

Results: Congenital coronary arteriovenous fistulas were originated from the left coronary artery in six patients, right coronary artery in three patients, and the left circumflex artery in one patient. The drainage site of most CAVFs was the right ventricle, as expected and drainage to pulmonary artery (2) and right atrium (1) were other common anatomical locations, respectively. We followed the patients for the degree of shunt and for ongoing enlargement of coronary arteries for six months. Proper intervention for surgery or transcatheter occlusion was based on individual basis. Surgical ligation was the preferred management in three patients, as the anatomic features of fistulas were not appropriate for a catheter-based intervention. A catheter-based attempt for interventional closure was unsuccessful in a patient and underwent surgical ligation.

Conclusion: Our study results suggest that it is important to diagnose CAVF in childhood due to the high risk of complications seen in adulthood, particularly, such as heart failure, myocardial ischemia, infective endocarditis and arrhythmias.

Keywords: Congenital; coronary arteriovenous fistula; echocardiography.

ÖZ

Amaç: Bu çalışmada doğuştan koroner arteriyovenöz fistüllerin (KAVF) özellikleri değerlendirildi ve tedavi zamanı tartışıldı.

Çalışma planı: Ocak 2008 - Aralık 2012 tarihleri arasında hastanemizde 10 pediyatrik hastaya KAVF tanısı konuldu. Demografik özellikler, semptomlar, klinik bulgular, elektrokardiyografi, ekokardiyografi, anjiyografi bulguları ve tedavi sonrası klinik seyir geriye dönük olarak değerlendirildi.

Bulgular: Doğuştan koroner arteriyovenöz fistüller altı hastada sol koroner arterden, üç hastada sağ koroner arterden ve bir hastada sol sirkümfleks arterden köken alıyordu. Çoğu KAVF'lerin drenaj yeri beklenildiği gibi sağ ventriküle olurken, sırasıyla 2 ve 1 olmak üzere pulmoner arter ve sağ atriyum sık olan diğer drenaj yerleriydi. Şant miktarının derecesi ve koroner arter genişlemesinin sürekliliği açısından hastalar altı ay süreyle takip edildi. Cerrahi veya transkateter kapatmaya dayalı uygun girişim hastaya göre seçildi. Üç hastada fistüllerin anatomik özelliklerinin kateter dayalı kapama için uygun olmamasından dolayı, tedavi seçeneği olarak cerrahi ligasyon tercih edildi. Kateter dayalı girişimsel kapama bir hastada başarılı olmadı ve cerrahi ligasyon yapıldı.

Sonuç: Çalışma bulgularımız, özellikle ileri yaşta kalp yetmezliği, miyokard iskemisi, infektif endokardit ve aritmi gibi komplikasyon riskinin yüksek olması nedeniyle, KAVF'nin çocukluk çağında teşhis edilmesinin önemli olduğunu göstermektedir.

Anahtar sözcükler: Doğuştan; koroner arteriyovenöz fistül; ekokardiyografi.

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A coronary arteriovenous fistula (CAVF) is a rare anomaly that is defined by an abnormal connection of the coronary arteries to a cardiac chamber, large blood vessels, or other vascular structures that bypass the myocardial capillary network.[1] It is the most common congenital anomaly of the coronary arteries. In addition, it affects hemodynamic parameters and accounts for between 0.3 and 0.8% of all congenital heart defects.[2] The vast majority of CAVFs arise from either the right coronary artery (RCA) or left coronary artery (LCA), whereas the circumflex artery (Cx) is rarely involved. Furthermore, in most cases, fistulous drainage habitually flows into the right-sided chambers, in particular the right ventricle.[3] For children with CAVFs, reducing the high risk of complications customarily seen in adulthood, including heart failure, myocardial ischemia, infective endocarditis, and arrhythmias, is of the utmost importance.

PATIENTS AND METHODS

This study was composed of 10 pediatric patients (6 boys, 4 girls; 4.85±4.0 years; range 3 days-14 years) with CAVFs were diagnosed at our institution between January 2008 and December 2012. Those with anomalous origin of the right or left coronary artery from the pulmonary artery were excluded from study. All of the patients were followed up regularly via electrocardiography and echocardiography, and two-dimensional (2D) echocardiographic and color Doppler images were obtained from the parasternal long and short axes, apical four-chamber, subcostal, and suprasternal views with variable frequency transducers using the Vivid™ S6 cardiovascular ultrasound system (GE Healthcare, Milwaukee, WI, USA). The coronary artery was considered to be dilated when its diameter was higher than +2 standard deviation (SD). The demographic characteristics, symptoms, clinical findings, electrocardiographic results, and cardiac imaging survey results [i.e., echocardiography and multislice computed tomography angiography (CTA) as well as catheter angiographic imaging modalities on an as needed basis] were evaluated retrospectively. The clinical course, including the intervention procedure, was also examined.

RESULTS

All of the patients were asymptomatic except for one who presented with chest pain and fatigue during exertion. A continuous murmur heard during a physical examination was the only clue regarding these symptoms. Chest radiography revealed no abnormal findings in the patients, and the electrocardiography results were normal in the asymptomatic patients. However, the electrocardiography did reveal that the patient with chest pain and fatigue had signs of myocardial ischemia. All of the patients were initially examined via echocardiography, and then catheter angiography was performed. The multislice CTA and cardiac magnetic resonance imaging (MRI) modalities are technically difficult to perform on pediatric patients and are more expensive. Hence, multislice CTA was performed on just two patients (Figure 1). Our institution prefers cardiac catheterization as the primary choice over other modalities since it is the gold standard for determining the exact fistula course and hemodynamic status. In addition, we also prefer to perform an adequate closure procedure in the same session. For all of our patients, the angiographic findings were highly consistent with the echocardiography results. The CAVFs originated from the LCA in six (60%) patients while there was RCA involvement in three others. Additional, we found one rare case of a CAVF originating from the left Cx artery. As expected, the drainage site was the right ventricle in six patients (60%). The CAVFs were also draining into the pulmonary artery in three others and into the right atrium in another. Coincidentally, one of the CAVFs was associated with coarctation of the aorta, and another occurred in conjunction with an atrial septal

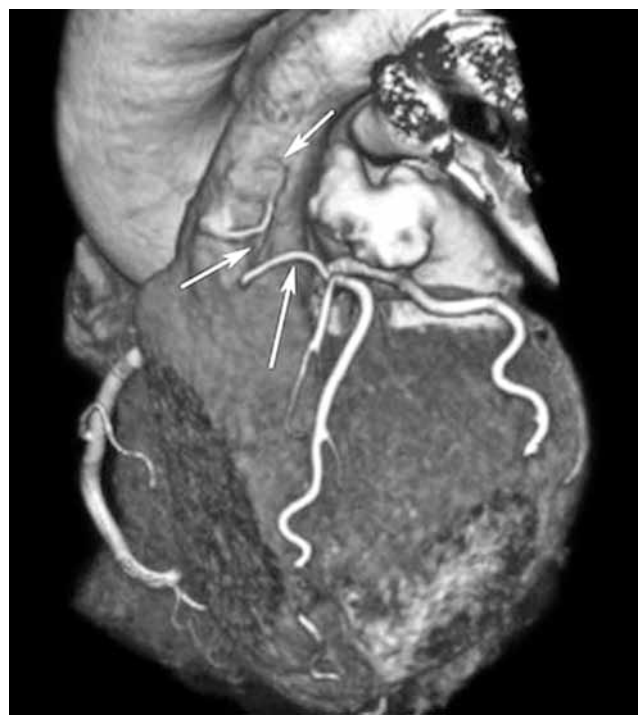

Figure 1. Three-dimensional, reconstructed, enhanced coronary angiographic image showing a small fistula (arrows) between the left anterior descending artery and the pulmonary artery (Patient no. 7).

defect (ASD). Furthermore, a single ostium coronary artery originating from the aorta was identified in two cases (Figure 2), and RCA agenesis was detected in one of these two patients (Figure 3). Unfortunately, the youngest patient died from neonatal sepsis, which was not connected to the CAVF. In three patients, surgical ligation of the CAVFs was the preferred treatment option because the anatomic features of the fistulas were not appropriate for catheter-based intervention. In patient number 10, the fistulous communications were closed with multiple pledgeted sutures, and the fistula was ligated via a right ventriculotomy with a beating heart and cardiopulmonary bypass (CPB) support. For patient numbers 4 and 6, the fistulas were ligated with a beating heart and CPB support. Unfortunately, in patient number 3, the catheter-based attempt at interventional closure was unsuccessful because we did not pass the guidewire from the left anterior descending coronary artery (LAD) branch at an acute angle forward into the right ventricle. Therefore, this patient also underwent surgical ligation with a beating heart and CPB support. The patients' demographic details along with the treatment options and follow-up results are summarized in Table 1.

DISCUSSION

A congenital CAVF is a rare anomaly because of the persistent presence of embryonic intratrabecular spaces and coronary sinusoids. In most cases, the fistulas occur in a single coronary artery and usually drain into the right side of the heart.[4] The majority of patients with CAVFs are asymptomatic at the time of diagnosis. The natural history and clinical course vary significantly and have not yet been clearly defined.[5]

There is a general consensus that symptomatic patients should be treated, although several reports have also demonstrated spontaneous closure.[6] Choosing between surgical intervention and transcatheter closure of the fistula is still tricky. However, trans-catheter closure seems reasonable in certain circumstances including fistulas with favorable anatomy (e.g. nontortuose vessel), the fistula in which distal portion is suitable for device closure, the fistula with narrowing distal end which avoid embolization, and the fistulas in which have no other indications for surgery.[6,7]

The ideal management strategy for patients with fistulas must be individualized.[8,9] Fistulas which originate from the proximal part of the coronary artery run the risk of a late giant aneurysm, dilation of the proximal coronary artery, and rupture. Therefore, in these cases, surgical or transcatheter closure seem reasonable at any age.[9,10] Transcatheter closure of fistulas has the advantage of less morbidity, a lower cost, and a shorter recovery time. In addition, there is no need for a thoracotomy and CPB.[9,11] In a report by Aydogan,[12] successful transcatheter embolization

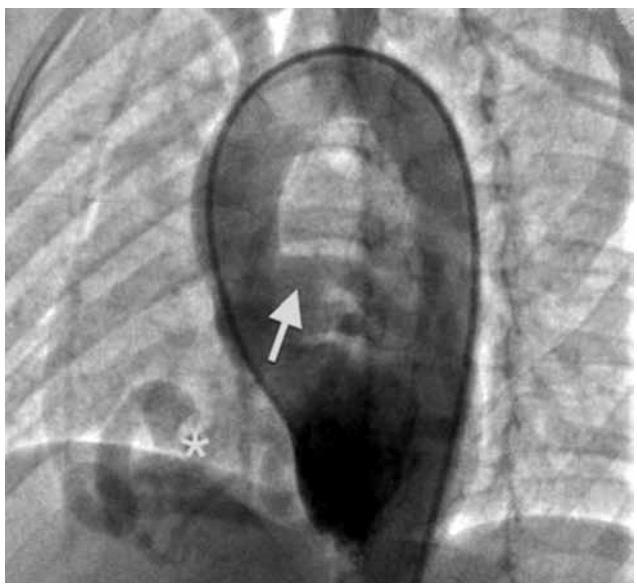

Figure 2. Angiographic image showing the single coronary ostium on the aorta (arrow) in the left anterior-oblique position and the drainage site of the fistula into the right ventricle (*) (Patient no. 3).

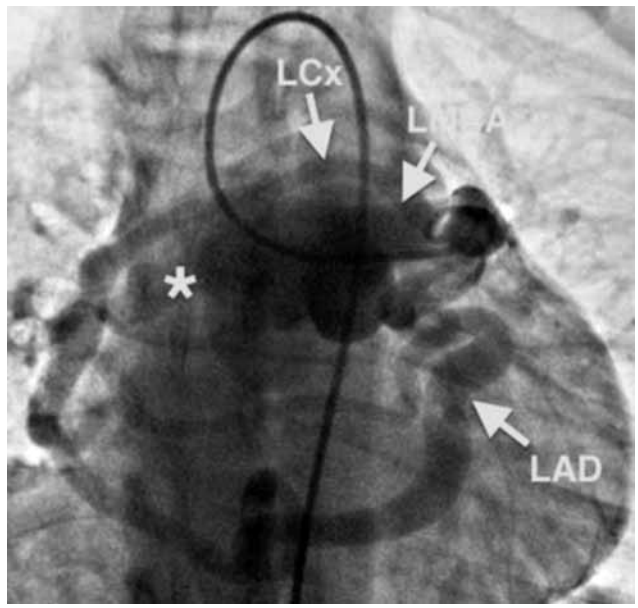

Figure 3. Angiographic image showing the dilated and tortuous left main coronary artery, left anterior descending artery, and left circumflex artery. The drainage site of the fistula was the superior margin of the right ventricle (*). The right coronary artery cannot be seen (Patient no. 10).

Table 1. Demographic characteristics, symptoms, diagnostic findings, treatment, and follow-up for 10 patients with coronary arterial venous fistulas

Patient no	Age	Gender	Symptom	Comorbid pathology	Qp/Qs	Tract of	Management	Follow-up fistula
1	3 days	Female	–	Coarctation of the aorta	1.0	LCA→RV	–	Exitus
2	13 months	Male	–	–	1.4	RCA→RV	–	Asymptomatic
3	18 months	Female	–	Persistent dilatation of the LCA	1.5	LCA→RV	Surgical ligation	Asymptomatic
4	3.5 years	Female	–	Single coronary ostium	1.88	LCA→RV	Surgical ligation	Asymptomatic
5	4 years	Female	–	–	1.0	Cx→PA	–	Asymptomatic
6	4 years	Male	–	Persistent dilatation of the RCA	-	RCA→RV	–	Lost to follow-up
7	6.5 years	Male	–	ASD; persistent dilatation of the RCA	2.0	RCA→RA	Surgical ligation	Lost to follow-up
8	7 years	Male	–	–	1.55	LCA→PA	–	Asymptomatic
9	7 years	Male	–	–	1.0	LCA→PA	–	Asymptomatic
10	14 years	Male	Exertional chest pain; fatigue	Persistent dilatation of the LCA; single coronary ostium; RCA agenesis	1.47	LCA→RV	Surgical ligation	ST depression in the inferolateral derivations

LCA: Left coronary artery; RV: Right ventricle; RCA: Right coronary artery; Cx: Circumflex artery; PA: Pulmonary artery; ASD: Atrial septal defect; RA: Right atrium.

(coil/balloon) was performed on five children with CAVFs, and Karagöz et al.[8,9] reported that they closed a CVAF with a ductal occluder in a symptomatic infant and used a vascular plug in a newborn baby who had been prenatally diagnosed with a large CAVF.

Furthermore, a review of 174 patients with CAVF revealed that patients should undergo percutaneous or surgical closure in childhood, even if they are asymptomatic and have a small fistula size with a low Qp/Qs ratio, because of serious complications that can occur in adults.[13] However, based on recent reports, the optimal management for asymptomatic patients with low Qp/Qs ratio is still not clear.[1]

An examination of case reports reveals the diverse spectrum of CAVFs. For example, in one report, a 46-year-old man presented with chest pain and a five-month history of palpitation, but no significant shunt was detected on angiography.[14] Moreover, a fifteen-month-old who presented with congestive heart failure and myocardial ischemia was diagnosed as having a CAVF. In our cohort, the only patient with any symptoms was a 14-year-old who presented with chest pain and fatigue during exertion. His past medical history was unremarkable. Electrocardiography revealed an ST segment depression in inferolateral derivations in this patient, and catheter angiography confirmed a fistula between the LCA and right ventricle. In addition, his Qp/Qs ratio of 1.47 was relatively low. Many reports have revealed that the majority of CAVF patients remain asymptomatic, especially during the first two decades of life,[5,13,15-17] whereas fewer have documented the occurrence of spontaneous closure.[6,18,19]

Consequently, when evaluating the relatively scarce number of patients in our study, we noted that a management strategy similar to the one used by Liang et al.[17] was employed. Their approach was to evaluate the symptomatic patients according to the degree of elevation of shunt magnitude and follow them up for six months to check for an enlarged persistency of dilatation of the coronary arteries. At that time, determined whether surgery or catheter-based occlusion is the best course of treatment is possible. With regard to asymptomatic patients, it might be prudent to closely follow them up via conservative methods until adulthood. Based on our findings, we suggest that multicenter, long-term, prospective studies be conducted to assess the optimal management strategy for patients with CAVFs.

Declaration of conflicting interests

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