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Coronary-cameral fistula between left anterior descending artery and right atrium in childhood: a case report

Çocukluk çağında sol ön inen arter ve sağ atriyum arasında koroner-kameral fistül: Olgu sunumu

Ünal Aydın, İsmihan Selen Onan, Onur Şen, Burak Onan, İhsan Bakır

Department of Cardiovascular Surgery, İstanbul Mehmet Akif Ersoy Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey

ABSTRACT

Coronary artery fistulas are abnormalities of artery termination. The fistulas involving the right heart chambers are more common than those involving the left heart chambers. The occurrence of a coronary-cameral fistula in pediatric patients is unusual. Patients are mostly asymptomatic and presenting symptoms may occur at older ages. The pathology is diagnosed by two-dimensional echocardiography, cardiac catheterization, and computed tomographic angiography. Early treatment is recommended due to the risk of congestive heart failure, endocarditis, myocardial ischemia/infarction, pulmonary hypertension, and coronary aneurysm formation with subsequent ruptures or embolization. Herein, we present an 11-year-old girl in whom a giant coronary artery fistula localized between the left anterior descending artery and the right atrium was successfully treated.

Keywords: Childhood; coronary-cameral fistula; left anterior descending artery; right atrium.

ÖZ

Koroner arter fistülleri koroner arterlerin sonlanma anormallikleridir. Sağ kalp boşluklarını tutan fistüller, sol kalp boşluklarını tutan fistüllerden daha sık görülmektedir. Koroner-kameral fistül oluşumu pediatrik hastalarda nadirdir. Hastalar genellikle asemptomatiktir ve eşlik eden semptomlar ileri yaşlarda görülebilir. Patoloji iki boyutlu ekokardiyografi, kalp kateterizasyonu ve bilgisayarlı tomografi ile tanılanır. Konjestif kalp yetersizliği, endokardit, miyokard iskemisi/enfarktüsü, pulmoner hipertansiyon ile koroner anevrizmaya sekonder rüptür ve embolizasyon riski nedeniyle, erken tedavi önerilmektedir. Bu yazıda, sol ön inen arter ve sağ atriyum arasında yerleşimli dev koroner arter fistülü olan ve cerrahi ile başarılı bir şekilde tedavi edilen 11 yaşında bir kız çocuğu sunuldu.

Anahtar sözcükler: Çocukluk çağı; koroner-kameral fistül; sol ön inen arter; sağ atriyum.

Coronary-cameral fistulas (CCFs) are defined as abnormalities of coronary artery termination.[1-3] The communication occurs between any coronary artery and chambers of the heart. Other termination centers are superior vena cava, coronary sinus, pulmonary artery and pulmonary veins. In general, CCFs are diagnosed incidentally during diagnostic coronary artery catheterization. The occurrence of a large CCF is rare in adult and pediatric patients.[1,2] The majority of patients are asymptomatic and presenting symptoms may occur at older ages.[1-3] In this article, we report a girl in whom a giant CCF localized between the left anterior descending artery and the right atrium was successfully treated.

CASE REPORT

An 11-year-old girl was referred for further evaluation of a continuous parasternal murmur over the fourth intercostal space. She was asymptomatic during daily physical activities. The murmur was incidentally diagnosed during physical examination for an upper respiratory tract infection. Cardiac auscultation showed a hyperkinetic precordium and a continuous murmur over the left parasternal area. The electrocardiogram

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Correspondence: Burak Onan, MD. İstanbul Mehmet Akif Ersoy Göğüs Kalp ve Damar Cerrahisi Eğitim ve Araştırma Hastanesi, Kalp ve Damar Cerrahisi Kliniği, 34303 Küçükçekmece, İstanbul, Turkey.
Tel: +90 553 - 622 38 78 e-mail: burakonan@hotmail.com

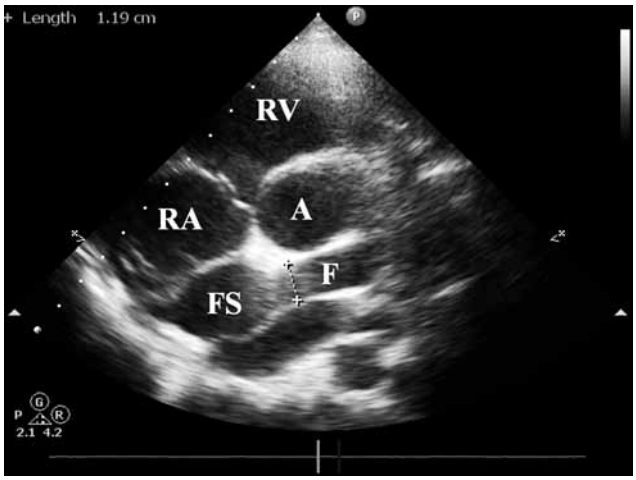

Figure 1. A parasternal short axis view of transthoracic echocardiography showing a coronary fistula draining into the right atrial cavity through a fistula tract.
RV: Right ventricle; RA: Right atrium; A: Aorta; F: Fistula tract; FS: Fistula sac.

showed sinus rhythm and right axis deviation. A chest X-ray revealed an increased cardiothoracic index and pulmonary vasculature. Transthoracic echocardiography showed a CCF originating from the left coronary artery draining into the right atrium through an intra-atrial sac (Figure 1). The fistula tract was 18 mm in its maximal diameter. Fractional shortening of the left ventricle was 34%. There was mild aortic valve regurgitation. Left cardiac chambers were dilated with a left ventricular end-diastolic diameter of 52 mm and end-systolic diameter of 34 mm. Cardiac catheterization confirmed a CCF from the proximal segment of the left anterior descending artery draining into the right atrium through a sac (Figure 2a). Qp/Qs ratio was 1.6. Once transcatheter coil embolization failed to close the fistula tract, surgical treatment was recommended.

The operation was performed using cardiopulmonary bypass at moderate hypothermia. After cardiac arrest and right atriotomy incision, intraatrial cavity was explored to find the distal end of the fistula (Figure 3). The fistula sac was observed on the anterolateral wall of the right atrium and it was opened through a more lateral incision on the right atrial wall. This was the second atriotomy incision. The fistula sac was internally opened and a common right atrial chamber was formed. Then, the proximal neck of the fistula tract was ligated internally. The distal outflow of the fistula sac was ligated through the right atrium. The operation was completed uneventfully.

No residual shunt was observed during intraoperative transesophageal echocardiography. Postoperative period was uneventful without myocardial ischemia or infarction. During follow-up, there was no residual fistula tract on cardiac catheterization (Figure 2b). The patient was discharged in the sixth postoperative day. Her clinical condition is good at three years during follow-up.

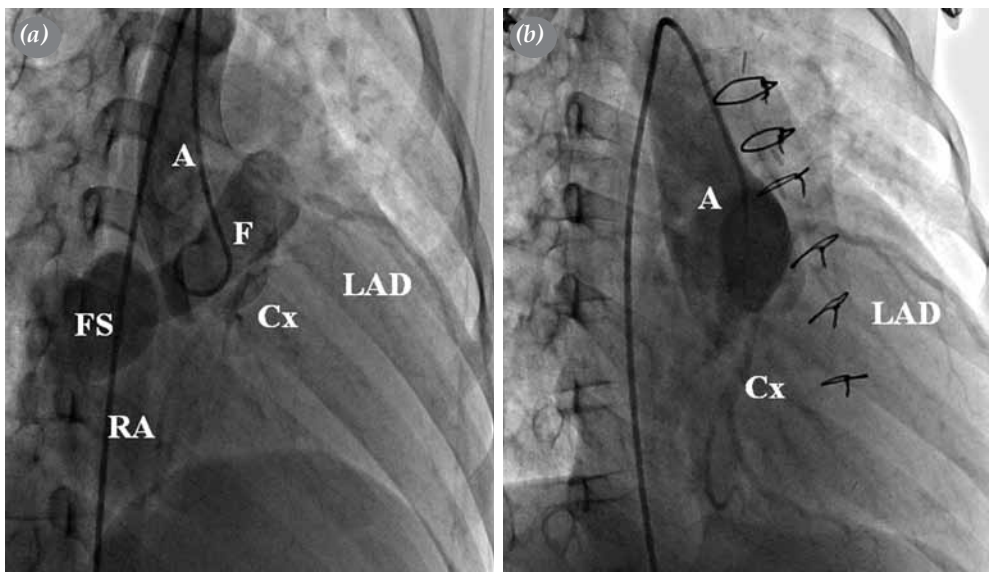

Figure 2. (a) Catheterization of coronary artery fistula showing a fistula between the left coronary system and right atrium. Fistula tract was draining a sac within the right atrial cavity. **(b)** Postoperative coronary angiogram showing no evidence for residual fistula tract after operation.
A: Aorta; F: Fistula tract; FS: Fistula sac; Cx: Circumflex coronary artery; LAD: Left anterior descending artery; RA: Right atrium.

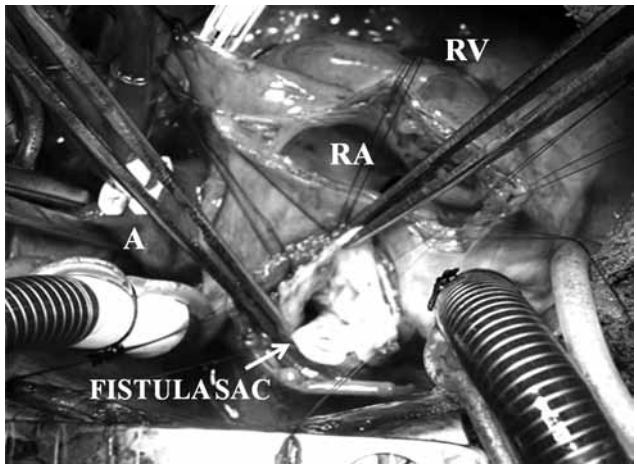

Figure 3. A surgical view showing the fistula sac within the right atrial cavity.

RV: Right ventricle; RA: Right atrium; A: Aorta.

DISCUSSION

Coronary-cameral fistula is an abnormal termination of coronary arteries.[1-8] This type of abnormal communication arising from a coronary artery and draining into one of the cardiac chambers is termed as a CCF, rather than a coronary artery fistula. These CCFs usually arise from the RCA and commonly draining into right-sided chambers. Origination of the fistula from the left coronary system is less frequent. In addition, CCFs are present in 0.002% of the overall population and represents 0.4% of all cardiac malformations.[3] It has been reported that 40 to 55% of cases with CCF are asymptomatic.[4] The number of symptomatic patients tends to increase after the age of 20 years. However, the incidence of CCF in pediatric cases is unknown.

Children are mostly asymptomatic and presenting symptoms may usually develop at older ages.[1,2] Patients typically present with dyspnea, easy fatigability, angina pectoris, and other non-localizing complaints. These symptoms are due to a left-to-right shunt or ischemia related to coronary steal phenomenon.[1-8] The degree of shunting determines the degree of clinical symptoms and the age of presentation. In large fistulas, congestive heart failure, myocardial ischemia, rupture of dilated coronaries, hemopericardium or peripheral embolization may also develop. From the clinical perspective, the presence of a continuous murmur at the right parasternal area and radiologic evidence of increased pulmonary vasculature in acyanotic children may suggest a cardiac fistula. The differential diagnosis includes patent ductus arteriosus, ruptured sinus of Valsalva, ventricular septal defect, and aortopulmonary window.[1-8] Electrocardiography, coronary artery imaging, and echocardiography are helpful modalities for the definite diagnosis.

The closure of CCFs is recommended in both symptomatic and asymptomatic patients.[1-8] Fistulas can be closed with percutaneous techniques using detachable balloons, platinum micro-coils or steel coils.[3,8] Factors favoring the success of transcatheter closure include the ability to cannulate the feeding branch adequately and safely, and the presence of a single, narrow restrictive drainage site. Multiple communications between the fistula and coronaries and the presence of large branches may decrease the success of transcatheter closure of CCF.[3,8] The diameter of the fistula tract determines the feasibility of closure using percutaneous techniques. Otherwise, surgical closure of large CCFs is recommended to avoid cardiac complications.[1-8]

Currently, surgical closure techniques include ligation of the fistula alone (with or without cardiopulmonary bypass) or combined coronary artery bypass grafting after ligation.[1,3,5-7] In addition, thoracotomy or median sternotomy incisions can be used. The goal is the closure of the inlet and outlet tract of the fistula and division of the communication between the coronary artery and the terminal outflow area. Anatomically, CCFs often present with two different characteristics: In the first type, coronary segment proximal to the origin of the fistula is dilated, but the distal segment is normal. These fistulas can be closed by epicardial ligation. Ligation site can be the origin of the fistula or both proximal and distal coronary segment around the origin of fistula tract. In the latter case, coronary bypass grafting is necessary after ligation. In the second type of presentation, coronary artery is dilated and fistula terminates in the right or left chambers of the heart. In these cases, CCFs are ligated by intracardiac purse-string sutures at the site of termination with cardiopulmonary bypass.[4,5,7] Although surgical closure is simple in most cases, surgeon should determine the precise location and tract of the fistula before the operation. The clear mental image of the fistula should be established before operation. From the surgical view, fistulas can be first exposed macroscopically during operation and are simply ligated. Second, arteriotomy is performed to expose the origin of the fistula and an intraarterial ligation of the origin site is made. Third, termination site is exposed within the cardiac chambers or great vessels and fistula is closed internally.

In the repair of large CCFs, the only disadvantage may be seen during delivery of cardioplegia solution.[7] As the fistula is giant in size, an excessive left-to-right shunt may cause coronary ischemia and infarction of the distal coronary system due to steal phenomenon during cardioplegia delivery.[7] To avoid this complication, total cardiopulmonary bypass was initially established and, then, the right atrium was opened and fistula outlet was immediately closed during the delivery of cardioplegia solution in our case. This allowed the proper delivery of cardioplegia to the left coronary system, particularly to the left ventricle. Alternatively, retrograde delivery of cardioplegia can be a safer alternative in certain cases.

In conclusion, coronary-cameral fistulas are rarely diagnosed in clinical practice, particularly in childhood. The closure is recommended to prevent endocarditis, coronary ischemia, and associated myocardial dysfunction. Of note, surgical closure is needed in the management of large coronary-cameral fistulas.

Declaration of conflicting interests

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