Right Coronary Artery Fistula Draining into the Right Atrium and Associated with Mitral Valve Stenosis: A Case Report

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ABSTRACT

Coronary artery fistula (CAF) is a rare congenital anomaly of the coronary arteries in which abnormal connections are present between the coronary artery branch and the cardiac chambers or a major vessel. The incidence of CAF is estimated at 1 in 50,000 live births, and it is detected in approximately 0.2% of the adult population during coronary angiography. Reports of the coincidence of mitral stenosis and CAF are rare in the literature. We report a case of CAF and mitral valve stenosis in a patient with dyspnea and fatigue before valve replacement and surgical radiofrequency ablation. Coronary angiography showed a connection between the right coronary artery and right atrium. A fistula opening into the right atrium is rare in patients with coronary artery anomalies and mitral valve disease. Coronary angiography of the patient 1 month after surgical repair showed that the coronary anatomy was normal and the fistula was occluded. CAF can be diagnosed more frequently if coronary angiography is performed simultaneously with cardiac catheterization to evaluate valve functions or nonatherosclerotic myocardial ischemia in each valvular heart disease case. Surgical repair of CAF is the first-choice treatment to prevent complications and improve quality of life.

INTRODUCTION

Coronary arterial fistula (CAF), first defined by Krause and Trevor in 1865, is an abnormal connection between a coronary artery and a cardiac chamber or major vessel (eg, vena cava, pulmonary vein, or pulmonary artery) [Luo 2006]. This rare congenital anomaly of the coronary arteries is usually discovered by chance during coronary arteriography, but these fistulas can cause serious coronary morbidity with increased mortality rates and can lead to angina, syncope, congestive heart failure, myocardial infarction, and sudden death [Gunduz 2003]. The association of mitral valve disease

with CAF is rare. In most cases, CAF is either asymptomatic or presents with angina-like symptoms, but symptoms and physical examination findings may be masked when CAF is associated with a valvular disease [Kayacıoglu 2002]. A fistula opening into the right atrium (RA), which occurred in our patient, has not been reported in other cases of CAF with coronary artery anomalies and mitral valve disease [McClung 1996; Kayacıoglu 2002]. We report successful treatment of a patient with mitral valve stenosis accompanied by a right coronary artery (RCA) fistula draining into the RA, a left atrial (LA) thrombus, and atrial fibrillation.

CASE REPORT

Our patient was a 55-year-old woman who was followed for mitral stenosis (MS) for 18 years and had suffered rheumatic fever 25 years previously. She was examined at our cardiology clinic for dyspnea and increasing fatigue. Her electrocardiogram (ECG) showed atrial fibrillation and LA dilatation. Chest x-ray revealed cardiomegaly, hilar congestion, and flattening of the pulmonary conus. Transthoracic echocardiography showed MS (mitral valve area, 0.8 cm^2), and the pressure gradient at the mitral valve was maximum at 18 mm Hg, with a mean of 12 mm Hg. The mitral valve leaflets were thick and calcific and their opening movement was extremely limited. The diameters of the LA (61 mm) and left ventricle (LV) (50/32 mm) were enlarged. The ejection fraction (EF) was 60%. Second-degree tricuspid valve insufficiency and pulmonary hypertension (pulmonary artery pressure [PAP], 55-60 mm Hg) were accompanied by hyperechogenic appearance correlated with an LA thrombus. Because the patient was middle aged, coronary arteriography (CAG) and cardiac catheterization were performed. RA, right ventricle (RV), PAP, LV, and aorta pressures were 9 mm Hg, 50/8 mm Hg, 55/32 mm Hg, 110/8 mm Hg, and 120/85 mm Hg, respectively. EF was confirmed to be 60%. CAG showed a CAF draining from the RCA into the RA (Figure 1). Coronary arteries were normal. The Qp:Qs ratio was 1:3. In light of these findings, the patient underwent surgery.

Surgical Technique

The patient underwent surgery while in a supine position under endotracheal general anesthesia. After the median
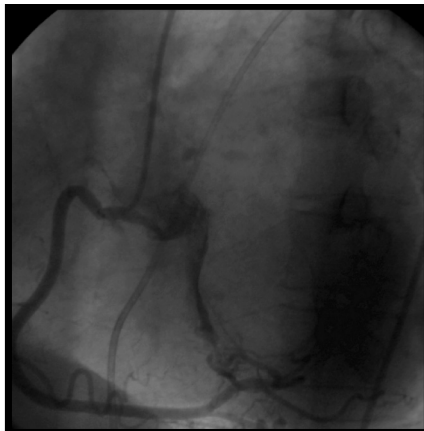
Figure 1. Angiographic image of the coronary fistula draining from the right coronary artery to the right atrium.

sternotomy, pericardial layers were opened longitudinally. Because the thrill originated from the mitral valve pathology, no fistula thrill could be palpated via external palpation of the heart. Cardiopulmonary bypass (CPB) was started with aortic and bicaval venous cannulation after heparinization. After the aorta was cross-clamped, the heart was arrested by intermittent antegrade isothermic blood cardioplegia. Moderate hypothermia was achieved (28°C). A vent was placed via the right superior pulmonary vein. Standard left atriotomy was made from the interatrial junction. An abandoned organized thrombus was evacuated from the LA (Figure 2). The mitral valve was severely calcified and stenosed, forming a block. Subvalvular structures were not convenient for repair. The native valve was resected and the posterior leaflet was preserved. Our standard radiofrequency ablation technique was performed with a Cardioblate BP (bipolar) surgical ablation device (Medtronic 60821; Medtronic, Minneapolis, MN, USA) (Figure 3). This procedure prolonged the normal cross-clamp duration by 5 minutes. Surgical closure of the left auricle was performed internally from the LA with primary sutures of 3/0 monofilament polypropylene. Subsequently, a right atriotomy was performed. The tricuspid valve was competent. With the help of cardioplegy via the aortic root, the fistula orifice on the coronary sinus was located and ligated with transfiction sutures (Figure 4). The mitral valve replacement procedure was performed by the insertion of a 29-mm Sorin bileaflet mechanical prosthetic valve (Sorin Biomedica, Via Crescentino, Italy) using a series of Teflon pledget 2/0 ti-cron U-sutures. The left atriotomy was closed according to the standard procedure. RA and RV permanent pacemaker electrodes were placed. Postoperative rhythm was sinusal. There were no signs of ischemia in daily ECG during the postoperative period. The patient was discharged on postoperative day 7 and considered cured. After surgical repair of the fistula, exercise ECG showed no abnormality, and no fistulas were detected via CAG (Figure 5). Transthoracic echocardiography showed normal function of the prosthetic mitral valve, and ECG showed sinus rhythm.

Prophylactic amiodarone treatment was continued for 6 months, and no complications emerged during the patient's follow-up in our outpatient clinic.

DISCUSSION

Congenital arteriovenous or arterioarterial fistulas of the coronary arteries are direct connections between a branch of the coronary artery and the lumen of 1 of the 4 cardiac chambers, the coronary sinus, the superior vena cava, the pulmonary artery, or the pulmonary veins without the intervening capillary system [Enç 1999]. Generally, these fistulas are small, and their incidence rate is 0.2% in the adult population [Tanriverdi 2006]. CAF incidence hovers between 0.08% and 0.3% in angiographic studies, and their presence is considered clinically insignificant. CAF are usually of congenital origin, making up 0.27% to 0.40% of all congenital cardiac defects [Shyam Sunder 1997], but may also appear as a result of chest trauma or iatrogenic procedures. Patients may be asymptomatic or show signs and symptoms of myocardial ischemia [Tirabassi 2006]. Clinical presentations are variable depending on the type of fistula, shunt volume, site of the shunt, and presence of other cardiac conditions [Luo 2006]. The Hobbs et al series included the greatest number of patients, whose fistulas were classified according to their origin [Yamanaka 1990; Kayacıoglu 2002]. In this patient series, 50% of fistulas were RCA, 42% were left coronary artery (LCA), and 19% were RCA + LCA. Moreover, the arrangement of the fistulas according to their openings were as follows: 41% RV, 26% RA, 17% PA, 5% LA, 3% LV, 7% coronary sinus, and 1% VCS. According to Fernandes et al, 73% of isolated coronary fistulas are asymptomatic [Fernandes 1992; Enç 1999]. Rates for congestive heart failure symptoms, myocardial infarction, rupture, and bacterial endocarditis were 12%, 4%, 1%, and 3%, respectively, in a study conducted with 174 patients [Liberthson 1979; Kimura 2006]. In this study, the rate of symptomatic patients was 30%. The same study showed that the rate of symptomatic patients was 9% in patients younger than 20 years and 55% in patients older than
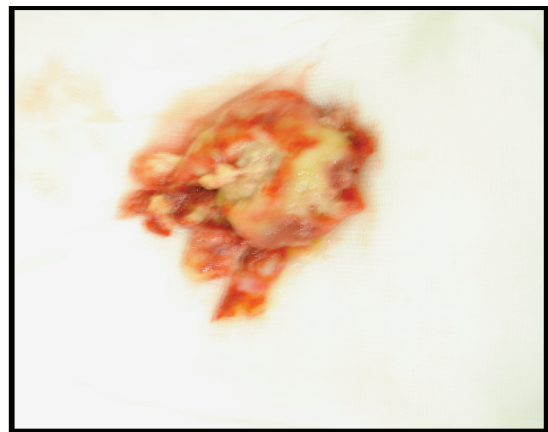

Figure 2. Organized thrombus removed from the cavity after the left atriotomy.
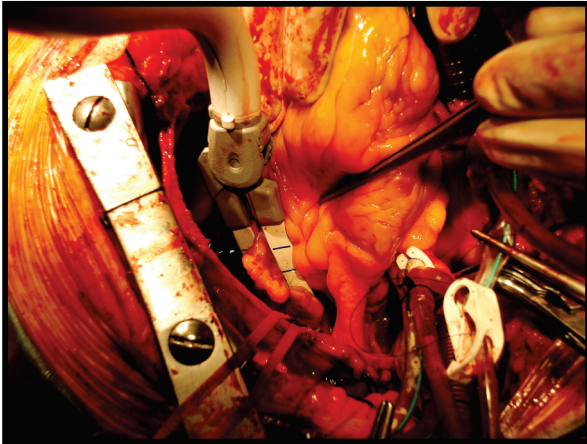
Figure 3. The application of radiofrequency ablation to the left auricle.

20 years. Bacterial endocarditis risk is high if fistula flow is high. Thus, antibiotic prophylaxis may be necessary [Kayacıoglu 2002].

The most frequent symptom of CAF is continuous murmur at the left parasternal region extending to the apex. Enç et al [1999] reported nonspecific ECG results in 54% of the cases. Cardiomegaly and increased pulmonary vascularity are present in 50% of cases. Cardiac catheterization and coronary angiography are still widely accepted diagnostic methods because they define the precise anatomy and reveal additional anomalies [Enç 1999; Kayacıoglu 2002; Huang 2006; Tanriverdi 2006].

Approximately 10 CAF cases with additional valve disease have been reported. In 2 of these, patients had aortic valve stenosis (AS) [McClung 1996; Kayacıoglu 2002]; there was a connection between the PA and both coronary artery systems in one patient and between the PA and RCA-RA fistula in the other. The remaining 8 reported cases were MS patients. Among these patients, MS was detected between the LCA and LA in 2 patients, between the LCA and PA in 1 patient, between the RCA and PA in 1 patient, and between both coronary systems and PA in the remaining 4 patients [Ayala 1996; Kayacıoglu 2002]. Kayacıoglu et al [2002] reported an RCA-RV case with MS for the first time. Our case is the first reported RCA-RA fistula diagnosed in an MS patient.

CAF occlusion is an event rarely reported in the literature. Among all cases, the spontaneous occlusion rate for CAF hovers between 1% and 2%. The most effective factor for spontaneous occlusion is the diameter of the fistula and flow rate [Shyam Sunder 1997]. The first-choice treatment for CAF occlusion is surgery [Liberthson 1979; Tengiz 2002]. For CAF without occlusion, which is generally asymptomatic, there are 2 treatment alternatives [Kayacıoglu 2002], medical follow-up without treatment unless hemodynamic problems occur and symptoms appear or elective fistula ligation before the patient reaches 20 years of age, without taking into consideration whether symptoms appear or not. The aim of this procedure is preventing the development of fistula-dependent complications. If there is a moderate or high outflow of shunt or the patient is symptomatic, regardless of the age of the patient, surgery is recommended. Surgery can be performed without CPB if the chamber into which the fistula drains will not be exposed. Some surgeons recommend CPB for all cases in order to avoid ligation from the wrong artery and to definitely occlude the fistula [Enç 1999]. Enç et al studied approximately 9 cases in 10 years and performed ligation from RA with CPB in a case with CAF between the RCA and RA [Enç 1999].

Recently, new methods have been developed to occlude the CAF by transcatheter intervention [Tengiz 2002; Spektor 2006]. These methods include coil embolization, intravascular embolization with chemical materials, and balloon occlusions. There are no reported studies that cover the long-term results obtained through transcatheter occlusion interventions in detail. Although transcatheter occlusion interventions are less invasive, they cannot be used in all cases. Coil embolization of the largest fistula was not found to be an appropriate technique for management [Tengiz 2002]. Transcatheter closure of CAF with coils is well described. The use of newer devices may offer advantages such as improved control of device placement, the use of a single device instead of multiple devices, and high rates of occlusion [Behera 2006]. Okubo et al [2001] performed a study on 13 patients and successfully occluded the lesions in 9 patients. However, during the angiographic examination of these 9 patients, which was performed 14 months later, hemodynamically insignificant residual shunts were found in 4 patients.

Atrial fibrillation is the most frequent rhythm disorder in the general population, with an overall incidence rate of 1% that reaches 4% in patients older than 60 years. The rate is 50% in patients who have undergone mitral valve surgery [Vaturi 2001]. In those patients with previous atrial fibrillation, the persistence of atrial fibrillation after mitral valve surgery may hinder the satisfactory functioning of the heart and cause thromboembolic events. Radiofrequency ablation during mitral valve surgery is a simple technique that has shown satisfactory early results in a study cohort in which
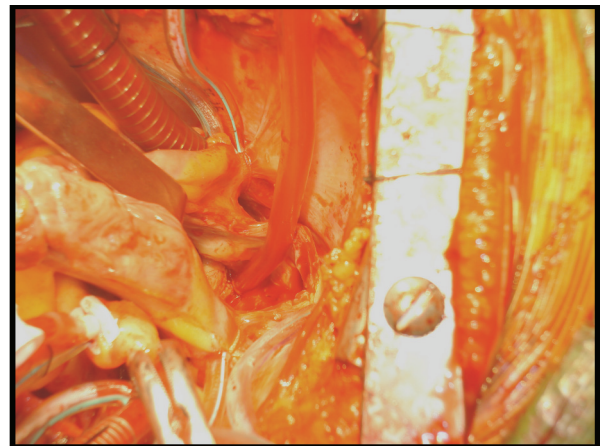

Figure 4. Coronary artery fistula in the right atrium with its opening orifice located over the coronary sinus.
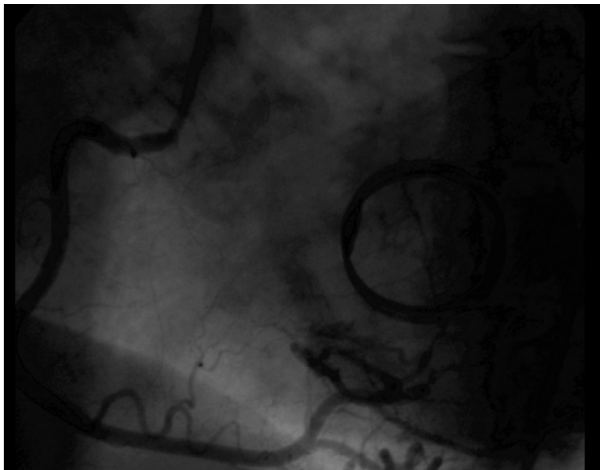
Figure 5. The image obtained via coronary angiography at the end of the first postoperative month shows that the fistula, which was previously draining from the right coronary artery to the right atrium, is occluded.

sinus rhythm was maintained in 88.9% of the patients during a 1-year follow-up period [Demirkılıç 2003].

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