# Coronary Artery Fistula within Large Left Atrial Thrombus in a Patient with Rheumatic Mitral Stenosis: A Case Report and Review

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## ABSTRACT

Coronary artery fistulae, which are connections between coronary artery branches and cardiac chambers or major vessels, are usually congenital forms of coronary artery anomalies, but rarely may be acquired because of cardiac trauma, neovascularity in association with cardiac neoplasm, mural thrombus formation, or iatrogenic causes. Although half of the patients with coronary artery fistula remain asymptomatic, these fistulae may lead to angina, syncope, congestive heart failure, myocardial infarction, and sudden death. Here we present a case of coronary artery fistula between right coronary artery and left atrium through a large left atrial thrombus in a patient with rheumatic mitral stenosis. The diagnosis was confirmed using echocardiography, conventional and multislice computed tomography, and coronary angiography. To the best of our knowledge, this interesting and unique entity is the first to be reported in the present literature.

**Key Words:** Coronary artery fistula; left atrial thrombus; rheumatic mitral stenosis

### Romatizmal Mitral Stenozu Olan Hastada Büyük Sol Atriyal Trombüs İçine Doğru Seyreden Koroner Arter Fistülü

## ÖZET

Koroner arter dalları ile kardiyak boşluklar veya büyük damarlar arasındaki normal dışı bağlantılar olan koroner arter fistülleri (KAF) genellikle koroner arter anomalilerinin konjenital formlarıdır. Fakat nadiren kardiyak travma, iyatrojenik nedenler veya kardiyak neoplazm ve mural trombüs oluşumu ile ilişkili neovaskülarizasyona ikincil edinsel olabilir. KAF'ı olan hastaların yaklaşık yarısının asemptomatik kalmasına rağmen bu fistüller anjina, senkop, konjestif kalp yetmezliği, miyokart infarktüsü ve ani ölüme yol açabilirler. Bu yazıda romatizmal mitral kapak stenozu olan bir hastada sağ koroner arter (RCA) ile sol atriyum (LA) arasında, büyük LA trombüs içine açılan KAF'ı sunuyoruz. Tanı 2 boyutlu ekokardiyografi, konvansiyonel koroner anjiyografi ve çok kesitli bilgisayarlı tomografik koroner anjiyografi kullanılarak doğrulandı. Bildiğimiz kadarı ile bu ilginç ve benzersiz durum mevcut literatürde ilk defa raporlanmıştır.

**Anahtar Kelimeler:** Koroner arter fistülü; sol atriyal trombüs; romatizmal mitral kapak darlığı

## INTRODUCTION

Coronary artery fistulae (CAF) are abnormal connections between coranary arteries and cardiac chambers, great vessels (vena cava, and pulmonary vein and artery) or other vascular structures (mediastinal vessels and coronary sinus). CAF are usually of congenital origin and are not gender specific; however, they sometimes occur secondary to cardiac surgery, cardiac invasive procedures, chest trauma, neovascularity in association with cardiac neoplasm, and formation of mural thrombus[1-4]. Although half of the patients with CAF remain asymptomatic, these fistulae are important causes of coronary morbidity and mortality leading to syncope, congestive heart failure, infective endocarditis, myocardial infarction, and sudden death[3]. They may be seen at any age, but some studies have emphasized that the incidence of symptoms and complications increases with age, particularly after the age of 20 [5,6]. This rare anomaly of coronary arteries is usually incidentally diagnosed using coronary angiography. CAF arise from right coronary artery (RCA) or its branches in approximately half of the cases, whereas in the remaining cases, they arise from left anterior descending (LAD) and circumflex (CX) coronary arteries or their branches. Drainage most frequently occurs into the right side of the heart or pulmonary artery[5]. The association of rheumatic mitral stenosis (MS) with CAF has rarely been reported. To our knowledge, our study is

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the first to report a fistula formation that originated from RCA branches and drainaged through a large left atrial thrombus into the left atrium in a patient with rheumatic MS.

## CASE REPORT

A 57-year-old female was admitted to our outpatient clinic with exertional dyspnea and chest pain. These symptoms had increased during the last month prior to admission. She had a history of hypertension and atrial fibrillation (AF). At physical examination, blood pressure was 135/85 mmHg, heart rate was 96 beats/min and arrhythmic, and body temperature was 36.6°C. Cardiac auscultation revealed an opening snap, soft diastolic (murmur) rulman at the apex, and a loud mid-systolic murmur was heard along the left sternal border.

Electrocardiography revealed AF, left ventricular hypertrophy, and ST segment depression on V5-V6. Transthorasic echocardiography (TTE) revealed rheumatic mitral valve with moderate to severe stenosis and mild mitral regurgitation (Figure 1). All cardiac chambers were enlarged with the left ventricular ejection fraction of 35%. Transesophageal echocardiography (TEE) revealed a large mural thrombus formation and spontaneous echo-contrast images in the left atrium (LA). The patient underwent coronary angiography prior to surgery, which revealed normal LAD and CX coronary arteries. The conus and posterolateral branches of RCA was fistulized to mural thrombus adherent to the LA wall (Figure 2). RCA, CX, and LAD did not contain any atherosclerotic lesion. A 64-slice multidetector computed tomography (MSCT)-coronary angiography confirmed the fistulization from the conus and posterolateral branches of RCA to the mural thrombus adherent to the LA wall (Figure 3A,B,C). Subsequently, she underwent surgery and was followed up at different health centers.

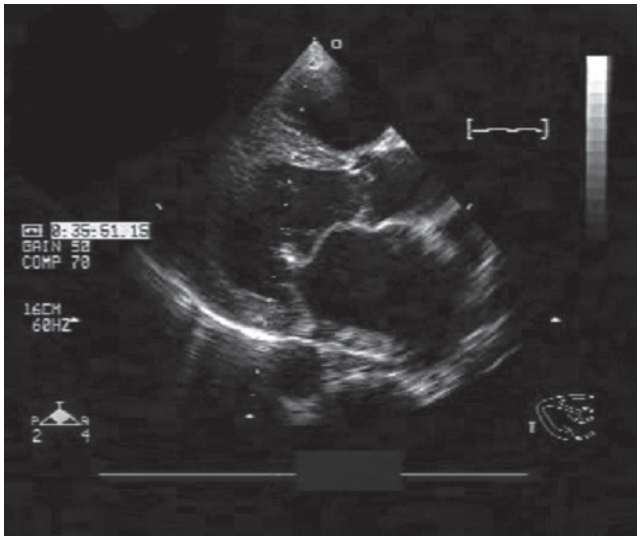



**Figure 1.** Rheumatic mitral valve with moderate to severe stenosis and mild mitral regurgitation on transthoracic echocardiography.

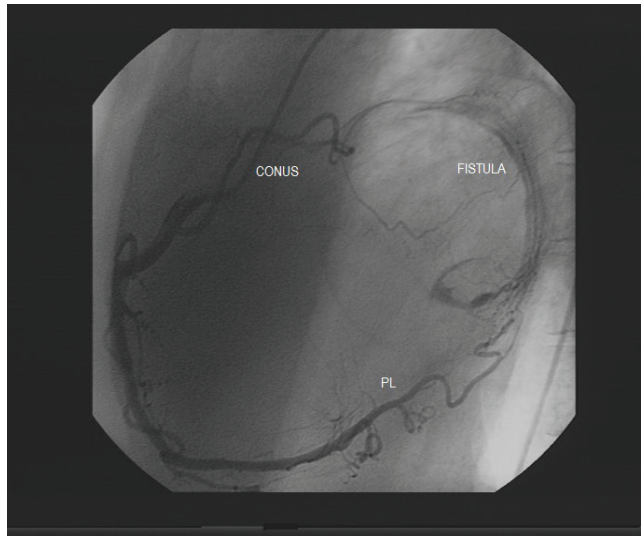



**Figure 2.** The conus and posterolateral branches of right coronary artery was fistulized to mural thrombus adherent to the left atrial wall.

## DISCUSSION

The first description of CAF was reported by the Austrian anatomist Josef Hytril in 1865, and the first surgical treatment was reported in 1958 by Fell[2,6]. Congenital CAF may develop when enlargement of the capillary network occurs during the embryogenesis of coronary circulation or when the main coronary arteries remain attached to the pulmonary trunk at the time of their separation[7]. The incidence of CAF is estimated at 1 in 50,000 live births, and 0.1% in the adult population are referred for cardiac catheterization[8,9]. Acquired CAF may occur secondary to myocardial infarction, chest trauma, or iatrogenic causes such as recurrent endomyocardial biopsies following heart transplantation, percutaneous transluminal coronary angioplasty, septal myectomy, permanent pacemaker implantation, closed-chest ablation of accessory pathways, or transbronchial lung biopsy[10]. In acquired CAF, fistula formation have been associated with an increased expression of some growth factors such as vascular endothelial growth factor, which has the most important role in angiogenesis and vascular migration[11]. The incidence of symptoms and complications increases with age, particularly after the age of 20[12,13]. The most frequent symptoms of CAF are dyspnea at rest or on exertion, fatigue, and angina[1,14].

In approximately half of the cases, CAF arise from RCA and its branches. Hobbs et al. reported the greatest number of patients with CAF and classified the fistulae according to their origin. In their patient series, 50% of fistulae arise from RCA, 42% from left main coronary artery, and 19% from both[15]. Drainage most frequently occurs into the right side of the heart and pulmonary artery. Lowe et al. reported that 39% fistulae drained into the right ventricle (RV), 33% into the superior vena cava/right atrium/coronary sinus (SCV/RA/CS), 20% into the pulmonary artery (PA), 6% into the LA, and 2% into the left

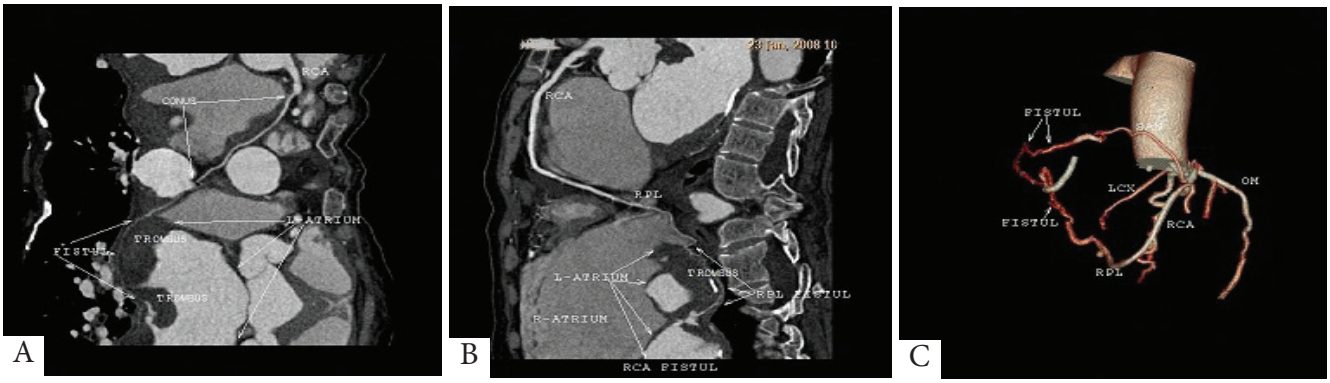



**Figure 3. (A, B, C)** 64-slice multidetector computed tomography-coronary angiography confirmed the fistulization from the conus and posterolateral branches of right coronary artery to the mural thrombus adherent to the left atrial wall.

ventricle (LV)[2]. In our case report, two CAF arose from conus and right posterolateral branches of RCA and drained into the LA through a large mural thrombus adherent to the LA wall in a patient with rheumatic MS. Only few CAF cases associated with additional mitral valve disease have been reported in the literature.

Whether MS and CAF incidentally occurs simultaneously or occurs as a result of hemodynamic changes in MS remains unclear. Left atrial thrombus is a frequent complication of MS. In 1975, Standen first described a fistulous tract leading from CX to the LA in a patient with severe MS and left atrial thrombus and suggested that the fistula formation resulted from the necrosis of the thrombus with ulcerated surface[16]. In 1981, Colman et al. evaluated coronary angiograms of 507 patients prior to open mitral valve surgery for the diagnosis of neovascularization and its relation with atrial thrombi. Angiographic neovascularization was observed in 30 patients, and all of them arose from the CX coronary artery. This retrospective study showed that coronary neovascularization with fistula formation was a specific sign for the presence of left atrial thrombus[17]. Fu M et al. also reported that coronary neovascularization was a specific sign of thrombus in 75 patients with MS[18]. In this study, 21 patients showed positive echocardiography findings, which suggested LAA thrombus, and 20 patients showed abnormal vessel or neovascularization; in 18 patients, neovascularization arose from LCA to LAA, and in the remaining, it arose fom RCA to LAA. In 1996, Mclung JA et al. reported a case with angiographically documented fistula between CX and LAA in a patient with MS, which demonstrated TEE[19]. In 2009, a case with neovascularization from RCA and CX to LA thrombi in an elderly patient with severe degenerative MS was reported[20]. In 2012, Hammami et al. reported a case of neovascularization between CX and LA thrombi in a patient with severe rheumatic MS[21]. Most neovascularization to LA thrombi reported cases arose from CX and only few cases had additional rheumatic MS. Our case is the first to report neovascularization or fistula between RCA and LA in a patient diagnosed with rheumatic MS and myocardial bridging in the LAD.

Various cardiac imaging modalities are used for the diagnosis and treatment of CAF. Combined two-dimensional and pulsed Doppler echocardiography can demonstrate a dilated coronary artery and turbulent flow in the fistula and recipient chamber[22]. TEE, particularly with a multiplane probe, is superior to TTE for detecting the origin, course, and drainage site of CAF[23]. Conventional angiography is an invasive procedure and sometimes fail to demonstrate the spatial relationship and connections of CFA. New-generation MSCT has become a fundamental tool for several coronary artery diseases. This technology may be applied for the evaluation of CAF, including the origin, course, and drainage of the fistulae. The three-dimensional geometry of MSCT can show more details regarding the spatial relationship with contiguous and surrounding structures and can provide details about the tortuous anatomy of the vascular malformation as an important advantage over other mentioned diagnostic modalities, particularly in volume-rendered views[24]. Magnetic resonance imaging have also become alternative methods to evaluate the anatomy, flow, and function of CAF.

## CONCLUSION

CAF are rare coronary artery abnormalities, which are usually congenital in origin. Neovascularization in association with mural thrombus is one of the acquired causes of CAF in patients with MS. Although the combination of various cardiac imaging modalities are used for the diagnosis and treatment of CAF, MSCT can provide more details about CAF.

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