

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 (CAF) which are abnormal connections between the coronary artery branches and cardiac chambers or major vessels are usually congenital forms of coronary artery anomalies but rarely may be acquired due to cardiac trauma, neovascularity in association with cardiac neoplasm, mural thrombus formation or iatrogenic causes. Although half of the patients with a coronary artery fistula remain asymptomatic, these fistulae may lead to angina, syncope, congestive heart failure, myocardial infarction and sudden death. Here we present a coronary artery fistula which is between Right Coronary Artery (RCA) and Left Atrium (LA), drainage into a large LA thrombus in a patient with rheumatic mitral stenosis. The diagnosis was confirmed by 2 Dimensional Echocardiography, Conventional Coronary Angiography and MultiSlice Computed Tomography(MSCT) coronary angiography. To the best of our knowledge this interesting and unique entity is first to be reported in the current literature.

Keywords: Coronary Artery Fistula, Left Atrial Thrombus, Rheumatic Mitral Stenosis

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

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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, iatrojenik 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 enfarktüsü 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 trombüsüne açılan Koroner Arter Fistülünü sunuyoruz. Tanı 2 Boyutlu Ekokardiyografi, Konvansiyonel Koroner Anjiyografi ve Çokkesitli 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

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Introduction

Coronary artery fistulae (CAF) are abnormal connections between coronary arteries and cardiac chambers, great vessels (vena cava, pulmonary veins, pulmonary artery) or other vascular structures (mediastinal vessels, coronary sinus). CAF are usually of congenital origin and not gender specific [1,2] however sometimes occur secondary to cardiac surgery, cardiac invasive procedures, chest trauma, neovascularity in association with cardiac neoplasm and formation of mural thrombus [3,4]. Although half of the patients with CAF remain asymptomatic, these fistulas 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 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 diagnosed incidentally during coronary angiography. CAF arise from right coronary artery (RCA) or its branches in about half of the cases. The remainings arise from the left anterior descending (LAD) and circumflex (CX) coronary arteries or their branches. Drainage occurs most frequently into the right side of the heart or pulmonary artery [5]. The association between rheumatic mitral stenosis (MS) and CAF is rare in the current literature. According to our knowledge, a fistula formation that originated from RCA branches and drained through a large left atrial thrombus to the left atrium in a patient with rheumatic mitral stenosis has not been reported yet.

Case Presentation

A 57 years old female patient 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, arrhythmic and body temperature was 36.6 °C. Cardiac auscultation revealed an opening snap, soft diastolic rumble at the apex. A loud mid-systolic murmur was heard along the left sternal border. The electrocardiogram showed AF, left ventricular hypertrophy and ST segment depression on V5-V6. Transthoracic echocardiography (TTE) showed 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 (EF) of 35%. Transesophageal echocardiography (TEE) revealed a large mural thrombus formation and spontaneous echo contrast images in the left atrium. She underwent coronary angiography before surgery which revealed normal LAD and CX coronary arteries. The conus and posterolateral branches of RCA was fistulized to mural thrombus adherent to the left atrial 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 3-4-5]. Subsequently she underwent surgery and was followed at different health center.

Discussion

The first description of CAF was reported by the Austrian anatomist Josef Hyrtl in 1865 [2] and the first surgical treatment was reported in 1958 by Fell. [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 1/50.000 live births [8] and 0.1% in adult population who are referred for cardiac catheterization [9]. Acquired CAF may occur secondary to myocardial infarction, chest trauma or iatrogenic causes such as recurrent endomyocardial biopsies after 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 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 about half of the cases CAF arise from the 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 arised from RCA, %42 arised from left main coronary artery (LMCA), and 19% arised from both [15]. Drainage occurs most frequently into the right side of the heart and pulmonary artery. Lowe et al reported that 39% fistulae drained into the right ventricle (RV), 33% drained into the superior vena cava/right atrium/coronary sinus (SCV/RA/CS), 20% drained into the pulmonary artery (PA), 6% drained into the left atrium(LA) and 2% drained into the left ventricle (LV) [2]. In our case report, there are two CAFs, they arise 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 mitral stenosis. Only few CAF cases associated with additional mitral valve disease have been reported in the literature.

The question of whether MS and CAFs togetherness occurs incidentally or as a result of the hemodynamic changes in MS has not been answered completely in the current literature yet. Left atrial thrombus is a frequent complication of MS. Standen first described a fistulous tract leading from CX to the left atrium in a patient with severe MS and left atrial thrombus in 1975 and also suggested that the fistula formation resulted from necrosis of the thrombus with ulcerated surface [16]. In 1981 Colman et al evaluated coronary angiograms of 507 patients before open mitral valve surgery for diagnosis of neovascularization and its relation with atrial thrombi. In 30 patients angiographic neovascularization was shown. All of them arised 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]. Also Fu M et al reported that coronary neovascularization was a specific sign of thrombus in 75 patients with MS [18]. In this study twenty-one patients had positive echocardiographic findings suggesting LAA thrombus and twenty patients had abnormal vessel or neovascularization. In 18 patients neovascularization arised from LCA to LAA, and in the remaining 2 patients neovascularization arised from RCA to the LAA. In 1996, Mclung JA et al reported a case with angiographically documented fistula between the CX and LAA in a patient with MS, which demonstrated by 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 with severe rheumatic MS [21]. The most of the neovascularization to LA thrombi cases in the current literature arised from CX and only few cases were reported to have additional rheumatic MS. Our case is the first to be reported with 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, turbulent flow in the fistula and the recipient chamber [22]. TEE particularly with a multiplane probe is superior to TTE in detection the origin, course, 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 becomes 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 3-dimensional geometry of MSCT can show more details regarding the spatial relationship with contiguous and surrounding structures, providing 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 also have 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 combination of various cardiac imaging modalities are used for diagnosis and treatment, MSCT can show more details about CAF.

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Conflict of Interest; All authors declare that there is no conflict of interest

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Figure Legends:



Figure 1: Rheumatic mitral valve with moderate to severe stenosis and mild mitral regurgitation on transthoracic echocardiographic examination.

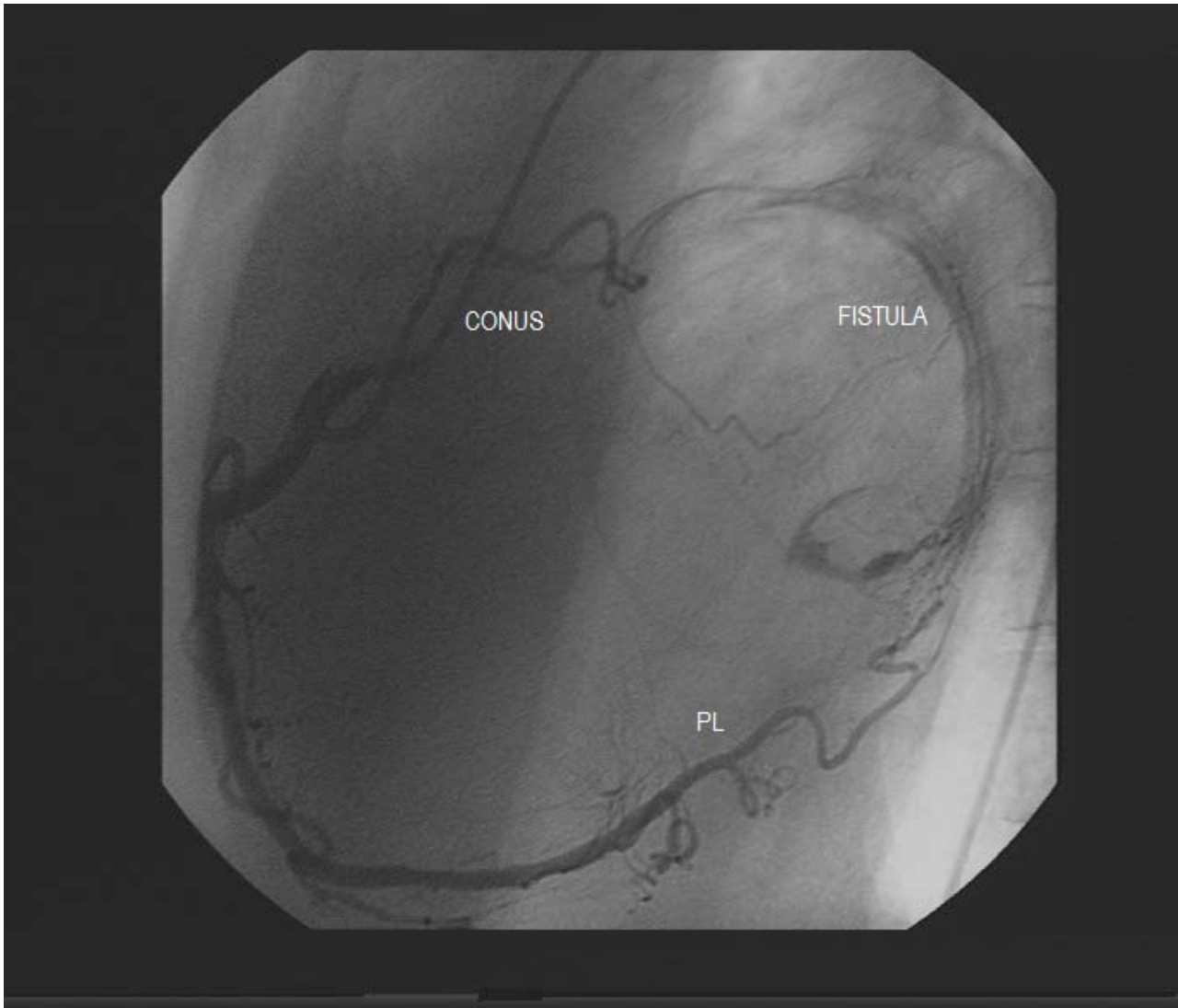


Figure 2: The conus and posterolateral branches of right coronary artery was fistulized to mural thrombus adherent to the left atrial wall

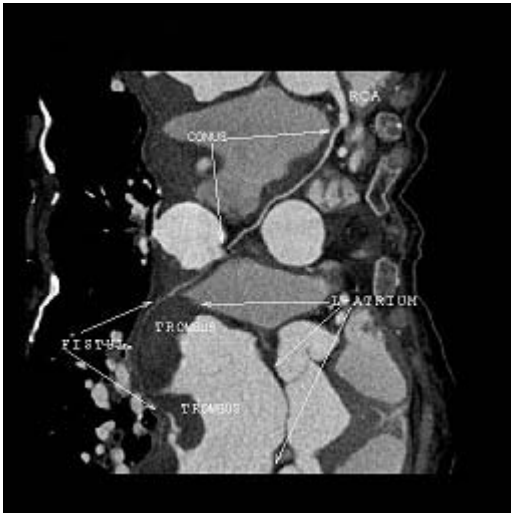


Figure 3-5: 64 Slice Multidetector Computed Tomography coronary angiography confirmed the fistulization from the conus and posterolateral branches of right coronary artery to the mural thrombus adherent to the left atrial wall



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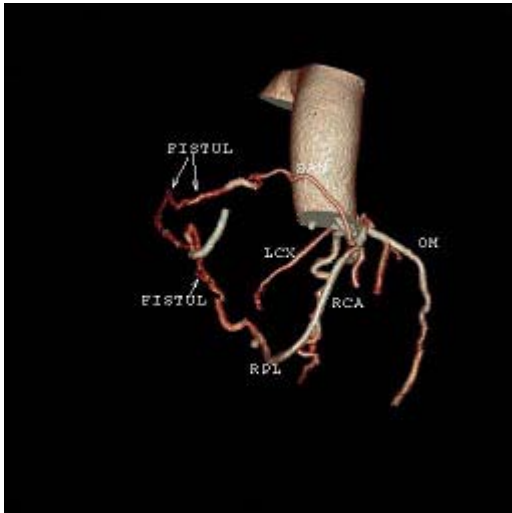


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