

Right Coronary Artery to Right Atrial Fistula: Role of Multi-Modality Imaging and Percutaneous Closure

Review began 07/04/2022

Review ended 07/07/2022

Published 07/10/2022

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Abstract

Coronary artery fistula (CAF) is a connection between a coronary artery and a cardiac chamber or nearby vessel. Our case represents a fistula arising from the right coronary artery and terminating in the right atrium, presenting as atrial fibrillation. CAF closure options include surgical and percutaneous approaches.

Categories: Cardiology, Internal Medicine

Keywords: right atrium, percutaneous closure, cardiac magnetic resonance imaging, atrial fibrillation, coronary artery fistula

Introduction

Coronary artery fistula (CAF) is defined as a connection arising between a coronary artery and a cardiac chamber or another nearby vessel. CAF can be congenital, iatrogenic, or acquired secondary to trauma. Pacemaker implantation, endomyocardial biopsy, and coronary angiography are considered invasive cardiac procedures in which CAF may arise [1]. In most cases, CAFs appear as an isolated finding (90.5%); however, CAF can be associated with other congenital cardiac malformations, including atrial septal defects, patent foramen ovale, pulmonary artery atresia, tetralogy of Fallot, ventricular septal defects, and patent ductus arteriosus [2]. We present a case of a patient who presented with atrial fibrillation and, on further investigation, was found to have a CAF. The patient was treated using the percutaneous approach.

Case Presentation

History of presentation

A 60-year-old male patient with a past medical history of type 2 diabetes mellitus was referred to the cardiology clinic for dyspnea of six months duration and a new diagnosis of atrial fibrillation (AF). Due to worsening symptoms, a transesophageal echocardiogram (TEE)-guided cardioversion was planned to attempt restoration of normal sinus rhythm (NSR).

Differential diagnosis

Differential diagnoses included aortic diverticulum, aortic dissection pouch, Kawasaki disease, and a coronary artery fistula.

Investigation

The patient underwent TEE, which revealed normal left ventricular systolic and right ventricular systolic functions. Atria was mildly enlarged. The left atrial (LA) appendage was free of thrombus. The most striking abnormality was an abnormal vascular structure of 4 x 4 cm adjacent to the ascending aorta (Figure 1).

How to cite this article

Jabri A, Shahrory Z, Nasser M, et al. (July 10, 2022) Right Coronary Artery to Right Atrial Fistula: Role of Multi-Modality Imaging and Percutaneous Closure. Cureus 14(7): e26716. DOI 10.7759/cureus.26716

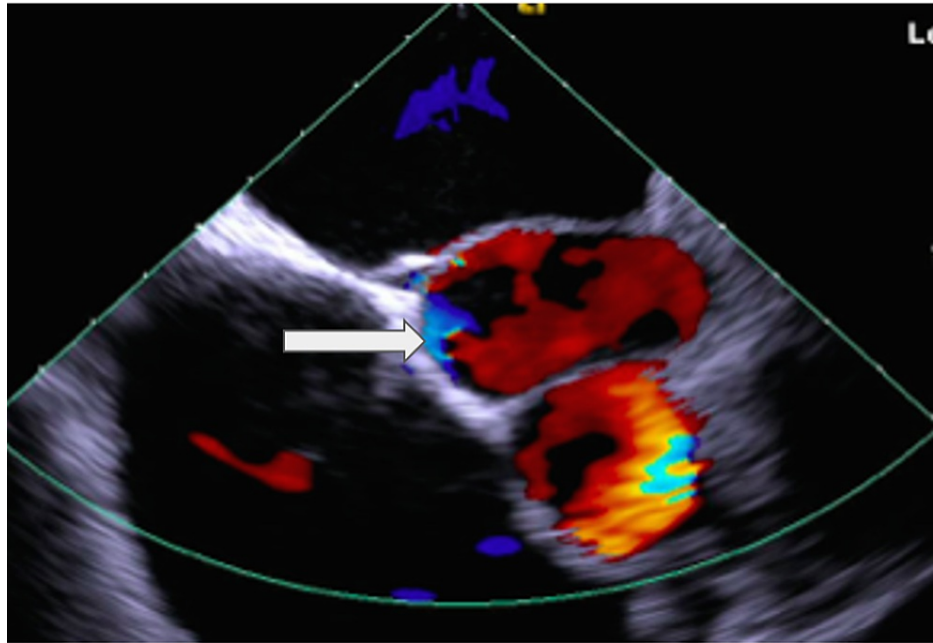


FIGURE 1: A transesophageal echocardiogram (mid-esophageal short-axis view) shows an abnormal structure (4 x 4 cm) adjacent to the aorta (white arrow).

The patient was cardioverted to NSR with 200 joules of synchronized shock. Chest computed tomography (CT) was performed next for further evaluation. It revealed a large, very tortuous, and saccular fistula measuring 3.6 x 3.3 cm connecting the right coronary artery (RCA) to the right atrium (RA) (Figure 2). A cardiac MRI was also performed for further assessment confirming the presence of fistula and significant shunting (Figure 3). The shunt fraction of pulmonary blood flow to systemic blood flow ($Q_p:Q_s$) was 1.7, indicating a significant left-to-right shunt. Afterward, coronary angiography was performed, which revealed a large fistula arising from the ostium of the RCA and emptying into the RA (Figure 4). A significant amount of contrast media passed from the aorta into the RA through the fistulous connection.

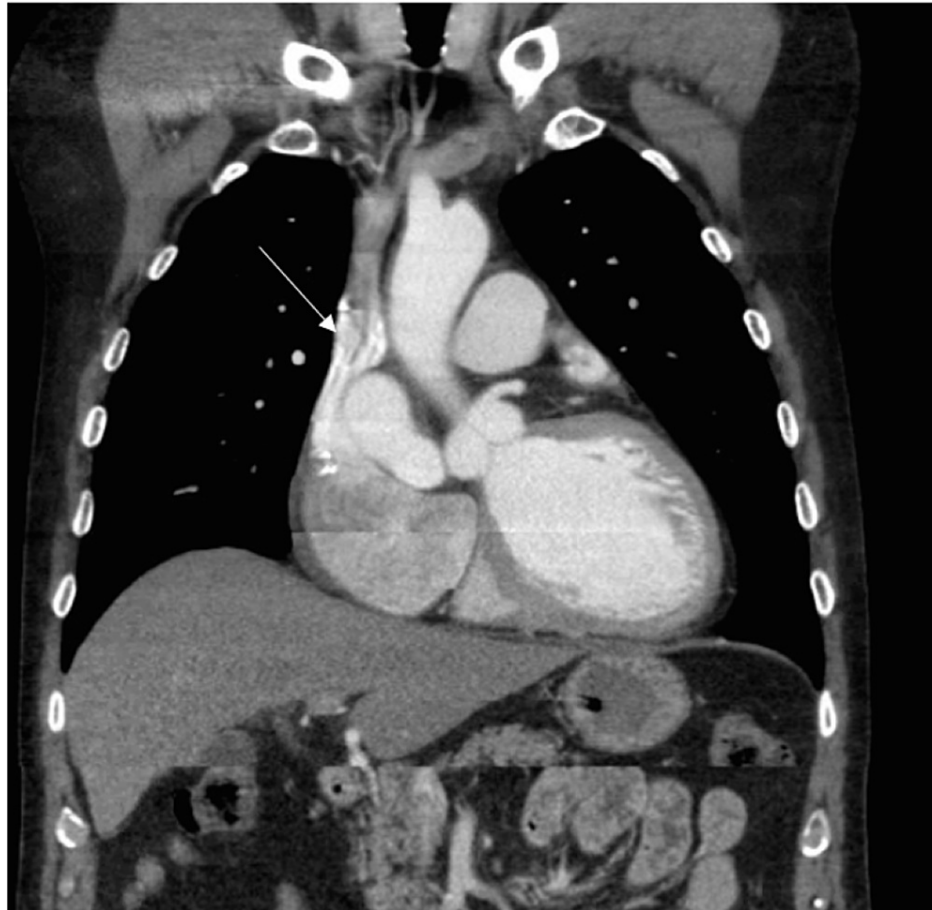


FIGURE 2: Chest CT showing saccular fistula (white arrow) measuring 3.6 x 3.3 cm connecting the right coronary artery to the right atrium.

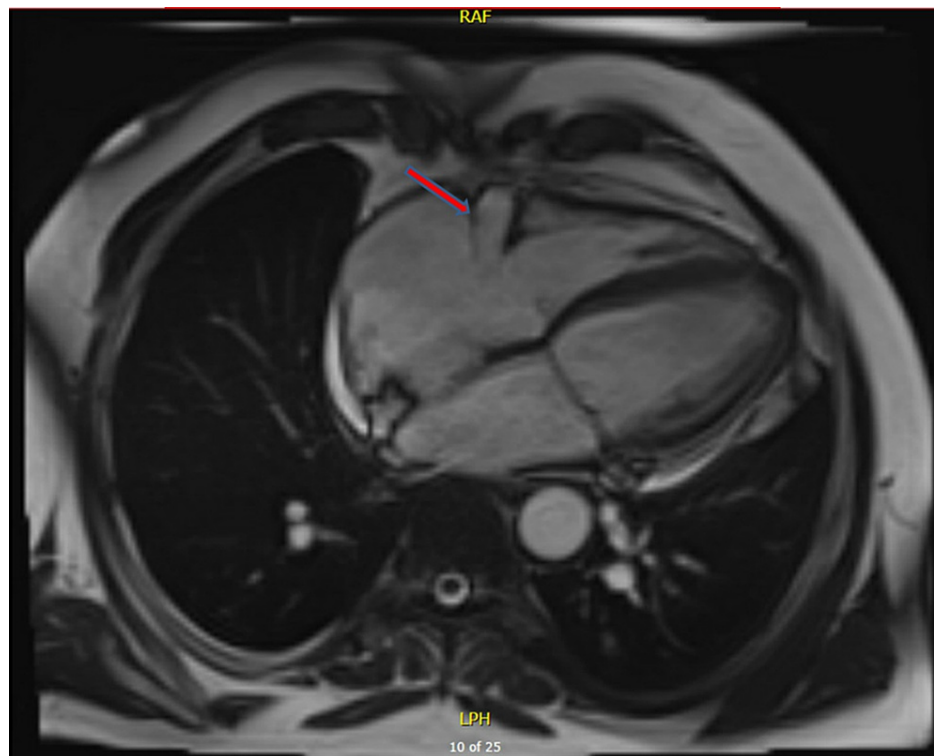


FIGURE 3: Cardiac MRI showing the fistula and shunting (red arrow).

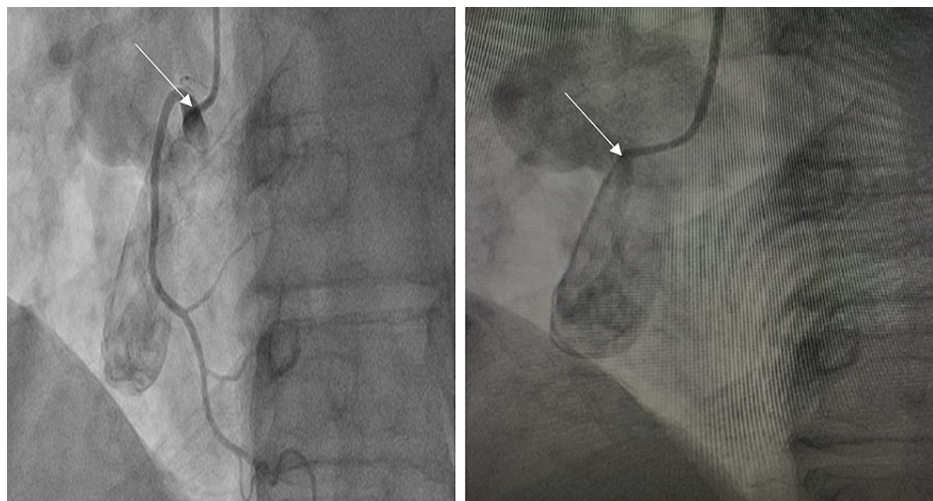


FIGURE 4: Fistula arising from the ostium of the right coronary artery and emptying into the right atrium (white arrow).

Management

The structural heart team was consulted to evaluate percutaneous therapeutic options instead of more invasive surgical options. The patient later underwent percutaneous closure with a 16 mm Amplatzer Vascular Plug II (Abbott Laboratories, Chicago, IL) and coils (Figure 5). Angiography following the procedure showed occlusion of the fistula. The patient was discharged on apixaban, aspirin, and clopidogrel with a plan to discontinue clopidogrel after one month and continue aspirin and apixaban indefinitely. The patient reported improvement in symptoms in the one-month follow-up visit.

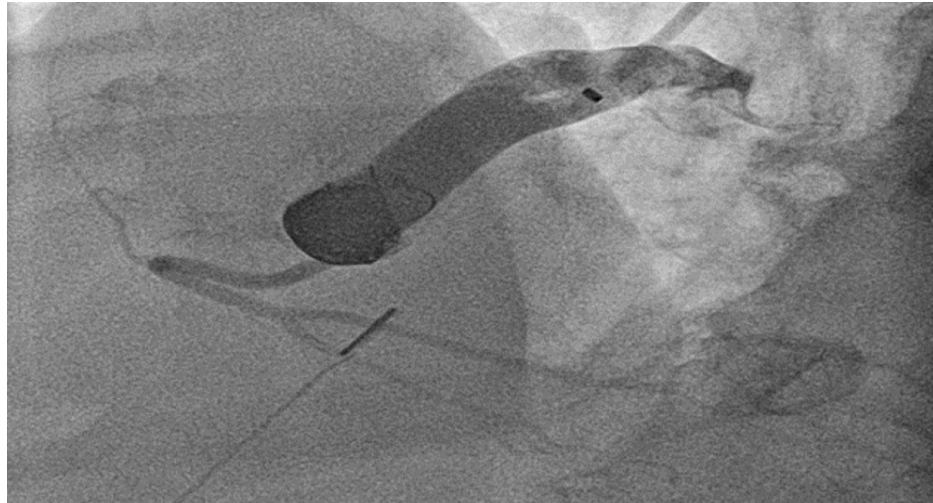


FIGURE 5: The patient undergoing percutaneous closure with a 16 mm Amplatzer Vascular Plug II and coils.

Discussion

The most common site of origination of CAFs is the RCA with an incidence of 33%, followed by the left coronary artery (LCA) (34.9%), left anterior descending artery (6.3%), circumflex artery (4.8%), and finally, both RCA and LCA (1.6%) [3]. CAFs do not only arise from different coronaries but also terminate in different cardiac chambers and nearby vessels. The right ventricle (RV) is the most prevalent drainage site for CAFs, making up 34.9% of cases. The RA (27%) and pulmonary artery (PA) (27%) are the second most common sites of drainage and are followed by the left ventricle (LV) (6.3%), coronary sinus (CS) (3.2%), and finally LA (1.6%) [3].

The clinical manifestations of CAFs rely on the diameter of the fistula and the difference between the systemic blood pressure affecting the coronary artery and the blood pressure in the draining chamber. In most circumstances, CAFs are relatively small, and patients are asymptomatic. However, CAFs can result in the coronary artery steal phenomenon and deviate blood flow through the coronary arteries into the terminating chambers. Symptoms may arise because of oxygen supply deprivation to the myocardium. Heart failure, myocardial infarction, pulmonary hypertension, endocarditis, arrhythmias, thrombosis of the fistula, or fistula rupture may also occur. A loud continuous murmur at the lower sternal or midsternal border is the most prevalent finding of CAFs. Few cases have been reported in the literature of CAFs presenting with atrial fibrillation. Two previously reported atrial fibrillation cases were secondary to CAF arising from the RCA and ended in either the superior vena cava or the RA. The location of the fistula likely contributed to the evolution of AF in those scenarios and our case [4].

The diagnosis of CAF can be challenging. Initial evaluation of the patient should involve an electrocardiogram, which could be normal, show chamber enlargement, arrhythmias, or signs of ischemia, depending on the site and flow of the fistula. Two-dimensional and color Doppler transthoracic echocardiogram (TTE) or TEE are valuable tools for evaluating patients with CAF [5]. Moreover, cardiac MRI will allow identification of the site, size, and shunt flow. A cardiac CT angiogram is another modality for diagnosis. However, coronary angiography is still the gold standard of diagnosis.

CAF management is mainly related to the presence or absence of symptoms and the amount of shunting. Symptomatic patients can be candidates for either surgical or percutaneous closure. On the other hand, the management of asymptomatic CAFs varies depending on the risk of complication, for which high risk will require treatment versus lower risk may only require monitoring [6]. The course, tortuosity, and the coexistence of aneurysmal dilation dictate the approach used in the fistula closure. Patients with CAF who require a bypass graft or surgical valve repair/replacement should undergo surgical closure.

Moreover, CAFs with high flow, branching course, tortuosity, or coexistent aneurysmal enlargement are better approached with the surgical technique. Patients with CAFs who did not meet the above characteristics can better tolerate percutaneous closure [7]. Transcatheter closure is preferred when applicable due to the lower risk of bleeding, infections, wound healing, general anesthesia adverse effects, and cardiopulmonary bypass complications. Post-closure medical therapy recommendations include antiplatelet medications and prophylactic antibiotics for infective endocarditis for the first six months before undergoing high-risk procedures [8].

Complications of CAF include recurrence rates of 9-19% with transcatheter closure versus 25% with surgical ligation [9]. Other complications include myocardial infarction, tricuspid regurgitation, thromboembolism, or death. Post repair, the recorded survival rates of coronary artery fistulas in the literature were 93%, 74%, and 68% at one, five, and 15 years, respectively [10]. In the case presented by Jamali et al., the author mentioned complete resolution of atrial fibrillation post obliteration of the CAF [4].

Conclusions

CAFs have a broad spectrum of presentations. The characteristics of the fistula dictate the severity of signs and symptoms and will later influence the treatment approach. Arrhythmias, including AF, can be the presenting feature of CAFs and may be eradicated post obliteration of the CAF. Percutaneous and surgical approaches have been associated with recurrence, and therefore long-term follow-up is more appropriate.

Additional Information

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

Acknowledgements

Ahmad Jabri and Zaid Shahreri are both first authors.

References

1. Spaedy TJ, Wilensky RL: Coronary artery fistulas: clinical implications. *ACC Curr J Rev.* 1994, 3:24-5.
2. Levin DC, Fellows KE, Abrams HL: Hemodynamically significant primary anomalies of the coronary arteries. Angiographic aspects. *Circulation.* 1978, 58:25-34. [10.1161/01.cir.58.1.25](https://doi.org/10.1161/01.cir.58.1.25)
3. Xie M, Li L, Cheng TO, et al.: Coronary artery fistula: comparison of diagnostic accuracy by echocardiography versus coronary arteriography and surgery in 63 patients studied between 2002 and 2012 in a single medical center in China. *Int J Cardiol.* 2014, 176:470-7. [10.1016/j.ijcard.2014.07.198](https://doi.org/10.1016/j.ijcard.2014.07.198)
4. Jamali HK, Raza U, Waqar F: Idiopathic atrial fibrillation and coronary arteriovenous fistulae: is there a link?. *Cardiology.* 2016, 134:433-5. [10.1159/000444846](https://doi.org/10.1159/000444846)
5. Shakudo M, Yoshikawa J, Yoshida K, Yamaura Y: Noninvasive diagnosis of coronary artery fistula by Doppler color flow mapping. *J Am Coll Cardiol.* 1989, 13:1572-7. [10.1016/0735-1097\(89\)90351-3](https://doi.org/10.1016/0735-1097(89)90351-3)
6. Buccheri D, Dendramis G, Piraino D, et al.: Coronary artery fistulas as a cause of angina: how to manage these patients?. *Cardiovasc Revasc Med.* 2015, 16:306-9. [10.1016/j.carrev.2015.04.011](https://doi.org/10.1016/j.carrev.2015.04.011)
7. Buccheri D, Chirco PR, Geraci S, Caramanno G, Cortese B: Coronary artery fistulae: anatomy, diagnosis and management strategies. *Heart Lung Circ.* 2018, 27:940-51. [10.1016/j.hlc.2017.07.014](https://doi.org/10.1016/j.hlc.2017.07.014)
8. Said SA, Lam J, van der Werf T: Solitary coronary artery fistulas: a congenital anomaly in children and adults. A contemporary review. *Congenit Heart Dis.* 2006, 1:63-76. [10.1111/j.1747-0805.2006.00012.x](https://doi.org/10.1111/j.1747-0805.2006.00012.x)
9. Said SA, Nijhuis RL, Op den Akker JW, et al.: Diagnostic and therapeutic approach of congenital solitary coronary artery fistulas in adults: Dutch case series and review of literature. *Neth Heart J.* 2011, 19:183-91. [10.1007/s12471-011-0088-2](https://doi.org/10.1007/s12471-011-0088-2)
10. Said SM, Burkhart HM, Schaff HV, et al.: Late outcome of repair of congenital coronary artery fistulas—a word of caution. *J Thorac Cardiovasc Surg.* 2013, 145:455-60. [10.1016/j.jtcvs.2012.11.028](https://doi.org/10.1016/j.jtcvs.2012.11.028)