

REVIEW ARTICLE

Coronary Fistula Between Left Anterior Descending Artery (LAD) and Pulmonary Artery (PA) Leading to Sudden Cardiac Death: Case Report with Literature Review

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Abstract: Coronary Artery Fistulas (CAF) are inappropriate connections between a coronary artery and a major vessel or a cardiac chamber. These fistulas may or may not present with symptoms, but they need to be detected as early as possible in order to decide the most appropriate treatment methodology (*i.e.* surgery vs. conservative management). We report the case of a 67-year-old female with no modifiable cardiovascular risk factors who had an unwitnessed sudden death at home during her ongoing evaluation of a fistula detected incidentally between the Left Anterior Descending Artery (LAD) and the Pulmonary Artery (PA). This case highlights that early diagnosis and treatment of symptomatic CAF are crucial to minimize the risk of sudden cardiac death. Although the symptomatic fistula of LAD to PA has been reported in the literature multiple times, it has been rarely reported that this fistula can result in sudden cardiac death.

Keywords: Coronary fistula, fistula between LAD and pulmonary artery, congenital coronary artery disease, angina, dyspnea, heart failure.

1. INTRODUCTION

Coronary Artery Fistulas (CAFs) are rare congenital or acquired coronary artery anomalies that occur in about 0.88% of the general population as demonstrated in retrospective studies on patients who have undergone coronary CTA [1] with prevalence rates for coronary-pulmonary artery fistula (CPAF) ranging from 0.32% to 0.68% [2, 3]. Most patients are asymptomatic, but angina, dyspnea, heart failure, arrhythmias or sudden cardiac death may arise as a consequence of coronary fistula [4].

We report the case of a 67-year-old woman with a history of atrial fibrillation who presented with shortness of breath. Cardiac catheterization revealed a large aneurysmal Left Anterior Descending Artery (LAD) draining into the Pulmonary Artery (PA). A few days afterward, the patient died suddenly at home. In this article, we will highlight the importance of diagnosis and treatment of PA to LAD fistula

and its association with congestive heart failure and sudden cardiac death. Most CAFs are asymptomatic, but once symptoms arise, clinicians should treat it as early as possible in order to minimize serious consequences, such as mortality which was demonstrated in our case.

2. REPORT OF THE CASE

A 67-year-old obese female with a past medical history of atrial fibrillation on anticoagulation, daily alcohol consumption, panic attacks, and generalized anxiety disorder, is presented to the emergency department with shortness of breath, heart racing, and decreased exercise tolerance for the last week. She reported that she developed dyspnea with minimal activity, worsening progressively to the point of not being able to perform activities of daily living. She described her exercise tolerance as less than one block limited by dyspnea. She denied chest pain or dizziness. Physical examination revealed a well-developed female, in mild distress due to dyspnea. She was afebrile with blood pressure within normal range and a heart rate of 84 with irregularly irregular rhythm. Her electrocardiogram demonstrated atrial fibrillation with the episode of premature ventricular contractions

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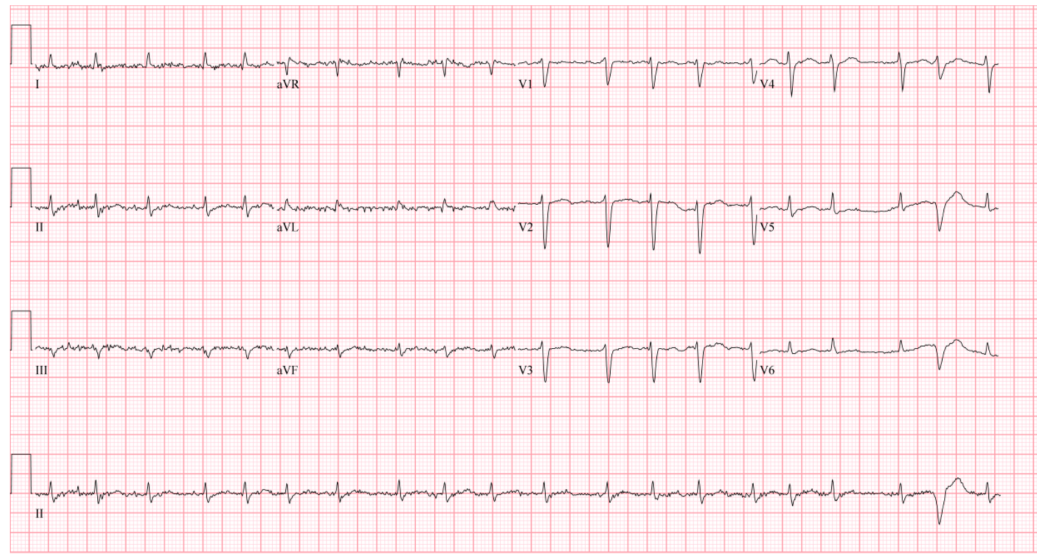


Fig. (1). EKG demonstrated atrial fibrillation with episode of premature ventricular contractions. (A higher resolution / colour version of this figure is available in the electronic copy of the article).

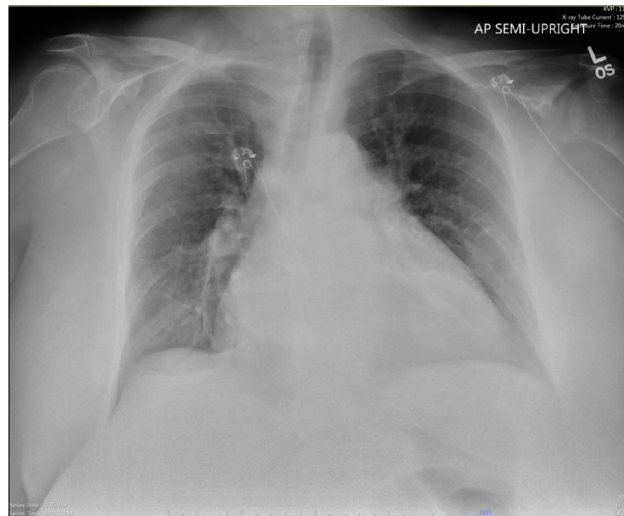


Fig. (2). Chest X ray showed cephalization of pulmonary vasculature and cardiomegaly. (A higher resolution / colour version of this figure is available in the electronic copy of the article).

(Fig. 1). Chest X-ray showed the cephalization of pulmonary vasculature and cardiomegaly (Fig. 2). Transthoracic echocardiography revealed an ejection fraction of 25% to 30% with severe global hypokinesia with regional variation; diastolic function was not assessed due to atrial fibrillation. The echocardiogram also showed mitral and tricuspid regurgitation (Fig. 3). There is no record of a previous echocardiogram. Blood tests revealed elevated N-terminal pro B-type natriuretic peptide (NT-pro BNP) at 5,975 pg/mL [reference range < 300 pg/mL], aspartate aminotransferase (AST) 200 IU/L [8 to 48 IU/L] and alanine aminotransferase (ALT) 212 IU/L [7 to 55 IU/L]. She was admitted for cardiac monitoring due to new-onset acute decompensated systolic heart failure with elevated transaminases likely due to congestive hepatopathy. She started diuretics, beta-blockers for rate control, and angiotensin-converting enzyme inhibitors. During her stay, cardiac monitoring showed episodes of nonsustained ventricular tachycardia. As part of the work up to de-

termine the cause of new-onset acute heart failure, she was taken to cardiac catheterization which revealed non-obstructive epicardial coronary disease with large aneurysmal Left Anterior Descending Artery (LAD) connection draining into the Pulmonary Artery (PA) (Fig. 4). The occurrence of systolic heart failure was attributed to tachyarrhythmia-induced cardiomyopathy, fistula-induced high-output heart failure, and alcohol-related cardiomyopathy. The patient was scheduled to undergo coronary artery CTA as an outpatient, but the study was postponed with a goal of doing it when the heart rate was brought under control for optimal imaging. She was discharged a few days later once euvolemic status was achieved. A few days after being discharged, the hospital notified that the patient died suddenly at home with failed attempts at resuscitation. Upon our knowledge, no autopsy was done. The cause of death was presumed due to complications from abnormal connections between LAD and PA.

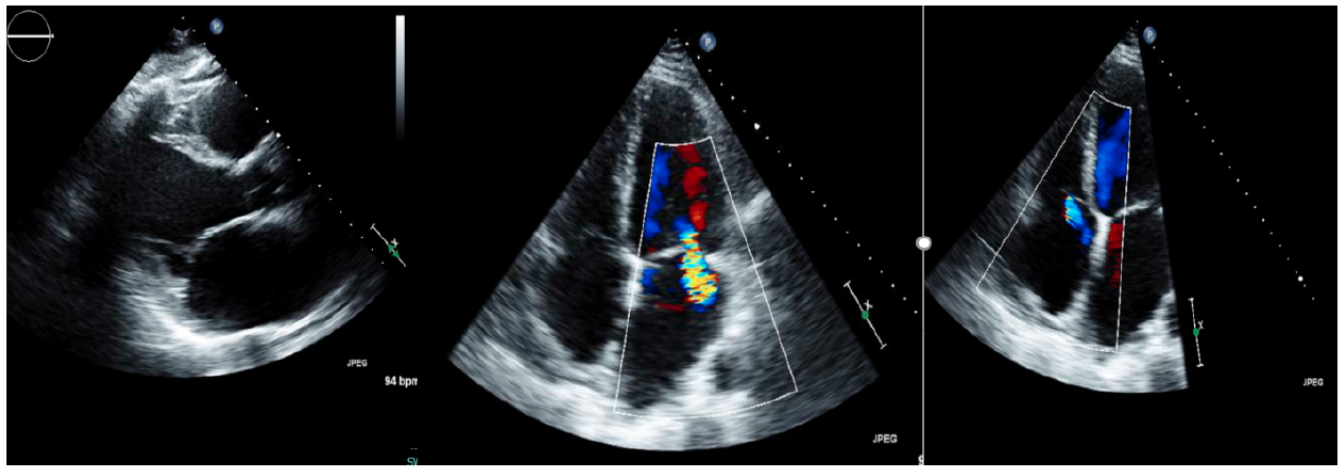


Fig. (3). Transthoracic echocardiography revealed an ejection fraction of 25% to 30% with severe global hypokinesia with regional variation, diastolic function was not assessed due to atrial fibrillation. It showed also mitral and tricuspid regurgitation. (A higher resolution / colour version of this figure is available in the electronic copy of the article).

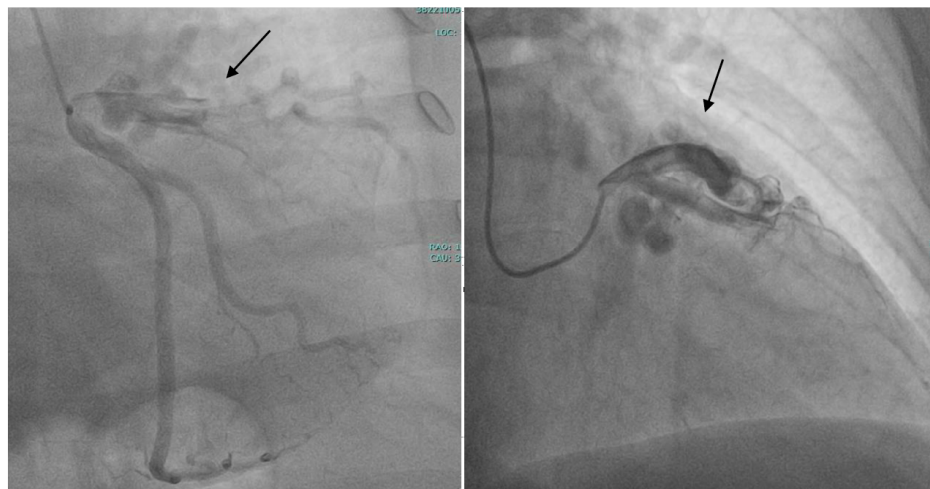


Fig. (4). Cardiac catheterization which revealed non obstructive epicardial coronary disease with large aneurysmal left anterior descending artery (pulmonary artery (PA)) connection draining into pulmonary artery (PA) - arrows indicates the fistula-. (A higher resolution / colour version of this figure is available in the electronic copy of the article).

3. DISCUSSION

The accurate incidence of Coronary Artery Fistulas (CAF) in the population is difficult to be estimated because they are usually asymptomatic. However, the available data suggest that its prevalence ranges from 0.09 to 0.5% [5]. Another study showed that the most common type of CAF found by MDCT studies is coronary-to-pulmonary artery fistula (CPAF) with the prevalence of $0.17 \pm 0.67\%$ [6]. One study showed that the global incidence of CAF is closer to 0.002% [7].

There are two types of coronary fistulas: type I is a single artery to artery connection, and type II is comprised of multiple CAFs [7]. The shared characteristic between both types is their high tendency to have a tortuous course [7]. In addition to the varied anatomy, the underlying pathophysiological mechanism in symptomatic patients appears to be coronary steal syndrome, whereby shunting of blood to the areas of least resistance to blood flow causes ischemia of the areas that have the highest resistance to blood flow [6]. In the ab-

sence of coronary artery disease, it is hypothesized that ischemic symptoms occur as a result of coronary steal [6]. However, not all researchers agree that coronary steal occurs in patients with CAF.

The most commonly reported type of fistula is where the Right Coronary Artery (RCA) fistulizes into the right heart chambers. Left heart coronary fistulas are less common and happened to occur in our patient. The vast majority of CAFs is present at birth [7]. However, they can develop as a result of iatrogenic intervention like in coronary angioplasty, coronary artery bypass surgery, or after myocardial biopsy and cardiac transplantation [8]. In another study, a larger proportion of fistulas emerged from either the right coronary artery or the left anterior descending artery [9].

The presentation of CAF is mostly non-specific and includes angina, coronary steal, endocarditis, and heart failure which make the diagnosis quite challenging [9]. Coronary artery fistulas can terminate into a cardiac chamber or into the pulmonary artery [10]. These abnormal connections are

typically detected incidentally on coronary angiography and carry serious risk of sudden cardiac death [10]. Their presence can also be detected on radiographic imaging that is done for other clinical indications, such as quantitation of cardiomegaly in heart failure or identification of masses. The suspicion for these anomalies should be elevated when there is no obstructive coronary artery disease to explain myocardial wall motion abnormalities or reduced left ventricular systolic function seen on echocardiography. CAFs can also be diagnosed during the evaluation of a precordial continuous murmur [10]. Another clue to the possibility of CAF is the presence of premature ventricular contractions [11]. Determinants of pretest probability of CAF are not currently known. A full cardiac workup through cardiac catheterization is necessary to find out whether there are coronary artery anomalies that can explain symptoms, as was the case in our patient. All the tests that were performed on our patient were necessary to arrive at the proper diagnosis, especially cardiac catheterization. Echocardiography, coronary angiography, and CT angiography are used to characterize the course of a fistula in symptomatic patient who may benefit from percutaneous closure of the fistula [12].

There are no established guidelines for the treatment of CAF. However, the current management is based on the presence or absence of symptoms. Symptomatic patients need surgery in order to reduce CAF complications, such as sudden cardiac death, while asymptomatic patients may benefit from conservative management with close outpatient monitoring. The indications for surgical intervention include pulmonary-systemic flow ratio exceeding 1.5:1, side branch obstruction, coronary rupture, or aneurysmal degeneration [10]. Patients who undergo fistula ligation need post-operative angiography or other imaging modality to ensure that the fistula does not recanalize [8]. Our patient had the fistula with a concomitant coronary aneurysm. Surgical treatment was found to be beneficial in a patient with simultaneous fistula and aneurysm [11]. The literature showed that patients who are at risk of hemodynamic compromise may benefit from coil embolization of the fistula [13-15]. The following cases illustrate the therapeutic benefit of endovascular coiling.

One case discussed a 24-year-old female who suffered blunt trauma while playing a sport and sustained what was most likely a hematoma that occluded a left main fistula for two weeks, with retrograde embolization of a clot into the left circumflex artery, causing a posterior wall MI [13]. She underwent percutaneous endovascular coiling in order to occlude the fistula and prevent embolization from recurring. As a result of the coiling, the patient remained symptom-free for 9 months thereafter. The study advocated for the use of coronary angiography for all patients who survive sudden cardiac death [13]. Another case report showed a 60-year-old symptomatic female who was found to have bilateral coronary to pulmonary artery fistulas, with one originating from the RCA and the other originating from the Left Main Coronary Artery (LMCA). Both were ligated in a single surgical setting, and the patient had a favorable outcome [14]. Lastly, one article presented the case of a 65-year-old male who had dominant multivessel fistulas and a non-dominant fistula. After undergoing percutaneous coiling of the dominant multivessel fistulas, with no intervention for the non-dominant

one, he remained asymptomatic up to 18 months of follow up [15].

With regard to sudden cardiac death, Lau *et al.* [16] reported the case of a 29-year-old young male who had a history of unknown congenital cardiovascular disease, for which no surgery was performed [16]. This man collapsed suddenly at home, after which he was transferred to the emergency room, where resuscitation attempts were unsuccessful. On autopsy, the cause of death was found to be myocardial ischemia related to a right-sided fistula, in which the right coronary artery was involved, along with left ventricular hypertrophy. By contrast, our patient's sudden cardiac death resulted from a left-sided fistula coursing from the LAD to the PA.

CONCLUSION

CAF is a relatively rare condition that can have catastrophic outcomes if not promptly identified and treated. This case report highlights the need for rapid diagnosis and therapeutic intervention for patients with the uncommon fistula anomaly, where symptomatology is deceptively benign but can result in disastrous consequences and include mortality, as in our patient. Sudden death was unexpected because she was risk-stratified using the information available to us, and that stratification turned out to be insufficient to account for the actual risk the patient faced when she was sent home. There is currently no standardized approach to the treatment of symptomatic vs. asymptomatic CAF. This case might contribute to a shift in paradigm, whereby an assessment of coronary arteries should be assessed not only with a pretest suspicion for atherosclerotic disease, but also with a pretest consideration for anomalous coronary paths. Further studies are needed to address this question.

LIST OF ABBREVIATIONS

AST	=	Aspartate Aminotransferase
ALT	=	Alanine Aminotransferase
CTA	=	Computed Topography Angiography
CAF	=	Coronary Artery Fistula
CPAF	=	Coronary-to-Pulmonary Artery Fistula
LAD	=	International Unit per Litre
LAD	=	Left Anterior Descending Artery
NT-pro BNP	=	N-terminal Pro B-type Natriuretic Peptide
pg/ml	=	Picogram/milliliter
PA	=	Pulmonary Artery

CONSENT FOR PUBLICATION

Not applicable.

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CONFLICT OF INTEREST

The authors declare no conflict of interest, financial or otherwise.

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