Failure of Computed Tomography Angiography to Detect a Large Fistula Connecting the Left Anterior Descending Artery to the Left Ventricle Circumflex Artery

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April 16, 2024

Abstract

41-year-old male with no significant past medical history presented as an outpatient with atypical chest pain and had positive nuclear stress test. The patient subsequently underwent left heart catheterization, which revealed a fistula between the LAD and LV circumflex artery; however, CTA failed to show any evidence of a fistula.

Introduction:

Fistula is defined as an inappropriate connection between two body parts, such as an organ or blood vessel and another structure. More specifically, a coronary artery fistula (CAF) is an abnormal connection between a coronary artery and a major vessel or cardiac chamber.

CAFs are extremely rare, can be asymptomatic, and are often detected incidentally; thus, the true incidence remains elusive [1]. CAFs can either be congenital or acquired.

Congenital CAFs are theorized to be secondary to incomplete embryonic development, in which the sinusoids that normally communicate the great vessels and chambers of the heart with the coronary arteries are incompletely closed. Acquired CAFs can occur as a result of atherosclerosis, procedure-induced trauma, or collagen vascular disease [2].

Previous literature has demonstrated that CAFs are present in only 0.05-0.25% of patients who undergo coronary angiography and in 0.9% of patients who undergo CTA [3].

Fistulas arising from the LAD constitute only 30% of CAFs, and those draining into the left side of the heart constitute a mere 10%, which was the case illustrated herein [4].

The American College of Cardiology recommends closure of all large fistulas regardless of symptoms due to the vast potential for and severity of complications, including 'theft' from the adjacent myocardium, thrombosis and embolism, cardiac failure, atrial fibrillation, rupture, endocarditis/endarteritis, arrhythmias, ST-elevation myocardial infarction/non-ST elevation myocardial infarction, and even sudden cardiac death [5].

In this present case study, we describe a 41-year-old male with no significant past medical, surgical, or family history who presented with atypical chest pain.

The patient had a positive nuclear stress test and subsequently underwent installation of a left heart catheter, which revealed a large fistula between the LAD and the LV circumflex artery; however, subsequent CTA failed to demonstrate any evidence of a fistula being present.

We discuss the current imaging modalities and their respective advantages and limitations, in addition to providing an overview of disease management.

Case Presentation:

A 41-year-old man with no previous medical history comprising cardiovascular diseases, hypertension, oral hypoglycemic-treated diabetes, or dialysis-treated renal failure presented as an outpatient with atypical chest pain.

The patient had no known family history of heart disease or any personal history of cardiovascular procedures. Physical examination of the chest was unremarkable, and no murmurs, rubs, or gallops were detected upon auscultation. His ECG was normal as well.

The patient was sent for a nuclear myocardial perfusion stress test, which showed a small reversible perfusion defect in the basal-to-mid inferior wall, suggestive of inducible ischemia in the right coronary artery.

There was also a small, mild-intensity, fixed perfusion defect involving the mid-to-apical anterior segments. According to nuclear imaging results, the patient was a candidate for an outpatient left heart catheterization, which revealed no evidence of coronary artery disease but did show a moderate-sized fistula arising from the LAD to the LV circumflex artery (Figure-1) and luminal irregularity of RCA (Figure-2).

CTA of the heart was subsequently performed but exhibited no evidence of coronary artery disease or a fistula from the LAD or OM branches (Figure-3).

The patient was discharged with instructions to follow up as an outpatient to discuss disease management. Nevertheless, he unfortunately never returned to the clinic.

Since being diagnosed with coronary AV fistula, the patient has been following up with the DMC Sinai Grace Primary Care Center and his private cardiologist on an outpatient basis. He has had three admissions to the hospital with chest pain and shortness of breath. In 2019, the patient was admitted with a troponin level of 0.04 and with ECG findings of T wave inversions in the lateral leads. A transthoracic echocardiogram (TTE) was obtained, which showed no regional wall motion abnormalities but did indicate the interval development of moderate pulmonary hypertension and a small generalized pericardial effusion. As there was no significant increment in troponin level, no cardiac intervention was performed. At his last outpatient clinic visit with the primary care physician, the patient reported having exertional dyspnea and pedal edema but denied any chest pain, palpitations, diaphoresis, or vomiting. On physical exam, the patient had bilateral pitting pedal edema but there were no biochemical abnormalities.

Discussion:

When a fistula drains into the right side of the heart, the volume overload is occurring there, as well as in the pulmonary vascular bed, left atrium, and left ventricle. When the fistula drains into the left atrium or ventricle, volume overloading takes place in these chambers, but pulmonary blood flow does not dramatically increase. The size of the shunt is determined by the size of the fistula and the pressure difference between the coronary artery and the chamber into which the fistula drains. Most patients display no symptoms, especially during the first two decades of life, and are referred for testing due to an asymptomatic murmur; however, older patients may experience dyspnea or angina and occasionally arrhythmias. Complications that may arise include 'theft' from the adjacent myocardium, which can lead to myocardial infarction (MI), thrombosis and embolism, heart failure, atrial fibrillation, rupture, endocarditis/endarteritis, and even sudden death [4,5]. When diagnosing an AV shunt, CXR and EKG do not generally have significant value [6]. When the initial EKG is normal, treadmill or nuclear stress tests often reveal ischemic changes. 2D Doppler echocardiography and TEE may also show the site of drainage, but it is difficult to understand the detailed anatomy using these techniques. [4,5,6] Cardiac MRA is limited because the information regarding the origin and drainage

site of the fistula are less clear in comparison with those observed with CTA and conventional angiography [7]. Previously, cardiac catheterization was the first-line modality for diagnosing CAFs and was thought to have the highest efficiency for detailing the anatomy, size, origin, course, and presence of any stenosis and/or drainage; however, more recent studies have shown that CTA actually produces the best results. It has been demonstrated that the exact 3D course of the artery is difficult to obtain by conventional angiography, the drainage sites may not be as well visualized, and the invasive nature yields a higher mortality rate than CTA [7]. CTA is a relatively new imaging tool that was introduced 20 years ago for non-invasive cardiac imaging, and with the introduction of multi-detector computed tomography (MDCT), many problems related to image quality have been resolved [8]. CTA can produce high quality images that clearly delineate the cardiac chambers and coronary arteries and veins [5], in addition to having a shorter acquisition time and higher spatiotemporal resolution. The multiplanar reformation clearly demonstrates the sites of origin and termination of abnormal blood vessels, and 3D data provide a more precise overview of the heart, its vascular anatomy, and information pertaining to the diagnosis of coexistent abnormalities as compared with left heart catheterization [7]. However, limitations include less hemodynamic information, which we speculate to be the rationale for its failure to detect the fistula in our case.

According to the 2008 ACC/AHA guidelines, all large fistulas and small-to-moderate fistulas associated with myocardial ischemia, systolic/diastolic dysfunction, or ventricular dilation should undergo closure irrespective of the symptoms of lack thereof [9]. Patients diagnosed with CAFs should be educated prior to treatment. In contrast to patients with acquired heart disease, those with CAFs or adult congenital heart disease (ACHD) may have never experienced "normal" function. Cardiopulmonary exercise testing provides an objective assessment and has been shown to possess prognostic value for a wide variety of congenital conditions. There exist conflicting arguments regarding physical activity in these patients, with some suggesting restricted exercise, which will inevitably lead to a greater prevalence of obesity and other forms of heart disease. The most recent guidelines recommend safe exercise in patients across the spectrum of CAF severity, being individualized depending on patient clinical status. Self-directed activity is usually at 40-60% of the maximal exercise capacity, whereas fitness training occurs at 60-80% [9]. The treatment options for CAFs include surgery or catheter closure [5]. The main surgical options are epicardial fistula mobilization with ligation or division, arteriotomy of the dilated proximal coronary artery with suturing close to the fistula origin, and exposure of the fistulous connection and closure by direct suture or autologous pericardial patch. However, surgery is associated with a morbidity and mortality rate ranging from 0-6%, and there is a risk of MI and recurrence of the fistula [10]. Catheter closure is considered an effective and safe alternative, with the aim of occluding the fistula artery as distally and close to its termination point as possible to avoid occlusion of the branches supplying the normal myocardium [10]. There are many tools available for closure, including occlusion coils, detachable balloons, and deployable stents and balloons. The techniques and tools used for catheter closure are influenced by several different factors such as the morphology of the feeding arteries, patient age, and location of the fistula [10]. With catheter closure techniques, complete occlusion of the fistula may be achieved in >95% of patients. In the remaining patients, either further procedures are required to close the fistulas or conservative management can be undertaken if the residual fistulas are small [11]. Selection of percutaneous or surgical management is influenced by the anatomy and physiology of the CAF. Indications for surgical management are large, symptomatic, and/or tortuous fistulas with or without multiple communications and drainage; whereas, percutaneous transcatheter closure is considered when the origin of the fistula is in the proximal portion of the feeding coronary artery, is non-tortuous, or is in an older patient with high preoperative risk [7]. After closure, these patients should be placed on Coumadin, with a goal therapeutic INR of 2.5 for 6-12 months; and should be on anti-platelet drugs indefinitely.

Conclusions: The present case illustrated a rare CAF, in which the drainage is into the left circulation. CAFs are only detected in 0.05-0.25% of patients who undergo coronary angiography and left-sided drainage is seen in a mere 10% of these cases. Although current literature states that CTA is the first-line modality for the detection of CAFs and provides a more precise overview of the heart, its vascular anatomy and information pertaining to coexisting abnormalities via 3D data, we found that conventional angiography was able to detect the fistula when CTA was not. Further research should be elicited to directly compare the

sensitivity and specificity of coronary angiography with that of CTA for the detection of CAFs.

Declaration of Interest:

There was no identifiable harm or risk to the patient during the formation of this report, this was a retrospective case report and medical testing performed was strictly for diagnostic purposes. No PHI information was included in this manuscript. None of the authors had any conflicts of interest or financial incentives in the writing in this paper.

Ethics approval and consent to participate

Approved by the Institutional ethics committee. Published with written consent of the patient.

Financial support

This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Author Contributions:

SZND was responsible for conceptualization of data. DP and NK gathered data and wrote the initial draft. APM revised the manuscript and prepared figures. MGM revised and prepared the final draft.

References:

[1] P, Yelisetti R: Multiple Left Anterior Descending Coronary Artery to Left Ventricular Fistula - A Case Series and Literature Review. J. Community Hosp. Intern. Med. Perspect. 2017, 7:258-261. 10.1080/20009666.2017.1369380

[2] Abe T, Kamata K, Nakanishi K, et al.: Successful Repair of Coronary Artery-Coronary Sinus Fistula with Aneurysm in an Adult.. Ann. Cardiothorac. Surg. 1996, 61:1520-1523.

[3] Lim JJ, Jung JI, Lee BY, et al.: Prevalence and types of coronary artery fistulas detected with coronary CT angiography. AJR Am. J. Roentgenol. 2014, 203:237-43.

[4] Al-Douri A, Cedars A, Tran D, et al.: Coronary Artery Fistula between the Left Anterior Descending Artery and Pulmonary Artery.. Proc (Bayl Univ Med Cent). 2018, 31:64-66. 10.1080/08998280.2017.1401380

[5] Qureshi SA: Coronary Arterial Fistulas.. Orphanet J Rare Dis. 2006, 1:51-1764722. 10.1186/1750-1172-1-51

[6] Early SA, Meany TB, Fenlon HM, et al.: Coronary Artery Fistula; Coronary Computed Topography-The Diagnostic Modality of Choice. J. Cardiothorac. Surg. 2008, 3:41-2491619. 10.1186/1749-8090-3-41

[7] Yun G, Nam T, Chun E, et al.: Coronary Artery Fistulas: Pathophysiology, Imaging Findings, and Management.. RadioGraphics. 2008, 38:688-703. 10.1118/1.597705

[8] Liang Y, Kruger RA: Dual-Slice Spiral versus Single-Slice Spiral Scanning: Comparison of the Physical Performance of Two Computed Tomography Scanners.. Medical Physics. 1996, 23:205-220. 10.1118/1.597705

[9] 2008 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines.. (2008). Accessed: 02/01/2021: http://www.ahajournals.org/doi/full/10.1161/CIRCULATIONAHA.108.190690.

[10] Mavroudis C, Backer CL, Rocchini AP, et al.: Coronary Artery Fistulas in Infants and Children: A Surgical Review and Discussion of Coil Embolization.. Ann. Cardiothorac. Surg. 1997, 63:1235-1242. 10.1016/s0003-4975(97)00251-8

[11] Qureshi SA, Reidy JF, Alwi MB, et al.: Use of interlocking detachable coils in embolization of coronary arteriovenous fistulas.. Am J Cardiol. 1996, 78:110-113. 10.1016/s0002-9149(96)00239-1

Figure Legends:

Fig. 1 The coronary angiogram demonstrates LAD collateralization to OM branches of the LCFX finally fistulizes to the left atrium

Fig. 2 The coronary angiogram demonstrates mild luminal irregularity of RCA

Fig. 3 CTA shows no evidence of a fistula from the LAD or OM branches