EJCRIM
European Journal
of Case Reports
in Internal Medicine

LARGE CORONARY CAMERAL FISTULA TO THE LEFT VENTRICLE PRESENTING AS CONGESTIVE HEART FAILURE

Yashitha Chirumamilla, Ajit Brar, Farouk Belal, Philip McDonald

Department of Internal Medicine, Hurley Medical Center, Flint, USA

Corresponding author: Yashitha Chirumamilla **e-mail:** yashi.c527@gmail.com

Received: 08/02/2024 **Accepted:** 12/02/2024 **Published:** 28/02/2024

Conflicts of Interests: The Authors declare that there are no competing interests.
Patient Consent: The patient has given her verbal and written consent to use images and details regarding her medical condition to be published in a medical journal.

How to cite this article: Chirumamilla Y, Brar A, Belal F, McDonald P. Large coronary cameral fistula to the left ventricle presenting as congestive heart failure. *EJCRIM* 2024;**11**:doi:10.12890/2024_004364.

ABSTRACT

A 56-year-old African American female was under evaluation for coronary artery disease by a cardiologist due to her complaints of intermittent chest pain. She underwent an outpatient echocardiogram and was found to have an ejection fraction of 20–25% with global left ventricular hypokinesis. Due to this finding along with her ongoing chest pain, she was referred to the emergency department for further evaluation. Her electrocardiogram showed changes suggestive of ischaemia and her cardiac troponins were mildly elevated, so she underwent an urgent cardiac catheterisation. The angiography confirmed the reduced ejection fraction and global left ventricular hypokinesis, but also demonstrated a large coronary cameral fistula (CCF) extending from the first septal branch into the left ventricle. She was then diagnosed with non-ischaemic cardiomyopathy and heart failure with reduced ejection fraction secondary to the CCF. In this report, we illustrate a frequently encountered clinical scenario in which a patient presented with chest pain and EKG findings indicative of ischaemic cardiomyopathy. The patient also had several risk factors for coronary artery disease, however further investigation revealed an alternative diagnosis.

KEYWORDS

Coronary fistula, coronary cameral fistula, congenital coronary fistula

LEARNING POINTS

- A description of rare coronary anomalies adds to the fund of medical knowledge and can guide physicians to make evidence-based decisions regarding its management.
- Increasing description of coronary cameral fistula will alert clinicians to suspect it as a cause for worsening heart failure and as a treatable cause of non-ischaemic cardiomyopathy.

CASE DESCRIPTION

A 56-year-old African American female with a past medical history of hypertension, dyslipidaemia, obstructive sleep apnoea, chronic obstructive pulmonary disease, asthma, and chronic tobacco use. She was referred to the emergency department by her cardiologist due to complaints of intermittent chest pain. She described the chest pain to be centrally located, non-radiating and occasionally

EFIM
European Federation of Internal Medicine

Publisher
smc media srl

associated with shortness of breath. She had an outpatient echocardiogram performed due to these symptoms and the results revealed an ejection fraction of 20%–25% with global left ventricular hypokinesis, as well as mild mitral regurgitation, tricuspid regurgitation, and aortic regurgitation. Upon arrival, the patient was tachycardic with a pulse of 129 and otherwise stable vitals. She had normal breath sounds bilaterally with no audible murmur or jugular venous distension. Her electrocardiogram (EKG) revealed T-wave inversions in the anterior leads and Q-wave changes indicative of a myocardial infarction *(Fig. 1)*. Her cardiac troponins were mildly elevated at 0.140 ng/ml (reference range <0.04 ng/ml).

Due to concerning clinical findings and EKG results, a preliminary diagnosis of acute coronary syndrome was made and the protocol was promptly followed. She was given oral aspirin, started on an intravenous heparin infusion and underwent coronary angiography through a radial approach. The angiography confirmed the reduced ejection fraction and global left ventricular hypokinesis and demonstrated mild calcifications in the right coronary artery (RCA) with minimal to no calcifications in the left anterior descending (LAD) artery and left circumflex (LCX) artery. A large coronary cameral fistula (CCF) was also identified, extending from the large first septal branch into the left ventricle, and was deemed to be the culprit for the patient's symptoms *(Fig. 2)*. The LCX and RCA were found to be angiographically normal. Her left ventricular end diastolic pressure was noted to be 26 mmHg during the angiography.

She was ultimately diagnosed with non-ischaemic cardiomyopathy and new onset systolic heart failure, secondary to the presence of a CCF. She was managed with guideline-directed medical therapy for her congestive heart failure, provided with a LifeVest and scheduled to undergo an elective CCF closure.

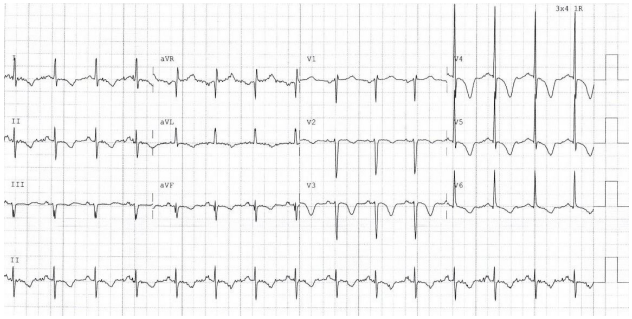

Figure 1. Electrocardiogram depicting new onset T-wave inversions in the anterior leads along with Q waves, indicative of myocardial ischaemia.

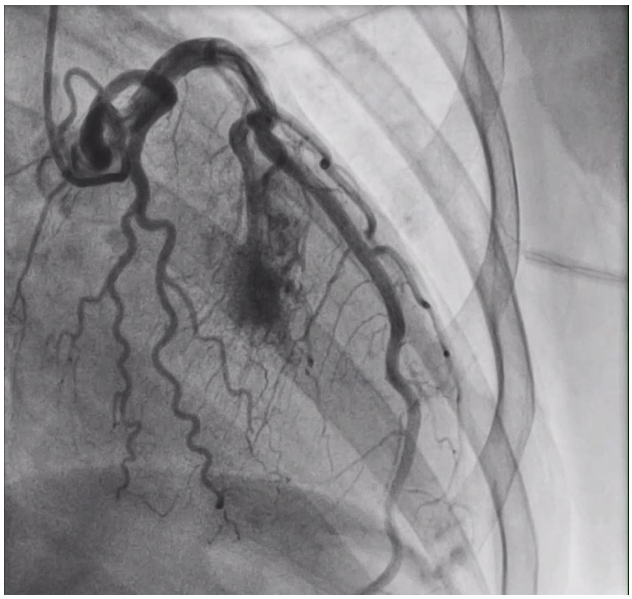

Figure 2. Cardiac catheterisation demonstrating a large coronary cameral fistula extending from the large first septal branch into the left ventricle.

DISCUSSION

Coronary anomalies were initially described by Ogden on the basis of anatomical properties after observing variations present in 224 patients[1]. Angelini updated this classification by taking into consideration the embryogenesis of coronary arteries and defined coronary anomalies to be patterns encountered in less than 1% of the cases. The need to differentiate between anatomical and pathophysiological anomalies was also stressed as the latter is more clinically relevant[2]. Dodge-Khatami et al. further established a standardised nomenclature with seven major categories for coronary anomalies, namely: anomalous pulmonary artery origins of the coronaries, anomalous aortic origins of the coronaries, congenital atresia of the left main coronary artery, coronary artery fistulas, coronary artery bridging, coronary aneurysms and coronary stenosis[3]. While congenital coronary anomalies are identified in 1% of the population, 0.2 – 0.4% are attributed to CCFs[4]. Our report aims to shed light on the clinical significance of this rare entity.

Coronary arterial fistulas are most often caused by embryological abnormalities but can rarely be due to trauma, invasive procedures, or cardiac surgeries. Congenital coronary artery fistulas are caused by abnormally persistent sinusoidal connections to cardiac structures; CCFs occur when the connection is to a cardiac chamber. The most common are the lower pressured right atrium and right ventricle at 41% and 26% respectively. The third most common is the pulmonary arteries at 16%. Less than 10% of the fistulas drain into the left atrium and left ventricle as seen in our patient[5,6].

A review study including 76 patients with coronary arteriovenous fistulas identified that 55% of them were asymptomatic while 34% presented with angina, and 13% presented with congestive heart failure[7]. CCFs which drain into the right heart chamber follow the pathway of least resistance and create left-to-right intracardiac shunts. However, CCFs that drain into the left chamber such as in our patient, develop left-to-left shunts which ultimately leads to a state of left ventricular volume overload. A large enough CCF can disrupt the normal coronary blood flow enough to cause demand ischaemia and left ventricular dysfunction

which can eventually lead to arrhythmias. A case report describes the presentation of a CCF with repetitive torsade de pointes[8]. Another case report outlines a CCF presenting with angina and new onset atrial fibrillation[9].

Management of CCFs is highly dependent on the clinical significance and patients' comorbidities. According to the ACC/AHA 2008 guidelines, for management of coronary arteriovenous fistulas, a large fistula regardless of symptoms should be closed with either transcatheter embolisation or surgical closure. Smaller or moderated sized fistulas should be managed the same way if there are accompanying complications of ventricular dysfunction, angina or arrythmias[4]. Beta blocker therapy has also been described, especially in inoperable conditions[8].

REFERENCES

1. Ogden JA. Congenital anomalies of the coronary arteries. *Am J Cardiol* 1970;**25**:474–479.
2. Angelini P. Normal and anomalous coronary arteries: definitions and classification. *Am Heart J* 1989;**117**:418–434.
3. Dodge-Khatami A, Mavroudis C, Backer CL. Congenital heart surgery nomenclature and database project: anomalies of the coronary arteries. *Ann Thorac Surg* 2000;**69**:S270–S297.
4. Mansour MK, Nagalli S. Coronary cameral fistula. [Updated 2022 Sep 19]. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2023 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK553211/
5. Kiefer TL, Crowley AL, Jaggers J, Harrison JK. Coronary arteriovenous fistulae: the complexity of coronary artery-to-coronary sinus connections. *Tex Heart Inst J* 2012;**39**:218–222.
6. Nkya DA, Sinyangwe G, Takawira FF. Transcatheter occlusion of giant congenital coronary cameral fistulae: a case series. *J Med Case Rep* 2019;307.
7. Said SA, el Gamal MI, van der Werf T. Coronary arteriovenous fistulas: collective review and management of six new cases – changing etiology, presentation, and treatment strategy. *Clin Cardiol* 1997;**20**:748–752.
8. Ghumman GM, Khan A, Shafqat M, Shah J, Singh H. A coronary cameral fistula associated with incessant ventricular arrhythmias. *Cureus* 2023;**15**:e35847.
9. Yuksel M, Yildiz A, Oylumlu M, Polat N, Acet H, Ozaydogdu N. Three vessel coronary cameral fistulae associated with new onset atrial fibrillation and angina pectoris. *Case Rep Vasc Med* 2014;**2014**:475325.