

Coronary Cameral Fistula

NIRMAL KUMAR MOHANTY¹, BIJAY KUMAR DASH², CHHABI SATPATHY³,
SATYA NARAYAN ROUSTRAY⁴, SATYASARATHI JENA⁵



ABSTRACT

Coronary Cameral Fistulas (CCFs) are rare arteriovenous malformations, defined as a connection between a coronary artery and a cardiac chamber or any segment of the systemic or pulmonary circulation. These may be congenital or acquired, and show a low incidence in both angiographic studies, and the general population. The presentation of CCFs varies depending upon their size and location. These can be majorly asymptomatic or show symptoms and complications like congestive heart failure, myocardial infarction, and bacterial endocarditis. Although percutaneous closure with embolisation can be done, surgical closure of CCF is a gold standard of treatment. The authors reported a case of a 57-year-old male patient who presented with exertional chest pain and dyspnoea. Coronary angiography revealed the presence of dilated tortuous Left Main Coronary Artery (LMCA), Left Anterior Descending artery (LAD), Left Circumflex artery (LCX) and CCF between LAD to Right Ventricle (RV), Obtuse Marginal (OM) to RV and Posterior Left ventricular Branch (PLB) to RV. No significant obstructive coronary artery disease was present. Coronary artery calcium score was 15. The patient was managed conservatively with antiplatelets, statin, and heparin and responded well to the treatment.

Keywords: Congestive heart failure, Coronary angiography, Myocardial infarction, Right ventricle

CASE REPORT

A 57-year-old male diabetic, non hypertensive, chronic smoker patient presented with exertional chest pain and breathlessness of three days duration in the Cardiology Outpatient Department (OPD). He had one episode of syncope during this period, which lasted for a few seconds with spontaneous and complete recovery. Chest pain was not associated with orthopnoea, Paroxysmal Nocturnal Dyspnoea (PND), palpitation or swelling of face or legs. The patient had type 2 diabetes with good glycemic control (Glycated hemoglobin (HbA1c) 6.9%) and was on oral antidiabetic drugs since 2008.

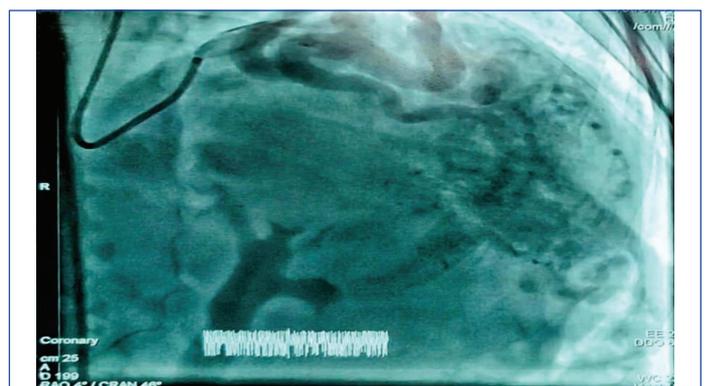
On examination, the pulse rate was 58 beats per minute (bpm), blood pressure was 110/70 mmHg. All peripheral pulsations were well felt. Heart sounds were normal. There was an ejection systolic murmur of grade II/VI in pulmonary area. Electrocardiogram (ECG) showed sinus bradycardia with ST depression in inferior and lateral leads (LII, LIII, augmented Vector Foot and augmented Vector Left (aVL), LI, V4 to V6). Echocardiography revealed the presence of mildly dilated Right Atrium (RA) and Right Ventricle (RV), ostium secundum-Atrial Septal Defect (os-ASD) of 7 mm size with a left to right shunt, interatrial septal aneurysm, mild Pulmonary Arterial Hypertension (PAH) with Tricuspid Regurgitation (TR). No Regional Wall Motion Abnormality (RWMA) was detected. Troponin-I was raised to nearly twice the upper limit of normal. A clinical diagnosis of Non ST Elevation Myocardial Infarction (NSTEMI) was made and the patient was managed with aspirin 150 mg daily, clopidogrel 75 mg daily, atorvastatin 40 mg daily and heparin (fondaparinux)-2.5 mg subcutaneous daily.

Elective coronary angiography was done on the third day of admission, which showed dilated Left Main Coronary Artery (LMCA) and dilated tortuous Left Anterior Descending (LAD) and Left Circumflex Artery (LCX) with fistulous connection between LAD and RV, Obtuse Marginal (OM1) and RV and Posterior Left ventricular Branch (PLB) and RV. Right Coronary Artery (RCA) was only mildly dilated [Table/Fig-1-3]. For better delineation of coronary anatomy, CT-coronary angiography was done. This showed dilated LMCA, dilated tortuous LAD and LCX and their branches. LAD was divided into two nearly similarly calibre anterior (diagonals) branch with a fistulous connection with posterior

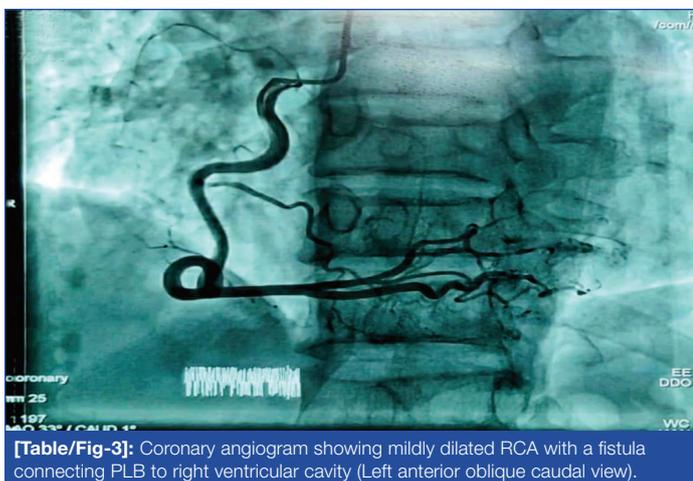
aspect of RV. A thin communication between PLB and adjacent posterior aspect of RV was seen, suggestive of Coronary Cameral Fistula (CCF) [Table/Fig-4,5]. LA and RA appeared dilated. Main pulmonary artery was dilated. A saccular out pouching of interatrial septum from LA to RA was seen (aneurysm). A small ASD was also seen. Coronary artery calcium score was 15 (mild degree of coronary artery disease). The patient was discharged three days later, on dual



[Table/Fig-1]: Coronary angiogram showing dilated LMCA and dilated tortuous LAD and LCX with a fistula connecting LAD and OM1 to right ventricular cavity (Right anterior oblique-caudal view).



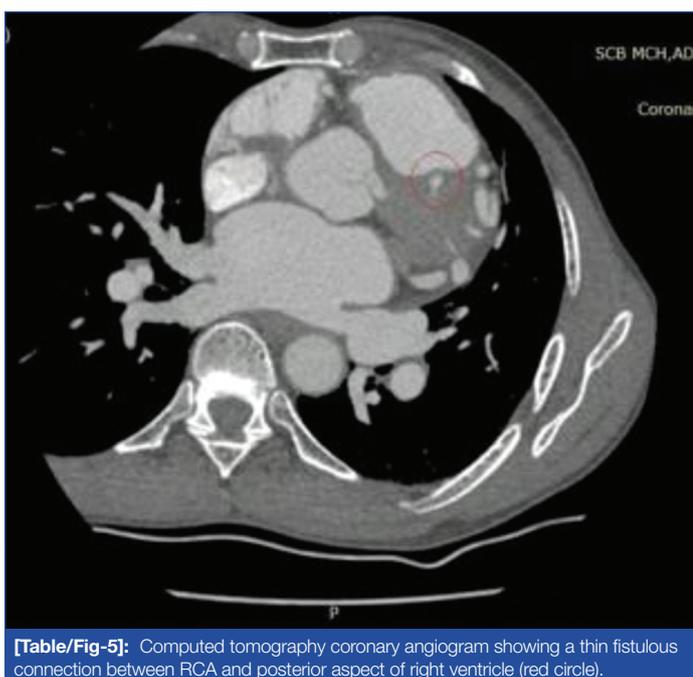
[Table/Fig-2]: Coronary angiogram showing dilated LMCA and dilated tortuous LAD and LCX with a fistula connecting LAD and OM1 to right ventricular cavity (Right anterior oblique-cranial view).



[Table/Fig-3]: Coronary angiogram showing mildly dilated RCA with a fistula connecting PLB to right ventricular cavity (Left anterior oblique caudal view).



[Table/Fig-4]: Computed tomography coronary angiogram showing a fistulous connection between LAD and right ventricular cavity (red circle).



[Table/Fig-5]: Computed tomography coronary angiogram showing a thin fistulous connection between RCA and posterior aspect of right ventricle (red circle).

DISCUSSION

Coronary artery anomalies include anomalies at the origin, during its course and at the site of termination. Fistulae result from its abnormal communication at the site of termination. Broadly, they are classified as Coronary Arterio-venous Fistula (CAVF) when the communication is between a coronary artery or its branches and any part of the systemic or pulmonary vasculature. Coronary cameral fistula is the abnormal communication between a coronary artery and any cardiac chamber [1]. In this case, all three major coronary arteries were involved and were draining to the right ventricle, the most common chamber to be involved [1].

The pathophysiology of fistula relates to the amount of blood flowing through it and its pressure gradient. Myocardial ischaemia may be due to decreased flow of blood through the coronary segment distal to the site of fistulous connection due to coronary steal phenomenon. If fistulae are long and multiple, the intracoronary diastolic perfusion pressure could drop below the critical level and during physical activity, leads to increased myocardial oxygen demand causing myocardial ischaemia [2].

Clinical presentation is dependent on the haemodynamic status of the anomaly. Factors like size the fistula, the resistance of the receiving chamber and myocardial ischemia influence the clinical outcome [3]. Most fistulae are small and clinically silent. However, common symptoms are exertional dyspnoea, fatigue, angina pain and sometimes congestive heart failure [4]. Clinically the cardiovascular examination findings are unremarkable in presence of haemodynamically insignificant fistula. The mechanism of symptoms is due to coronary steal phenomenon and diastolic overload [5,6].

Volume overload of cardiac chambers may cause congestive heart failure and arrhythmias. Other complications, like, intravascular thrombosis, infective endocarditis may occur [7]. The present patient had presented with exertional chest pain with dyspnoea. Though coronary angiography showed CCF involving all three major coronary artery branches, there was no significant obstructive lesion. So the anginal pain could be due to coronary steal phenomenon, which is due to the diversion of blood through the fistulous tract causing myocardial ischemia distal to it. In a study by Wilde P and Watt I, 57.4% were asymptomatic, 24.2% had dyspnoea and 18.7% with chest pain [8]. Said SA et al., showed the symptoms of fistula draining to left ventricle could be similar to the signs and symptom of aortic regurgitation [7].

Chest X-ray and Electrocardiography (ECG) changes and even echocardiology may not be of much help in the diagnosis of coronary artery fistula. No specific ECG changes point towards a coronary fistula. Chest radiographs may show cardiomegaly in the presence of significant shunt flow and ECG may reveal the effect of volume overload in larger fistulas. But these are non specific. Echocardiography may only detect dilated coronary artery ostium, not the entire course of the coronary arteries. Intraventricular turbulence may sometimes raise a suspicion of fistulous tract, which can be confirmed by an angiographic study [9].

Coronary angiography establishes the diagnosis of coronary artery fistula. In addition, the severity of atherosclerotic coronary artery disease is also simultaneously diagnosed. Presently, Multidetector Cardiac Computed Tomography (MDCT) is widely used as it is non invasive and it provides three dimensional visualisations making better delineation of coronary anatomy. Lim JJ et al., have shown MDCT to be better in detecting coronary anomalies than traditional CT [10].

Treatment of coronary cameral fistula depends on its hemodynamic significance. Those fistulae being clinically silent without other coexisting abnormalities do not require further treatment. Various treatment options include surgical or catheter closure. Surgery is

antiplatelet therapy with aspirin and clopidogrel as well as statin and Angiotensin-converting Enzyme (ACE) inhibitors. At 1 month follow-up, he remained asymptomatic.

associated with low mortality and morbidity. Most of the cases described in past, concern the surgical correction of primary coronary cameral fistula of congenital aetiology [11,12].

Transcatheter closure has become the method of choice though surgical intervention gives good result [13]. Percutaneous techniques like interlocking detachable coils, detachable balloons, polyvinyl alcohol foam and amplatzer duct occluder are other treatment options. Device closure is having its own complications like myocardial infarction and migration of closure devices to extracoronary vascular structures or elsewhere within coronary branches [14].

More recently, Yu X et al., [15] described the spontaneous closure of a secondary RCA-RV fistula, and Mertens A et al., [16] reported the case of an acquired LAD-RV fistula treated with coil embolization. The index case was reviewed at the multidisciplinary team meeting by interventional cardiologist, cardiothoracic surgeon and radiologist and was concluded that serial imaging with annual echocardiography and regular clinical review would be the best approach for the patient and that there was no current indication for invasive management.

CONCLUSION(S)

Coronary cameral fistula, mostly an incidental finding while doing coronary angiography is many a times asymptomatic. Now-a-days, it is detected while doing CT angiography for the diagnosis to 'rule out' coronary artery disease. Serial follow-up for asymptomatic CCF is required and definite therapeutic intervention either with surgery or catheter based techniques is required for symptomatic fistulae. Its association with ostium secundum ASD is very rare as in the present case. CCF needs serial follow-up and treatment when symptoms of myocardial ischemia or congestive cardiac failure warrants active intervention.

REFERENCES

- [1] Sagar D, Hernandez A, Heimowitz T. Coronary artery-Left ventricle fistula: A case report of a rare connection error! *Cureus*. 2015;7(4):e266. PMID: 26180690 PMID: PMC4494526. Doi: 10.7759/cureus.266.
- [2] Heper G, Kose S. Increased myocardial ischemia during nitrate therapy: Caused by multiple coronary artery-left ventricle fistulae? *Tex Heart Inst J*. 2005;32(1):50-52. PMC555822.
- [3] Majidi M, Shahzamani M, Mirhoseini M. Clinical features of coronary artery fistula. *J Tehran Heart Cent*. 2011;6(3):158-62.
- [4] Kugelmass AD, Manning WJ, Piana RN, Weintraub RM, Baim DS, Grossman W. Coronary arteriovenous fistula presenting as congestive heart failure. *Cathet Cardiovasc Diagn*. 1992;26(1):19-25.
- [5] Mangukia CV. Coronary artery fistula. *Ann Thorac Surg*. 2012;93(6):2084-92.
- [6] Sommer RJ, Hijazi ZM, Rhodes JF Jr. Pathophysiology of congenital heart disease in the adult: Part I: Shunt lesions. *Circulation*. 2008;117(8):1090-99.
- [7] Said SA, Schiphorst RH, Derksen R, Wagenaar LJ. Coronary-cameral fistulas in adults: Acquired types (second of two parts). *World J Cardiol*. 2013;5(12):484-94.
- [8] Wilde P, Watt I. Congenital coronary artery fistulae: Six new cases with a collective review. *Clin Radiol*. 1980;31(3):301-11.
- [9] Mansour MK, Nagalli S. Coronary Cameral Fistula. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing; 2022.
- [10] Lim JJ, Jung JI, Lee BY, Lee HG. Prevalence and types of coronary artery fistulas detected with coronary CT angiography. *AJR Am J Roentgenol*. 2014;203(3):W237-43.
- [11] Abrams LD, Evans DW, Howarth FH. Coronary artery-right ventricular fistula treated surgically. *Br Heart J*. 1967;29(1):132-34.
- [12] Morgan JR, Forker AD, O'Sullivan MJ, Fosburg RG. Coronary arterial fistulas: Seven cases with unusual features. *Am J Cardiol*. 1972;30(4):432-36.
- [13] Armsby LR, Keane JF, Sherwood MC, Forbess JM, Perry SB, Lock JE. Management of coronary artery fistulae. Patient selection and results of transcatheter closure. *J Am Coll Cardiol*. 2002;39(6):1026-32.
- [14] Minhas AM, Ul Haq E, Awan AA, Khan AA, Qureshi G, Balakrishna P. Coronary-cameral fistula connecting the left anterior descending artery and the first obtuse marginal artery to the left ventricle: A rare finding. *Case Rep Cardiol*. 2017;8071281. Doi: 10.1155/2017/8071281.
- [15] Yu X, Wang X, Zhang R, Xu F, Ji F. Spontaneous closure of an iatrogenic coronary artery fistula during recanalization of a chronic total occlusion lesion: A case report. *Medicine*. 2019;98(3):e14068.
- [16] Mertens A, Dalal P, Ashbrook M, Hanson I. Coil embolization of coronary-cameral fistula complicating revascularization of chronic total occlusion. *Case Rep Cardiol* 2018;6857318. Doi: 10.1155/2018/6857318.

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Cardiology, Srirama Chandra Bhanja Medical College, Cuttack, Odisha, India.
2. Assistant Professor, Department of Cardiology, Srirama Chandra Bhanja Medical College, Cuttack, Odisha, India.
3. Associate Professor, Department of Cardiology, Srirama Chandra Bhanja Medical College, Cuttack, Odisha, India.
4. Professor, Department of Cardiology, Srirama Chandra Bhanja Medical College, Cuttack, Odisha, India.
5. Senior Resident, Department of Cardiology, Srirama Chandra Bhanja Medical College, Cuttack, Odisha, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Satyasarithi Jena,
Senior Resident, Department of Cardiology, Srirama Chandra Bhanja Medical College,
Cuttack, Odisha, India.
E-mail: satya.sarithi@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Apr 27, 2022
- Manual Googling: May 25, 2022
- iThenticate Software: Aug 04, 2022 (17%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Apr 10, 2022**

Date of Peer Review: **May 25, 2022**

Date of Acceptance: **Aug 05, 2022**

Date of Publishing: **Nov 01, 2022**