

Coronary Artery Fistula As An Unexpected Cause of Heart Failure: A Case Report

Andrianto, Alqi Yutha, Primasitha Maharany Harsoyo

Department of Cardiology and Vascular Medicine, Faculty of Medicine Universitas Airlangga – Dr. Soetomo General Hospital, Surabaya, East Java, Indonesia

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SOUHRN

Koronární píštěl (coronary artery fistula, CAF) je nepříliš často se vyskytující spojka mezi epikardiální částí koronární tepny na jedné straně a buď srdečním oddílem (anglicky coronary cameral fistula, CCF), nebo s velkou srdeční cévou, například plicními tepnami nebo žilami na straně druhé. Protože je většina pacientů v prvních dvou dekádách života asymptomatických, zůstává tato anomálie dlouhodobě skryta; symptomů a komplikací začíná přibývat až po třetí dekádě života. Soudí se, že primárním patofyziologickým problémem u koronárních píštělí je jev označovaný jako koronární steal syndrom. Symptomy závisejí na rozsahu steal („vykrádání krve určené pro srdce“) nebo shuntu. Popisujeme případ 70letého muže se symptomy srdečního selhání, u něhož byla následně pomocí angiografického vyšetření stanovena diagnóza koronární píštěle.

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ABSTRACT

Coronary artery fistulae (CAF) are uncommon epicardial connections between coronary vessels and either a heart chamber, known as coronary cameral fistula (CCF), or the major vessels of the heart, such as pulmonary arteries or veins. Most patients remain asymptomatic in the first two decades, thus making their presence all the more concealed. Following the third decade, occurrence of both symptoms and complications rises. The primary pathophysiological issue in coronary fistulae is believed to be the coronary steal phenomenon. The symptoms experienced are contingent on the extent of steal or shunt. We report the case of a 70-year-old man presenting with symptoms of heart failure, subsequently diagnosed with a coronary artery fistula through angiography.

Introduction

Coronary artery fistulae (CAF) are uncommon epicardial connections between coronary vessels and either a cardiac chamber, known as coronary cameral fistula (CCF), or the major vessels of the heart, such as pulmonary veins or arteries. Coronary artery fistulae (CAF) constitute a rare and major coronary anomaly, found in approximately 0.1% of the general population, accounting for 0.4% of all congenital cardiac malformations. Typically, these anomalies are incidentally discovered during coronary angiography. Most patients remain asymptomatic in the first two decades, thus making their presence all the more concealed. Following the third decade, occurrence of both symptoms and complications rises. The primary pathophysiological issue in coronary fistulae is believed to be the coronary

steal phenomenon. The symptoms experienced are contingent on the extent of steal or shunt. We present the case of 54-year-old man with heart failure symptoms who was later found to have a coronary artery fistula on angiography.

Case report

A male patient, aged 70 years, presented at the clinic with complaints of fatigue and shortness of breath during moderate to vigorous activity over the last few months. These complaints manifest during activities such as ascending and descending stairs and diminish with rest. The patient has a history of diabetes mellitus and atrial fibrillation. Despite the current therapy, the patient did not experien-

Address: Alqi Yutha, MD, Department of Cardiology and Vascular Medicine, Faculty of Medicine Universitas Airlangga – Dr. Soetomo General Hospital, Jl. Mayjend Prof. Dr Moestopo No 4-6, Surabaya, East Java 60285, Indonesia

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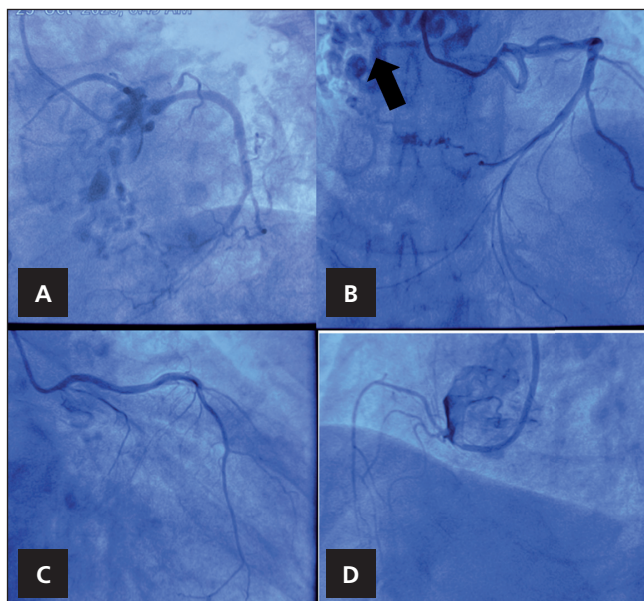


Fig. 1 – Coronary angiography. (A) LM; (B) LCx accompanied by fistula (black arrow) from proximal segment; (C) LAD; (D) RCA.

ce significant improvement in their symptoms, prompting them to seek a change in healthcare providers. Transthoracic echocardiography revealed a reduction in left ventricular function with global hypokinesis (EF 40%). Valves were within normal limits, and no abnormal color jet turbulence was observed. Subsequently, the patient received a diagnosis of ischemic cardiomyopathy (ICM). Following that, the patient underwent a cardiac catheterization examination, revealing challenges in evaluating the left main (LM). The left anterior descending (LAD) was found to be normal, while the left circumflex (LCx) was dominant and exhibited a fistula to a major blood vessel. Additionally, the right coronary artery (RCA) was noted to be small (Fig. 1).

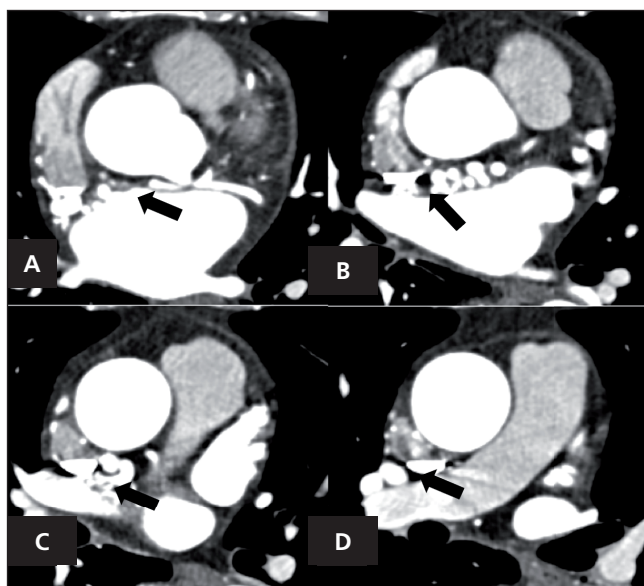


Fig. 2 – Sequential CTA (A–D) images showing a fistula (black arrow) extending from the LCx towards the RPA.

For a more detailed view of the coronary anatomy and fistula, a computed tomography angiography (CTA) examination was performed (Fig. 2). The results indicated left coronary artery dominance, a short LM, mild stenosis in the LAD, and a vascular malformation with the presence of a fistula extending from the LCx to the right pulmonary artery (RPA).

The patient then also underwent an MRI examination with results of late gadolinium enhancement (LGE) non specific mid myocardial fibrosis at basal septum with regional hypokinesia of the septum. MRI impression as non ischemic cardiomyopathy with a global reduced left ventricle ejection fraction (EF) of 39.8%. In conclusion, patients that previously diagnosed with Ischemic Cardiomyopathy (ICM) exhibiting symptoms of heart failure may not necessarily have coronary artery disease. Instead, the patient's symptoms seem to be attributed to a coronary steal phenomenon induced by coronary arteriovenous fistula (CAF). Subsequently, a transcatheter coiling closure procedure was carried out. Upon evaluation three months post-coiling, the patient reported an improvement in symptoms, and there was an observed increase in the EF to 45%.

Discussion

A fistula of coronary artery is characterized by the formation of an abnormal connection between the coronary arteries of the heart and either the pulmonary or systemic circulation. The term "coronary-cameral fistula" specifically refers to the communication between the coronary arteries and heart chambers.¹ Although most coronary fistulas are congenital anomalies, acquired fistulas have been documented in cases of trauma, as complications arising from coronary artery bypass surgery, and in certain instances of Takayasu's arteritis.²

Coronary artery fistula's incidence is often underreported, with some studies suggesting it to be less than 1%. Said et al. observed that 69% of unilateral fistulae originated from the left coronary artery (LCA), while 31% originated from the RCA. Among the LCA-originating fistulae, approximately 29% were traced back to the left circumflex artery (LCx). Notably, only 4% of LCx-originating fistulae terminated in the left ventricle, whereas the majority (81%) concluded in the pulmonary artery/right atrium/right ventricle, aligning with the findings in our case.³

Coronary artery fistulas typically remain asymptomatic during the initial two decades, particularly if they have a minor hemodynamic impact. However, beyond this timeframe, the frequency of symptoms and complications tends to increase.⁴ The hemodynamic consequences are contingent upon factors such as the pressure gradient across it, the volume of shunt flow, and the size of the fistula.¹ While the majority of fistulas is small and does not induce significant alterations, a sizable fistula with an excessive flow into the right heart has the potential to cause volume overload, leading to pulmonary hypertension. The redirection of blood through a fistula can also trigger a coronary steal syndrome/phenomenon, resulting in a reduction of blood flow to the myocardium

and leading to myocardial ischemia with accompanying symptoms.⁵

Complications associated with coronary artery fistulas encompass atrial fibrillation, heart failure, thrombosis and embolism, endocarditis/endarteritis, aneurysm, and fistula rupture, arrhythmia, and sudden death. Although thrombosis within the fistula is uncommon, it has the potential to precipitate acute myocardial infarction.⁴ Moreover, there have been reported instances of the spontaneous rupture of an aneurysmal fistula, resulting in hemopericardium.⁶

A study conducted by Said and Werf in 2007 included 13 patients experiencing angina pectoris in the absence of coronary heart disease (CHD), with seven of them exhibiting objective evidence of coronary insufficiency. The research highlighted that increased myocardial work during exercise leads to microvasculature dilation, up to 4-5 times, to augment coronary blood flow, known as coronary flow reserve (CFR). In CAF-Associated Angina, there is an elevated basal coronary blood flow, resulting in decreased CFR. At rest, myocardial perfusion is sufficient. However, during exercise, the myocardium may experience inadequate perfusion because CFR cannot be increased. The study clarifies a continuous flow pattern characterized by higher velocity during the systole phase compared to diastole in the CAF originating from the LAD coronary artery to the pulmonary artery. This observation was made after assessing the flow velocity within the coronary arteries and the fistula. A high average peak flow velocity (APV) at baseline was found by intracoronary Doppler flow monitoring, along with a loss of CFR.⁷

Following successful percutaneous transcatheter embolization (PTE) from the right coronary artery (RCA) to the fistula-associated angina (FAK), CFR could recover to normal, accompanied by a reduction in baseline APV. As a result, the study concludes that persistently elevated basal coronary flow, even in the absence of coronary artery narrowing, has adverse effects on CFR. This condition can potentially lead to angina pectoris in individuals with relatively normal coronary artery conditions. Moreover, in patients with CAF accompanied by mild coronary stenosis, angina symptoms may develop prematurely.^{7,8}

Physical examinations often reveal a continuous murmur, the degree and location of which can vary significantly depend on the position and size of the coronary fistula. Many patients, however, may have a normal physical examination. Large fistulas with substantial flow/shunts may manifest signs and symptoms of early congestive heart disease. Angina, exertional dyspnea, syncope, and palpitations are among the various symptomatic manifestations.⁹

Doppler echocardiography-2D proves highly beneficial in revealing the dilatation of coronary arteries that are impacted, and color flow mapping is a useful tool for locating drainage. However, determining the anatomical details of the fistula can be challenging with this technique. In color Doppler flow imaging, significant flow can be observed at the origin or even along the blood vessels, with the flow into the right heart chamber it can also be visualized.¹⁰

Cardiac catheterization and angiography serve as methods for evaluating fistulas. Catheterization is em-

ployed to assess the hemodynamics of the fistula, while cardiac catheterization is the preferred modality for visualizing the structure and flow patterns of the coronary arteries. In most cases, intracardiac pressure is normal, and shunt flow is typically small. Using selective coronary arteriography or aortography, the anatomy of the fistula can be examined in more detail, providing information about its size, origin, course, presence of stenosis, and drainage sites.¹¹

Although coronary angiography remains the gold standard for coronary artery imaging, there are situations in which it may not always be clear where the fistula originated or how it relates to the surrounding cardiac structures. The ability to observe and measure abnormal tortuous vessels with angiography in a single plane can be challenging. In such cases, noninvasive methods such as CT and MRI can be employed to complement coronary angiography.¹² CT can offer high-resolution anatomical images through ECG-gated reconstruction. These images can reveal an enlarged fistula and assess for thrombus formation or aneurysmal dilatation in the blood vessels. Three-dimensional CT volume-rendered pictures offer a thorough view of the vascular and heart anatomy. CT is regarded as a beneficial alternative for coronary angiography. However, it's important to note that one of the main limitations of CT is radiation exposure.¹³

In this particular case, echocardiography failed to detect a fistula, while a CT examination revealed a clear image of a coronary artery fistula. CT enables excellent anatomical imaging with high resolution, facilitating the identification of the fistula's origin, course, location, and diameter. The discovery of a coronary artery fistula in this case was unexpected, aligning with existing literature that often characterizes this lesion as being diagnosed incidentally.¹⁴

Therapeutic intervention, such as transcatheter closure (TCC) or surgical ligation is typically undertaken in patients with symptomatic fistulas or those experiencing complications. These complications may include left ventricular volume overload, left ventricular dysfunction, large left-to-right shunting, congestive heart failure, myocardial ischemia, and efforts to prevent endocarditis or endarteritis.¹⁵

The preferred method for closing a fistula is percutaneous transcatheter closure, as it is a less invasive procedure that can help avoid potential surgical complications. However, in cases where the anatomy of the fistula is not conducive to transcatheter closure, especially if the fistula is tortuous or large in size, surgical closure is recommended.¹⁴ Presently, there is no unanimous agreement on the treatment of asymptomatic patients. However, elective closure of fistulas is advised in children aged older than 3 to 5 years due to the heightened risk of complications in this age group. Additionally, elective closure is recommended for cases in which fistulas arise from the proximal segments of the coronary arteries. This recommendation is based on the susceptibility of these fistulas to aneurysm formation and rupture.¹⁶

Having a comprehensive understanding of the fistula's anatomy is critical when considering closure via transcatheter. The fistula's body usually acts as the designated landing zone for the selected closure device. It is impera-

tive to avoid specific sites, such as the fistula ostium, the fistula termination, or the native coronary vessel. Landing the device in or interfering with the native coronary vessel can lead to complications such as *in situ* thrombosis, spasm, or dissection. The choice of closure devices is determined by the size of the fistula. Hydrophilic vascular coils are usually the preferred initial option when dealing with fistulas that have a diameter of less than 5 mm. In cases where the fistula measures more than 5 mm in diameter, the use of patent ductus occluders or vascular coils is a common practice. This careful consideration of anatomy and device selection is crucial for the success and safety of the transcatheter closure procedure.

Conclusion

A coronary artery fistula is an anomaly of the coronary arteries that can be either congenital or acquired. While most patients may remain asymptomatic, large and high-flow fistulas can lead to symptomatic presentations and even life-threatening complications. Imaging modalities, such as contrast-enhanced CT-angiography, can accurately depict the anatomy of the fistula, although the gold standard for diagnosis remains diagnostic coronary catheterization. In the management of symptomatic patients, transcatheter fistula closure is a preferred treatment modality. The techniques employed should be appropriate for the size and location of the coronary artery fistula, taking into consideration the patient's size. The successful closure of coronary artery fistulas through transcatheter interventions is crucial in addressing symptoms and preventing potential complications.

Conflict of interest

No conflict of interest.

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