

Congenital Coronary Artery Anomaly in an Adult: Double Barrel Left Coronary System and Coronary Artery Fistula Connecting a Giant Left Circumflex Artery to the Coronary Sinus

Mary Michelle Z. Peñaranda, M.D.*; Jun Maximo F. Lasco II, M.D.**; and Eduardo I. Lee, M.D.***

Abstract

Introduction: Coronary anomalies are rare congenital anatomic patterns affecting approximately one percent of the general population. Most of the cases may be asymptomatic and are never detected. Rarely, they result in severe life threatening consequences, thus, the diagnostic challenge is recognizing the abnormality early. Cardiac imaging modalities such as echocardiography, coronary computed tomography angiography (CTA), and coronary angiography can help us in the diagnosis.

Case: A 49-year-old female who initially presented with dyspnea and managed as rheumatic heart disease and hypertension. An echocardiogram finding prompted further investigation with coronary CTA and coronary angiography which identified the coronary artery anomalies. She was advised surgery but was unable to comply. Accordingly, medical management of her heart failure symptoms was pursued. She eventually succumbed to multiorgan failure as the complications of the fistula set in, three years after she had her initial presentation.

Discussion: The normal coronary anatomy consists of the the right and left coronary arteries that originate from the

aorta. Our patient has two coronary artery anomalies: one is the double barrel left coronary system - her left anterior descending artery and left circumflex artery (LCX) originate directly from the aorta instead of from the left coronary artery. Second, is the coronary artery fistula (CAF), with her LCX draining into the right atrium through the coronary sinus. CAFs tend to grow with age, and if untreated, can cause clinical symptoms in older patients. The consequences of the patient's abnormal connection may have caused volume overload to the right sided chambers resulting to heart failure. In adults with a congenital heart disease such as the CAF, it is recommended that if a continuous murmur is present, it should be defined either by echocardiography, magnetic resonance imaging, CTA, or cardiac catheterization.

Conclusion: A high level of suspicion for a CAF as a cause of an adult patient's progressive heart failure can lead to timely diagnosis and optimal interventions.

Keywords: case report, coronary artery fistula, heart failure, echocardiography, coronary angiography, coronary computed tomography angiography

Introduction

Coronary artery anomalies (CAAs) are a diverse group of congenital disorders defined as rare anatomic patterns of the coronary artery that are generally asymptomatic, but with some cases causing symptoms of angina, dyspnea, or sudden cardiac death.^{1,2} These cases are exceptionally rare in the adult population, with some cases that are never detected, thus, their true incidence is highly speculative.³

An anomaly of the coronary artery origin coexisting with an anomaly on its termination, as in the case of our

* Medical Resident, Department of Internal Medicine, Cebu Velez General Hospital, Cebu City, Philippines

** Adult Cardiology/Interventional Cardiology, Chong Hua Hospital, Cebu City, Philippines

*** Department of Internal Medicine, Cebu Velez General Hospital, Cebu City, Philippines

Corresponding author: Mary Michelle Z. Peñaranda, M.D., Cebu Velez General Hospital, Cebu City, Philippines
Email: mmzpenaranda@gmail.com

patient, has not been reported in all the literature we have exhaustively searched.

This is a case of a 49-year-old woman who presented with progressive heart failure symptoms that was brought about by a double barrel left coronary system and a fistula from the left circumflex artery to the coronary sinus draining into the right atrium. She was advised surgical repair but was not able to comply. We are reporting this case to highlight the diagnostic modalities that can help us identify these cases earlier, prompting timely interventions. This study also presents end stage heart failure as a clinical outcome of a coronary artery fistula.

Case Presentation

Three years prior to her most recent admission, the patient was brought for consultation due to cough and shortness of breath upon minimal exertion and upon auscultation a

continuous murmur was noted. An echocardiogram was then taken which showed mild regurgitation, mild mitral stenosis, concentric left ventricular hypertrophy, dilated left atrium and mild pulmonary hypertension. She was then diagnosed with heart failure and rheumatic heart disease. She was started on losartan and furosemide+potassium chloride tablet, for which she noted an improvement in her shortness of breath. A repeat echocardiogram was taken nine months later and results showed valvular heart disease: moderate mitral regurgitation, with no significant interval change. Losartan was shifted to irbesartan and the patient noted resolution in her shortness of breath.

Two years prior to her most recent admission, patient had no shortness of breath. Follow-up check-up showed atrial fibrillation with moderate ventricular response. She was given amiodarone and warfarin tablets and a repeat echocardiography showed no significant change. Irbesartan was continued.

In the interim, patient felt well, was compliant with her medications, and regular check-ups for protime monitoring. Three months prior to her most recent admission, cough and shortness of breath recurred. Chest x-ray posterioranterior (PA) view showed marked cardiomegaly with a prominent pulmonary segment without signs of failure. Ivabradine, cefuroxime, and montelukast+levocetirizine were added to her medications.

Two months prior to her recent admission, she reported worsening shortness of breath on exertion prompting admission. She was noted to be slightly tachypneic with bilateral rales. Chest X-ray PA view showed cardiomegaly, signs of congestion, and pneumonia. (Figure 1)

A repeat 2D echocardiography with Doppler was taken which showed multichamber dilatation with preserved left ventricular systolic function with ejection fraction of 61% and a preserved right ventricular systolic function; a dilated left aortic cusp; with the presence of a rounded structure in the superior portion of the right atrium close to the interatrial septum; consideration was an intracardiac cyst and a vascular structure due to the presence of occasional doppler color flow in some views. Severe pulmonary hypertension with estimate pulmonary artery systolic pressure of 80mmHg by TR jet and a dilated main pulmonary artery trunk were also noted.

To confirm the echocardiography findings that was suggestive of a fistula, coronary computed tomography angiography (CTA) (Figure 2) was done which showed the following findings. (Table 1)

In the institution, the patient was managed as a case of community acquired pneumonia, moderate risk, and right sided heart failure. Patient was transferred to another

hospital to confirm the suspected coronary artery anomaly and in anticipation of a surgical period, A coronary arteriography was done revealing findings summarized in Table 1 and Figure 3.

She was referred to a thoracic and cardiovascular specialist for surgical eligibility, however, it was the opinion

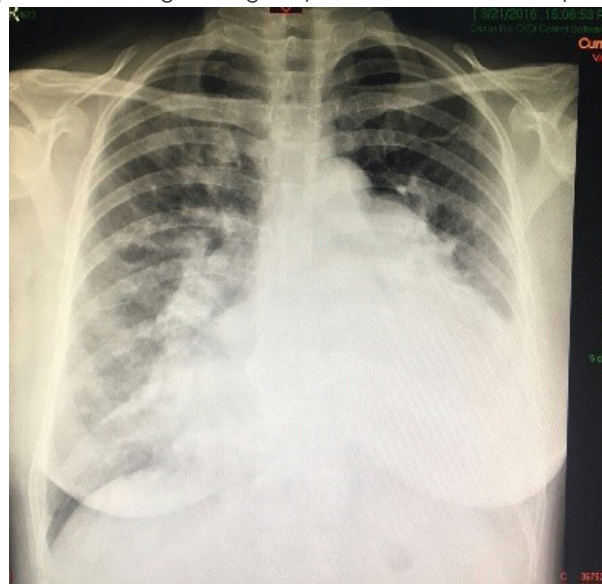


Figure 1. Posteroanterior chest radiograph shows cardiomegaly with accentuated pulmonary vascular markings and inflammatory process at the right lung and left lower lobe

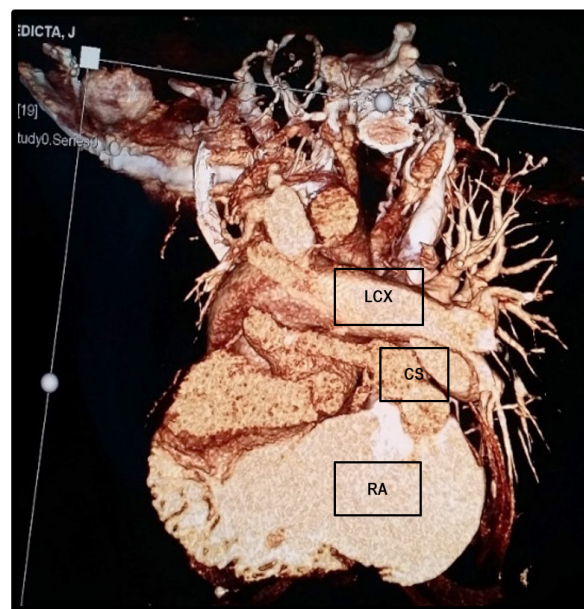


Figure 2. Coronary computed tomography angiography posteriorbasal view dilated right atrium (RA); coronary sinus (CS); left circumflex artery (LCX)

Table 1. Coronary computed tomography angiography findings

1. Cardiomegaly with gross enlargement of the right atrium
2. Tortuous and serpiginous left circumflex artery arising from the left coronary cusp of the aorta, draining into a grossly dilated coronary sinus protruding into the dilated right atrium. Consider left circumflex artery to coronary sinus fistula
3. Small amount of pericardial effusion at the posterior inferior aspects.

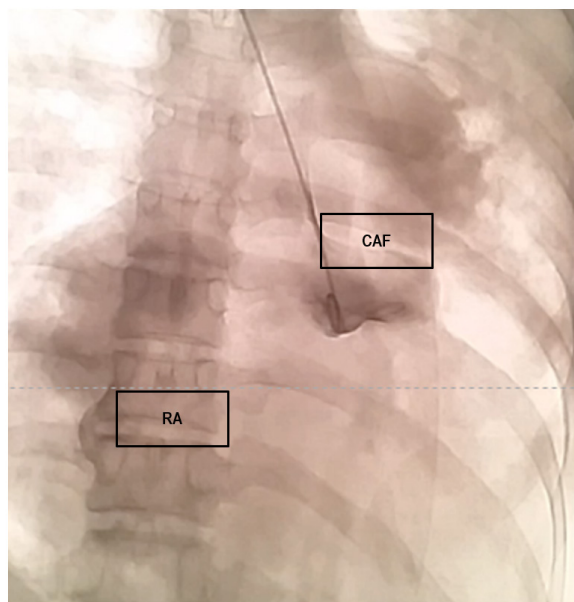


Figure 3. Coronary arteriography (CAF); Coronary artery fistula right atrium(RA)

Table II. Coronary arteriography findings

Left coronary artery: left anterior descending artery appears to originate directly from the aorta. it is a big artery which is free of disease. diagonal branches are fair sized normal vessels.

Left circumflex artery originates from the aorta and is a giant vessel. selective power injection revealed contrast material crossing through the giant left circumflex artery with tortuous course posteriorly and draws into the right atrium suggesting a fistula connecting the left circumflex artery and coronary sinus.

Conclusion: Congenital heart disease; double barrel left coronary system with coronary artery fistula-connecting a giant left circumflex artery to the coronary sinus draining into the right atrium

of that surgeon that she was already inoperable due to her high pulmonary pressure with signs of right sided heart failure. She was advised to be seen by another specialist who, upon knowing her case, might deem her as surgically eligible, however, she was unable to comply. Symptoms of dyspnea were noted to have temporarily improved and she was discharged. She then had subsequent admissions for progression of signs of right sided heart failure.

Upon admission, the patient manifested generalized edema, resting dyspnea, orthopnea, and decreased urine output. She was tachypneic, had icteric sclerae, neck vein distention, bibasal rales, and had a continuous Grade 5 murmur at the back between the spine and left scapula. Occasionally she would have hypotensive episodes and hemoptysis. The patient and family decided to defer definitive management and to continue palliative treatment only. Patient eventually succumbed to multiorgan failure.

Discussion

Normally, the two main coronary arteries originate from the aorta. The right coronary artery provides branches to the right ventricular free wall. The left coronary ostium is

usually single, giving rise to a short, common left coronary artery trunk that branches into the left anterior descending artery and left circumflex artery. The left anterior descending artery extends toward the cardiac apex, the anterior wall of the right ventricle, and the anterolateral free wall of the left ventricle. The left circumflex artery courses along the left atrioventricular groove and posteriorly toward the crux of the heart.⁵

Our patient's coronary artery anomalies comprise of two distinct morphologic features: one is the anomaly in its origin, also known as a double barrel left coronary system. Her left anterior descending artery and left circumflex artery originate directly from the aorta instead of from the left coronary artery. Second, is the anomaly in termination, a coronary artery fistula, with its left circumflex artery fistula draining into the right atrium through the coronary sinus.

The embryogenesis of these fistulas is uncertain, however, fistulas that enter the right ventricle have been related to persistence of primitive intramyocardial sinusoids or to the development of a rectiform vascular network in the distal branches of the involved coronary artery. Fistulas that enter the left ventricle, on the other hand, are thought to result from direct flow through the besian venous channels.⁶

In a review of 50 adults in a tertiary cardiac specialization hospital, the incidence of coronary artery anomalies is 0.9%. 52% of which detailed a separate origin of the left anterior descending artery and left circumflex artery, described as a double barrel anatomy of the left coronary system.⁷

Isolated reports have also appeared of coronary artery anomalies of termination, such as coronary artery fistulas (CAFs) from the conus artery to the right atrium, from the coronary sinus to the left ventricle, from the left circumflex to the coronary sinus or to the pulmonary artery, or from the microfistulae to the left ventricle.⁶ The incidence of multiple or large fistulae is approximately 0.05% and the incidence of small fistulae is 0.1 to 0.4%.⁸

In the latest report from the Philippine Heart Center, a review was done from 1985-1997 in a total of 20 patients, with ages ranging from two months to 71 years old, who were diagnosed to have CAFs. Rheumatic heart disease was identified in five those patients. A fistulous tract originated from the left coronary artery in 17 patients, two patients had their tract arising from the right coronary artery and only one patient had the fistulous tract arising from both left and right coronary artery. The sites of termination of the tract in descending order were into the left ventricle, right atrium, pulmonary artery, left atrium and right ventricle. The most common complaint found was easy fatigability and shortness of breath. The presence of continuous murmur was the main indication for performing cardiac catheterization in twelve patients. Eleven patients underwent surgery while

nine patients were medically managed (two had a small fistula while the rest refused surgery). No long term follow-up was done.⁹

Overall, anomalies of the coronary arteries are rare, occurring in 0.2% to 1.2% of the general population. But the true incidence is difficult to evaluate because most of the cases may be asymptomatic.¹ CAFs tend to grow with age, and if untreated, they cause clinical symptoms in 63% of older patients usually beginning in the fifth or sixth decade.¹⁰ This may have been the case of our patient when she presented her with symptoms at her fourth decade of life.

In adult patients with no previous diagnosis of heart disease, a congenital defect often is not considered as a potential cause of symptoms hence the diagnosis may be delayed.¹¹

The initial suspicion of a congenital coronary artery fistula in an adult is a continuous murmur from the continuous pressure differences between the aortic root and the low-pressure receiving chambers. This murmur is determined by the drainage site of the fistula but it is commonly missed.⁶ The patient's Grade 5 continuous murmur heard in the back between the spine and left scapula is consistent with a left circumflex coronary arterial fistula that drains into the coronary sinus.

Adult patients with a coronary artery fistula can present with angina, myocardial infarction, heart failure, arrhythmias, and endocarditis.¹² A coronary artery fistula's physiologic consequences depend on the volume of blood flowing through it, the vascular bed into which it drains, and the myocardial ischemia that results from a coronary steal caused by low-resistance vascular channels. A fistula that drains into the right atrium, right ventricle, or coronary sinus constitutes a left-to-right shunt, creating a volume overload to the right ventricle as well as in the pulmonary vascular bed, the left atrium and the left ventricle.⁶

It is the opinion of the authors that the consequences of the abnormal connection between the left circumflex artery and the right atrium via the coronary sinus may have caused volume overload to the right sided chambers, ultimately resulting to severe pulmonary hypertension. This was clinically evident with the patient's symptoms of hepatomegaly, ascites, jaundice, and bipedal edema.

The American College of Cardiology/American Heart Association (ACC/AHA) guidelines for adults with congenital heart disease class I recommendation for coronary artery fistula include that if a continuous murmur is present, its origin should be defined either by echocardiography, magnetic resonance imaging, CTA, or cardiac catheterization. Furthermore, they recommended that clinic follow-up

with echocardiography every three to five years can be useful for patients to exclude development of symptoms or arrhythmias, or progression of size or chamber enlargement that might alter management.¹³

In echocardiography, a coronary fistula is recognized as an abnormal area of dilation with diastolic or continuous flow plus a flow disturbance at the site of entry into the cardiac chamber.¹¹ The entire course of the fistula can sometimes be tracked using imaging and color Doppler. Cardiac chamber dilatation is unusual but may be present in the patient with a large left-to-right shunt.¹⁴ In the initial echocardiography findings of the patient, the coronary artery fistula wasn't visible yet. It was only in the most recent echocardiography when findings of multi-chamber dilatation and occasional Doppler color flow close to the interatrial septum were seen and this was suggestive of a large coronary artery fistula. Transthoracic echocardiography has a relatively low sensitivity for the detection of coronary artery anomalies in adults and fails to show the origin of the coronary artery in many patients. Its role in identifying drainage sites of fistulas may be controversial.¹⁹ That is why it is imperative to maintain a high degree of suspicion that such anomalies exist.

Coronary CTA is the main diagnostic tool to depict the entire course of coronary artery anomalies of termination.⁸ CTA allows simultaneous 3D visualization of the coronary vessels. Often data obtained during CTA are incremental to those to those found invasively in angiography. In a study done, the sensitivity and specificity of the CTA to determine anomalous coronaries was 100% and the specific anatomic course of the coronary anomalies was correctly classified in 97% of their patients.¹⁵ In coronary fistulas, it can show the details of fistula source, course, and site of termination and its principal advantage is that it can noninvasively image the surrounding anatomy directly.⁸ A left circumflex artery to coronary sinus fistula was considered in this patient because on coronary CTA, a tortuous and serpiginous left circumflex artery arising from the left coronary cusp of the aorta, draining into a grossly dilated coronary sinus protruding into the dilated right atrium was seen.

Coronary angiography remains the gold standard method in diagnosis of coronary artery fistulas. It can be used to evaluate the size, number of fistulas, anatomical features, and hemodynamics of the fistulous tract.¹⁰

However, the precise path of an anomalous vessel may be difficult to delineate because of restricted 2D view display. In about 50% of the patients, coronary artery anomalies might be incorrectly classified during a coronary angiography.¹⁵ Under such conditions, non-invasive methods such as transthoracic echocardiography combined with Doppler and color flow imaging, transoesophageal echocardiography, magnetic resonance imaging and CTA can be used as adjunct to coronary angiography.¹⁶

With the complementary findings of the transthoracic echocardiography, coronary CTA, and coronary angiography, the diagnosis of the large coronary artery fistula was conclusively made, three years after the patient had her initial presentation.

Coronary artery fistulas (CAFs) have a class II indication for device occlusion therapy. ACC/AHA consensus guidelines indicate that a large CAF, regardless of symptomatology, should be closed via either a transcatheter or surgical route after delineation of its course and its potential to fully obliterate the fistula.¹³

Relative contraindications to transcatheter treatment of CAF include a fistula size that is too small, extreme tortuosity preventing delivery of a catheter far enough distally for embolization, presence of multiple fistula, proximity of normal coronary branches to CAF, and a grossly enlarged proximal coronary associated with a distal fistula.¹⁷ The preferred method of approach for any patient depends on the anatomy of the fistula, the presence or absence of associated defects and the experience of the interventional cardiologists and surgeons.¹⁸

For patients whom surgical and invasive interventions are not an option, as in the case of our patient, symptomatic treatment for her end stage heart failure symptoms ensues which mostly involves palliative treatment.

Conclusion

In the appropriate clinical setting, a high level of suspicion for a coronary anomaly as a cause of a patient's progressive heart failure can lead to timely diagnosis. Several diagnostic modalities such as echocardiography, coronary CTA, and coronary angiography can help us identify these rare cases of anomalous coronaries to provide optimal interventions.

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