

A Curious Case of an Anomalous Right Coronary Artery

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Abstract

Single coronary artery (SCA) is an unusual congenital anomaly, seen by diagnostic coronary angiography. Absence of the right coronary artery (RCA) is one of the rarest coronary artery anomalies occurring when the artery fails to develop in the right atrioventricular (AV) groove. Herein, we describe the case of a 58-year-old man presenting with new onset decompensated congestive heart failure found to have a congenitally absent right coronary ostium. The AV groove extended such that the left circumflex artery supplied the domain of the RCA. Such coronary artery anomalies are rare, and we aim to shed further insight into these congenital processes so that operators may remain vigilant of them in their practice.

Keywords: Single coronary artery; Coronary angiography; Coronary artery anomaly; Right coronary artery

Introduction

Single coronary artery (SCA) is an unusual congenital anomaly that is sparsely reported in literature, seen by diagnostic coronary angiography in about 0.024% to 0.066% of cases [1-3]. It is commonly associated with other congenital cardiac malformations such as coronary arteriovenous fistula, bicuspid aortic valve [4], or transposition of the great vessels [5]. Absence of the right coronary artery (RCA) is one of the rarest coronary artery anomalies occurring when the artery does not develop in the right atrioventricular (AV) groove.

We report the case of a patient with new onset combined decompensated heart failure found to have an absent RCA with a left circumflex artery (LCX) that coursed through the

AV groove extended, supplying the RCA territory.

Case Report

A 58-year-old male patient with history of type II diabetes mellitus and hypertension, not on any medications, presented to the emergency department with progressively increasing lower extremity edema, shortness of breath and dyspnea on minimal effort for the past 2 weeks. He has not been following with his physician for over a year.

Troponins were 0.01 for three consecutive sets, serum pro-brain type natriuretic protein (pro-BNP) was 12,066. Chest X-ray showed small bilateral pleural effusions and subsegmental left atelectasis. Electrocardiogram (ECG) revealed normal sinus rhythm and nonspecific T wave abnormalities. Total cholesterol was 221, triglycerides were 221, high-density lipoprotein (HDL) was 48 and low-density lipoprotein (LDL) was 171. Transthoracic echocardiogram showed a left ventricular ejection fraction (LVEF) of 25-30%, along with grade II diastolic dysfunction. The mitral valve leaflets were tethered due to reduced systolic function and elevated left ventricular diastolic pressure. Mild mitral regurgitation and a left pleural effusion were also visualized.

Cardiology evaluated the patient and started aspirin 81 mg, carvedilol 3.125 mg twice daily, Imdur 30 mg daily, and Lasix 40 mg intravenous (IV) twice daily. The patient was found to have subacute on chronic combined decompensated congestive heart failure and a left heart catheterization was recommended to rule out ischemic cardiomyopathy. Right radial arterial access was performed and indicated a left dominant coronary circulation with normal left main, medium sized left anterior descending artery (LAD) with mild atherosclerosis but no flow limiting lesions. The LCX was large and showed no angiographic evidence of disease. The AV groove extended and the LCX artery supplied the RCA domain. The ostial RCA was congenitally missing and not visualized on angiography (Fig. 1). The aortic pressures were found to be 108/45/76 mm Hg and the left ventricular pressures were found to be 114/4 mm Hg. The patient was discharged from the coronary catheterization laboratory with continuation of medications for aggressive medical management and risk factor modification.

Discussion

We discuss a rare case of SCA, particularly an absent RCA. The LCX extended down the AV groove thereby supplying the

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Figure 1. Coronary angiography showing a congenitally absent right coronary ostium and a large left circumflex artery extending down the atrioventricular groove and supplying the right coronary artery myocardial domain.

RCA territory. The ostial RCA was congenitally absent and not visualized on angiography. There were no other associated cardiac anomalies and only mild atherosclerosis of the LAD. Incidence of the RCA arising from the left coronary branches varies between 0.1% and 0.9%. To our knowledge, most reported cases involve the proximal or middle portion of the LAD as an origin of the RCA [6]. What is dangerous in this entity, is that patients with similar condition, have only one coronary blood flow system, which in case got affected, might lead to a catastrophic death, arrhythmia or subsequent heart failure. Prognosis for patients with SCA ranges widely from normal life span to sudden cardiac arrest [1]. In the presence of other concomitant congenital anomalies, these patients might have higher risk of atherosclerosis, heart failure, or associated congenital cardiomyopathy [1]. In our case, the patient's heart failure was non ischemic in origin; however, the patient did not follow up with our team; therefore, complete workup was not done, including cardiac magnetic resonance imaging. The SCA in this case was an incidental finding and was unrelated to the patient's reduced ejection fraction.

There are numerous stratifications of SCA anomalies. The most employed is created by Lipton et al [1], describes the SCA according to origin and distribution of the coronary arteries. The anomalous coronary artery system is classified as either right or left depending on the origin of the dominant artery. When the RCA is arising from the terminal extension of the LCX artery, it is classified as LI. However, if the RCA arises from the left main (LM), LCX and LAD arteries, it will be classified as LIIA, LIIB and LIIP subgroups, respectively. Type III represents the LAD and LCX arteries that originate from the proximal part of the RCA [1]. This grouping was later modified by Yamanaka et al [7] with the addition of the septal

and combined types, designated as "S" and "C", for further classification of the anatomical variation [6]. Our case belongs to the LI type, due to the angiographic finding of the AV groove extension so that the LCX supplied the RCA territory in the absence of an RCA.

Absence of the RCA was found by Yamanaka et al [7] in only 20 patients who underwent cardiac catheterization, out of 126,595 with a frequency of only 0.016% [7]. The exact incidence is unknown, but there have been other reported cases [8-12], with at least 24 cases reported by Pourafkari et al [13].

Patients with congenital absence of the RCA can present with nonspecific symptoms and may be mislabeled as coronary artery disease (CAD) or remain undetected which may explain the low prevalence [14]. SCA, including group I anomalies, have been shown to have a variable clinical course, from benign to serious [6].

Patients can present with atypical chest pain without a significant CAD [15, 16], typical chest pain, unstable angina, dyspnea, and myocardial infarction [12] and sudden cardiac arrest [17]. It has also been reported that myocardial ischemia is a direct result of SCA in the absence of CAD in 15% of patients [18]. One-third of patients with type LI (four out of 12) have required coronary artery bypass graft (CABG) surgery according to Chou et al [19]. Pourafkari et al reported two out of 24 patients who required CABG and six out of 24 cases requiring percutaneous coronary intervention (PCI), with the remaining patients having benign courses [13].

Coronary angiography is the gold standard for diagnosis and classification of SCA [19]. SCA causes diagnostic difficulty during cardiac catheterization and cardiac surgery. Interventional cardiologists need to be familiar with the various categories of this congenital anomaly [17]. To our knowledge this case of a SCA presenting as dyspnea is barely reported [13]. While most patients improve with medical treatment, others require intervention. PCI in similar cases poses technical challenges, difficulties in cannulation of the coronary ostium as well as challenges with providing adequate catheter support due to abnormal origin and course [20].

During cardiac catheterization, when there are difficulties when visualizing the coronary arteries, clinicians should be aware of the potential for aberrant vessels. Being of aware of these abnormalities allows for prompt diagnosis and therapeutic decisions. Donaldson et al reported that in 28 of 75 cases (37%) cardiac catheterization was repeated due to inadequate visualization of the aberrant vessel. Of the patients studied, 15 were reinvestigated once, nine twice, three cases three times, and one four times until a correct diagnosis could be confidently made [21]. These anomalies, when observed in the index case, result in shorter procedure time, lesser quantity of dye, fewer catheter manipulations, and better angiographic images in subsequent cases as the anomalous course of the RCA was known [22].

Conclusions

We present a case of a patient presenting with signs and symptoms of subacute on chronic combined decompensated congestive heart failure, found to have a congenitally absent right

coronary ostium on coronary angiography during left heart catheterization, to rule out ischemic cardiomyopathy as the etiology of heart failure. The LCX extended through the AV groove and supplied the territory of the RCA. Such coronary artery anomalies are infrequent, and we aim to shed further insight into these congenital anomalies so that operators may remain vigilant of them in their practice. Standard angiography is affected with these infrequent anomalies, and procedural complications might be higher.

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Conflict of Interest

The authors declare that there is no conflict of interest.

Informed Consent

Patient consent for publication was obtained.

Author Contributions

All authors have participated in writing this case report. Hamfrehth Rahming, Chetan Singh, Marc Assaad and Hussam Aridi helped drafting the manuscript. Michael Cinelli, Boutros Karan, and Ruben Kandov not only participated in writing the manuscript, but also followed the patient under their cardiology service, and were responsible for the coronary angiography along with the images that were obtained. Michael Cinelli is also responsible of the concept and Ruben Kandov has also a supervision role.

Data Availability

The authors declare that data supporting the findings of this paper are available within the article.

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