Case Report

Double right coronary artery diagnosed by computed tomographic angiography: A case report and systematic review of the literature

Mohammadreza Zarisfi, MDa, Sina Tavakoli, MD, PhDab,∗

a Department of Radiology, University of Pittsburgh, Pittsburgh, PA
b Heart, Lung, Blood, and Vascular Medicine Institute, University of Pittsburgh Medical Center (UPMC), Pittsburgh, PA

Article history:
Received 2 March 2022
Accepted 9 March 2022

Keywords:
Congenital coronary anomaly
Conventional coronary angiography
Double right coronary artery
Coronary computed tomographic angiography

ABSTRACT

Double right coronary artery (RCA) is an extremely uncommon anomaly that is mostly detected incidentally in patients undergoing coronary angiography. It can be a benign and isolated anomaly or associated with other congenital abnormalities, mostly other coronary anomalies. Although atherosclerosis and myocardial ischemia have been frequently reported in patients with double RCA, this likely reflects that the patients were evaluated for chest pain rather than the predisposition to atherosclerosis in double RCA. Paralleling the increased awareness of this entity and the availability of non-invasive and cost-effective imaging of the coronary arteries, the diagnosis of double RCA has increased recently. Here, we present a case of double RCA diagnosed by coronary computed tomographic angiography, and provide a mini-review on the demography, anatomic variants, and clinical significance of double RCA.

© 2022 Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

Introduction

The incidence of coronary artery anomalies is ~1.5% in patients undergoing multi-detector computed tomography (MDCT) [1]. The majority of these anomalies are the abnormal origin of the right or left coronary artery (or their branches) from the opposite or non-coronary sinus [1]. Double right coronary artery (RCA) is a rare anomaly in which the right coronary system is formed of 2 distinct branches [2]. Although reported mostly as a benign entity, double RCA has been linked to myocardial infarction (MI) and atherosclerosis [3,4]. Several studies have reported variants in anatomical details, including branching patterns and geometric shapes, of double RCA [5]. However, these anatomic variants are far less understood and reviewed in the radiology literature com-

Abbreviations: AMB, acute marginal branch; CA, conus artery; CTA, computed tomographic angiography; LAD, left anterior descending; MDCT, multi-detector computed tomography; MI, myocardial infarction; PDA, posterior descending artery; RCA, right coronary artery.

∗ Acknowledgments:

∗ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

∗ Corresponding author.

E-mail address: sit23@pitt.edu (S. Tavakoli).

1930-0433/© 2022 Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)
pared to double left anterior descending (LAD) coronary artery. Here, we present a patient with double RCA, and provide a mini-systematic review on anatomical characteristics, imaging modalities of choice, and atherosclerotic involvement of double RCA which may assist the detection and classification of this rare anomaly.

Case Presentation

A 54-year-old male patient with a complaint of intermittent non-exertional sharp pain in the left chest was evaluated by coronary computed tomographic angiography (CTA). The patient underwent electrocardiographic-gated single heartbeat coronary CTA (kVp: 100, mA: 1028) using a Revolution CT scanner (GE Healthcare) after intravenous administration of 75 mL of Isovue-370. Heart rate control was achieved by two 25 mg oral doses of metoprolol taken the night before and in the morning of CTA. Sublingual nitroglycerin (0.4 mg) was administered 3-4 minutes prior to the scan. The heart rate during the scan was 49 beats-per-minute.

Coronary CTA delineated the normal origin of the RCA from the right sinus of Valsalva (Fig. 1). There was an immediate take-off of the conal branch from the RCA. RCA was then divided into 2 branches of almost equal size traveling relatively in parallel, 1 in the right atrioventricular groove and the other slightly anterior to it. The anteriorly located RCA gave rise to an acute marginal branch. Both RCAs terminated in separate posterior descending arteries (PDAs) and posterolateral branches. The origin and course of the left coronary system was normal, and there was no significant atherosclerotic involvement of the right or left coronary arteries.

Discussion

Data Extraction and Demographic Data

We performed a systematic search for publications using Medline, Embase, and Web of Science in any language up to January 2022 by using Medical Subject Heading terms: “double right coronary artery,” “dual right coronary artery,” “split right coronary artery,” and “duplicated right coronary artery.” (Fig. 2) The bibliographies and review articles were reviewed manually for additional citations. After removing duplicates and non-relevant studies, our search identified 61 published articles; we retrieved the full text of 54 articles, demographic data, and atherosclerosis involvement of RCAs for 4 studies were collected from other publications. We identified a total of 65 cases, 52 men and 13 women with a predominance of men (4:1 male-to-female ratio). Although about 50% of cases (31/65) were from Turkey, the extremely rare occurrence of this anomaly precluded any conclusion about the role of ethnicity as a potential cause for double RCA. The mean age (± standard deviation) at the time of diagnosis was 55.3 ± 11.4 years. The most common (75.4%) initial presenting symptoms was chest pain (46/61, data was not available for 4 cases).

Anatomical Variations

Double RCA can be categorized according to the number of ostia from aorta, as single ostium or separated ostia. Among 65 cases identified in the literature, 31 cases (47.7%) had a single-ostium RCAs which were bifurcated after a short course, similar to the case presented here. The remaining 34 cases (52.3%) had 2 separate RCA ostia adjacent to each other (Table 1). Double RCA was an isolated finding in 52 cases (85.3%). In 9 cases (14.7%), other congenital cardiac anomalies, mostly other coronary anomalies, were present. In 3 cases, 1 branch of the RCA anomalously originated from the left main or LAD [6] coronary artery. In 2 other cases, RCA originated from the left [7] and non-coronary [8] sinuses of Valsalva. In 65.6% of patients (40/61 with available images to review), the RCAs were of almost the same caliber. However, in 34.4% (21/61) of cases, 1 RCA was larger than the other.

Atherosclerotic Involvement

Our review of the reported cases demonstrates that 39.0% of cases (25/64) were associated with RCA atherosclerosis (summarized in Supplemental Table 1), among them both RCAs were involved in 68% (17/25) cases and a single RCA branch was involved in the remaining 32% (8/25) of patients. Moreover, atherosclerosis in double RCA originating from single ostia was found in 60% of cases (15/25) cases versus 40% of cases (10/25) with separated ostia. Interestingly, atherosclerotic lesions of the left-sided coronary system were also detected in 40.6% (26/64) of patients with double RCA cases. The co-occurrence of atherosclerosis in both double RCA and left-sided coronary system was reported in 26.5% (17/64) of cases, while isolated double RCA atherosclerosis was observed in 24.1% (15/64) of cases. Overall, in 25% (16/64) of patients, the double RCA was accompanied by myocardial infarction. In addition to atherosclerotic involvement, hemodynamic changes due to ostial narrowing, such as slit-like take off, have been considered as a possible mechanism of myocardial ischemia [9,10].

Imaging Modalities

Three main imaging modalities have been used for the evaluation of double RCA, each with advantages and drawbacks. Conventional coronary angiography has long been considered as the gold standard technique in imaging coronary arteries, particular for the assessment of luminal stenosis [11,12]. Conventional angiography provides the opportunity for intervention in the presence of ischemic lesions. However, differentiating a double RCA from the high take-off of a large right ventricular branch would be difficult due to its 2 dimensional depiction of the coronary arteries [11]. Over the past decade, coronary CTA has emerged as a complementary or alternative modality to invasive angiography. Coronary CTA allows for a more accurate 3 dimensional depiction of RCA origin, pathway, and branching [1,13]. The main drawback of coronary CTA is exposure to ionizing radiation, though the implementation of effective dose-reduction techniques, such as prospective electrocardiography-gating, reduces this concern compared to
Fig. 1 – Coronary CTA demonstrates bifurcation of RCA after the take off of the conal branch from the common trunk arising from the right sinus of Valsalva. A. RCA: anterior right coronary artery; AMB: acute marginal branch; CA: conus artery; LAD: left anterior descending artery; PDA: posterior descending artery; P. RCA: posterior right coronary artery

Table 1 – Summary of patients’ characteristics in the review.

| Characteristic                                                                 | Count   |
|------------------------------------------------------------------------------|---------|
| Age (y)                                                                       | 55.3 ± 11.4 |
| Gender                                                                        |         |
| Male                                                                          | 13/65 (20%) |
| Female                                                                        | 52/65 (80%) |
| Most common presenting symptoms                                              |         |
| Chest pain                                                                    | 46/61 (75.4%) |
| Dyspnea                                                                       | 8/61 (13.1%) |
| Origin of double RCA                                                          |         |
| Separate                                                                      | 34/65 (52.3%) |
| Single                                                                        | 31/65 (47.7%) |
| RCAs size/caliber                                                             |         |
| Equal                                                                         | 40/61 (65.6%) |
| Unequal                                                                       | 21/61 (34.4%) |
| Double RCA association with other congenital anomalies                        | 9/61 (14.7%) |
| Double right coronary artery and atherosclerosis                              |         |
| Overall incidence of atherosclerosis involving the double RCAs                | 25/64 (39.0%) |
| Overall incidence of atherosclerosis involving LCA                            | 26/64 (40.6%) |
| Atherosclerosis involving the double RCA with single ostium                  | 17/25 (68%) |
| Atherosclerosis involving the double RCA with separate ostium                | 8/25 (33.3%) |
| Atherosclerosis involving both RCA branches                                   | 15/25 (60%) |
| Atherosclerosis involving only 1 RCA                                          | 10/25 (40%) |
| Atherosclerosis involving both RCA and LCA                                    | 17/64 (26.5%) |
| Atherosclerosis involving only RCA                                            | 8/64 (12.5%) |
| Double RCA accompanied with myocardial infarction                             | 16/64 (25%) |

LCA: left coronary artery; RCA: right coronary artery
Fig. 2 – Flow-diagram of study selection

the early days of cardiac imaging [10,14]. More recently, coronary magnetic resonance angiography (MRA) has been considered as a safe and non-invasive method for assessment of the coronary artery anomalies [15]. While the lack of ionizing radiation is a clear advantage of MRA, the lower spatial resolution and motion degradation limit the utility of coronary MRA to the assessment of the proximal segments of the coronary arteries [15,16].

**Conclusion**

Double RCA is a rare anomaly of the coronary artery system that often remains undetected until patients undergo invasive or non-invasive assessment of the coronary arteries for the work up of chest pain. Identifying this anomaly can help prevent unexpected complications during the coronary intervention or cardiac operation. In addition, double RCA can be affected with atherosclerosis that could be a possible culprit for patients presenting with angina and evidence of MI. While atherosclerotic involvement of RCA was present in a significant fraction of the reported cases who underwent conventional angiography or CTA for chest pain, it is unclear if RCA is predisposed to higher rate or development of more severe atherosclerotic lesions in this anomaly. Abnormalities in the origin of the RCA from aorta (if present), such as slit-like proximal course of the artery, has been considered to contribute to myocardial ischemia, though this requires further assessments.

The growing utilization of coronary CTA, as a non-invasive modality, provides a high pre-intervention diagnostic accuracy for the diagnosis of coronary artery anomalies. A priori knowledge of this anomaly through radiology reports, similar to other coronary anomalies, would provide a helpful roadmap to interventional cardiologists for intervention planning and may reduce the potential for complications.

**Patient Consent**

This case report does not meet the definition of human subject’s research requiring IRB review. Informed consent was not required, as no identifiable information is included in the report.

**Supplementary materials**

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2022.03.042.

**REFERENCES**

[1] Fujimoto S, Kondo T, Orihara T, Sugiyama J, Kondo M, Kodama T, et al. Prevalence of anomalous origin of coronary artery system outside aorta. J Radial Case Rep 2016;8(11):5.
artery detected by multi-detector computed tomography at one center. J Cardiol 2011;57(1):69–76.

[2] Chien T-M, Chen C-W, Chen H-M, Lee C-S, Lin C-C, Chen Y-F. Double right coronary artery and its clinical implications. Cardiol in the Young 2014;24(1):5–12.

[3] Acet H, Ozyurtlu F, Bilik MZ, Ertas F. A rare coronary anomaly: Atypical double right coronary artery with an acute inferior myocardial infarction. Korean Circ J 2012;42(3):208–11. doi: 10.4070/kcj.2012.42.3.208.

[4] Ermis E, Ipek E, Demirelli S. Double right coronary artery arising from separate ostia. Am J Cardiol 2014;113(7):S105–S106. doi: 10.1016/j.amjcard.2014.01.274.

[5] Singh S, Ajayi N, Lazarus L, Satyapal KS. Anatomic study of the morphology of the right and left coronary arteries. Folia Morphol 2017;76(4):668–74.

[6] Andreou AY, Tryfonos A, Christodoulou C, Theodorou S, Avraamides PC. Isolated single coronary artery with dual right coronary artery distribution. A rare anatomical variation. Herz 2012;37(4):432–5. doi: 10.1007/s00059-011-3529-1.

[7] Andreou AY. Split right coronary artery. J Cardiovasc Med (Hagerstown) 2010;11(5):399–400. doi: 10.2459/JCM.0b013e328335757c.

[8] Huang ZQ, Chen SJ, Chen J. Dual right coronary artery associated coronary artery fistula. Eur Heart J 2008;29(8):968. doi: 10.1093/eurheartj/ehm533.

[9] Abolbashari M, Kar S. Symptomatic double right coronary artery with inferior wall ischemia. J Am Coll Cardiol 2019;73(051):2875.

[10] Abolbashari M, Kar S, Marmol-Velez A, Ramos-Duran L, Mullins C, Mukherjee D, et al. Double right coronary artery and its clinical significance: review of the literature. Cardiovasc Revasc Med 2017;18(8):632–5. doi: 10.1016/j.carrev.2017.06.016.

[11] Altun A, Akdemir O, Erdogan O, Ozbay G. An interesting diagnostic dilemma: double right coronary artery or high take off of a large right ventricular branch. Internat J Cardiol 2002;82(1):99–102.

[12] Barthe J-E, Benito M, Sala J, Houbani AJ, Quintana E, Esplugas E, et al. Double right coronary artery. Am J Cardiol 1994;73(8):622.

[13] Kunimasa T, Sato Y, Ichikawa M, Ito S, Takagi T, Lee T, et al. MDCT detection of double right coronary artery arising from a single ostium in the right sinus of Valsalva: report of 2 cases. Int J Cardiol 2007;115(2):239–41. doi: 10.1016/j.ijcard.2006.01.060.

[14] Alfakih K, Budoff M. MDCT coronary angiography: does the benefit justify radiation burden. Br J Cardiol 2010;17:207–8.

[15] Welker M, Salanitri J, Deshpande VS, Shea SM, Li D, Pereles FS. Coronary artery anomalies diagnosed by magnetic resonance angiography. Australasian Radiol 2006;50(2):114–21.

[16] Mavrogeni S, Markouzis-Mavrogenis G, Kolovou G. Contribution of cardiovascular magnetic resonance in the evaluation of coronary arteries. World J Cardiol 2014;6(10):1060–6. doi: 10.4330/wjc.v6.i10.1060.