

Congenital absence of the right coronary artery

A case report and literature review

Gao-Wu Yan, MD^a, Anup Bhetuwal, MD^b, Guo-Qing Yang, MD^{a,*}, Quan-Shui Fu, MD^a, Na Hu, MD^a, Lin-Wei Zhao, MD^a, Hong Chen, MD^a, Xiao-Ping Fan, MD^a, Jing Yan, MD^a, Hao Zeng, MD^a, Qing Zhou, MD^a

Abstract

Rationale: Congenital absence of the right coronary artery (RCA) is a rare congenital malformation of the cardiovascular system which may have fatal consequences.

Patient concerns: A 63-year-old man with a 5-year history of chest pain after exertion which had aggravated for >1 month was advised for admission and computed tomography angiography (CTA) examination of the coronary artery to screen for coronary artery disease (CAD).

Diagnoses: The coronary artery CTA showed absence of RCA arising from the aortic root after which a selective coronary angiography (SCA) examination was done that confirmed the diagnosis of congenital absence of RCA.

Interventions: As the patient refused to receive a coronary artery stent implantation citing his financial condition, only symptomatic treatment was given.

Outcomes: The patient requested to be discharged from the hospital against the advice of his doctors 1 week later. A query made by the telephone suggested that the patient's symptoms were under control by use of prescribed medications only.

Lessons: Although being a rare condition, a coronary artery CTA examination can be utilized to screen for congenital absence of RCA and other varieties of cardiovascular malformation whereas SCA can be performed to confirm the diagnosis.

Abbreviations: AST = aspartate aminotransferase, CAD = coronary artery disease, CK-MB = creatine kinase isoenzyme, CT = computed tomography, CTA = computed tomography angiography, cTnI = troponin I, ECG = electrocardiogram, LAD = left anterior descending, LCA = left coronary artery, LCX = left circumflex artery, LMCA = left main coronary artery, NSTEMI = non-ST-segment elevated myocardial infarction, RCA = right coronary artery, SCA = selective coronary angiography, UAP = unstable angina pectoris.

Keywords: computed tomography, computed tomography angiography, coronary artery, coronary artery disease, malformation, non-ST-segment elevated myocardial infarction, selective coronary angiography, unstable angina pectoris, variation

1. Introduction

Congenital absence of the right coronary artery (RCA) is a rare congenital malformation of the cardiovascular system with risk

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^aDepartment of Radiology, Suining Central Hospital, Suining, ^bSichuan Key Laboratory of Medical Imaging and Department of Radiology, Affiliated Hospital of North Sichuan Medical College, Nanchong, Sichuan Province, China.

*Correspondence: Guo-Qing Yang, Department of Radiology, Suining Central Hospital, Suining 629000, Sichuan Province, China (e-mail: 13890893057@163.com).

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involvement that can lead to death.^[1–3] It is presumed that congenital absence may be caused by agenesis or congenital occlusion of the RCA during the embryonic period which may also have an association with other congenital heart disease (CHD).^[3,4]

In this study, the authors report a case of congenital absence of RCA which was initially noticed on coronary artery computed tomography angiography (CTA) examination and confirmed by selective coronary angiography (SCA). In addition to the case report, published literature on single coronary artery has been reviewed to help us better understand this type of cardiovascular malformation, which as far as the authors are concerned, has not been specially published before.

The clinical material was available in the electronic medical database of Suining Central Hospital, thus, this study was approved by the institutional review board of our hospital with waiver of patient's informed consent.

2. Case report

A 63-year-old man was admitted to our hospital with chief complaint of repeated chest pain after exercise for >5 years. The chest pain was retrosternal, which would last for 10 to 20 minutes and could be relieved by rest. Since last 1 month, the frequency and severity of the chest pain had increased significantly and now not even being relieved by rest which prompted the patient to seek the medical attention for the first time.

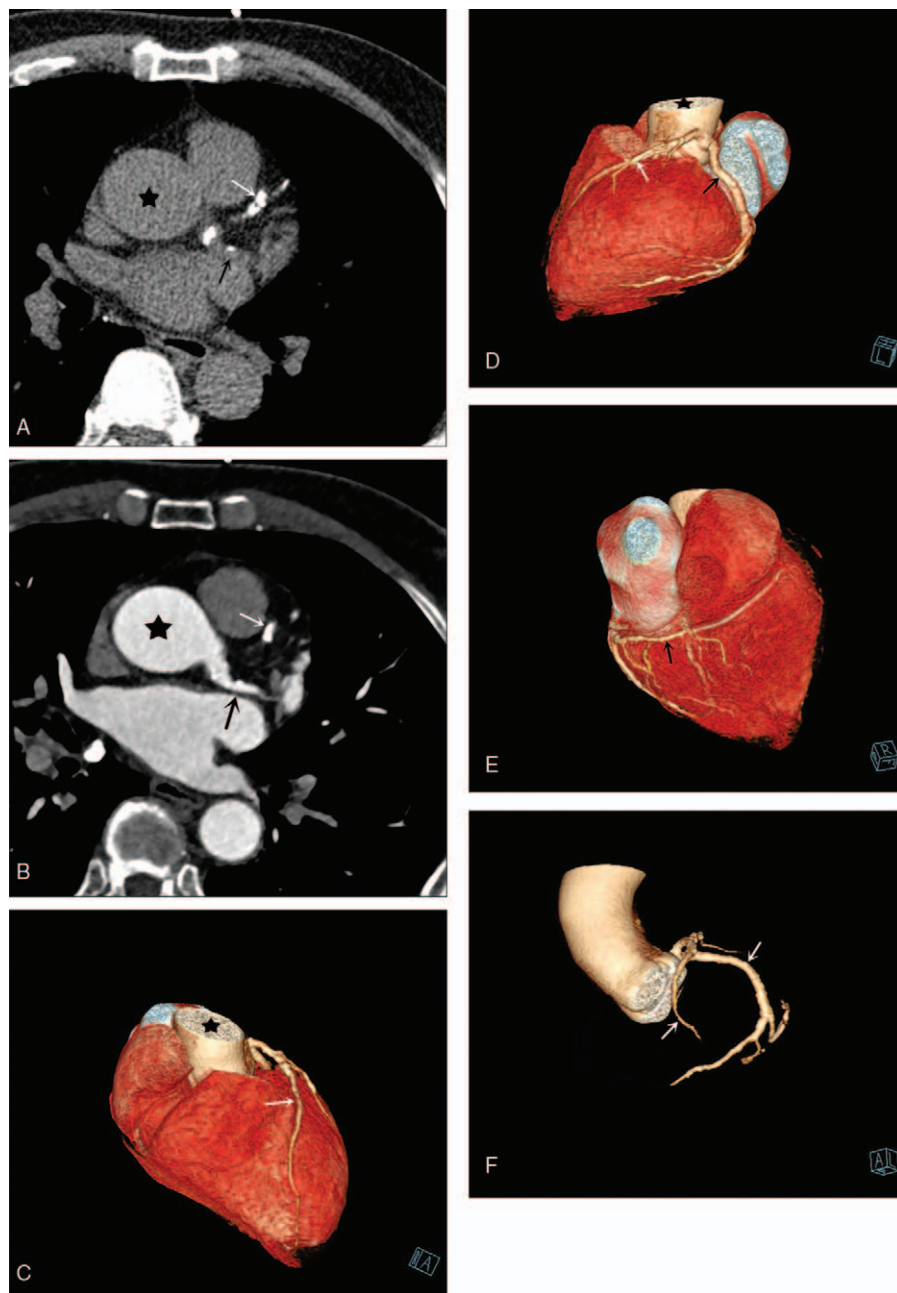


Figure 1. CTA examination of the coronary artery (A–F) showing LCA and its branches (white and black arrows) and absence of RCA at the aortic root (black star). Numerous calcified plaques are visible in the left coronary artery. LCA=left coronary artery.

He denied having any known chronic diseases in the past including any cardiovascular disease. On physical examination, the patient's blood pressure was 168/110 mmHg, respiratory rate—18/min, and heart rate—75/min. Cardiac examination was normal. The patient was then admitted with a provisional diagnosis of coronary artery disease (CAD), unstable angina pectoris (UAP), and hypertension for further investigation and treatment. Electrocardiogram (ECG) revealed sinus rhythm with ST-T changes. Troponin I (cTnI), creatine kinase isoenzyme (CK-MB), and aspartate aminotransferase (AST) level were 1659.4 pg/mL (reference range, 0–34.2 pg/mL), 17.85 ng/mL (reference range, 0–5.2 ng/mL), and 150 U/L (reference range, 15–40 U/L), respectively. A diagnosis of non-ST-segment elevation myocardial infarction (NSTEMI)

was made. The patient was then advised to have a coronary artery CTA examination (SOMATOM Definition Flash, Siemens Healthcare, Germany). In CTA images, RCA was not visible from its origin of aortic root (Fig. 1A–F). Numerous calcified plaques in the left coronary artery (LCA) (the coronary artery Agatston calcification score was 844) leading to different degrees of stenosis at the left main (LMCA), left anterior descending (LAD), and left circumflex (LCX) artery was noted. The maximum degree of stenosis was >50% at the LAD. A SCA (Fig. 2) confirmed the diagnosis of congenital absence of the RCA and stenosis at the LCA and its branches with maximum stenosis at the LAD (>50%). Following the report, the patient was advised to have a coronary artery stent implantation. However, citing the financial problems in his family, the patient



Figure 2. SCA showing absence of RCA and different degrees of stenosis at the LCA and its branches (white and black arrows) with 50% stenosis at the LAD branch (white arrow) thus confirming CTA findings. CTA=computed tomography angiography; LAD=left anterior descending, LCA=left coronary artery; RCA=right coronary artery, SCA=selective coronary angiography.

refused to receive the intervention. He was then offered symptomatic treatment only.

During his hospitalization, the patient was treated with aspirin (100 mg, qd), clopidogrel (75 mg, qd), atorvastatin (20 mg, qn), metoprolol (12.5 mg, bid), and isosorbide mononitrate (20 mg, bid). The hospital stay was uneventful. One week later, against the advice of his doctors, the patient discharged himself from our hospital. The patient was inquired about his current condition via the telephone where he stated that he is taking the medicines regularly. He also denied any further chest pain and discomfort.

3. Discussion

3.1. Congenital absence of the right coronary artery and its prevalence

As defined by Smith, a single coronary artery is an “isolated artery arising from the aortic trunk by a single coronary ostium and supplying the entire heart regardless of its distribution.”^[5] In general, single coronary artery has a low prevalence in population. Various literature has reported to be at 0.014% to 0.066%^[6–8] while Lipton et al^[6] noted that it occurred in approximately 0.024% of the population. A recent study by Türkmen et al^[8] recorded prevalence of single coronary artery at 0.031% (67/215140).

Congenital absence of the RCA or RCA extending from LCX is a special type of the single coronary artery.^[6,9] Its prevalence in patients with single coronary artery is low, hence, only few cases has been reported until now.^[9–11] Furthermore, patients with congenital absence of the RCA often present with non-specific clinical features and ECG manifestation who are often misdiagnosed as CAD or may remain undiagnosed. This might be one of the reasons for low prevalence of congenital absence of RCA.

3.2. Angiographic classification of single coronary artery

In 1979, Lipton et al^[6] first proposed an angiographic classification of single coronary artery which can be used for cases of single coronary artery without additional congenital cardiac anomalies. In brief, according to the site of origin (right or left coronary sinus), a single coronary artery is firstly divided into types R and L. Then secondly, according to its anatomical distribution of the branches, it is divided into groups I, II, and III, in which, group I represents the course of a single coronary artery extending to supply the heart that should have been supplied by the contralateral coronary artery, group II represents a single coronary artery giving off a transverse branch from its proximity to supply the heart that should have been supplied by the contralateral coronary artery, and group III represents a LCX and LAD branch arising separately from the trunk of a coronary artery. Lastly, according to the route undertaken by the large transverse trunk while crossing the base of the heart to arrive in the vicinity of the normal contralateral coronary artery, it is classified into A (anterior), B (between), and P (posterior) categories in relation to aorta and the pulmonary artery.

According to the course of the coronary arteries involved, Smith's classification has 3 groups. Group I consists of single coronary artery coursing as RCA, LCX or as the LAD artery, or a single left main coronary artery which divides into 2 branches as the LAD and LCX whereby RCA is formed by the extension of the circumflex traversing the base of the heart; Group II consists of a chief single artery giving off right and left main coronary arteries, or separate RCA, LAD, and LCX which then courses through their original paths; in group III, the single coronary artery will have an atypical branching leading to significant differences in the original routes of the 3 main coronary arteries.^[8]

Additionally, Yamanaka and Hobbs^[1] also divided coronary artery abnormality into benign and potentially serious anomalies in relation to clinical features. Single coronary artery is considered as one of the potentially serious anomaly which may be related with complications such as syncope, cardiac arrhythmias, angina pectoris, myocardial infarction, congestive heart failure, and even sudden death.

Among the 3 classification systems described above, the angiographic classification proposed by Lipton et al^[6] is now used widely in the clinical practice. According to Lipton's classification, the case reported in this present study is categorized as type L-I.

3.3. Clinical and ECG manifestation

The clinical manifestations of a patient who has a single left coronary artery may vary from asymptomatic to symptoms associated with myocardial ischaemia.^[12] During their early years patients may not show any sign or symptoms from this malformation. With aging, atherosclerosis in a single left coronary artery can lead to symptomatic clinical features that can vary from simple angina to grave consequences such as fatal arrhythmias, myocardial infarction, and sudden cardiac death.^[1] The relationship between the clinical manifestations and congenital absence of the RCA is still unknown.^[12] The authors presume that during an early age, absence of risk factors such as atherosclerosis, hypertension, and diabetes may be the reason behind the lack of clinical symptoms. Patients are symptomatic only when the disease of coronary artery begins to occur.

However, the relationship between the clinical manifestations and atherosclerosis of a single coronary artery is still controversial^[13–15] and use of prophylactic lipid lowering medications in such patients has not been clarified. We assume that because single coronary artery is responsible to meet all the metabolic demand of the heart, it may be excessively dilated causing increase tension on the wall of the vessel. This may lead to endothelial injury of the involved single coronary artery which can trigger the atherosclerotic process early.

The ECG manifestations of a patient with single left coronary artery may vary from no changes to various findings^[11] such as nonspecific ST-T wave changes to supraventricular arrhythmia. To a certain extent this may be explained by the blood supply to sinoatrial (SA) and atrioventricular (AV) node. While RCA normally supply these nodes it is done so by a single left coronary artery and its branches. Lack of adequate blood supply from this anomaly may lead to ischaemia in SA and AV node with eventual fibrosis and dysfunction which might be manifested in ECG varying from normal to abnormal readings related to ischaemia or arrhythmia.

The patient in our study denied any symptoms during his early age whereas since these last 5 years (present age 63 years) he had been experiencing chest pain.

3.4. Value of coronary artery CTA in the diagnosis of single coronary artery

Not only SCA examination is regarded as the “gold standard” for the diagnosis of diseases such as CAD and coronary artery malformation at present, it is also utilized for the treatment and prognostic follow-up.^[16] However, the shortcomings of SCA is that it is an invasive procedure with hazards of radiation exposure including risk of various malignancy to both the operator and the patient.^[17] Furthermore, during SCA examination that is intended to diagnose coronary artery malformation such as congenital absence of RCA as concerned in this study, an operator needs to experiment with numerous non-standard projection positions that may ultimately lead to increased consumption of contrast agent and longer procedure time while difficult manipulation of the catheter can predispose to increased risk of complications and wrongful diagnosis.

On the contrary, as a non-invasive examination, coronary artery CTA is being performed widely for diagnosis, treatment and follow-up of CAD, coronary artery malformation, and other diseases.^[18] In a meta-analysis conducted by Li et al,^[18] with SCA as the reference standard, diagnostic accuracy of multislice CTA for detection of coronary artery stenosis was evaluated at a patient-, vessel-, and segment-level. The pooled results showed that multislice CTA can effectively identify most of the patients who has CAD and the high negative predictive value (90%, 97%, and 97%, respectively) makes it an effective non-invasive alternative to SCA for the exclusion of stenosis. In addition, many post-processing techniques of coronary artery CTA examination such as volume rendering (VR), maximum density projection (MIP), and curved planar reformation (MPR) make it easier to evaluate during the pre- and post-procedure.^[19] Another added benefit CTA has over SCA is that it can evaluate the left and/or right ventricle and/or atrium at the same time in a “one-stop shop” examination.^[20]

In this case study, our patient was first diagnosed by a coronary artery CTA which was then confirmed by SCA. CTA revealed absent RCA at the aortic root and those areas of heart which should have been supplied by a RCA, now being supplied by an

elongated and dilated LCA and its branches. Among the various differential diagnosis of this condition the important and more clinically significant are complete obstruction or hypoplasia of one major coronary artery, a very dominant left system, an anomalous origin of one coronary artery, and others as reported in detail by Lipton et al.^[6]

3.5. Treatments

When it comes to the treatment of congenital absence of the RCA, currently, there is no standardized procedure or guideline from the evidence-based medicine.^[12] Choice of treatment may include either conservative treatment with anti-platelets, lipid-lowering, anti-hypertensive therapy etc. or with interventional therapy such as coronary artery revascularization, pacemaker implantation, and other cardiac surgical procedures.^[12] The patient concerned in this paper was first treated conservatively to control his symptoms. A coronary artery stent implantation was advised. However, because of his economical constraints the patient refused to receive the procedure following which he received only pharmacological treatment.

4. Conclusion

In conclusion, congenital absence of RCA is a rare condition. However, a coronary artery CTA can be applied to screen similar cardiovascular system malformations followed by a SCA examination to confirm the diagnosis.

Author contributions

Conceptualization: G-W. Yan, G-Q. Yang, Q-S. Fu.

Data curation: G-W. Yan, G-Q. Yang, Q-S. Fu, L-W. Zhao, H. Chen, J. Yan, H. Zeng, Q. Zhou.

Investigation: N. Hu, X-P. Fan.

Resources: G-Q. Yang

Supervision: G-Q. Yang

Writing – original draft: G-W. Yan, A. Bhetuwal, G-Q. Yang, Q-S. Fu.

Writing – review & editing: G-W. Yan, A. Bhetuwal, G-Q. Yang, Q-S. Fu.

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