An Extremely Rare Coronary Anomaly: Right Single Coronary Artery or Agenesis of Left Main Coronary Artery? A Classification Dilemma

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Research Article

Keywords:

Posted Date: December 16th, 2023

DOI: https://doi.org/10.21203/rs.3.rs-3727316/v1

Additional Declarations: No competing interests reported.

Abstract

Purpose

Single coronary artery is an extremely rare anomaly where all the coronary circulation relies upon a single coronary trunk from either the right or the left sinus of Valsalve.The purpose of the present case report is to describe an extremely rare coronary artery anomaly which causes a classification dilemma.

Methods

A 67-year-old male patient admitted with acute coronary syndrome underwent a coronary angiography performed with Judkins technique to obtain standard angiographic views.

Results

We hereby present a very rare case of right single coronary artery anomaly diagnosed with invasive coronary angiography and computed tomography scan in a 67-year-old male patient with chest pain and share a classification dilemma.

Conclusion

We presented an extremely rare case of right SCA anomaly which causes classification dilemma. Furthermore, such coronary anomalies should be kept in mind before performing percutaneous coronary intervensions.

Introduction

The term "coronary artery anomaly" encompasses a broad spectrum of congenital abnormalities that affect the origin, path, and structure of the epicardial coronary arteries.

With an estimated occurence rate of 0.024–0.066% in the general population, single coronary artery (SCA) is a very uncommon congenital anomaly characterized by the presence of a solitary coronary artery originating from a single coronary ostium, providing blood supply to the entire heart (1–3). While the affected cases may be asymptomatic, they also are likely to be admitted with a varity of symptomology including typical or atypical anginal chest pain, ventrivular arrhytmias, syncope, acute coronary syndrome or sudden cardiac death(2).

Hereby we present a unique case of right SCA which has not been fit to the current classification system.

Case report

A 67 year-old male patient was admitted to our cardiology clinic with a complaint of a burning sensation in his chest after exercising. He had a history of smoking for over 25 years and also exhibited hypertension. ECG was unrevealing other than 1.5 mm ST-segment elevation in aVR. Echocardiographic evaluation revealed a mild mitral regurgidation with a global left ventricular ejection fraction of 55%. During the treadmill test, the patient had to stop after five minutes due to dyspnea and high blood pressure. It was then decided to proceed with invasive coronary angiography in order to evaluate the patient further, which depicted an absent left mian stem (LMS) and normal right coronary artery (RCA). Of note, the left anterior destending artery (LAD) was in its inheritedcourse and seen as a continuation of RCA in the posterior interventricular groove near the cardiac apex rather than as otherwise a bifurcation of LMS in the base of the heart (Fig. 1–2, Video 1–2). The left circumflex artery (LCX) was observed to originate from the very proximal part of the LAD and to traverse its inherent course.A subsequent aortography was unable to localize the LMS (Video 3). At this point, it was considered that a chronic total occlusion might have been present in the LMS. Subsequently, a coronary CT scan was planned to better delineate the ultimate coronary anatomy, which revealed LMS agenesis and a patent single right coronary artery supplying the whole heart by giving off the LAD and subsequently the LCX (Fig. 3–4).We decided to follow up the patient conservatively with life style modifications and medical therapy as appropriate.

Discussion

SCA is an extremely rare coronary anomaly, in which only one coronary artery provides blood supply to the entire heart.Lipton et al. (3)had introduced a valuable angiographic classification system for SCA based on the site of origin and anatomical distribution of its branches. This *Lipton classification* divides cases into 'R' right-type and 'L' left-type based on the origin of the single coronary artery in the right or left sinus of Valsalva. Additionally, each case is further categorized into group I, II, or III based on the specific anatomical course of the artery. Type I denotes a right SCA arising from the right coronary sinus, and then then continues in the left atrioventricular groove as LCX before it terminates as LAD, or as the LMS that gives LCX and LAD; the LCX then terminates as RCA. In Type II, a SCA originates from either the right or left coronary sinus. From the proximal segment of this SCA, there is an additional anomalous artery that arises and crosses the base of the heart before returning to its original path. Type III corresponds to a scenario where LCX and LAD originate separately from the proximal right coronary artery (RCA). Furthermore, Lipton et al. classified SCA into three categories based on its relationship to the aorta and the main pulmonary artery. Category A refers to the anomalous artery passing in front of the main pulmonary artery, Category B is characterized by the anomalous artery passing between the ascending aorta and the main pulmonary artery. On the other hand, category P indicates that the anomalous artery passes behind the ascending aorta. In 1990, Yamanaka et al.(4)further modifiedthis modelinto a more useful framework for understanding and categorizing different variations of a SCA and introduced two additional groups: S and C. The group S denotes a transseptal course of the anomalous artery, while the group C represents a combined course that combines elements from other categories.

As one can appreciate, the anomaly in our case report does not fit into the modified Lipton classification, and it may be argued that it is not a true SCAexample. Therefore, we may propose a unique combination

of coronary anomalies where there is a left main stem agenesis together with single RCA connected to the LAD artery through an interarterial communication near the apical part of the left ventricle. Interarterial communications are the persistance of fetal coronary circulation patterns andare generally larger than 1 mm in diameter, possess strait routes and gentle curvatures, which are their main differentiating factors from the coronary collaterals(7). Morover, most of the coronary interarterial communications occur between RCA and LCX in the posterior interventricular groove and between RCA and LAD in the distal interventricular groove(8). Hence, it would also be prudent to regard the connection between LAD and RCA in our case report as a true interarterial communication. To our knowledge, this is the first case report to suggest such aclassification dilemma in coronary anomalies.

The optimal treatment approach for SCA is not well-defined. The decision on therapy should be guided by the specific course of the anomalous coronary artery and any associated coronary atherosclerosis. In cases where the anomalous artery courses between the aorta and main pulmonary artery, or when there is significant atherosclerosis, coronary artery bypass surgery may be beneficial. Revascularization strategies, such as percutaneous coronary intervention, have also been reported as successful in certain cases(4, 5). Due to the fact that the entire coronary circulation relies on a SCA, any complication would be inadequately tolerated. Therefore, the treatment plan should be tailored to the individual patient's condition and needs. However, it is of paramount importance for the cardiologist to be aware of this uncommon condition, especially before considereing any percutaneous coronary intervention to chronic total occlusions, wherein unawareness of the types of SCA anomalies may possibly lead to catastrophic coronary or cardiac perforations.It is for this reason that coronary CT angiography and magnetic resonance imaging provide perfect depiction of the ultimate coronary anatomy for the definitive diagnosis and risk stratification before implementation of a treatment modality.

Conclusion

Herein we presented an extremely rare case of right SCA anomaly. However, failure of our case to comply with the classification criteria proposed previously in the medical literature may lead to a classification dilemma. Such coronary anomalies should be kept in mind before performing percutaneous coronary intervensions, and further imaging modalities should be used to depict the exact coronary anatomy to any procedural complication.

Declarations

Ethical Approval: Not applicable.

Conflict of Interest: The authors declare that there is no conflict of interests for this manuscript.

Funding: The authors declare that there is no funding for this manuscript.

Consent: Written informed consent was obtained from the patient for this case report.

Authors' Contributions: Kök, Zafer: Data collection. Sokmen, Erdogan: Data collection and witing the manuscript. Bingöl, Bilge: Data collection. Kalay, Nihat: Critical review of the manuscript.

Availability of Data: Datasets regarding the current case report shall be available upon reasonable requests.

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Figures

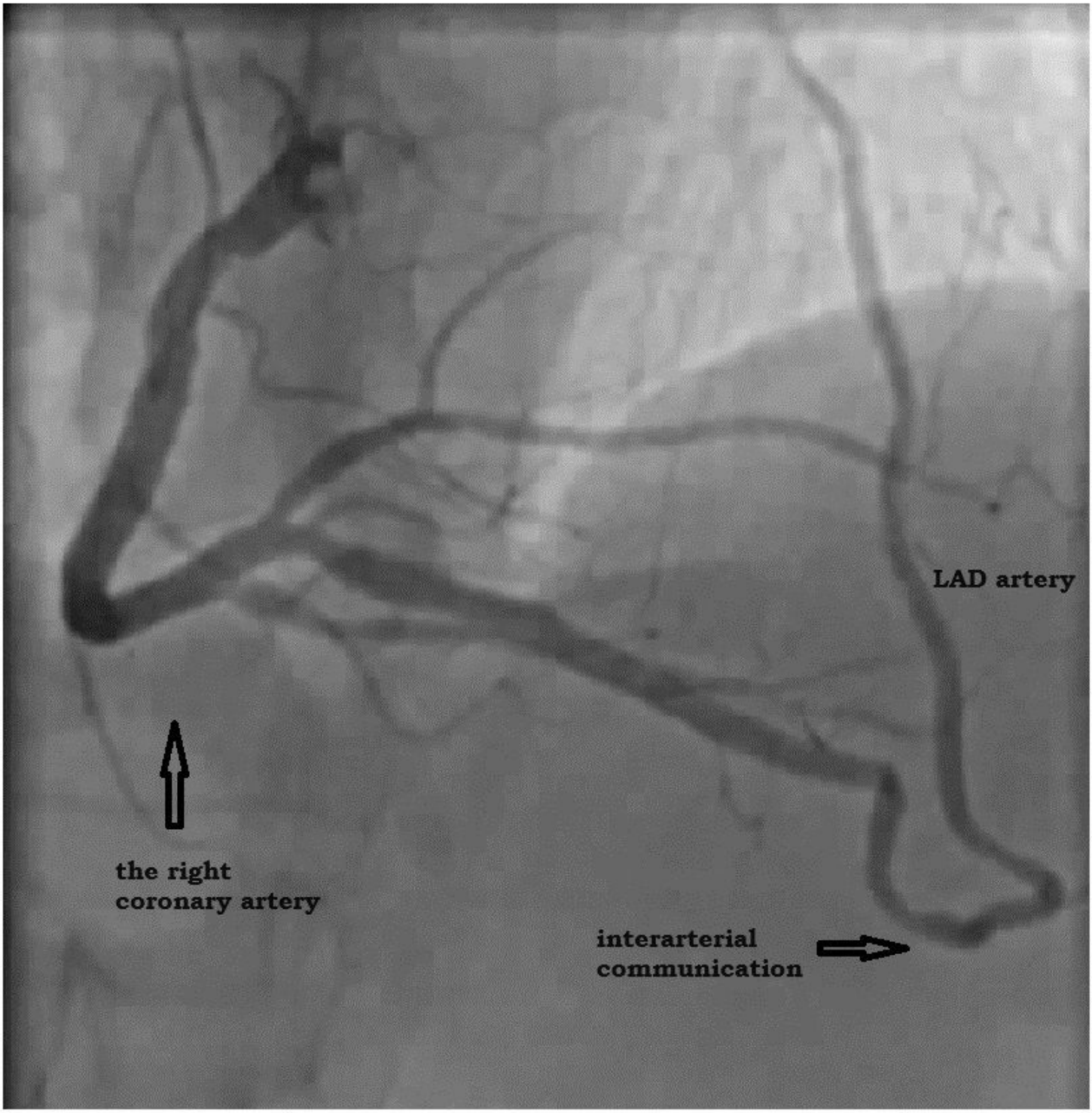

Figure 1

Left anterior oblique view depicting the interarterial communication between the body of LAD artery and the right coronary artery. Note that there is no filling of the left main coronary artery.

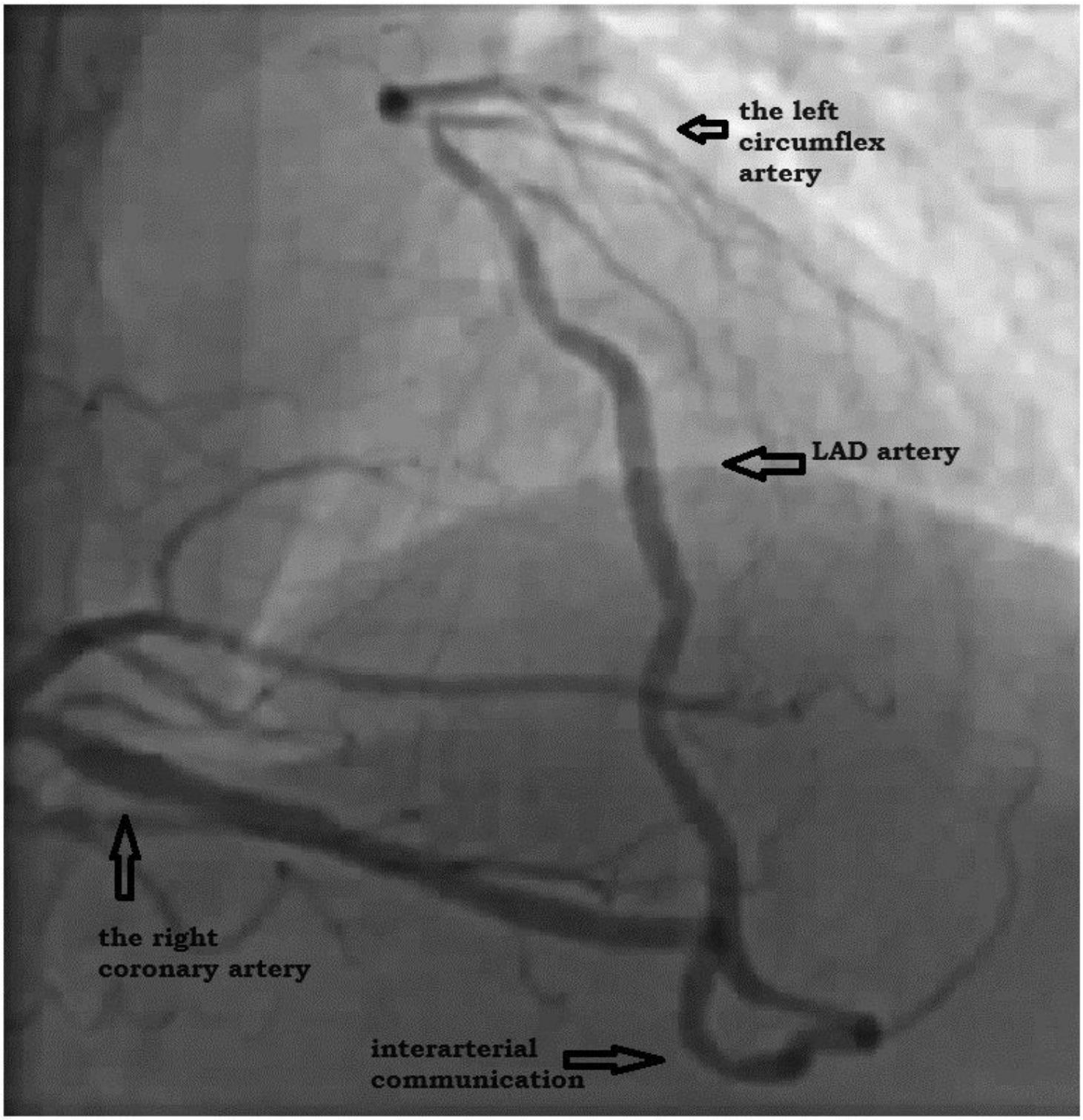

Figure 2

Left anterior oblique view with cranial angulation showing lack of the left main coronary artery.

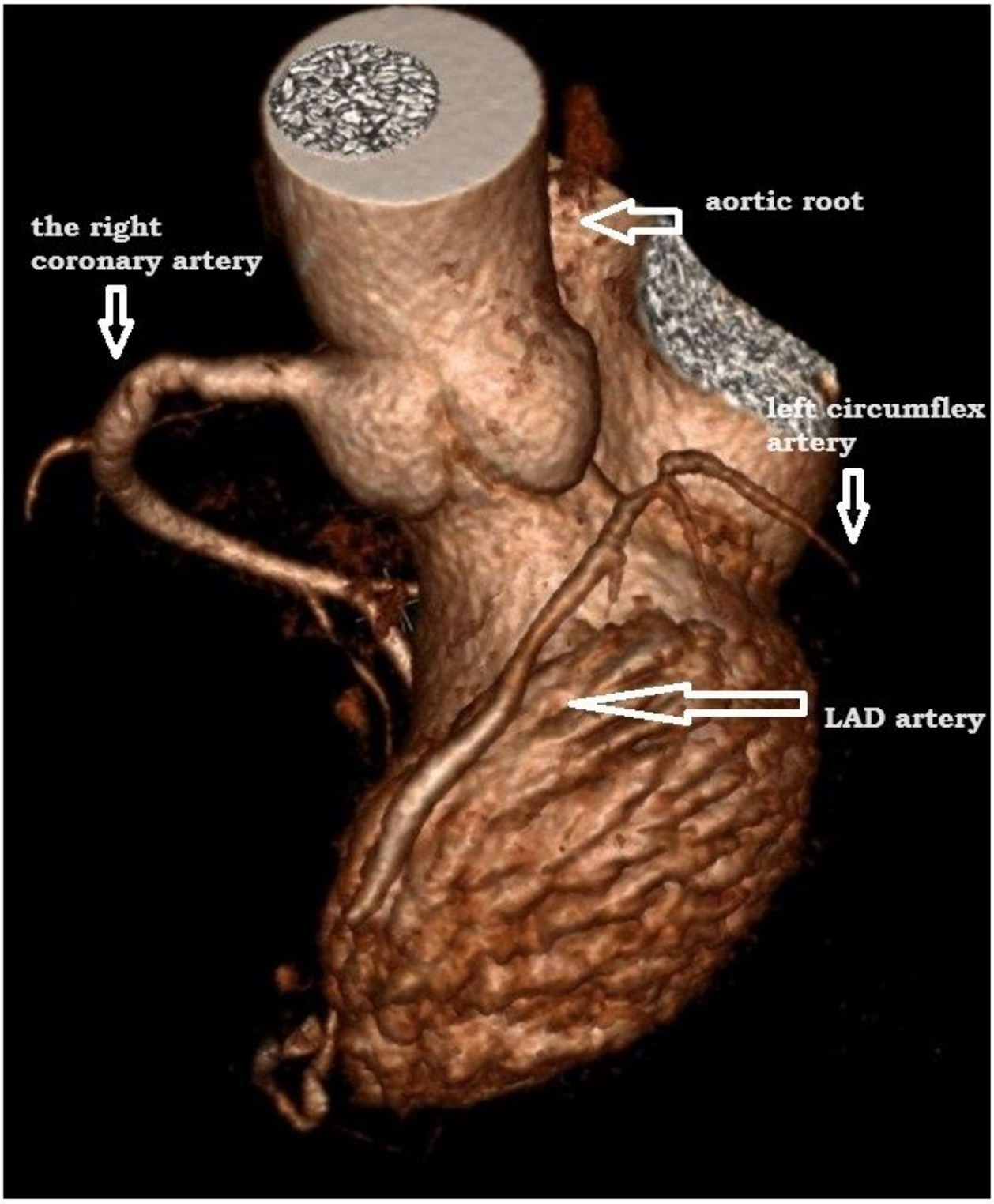

Figure 3

Coronary computed tomographic image depicting the agenesis of the left main coronary artery.

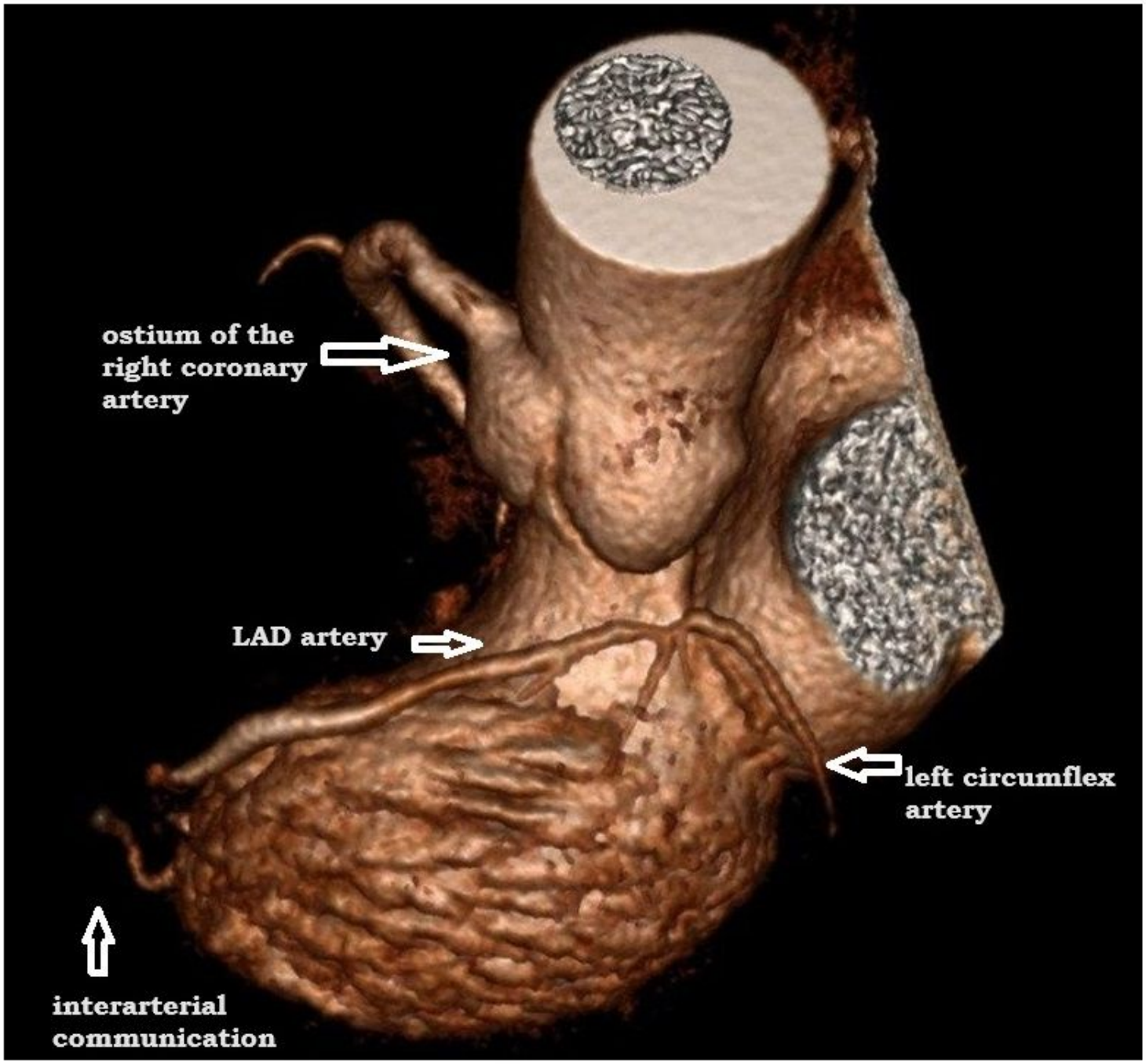

Figure 4

Coronary computed tomographic image showing origination of the right coronary artery from the aortic sinus of valsalva and agenesis of the left main coronary artery.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- Video1.mp4
- Video2.mp4

- Video3.mp4