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**CASE REPORT**
*Congenital heart disease*

# Anomalous origin of left main coronary artery from right coronary artery in a patient presenting with inferior wall myocardial infarction: a case report and literature review

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**Background** Anomalous origin of the coronary arteries is seen in less than 1% of the general population. Single coronary artery (SCA) is a congenital anatomic abnormality identified by a single coronary ostium giving rise to one coronary artery. We present an extremely rare variant of the left main coronary artery (LMCA) branching off from the right coronary artery (RCA) and following a prepulmonic course.

**Case summary** A 72-year-old woman presented due to ongoing chest pain with associated ST-segment elevation involving the inferior leads. Emergent cardiac catheterization revealed a 99% ulcerated lesion in distal RCA, which was intervened on with angioplasty and stent placement. The RCA was noted giving rise to LMCA, which followed a prepulmonic course (anterior to pulmonary artery) before trifurcating into a small caliber left anterior descending, ramus intermedius, and hypoplastic left circumflex arteries. The non-malignant course of the aberrant LMCA was confirmed on the coronary computed tomography angiogram. The patient was discharged home on guideline-directed medical therapy.

**Discussion** The patient illustrated congenital SCA with type RIIA pattern of the aberrant vessel based on the Lipton anatomic classification for SCA. The prepulmonic course of SCA is usually benign and can be managed conservatively.

**Keywords** Single coronary artery • Coronary artery anomaly • Prepulmonic • Anomalous left main coronary artery • Case report • Congenital heart disease

## Learning points

- In cases with difficult or unsuccessful coronary ostial engagement, coronary artery anomaly should always be considered.
- A potential malignant course of anomalous coronary artery should be ruled out with coronary computed tomography angiography.
- Single coronary artery with interarterial course is associated with high risk of sudden cardiac death.

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## Introduction

The term 'coronary artery anomaly' (CAA) is used when the observed coronary pattern is seen in less than 1% of the general population.[1] The overall incidence of CAA has been estimated between 0.9% and 5.6%.[2–4] Based on the origin and course of the anomalous artery, CAA can either represent a benign incidental finding or can have severe cardiovascular sequelae. Coronary computed tomography angiography (CTA) is a reliable non-invasive tool for diagnosing CAA. The management varies based on the nature of symptoms. Here, we present a case of inferior wall ST-elevation myocardial infarction due to distal right coronary artery (RCA) occlusion with incidental findings of anomalous left main coronary artery (LMCA) branching off the RCA.

## Timeline

| Day | Timing | Events |
|---|---|---|
| Day 1 | 5:25 a.m. | Patient presents with chest pain. |
| | 5:40 a.m. | Repeat electrocardiograms eventually demonstrated ST-segment elevation involving leads II, III, and aVF. Overhead 'cardiac alert' is called and patient is taken emergently for cardiac catheterization. |
| | 6:16 a.m. | Cardiac catheterization showing 99% distal right coronary artery (RCA) lesion and anomalous origin of left main coronary artery (LMCA) from RCA. |
| | 10:39 a.m. | Transthoracic echocardiography is performed which shows mild to moderately reduced ejection fraction. |
| Day 2 | 11:00 a.m. | Patient started on dual antiplatelet, statin therapy, and transferred out of cardiac care unit. Patient is scheduled for coronary computed tomography (CT) angiography. |
| Day 3 | 8:00 a.m. | Coronary CT angiography demonstrates benign prepulmonic course of aberrant LMCA. |
| | 11:00 a.m. | Patient discharged on aspirin, clopidogrel, atorvastatin, metoprolol-XL, and furosemide. |

## Case presentation

A 72-year-old Caucasian female presented to our facility with complains of new-onset substernal chest pain and shortness of breath. Other relevant medical history included active tobacco use and family history of premature coronary artery disease. On presentation, the patient was haemodynamically stable and physical examination including cardiovascular auscultation was unremarkable. Lab studies revealed leucocytosis 16 500 cells per cubic millimetre of blood (4000–11 000 cells/mm$^3$), normal serum chemistry, and elevated cardiac troponin I greater than 40 ng/mL (normal less than 0.04 ng/mL). Repeat electrocardiograms (EKG) eventually displayed ST-segment elevation in the leads II, III, and avF with concomitant ST-depression in reciprocal leads I and avL (*Figure 1A*). The patient was immediately transferred to the cardiac catheterization lab where she underwent coronary angiogram. After an initial failed attempt to first cannulate the left coronary ostium to evaluate for concomitant left anterior descending (LAD) and circumflex disease with the suspected culprit being the RCA based on EKG, the right coronary ostium was engaged instead, which revealed a large caliber RCA descending through the coronary sulcus to the crux, before bifurcating into the posterior descending artery and posterolateral artery. Contrast injection into the RCA also showed the anomalous origin of the LMCA stemming from the proximal segment of the RCA. The distal segment of the RCA showed 99% ulcerated lesion just prior to bifurcation, which was predilated and a 3.0 × 18 mm Xience Sierra drug-eluding stent was deployed with no residual stenosis. The aberrant LMCA reached the left side of the heart anterior to the pulmonary artery and trifurcated into a small caliber LAD, ramus intermedius (RI), and hypoplastic left circumflex arteries (*Figure 2*). There was less than 25% stenosis near the origin of the LAD and stable 50% atherosclerotic narrowing at the origin of diagonal branches. Transthoracic echocardiogram (TTE) performed post-procedure showed an ejection fraction (EF) between 40% and 45%, inferior and inferolateral wall hypokinesis and no valvular abnormalities (*Figure 1B*). A coronary CTA confirmed the findings of coronary angiogram and a potential malignant course of anomalous LMCA was ruled out (*Figure 3*). Further hospital stay remained uneventful, and the patient was discharged home on guideline-directed medical therapy with a beta-blocker, statin, and dual antiplatelet agents. At 1-month follow-up visit, the patient remained asymptomatic and a repeat TTE showed no new findings.

## Discussion

Single coronary artery anomaly (SCA) is a congenital anatomic abnormality identified by a single coronary ostium giving rise to all arteries supplying the heart. Single CAA is uncommon and seen in only 0.024–0.06% of cases.[5] In 1979, Lipton *et al.*[6] provided the angiographic classification of SCA based on the site of ostial location, the anatomical course of the single vessel and relationship of the aberrant transverse branch with respect to the great arteries of the heart. This was further modified by Yamanaka *et al.*[3] in 1990. Single CAA can either be right (R) or left (L) sided corresponding to the ostial location in the right or left sinus of Valsalva. Single CAA are classified into three groups from I to III with Group II consisting of various subtypes based on the course of the aberrant vessel (*Tables 1* and *2*). Villa *et al.*[7] in 2016 classified CAA functionally as (i) anomalies with obligatory ischaemia, (ii) anomalies without ischaemia, and (iii) anomalies with episodic ischaemia that occasionally cause severe events but are otherwise compatible with normal life.

In our case, the patient had congenital SCA; however, her acute presentation was unrelated to the aberrant vessel pathology. Based on the Lipton anatomic classification, the patient illustrated SCA type RIIA with the prepulmonic course of the aberrant vessel (LMCA). Potentially LMCA, RCA, and LAD can all branch from the SCA and take an aberrant course. Type II SCA involving the LMCA is fairly common and is seen in 5% of patients with tetralogy of

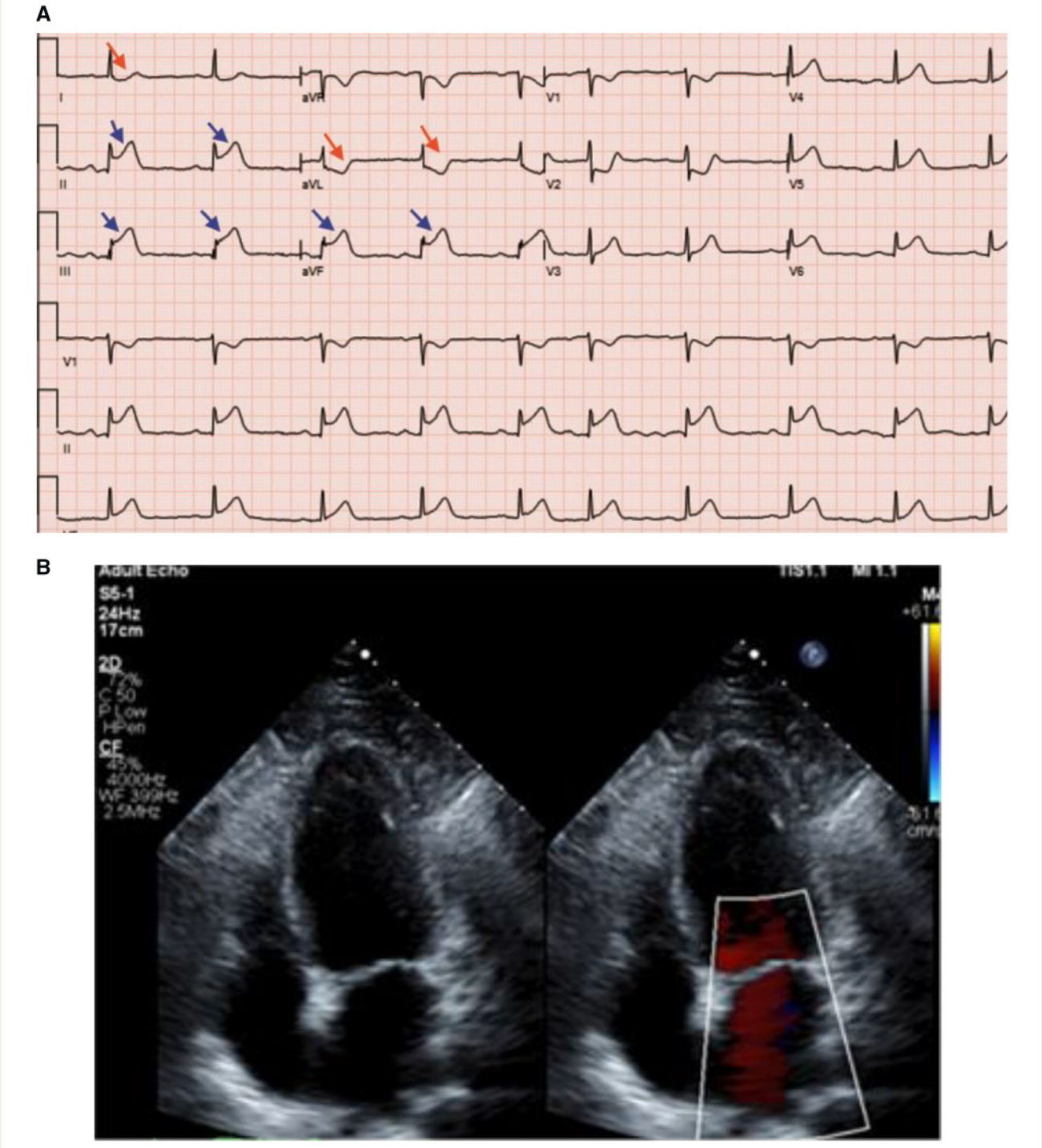


**Figure 1** (*A*) ST-segment elevation involving leads II, III, and aVF (blue arrow). Reciprocal ST-depression in leads I and aVL (red arrow). (*B*) Transthoracic echocardiogram (apical view) with inferolateral wall hypokinesis and no valvular abnormalities.

Fallot which can often complicate valve repair. As the vessel crosses the right ventricular outflow tract anteriorly, there are no significant haemodynamic consequences, however, isolated cases of angina have been reported.[8] The aberrant vessel can also take an interarterial, retroaortic, or septal (sub-pulmonic) route before reaching the left side of the heart. The five potential paths of the aberrant vessel before reaching the perfusion territory, correlate directly with the risk of SCA. Inter-arterial branch coursing between the pulmonary trunk and aorta has an aberrant intramural course within the aortic wall resulting in hypoplasia and lateral

compression.[9] Inter-arterial course can lead to sudden cardiac death (SCD). The mortality rates of left interarterial arteries (LAD or LMCA) are higher (57%) compared to right interarterial arteries such as RCA (25%). In the retroaortic subtype, the aberrant branch courses between the posterior aorta and the interatrial septum which can often complicate aortic valve surgery. This anomaly usually arises from RCA and is not haemodynamically significant. In the subpulmonic type of SCA, the aberrant branch travels anteriorly and inferiorly through the interventricular septum before coursing through septal myocardium. The subpulmonic type has a lower position, is surrounded by septal myocardium and does not have slit like orifice differentiating it from interarterial course.

When diagnosing suspected CAA, it is imperative that an exact anatomical course is established to assess the risk of SCD. The role of TTE in diagnosing CAA is limited. Inclusion of two new screening views by Thankavel *et al.*[10] improved the diagnostic ability of TTE in anomalous coronary artery from opposite sinus from 0.02% to 0.22%. Currently, coronary CTA and magnetic resonance angiography are Class I indications for diagnosing congenital CAA[11] Cardiac

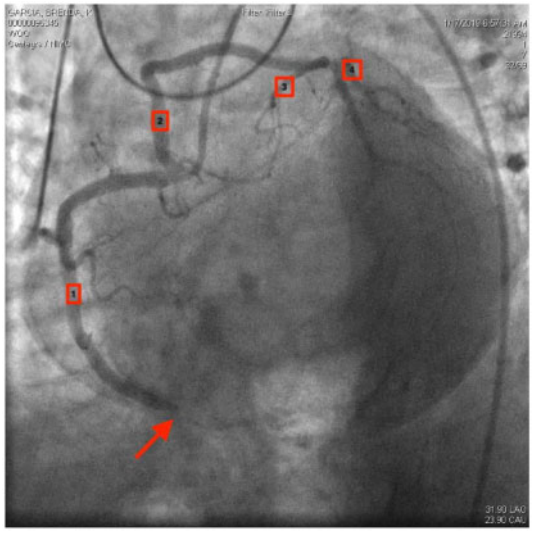

**Figure 2** Cardiac catherization showing distal 99% right coronary artery occlusion (red arrow). The left main coronary artery branching off the right main coronary artery and trifurcating into a small caliber left anterior descending, ramus intermedius, and hypoplastic left circumflex arteries. 1, right coronary artery; 2, left main coronary artery; 3, left circumflex artery; 4, left anterior descending artery.

**Table 1 Lipton's classification for single coronary artery**

| Ostia location | Anatomic distribution subtypes |
|---|---|
| Right sinus of Valsalva (R) | RI—SCA follows the course of a normal RCA |
| | RII—SCA from the right sinus gives off an anomalous transverse branch that crosses the base to reach the contralateral side |
| | RIII—SCA from the right sinus, with LAD and LCx arising from separate coronary trunks instead of single trunk |
| Left sinus of Valsalva (L) | LI—SCA follows the course of a normal LMCA |
| | LII—SCA from the left sinus gives off an anomalous transverse branch that crosses the base to reach the contralateral side |

LAD, left anterior descending artery; LCx, left circumflex artery; LMCA, left main coronary artery; RCA, right coronary artery.

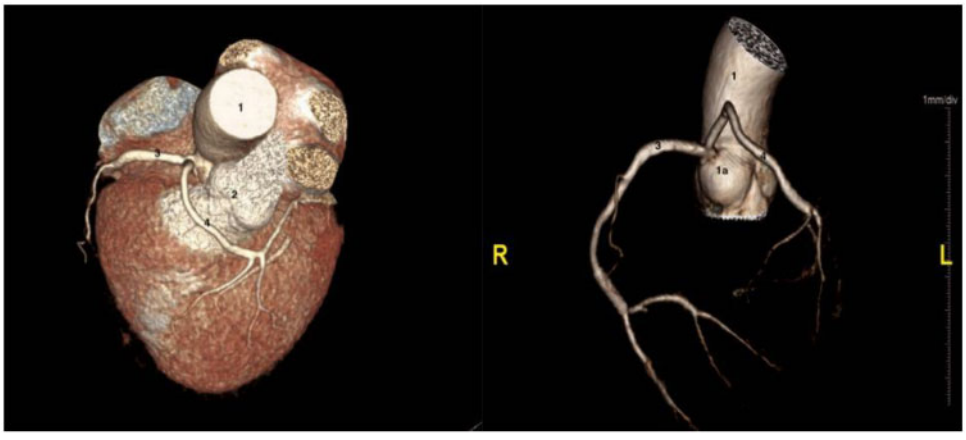


**Figure 3** Right anterior view of the three-dimensional constructed computed tomography image of the heart (left) showing the right coronary artery giving rise to the left main coronary artery and the latter's course anterior to the main pulmonary trunk. Constructed computed tomography coronary tree (right) showing single coronary ostium in the right aortic sinus giving rise to right coronary artery (with stent). 1, ascending aorta; 1a, right aortic sinus; 2, pulmonary artery; 3, right coronary artery; 4, left main coronary artery.

**Table 2 Subtypes of type II single coronary artery based on the course of the aberrant vessel**

| Anatomic distribution | SCA subtypes based on course of anomalous transverse branch |
|---|---|
| RII or LII | Type A—courses anterior to the pulmonary trunk |
| | Type B—courses between pulmonary artery and aorta |
| | Type P—courses posterior to the aorta |
| | Type S—septal type courses above the interventricular septum |
| | Type C—combined |

CTA allows for improved, non-invasive visualization of CAA albeit at the expense of radiation and contrast exposure. Superiority of coronary CTA was depicted by Shi *et al.*[12] in a report that showed conventional angiography was diagnostic in only 53% of CTA proven CAA cases. Magnetic resonance angiography is an alternative that can simultaneously assess structural abnormalities without requiring contrast or harmful radiation exposure however the use remains limited. Coronary angiography remains a useful test to diagnose and classify CAA and is the gold standard for identifying associated coronary artery disease. Traditional angiography offers limited visualization of the coronary ostia, proximal course, and surrounding structures. Ali *et al.*[13] recommended looking for the presence of two signs that should raise suspicion for coronary anomaly (i) the unperfused myocardium sign where the vessel supplying that myocardial territory is not visualized. (ii) The 'aortic root' sign where the vessel appears to cross the aorta and the pulmonary artery at the level of aortic root. In cases with difficult LMCA cannulation, a cuspogram can be performed to visualize the partly opacified artery. When the cuspogram is unsuccessful, right, and non-coronary sinus should be engaged to look for the anomalous origin of LMCA. The 'dot and eye method' was tested by Ishikawa *et al.*[14] to determine the true course of CAA radiologically.

The role of non-invasive functional testing to assess the haemodynamic effect of SCA on myocardial perfusion remains controversial. Both exercise treadmill and stress myocardial perfusion scan can yield false-positives and false-negative results. The use of fractional flow reserve (FFR) and intravascular ultrasound (IVUS) to guide therapy is increasingly gaining attention. Fractional flow reserve can help assess the haemodynamic flow of the aberrant vessel and IVUS can provide an insight on the ostial anatomy. Intravascular ultrasound can help identify slit like or stenosed orifice, acute angle takeoff, and intramural aortic segment (coursing through aortic wall) all of which can increase risk of SCD. Together FFR and IVUS can helps establish the need for intervention especially when the presenting symptoms are atypical.[15]

Treatment options include observation with conservative medical management, percutaneous intervention, or surgical repair. Proximal stenosis in cases with SCA can be devastating if adequate collaterals do not exist. The 2008 guidelines for adults with congenital heart disease recommend revascularization or surgical repair of interarterial course regardless of symptoms due to higher rates of myocardial infarction and surgical revascularization on follow-up[11] Aberrant vessels with prepulmonic, retroaortic, or transseptal courses have excellent prognosis. Asymptomatic patients with high risk anatomic features and absence of ischaemia on stress testing need multidisciplinary approach.

## Conclusion

We report a rare case of an incidentally identified SCA with Lipton type RIIA pattern. Although the majority of the patients are asymptomatic further evaluation with coronary CTA is warranted to rule out potential malignant pathology. The risk of SCD is highest with the interarterial course of the aberrant vessel. Stress testing is often not reliable in assessing the functional status of the patient. The optimal approach involves risk stratification, multidisciplinary management, and surgical intervention in appropriate patients.

## Lead author biography

Dipesh Ludhwani completed his medical school in India before working as a research assistant at Rutgers New Jersey Medical School. He started his Internal Medicine residency at Chicago Medical School, Northwestern McHenry Hospital in 2017. Currently, a second-year resident, Dr Ludhwani has been unanimously selected as the future third-year chief resident for his residency programme. He is interested in pursuing Cardiology fellowship and has published and presented numerous abstracts at regional and national conferences.

## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

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