Anomalous Coronary Arteries: Two Unique Cases as Viewed by Invasive Angiography with Brief Review of Literature

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Abstract: Coronary artery anomalies have a prevalence of less than 1% in the general population with an incidence ranging from 0.17% in autopsy cases to 1.2% in angiographically evaluated cases [1]. A single coronary artery is among the rarest coronary artery anomalies, seen in only 0.024-0.044% of the population [2]. Here, we report two cases of single coronary artery anomalies with two different presentations and classifications.

INTRODUCTION

Coronary artery anomalies have a prevalence of less than 1% in the general population with an incidence ranging from 0.17% in autopsy cases to 1.2% in angiographically evaluated cases [1]. A single coronary artery is among the rarest coronary artery anomalies, seen in only 0.024-0.044% of the population [2]. Here, we report two cases of single coronary artery anomalies with two different presentations and classifications.

CASE 1 DESCRIPTION

A 61-year-old female with a history of right renal artery stenosis, hypertension, and recurrent pneumonia presented with a one-day history of worsening dyspnea. Electrocardiography (ECG) demonstrated sinus tachycardia and an inferior infarct. Transthoracic echocardiography demonstrated decreased systolic function with an ejection fraction of 40-45% with severe hypokinesis of mid-apical, midanteroseptal, mid-inferoseptal, and apical septal walls. Coronary angiography visualized the right coronary artery (RCA) originating from the distal left circumflex artery (LCX) without any stenosis (Figure **2a**, **c**). There was no coronary artery originating from the right coronary sinus (Figure **2b**).

CASE 2 DESCRIPTION

A 66-year-old female with a past medical history of hypertension, diabetes, and a remote history of pulmonary embolism presented with a two-hour history of chest pain with radiation to her bilateral upper extremities, shortness of breath, lightheadedness, and diaphoresis. The patient initially was found to be bradycardic with a heart rate of 42bpm. ECG showed sinus bradycardia with 3-4mm inferior ST elevations and reciprocal depressions in the lateral leads. Coronary angiography visualized an occluded right posterior lateral (RPL) artery supplying the posterior left ventricular wall which was the culprit stenosis of the acute myocardial infarction. Interestingly, her left main coronary artery (LM) has an anomalous origin from the right coronary sinus. There was also severe stenosis in the left anterior descending (LAD) and LCX arteries. After PCI (percutaneous coronary intervention) to the RPL, she had staged PCI to the LAD and LCX (Figure **3a-c**). Coronary CT angiogram with 3D imaging illustrated the LM arising from the right coronary cusp, sharing its ostium with the RCA (Figure **1**). The LM has a benign posterior course between the right atrium and the aorta.

DISCUSSION

A single coronary artery is among the rarest coronary artery anomalies, seen in only 0.024-0.044% of the population [2]. Lipton *et al.* established nine patterns of single coronary arteries based on site of origin and anatomical circulation seen in Table **1a** [3]. Yamanaka and Hobbs in 1990 modified the classification previously described by Lipton *et al. and* designated each coronary anomaly with an R or L according to where the coronary anomaly originates [4]. The next level of classification is based on anatomical distribution. In type I, after the anomalous origin, the vessel follows the course of normal left or

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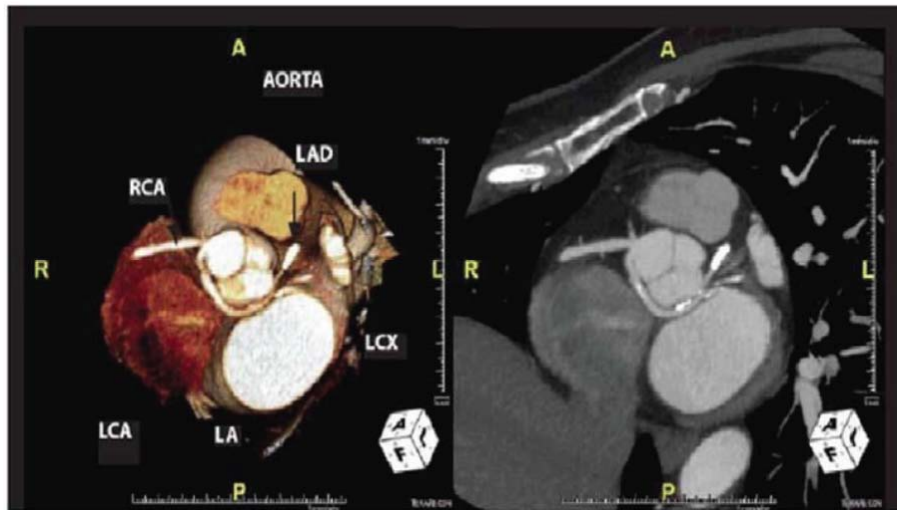
Figure 1: 3D reconstruction of CT images showing anomalous origin of left main coronary artery originating from right coronary cusp which is the R III subtype (RCA: Right coronary artery, LCA: Left coronary artery, LAD: Left anterior descending, LCX: Left circumflex artery, LA: Left atrium).
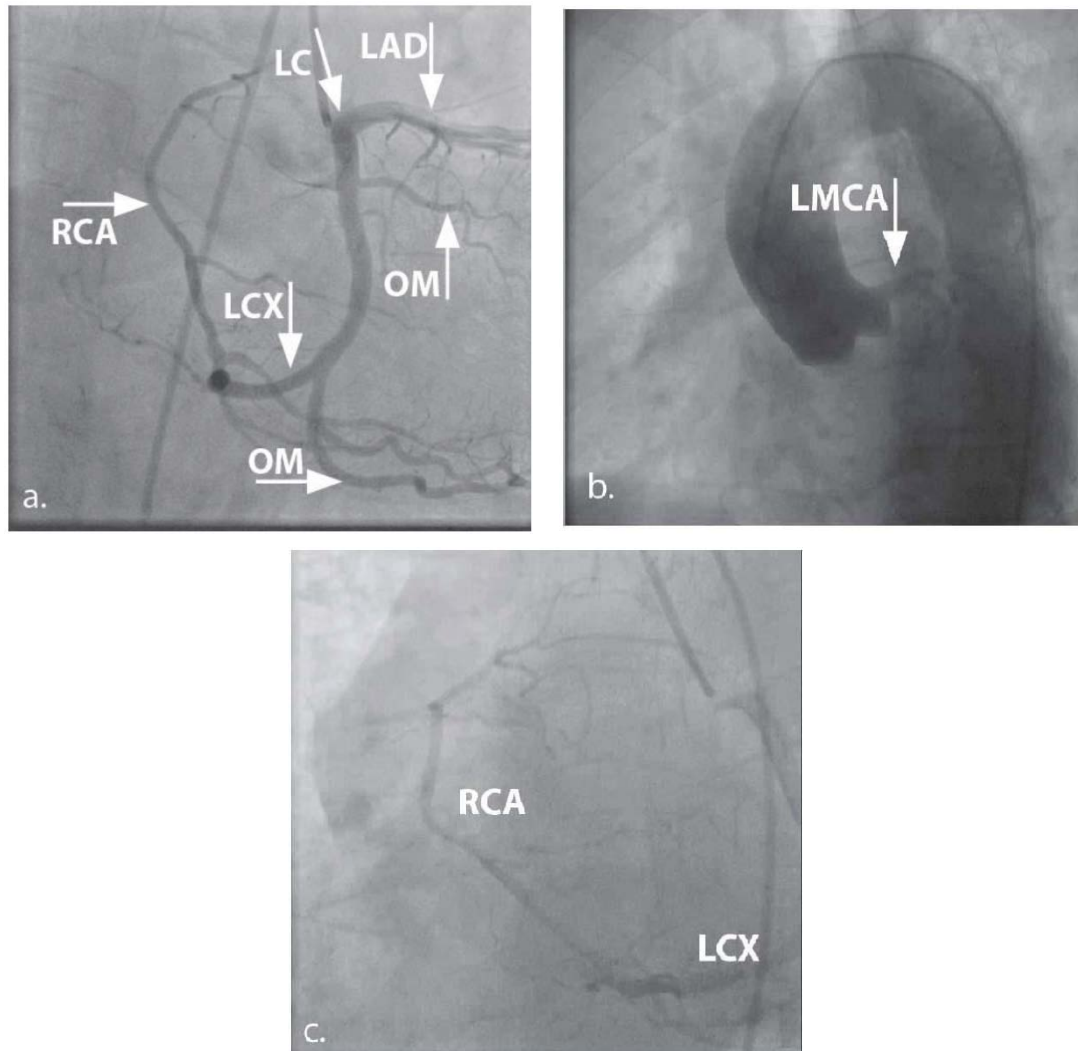

Figure 2: a-c: View of the RCA from the distal circumflex with no right coronary ostium which is the L-1 subtype. (pRCA: Proximal right coronary artery, mRCA: Middle right coronary artery, dRCA: Distal right coronary artery, LMCA: Left main coronary artery, LCX: Left circumflex artery, LM: Left atrium).
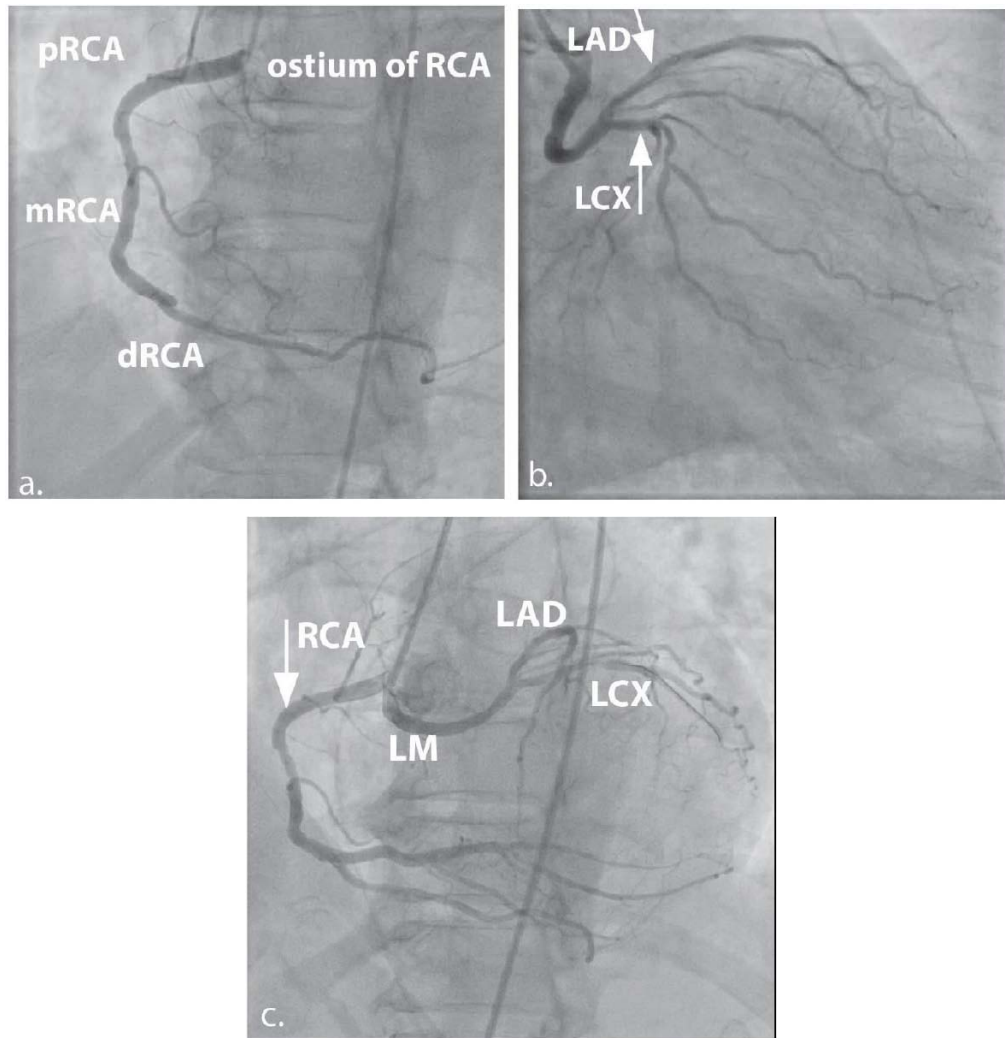
Figure 3: a-c: View of the left main coronary artery originating from right coronary cusp which is the R III subtype. (RCA: Right coronary artery, LAD: Left anterior descending, LCX: Left circumflex artery, LM: Left atrium).

Table 1: Coronary Artery Anatomical Distribution and Location- Nomenclature of origin, ostium, and course adapted from Lipton [3]

	Classification	Description
Ostial Location	O	Right sinus of valsalva
	O	Left sinus of valsalva
Anatomical Distribution	I	Solitary dominant vessel follows the course of either a normal right or left coronary artery
	II	One coronary artery arises from the proximal portion of the normally located other coronary artery
	III	LAD and CX arise separately from a common trunk originating from the right sinus of valsalva
Course of the Transverse Trunk	A	Anterior to the great vessels
	B	Between the aorta and pulmonary artery
	P	Posterior to the great vessels
	S	Septal type passes through the interventricular septum
	C	Combined type: A combination of diverse routes
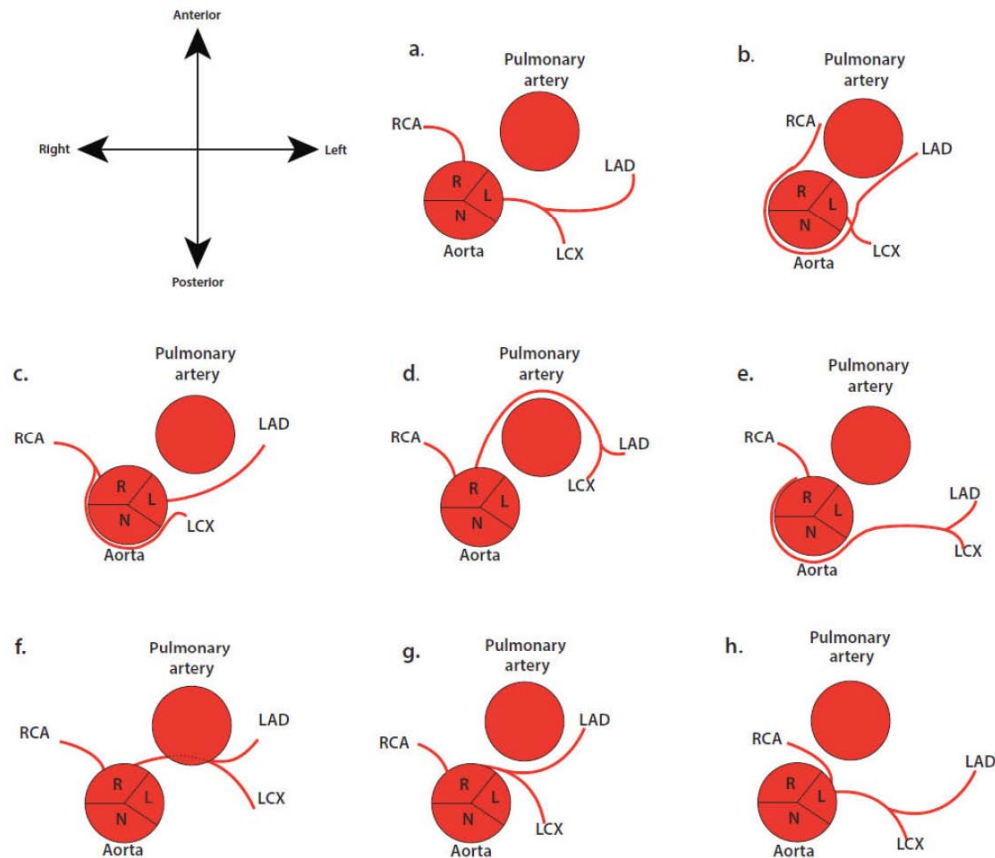
Figure 4: Normal coronary artery origin and course contrasted with several variants anomalous aortic origin of a coronary artery (AAOCA). a) Normal coronary artery origin and course. **b**) Anomalous origin of the LCX from the RCA with a posterior (retroaortic) course; this is more commonly encountered than AAOCA of the LCX from the right sinus of Valsalva. **c**) AAOCA of the LM with an anterior (prepulmonic) course. **d**) AAOCA of the LM with a posterior (retroaortic) course. **e**) AAOCA of the LM with a subpulmonic (intramyocardial) course. **f**) AAOCA of the LM with an interarterial course. **g**) AAOCA of the RCA with an interarterial course. **h**) *LCX* left circumflex coronary artery, *RCA* right coronary artery, *LAD* left anterior descending artery.

right coronary artery with a continuation into the missing artery's territory [5]. Type II shows the anomalous artery budding from the proximal part of the other normal artery and courses the base of the heart before taking the native course [5]. Lastly, in type III the LAD and CX arteries originate from the proximal part of the RCA [5]. Coronary artery anomalies are also differentiated in relation to the route taken to reach its vascular territory. Letters A, B, P, S, and C are used to delineate the course of the vessel with respect to the pulmonary artery and the aorta which is illustrated in Figure **4**. Certain anomaly classifications usually have a benign clinical course, such as RI, LI types. Meanwhile, the anomalous coronary artery courses between the outflow tracts such as R/LIIB or RIII are more prone to serious clinical complications like sudden cardiac arrest [6].

Case 1 illustrates an anomalous RCA from the distal circumflex with no right coronary ostium which is the L-1 subtype (Figure **4b**). The RCA coming from the distal circumflex is extremely rare [7]. The majority of

reported cases of RCA anomalies are from the proximal or middle portion of the LAD, whereas ours is the distal portion. Such an anomaly is not expected to have any severe complications or cause any myocardial ischemia in the absence of atherosclerotic disease. Case 2 demonstrates the left main coronary artery originating from right coronary cusp which is the R III subtype, also extremely rare (Figure **4e**). Due to its course between the RA and aorta this is benign with no need for surgical intervention.

CONCLUSION

The majority of patients do not exhibit symptoms when diagnosed with SCA during coronary angiography. Numerous patients will have negative cardiac workup or atypical chest pain, but a few will have typical chest pain, syncope, palpitations, ventricular tachycardia, and even myocardial infarction [6]. Here, we report two cases of rare congenital coronary artery anomalies with two different presentations and classifications. Many of these

patients usually present with no specific symptoms. However, in patients with previous undiagnosed anomalies, physicians should know about these uncommon peculiarities as some of these patients are at a high risk of cardiac complications. Coronary CTA is the gold standard imaging modality to demarcate the coronary artery anomalies and practitioners will have a better understanding of the clinical significance to appropriately diagnosis and treat these patients [2].

CONFLICTS OF INTERESTS

No Funding was received for this work; No conflicts of interests exist from any of the authors.

CONFIDENTIALITY AND RECORDS

Patient consent was secured from both patients to be a part of this paper. Any identifying information was removed, and informed consent was obtained from the patients throughout the entire process.

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