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CASE REPORT Open Access

Anomalous origin of the left main coronary artery in a 62-year-old woman: a case report and review of the literature

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Abstract

Introduction Abnormalities of the coronary arteries, including abnormal origins, are often detected in heart patients who undergo coronary angiography. Although only a small percentage of these abnormalities lead to serious complications, the consequences can be extremely fatal.

Case presentation We report the case of a 62-year-old woman who presented with chest pain. The patient has a history of hypertension and complains of chest pain that worsens with walking and improves with rest. A basic electrocardiogram (ECG) showed sinus rhythm with no signs of arrhythmia or ischemic changes. Coronary angiography revealed an anomalous left coronary artery originating from the right sinus of Valsalva (RSoV).

Conclusions We report a rare case of an anomalous left coronary artery originating from the RSoV without specific clinical symptoms. The patient presented with unstable angina, and after further investigation, this anomaly was diagnosed. Investigating these abnormalities, especially in young individuals with cardiac symptoms, should be prioritized. Cardiologists should also be aware of this rare condition that can have fatal consequences.

Keywords Coronary vessel anomalies, Diagnosis, Complications, Therapy, Left main, Coronary artery

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Introduction

Studies have shown that coronary artery anomalies are present in 0.64–1.3% of patients undergoing coronary angiography. Although most reported anomalies are benign, a small percentage can cause serious complications for patients [1]. It is very rare for three coronary arteries to originate from the same coronary orifice. In some cases, a coronary artery may originate from an ectopic aortic sinus [2]. In such cases, catheterization of both coronary arteries can be challenging due to the absence of one of the arteries from the usual aortic sinus ostium.

The clinical manifestations of coronary artery anomalies largely depend on the location of the anomaly. Taking a comprehensive history and considering the possibility of a coronary artery anomaly, especially when the patient's symptoms are otherwise unexplained, is critical for physicians to diagnose and prevent further complications. A review of the literature indicates that the anomalous origin of the left main coronary artery (LMCA) is particularly rare and warrants closer attention.

We describe a 62-year-old woman whose evaluation for chest pain revealed an anomalous left coronary artery arising from the RSoV. We discuss the clinical significance of this anomaly by reviewing the literature, exploring the views and complications associated with this condition, and outlining the current treatment approaches.

Case report

A 62-year-old woman (height: 155 cm, weight: 80 kg, BMI: 29.8) with a history of typical chest pain was referred to the angiography department for further investigation and angiography after visiting the cardiologist. The patient has a history of hypertension and complains of chest pain that worsens with walking and improves with rest (exertional chest pain, Functional Class I-II). She uses valsartan 160 mg and amlodipine 5 mg tablets to control her hypertension.

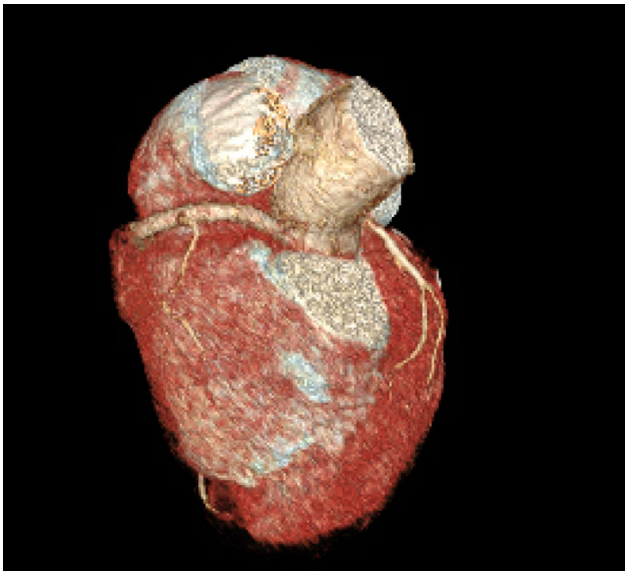

Fig. 1 In the sagittal view in the reconstructed image of CT scan angiography, it was found that the LMCA was separated from the RSoV

The patient was fully conscious with a pulse rate of 80 beats per minute, blood pressure of 135/80 mmHg, body temperature of 36.5 °C, respiratory rate of 16 breaths per minute, oxygen saturation of 98%, and a Glasgow Coma Scale (GCS) score of 15. A basic ECG showed sinus rhythm with no signs of arrhythmia or ischemic changes. An echocardiogram was performed on the patient, and the findings are as follows: normal LV size and systolic function (LV EF = 55%), mild diastolic dysfunction (DDG1), normal RV size and function, no valvular heart disease, and normal SPAP. Although her exertion stress test was negative, she experienced chest pain two- or three-times during exercise despite medical treatment, including bisoprolol 5 mg daily and nitroglycerin 2.6 mg bid. Therefore, angiography was decided. Given the patient's symptom (exertional chest pain), the patient underwent angiography, during which this anomaly was incidentally discovered. To further determine its type, a coronary CT angiography was subsequently performed, which fortunately revealed that it was not of the intra-arterial type. Given the non-significant lesions in the coronary arteries (mild CAD) and the presence of hypertension, the patient was started on treatment with statins, ASA, amlodipine, and valsartan.

Coronary angiography

In our patient, despite considerable effort and time, the left sinus of Valsalva and the LMCA were not initially observed. However, upon entering the RSoV and injecting the contrast agent, both the LMCA and the right coronary artery (RCA) were observed simultaneously. The LMCA was found to originate from the upper and left part of the RCA. None of the coronary arteries had significant stenosis; the left anterior descending (LAD) and left circumflex (LCX) arteries exhibited mild stenosis below 50%. To accurately diagnose the type of anomaly, a Computed Tomography Angiography (CTA) of the coronary arteries was performed, which confirmed a non-malignant form of the anomaly (Fig. 1).

The angiographic film showed an anomalous left coronary artery arising from the RSoV (Fig. 2). The patient's vital signs were monitored every hour after angiography. Additionally, the patient received 1000 cc of normal saline intravenously over 6 h.

CT angiography

CT angiography examination of the patient showed that the RCA and LMCA are both separated from the right

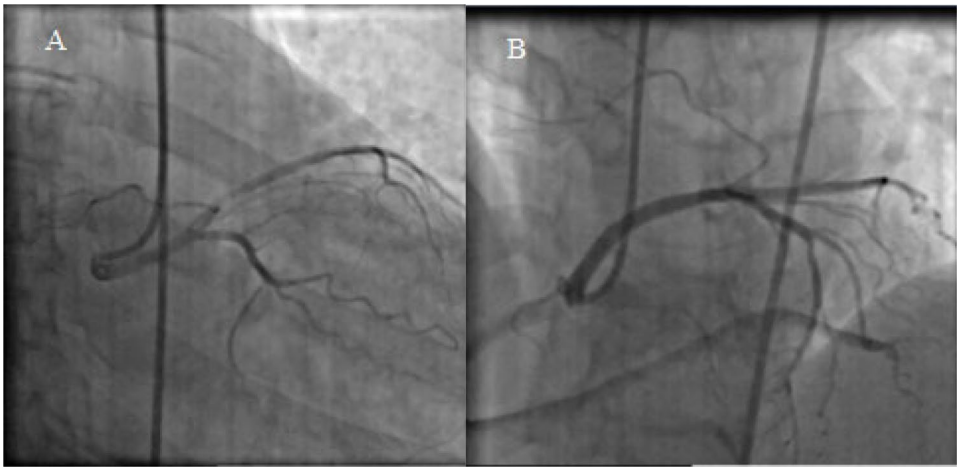

Fig. 2 Angiography revealed simultaneous visualization of the RCA and LMCA during contrast injection into the right sinus of Valsalva. (**A**: AP view, **B**: LAO view)

sinus of Valsalva. The RCA originates superiorly, while the LMCA originates inferiorly from the right sinus of Valsalva (Figs. 3 and 4). Also, the LMCA does not pass between the pulmonary artery and the aorta (non-malignant form) (Fig. 5).

Discussion

Congenital anomalies of the coronary arteries are among the rare causes of sudden death, which occur mostly during or immediately after intense physical activity [3]. These congenital coronary anomalies come in various types, with one of the most significant and sensitive being the origin of the LMCA from the RSoV. This anomaly is extremely rare, with an estimated prevalence ranging from 0.02 to 0.05% [4]. To better understand the various dimensions of such abnormal origins of coronary arteries, we examined cases reported in the literature (Table 1).

Type of anomaly

There are four subtypes of anomalies where the LMCA originates from the RSoV: (1) Interarterial: In this subtype, the LMCA passes between the aorta and the pulmonary trunk. Due to this positioning, during physical activity when blood flow increases in the aorta and pulmonary artery, the LMCA is compressed, leading to ischemia and consequently severe chest pain or cardiac arrhythmia; (2) Prepulmonic: Here, the LMCA travels anteriorly towards the right ventricular outflow tract; (3) Subpulmonic: The LMCA moves beneath the pulmonary artery along the proximal interventricular groove; (4) Retroaortic: The LMCA originates from the RCA and encircles the aortic root [5]. The intraarterial form is malignant and can cause severe ischemia or dangerous arrhythmias. The exact mechanism of coronary ischemia in these patients is not well understood. Several hypotheses include compression of the smaller, slit-like LMCA in the intraarterial form leading to loss of blood flow, vasospasm due to endothelial dysfunction, intramural compression due to high blood pressure, and bulging that blocks blood flow during increased demand [6].

The other three forms are generally benign and do not usually cause cardiac symptoms or arrhythmias. In a study by Moodie et al., angiographic reports of two patients showed LMCA originating from the RSoV and passing between the aorta and the pulmonary trunk (Type 1). In one of these patients, it led to an anterior myocardial infarction [7]. However, in other studies, the specific type of anomaly was not detailed, and most references broadly mentioned the origin of the LAD from the RSoV. In our patient, the LMCA was separated from the upper and left part of the RCA, and none of the coronary arteries had significant stenosis. To accurately diagnose the type of anomaly and assess potential risks, coronary CTA is necessary. In this case, coronary CTA reported a non-malignant form.

Symptoms

Our patient presented with chest pain while walking, which improved with rest. She also reported chest pain that did not respond to medication. Patients with coronary artery anomalies exhibit various symptoms, the most common being chest pain [7–11]. Other symptoms include shortness of breath [7–9] and syncope [7, 12]. In some cases, patients are asymptomatic, leading to sudden death [9]. Studies indicate that only 20% of patients present with symptoms such as angina induced by exertion, shortness of breath, or syncope, while the majority remain asymptomatic [13]. It is crucial to consider warning signs such as chest pain and syncope, especially in young individuals during physical activity and sports, as these symptoms could indicate conditions like

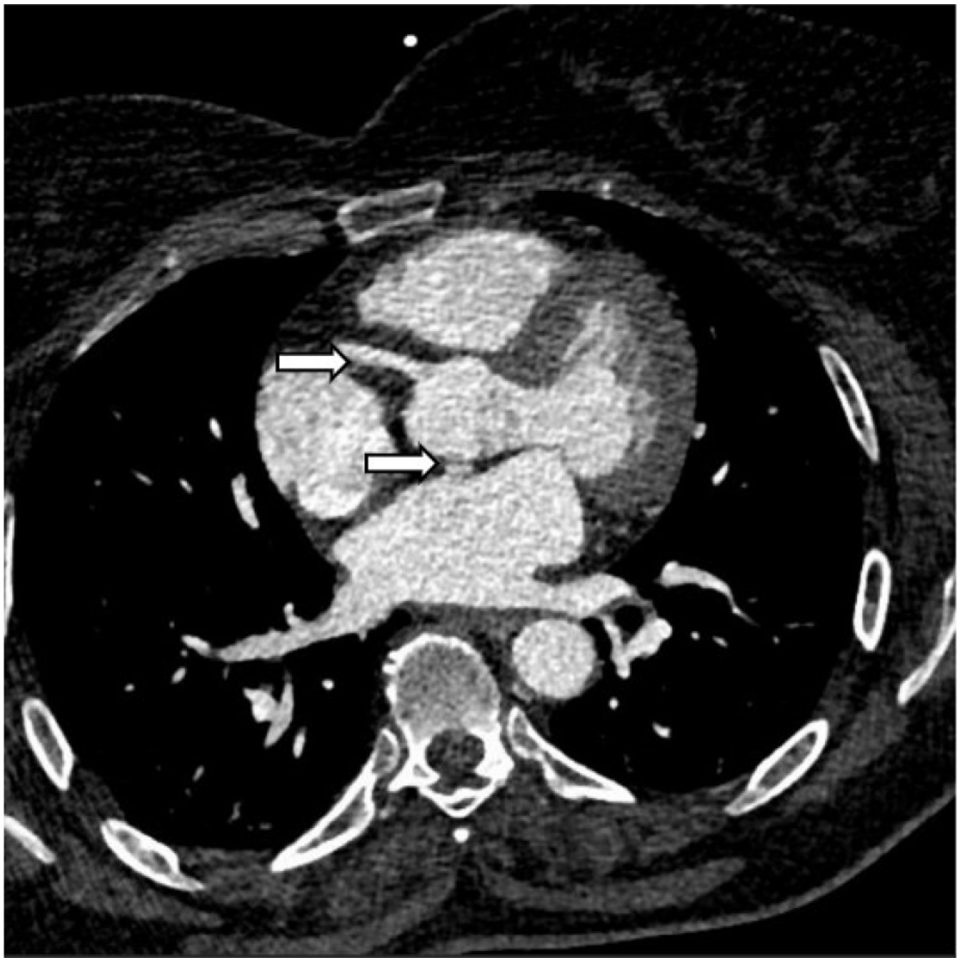

Fig. 3 Axial CT angiography section of the aorta and coronary arteries. The RCA and LMCA are both separated from the right sinus of Valsalva. The RCA originates superiorly, while the LMCA originates inferiorly from the right sinus of Valsalva

hypertrophic cardiomyopathy, long QT syndrome, and coronary artery anomalies [14]. Given the non-specific nature and wide range of symptoms associated with coronary artery anomalies, evaluating patients with various symptoms, particularly younger individuals, is essential.

Gender and age

Our patient is a 62-year-old woman. In a review of existing studies, out of 16 reported cases, 15 were men and only 5 were women. The oldest individual reported was an 81-year-old man [9]. Most cases of sudden death occurred in individuals under 20 years old, with clinical manifestations typically following intense physical exercise. Only two cases of cardiac arrest in older individuals have been reported, one of whom was a known CHF patient [9, 11]. No definitive correlation between gender and age with coronary artery anomalies has been reported, indicating the need for further studies to better understand the impact of gender on these anomalies.

Changes in ECG and exercise test

In our case study, the patient presented with exertional chest pain that was unresponsive to medical treatment, while her ECG showed normal findings without any ischemic changes or evidence of myocardial infarction. Studies on ECG findings in symptomatic patients revealed various changes ranging from normal to bundle branch block [7–9, 11] and ST elevation [7, 9, 15]. Additionally, cardiac arrest [9] was reported as a variable symptom. The exercise stress tests performed in only four cases were all reported as positive [7, 8, 10]. Despite a negative exercise test in our patient, she continued to experience chest pain during activity despite medical management, prompting the decision to proceed with angiography.

It is important to note that specific ECG changes indicative of coronary artery anomalies is not always present. Even in studies where patients had normal ECGs, anomalies were detected. Therefore, relying solely on ECG and exercise test results may not be sufficient. If symptoms persist, further comprehensive evaluations are necessary

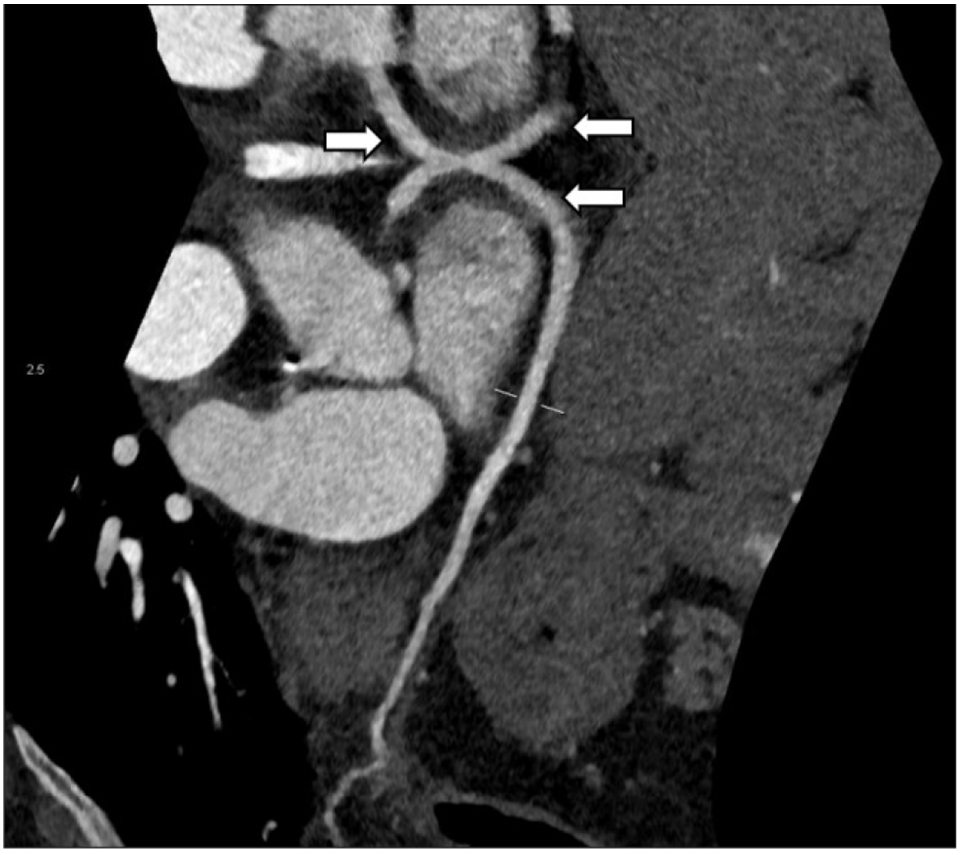

Fig. 4 Axial CT angiography section of the aorta and coronary arteries. The RCA and LMCA are separated from the right sinus of Valsalva

to detect coronary artery anomalies promptly and prevent irreversible complications.

Angiographic findings

Coronary angiography in our patient did not reveal any coronary artery involvement. Among patients who underwent angiography, seven cases showed coronary artery involvement in addition to the reported anomalies [7, 10, 16]. These findings significantly influence treatment decisions. In cases of sudden cardiac death, post-mortem examinations have shown coronary artery anomalies [9, 11]. Given that coronary angiography is the diagnostic method of choice for detecting coronary artery anomalies, it is recommended to perform this procedure in symptomatic individuals whose symptoms remain unexplained, ensuring thorough evaluation for the presence of anomalies. However, for a definitive diagnosis and understanding of the type of anomaly in these patients, CT coronary angiography is necessary.

Recommendations and treatment

The treatment of coronary artery anomalies remains controversial and depends on the identified anomaly type. Management decisions are guided by ischemic symptoms and the risk of sudden death. Surgical intervention is the primary treatment modality, although beta-blockers and calcium channel blockers are used to alleviate ischemic symptoms [17]. For instance, patients presenting with angina, syncope, or malignant ventricular arrhythmias, especially young individuals under the age of 35, should undergo surgical correction due to the high risk of sudden death [18]. In individuals over 35 years old, periodic examinations and exercise testing are recommended for the diagnosis of ischemia, ventricular arrhythmias, or abnormal coronary perfusion [19]. Patients with untreated LMCA anomalies arising from the RSOV should generally avoid vigorous exercise due to the increased risk of sudden death during exertion [14].

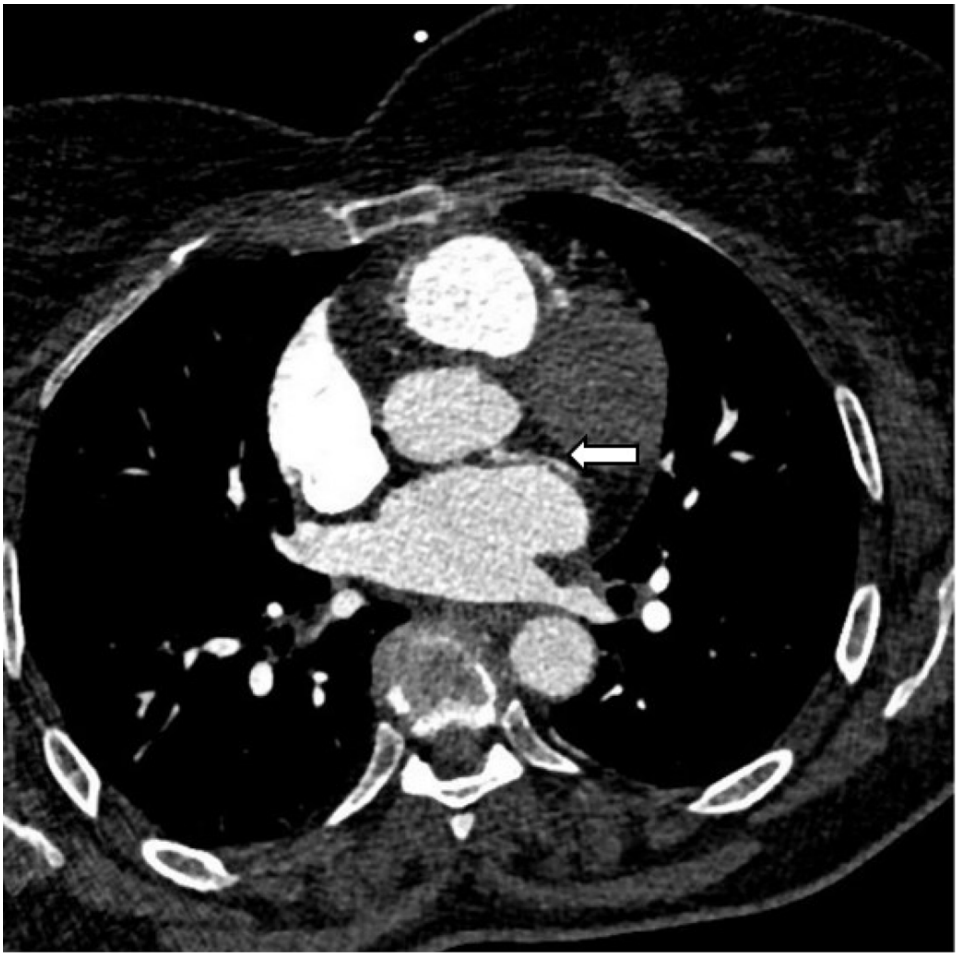

Fig. 5 Axial CT angiography section of the aorta and coronary arteries. The LMCA does not pass between the pulmonary artery and the aorta, indicating a non-malignant form

Table 1 Characteristics of 9 included case reports

Author, year	Country	Age, gender	Symptoms	ECG	Cardiac history	Final Clinical Diagnosis/ out put	exercise testing/ 99 m sesta-mibi study	Coronary angiography
Waters et al. 1992 [5]	USA	Female, 51	Chest pain and palpitations	-	-		The myocar-dium and significant ST-T wave changes in the lateral leads	-
		Male, 59	Chest dis-comfort and exertional dyspnea	Left anterior fascicu-lar block	-	Left ventricular hypertrophy and aortic stenosis		The LMC artery originated from the right sinus of Valsalva

Table 1 (continued)

Author, year	Country	Age, gender	Symptoms	ECG	Cardiac history	Final Clinical Diagnosis/ out put	exercise testing/ 99 m sesta-mibi study	Coronary angiography
Barth III and Roberts 1986 [6]	USA	13, Female	No	-	-	Sudden death (Shortly after exertion)	-	-
		14, Male	AP = angina pectom	Normal	-	Sudden death (Shortly after exertion)	-	-
		19, Male	No	VPC = ventricular premature com-plexe, LAH = left antenor hemiblock	-	Sudden death (Dunng exertion)	-	-
		64, Female	D = dyspnea	AMI = acute myo-cardial infarction, LBBB = left bundle branch block	CHF = conge-tive heart failure	Sudden death (CHF)	-	-
		81, Male	No Information		-	Sudden death (Alcoholism)	-	-
Hussam Eddin T. Al Hennawi [7]	Saudi Arabia	43, female	Cardiac arrest	Diffuse ST-segment elevation (STEMI) in the anterior and lateral leads, with reciprocal changes in the inferior leads	-	The patient recovered and was dis-charged for rehabilitation therapy.	-	The RCA with an initial failed attempt to engage into the LMCA
Pankaj Jariwala [8]	India	56, male	Chest discomfort	Normal	Chest discom-fort on exer-tion for the last 3 months	CAD	Positive for inducible ischemia at low workload.	The origin of the left coro-nary artery was found to be from the right coronary sinus.
Frederick Chua [9]	USA	22, male	Syncopal episode	-	No prior car-diac history		-	Anomalous origin of the LMCA that originates from a comomon ostium with the RCA
Vinay Kukreti [10]	United Kingdom	14 females	Chest pain, shortness of breath and cyanosis	Bifascicular block (RBBB) and left posterior fascicu-lar block) as well as significant ST changes suggestive of ischemia	One previous seizure like episode fol-lowing intense physical exertion.	Sudden death	-	Anomalous origin of the LMCA from the right sinus of Valsalva
Biana Trost [11]	USA	37- male	Chest pain	Normal	In 6 months, the pa-tient had experience stable angina pectoris	CABG	-	The LMCA originated from the RSoV

Table 1 (continued)

Author, year	Country	Age, gender	Symptoms	ECG	Cardiac history	Final Clinical Diagnosis/ out put	exercise testing/ 99 m sesta-mibi study	Coronary angiography
Doug-las S. Moodie [12]	USA	17- male	Collapsed	Anterior infarction	-	CABG	-	Anomalous left coro-nary artery arising from the RSoV and directed between the pulmonary artery and the aorta
		71- male	Shortness of breath, but no history of chest pain	Left anterior hemi-block with left atrial enlargement	Difficulty breathing and had an episode of syncope that lasted for 30 s	CAD	-	The LMCA originated anomalously from the right coronary sinus and passed between the pulmonary artery and the aorta
		57- male	Chest pain	-	Chest pain and episode of syncope while running	CAD	Positive	Anomalous origin of the LMCA and 70% narrowing of the right coronary artery
Anish Hirachan [13]	Egypt	49- male	chest pain	ST depression in inferolateral leads	-	CAD	-	Common origin of both the right main coronary artery and LMCA arising from RSoV with obstructive disease in middle segment of right coronary artery

AV = aortic valve; CAD = coronary artery disease; LM = left main coronary artery; MV = mitral valve; CABG = coronary artery bypass graft

Conclusion

Despite the rarity of coronary artery anomalies, they are of significant importance as they can lead to angina, syncope, and even sudden cardiac death. Therefore, investigating these anomalies, especially in young individuals with cardiac symptoms, should be prioritized. Angiography and CTA are currently considered reliable diagnostic methods, and surgical intervention remains the standard treatment approach, recommended for patients.

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Author contributions
MS, FJ and MZ write the initial report about patient, MZ, SP, FJ and NM were involved in investigation and data collection. NM, MZ and VM complete research and discussion, MZ, XX, AH, and MA drafted and corrected the manuscript. All authors read and approved the final manuscript for publication.

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Data availability
The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
This research has a code of ethics No. IR.RUMS.REC.1403.004 from Rafsanjan University of Medical Sciences.

Competing interests
The authors declare no competing interests.

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