Case Report

A rare case of single coronary artery with congenital absence of right coronary artery

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Abstract

A single coronary artery (SCA) is a relatively rare coronary artery anomaly. Absent right coronary artery (RCA) is the rarest of coronary artery anomalies occurring when the right coronary artery doesn't develop. In the past 14 years till 2020, only 26 cases of congenital absence of RCA have been reported in 24 articles. We report a rare case of single left coronary artery with congenital absence of right coronary artery detected by coronary angiography and confirmed by coronary CT angiography. The RCA territory was being supplied by a branch from left circumflex (LCX) artery. The patient presented with nonspecific central chest pain. We aim to shed light on this rare anomaly and raise awareness among medical practitioners.

Introduction

A single coronary artery (SCA) with congenital absence of right coronary artery (RCA) is a relatively rare coronary artery anomaly. Its prevalence is approximately 0.024%- 0.066% among patients who have undergone coronary angiography[1,2]. Absent right coronary artery (RCA) is one of the rarest coronary artery anomalies, which occurs due to failure to develop the right coronary artery in the atrioventricular groove. Between January 2010 - December 2020, only 26 cases of congenital absence of RCA have been reported in 24 articles[3]. Most of the patients with congenital coronary anomalies are asymptomatic and are often found incidentally. Some of these patients present with symptoms of myocardial ischemia, such as angina pectoris, chest heaviness or atypical chest pain. Only a small proportion of these patients suffer acute myocardial infarction or sudden death.

Case Report

A 70-year-old female, with a known history of CAD under anti-platelet, beta-blocker, angiotensin receptor blocker and oral nitrates presented to our hospital with the complaints of occasional nonspecific chest pain. The pain was central, not associated with exertion and increased in severity at night-time. She also complained of shortness of breath during exertion. She gave a history of severe chest pain four years prior to her current visit and was admitted to a hospital but documents were not available. Blood parameters like fasting lipid profile revealed an LDL cholesterol of 110 mg/dl, direct

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HDL cholesterol 86 mg/dl, triglyceride of 117 mg/dl. Complete blood count, renal functions and blood glucose levels were all in the normal range.

12 -lead electrocardiogram (ECG) showed (Figure 1) a sinus bradycardia with ST-T changes in chest leads V1 to V4. A transthoracic echocardiogram was done which showed mild to moderate mitral regurgitation, mild to moderate tricuspid regurgitation with mild pulmonary artery hypertension, mild to moderate aortic regurgitation, grade-1 left ventricular diastolic dysfunction with a left ventricular ejection fraction of 60% and no any regional wall motion abnormalities.

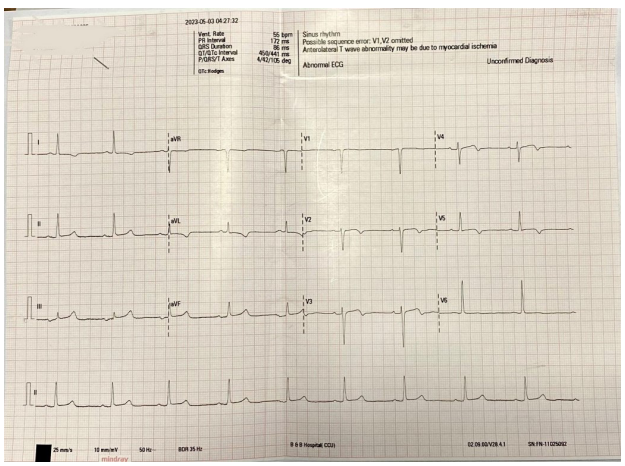

Figure 2: ECG shows sinus rhythm with ST-T changes in leads V1 to V4

During the course of treatment, the doses of beta-blocker and nitrates were increased with no improvement in symptoms. Then coronary angiogram was done which revealed 60% stenosis in mid left anterior descending artery (LAD), 70% stenosis of ostium of first diagonal branch of LAD (D1) with a dominant LCX. The RCA could not be visualized and anomalous absence was suspected. Coronary CT angiogram (Figure 3) showed single coronary artery with absent RCA. The RCA territory was being supplied by a branch from LCX. Rest of the coronary arteries had a normal origin and course with atherosclerotic plaques in proximal LAD leading to 70-80% stenosis.

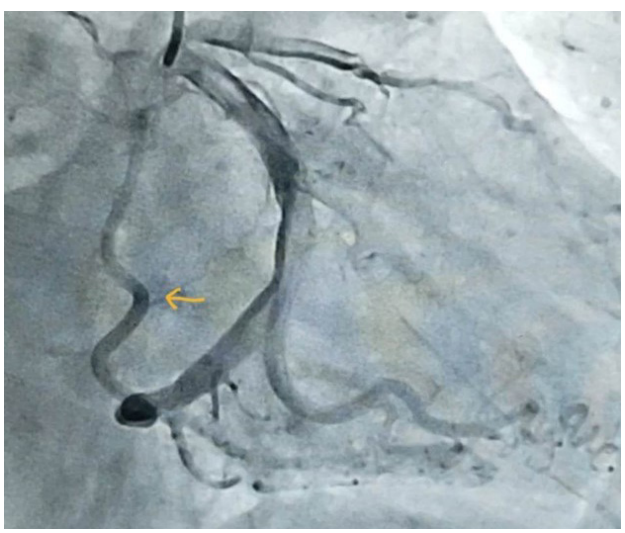

Figure 2: Coronary Angiography showing RCA territory being supplied by a branch of left circumflex artery

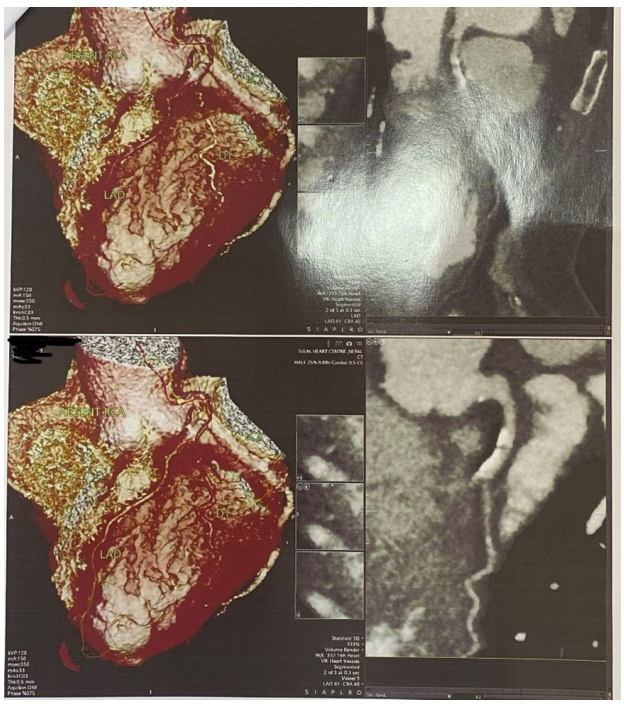

Figure 3: Coronary CT Angiography showing absent RCA and RCA territory being supplied by a branch from LCX.

Discussion

Congenital absence of RCA is rare with only 24 cases being reported in the last decade[3]. The underlying mechanism of this anomaly may be related to defects in fetal developmental process or congenital occlusion of coronary artery.

Most of the patients with congenital SCA anomaly are asymptomatic and found to present with non-specific clinical features and ECG changes. Some of SCA patients present with chest pain, myocardial infarction, ventricular tachycardia, syncope and even sudden cardiac death[4]. These patients largely remain undiagnosed and are often found incidentally[5]. This may be the reason for the low prevalence of this condition in the population.

The dangers of SCA lies in the fact that the patient has a single coronary blood flow system, which if gets affected might lead to arrhythmias, subsequent heart failure or catastrophic death. Few cases of SCA have presented with acute myocardial infarction[3].

According to Yamanaka and Hobbs[6], this case falls into a benign category as our patient had no features of syncope, cardiac arrhythmia, myocardial infarction or congestive heart failure.

Conclusions

The SCA with congenital absent RCA is a very rare coronary artery anomaly and is usually an incidental finding in a patient undergoing routine coronary angiography. Despite the absence of the RCA, patients with SCA have been reported to have normal cardiac function and carry a relatively low risk for cardiac events.

References:

1. Desmet W, Vanhaecke J, Vrolix M, Van de Werf F, Piessens J, Willems J, et al. Isolated single coronary artery: a review of 50,000 consecutive coronary angiographies. Eur Heart J [Internet] 1992;13(12):1637–1640.
2. Katekaru-Tokeshi DI, Jiménez-Santos M, Koppel CJ, Vliegen HW, Díaz-Zamudio M, Castillo-Castellón F, et al. Applicability of the Leiden convention and the Lipton classification in patients with a single coronary artery in the setting of congenital heart disease. J Cardiovasc Dev Dis [Internet] 2021;8(8):93.
3. Chen Z, Yan J, Han X, Adhikari BK, Zhang J, Zhang Y, Sun J, Wang Y. Congenital absence of the right coronary artery with acute myocardial infarction: report of two cases and review of the literature. J Int Med Res. 2020 Dec;48(12):300060520971508. doi: 10.1177/0300060520971508.
4. Said SA, de Voogt WG, Bulut S, Han J, Polak P, Nijhuis RL, Op den Akker JW, Slootweg A (2014) Coronary artery disease in congenital single coronary artery in adults: a Dutch case series. World J Cardiol 6:196–204.
5. Akcay A, Tuncer C, Batyraliev T, Gokce M, Eryonucu B, Koroglu S, Yilmaz R. Isolated single coronary artery. Circ J. 2008;72:1254–1258. doi: 10.1253/circj.72.1254.
6. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126595 patients undergoing coronary angiography. Catheter cardiovascular diagnostic procedure ?. 1990:21:28-40