

# Rare Case of Coronary Anomaly, Overview of Hypertrophic Cardiomyopathy with A Different Presentation

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## Abstract

Coronary artery anomalies are uncommon cardiac diseases. It is mostly detected incidentally in the adult population as well as being usually asymptomatic. Although its prevalence is low, its association with atherosclerosis, arrhythmias, and hypertrophic cardiomyopathy is important for sudden cardiac death. According to the literature, the association of hypertrophic cardiomyopathy with the coronary anomaly is rare. No case of hypertrophic cardiomyopathy accompanied by the absence of the left anterior descending artery has been reported. Therefore our case is special and valuable.

**Keywords:** Atrial Fibrillation, Coronary Anomaly, Hypertrophic Cardiomyopathy, Single Coronary Artery

## Introduction

Coronary artery anomalies are uncommon cardiac diseases. Although its prevalence is low, its association with atherosclerosis, arrhythmias, and hypertrophic cardiomyopathy is important for sudden cardiac death. (1,2,3) In order to talk about the coronary anomaly, we need to start by knowing the normal path of the coronary arteries. The left anterior descending artery (LAD) is the subepicardial artery, located in the anterior interventricular sulcus, giving septal penetrating branches. The circumflex artery (Cx) is the subepicardial artery, located in the left atrioventricular sulcus, giving at least one obtuse marginal branch. The right coronary artery (RCA) is the subepicardial artery, located in the right atrioventricular sulcus, giving at least an acute marginal branch. (4) Coronary artery anomalies can be classified in different ways. Shriki et al. grouped them under two headings as anomalies with hemodynamic severity and anomalies without hemodynamic severity. (5) The classification developed by Angelini and later Khatami et al. can be accepted as the most widely used classification. Based on these classifications, coronary artery anomalies can be divided into three or four large groups. Group A: coronary artery outflow abnormalities and distribution abnormalities, Group B: Intrinsic coronary artery anatomy abnormalities and Group C: Coronary artery termination

abnormalities, Group D: Abnormal collateral vessels. (6,7) The most common coronary anomaly is the atypical origin of CX from the right coronary circulation. However, data on left anterior descending artery anomalies are still insufficient in the literature. (4,8)

Hypertrophic cardiomyopathy is a genetic disease resulting from the thickening of the myocardium. It can cause complications such as heart failure, mitral valve disorders, atrial fibrillation, and sudden cardiac death. According to the literature, its association with coronary anomalies is rare. (2,3,9,10,11) Hypertrophic cardiomyopathy can cause sudden cardiac death, this risk increases when it is related to coronary anomaly.(11) For this reason, patients with hypertrophic cardiomyopathy with coronary anomaly should be observed closely.

## Case Report

A 37-year-old female patient presented to the emergency department with complaints of constricting chest pain and palpitations for several days. We were consulted by the emergency department with the initial diagnoses of atrial fibrillation with rapid ventricular response and acute coronary syndrome. It has been learned from her anamnesis that she had no known systemic disease or rhythm disorder and that she had not experienced syncope before. When her

family history was examined, it was revealed that there was no sudden cardiac death in any of her relatives. On physical examination, rales were heard in the bilateral lower zones of the lungs and two positive pretibial edemas were observed. In laboratory results, troponin value was 0.4 ug/L (normal range 0- 0.16 ug/L), C-reactive protein (CRP) value was 16 mg/dL (normal range 0- 5 mg/dL), D-Dimer value was 1.5 mg/L (normal range 0- 0,55 mg/L ), creatinine value was 0.7 mg/dl (normal range 0.5-0.9 mg/dl). Electrocardiography (ECG) showed atrial fibrillation rhythm with rapid ventricular response and ST depressions in leads V2-5.

(Figure 1) Echocardiography (ECHO) showed 40% ejection fraction, global hypokinesia, and septal hypertrophy. The septum thickness was 17 mm and the posterior wall was 18 mm. (Figure 2) In the patient's medical history, when he applied to an external center due to shortness of breath 2 years ago, on the cardiology consultation at that time, the ejection fraction was normal in ECHO and she was not taking any medication. Pulmonary computed tomography (CT) angiography was performed with the prediagnosis of pulmonary embolism in the patient who had D-dimer elevation and newly diagnosed atrial fibrillation. No image

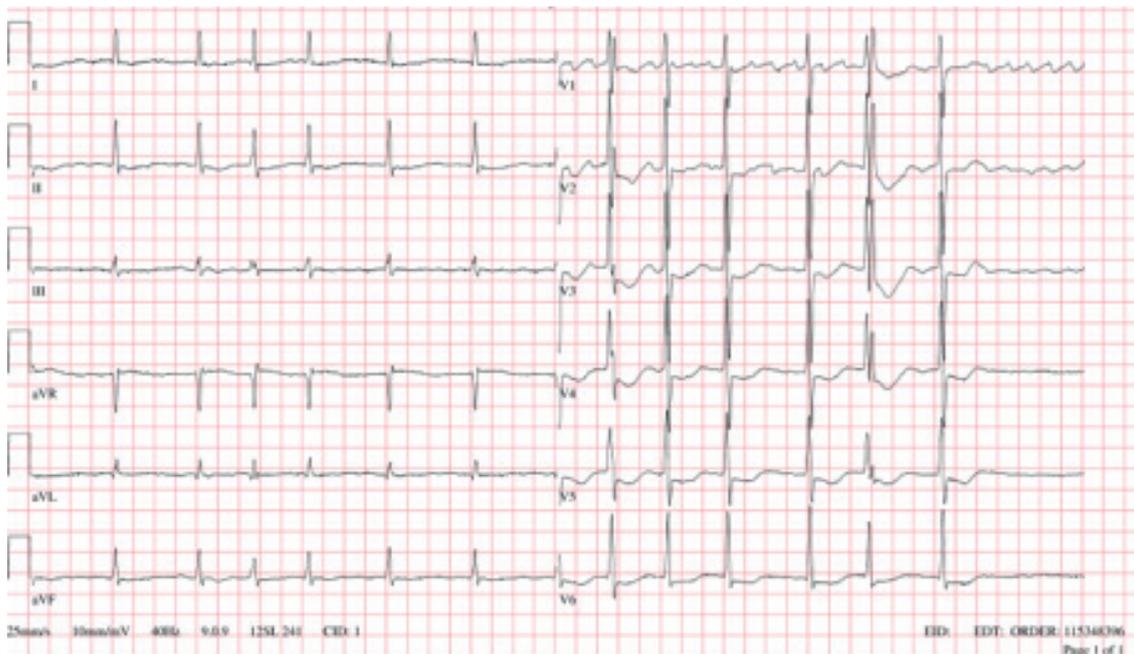


Figure 1. ECG atrial fibrillation.

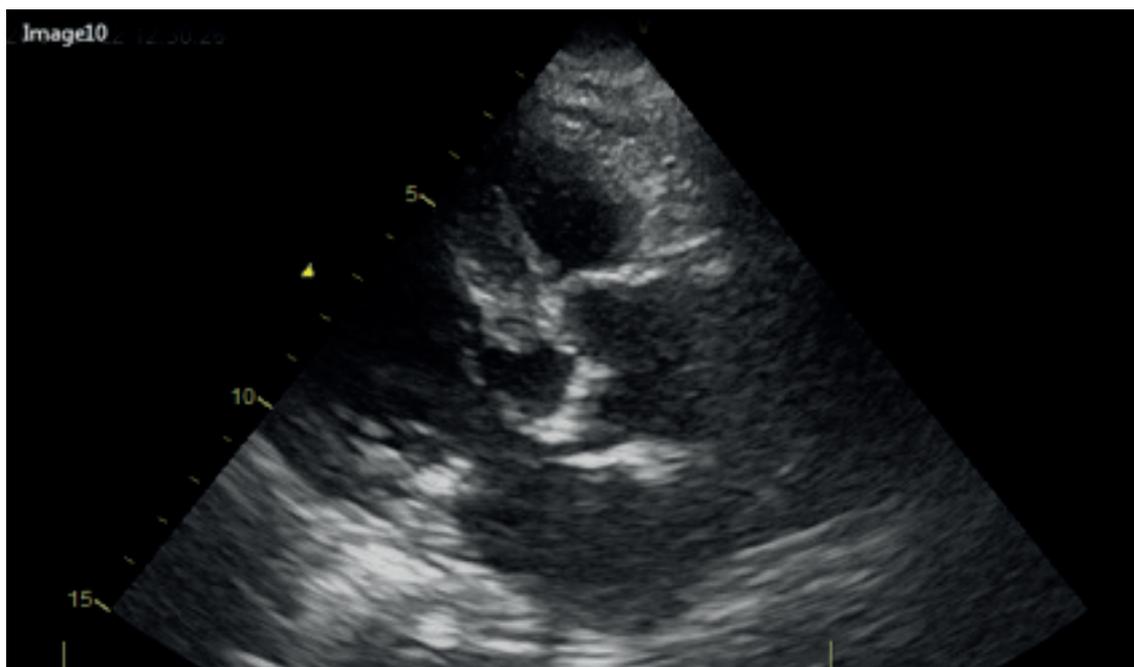


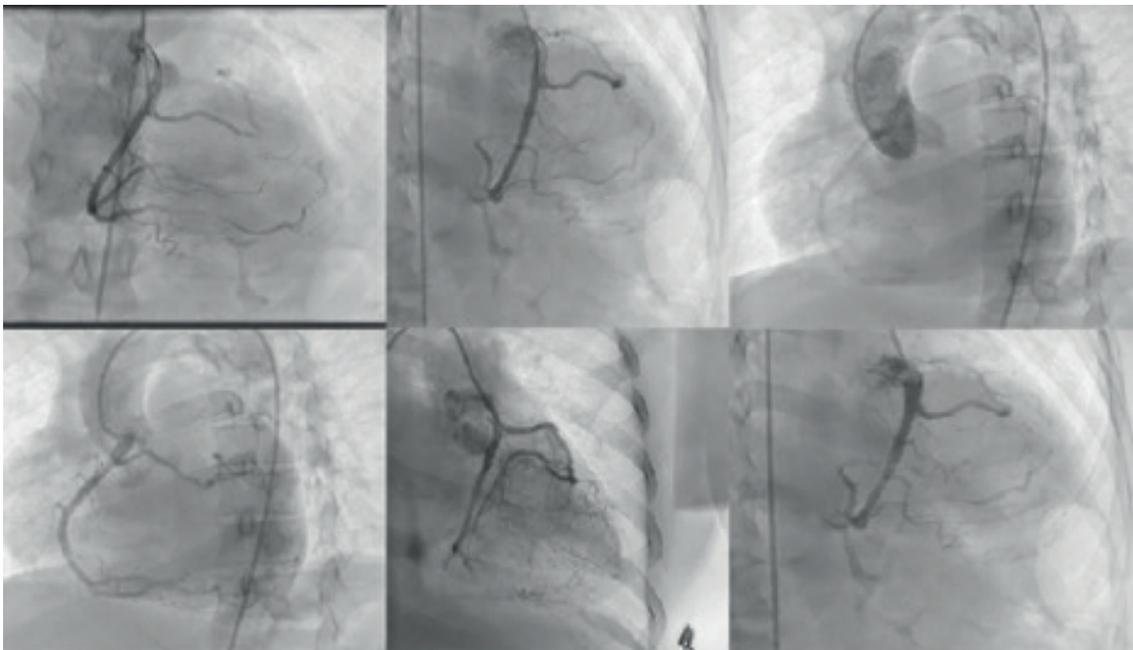
Figure 2. Transthoracic echocardiography image,septal hypertrophy.

compatible with embolism was detected in CT angiography. First of all, rate control was achieved and attempted to treat symptoms of decompensated heart failure. Upon detection of ST depressions in ECG and wall motion defect in ECHO, the patient was taken to coronary angiography after the patient's loading regressed. Coronary angiography of the patient revealed left main coronary artery agenesis, RCA, and CX originating from the same ostium and feeding the LAD area together, and a muscular bridge causing 80% stenosis in the distal RCA. (Figure 3) Afterward, cardioversion was planned to provide rhythm control to the patient. In the transesophageal echocardiography (TEE) performed before, apical hypertrophy and 0.8x2.0 cm thrombus in the left atrium were seen.(Figure 4) Cardioversion was delayed due to the detection of a thrombus in the left atrium. We started rivaroxaban 20 mg and arranged the heart failure treatment.

We discharged the patient to be re-evaluated with TEE and cardioversion, after using anticoagulant therapy for six months.

## Discussion

Although coronary anomalies are usually innocent, they have been shown to cause sudden cardiac deaths, although rarely. Many anomalies are found incidentally in coronary angiography and autopsies (12). The majority of these anomalies are incidentally detected benign (81%) anomalies that do not induce a major threat to myocardial perfusion (13,14). The spectrum of symptoms thought to be caused by coronary anomalies includes angina, syncope, congestive heart failure, and sudden death. Coronary artery anomalies are the second cardiovascular reason of sudden death in



**Figure 3.** Coronary angiography image (LMCA agenesis Absence of LAD , RCA and CX originate from the same ostium)



**Figure 4.** TEE image of left atrial appendix thrombus (red arrows) and left atrium SEC (blue arrows)

young people after hypertrophic cardiomyopathy.(13) Therefore, the combination of hypertrophic cardiomyopathy and coronary anomaly has great importance in terms of sudden cardiac death. These patients require close clinical follow-up and they may need implantable cardioverter-defibrillators (ICD). When we look at the literature, the scarcity of anomalies and the absence of LAD in our patient makes our case special. At the same time, considering that the detection of these cases are mostly coincidental, the fact that our case was diagnosed with atrial fibrillation with rapid ventricular response has made our case more interesting. We hope that this will put our case at the one of the top of the list of coronary anomalies in the literature and we hope to discover different coronary anomalies with new presentations.

**Informed consent:** *Informed consent was obtained from the patient for the publication of the case report and the accompanying images.*

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