Multivessel coronary-cameral fistulas associated with ventricular fibrillation: an unusual case

Ventrikül fibrilasyonuyla ilişkili çok sayıda korono-kameral fistül: Nadir bir olgu

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**Summary**—Coronary-cameral fistula (CCF) is a rare connection between a coronary artery and a chamber of the heart. It most often derives from the right coronary artery and drains into the right ventricle. CCF originating from all 3 major coronary vessels and draining into the left heart is an extremely rare coronary artery malformation. A 47-year-old man who was admitted to the clinic with angina pectoris and positive cardiac markers suddenly developed ventricular fibrillation (VF) while being prepared for coronary catheterization. He was successfully defibrillated and sinus rhythm was restored. The coronary angiography revealed normal epicardial coronary arteries with multiple diffuse fistulas originating from both the right and left coronary artery systems, terminating in the left ventricle. This is a very rare case of multiple CCFs originating from the epicardial coronary arteries and associated with myocardial ischemia and VF.

**CASE REPORT**

A 47-year-old man who had experienced new-onset angina pectoris for 2 days without any cardiac risk factors other than smoking and a family history of coronary heart disease was admitted to the cardiology clinic. There was no remarkable finding in the physical examination. His baseline 12-lead electrocardiogram (ECG) revealed a sinus rhythm without any ST-T wave abnormalities (Fig. 1). A transthoracic echocardiogram examination was normal in terms of with sudden cardiac death or survival of sudden cardiac arrest related to CFs have also been reported.[3] CCF originating from all 3 major coronary arteries that terminate in the left ventricle is an extremely rare phenomenon.[4]
ejection fraction, wall motions, valvular functions, and left/right heart chamber sizes. The patient’s serum troponin level was mildly elevated (high-sensitive troponin T: 75 pg/mL, upper reference level: 14–100 mg/mL) with a creatine kinase-MB level in the normal range. Coronary angiography was performed as the patient continued to experience chest pain despite medical therapy. The patient lost consciousness upon admission to the catheterization laboratory, with a VF rhythm observed on the monitor. He was promptly defibrillated with a single shock of 300 J, and sinus rhythm was restored. Unfortunately, the ECG demonstrating VF was not obtained. Coronary angiography was immediately performed to rule out significant coronary artery disease. The epicardial coronary arteries were normal; however, the angiography revealed widespread drainage of the contrast agent to the left ventricular cavity through many, small, diffuse fistulas, resulting in left ventricular contrast opacification (Fig. 2a, b and Video 1, 2*). A left ventriculography test was performed, which indicated normal left ventricular size and function.

No intracardiac shunt-related fistulas were detected and the coronary sinus size was normal. A diagnosis of CCF from all 3 coronary arteries was made. No obvious reason for the VF and chest pain, such as structural heart disease, myocarditis, electrolyte disturbance, congenital conduction system abnormality, drug abuse, or pulmonary embolism was found during the patient’s hospital follow-up.

![Baseline electrocardiogram without any ST-T changes and with normal PR/QRS/QTc intervals.](image1)

![Coronary-cameral fistulas and ventricular fibrillation](image2)

**Figure 1.** Baseline electrocardiogram without any ST-T changes and with normal PR/QRS/QTc intervals.

**Figure 2.** (A) Coronary-cameral fistulas can be seen between the distal segment right coronary artery and left ventricle. (B) Coronary-cameral fistulas are revealed between the distal segment of the left anterior descending artery, the circumflex artery, and the left ventricle.
Coronary artery fistula is primarily a congenital connection between a coronary artery and any of the great vessels (coronary-arteriovenous fistula) or one of the heart chambers (coronary-cameral fistula). CCF is a rare cardiac anomaly reported in approximately 0.08% to 0.3% of unselected patients who underwent diagnostic coronary catheterization.\(^5\) This type of fistula commonly originates from the right coronary artery and drains into the right-sided chamber of the heart. A connection between the left coronary arteries and the left ventricle is visualized in only 10% of CCF cases, and CCF from all 3 major coronary arteries terminating in the left ventricle is an extremely rare type of CF.\(^5\) Patients with CF are commonly asymptomatic and CF is usually detected incidentally. Patients with multiple CCF, however, are more symptomatic.\(^3,6\) The presence of symptoms is related to the size and direction of the fistula. These fistulas may cause angina through coronary steal phenomenon and diastolic overload. Exertional dyspnea, congestive heart failure, syncope, endocarditis, myocardial infarction, cardiac arrhythmias, and sudden cardiac death have been reported in this population.\(^3,7\) A study of 152 CF patients reported 1 sudden cardiac arrest case with VF; however, the type of fistula is not stated.\(^3\) Yuksel et al.\(^8\) reported a case of 3-vessel CCF with new onset atrial fibrillation, and the case of a 24-year-old women who survived sudden cardiac arrest due to myocardial ischemia with subsequent VF caused by thrombosis of a coronary artery fistula deriving from the left main coronary artery has also been reported.\(^9\) It was suspected to be the result of a post-traumatic hematoma causing total occlusion of the fistula and then initiating retrograde thrombosis of the coronary artery and resulting in myocardial infarction. To date, to the best of our knowledge, there is no reported case of 3-vessel corona-cameral fistulas associated with VF. It is suspected that VF occurred in this case due to myocardial ischemia caused by coronary flow steal through multiple micro-fistulas.

Several conditions and diseases may be responsible for VF in a patient with coronary arteries that appear normal on an angiogram. Coronary artery spasm (Prinzmetal’s variant angina) may play an important role in acute coronary syndromes and life-threatening arrhythmias due to vasoconstriction of the coronary arteries and myocardial ischemia.\(^{10}\) As there were no ST–T changes on baseline/follow-up ECGs and no vessel diameter reduction after the administration of intracoronary nitroglycerin, Prinzmetal’s variant angina was ruled out in our case. A detailed evaluation revealed that the levels of electrolytes (including potassium and magnesium), D-dimer, and oxygen saturation on admission were within the normal range and there was no right heart dilation; thus, acute pulmonary embolism and electrolyte disturbances did not account for the VF. Ventriculography and repeated echocardiography revealed normal size and function of the heart chambers and normal wall thickness without valvular disease; cardiomyopathy and severe valvular disease were also ruled out. The patient stated that there was no known family history of coronary artery disease or sudden cardiac death. In addition, the PR and QTc intervals were within the normal range and there were no delta or epsilon waves or indication of bundle block observed on the baseline ECG, which excluded Wolff–Parkinson–White syndrome, right ventricular dysplasia, Brugada syndrome, and long/short QT syndromes.

Patients with multiple and diffuse coronary artery fistulas may have symptoms due to compromised myocardial blood flow resulting in myocardial ischemia. In addition to clinical symptoms, such as angina, exertional dyspnea, and syncope, rarely, life-threatening cardiac arrhythmias, like VF, may occur in patients with multiple CCFs. A potential CF abnormality should be kept in mind for patients who present with angina pectoris and who develop ventricular fibrillation, after the more common and well-known causes of life-threatening arrhythmia have been excluded.

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

*Supplementary video file associated with this article can be found in the online version of the journal.*

REFERENCES


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Anahtar sözcükler: Koroner anjiyografi; fistül; ventriküler fibrilasyon.