A very rare combination of four coronary artery anomalies in a patient with acute inferior myocardial infarction

Akut inferiyor miyokart enfarktüslü bir hastada nadir görülen dört koroner arter anomalisinin birlikteliği

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Summary – We present a combination of four rarely seen coronary anomalies: double right coronary artery originating from the right coronary sinus (RCS) and left main coronary artery, respectively, and separate origination of the left anterior descending (LAD) artery, circumflex artery (Cx), and septal perforator artery from the RCS. These anomalies were encountered in a 46-year-old male patient who had a previous diagnosis of spina bifida occulta and renal pelvis and presented with the complaint of chest pain of two-hour onset. He had no conventional coronary risk factors and no history of chest pain or syncope. Electrocardiography showed ST-segment elevation and cardiac enzyme levels were elevated. Coronary angiography was performed with the diagnosis of acute inferior myocardial infarction, which showed a severe stenosis in the mid portion of the LAD and total occlusion in the proximal part of the Cx. Balloon dilatation and stent implantation were performed for the Cx lesion and TIMI 3 flow was achieved. One month after the procedure, percutaneous coronary intervention was repeated for the LAD lesion and patency was achieved with balloon dilatation and stenting. Since visualization of all the coronary anomalies mentioned above posed some difficulties during coronary angiography, cardiac computed tomography angiography was also used to reveal the ostia and the courses of coronary arteries. This combination of four rare coronary anomalies has not been reported before.

Özet – Bu yazıda, nadir görülen dört koroner anomalinin bir arada görüldüğü bir olgu sunuldu. Bu anomaliler şunlardı: Sağ koroner sinüsten ve sol ana koroner arterden köken alan çift sağ koroner arter ve yine sağ koroner sinüsten ayrı çıkışları olan sol ön inen arter, sirkumfleks arter ve septal perforatör arter. Anomaliler, daha önceden gizli spina bifida ve renal pelvis tanıları olan ve iki saatlik göğüs ağrısıyla başvuran 46 yaşında erkek hastada görüldü. Hastada bilinen koroner risk faktörleri ve göğüs ağrısı ve bayılma öyküsü yoktu. Elektrokardiyografide ST yükselmesi ve kardiyak enzim düzeylerinde artış görülmesi üzerine, akut inferiyor miyokart enfarktüsü tanısıyla hastaya koroner anjiyografi yapıldı ve sol ön inen arterin orta bölümünde ciddi darlık ile sirkumfleks arterin proksimal bölümünde tam tıkanma saptandı. Sirkumfleks arterdeki lezyona balonla genişletme ve stent yerleştirme uygulandı ve TIMI 3 akım elde edildi. Bu işlemden bir ay sonra, perkütan koroner girişim sol ön inen arterdeki lezyon için tekrarlandı ve balonla genişletme ve stent yerleştirme ile bu arterde de açıklık sağlandı. Koroner anjiyografi sırasında yukarıda belirtilen koroner anomalilerin görüntülenmesinde güçlükler yaşandığından, koroner arterlerin ağızlarının ve seyirlerinin daha iyi anlaşılabilmesi için hasta kardiyak bilgisayarlı tomografi anjiyografi ile de değerlendirildi. Bu dört nadir koroner anomalinin bir arada bulunması daha önce bildirilmemiştir.

The incidence of coronary artery anomalies is 1.3% in angiography series.[1] Although most CAAs are benign, some may cause arrhythmias, syncope, heart failure, or sudden death, even in the absence of atherosclerosis.[2] Furthermore, CCAs are the second most common cause of sudden death in young athletes.[3] Transthoracic and transesophageal echocardiography, contrast-enhanced electron-beam tomography, magnetic resonance imaging, cardiac computed tomography angiography, and conventional coronary angiography may reveal anomalous coronary arteries and their courses.[2,4]

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We present a combination of four rare CAAs in a patient suffering from acute inferior myocardial infarction. To the best of our knowledge, this complex coronary anomaly has not been reported before.

CASE REPORT

A 46-year-old male patient was admitted to our hospital after two hours of chest pain. Rest electrocardiography showed ST-segment elevation in D2, D3, and aVF, and ST depression in the precordial leads. The patient was diagnosed with spina bifida occulta and renal pelvis five years before. He had no conventional coronary risk factors and the rest of the examination findings were normal. Biochemical analysis was normal except for elevated cardiac enzyme levels. The patient was taken to the catheterization laboratory for primary percutaneous coronary intervention with the diagnosis of acute inferior myocardial infarction. Coronary angiography was performed using the Judkins technique. The right coronary artery was visualized in the normal position by the right Judkins catheter. Some retrograde collateral flow arising from the distal part of the RCA was observed. The engagement of the ostium of the left main coronary artery was unsuccessful with the left Judkins catheter. We proceeded with aortography to assess the LM and its course, which revealed that the LM was arising from the right coronary sinus (Fig. 1a). Engagement of the LM was successful with the left Amplatz 2 catheter. The LM was long and branching out of the RCA and left anterior descending artery. A severe stenosis in the mid portion of the LAD was noted (Fig. 2a). While trying to engage the LAD, a septal perforator artery that arose from the RCS and was free of atherosclerosis was accidentally cannulated. Because the left circumflex (Cx) artery was still not visualized, the angiography runs were carefully reviewed, and a tiny ostium was noticed just below the ostium of the RCA. The Amplatz 2 catheter was placed inferior to the right coronary ostium, and finally, cannulation of the Cx, which was totally obstructed in the proximal part, was successfully performed. Just after the cannulation of the Cx, severe bradycardia developed, and a temporal pacemaker was placed through the right femoral vein. Balloon angioplasty was performed with a 2.0 x 20-mm balloon at 10 atm and was followed by implantation of a 3.0 x 20-mm Liberte stent at 16 atm. Vessel patency with good TIMI 3 flow was achieved (Fig. 3).

Abbreviations:

CAA *Coronary artery anomaly*
CTA *Computed tomography angiography*
LAD *Left anterior descending*
LM *Left main*
PCI *Percutaneous coronary intervention*
RCA *Right coronary artery*
RCS *Right coronary sinus*
SPA *Septal perforator artery*

During hospitalization, cardiac CTA was performed to reveal the ostia and the courses of the coronary arteries. This scan showed that the LM, Cx, SPA, and RCA were originating separately from the RCS (Fig. 1b). Another smaller RCA was coming from the

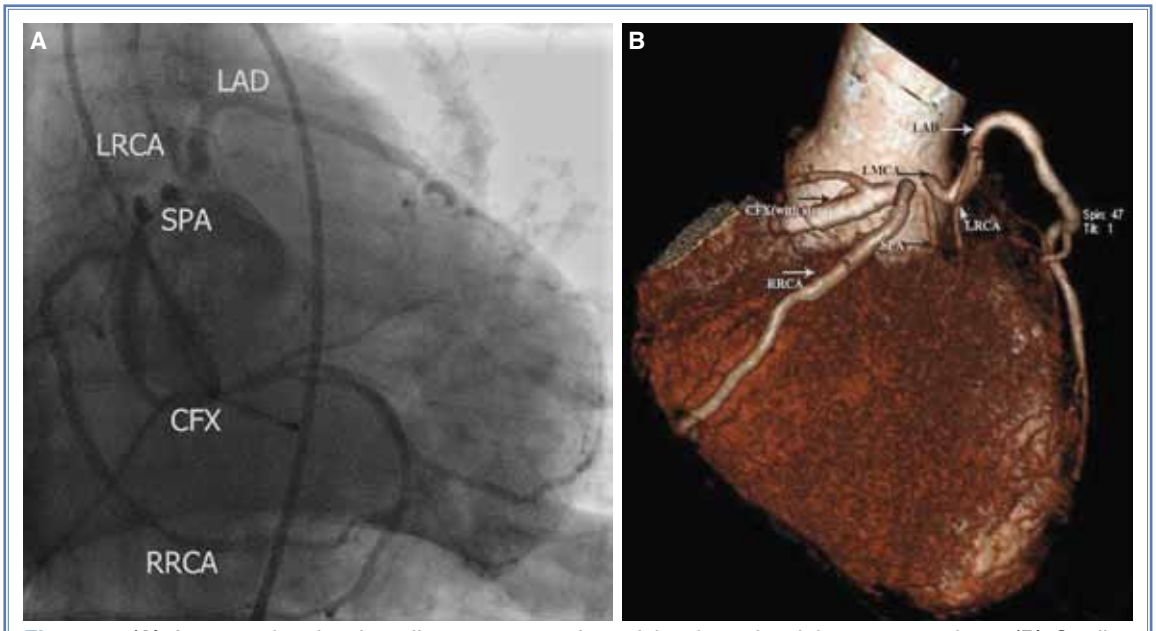

Figure 1. **(A)** Aortography showing all coronary arteries arising from the right coronary sinus. **(B)** Cardiac computed tomography showing origins of all coronary arteries and their courses.

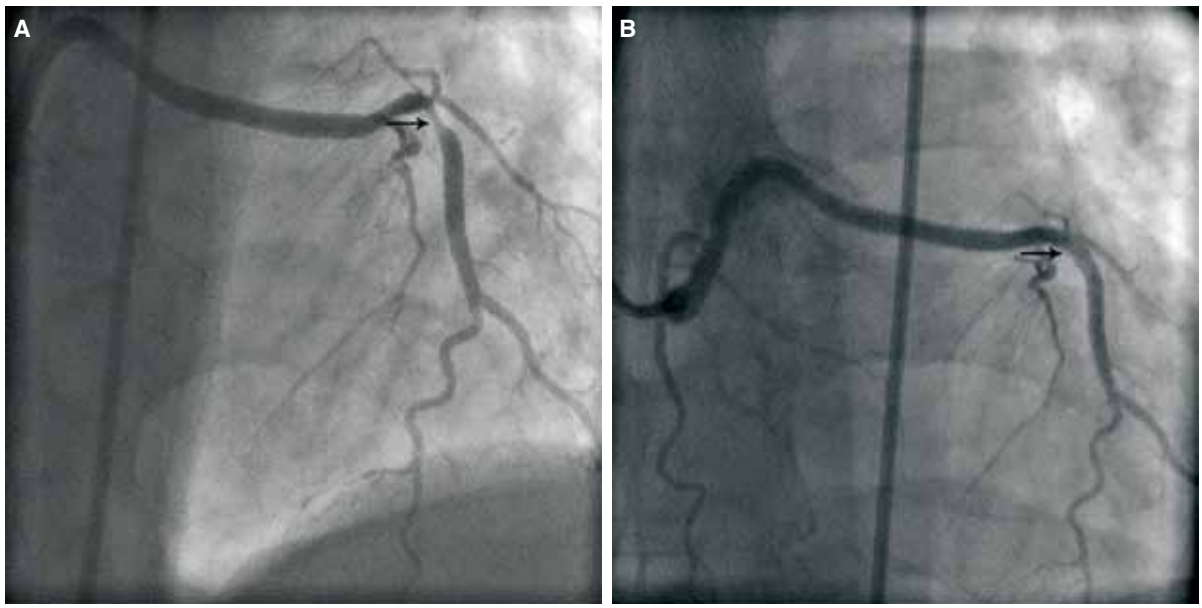

Figure 2. **(A)** Severe stenosis in the mid portion of the left anterior descending artery and **(B)** its patency achieved with balloon angioplasty and stenting one month after the index procedure.

proximal part of the LM, giving off small right ventricular branches.

The patient did not have chest pain or syncope prior to this hospitalization. After an extensive discussion with the patient and his family about the treatment options, we decided to perform PCI to the LAD one month after the index procedure. The clinical course of the patient was uneventful, and he was discharged on the fourth day in good condition. One month after the index procedure, the patient was hospitalized for PCI. Preprocedural transthoracic echocardiography showed no regional wall motion abnormality and normal left ventricular systolic function. The LM was cannulated with a Hockey Stick (HS2) catheter. After predilatation with a 3.0 x 15-mm balloon at 12 atm, a 3.5 x 22-mm Liberte stent was implanted at 16 atm. Postdilatation with the stent balloon at 20 atm was also performed. Vessel patency was achieved without any residual stenosis in the LAD (Fig. 2b). The patient was discharged two days after PCI in good condition, with a treatment regimen including clopidogrel, aspirin, metoprolol, ramipril, and statin.

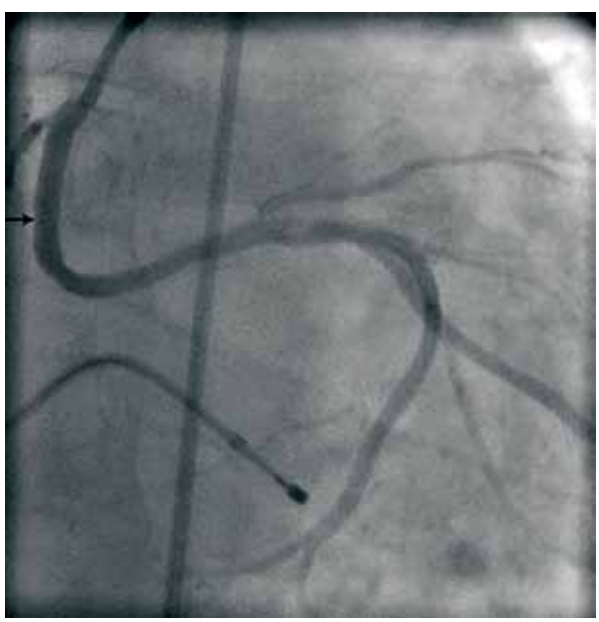

Figure 3. Circumflex artery after balloon angioplasty and stenting.

DISCUSSION

We presented a combination of four rare coronary anomalies: double RCAs originating from the RCS and LM, and separate origination of the LAD, Cx, and SPA from the RCS in a patient with acute inferior myocardial infarction. Although individual occurrences of the LAD arising from the RCS, dual RCAs, Cx originating from the RCS, and an SPA originating separately from the aorta have been reported,[5] the combination of all these CAAs in the same patient has not been described previously.

The importance of a septal coronary artery directly originating from the aorta has not been clearly shown.[5]

The Cx artery arising from the RCS or RCA is one of the most common coronary anomalies and is thought to be benign. However, it might be compressed or damaged during valvular surgery.[6] In the CASS study, it was shown that abnormally originating Cx arteries had more atherosclerosis than those arising normally.[7]

Most reported double RCA cases in the literature originate from the same ostium or RCS.[8] Double RCAs originating from both the LM and RCS are rarely reported. The RCA coming from the LM could be compressed while coursing between the aorta and pulmonary artery, and this may cause angina pectoris, acute myocardial infarction, syncope, and sudden death.[9] In our case, a second RCA arose from the proximal part of the LM and coursed anterior to the aorta. This course might prevent compression from the aorta or pulmonary artery. The occurrence of the LM or LAD arising from the RCS is a very rare congenital anomaly. Even in the absence of atherosclerosis, this anomaly may cause chest pain, syncope, arrhythmias, heart failure, or sudden death, especially in young people.[9]

Our patient had a five-year diagnosis of spina bifida occulta and renal pelvis. We could not find any case reports suggesting a relationship between coronary anomalies and neurological or genitourinary systems. It might be a coincidence rather than a syndrome. The asymptomatic nature of coronary anomalies might prevent diagnosis of these anomalies in patients with neurological or genitourinary system anomalies or vice versa.

Even though invasive coronary angiography is the standard imaging modality, CTA, magnetic resonance imaging, and echocardiography may reveal coronary anomalies and provide information about their courses. We used CTA to show the ostia and the courses of the coronary arteries. It also showed double RCAs and the RCS as the origin of the LM, Cx, and SPA.

Treatment options for coronary anomalies include medical, percutaneous, and surgical revascularization. Due to the low incidence of single coronary arteries, it is difficult to recommend a definitive treatment. Specifically, coronary arteries arising from the wrong sinus have been linked to adverse clinical events. Ischemia on myocardial perfusion scintigraphy or the presence of symptoms such as syncope or chest pain may help in the classification of patients with a coronary artery arising from the wrong sinus of Valsalva.[9]

In our case, primary PCI was performed for the abnormally originating Cx, and the patient was discharged with medical therapy. For the stenosis in the LAD, we preferred PCI as the treatment strategy after extensive discussion with the patient and his family. There are case reports showing that, despite some technical difficulties, especially related to engagement of the anomalous artery, PCI can be performed safely and effectively as a method of revascularization for atherosclerotic disease of a single coronary artery accompanied by an anomalously originating Cx.[10] We also had some difficulties in cannulating the LM and passing the balloon and stent. The buddy wire technique and a Hockey Stick 2 guiding catheter were used to overcome these difficulties.

Considering the possibility of more than two CAAs, interventional cardiologists should screen patients with an CAA very carefully for additional CAAs during diagnostic and interventional procedures. Increased awareness of these anomalies might also help us achieve vessel patency through PCI or other procedures, decrease procedure time, or prevent possible complications.

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Key words: Angioplasty, balloon, coronary; coronary angiography; coronary vessel anomalies/diagnosis; myocardial infarction; tomography, X-ray computed.

Anahtar sözcükler: Anjiyoplasti, balon, koroner; koroner anjiyografi; koroner damar anomalisi/tanı; miyokart enfarktüsü; bilgisayarlı tomografi.