# Extensive, Idiopathic, Recurrent, Spontaneous Coronary Artery Dissection Healed With Medical Treatment

Medikal Tedavi ile İyileşen Yaygın, İdiyopatik, Tekrarlayan Spontan Koroner Arter Diseksiyonu

Ertan AKBAY<sup>1</sup> © 0000-0002-9146-0621 Orhan ÖNALAN<sup>2</sup> © 0000-0001-9780-7051 Adem ADAR<sup>2</sup> © 0000-0002-2404-6447 Fahri ÇAKAN<sup>2</sup> © 0000-0002-5427-3480

<sup>1</sup>Department of Cardiology, Başkent University Hospital, Alanya Medical and Research Center, Antalya, Turkey

<sup>2</sup>Department of Cardiology, Karabük University Faculty of Medicine, Karabük, Turkey

### ABSTRACT

Spontaneous coronary artery dissection is a rare cause of sudden cardiac death and acute coronary syndrome, and mainly affects young female. Treatment of this condition depends on the clinical features, hemodynamic and angiographic findings of the patient, but due to the rarity of the disease, guideline support was insufficient until the last period. Depending on the condition of the patient and the coronary blood flow, medical observation, percutaneous coronary intervention or coronary artery bypass graft surgery are used in the treatment. We present a young female patient who underwent emergency coronary artery bypass graft surgery after spontaneous left main coronary artery dissection four years ago and was recently diagnosed with extensive spontaneous coronary artery dissection in her right coronary artery. After the diagnosis, the patient was given medical treatment and recovered completely.

Keywords: Spontaneous coronary artery dissection, acute coronary syndrome; coronary artery disease.

#### ÖΖ

Spontan koroner arter diseksiyonu, ani kardiyak ölüm ve akut koroner sendromun nadir bir nedenidir ve özellikle genç kadınları etkiler. Bu durumun tedavisi hastanın klinik özelliklerine, hemodinamik ve anjiyografik bulgularına bağlıdır, ancak hastalığın nadir olması nedeniyle son döneme kadar kılavuz desteği yetersizdi. Hastanın ve koroner kan akımının durumuna göre tedavide medikal gözlem, perkütan koroner girişim ya da koroner arter bypass greft cerrahisi kullanılmaktadır. Dört yıl önce spontan sol ana koroner arter diseksiyonu sonrası acil koroner arter bypass greft cerrahisi uygulanan ve yakın zamanda sağ koroner arterinde yaygın spontan koroner arter diseksiyonu tanısı alan genç bir kadın hastayı sunuyoruz. Teşhisin ardından hasta medikal tedavi ile takip edildi ve tamamen iyileşti.

Anahtar kelimeler: Spontan koroner arter diseksiyonu; akut koroner sendrom; koroner arter hastalığı.

#### **INTRODUCTION**

**Corresponding Author Sorumlu Yazar** Ertan AKBAY drertanakbay@gmail.com

Received / Geliş Tarihi : 11.07.2021 Accepted / Kabul Tarihi : 13.11.2021 Available Online / Çevrimiçi Yayın Tarihi : 02.12.2021 A spontaneous coronary artery dissection (SCAD) occurs when a tear is formed in the arterial wall, which results in the separation of intimal and medial layers and the formation of a pseudo lumen (1,2). Coronary blood flow in the real lumen deteriorates due to hematoma formation inside the pseudo-lumen which compresses the actual lumen (1). This may lead to myocardial ischemia, acute coronary syndrome, cardiogenic shock and even sudden cardiac death (3,4). The peripartum state, oral contraceptive use, connective tissue disease, and notably fibromuscular dysplasia are some of the etiological factors of SCAD (1). This case pertains to a young female who had idiopathic SCAD in the right artery and fully recovered with the non-invasive treatment.

## **CASE REPORT**

A thirty-six-year-old female was admitted to the emergency service with excruciating chest pain and numbness in the left arm. Her anamnesis revealed the history of coronary artery bypass graft surgery (CABG) four years ago. The patient had no cardiovascular risk factors; she had three children and seven years have passed since her last birth. Although no ischemic changes were observed in the ECG, the patient stated that her symptoms were similar to what she had experienced prior to her CABG (Figure 1). Her vital signs were stable (blood pressure: 120/170 mmHg, breathing rate: 12/min, heart rate: 60bpm and SpO2: 95%), laboratory tests revealed high levels of CK-MB (26 U/L, N: 0-25 U/L) and normal levels of troponin (0.000 ng/ml, N: 0.000-0.006 ng/ml). Other laboratory parameters were completely normal (creatinine: 0.54 mg/dl, hemoglobin: 12.1 g/dL, TSH: 0.65 uIU/ml). When we detailed the anamnesis, it was learned that the patient presented with chest pain similar to the emergency department four years ago and had a cardiac arrest in the follow-up. Since anterior wall myocardial infarction had been observed in her ECG after defibrillation, she had been transferred to the nearest interventional cardiology center. Coronary angiography had showed a dissection extending from the left main coronary artery (LMCA) to the left anterior descending artery (LAD), following which the patient was given an emergency CABG at that time (5). With her current clinical symptoms, the patient was admitted to coronary intensive care unit and given anti-platelet and anticoagulation treatment. The coronary angiography was performed twelve hours after hospitalization and revealed non-critical obstructions in the LAD and circumflex artery (CX) (Figure 2). The flow in the distal portion of the LAD was competitive between native LAD and left internal mammary artery (LIMA). LIMA was selectively imaged and seen as normal. Extensive dissection line was seen in the right coronary artery (RCA), starting from the right coronary sinus and extending to the posterior descending artery (Figure 3). The previous dissection extending from LMCA to LAD, which had been present in her previous angiography, was seen as recovered totally (Figure 2). Since the clinical condition of the patient was stable, she was given dual antiplatelet and heparin infusion. Thorax CT angiography was performed whether to detect the aortal extension of the dissection in the right coronary sinus. No dissection was observed in the ascending aorta at its distal end. As the condition of the patient remained stable, her medication

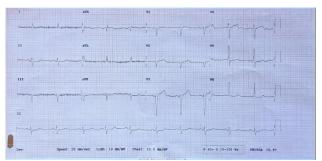


Figure 1. Electrocardiography at hospital admission

was changed to low molecular weight heparin and she was monitored at the service unit. The control coronary artery angiography was performed two weeks later and revealed that the right coronary sinus dissection had regressed, the hazy appearance of the RCA right ventricle branch had improved and the distal flow became better (Figure 4).

The patient was discharged after being electrically and hemodynamically stable at mobilization. At the 6<sup>th</sup> month follow-up, she did not report any complaints and there was no ischemic event in this period.

## DISCUSSION

Although SCAD is rare in the general population, it is more common in female, younger than 50 years of age, pregnancy and peripartum period (6,7). While the treatment was planned with case reports and retrospective study information before, it found a place in the "2020 European Society of Cardiology, Acute Coronary Syndromes in Patients Presenting without Persistent ST-Segment Elevation Guidelines" (8). Our case differs since she had a coronary bypass surgery due to SCAD and this time presented with spontaneous dissection of a different coronary artery and recovered without complications with medical treatment.

SCAD is classified according to the occlusion and severity of the occlusion in the coronary. SCAD types have been identified, and the visual appearance of multiple lumen in the coronary is determinant of type 1 dissection (6). Although coronary CT angiography (CCTA) is useful in diagnosis, its negative nature is not sufficient to exclude the diagnosis of SCAD (9). In addition, intravascular ultrasound (IVUS) and intracoronary imaging (optical coherence tomography, OCT) are used in its diagnosis (10).

Non-invasive treatment, percutaneous coronary intervention (PCI) and CABG are some clinical options for



**Figure 2.** The main coronary and proximal left anterior descending artery area with previous dissection and requiring coronary artery bypass surgery

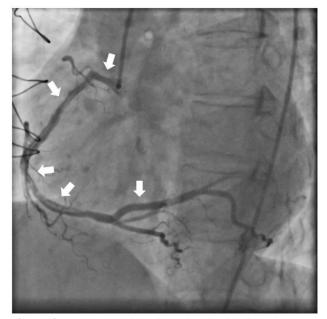


Figure 3. Long and wide dissection in the right coronary artery starting from the coronary sinus

SCAD (6,8). The hemodynamic status of the patients, whether the lesion is obstructive or not, and distal coronary flow status affect the treatment strategy (6,8). In addition to patient-specific treatment, recent studies suggest that non-invasive treatment is superior to PCI and CABG in terms of mortality and recurrence (11-13). In her previous admission, the patient had applied to the emergency department with similar symptoms and had experienced a cardiac arrest during her follow-up. Emergency CABG had been performed after LMCA dissection was observed in her coronary angiography (5). This time, patient's hemodynamics were stable and dissection in LMCA was observed healed by the time and a new diffuse dissection arose in RCA. Since the vital signs of the patient were stable, there was no chest pain and TIMI 3 flow was present in the distal proportion of RCA, we decided to continue medication with non-invasive treatment. In addition, since the RCA is dissected from the initiation to the distal, we decided not to perform PCI as it requires penetration through the pseudo-lumen, which may increase the hematoma inside the pseudo-lumen and disrupt the flow in the true lumen. If PCI is unavoidable, it should be performed together with IVUS or OCT (14). Increased complication risk of a second CABG led us to primarily consider non-invasive treatment.

When the clinic is unstable, as in the first application of our patient, we frequently apply to interventional treatment strategies (13,15). Cardiogenic shock is observed more frequently in SCAD patients presenting with ST elevation compared to atherosclerotic lesions (15). In addition, the dissected area is observed most frequently in the LMCA and proximal LAD (15). However, survival is better in SCAD patients presenting with ST elevation (13,15).

Coronary angiography performed on the 15<sup>th</sup> day of hospitalization showed that the dissection regressed. Our case showed that even a diffused dissection could be treated without any complications by non-invasive treatment, consistent with current reports (8,16). In addition,

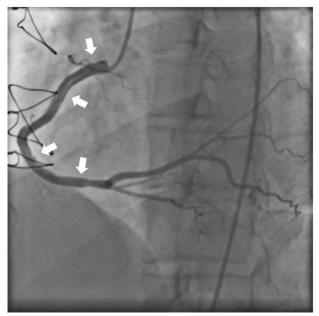


Figure 4. Control coronary angiography performed two weeks later

due to the absence of common predisposing factors like vasculitis, systemic inflammatory disease, pregnancy, substance use and stress, we identified this case as idiopathic SCAD. Studies similar to our patient showed that SCAD recurs with a frequency of more than 10% and can often occur in a different coronary artery (17,18). In addition, it suggests that recurrence may be associated with hypertension and coronary tortuosity (18). Although long-term treatment is not clearly known, medical treatment and follow-up seem to be a good alternative in patients with recurrent SCAD (13,18).

In conclusion, even in the presence of extensive dissection, if the clinical characteristics of SCAD patients are stable, the dissection can regress with a non-invasive treatment.

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

Conflict of Interest: None declared by the authors.

Financial Disclosure: None declared by the authors.

Acknowledgements: None declared by the authors.

**Author Contributions:** Idea/Concept: EA, OÖ; Design: EA, AA; Data Collection/Processing: EA, FÇ; Analysis/Interpretation: EA; Literature Review: EA; Drafting/Writing: EA; Critical Review: OÖ, AA.

## REFERENCES

- Gilhofer TS, Saw J. Spontaneous coronary artery dissection: update 2019. Curr Opin Cardiol. 2019;34(6):594-602.
- Di Fusco SA, Rossini R, Zilio F, Pollarolo L, di Uccio FS, Iorio A, et al. Spontaneous coronary artery dissection: Overview of pathophysiology. Trends

Cardiovasc Med. 2021;[Epub ahead of print]. doi: 10.1016/j.tcm.2021.01.002.

- 3. Nishiguchi T, Tanaka A, Ozaki Y, Taruya A, Fukuda S, Taguchi H, et al. Prevalence of spontaneous coronary artery dissection in patients with acute coronary syndrome. Eur Heart J Acute Cardiovasc Care. 2016;5(3):263-70.
- 4. Hill SF, Sheppard MN. Non-atherosclerotic coronary artery disease associated with sudden cardiac death. Heart. 2010;96(14):1119-25.
- 5. Cetin E, Ozyuksel A. Beating heart myocardial revascularisation of a sudden cardiac death survivor with spontaneous coronary artery dissection: pitfalls from diagnosis to surgery. BMJ Case Rep. 2014;2014:bcr2014207188.
- Kim ESH. Spontaneous coronary-artery dissection. N Engl J Med. 2020;383(24):2358-70.
- Iismaa SE, Hesselson S, McGrath-Cadell L, Muller DW, Fatkin D, Giannoulatou E, et al. Spontaneous coronary artery dissection and fibromuscular dysplasia: vasculopathies with a predilection for women. Heart Lung Circ. 2021;30(1):27-35.
- 8. Collet JP, Thiele H, Barbato E, Barthélémy O, Bauersachs J, Bhatt DL, et al. 2020 ESC Guidelines for the management of acute coronary syndromes in patients presenting without persistent ST-segment elevation. Eur Heart J. 2021;42(14):1289-367.
- 9. Eleid MF, Tweet MS, Young PM, Williamson E, Hayes SN, Gulati R. Spontaneous coronary artery dissection: challenges of coronary computed tomography angiography. Eur Heart J Acute Cardiovasc Care. 2018;7(7):609-13.
- 10. Takahashi T, Okayama H, Matsuda K, Yamamoto T, Hosokawa S, Kosaki T, et al. Optical coherence tomography-based diagnosis in a patient with STelevation myocardial infarction and no obstructive coronary arteries. Int J Cardiol. 2016;223:146-8.

- 11. Tweet MS, Eleid MF, Best PJ, Lennon RJ, Lerman A, Rihal CS, et al. Spontaneous coronary artery dissection: revascularization versus conservative therapy. Circulation. Circ Cardiovasc Interv. 2014;7(6):777-86.
- 12. Alfonso F, Paulo M, Lennie V, Dutary J, Bernardo E, Jiménez-Quevedo P, et al. Spontaneous coronary artery dissection: long-term follow-up of a large series of patients prospectively managed with a "conservative" therapeutic strategy. JACC Cardiovasc Interv. 2012;5(10):1062-70.
- 13. Gómez-Hospital JA, Ariza-Solé A. Spontaneous coronary artery dissection: a benign entity? Rev Esp Cardiol (Engl Ed). 2021;74(1):2-4.
- 14. Lempereur M, Fung A, Saw J. Stent mal-apposition with resorption of intramural hematoma with spontaneous coronary artery dissection. Cardiovasc Diagn Ther. 2015;5(4):323-9.
- 15. Lobo AS, Cantu SM, Sharkey SW, Grey EZ, Storey K, Witt D, et al. Revascularization in patients with spontaneous coronary artery dissection and STsegment elevation myocardial infarction. J Am Coll Cardiol. 2019;74(10):1290-300.
- 16. Shoja T, Basman C, Jain S, Mangla A, Lasic Z. Postpartum sudden cardiac death after spontaneous coronary artery dissection in a patient with fibromuscular dysplasia. Cardiol Res. 2017;8(6):327-30.
- 17. Saw J, Aymong E, Sedlak T, Buller CE, Starovoytov A, Ricci D, et al. Spontaneous coronary artery dissection: association with predisposing arteriopathies and precipitating stressors and cardiovascular outcomes. Circ Cardiovasc Interv. 2014;7(5):645-55.
- Kok SN, Tweet MS. Recurrent spontaneous coronary artery dissection. Expert Rev Cardiovasc Ther. 2021;19(3):201-10.