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***Case Report** / Olgu Sunumu*

Spontaneous idiopathic pulmonary artery dissection with ST segment elevation in Leads aVR and V1

aVR ve V1 derivasyonlarında ST segment elevasyonu ile birliktelik gösteren spontan idiyopatik pulmoner arter diseksiyonu

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ABSTRACT

Pulmonary artery dissection is a rare condition that often occurs on the basis of pulmonary arterial hypertension and causes complications such as cardiogenic shock and sudden death. Additionally, this condition can be idiopathic. A 59-year-old male patient with no previous history of disease presented to our clinic with chest pain and shortness of breath. Coronary arteries were normal on coronary angiography in the patient who had a positive troponin test result and ST segment elevation in leads V1, V2, V3 and aVR. Pulmonary embolism was suspected in the patient whose condition worsened. Pulmonary artery dissection was diagnosed via the contrast-enhanced computed tomography and sudden cardiac death occurred. In conclusion, pulmonary artery dissection may cause aVR segment elevation on electrocardiography.

Keywords: Angina pectoris, dissection, pulmonary artery.

ÖZ

Pulmoner arter diseksiyonu, genellikle pulmoner arteriyel hipertansiyon zemininde gelişen ve kardiyojenik şok ve ani ölüm gibi komplikasyonlara neden olan nadir bir durumdur. Bununla birlikte, bu durum idiyopatik olabilir. Daha önce herhangi bir hastalık öyküsü olmayan 59 yaşında erkek hasta göğüs ağrısı ve nefes darlığı ile kliniğimize başvurdu. Koroner anjiyografide koroner arterler normaldi ve hastanın troponin test sonucu pozitif olup, V1, V2, V3 ve aVR derivasyonlarında ST segment elevasyonu izlendi. Durumu ağırlaşan hastada pulmoner emboliden şüphelenildi. Pulmoner arter diseksiyonu kontrastlı bilgisayarlı tomografide tanılandı ve ani kardiyak ölüm gelişti. Sonuç olarak, pulmoner arter diseksiyonu elektrokardiyografide aVR segment elevasyonuna neden olabilir.

Anahtar sözcükler: Göğüs ağrısı, diseksiyon, pulmoner arter.

Pulmonary artery dissection (PAD) is a rare and fatal complication of pulmonary hypertension (PHT), or pulmonary artery aneurysm (PAA), which is usually associated with congenital heart disease.[1] Chest pain, cardiogenic shock, and eventually sudden death are typically developed with PAD.[2] In this article, we present a rare case of an isolated PAD without associated PHT or PAA.

CASE REPORT

A 59-year-old male patient was admitted to the emergency service with new-onset retrosternal chest pain and shortness of breath. He had no medical history of disease. His blood pressure was 140/80 mmHg, heart rate was 95/min, and oxygen saturation was 94%. The first troponin value was 0.02 ng/mL (cut-off level: 0.01 ng/mL), the second one was 0.06 ng/mL, and the

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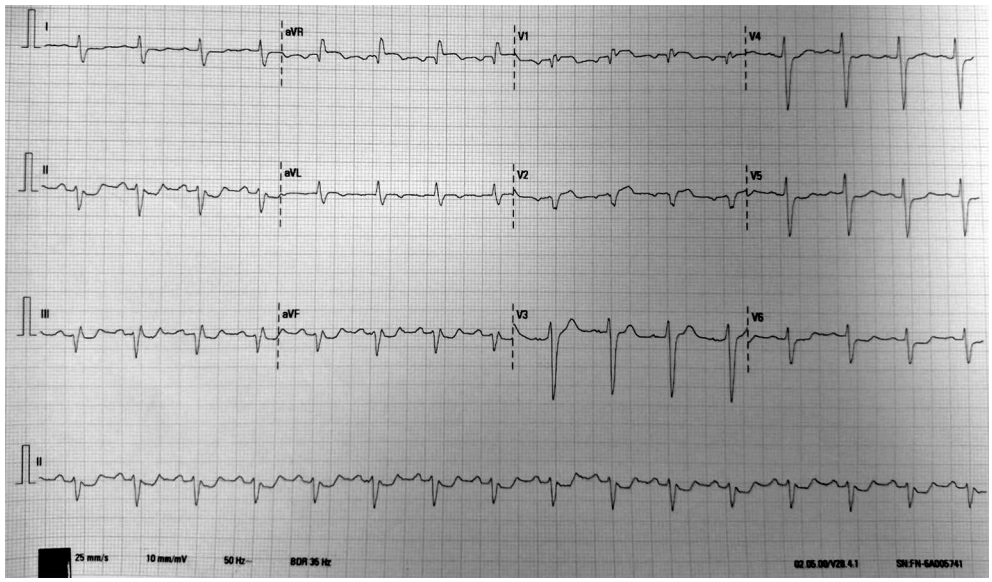

Figure 1. Electrocardiogram showing ST-segment elevation in Leads V1, V2, V3 and aVR.

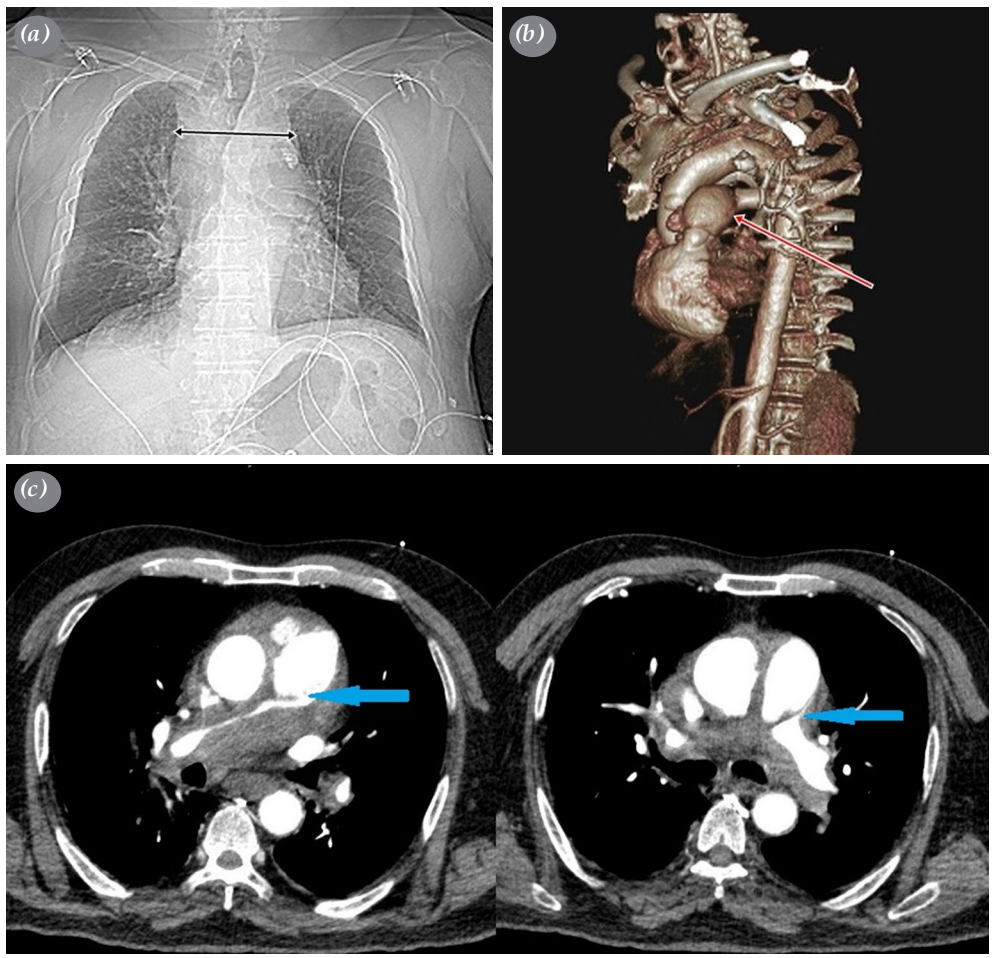

Figure 2. Computed tomography angiography. **(a)** Mediastinal widening, **(b)** three-dimensional image of pulmonary artery dissection, **(c, d)** dissection flap arising from pulmonary cusp level extending to both two main pulmonary arteries and mediastinal hematoma.

D-dimer value was within normal ranges. The elevation of consequent high sensitivity troponin-I levels and D-dimer value were evaluated using blood tests, and the elevation of V1-V2-V3 and aVR segments on electrocardiogram (ECG) was investigated. Coronary angiography was performed due to acute coronary syndrome diagnosis (Figure 1). Apart from 50% stenosis in the left anterior descending (LAD) artery, no other pathology was detected in the coronary arteries. Echocardiograms revealed ejection fraction as 55%, and no extra pathology was found.

The symptoms of the patient were not resolved and began to worsen. Blood pressure decreased and shortness of breath developed. Suspecting pulmonary embolism, emergent contrast-enhanced thoraco-abdominal computed tomography (CT) was performed, and PAD was diagnosed (Figure 2). On second echocardiogram, acute right ventricular insufficiency and pericardial effusion were detected. An emergent operation was planned; however, sudden cardiac death occurred during the preparation for surgical intervention.

DISCUSSION

Chest pain is one of the most common and complicated cause of emergency service admission all over the world. The most common causes of the chest pain include coronary artery disease, pneumonia, pleural effusions, pleurisy, pericarditis, chest wall trauma, aortic dissection, cardiac syndrome X, peptic ulcers, panic disorders and anxiety.[3] Due to these multiple causes, PAD cannot be diagnosed at first sight among all chest pain complaints. In addition, we did not consider PAD in our case, as no congenital heart disease existed in the history of the patient, and no suspicion of PHT was aroused based on the diagnostic tests.

Major changes on ECG initially suggested that the pathology was left main coronary artery (LMCA) occlusion. Basic findings on ECG that were considered as the indicators of left main coronary artery occlusion were ST depression in Leads I, II, aVL and V4-V6, ST elevation in aVR ≥1 mm and ST elevation in aVR ≥V1. Symptoms and ECG findings of the patient were similar to that of LMCA occlusion. This situation could also be a result of external compression of the enlarged pulmonary artery due to dissection on the LMCA. However, this possibility was ruled out depending on the absence of abnormal increase in the results of the repeated troponin tests, and additionally normal coronary angiography was excluded, as well.

On ECG, aVR derivation faced the basal segment of interventricular septum.[4] Diagnoses that can result in elevation in aVR derivation include acute pulmonary embolism, certain arrhythmias, and Takotsubo syndrome.[5] Possible mechanisms that cause ST segment elevation in acute pulmonary embolism were increased acute right ventricular failure, impairment of coronary blood flow, and increased myocardial oxygen consumption. In our case, ST segment elevation in Lead aVR could be a result of these mechanisms. In the presence of ST segment elevation on aVR derivation, pulmonary dissection must be kept in mind in the differential diagnosis. Among the causes that result in ST segment elevation on aVR derivation, PAD is not included in differential diagnosis in published reports.

Aortic dissection could have been considered in the differential diagnosis. However, due to the absence of any risk factors such as hypertension or connective tissue disease related to aortic dissection in the history of the patient, as well as the presence of troponin and ST segment elevation in the ECG leads, the diagnosis was directed to acute coronary syndrome. Therefore, coronary angiography was initially planned.

Right ventricular hypertrophy, dilated right atrium, dilated right ventricle, and elevated pulmonary artery pressure are important echocardiographic findings for early diagnosis of PAD.[6] Echocardiography (ECHO) was the first-line diagnostic feature for early diagnosis of rare cases such as PAA and PAD. In our case, ECHO was performed after coronary angiography. We concluded that two mechanisms played a major role as a cause of not evaluating the pulmonary dissection at first ECHO: the first one is that the dissection developed isolated from PAA and PHT and the second one is that the dissection flap was not widened at first time to cause right ventricular dysfunction. Due to the crescendo dyspnea accompanying chest pain, PAD was evaluated on CT and right ventricular dysfunction was evaluated on the second ECHO after the CT. Any finding in the right ventricle might have been overlooked in the patient who was evaluated with hand ECHO in the emergency service. Therefore, diagnosis of pulmonary embolism might not have been focused.

In contrast to aortic dissections, PAD progresses rapidly, and rupture usually occurs before developing a reentry zone. Subsequently, cardiogenic shock and sudden death occurs. From the first definition of the pathology suggested by Helmbrecht in 1842 to today, only a few of those that have been diagnosed with this pathology among 133 cases have survived.[1,3]

Since there are very limited cases similar to this case in the literature, there is no consensus on optimal treatment. Matsumoto et al.[7] reported medical follow up for two cases that were diagnosed with PAD. Mohammad et al.[8] followed an idiopathic PAD case that did not accept a surgical intervention with diuretic and vasodilator medications for one year. In all three cases, hemodynamic instability was not evaluated. No rupture was diagnosed in all three cases: a patient whom dissection of the distal segment of left pulmonary artery secondary to trauma was applied, a 97-year-old patient, and a patient that did not accept surgical intervention.

In particular, for the PAD cases that developed due to PHT and PAA and resulting in rupture and hemodynamic instability, early and urgent diagnosis and emergent surgical intervention after the diagnosis seem to be the only treatment option. In our case, we also planned to apply emergent surgical intervention; however, cardiogenic shock developed rapidly, and sudden cardiac death occurred.

In conclusion, early diagnosis is life-saving in the presence of pulmonary artery dissection. In particular for the patients admitted to emergency service with chest pain and dyspnea with the history of congenital heart disease and pulmonary hypertension with ST segment elevation of aVR derivation on electrocardiogram, pulmonary artery dissection is a complication that must be kept in mind for early diagnosis and prevention of possible early mortality and morbidity.

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