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Case Report

Left Main Coronary Dissection in a Patient with Takayasu's Arteritis

Takayasu Arteriti Olan Bir Hastada Sol Ana Koroner Diseksiyonu

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Takayasu's arteritis is a type of vasculitis which is characterized by chronic inflammation involving the aorta and its major proximal branches. The inflammatory process in the vessel wall due to intimal thickening and fibrosis may result in catastrophic events such as vascular stenosis and spontaneous dissection. However, spontaneous coronary dissection is a relatively rare outcome. The use of transthoracic echocardiography in the evaluation of thoracic aortic involvement has been shown to be advantageous since this method is relatively cheap and non-invasive. In this article, we report a 48-year-old female case who was previously diagnosed with Takayasu's arteritis with a spontaneous left main coronary artery dissection which was detected by coronary angiography following the sudden onset of severe chest pain.

Key words: Arteritis; coronary; dissection; Takayasu's.

Takayasu arteriti aort ve ana dalların proksimalini tutan, kronik inflamasyon ile karakterize bir vaskülit tipidir. İntimal kalınlaşma ve fibrozise bağlı olarak, damar duvarındaki inflamatuvar süreç, vasküler darlık ve spontan diseksiyon gibi katastrofik durumlara yol açabilir. Ancak, spontan koroner diseksiyon daha nadir rastlanan bir sonuçtur. Torasik aort tutulumunun değerlendirilmesinde transtorasik ekokardiyografi kullanımının, ucuz ve noninvaziv bir yöntem olması nedeniyle avantajlı olduğu gösterilmiştir. Bu yazıda daha önce Takayasu arteriti tanısı konmuş ve ani başlayan şiddetli göğüs ağrısı nedeniyle yapılan koroner anjiyografisinde spontan sol ana koroner arter diseksiyonu saptanan 48 yaşında bir kadın olgu sunuldu.

Anahtar sözcükler: Arterit; koroner; diseksiyon; Takayasu.

Takayasu's arteritis, which was first diagnosed by a Japanese ophthalmologist in 1908 based on ocular findings,[1] is characterized by organ ischemia as a result of chronic inflammation of the aorta and its main branches. Takayasu's arteritis predominantly occurs before the age of 40 and is more common in females. Coronary involvement, although rare, is often ostial and causes ischemia due to inflammatory stenosis. In addition to coronary stenosis, late-term vital complications, such as vascular dissection, should be monitored carefully. Furthermore, transthoracic echocardiography should be considered as an alternative non-invasive method to be used in the follow-up and evaluation of the thoracic aorta.

CASE REPORT

A 48-year-old female patient with no known cardiovascular risk factors was referred to our emergency department due to the sudden onset of severe, constricting chest pain that had been occurring for the previous four hours. On physical examination, blood pressure measurements from the right and left arms were 155/90 and 130/80 mmHg, respectively, and an apical 2/6 systolic heart murmur was heard.

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Electrocardiography (ECG) showed a 1 mm ST-segment depression in the precordial derivation and a 1.5 mm ST elevation in augmented vector right (aVR) (Figure 1). Laboratory tests revealed a creatinine level of 1.8 mg/dL, a urea level of 80 mg/dL, and slightly increased troponin/creatine kinase-MB fraction (CK-MB) levels. Based on the patient's medical history, she had been diagnosed with Takayasu's arteritis 12 years earlier due to renal artery stenosis, subclavian artery stenosis, and intermittent claudication. In addition, a pulmonary embolism had been detected four years previously. Transthoracic echocardiography showed normal left ventricular systolic function, mild aortic regurgitation, and moderate tricuspid regurgitation. The systolic pulmonary artery pressure was 35 mmHg. The ascending aorta was measured as 4.0 cm, and no dissection flap was detected. An echocardiographic examination revealed that the proximal aorta was thickened and hyperechogenic, and calcific plaques were commonly observed on the inner surface of the lumen (Figure 2). Using suprasternal imaging, it was determined that the arcus aorta was 4.3 cm in width, and moderate proximal brachiocephalic, common carotid, and subclavian artery stenoses were detected. Thickening of the arcus aorta wall was observed in addition to an increased echogenic area and a rough inner lumen (Figure 3). Due to ongoing severe chest pain, the patient was urgently transferred to the catheter laboratory. Spontaneous dissection causing severe stenosis in the proximal left main coronary artery (LMCA) was detected by coronary angiography (Figure 4), but all other coronary vessels were normal. In addition, there was no significant narrowing observed on the carotid Doppler ultrasound. The patient's hemodynamic values were normal, and an emergency operation was planned. Ascending aorta, upper, and lower vena cava cannulations were performed after a median sternotomy and then the patient entered into the pump. The heart was stopped by cold potassium while the patient was under general (rectal temperature 28 °C) and topical hypothermia. During an intraoperative evaluation of the thoracic aorta, it was noted that it was completely calcified and hardened. Aortocoronary anastomosis was performed using a saphenous venous graft because of the ostial involvement of the subclavian and left internal mammary arteries (LIMA). The postoperative course was uneventful, and the patient was discharged on postoperative day 10.

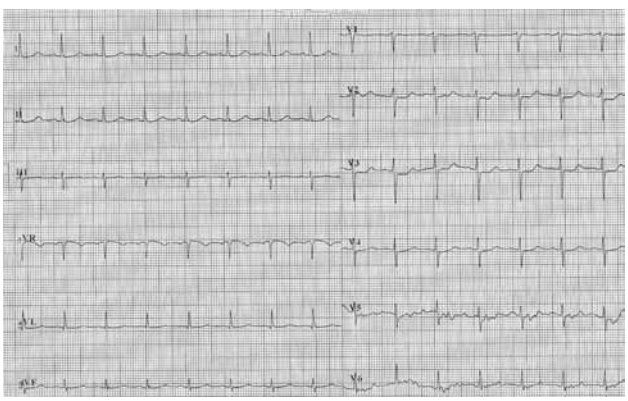

Figure 1. Electrocardiography showing precordial ST-segment depression and ST-segment elevation in lead aVR.

DISCUSSION

Takayasu's arteritis, also known as Martorell's syndrome, is an inflammatory disease that often affects the aorta and the proximal segments of the main aortic. It also may result in vascular occlusion due to intimal fibrosis and thickening. In addition to

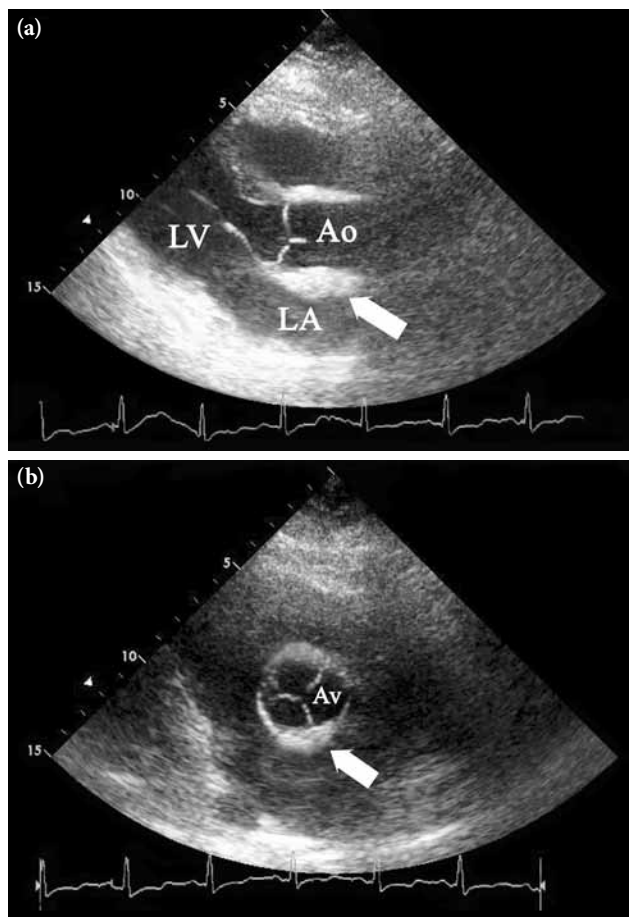

Figure 2. Transthoracic echocardiographic images indicating a thickened and hyperechogenic proximal aortic wall. **(a)** Parasternal long-axis view and **(b)** parasternal short-axis view showing increased thickness of the aortic root (shown by arrows). LV: Left ventricle; LA: Left atrium; Av: Aortic valve.

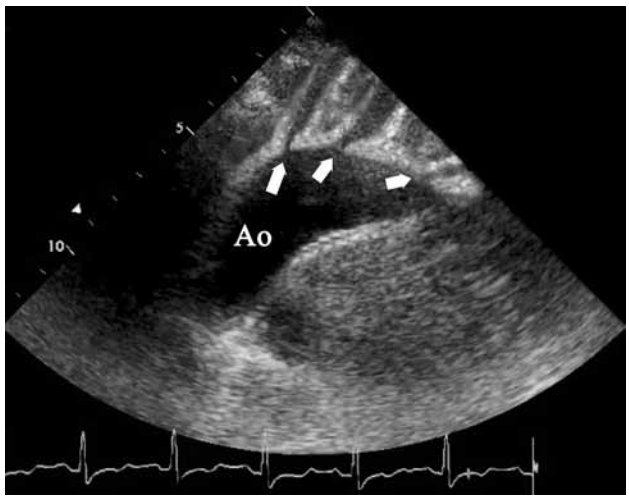

Figure 3. Suprasternal echocardiographic image of the arcus aorta. The arrows indicate the ostial involvement of the main branches. From left to right, the brachiocephalic, common carotid, and subclavian arteries can be viewed. The wall of the arcus aorta is thickened and has an increased echogenic area. Ao: Aorta.

ostial stenosis, aorta aneurysms may be encountered, depending on the existence panarteritis. This is especially true if there is destruction of the tunica media. The cause of Takayasu's arteritis is not yet known, but it is more common in young females. Symptoms vary depending on the involvement and severity of the functional impairment of the organs supplied by the stenotic vessels. During the early stage, rheumatic complaints such as fever, malaise, fatigue, general body pain, and weight loss may occur due to the systemic inflammatory condition. As a result of chronic vascular inflammation, intimal fibrosis may result in catastrophic outcomes, such as organ ischemia. Arm and leg pain, renovascular hypertension, and neurological symptoms may develop in patients during advanced stages of this disease. Pulmonary arterial hypertension and emboli are also often observed due to the involvement of the pulmonary artery. A diagnosis of Takayasu's arteritis is made clinically according to the presence of at least three of the American College of Rheumatology (ACR) 1990 criteria:[2] *(i)* disease onset prior to age 40, *(ii)* claudication of the extremities, *(iii)* reduction in the brachial pulse, *(iv)* more than a 10 mmHg pressure difference between the two arms, *(v)* detection of a murmur around the aorta, subclavian artery, or both, and *(vi)* abnormal arterial imaging. There are no specific blood tests for Takayasu's arteritis.

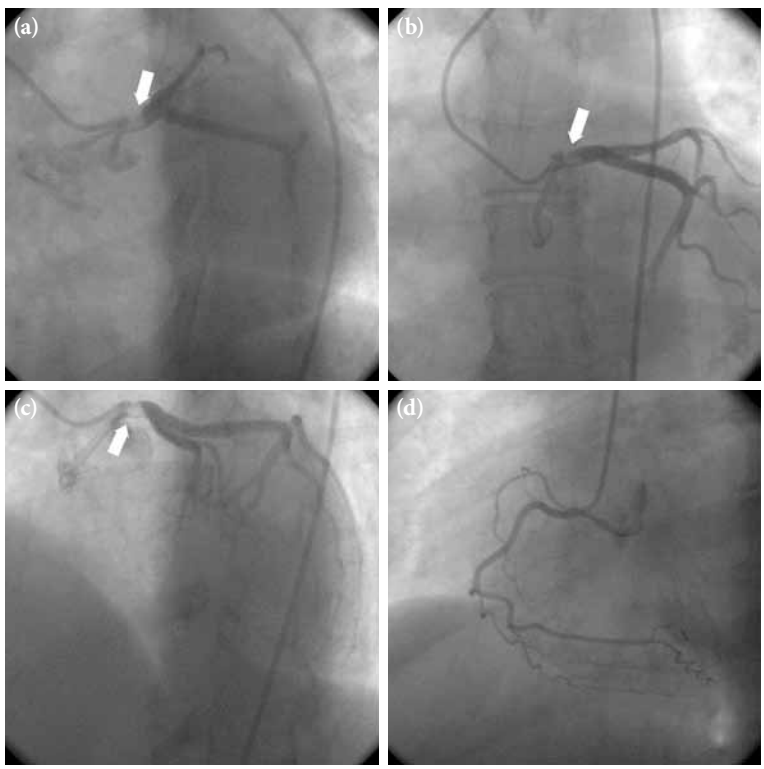

Figure 4. Coronary angiographic images. **(a)** Left anterior oblique caudal view of left main coronary artery. **(b)** Posterior anterior view of left main coronary artery. **(c)** Left anterior oblique cranial view of left main coronary artery. **(d)** Left anterior oblique view of right coronary artery.

Medical treatments include immunosuppressive therapy, such as steroids, cyclophosphamide, and methotrexate, but there is no current agreement on the best treatment option. Coronary artery involvement occurs in 5-15% of patients. Although Takayasu's arteritis often causes ostial stenosis, spontaneous LMCA dissection is an extremely rare complication.[3] In the case of sudden-onset chest pain in a patient with this disease, coronary artery dissection should be considered due to the need for early diagnosis and rapid treatment. However, dissection of the ascending aorta should be excluded. In our patient, transthoracic echocardiography (suprasternal view) did not show aortic dissection. Therefore, computed tomography (CT) and magnetic resonance imaging (MRI) were not performed because of poor renal function. An echocardiographic examination should be used during routine follow-up of all patients with Takayasu's arteritis to evaluate aortic involvement. Transthoracic echocardiography is non-invasive and inexpensive and has been proven to be an excellent method for patient diagnosis and follow-up. Intimal thickening and hyperechogenic appearance due to calcification of the aorta and its major branches are important and valuable observations. As in our patient, echocardiography may be useful as an alternative to MRI and CT for detecting ostial stenosis of the main branches. Percutaneous coronary intervention (PCI) for left main coronary stenosis has been described in the literature; however, invasive treatments should be decided upon according to the lesion location, number of stenotic vessels, and available technical equipment.[4] There are currently no clinical studies showing the superiority of percutaneous intervention over surgical therapy in patients with Takayasu's arteritis complicated by left main coronary dissection. Due to the risk of entering a false lumen, surgical management was planned. In our patient, an internal mammary artery (IMA) graft was not used because there were advanced fibrotic changes and calcification in the aorta and its major branches. A saphenous graft was preferred, as previous reports have shown lesser venous involvement with this procedure.[5] A careful preoperative echocardiographic examination with suprasternal views is suggested, which can provide the surgeon with adequate information regarding graft choice. When intimal thickening is intense and subclavian ostial stenosis is detected, saphenous graft selection should be given priority. No guidelines are currently available regarding percutaneous intervention to protect the LMCA following surgery for its dissection in patients with Takayasu's arteritis.

In conclusion, although Takayasu's arteritis is a rare disease, physicians should be mindful of serious complications such as coronary involvement and dissection. Transthoracic echocardiography with suprasternal views provides fast, reliable information and can be used as an alternative to CT and MRI. Future prospective echocardiographic studies examining the relationship between aorto-ostial involvement, aortic intimal thickness, and coronary involvement will be instrumental in providing information that can be used to detect patients at a high risk for coronary dissection.

Declaration of conflicting interests

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