De Bakey type I aortic dissection in a patient with idiopathic dilated cardiomyopathy

İdiyopatik dilate kardiyomiyopatili bir olguda De Bakey tip I aort diseksiyonu

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Aortic dissection is a very rare clinical entity in patients with dilated cardiomyopathy. A 30-year-old man with known dilated cardiomyopathy presented with complaints of dyspnea and fatigue. Physical examination showed increased central venous pressure and a continuous murmur on the right sternal border. Echocardiography revealed severe systolic dysfunction in both ventricles (left EF 20%, right EF 25%), dissection in the ascending aorta, and moderate aortic regurgitation. Thoracoabdominal computed tomography showed that the dissection flap extended from the sinotubular junction to the iliac bifurcation. A diagnosis of De Bakey type I aortic dissection was made. Following Bentall operation, the patient was discharged on the 15th postoperative day.

Key words: Aneurysm, dissecting/surgery; aortic aneurysm/complications/surgery; cardiomyopathy, dilated.

Dilate kardiyomiyopatili olgularda aort diseksiyonu oldukça nadir görülen bir durumdur. Dilate kardiyomiyopatili 30 yaşında erkek hasta dispne ve çabuk yorulma yakınmalarıyla başvurdu. Fizik muayenede santral venöz basıncın yükselmiş olduğu görüldü ve sternum sağ kenarı boyunca sürekli üfürüm işitildi. Ekokardiyografik incelemede her iki ventrikülde belirgin sistolik disfonksiyon (sol EF %20, sağ EF %25) ile birlikte kardiyak boşluklarda belirgin dilatasyon, çıkan aortta diseksiyon flebi ve orta derecede aort yetersizliği saptandı. Torakoabdominal bilgisayarlı tomografide diseksiyon flebinin, sinotübüler bileşkeden iliyak bifurkasyona doğru uzandığı görüldü. De Bakey tip 1 aort diseksiyonu tanısıyla Bentall ameliyatı uygulanan hasta ameliyat sonrası 15. günde taburcu edildi.

Anahtar sözcükler: Anevrizma, diseksiyon; aort anevrizması/komplikasyon/cerrahi; kardiyomiyopati, dilate.

Acute aortic dissection is an unusual clinical entity in patients with dilated cardiomyopathy. There are only a few cases of acute aortic dissection reported in patients with dilated cardiomyopathy, and, to our knowledge, the presented case is the first reported in Turkey.

CASE REPORT

A 30-year-old man with dilated cardiomyopathy was referred to our center for investigation into progressive dyspnea and severe exercise intolerance. Physical examination revealed raised central venous pressure, pitting edema of the lower limbs, and a loud systolic and diastolic murmur at the right sternal border. Electrocardiogram showed atrial fibrillation with ventricular rate of 120/minute and incomplete left bundle branch block. On chest X-ray, he had cardiomegaly and a dilated ascending aorta. Echocardiographic examination revealed dilated cardiac chambers and severe biventricular systolic dysfunction (left and right ventricular ejection fractions 20% and 25%, respectively). Dilatation of the ascending aorta with a dissection flap, false lumen, and moderate aortic regurgitation were seen in serial views (Fig. 1a-c). A thoracoabdominal computed tomographic scan showed a dissection flap extending from the sinotubular junction to the iliac bifurcation. The patient was diagnosed as having De Bakey type I aortic dissection and underwent complete replacement with a composite aortic valve-ascending aortic

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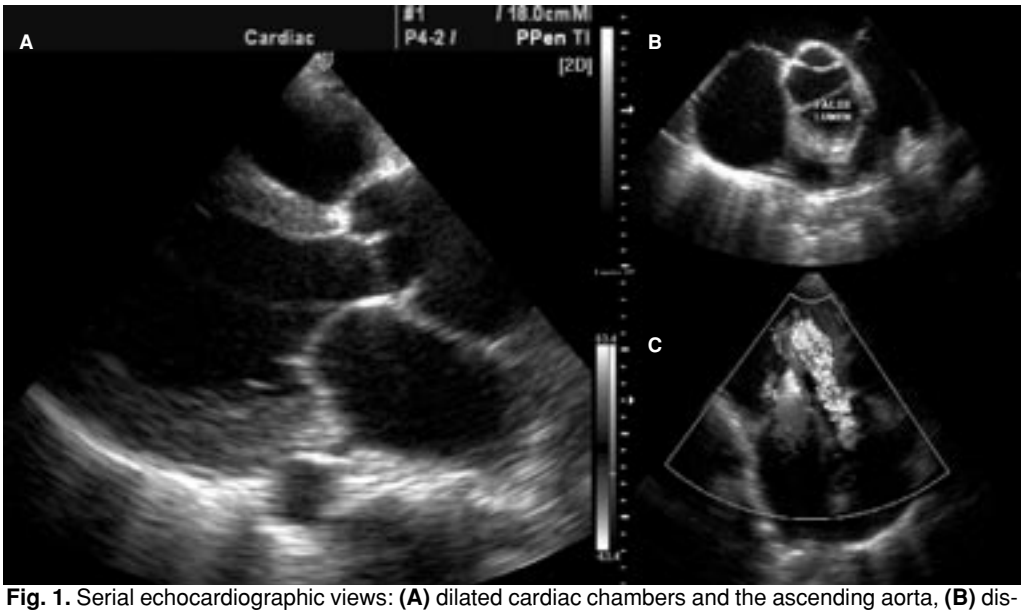

Fig. 1. Serial echocardiographic views: **(A)** dilated cardiac chambers and the ascending aorta, **(B)** dissection flap and false lumen, and **(C)** moderate aortic regurgitation.

conduit (Bentall operation) under elective settings. He had an uneventful postoperative course and was discharged on the 15th postoperative day.

DISCUSSION

The incidence of aortic dissection ranges from 5 to 30 cases per million per year. Systemic hypertension is the most common factor predisposing the aorta to dissection, the other factors being aortic dilatation and aneurysms, annuloaortic ectasia, chromosomal aberrations (e.g. Turner syndrome, Noonan syndrome), coarctation of the aorta, aortic arteritis, bicuspid aortic valve, and hereditary connective tissue disorders (e.g. Marfan syndrome, Ehlers-Danlos syndrome).[1] Marfan syndrome accounts for the majority of cases of aortic dissection in patients younger than 40 years of age.[1] Acute elevation of blood pressure secondary to cocaine use or abrupt discontinuation of beta-blocker therapy, which was prescribed to our patient, have been reported to be associated with aortic dissections and dilated cardiomyopathy.[2] None of the above-mentioned conditions were present in our patient.

The coexistence of aortic dissection and dilated cardiomyopathy is very rare and the co-occurrence of these conditions is equivocal.[3,4] The essential predisposing processes to acute aortic dissections include medial degeneration of the aortic wall, and acute, profound elevation of arterial blood pressure, causing a rapid rise in the first derivative of ventricular pressure (dP/dt) on the aortic wall.[1] The critical effect of advanced systolic heart failure is reduced aortic blood flow resulting from reduction in cardiac output and dP/dt. Reduced blood flow combined with increased heart rate causes a retrograde flow and negative shear stress along the aortic wall.[5] However, tendency to aortic dissection is still questionable despite the contribution of these hemodynamic effects. Association between the two diseases remains rather speculative, and aortic dissection may be considered a very rare cause of worsening clinical situation in a patient with dilated cardiomyopathy.

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