Acute aortic dissection due to long-term steroid therapy for Addison's disease

Addison hastalığı için uzun süreli steroid terapisi uygulanmasına bağlı olarak gelişen akut aort diseksiyonu

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A 19-year-old man was admitted to our clinic with a complaint of sudden-onset chest pain radiating to the back, which started two days ago. The patient, who had been receiving steroid therapy for Addison's disease for 17 years was diagnosed with acute type 2 aortic dissection. Bentall procedure was performed as an emergency surgical therapy under cardiopulmonary bypass by using hypothermic selective antegrade cerebral perfusion via right axillary artery. The patient was discharged with complete recovery on postoperative 11th day. Long-term steroid therapy may lead to a rise in blood pressure and subsequently to aortic dissection. While there are cases in the literature that developed acute aortic dissection associated with long-term steroid therapy in various diseases such as systemic lupus erythematosus, temporal arteritis, and aortitis syndrome, reports about the concurrent presence of Addison's disease and aortic dissection are very rare.

Key words: Addison disease; aortic dissection; steroid therapy.

On dokuz yaşında erkek hasta; iki günlük, ani başlangıçlı ve sırta yayılan göğüs ağrısı yakınması ile kliniğimize başvurdu. On yedi yıldır Addison hastalığı nedeniyle steroid tedavisi alan hastaya akut tip 2 aort diseksiyonu tanısı konuldu. Hastaya acil cerrahi tedavi olarak, sağ aksiller arter yoluyla hipotermik yarı seçici antegrad serebral perfüzyon kullanılarak kardiyopulmoner bypass altında Bentall işlemi uygulandı. Hasta ameliyat sonrası 11. günde tam iyileşmeyle taburcu edildi. Uzun süreli steroid tedavisi kan basıncında yükselmeye ve bunun sonucunda aort diseksiyonuna neden olabilir. Literatürde sistemik lupus eritamatozus, temporal arterit ve arterit sendromu gibi hastalıklarda uzun süreli steroid kullanımına bağlı aort diseksiyonu gelişen olgular olmasına rağmen Addison hastalığı ile aort diseksiyonu birlikteliğine ilişkin bildiriler çok nadirdir.

Anahtar sözcükler: Addison hastalığı; aort diseksiyonu; steroid tedavisi.

Acute aortic dissection is one of the most challenging diseases of the human aorta. It is mainly a disease of the elderly. It is rarely seen before the age of 40. Risk factors for aortic dissection are chronic hypertension, atherosclerotic disease, cardiovascular abnormalities, and genetic disorders.[1] Addison's disease, which is a result of primary adrenal failure, is associated with hypotension; however our patient had high blood pressure, which was probably related to chronic corticosteroid treatment.

In this report we present an acute aortic dissection case due to corticosteroid treatment in a young male who had Addison's disease.

CASE REPORT

A 19-year-old male was referred to our Emergency Department with acute-onset chest pain which radiated to the back that started two days before. He had been diagnosed with Addison's disease at the age of two years, and had been on steroid therapy with prednisone at 20 mg/day for 15 years, which was replaced with hydrocortisone of 30 mg/day. He had a history of left orchiectomy revealing adenoma on pathologic examination. Additionally, he had suffered from hypertension for two years just after beginning the hydrocortisone regimen. There was no family history of hypertension or genetically transmitted disease.

His body temperature was 37 °C, blood pressure was 140/65 mm Hg, heart rate was 85 beats per minute, respiratory rate was 24 breaths per minute and arterial oxygen saturation was 97% on admission. His weight was 54 kg and his height was 157 cm. On physical examination, aortic regurgitation murmur was

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prominent on auscultation. The lungs were normal. Peripheral pulses were equal in the lower extremities. Other system examinations were entirely normal.

Since his chest X-ray showed enlargement of the upper mediastinum, he was evaluated with echocardiography. A dilated ascending aorta with a dissection flap and severe aortic regurgitation were found (Fig. 1). Consequently, thoracoabdominal contrast enhanced computed tomography (CT) scan confirmed the echocardiographic findings. The diameter of the ascending aorta was 4.4 cm with a prominent intimal flap seen in the ascending aorta, showing an acute type 2 aortic dissection emergency surgery was performed when the diagnosis was confirmed.

At operation, a type 2 dissection was seen without any rupture to pericardium (Fig. 2a). After right axillary artery and two-stage atrial cannulation, cardiopulmonary bypass was instituted and maintained using semiselective antegrade cerebral perfusion method. Just after aortic cross-clamping, the ascending aorta was transected 2 cm distal to the coronary ostia, and cold blood cardioplegia was given via coronary ostia. The aortic valve was tricuspid and the cusps were thickened. An intimal tear was located in the ascending aorta 2 cm distal to the coronary ostia (Fig. 2b). We decided to replace the ascending aorta and the aortic valve. A Button Bentall procedure was performed using 30 mm Dacron composite graft (Fig. 2c). Administration of 20 mg twice a day prednisolone was started early after the operation and reduced to 20 mg/day over a period of three weeks. Histopathological examination of the resected aorta was performed. There were no findings of vasculitis or myxomatous degeneration in the media layer but medial degeneration and atherosclerosis including fibrosis of elastic structures, and necrosis of smooth muscle cells were found. He had an uneventful recovery and was discharged on the 10th postoperative day with prednisolone, warfarin and beta-blocker treatment.

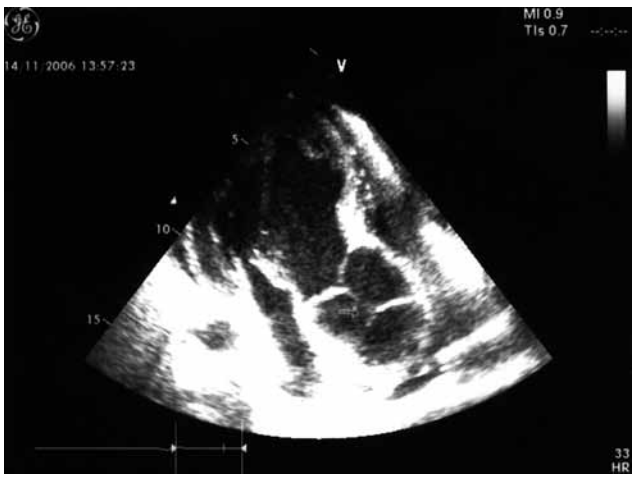

Fig. 1. Echocardiographic imaging revealed, a dilated ascending aorta with a dissection flap and a severe aortic regurgitation.

DISCUSSION

The most prominent finding of non-traumatic aortic dissection is medial degeneration of the aorta. It is generally

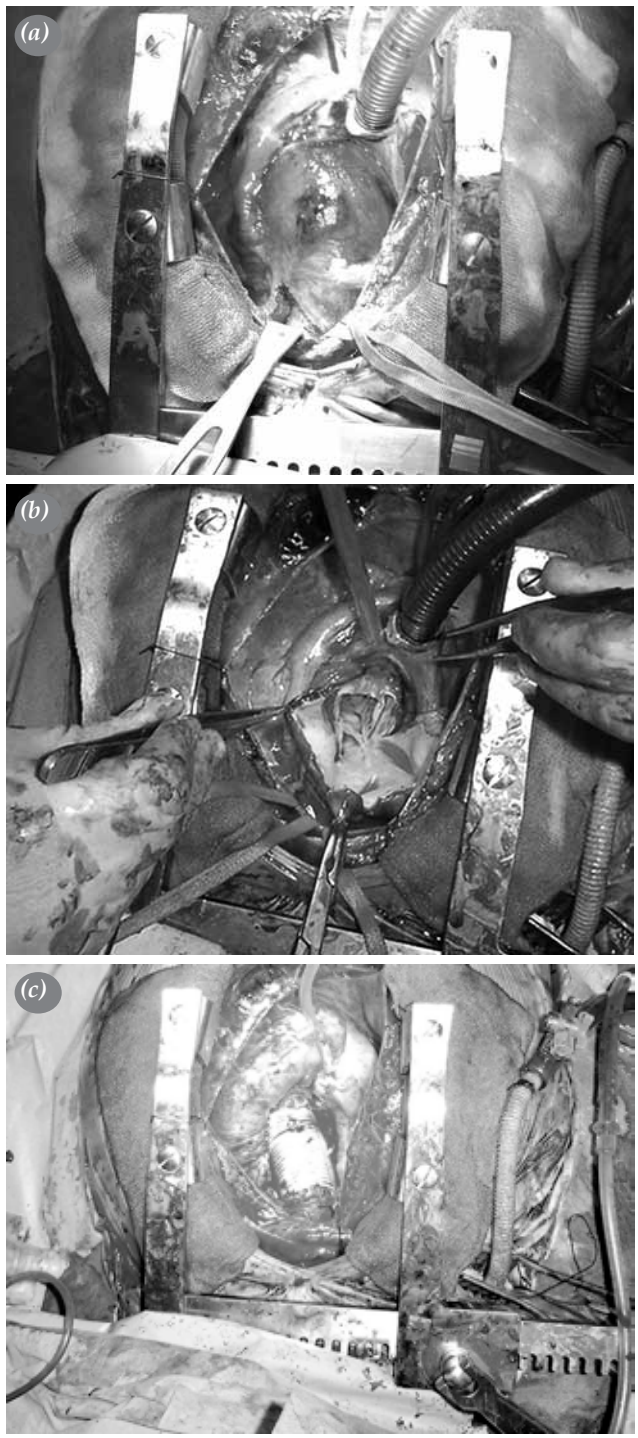

Fig. 2. (a) Operative view of dilated ascending aorta. **(b)** Intimal tear was located in the ascending aorta. **(c)** Button Bentall procedure (by using 30 mm Dacron composite graft).

due to underlying disease, like atherosclerosis, or hypertension.[1] There are several predisposing conditions for aortic wall weakening which result in a tendency for aortic dissection at a younger age, including genetic disorders like Marfan's disease, Ehlers-Danlos and Turner syndromes. Additionally physiologic factors including pregnancy, a dilated aortic root, coarctation of the aorta and a bicuspid aortic valve cause chronic stress that result in earlier weakening of the aortic wall.[2] The adrenal cortex produces and secretes insufficient amounts of glucocorticoids, mineralocorticoids, and androgens in Addison's disease. Corticosteroids, usually hydrocortisone or prednisone, are used for Addison's disease as it can be life-threatening.[3] Unfortunately, chronic steroid usage may cause fragility of the vessels due to its negative effect on collagen formation and connective tissue strength. Many studies reported that one of the most important side effects of long-term steroid therapy for the treatment of various diseases is the occurrence of atherosclerotic changes in the arterial system, which in fact is the probable cause or pathogenesis of aortic dissection or aneurysm due to inhibition of arterial injury repair. Choi et al.[4] reported that hypertension and long-term corticosteroid therapy were common features in younger patients with dissecting aneurysm. Cumulative steroid doses given in the treatment period are correlated with the severity of their atherogenic effects. In these patients, the presence of atherosclerosis is more important in surgical repair of aortic dissection because of the fragility of aortic tissue.[5] Furthermore, long-term steroid therapy can also stimulate increasing blood pressure therefore raising the incidence of hypertension. In most cases the aortic dissection induced by sudden hypertension is generally associated with mural thinning of the aortic wall or fixed atherosclerotic plaques due to long-term steroid therapy.[5]

The family history of the patient did not include any aortic dissection or connective tissue disorder like Marfan's syndrome. Additionally, he did not meet diagnostic criteria for Marfan's syndrome defined by De Paepe et al.[6] The patient described herein had been on steroid therapy for 17 years (prednisone of 20 mg/day for 15 years, and hydrocortisone 30 mg/day for the last two years). But it should be kept in mind that although many patients use long-term steroids for various clinical conditions, acute aortic dissection is not seen frequently among them. In our case, the presence of hypertension due to steroid usage possibly contributed to aortic aneurysm or intimal tearing.

In conclusion, the occurrence of aortic dissection in patients with Addison's disease is extremely rare. In addition to hypertensive effects, long-term steroid therapy may have promoted the development of aortic dissection in this patient by accelerating the atherosclerotic process. Close follow-up is necessary to prevent aortic aneurysm and aortic dissection formation in patients with long-term steroid therapy.

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REFERENCES

1. Erbel R, Alfonso F, Boileau C, Dirsch O, Eber B, Haverich A, et al. Diagnosis and management of aortic dissection. Eur Heart J 2001;22:1642-81.
2. Basso C, Frescura C, Corrado D, Muriago M, Angelini A, Daliento L, et al. Congenital heart disease and sudden death in the young. Hum Pathol 1995;26:1065-72.
3. Oelkers W. Adrenal insufficiency. N Engl J Med 1996; 335:1206-12.
4. Choi KH, Rim SJ, Lee SK, Jang BC, Cho SH. Dissecting aortic aneurysm with aortic-valve insufficiency in systemic lupus erythematosus. Nephrol Dial Transplant 1999;14:969-73.
5. Korkut AK, Wellens F, Foubert L, Goethals M. Successful treatment of acute dissection of the donor aorta after orthotopic heart transplantation. J Heart Lung Transplant 2003;22:701-4.
6. De Paepe A, Devereux RB, Dietz HC, Hennekam RC, Pyeritz RE. Revised diagnostic criteria for the Marfan syndrome. Am J Med Genet 1996;62:417-26.