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Case Report

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Successful Kidney Transplantation in a Patient with Marfan's Syndrome with a History of Aortic Dissection Operation and a Dissection Flap Up to the Iliac Bifurcation Level

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ABSTRACT

Marfan's syndrome is a collagen tissue disease characterized by aortic aneurysm and dissection. In our study, a case who was diagnosed with Marfan's syndrome had a dissection flap up to the iliac bifurcation level and underwent successful renal transplantation; is presented. A 27-year-old male patient had a history of operations due to 2 aortic dissections. In the physical examination, he had hyperflexible joints, arachnodactyly, a pansystolic murmur in the mitral focus, and severe myopia. Echocardiography revealed enlargement of the aortic root, and a dissection flap extending from the abdominal aorta to the iliac bifurcation was detected in tomographic angiography. Genetic analysis revealed Fibrillin-1 gene exon 66 region c.8384T > C (p.Ile2795Thr) missense heterozygous mutation. The patient, who was diagnosed with Marfan's syndrome, underwent kidney transplantation from his healthy and non-mutated sibling. There were no vascular complications in the 1-year-period after transplantation, and he is followed up with normal kidney function. In our study, it has been shown that successful kidney transplantation can be performed in a patient with Marfan's syndrome who has a dissection flap up to the iliac bifurcation level.

Keywords: Renal transplantation, Marfan's syndrome, dissection

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INTRODUCTION

Kidney transplantation (KTX) is a preferred treatment modality for end-stage kidney disease patients (ESKD).[1] The vascular anatomy of the donor and recipient is very important in KTX. Thoracoabdominal aortic aneurysm or dissection is one of the most important vascular problems in patients with Marfan's syndrome (MFS).

Marfan's syndrome with a reported incidence of approximately 1/5000 individuals is a predominantly autosomal dominant inherited disorder of connective tissue. It is caused by mutations in the extracellular matrix protein fibrillin-1 (FBN-1) that causes ocular, skeletal, and important cardiovascular manifestations.[2,3] The diagnosis of MFS is made using the Ghent criteria. It is largely based on clinical manifestations from various organ systems and genetic testing.[4] Marfan's syndrome displays a number of abnormalities of the thoracic and abdominal aorta, ranging from abnormal aortic stiffness to aortic aneurysm.[5,6]

In our study, a patient with MFS who had an aortic dissection flap up to the iliac bifurcation level and who later had a kidney transplant from his sibling is presented.

CASE PRESENTATION

A 27-year-old male patient presented to our center for kidney transplantation. He had a history of 2 operations for aortic dissection. It was learned that after the first operation (July 2018), kidney functions deteriorated and then chronic kidney disease developed and followed; dialysis was started 4 months ago, and the

patient had lost his mother (30 years old) because of the aortic dissection. Olmesartan, Nebivolol, and Doxazosine were used as antihypertensives.

Physical examination revealed a height of 183 cm, blood pressure was 140/90 mmHg, pansystolic murmur in the mitral focus, hyperflexible elbow joint, marfanoid appearance, and arachnodactyly. Except for myopia, no finding in favor of MFS was detected in the eye examination. Preoperative serum creatinine level was 7.7 mg/dL (eGFR: 8.5 mL/min). ECG: There was normal sinus rhythm and T wave abnormalities.

There was enlargement of the aortic root, left ventricle, and atrium dilatation and mitral regurgitation, anterior septum mid and apex hypokinesia with a normal ejection fraction in his echocardiographic examination. Evaluated by preoperative coronary computerized tomographic angiography (CTA), no pathology was detected except for operation, aneurysm, and dissection findings. Computerized tomographic angiography of the patient revealed a prosthetic graft extending from descending aorta to mid thoracic aorta, aneurysmatic dilatation of ascending aorta, and linear dissection flap extending from the distal thoracic aorta to the beginning of iliac bifurcation, and normal external and internal iliac arteries (Figures 1 and 2).

The genetic testing for MFS from the patient resulted in c.8384T > C (p.Ile2795Thr) missense heterozygous mutation of FBN-1 gene exon 66 region.

Our patient was diagnosed with MFS (within the scope of Ghent criteria) due to the history of aortic dissection and aneurysm operation, FBN-1 mutation, hyperflexible elbow joint, arachnodactyly, myopia, and a history of death in the mother due to aortic dissection at a young age (suspicious presence of family history).

The donor was a 31-year-old female, sibling, with no known disease, the systemic examination was normal, and there was 1 renal artery and vein for each kidney in CTA. No mutation was detected. Kidney transplantation was performed with left laparoscopic donor nephrectomy and external iliac artery and vein anastomosis.

MAIN POINTS

- Our case study has shown that successful kidney transplantation can be performed in patients with Marfan's syndrome (MFS).
- Our case is an MFS patient with a dissection flap up to the level of the iliac artery bifurcation, and no vascular complication developed at the end of the first year.
- In line with the literature recommendations, blood pressure control is provided with beta-blockers and renin angiotensin system blockers and is followed up with normal kidney function.

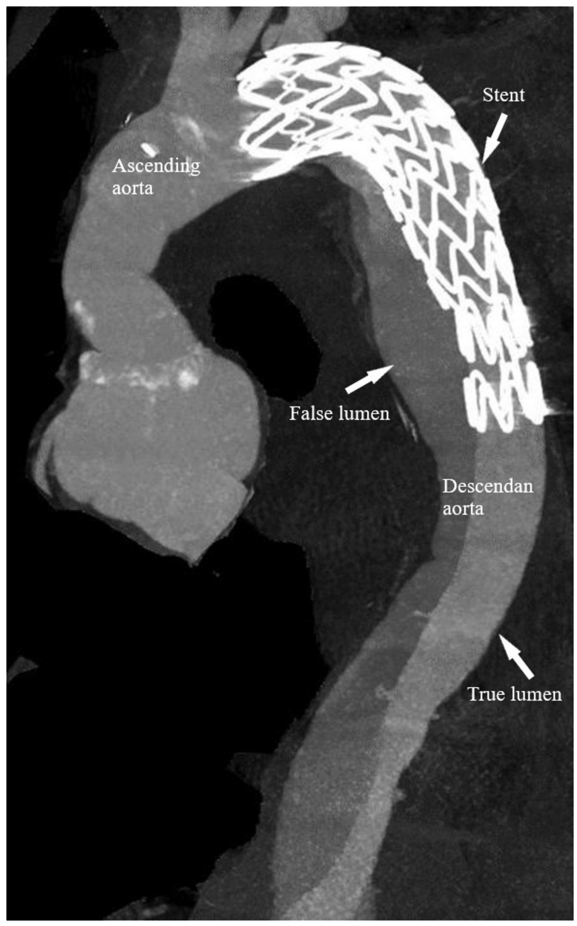

Figure 1. Computerized tomographic angiography; stent (extending from ascending aorta to descending aorta), intimal dissection extending to the abdominal aorta and true/false lumens.

Lymphocyte cross match and panel reactive antibody tests were negative, and there was 6/6 HLA matching. The initial immunosuppressive protocol consisted of tacrolimus, mycophenolate mofetil, and prednisolone. No vascular complications developed, and the patient was discharged on the 5th day with normal kidney function (serum creatinine level 1.2 mg/dL, CKD-EPI eGFR: 75 mL/min/1.73m^2) and blood pressure (130/80 mmHg). Nebivolol 5 mg/day and Olmesartan Medoksomil 20 mg/day were used in antihypertensive treatment.

In the postoperative renal Doppler examinations, it was shown that the renal artery flow velocity and forms were normal, the renal vein was open, the resistive index was 0.5-0.6 (upper, middle, and lower pole), and there was no dissection or aneurysm of the iliac artery. At the 5th month of iliac CTA, it was observed that the dissection flap was stable, did not exceed the iliac bifurcation level, and the iliac arteries were intact (Figure 3). In the follow-up, rejection attacks did not develop and kidney functions were normal. In the postoperative 1st year, blood pressure was 120/80 mmHg, serum creatinine level was 1.13 mg/dL, CKD-EPI eGFR: 87.3 mL/min/1.73 m^2.

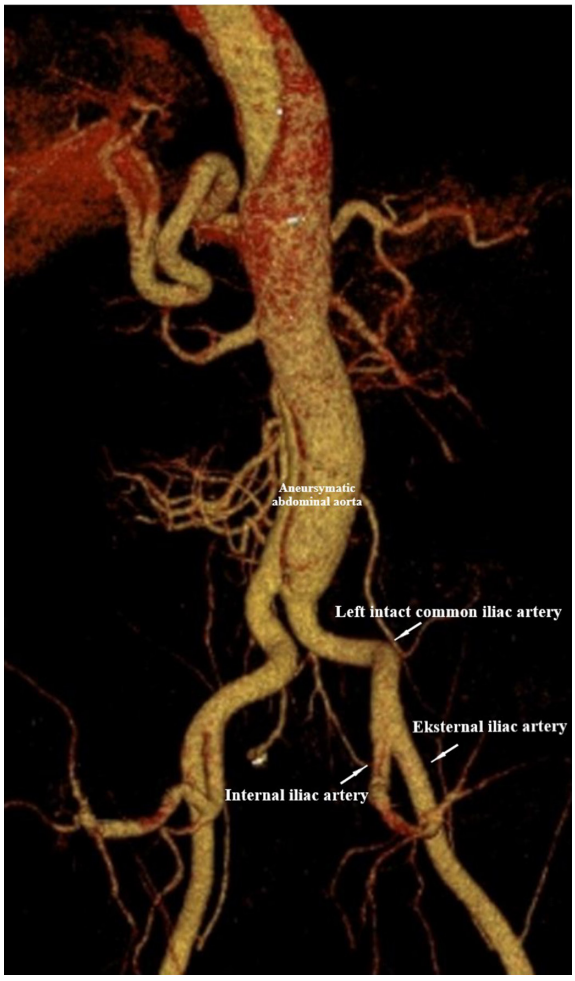

Figure 2. Computerized tomographic angiography; aneursymatic abdominal aorta, left intact common iliac artery, external and internal iliac artery.

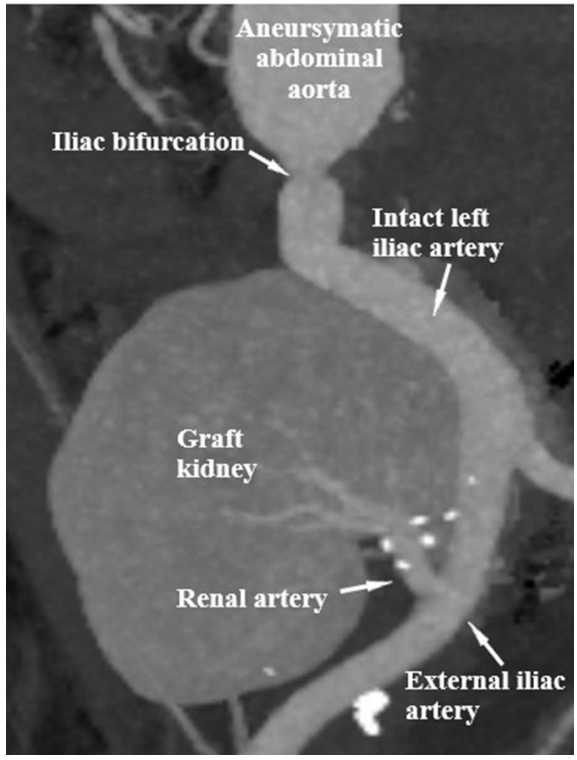

Figure 3. Postoperative computerized tomographic angiography; aneursymatic abdominal aorta, transplanted (graft) kidney, intact iliac bifurcation, intact left iliac artery, external iliac artery, and renal artery.

DISCUSSION

A case who was diagnosed with MFS had a dissection flap up to the iliac bifurcation level and underwent successful kidney transplantation is presented here. This MFS case is characteristic in 3 respects: (1) to the best of our knowledge, our patient is the second MFS patient who received a living donor kidney transplant, (2) having a dissection flap up to the iliac bifurcation, and (3) FBN-1 gene mutation, which has not yet been defined in data banks.

Regardless of whether it is related to MFS or not, kidney dysfunction due to kidney ischemia may develop after thoracoabdominal aortic aneurysm and dissection operations (approximately 30%) and rarely progress to ESKD.[7] In addition, there is a risk of dissection in vascular anastomosis operations in patients with MFS. Ryosaka et al[8] presented the first case of MFS with living donor kidney transplantation. The 2nd year results of successful kidney transplantation in a pregnant female patient who developed ESKD due to kidney ischemia due to massive hemorrhage after aortic dissection and aneurysm operation are presented. Vascular complications and rejection did not develop after the KTX, and it was stated that he was followed up with normal kidney function. In our case, due to the same reasons, ESKD developed, hemodialysis was started, and then a successful kidney transplant was performed from his sister. Our patient did not develop any vascular complications or rejection, and he is being followed up with normal kidney function.

The other important point in MFS is the control of blood pressure. Renin-angiotensin aldosterone system antagonists and β-blockers are generally suggested. Especially, β-blocker is suggested as it may decrease cardiac contractility, pulse pressure, and the risk of aortic dissection.[9] In our case, blood pressure control was obtained with Nebivolol and Olmesartan, and no complications were experienced.

This case study, together with the first case study, demonstrated that successful kidney transplantation can be performed in patients with MFS. In addition, although dissections involving the iliac artery are very rare in patients with MFS, it should be kept in mind that it is very important to periodically follow up the cases with CT or magnetic resonance imaging.

Ethics Committee Approval: The study was approved by the Akdeniz University Medical Faculty Clinical Research Ethics Committee (Date: July 7, 2021, Decison No: KAEK-530).

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

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Declaration of Interests: The authors declare that they have no competing interest.

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