

Renal Autotransplantation in a Patient with Bilateral Renal Artery Stenosis Secondary to Takayasu Arteritis

E. Mekik Akar^{1*},
F. Aydın²,
A. Tüzüner³,
S. Fitöz⁴,
S. Öztürk⁵,
E. D. Kurt Şükür¹,
U. Şanlıdilek⁶,
E. Çelikel²,
Z. B. Özçakar¹,
N. Çakar¹,
F. Yalçinkaya¹

¹*Pediatric Nephrology Department, School of Medicine, Ankara University, Ankara, Turkey*

²*Pediatric Rheumatology Department, School of Medicine, Ankara University, Ankara, Turkey*

³*General Surgery Department, School of Medicine, Ankara University, Ankara, Turkey*

⁴*Pediatric Radiology Department, School of Medicine, Ankara University, Ankara, Turkey*

⁵*Pediatrics Department, and School of Medicine, Ankara University, Ankara, Turkey*

⁶*Radiology Department, School of Medicine, Ankara University, Ankara, Turkey*

ABSTRACT

Involvement of the renal artery is common in Takayasu arteritis. We, herein, present on a patient with Takayasu arteritis causing severe renal failure and a successful auto-transplantation. This case shows that early diagnosis and immediate appropriate interventions are life-saving in patients with Takayasu arteritis. Renal auto-transplantation performed in selected cases increases dialysis-free survival.

KEYWORDS: Takayasu arteritis; Renal artery stenosis; Renal insufficiency; Transplantation, autologous

INTRODUCTION

Takayasu arteritis (TA) is a large-vessel vasculitis that primarily affects the aorta and its primary branches. The disease is characterized by granulomatous inflammation of the vessel wall with an elusive etiopathogenesis. Involvement of the renal artery is common in TA, which results in renal artery stenosis and renovascular hypertension. In severe cases, it can even cause total occlusion of the renal artery and if bilateral, result in renal failure [1, 2]. We, herein, present on a case of TA causing bilateral renal artery stenosis leading to renal failure and auto-transplantation.

CASE REPORT

A 17-year-old male patient was visited in a local hospital with periorbital and pretibial edema, and back pain. Due to severe hypertension, renal doppler ultrasound was performed that revealed bilateral renal artery stenosis. He was referred to our clinic for further investigation and treatment. On physical examination at admission, he had periorbital and pretibial edema and a periumbilical bruit. His blood pressure was measured at 185/115 mm Hg on both arms and 240/160 mm Hg on the lower extremity. He had an initial creatinine level of 1.2 mg/dL, ESR of 63 mm/hr, and CRP level of 52.4 mg/L. Renal doppler ultrasonography demonstrated total occlusion of the left renal artery, severe stenosis (nearly occlusion) of the right artery, and vascular wall thickening of the abdominal aorta and superior mesenteric artery (Fig 1).

MR angiography showed wall thickening and

*Correspondence: Ece Mekik Akar, Ankara University School of Medicine, Pediatric Nephrology Department, Dikimevi, Cebeci Çankaya/Ankara, Turkey 06590
Tel: +90-535-862-2883
E-mail: emekik@yahoo.com

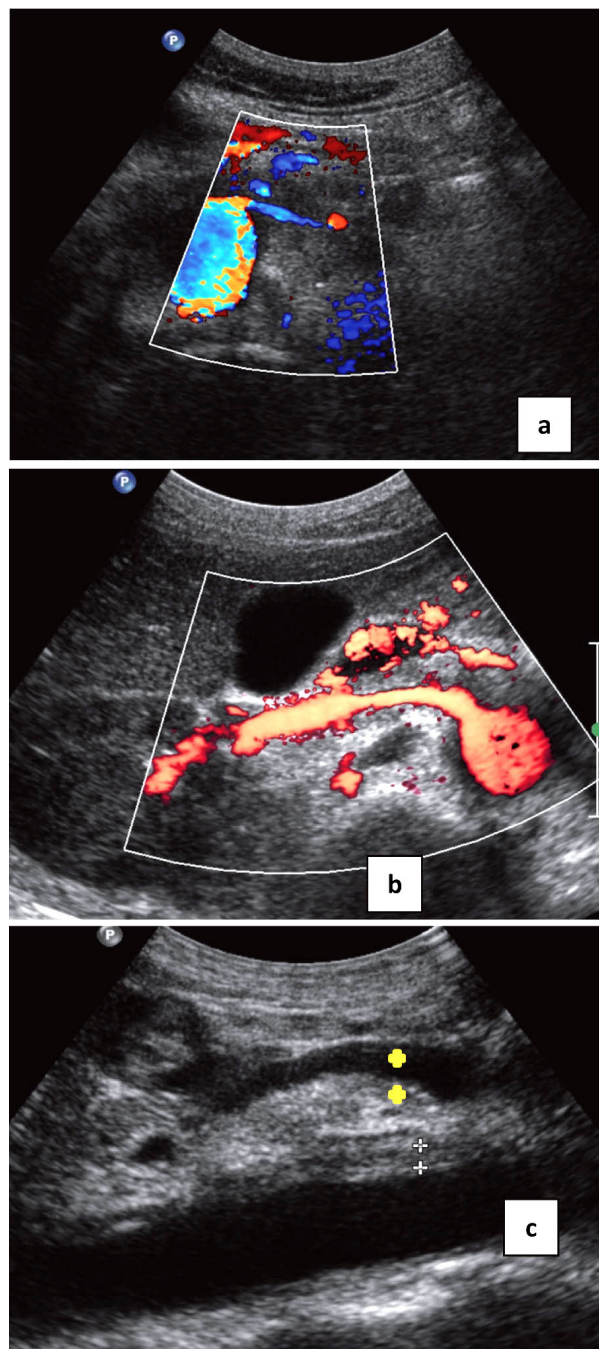


Figure 1: Renal doppler ultrasonography findings of the patient. (a) Total occlusion of left renal artery, (b) proximal narrowing of the right renal artery, and (c) irregular and thickened vascular wall of the abdominal aorta

perivascular inflammation of the aorta and its major branches, significant narrowing in the superior mesenteric artery and right renal artery, total occlusion of left renal artery and decreased T_2 signal density in the left kidney suggesting diffuse ischemia and infarct (Fig 2).

According to these findings, the diagnosis of TA was made. Pulse methylprednisolone therapy (1 g/dose) was immediately started at the first day of admission. After 12 hours of the treatment, he became anuric, his edema increased, and his serum creatinine level raised to 4 mg/dL. Urgent hemodialysis was performed. Due to the rapid progression of the disease, intravenous pulse cyclophosphamide (CYC) was given. Despite daily hemodialysis and intense immunosuppressive therapy with CYC and a 5-day course of pulse methylprednisolone, his creatinine levels continued to rise (7 mg/dL). On the sixth day of admission, the attempt for revascularization of renal arteries by percutaneous transluminal angiography (PTA) was unsuccessful. Considering his severe renal failure and ineffective medical and endovascular interventions, renal auto-transplantation was performed. His relatively preserved right kidney was dissected and directly anastomosed to the right external iliac artery. The right kidney was preferred as MR angiography revealed no perfusion in the left kidney. The right kidney was procured by pure laparoscopic donor nephrectomy technique. The kidney was retrieved through a right modified Gibson incision by opening peritoneum. After retrieval, the peritoneum was closed. The right renal artery was trimmed about 1 cm at back-table. The right kidney transplantation was performed through the same incision. Surgery was uneventful despite the use of methylprednisolone and CYC. Post-operatively, diuresis began immediately. The creatinine level started decreasing right after the surgery and returned back to normal (1.0 mg/dL) after 10 days. Anti-platelet therapy with aspirin was commenced after the surgery. Immunosuppressive therapy was continued with 60 mg/day oral corticosteroid; a second dose of CYC was given 3 weeks after the transplantation. Hypertension could be partially controlled with amlodipine, doxazosin, propranolol, and minoxidil. The patient was discharged on the 22nd day of the transplantation with normal creatinine, ESR, and CRP levels (0.8 mg/dL, 4 mm/hr, and 0.3 mg/dL, respectively), normal urinary output and fully functioning auto-transplanted right kidney. Tocilizumab therapy was initiated two

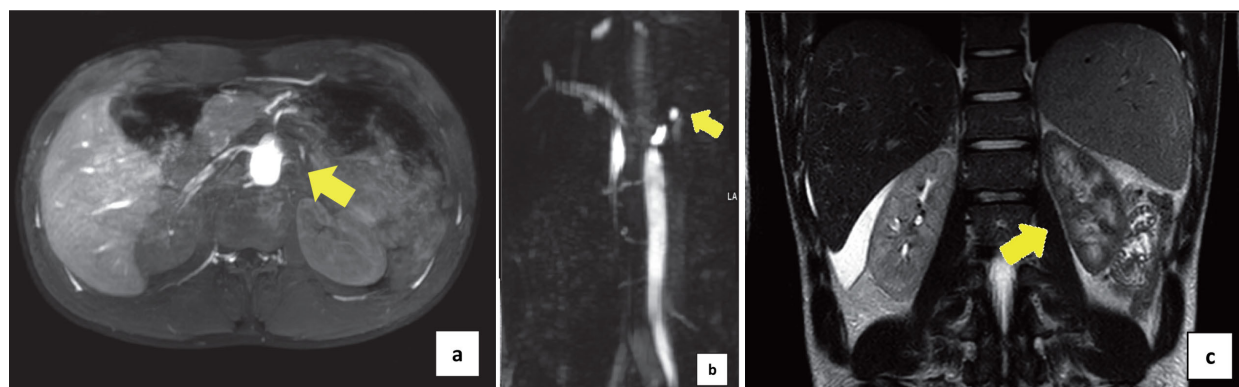


Figure 2: MR angiography images of the patient: (a and b) The left renal artery is totally occluded while the right renal artery shows significant narrowing; (c) ischemic changes in the left kidney

months after the operation. At the last visit (9 months of operation), anti-hypertensive drugs were reduced as severity of hypertension decreased; the patient was in complete remission with normal renal function.

DISCUSSION

The most common presentation of TA is renovascular hypertension, which results from the involvement of the main renal arteries. During the disease course, its incidence increases to 60% [3]. Generally, the stenosis is unilateral and proximal, but it can also be bilateral. In our patient, the initial symptoms were hypertension and renal failure, which indicated a severe disease course. Therefore, intense immunosuppressive therapies started immediately to preserve the perfusion of the kidneys.

In the advanced stages of the disease, main principles of treatment include revascularization of the affected organs in order to preserve the organ perfusion. Revascularization can be provided by endovascular interventions or surgery. Endovascular interventions include balloon angioplasty and stent placement; surgical options include aorta-renal bypass and auto-transplantation. Both surgical and endovascular interventions are not recommended during the acute phase of the disease [4]. As a general rule, inflammation on the vessel wall should be suppressed before any interventions [2]. Studies show that patients with high CRP, ESR and fibrinogen levels tend to have more arterial complications than others [5]. Al-

though our medical therapy attempts failed to preserve perfusion of the kidneys, we believe that they helped to reduce the inflammation in the vessel walls and enabled us to perform interventional modalities. Immunosuppressive treatments are also recommended in the post-interventional period [6].

Percutaneous transluminal angioplasty (PTA) is suggested to be the first line of treatment as it is a less invasive and safe method [2]. Our first attempt for revascularization was PTA, consistent with universal approach, but it was unfortunately not successful. As surgical options were reported to be considered in patients who are unresponsive to medical and endovascular treatments, we performed auto-transplantation for our patient, after medical therapy and PTA had failed [7].

Renal auto-transplantation was firstly performed by James Hardy in 1963 to repair a high ureteral injury [8]. Since then, this procedure has been used for ureteral injuries complex as well as renovascular diseases [9]. Auto-transplantation is a safe method with very low complication rates, especially in experienced transplant centers [10]. Auto-transplantation is an organ-sparing and dialysis-dependence-preventing treatment option for TA-induced renal artery stenosis in patients in whom medical care or PTA has failed or is technically impossible. As recurrent endovascular interventions with prolonged disease course have been reported to result in peri-arterial fibrosis, which makes the operation more difficult, early surgical interventions

increase the success rate of the method. Also, prolonged ischemia period of kidneys, lowers the chance of dialysis-free-survival [4].

Due to rarity of the disease, there is limited data about children with TA and auto-transplantation. However, almost all of them found auto-transplantation a successful surgical option. The first pediatric auto-transplantation case series was reported in 1985 where 16 patients with renal artery diseases (2 with TA) were studied. All ended up with excellent renal function and free of hypertension [11]. The most recent pediatric series was reported by Corbetta, *et al.*, which included 13 patients with renovascular hypertension, five of whom had TA; 61% of their patients was fully recovered; 39% had clinical improvement after a median follow-up of 53.4 months [12].

Rates of morbidity and mortality related to auto-transplantation were found to be very low. Starting from the early post-operative period, urine output, renal functions and hypertension improved; this well-being state was maintained in long-term follow-up. Although nearly six months might be needed for complete recovery of the hypertension, reduction in the number and dose of anti-hypertensive drugs can be achieved after a short while [4, 9, 12-14]. In our case, while diuresis and renal recovery occurred just after the surgery, hypertension continued which was presumably due to the remaining ischemic left kidney. Even so, a left nephrectomy simultaneously with renal auto-transplantation was not performed; it was a more aggressive procedure. Additionally, as left kidney had no perfusion on MR angiography, we believed that it would become atrophic soon. We could control the hypertension with drugs within nine months post-transplantation.

Long-term follow-up is recommended for patients with TA who undergo renal auto-transplantation for monitoring the patient for disease progression and restenosis [4].

In conclusion, it seems that early diagnosis and immediate appropriate interventions are life-saving in TA. Renal auto-transplantation

performed in selected cases is a kidney-saving method and provides dialysis-free-survival in experienced transplant centers.

CONFLICTS OF INTEREST: None declared.

REFERENCES

1. Chaudhry MA, Latif F. Takayasu's arteritis and its role in causing renal artery stenosis. *Am J Med Sci* 2013;**346**:314-8.
2. Keser G, Direskeneli H, Aksu K. Management of Takayasu arteritis: a systematic review. *Rheumatology (Oxford)* 2014;**53**:793-801.
3. Maritati F, Iannuzzella F, Pavia MP, *et al.* Kidney involvement in medium- and large-vessel vasculitis. *J Nephrol* 2016;**29**:495-505.
4. Haberal HB, Tonyali S, Peynircioglu B, *et al.* Renal Autotransplantation with Autologous Saphenous Vein Graft in a Patient with Takayasu Arteritis and Existing Renal Artery Stent in Her Solitary Kidney. *Urol Int* 2018;**100**:181-4. doi: 10.1159/000475509.
5. Saadoun D, Lambert M, Mirault T, *et al.* Retrospective analysis of surgery versus endovascular intervention in Takayasu arteritis: a multicenter experience. *Circulation* 2012;**125**:813-9.
6. Min PK, Park S, Jung JH, *et al.* Endovascular therapy combined with immunosuppressive treatment for occlusive arterial disease in patients with Takayasu's arteritis. *J Endovasc Ther* 2005;**12**:28-34.
7. Lacombe M. [Role of surgery in the treatment of renovascular hypertension in the child]. *Bull Acad Natl Med* 2003;**187**:1081-93; discussion 93-4. [in French]
8. Hardy JD, Eraslan S. Autotransplantation of the Kidney for High Ureteral Injury. *J Urol* 1963;**90**:563-74.
9. Hau HM, Bartels M, Tautenhahn HM, *et al.* Renal autotransplantation--a possibility in the treatment of complex renal vascular diseases and ureteric injuries. *Ann Transplant* 2012;**17**:21-7.
10. Yakupoglu YK, Aki FT, Kordan Y, *et al.* Renal Auto-transplantation at Three Academic Institutions in Turkey. *Urol Int* 2016;**97**:466-72.
11. Jordan ML, Novick AC, Cunningham RL. The role of renal autotransplantation in pediatric and young adult patients with renal artery disease. *J Vasc Surg* 1985;**2**:385-92.
12. Corbetta JP, Duran V, Burek C, *et al.* Renal auto-transplantation for the treatment of renovascular hypertension in the pediatric population. *J Pediatr Urol* 2011;**7**:378-82.
13. Kim HK, Lee JM, Cho MH, Huh S. Aortic stent and renal autotransplantation for the management of renovascular hypertension with Takayasu's arteri-

tis: report of a case. *Surg Today* 2014;**44**:944-7.
14. Chiche L, Kieffer E, Sabatier J, *et al.* Renal auto-

transplantation for vascular disease: late outcome according to etiology. *J Vasc Surg* 2003;**37**:353-61.