

CASE REPORT

A Rare Cause of Renovascular Hypertension: Takayasu Arteritis with Only Renal Artery Involvement

Ali Borazan

Mustafa Kemal University Faculty of Medicine, Department of Nephrology, Hatay, Turkey

Omur Gokmen Sevindik

Dokuz Eylul University Faculty of Medicine, Department of Nephrology, Turkey

Dilek Solmaz

Dokuz Eylul University Faculty of Medicine, Department of Romatology, Turkey

Aytaç Gulcu

Dokuz Eylul University Faculty of Medicine, Department of Radiology, Turkey

Caner Cavdar, Aykut Sifil, and Ali Celik

Dokuz Eylul University Faculty of Medicine, Department of Nephrology, Turkey

Servet Akar

Dokuz Eylul University Faculty of Medicine, Department of Romatology, Turkey

Yigit Goktay

Dokuz Eylul University Faculty of Medicine, Department of Radiology, Turkey

Taner Camsari

Dokuz Eylul University Faculty of Medicine, Department of Nephrology, Turkey

Takayasu arteritis is a chronic inflammatory disease that affects mainly the aorta, main branches of aorta, and pulmonary arteries with unknown etiology. Disease affecting solely the renal arteries is rare. We will present a case that had hypertension, hypokalemia, and metabolic alkalosis where the etiology was type 2 Takayasu arteritis, affecting renal arteries.

Keywords Takayasu arteritis, hypocalcemia, metabolic alkalosis, hypertension

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Address correspondence to Ali Borazan, Mustafa Kemal Üniversitesi Tıp Fakültesi, Nefroloji BD, 31100 Hatay, Turkey; Fax: +90 0(326) 2790929; E-mail: borazanali@hotmail.com

INTRODUCTION

Takayasu arteritis was first described by Japan ophthalmologist Dr. Mikito Takayasu in 1908. The incidence of the disease varies worldwide and is increasing over time. It is a rare disease affecting mainly the arcus aorta, the main branches of aorta, and, rarely, the peripheral arteries. It is seen more in women under age of 40. It's also called the pulseless arteritis because it affects main branches of aorta. A difference in both arms for systolic blood pressure of more than 10–20 mmHg and claudication of the extremities makes the physician consider a possible diagnosis of Takayasu arteritis.^[1–3]

In this report, we present the case of a 19-year-old girl in whom we diagnosed Takayasu arteritis while investigating the etiology responsible for hypertension, hypokalemia, and metabolic alkalosis.

CASE

A 19-year-old female, first complaining about headache mainly localized at the retro-orbital area, admitted to a state hospital two and one-half months ago. The computed tomography of the brain revealed no pathological findings. The physical examination revealed a 150/110 blood pressure. Patient was put on amlodipine 5 mg a day and referred to a nephrology clinic for further evaluation. She was charged to the nephrology ward in order to be investigated because of the accompanying hypokalemia and metabolic alkalosis.

In physical examination, the blood pressure was 140/90 mmHg and the heart rate was 78/min. No other pathological signs were detected. There was no difference between blood pressures measured from both arms. Routine laboratory tests revealed no pathology among complete blood count, urinalysis, blood urea nitrogen, creatinine, total protein and albumin, sodium, calcium, or chloride levels. Rheumatoid factor, anti-cardiolipin Ig M and G, and anti-nuclear antibody were all negative. Erythrocyte sedimentation rate and C-reactive protein level were 34 mm/hr (5–20 mm/h) and 1.31 mg/dL (<3 mg/dL), respectively. Serum potassium levels varied between 3.1 and 3.5 mEq/L. Arterial blood gas analysis was consistent with metabolic alkalosis (pH: 7.50, HCO₃: 32.4 mmol/L, PaCO₂: 30.8 mmHg).

In daily excreted urinalysis the Na, potassium, chloride, and calcium levels were Na: 13 mmol/day (40–220 mmol/day), K: 16.2 mmol/day (25–125 mmol/day), Cl: 18.2 mmol/day (110–250 mmol/day), and Ca: 142.4 mg/day (100–320 mg/day). Plasma renin activity was measured as 8.5 ng/mL/hr (0.2–3.4 ng/mL/hr). The plasma aldosterone level was 295 pg/mL (20–240 pg/mL). The ratio of renin/aldosterone was 28.

In abdominal ultrasonography, the sizes of kidneys were in normal ranges, and the echogenicity of parenchyma was not increased bilaterally. In Doppler ultrasonography, stenosis of left renal artery and arterial wall thickening among the aorta were detected. The resistive index of right renal artery was in normal ranges, but it was severely decreased at the left side due to the proximal stenosis of artery (0.29–0.3).

In abdominal tomography, nothing pathological was detected about the surrenal glands, but there was concentric wall thickening at the abdominal aorta that was consistent with arteritis.

MR angiography showed that the arch of aorta and major branches were all potent with normal calibration. At infrarenal abdominal aorta, there was obvious pathological involvement, and calibration became thinner. Nevertheless, the calibration returned to normal just beyond iliac bifurcation. There was severe stenosis just at the origin of left renal artery. Superior mesenteric artery and celiac trunk was potent with normal calibration (see Figure 1).

Intra-arterial aortography was applied to the patient, and pathological involvement was detected at infrarenal segment of abdominal aorta. The calibration became thinner at that segment and returned to normal just beyond iliac bifurcation. High-grade stenosis at the origin of left renal artery was seen, and other branches of the abdominal aorta were all normal (see Figure 2).

The definitive diagnosis of Takayasu arteritis, which involves the infrarenal segment of the abdominal aorta and left renal artery, was made by clinical and imaging data. The medications were modified as follows: amlodipine 5 mg, acetyl salicylic acid 100 mg 1*1, methotrexate 12, 5 mg 1/week, and pedinol 32 mg 1*1, and the patient was discharged. After six months, the aortography repeated and the stenotic lesion was still present (see Figure 3). Therefore, balloon dilatation and stenting were applied to the stenotic region. At the control angiography, after the procedure, there was 50% residual stenosis (see Figure 4). The patient is still being followed at our nephrology department.

DISCUSSION

Hypertension due to renal artery stenosis is especially seen in children under age of 18 and comprises 10% of etiologic causes of hypertension among that specific age group. Despite the fact that the most common cause of renal artery stenosis, is fibromuscular dysplasia, there are many others like Takayasu arteritis, neurofibromatosis, and renal transplantation. In India, Takayasu arteritis is the most common cause of renovascular hypertension in children. Renovascular hypertension is the most serious complication of Takayasu arteritis at the pediatric age group.^[1,2]

The etiology of Takayasu arteritis is still not known. Some infections and genetic and immunologic anomalies may play a role in the pathogenesis of the disease.^[3] In a recent study, Eichorn et al.^[4] showed that only anti-endothelial cell antibody levels were increased in 18 subjects out of 19, and no increased levels were detected among anti-neutrophil cytoplasmic antibody (ANCA), ANA, anti-DNA, extractable nuclear antibody (ENA), anti-Ro antibody, anti-cardiolipin antibody, and circulating immune complexes. In our patient, the ANA, anti-DNA, and antiphospholipid antibodies were all negative, but AECA was not studied.

Clinical features vary with the severity of inflammation at the acute phase and the localization and extension of the stenotic segment at the chronic phase.^[5] Initial symptoms are constitutional like fever, malaise, fatigue, sweating, loss of appetite, weight loss, headache, myalgia, arthritis, and commonly accompanying hypertension.^[6]

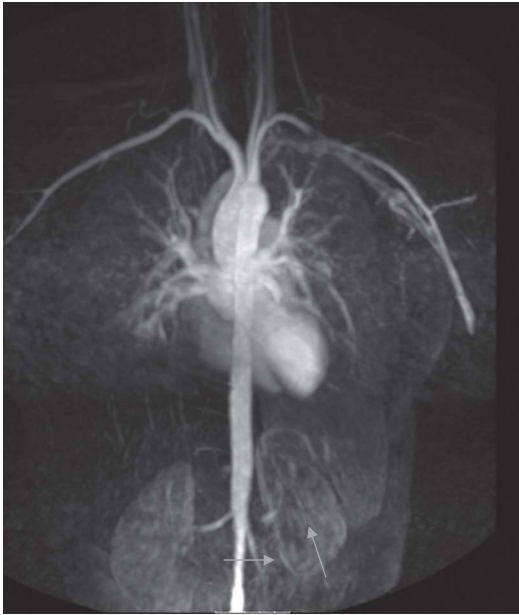


Figure 1. MR angiograph view of the case: severe stenosis at left renal artery and decreased calibration at infrarenal aorta.



Figure 3. Selective angiography of left renal artery: severe stenosis at left renal artery and tortuosity at infrarenal aorta.



Figure 2. The first aortography of the case: stenosis at left renal artery and decreased calibration at infrarenal aorta.

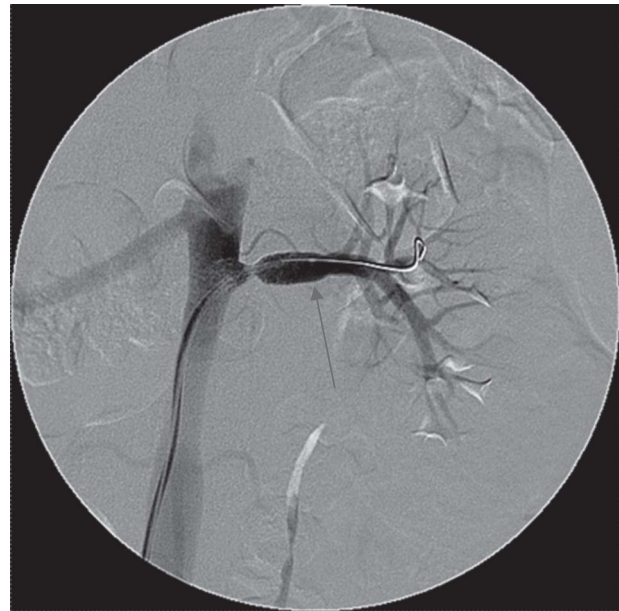


Figure 4. After angioplastic intervention to left renal artery, 50% residual stenosis still remains.

It has been shown that at the acute exacerbation periods of the disease, the acute phase reactants like erythrocyte sedimentation rate and c reactive protein are increasing. In this case, only erythrocyte sedimentation

rate was increased.^[7] However, c-reactive protein level was in normal ranges.

The diagnosis is made by the combination of clinical and angiographical signs. In order to detect which parts or

segments of aorta are involved, aortography has to be done.^[8,9]

In this case, because of accompanying hypokalemia, increased potassium excretion in urine, and metabolic alkalosis, primary hyperaldosteronism or renal artery stenosis was thought to be the possible etiology of hypertension. The ratio of plasma renin activity over serum aldosterone level was 30, but because of a four-fold increased renin activity and mildly increased aldosterone level, the ratio was not meaningful. The computed tomography, which was applied to detect any possible surrenal pathology, revealed concentric wall thickening and other signs of arteritis at abdominal aorta. After that, the diagnosis of Takayasu arteritis was done according to the findings that angiography and aortography revealed.

Takayasu arteritis is classified according to the angiographic findings (Lupi Herrera classification). These are as follows^[10]:

- *Type I.* Aortic archus or main branches of aorta involved;
- *Type II.* Thoracoabdominal aorta involved;
- *Type III.* Diffuse involvement; and
- *Type IV.* Pulmonary involvement.

A retrospective study previously reported by Lupi-Herrera et al.^[10] determined the incidences among the groups as type III 65%, type II 12%, and type I 8% of all cases. In this case, renal and infrarenal segment of abdominal aorta and proximal origin of left renal artery were involved, and the case was included in the type II class.

By effective immunosuppression with novel therapeutic agents and recent successive invasive techniques, long survival periods are reported.^[1] For treatment of the disease, steroids are widely used; however, unresponsive patients need further immunosuppression with other agents like cyclophosphamide and methotrexate.^[1,11] High-dose steroids suppress the acute phase of the disease and prevent arterial stenosis but have weak benefit among anti-fibrosis and anti-thrombosis. Aggressive treatment is a must in renovascular hypertension. Antiplatelet agents like low-dose aspirin and dipyridamole are indicated at the chronic occlusive phase.^[11] Aortic graft replacement and bypass are traditional surgical techniques used in the treatment of the disease. However, several other invasive procedures became available in the treatment, like percutaneous transluminal balloon angioplasty and endovascular stenting.^[12] In this case, because of the severe renal artery stenosis and successful blood pressure control with only one medication, methotrexate and prednisolone was begun as immunosuppressive treatment, and balloon angioplasty and stenting were performed for the stenosis.

The first percutaneous transluminal angioplasty in Takayasu arteritis with renal involvement was performed by Saddekni et al.^[13] in 1980. After that, it was seen as an alternative treatment to surgical approach and performed successfully several times. The response was especially better with the pediatric patients.^[14,15]

We may conclude that Takayasu arteritis with renal artery involvement must be kept in mind as an etiologic factor for secondary hypertension, even if there is no blood pressure difference between extremities. We present a case that had hypertension, hypokalemia, and metabolic alkalosis where the etiology was type 2 Takayasu arteritis, affecting only renal arteries.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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