

Renovascular Hypertension Due to Bilateral Renal Artery Stenosis and Surgical Treatment: A Case Report of a 14 Month-Old Infant

BİLATERAL RENAL ARTER DARLIĞINA BAĞLI RENOVASKÜLER HİPERTANSİYON VE CERRAHİ TEDAVİSİ: 14 AYLIK SÜT ÇOCUĞU OLGU SUNUMU

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Abstract

Fibromuscular dysplasia (FMD) has the first place among the etiologic factors of renovascular hypertension in children. Malignant hypertension unresponsive to medical treatment requires surgical intervention. However, young age and bilateral lesion may hold the indication for surgery as reflected by the small number of such reports.

Hereby, we presented the long-term results of both medical and surgical therapies of a case with renovascular hypertension due to FMD in infancy.

Key Words: Renovascular hypertension, infancy, fibromuscular dysplasia, renal artery stenosis

Özet

Çocukluk çağında renovasküler hipertansiyonun etiolojisinde fibromusküler displazi ilk sırada yer almaktadır. Renovasküler hipertansiyonda tıbbi tedaviye yanıt vermeyen ağır kan basıncı yüksekliklerinde cerrahi tedavi gerekebilir. Bununla birlikte litertürde küçük yaşta iki taraflı lezyonu bulunan ve cerrahi yaklaşım gerektiren çok az olgu bildirilmiştir.

Bu çalışmada, fibromusküler displaziye bağlı renovasküler hipertansiyonlu bir olgunun uzun dönem tıbbi ve cerrahi tedavileri sunulmuştur.

Anahtar Kelimeler: Renovasküler hipertansiyon, çocukluk çağı, fibromusküler displazi, renal arter darlığı

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The most important cause of renovascular hypertension is renal artery stenosis (RAS). Although the incidence of RAS in childhood is reported as 6-13%, the real incidence of RAS is not known exactly.^{1,2} Lawson et al reported 16 children with FMD out of 21 children whom they investigated for renovascular hypertension.³

Fibromuscular dysplasia (FMD) is the common leading cause of renovascular hypertension in children.¹ Malignant hypertension unresponsive to medical treatment may require surgical interven-

tion. However, young age and bilateral lesion may hold the indication for surgery as reflected by the small number of such reports. Hereby, we present the longterm results of both medical and surgical therapies of a case with renovascular hypertension due to FMD in infancy.

Case Report

A 14-month-old female infant was admitted to the local emergency department with headache, uneasiness, vomiting and nausea. Her urine output had decreased from 7 to 3 wet diapers per day. On physical examination; her weight was 25-50th percentile and height 75-90th percentile. Her cardiovascular examination was normal except for severe hypertension: Left arm 152/122 mmHg; right arm 147/119 mmHg, left leg 200/140 mmHg; right leg 170/135 mmHg. Grade I hypertensive retinopathy

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was observed in fundoscopic examination. All other systems were normal except central facial paralysis and epigastric bruit. She had unremarkable prenatal, birth and family histories.

On admission, blood analysis revealed: white blood cell count $22800/\text{mm}^3$, hemoglobin 11 g/dL, blood urea nitrogen 40 mg/dL, serum creatinine 1.1 mg/dL, sodium 130 mEq/L, potassium 5.3 mEq/L, vanillylmandelic acid (VMA), aldosterone, ACTH, cortisol and 17-OH progesterone levels were normal. Antinuclear antibodies, anti-dsDNA, antineutrophil cytoplasmic antibodies, erythrocyte sedimentation rate, IgA and complements factors C3 and C4 were within normal limits or negative. Urinalysis revealed 2+ protein, 8-10 red blood cells, pH: 7, specific gravity 1.015, trace glucose, and negative ketones. Urine electrolytes were normal. A spot urine protein/creatinine ratio was elevated at 42 mg/mmol (normal < 20) and creatinine clearance was 27 ml/min per 1.73 m^2 . At this point, the patient was thought to have in hypertensive crisis. Her fluid limited to insensible losses plus 1 : 1 replacement of her urine output, and she was placed on a low-sodium diet. Antihypertensive therapy was started with captopril perorally, with no effect on blood pressure. Application of other classes of antihypertensive drugs, including hydralazine, nifedipine and furosemid, had no effect on hypertension. Doppler ultrasonography of the kidneys was performed. The left kidney was 7.6 cm the right kidney 6.8 cm. in length. There were no masses or hydronephrosis. Doppler flow studies of both renal arteries and veins were normal; however, the Doppler waveform pattern of the interlobar vasculature could not be assessed adequately due to patient motion. MAG 3 (mercaptoacetyl-triglycine) scan showed that there was bilateral renal hypofunction significantly in the left kidney and no significant excretion. At this point, captopril therapy was discontinued due to highly likelihood of bilaterally renal artery stenosis. A renal angiogram was initiated but the catheter could not be passed to distal segments because of bilaterally 95% constrictive stenosis and angioplasty was failed. Venous renin sampling was taken during the angiogram. The renin levels of the renal veins were higher than the reference range. Instead of this an

aortogram was performed (Figure 1, 2). An echocardiogram demonstrated mild mitral valve insufficiency and left ventricular hypertrophy. Cranial MR and angiography were normal. Due to lack of angioplasty, surgical management was performed as the appropriate choice of therapy. She underwent bilateral PTFE aorta-renal by-pass procedure. Within 24 h of the procedure, the patient was normotensive. Conservative medical therapy and digitalization were also arranged.

Biopsy of the left kidney was performed during the surgery. Histologic examination of the left renal artery showed the thickening with fibromuscular dysplasia.

The patient was discharged without any anti-hypertensive medications 6 months after the surgery. The urine protein/creatinine ratio decreased

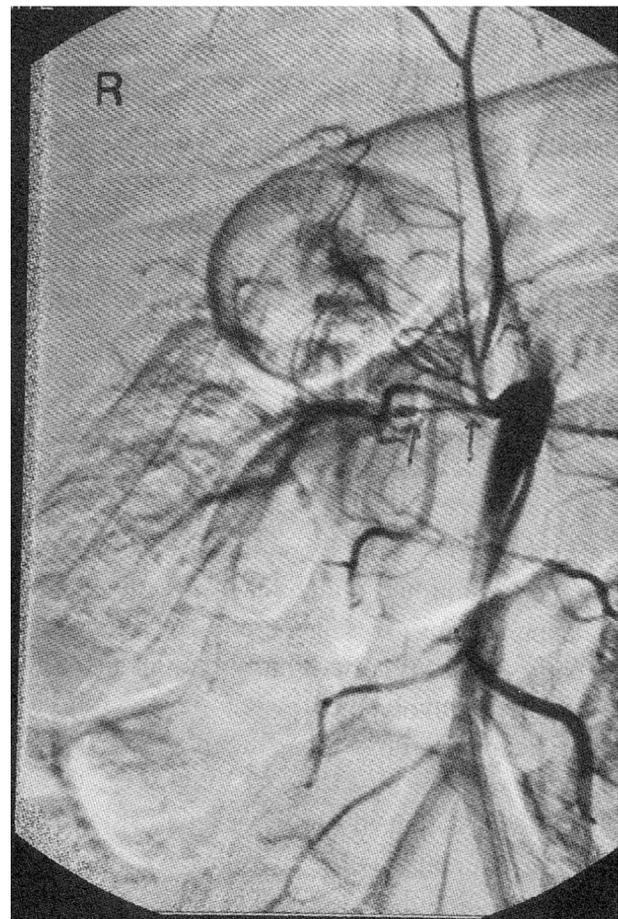


Figure 1. Aortogram showing right renal artery stenosis (Same patient).

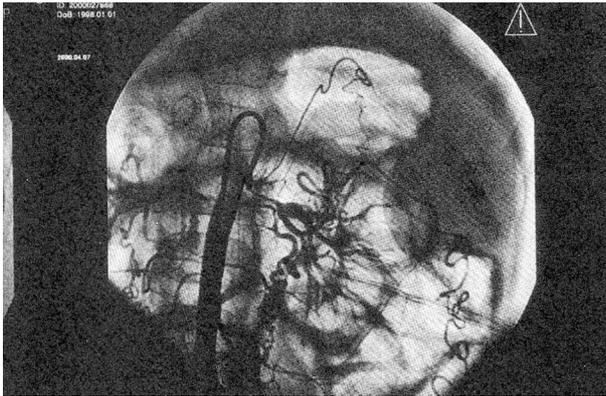


Figure 2. Aortogram showing left renal artery stenosis (Same patient).

to normal limits. All serum electrolytes, urea and creatinine and creatinine clearance were normal.

Discussion

Children with FMD and renovascular hypertension may be asymptomatic or may experience headache, vomiting, nausea, convulsions and also congestive heart failure or death.⁴⁻⁶ Our case had headache elevated blood pressure, vomiting-nausea and congestive heart failure were observed in the clinical course. It is evident that FMD is seen more commonly at the distal half of the renal artery. The “string of beads appearance” of medial dysplasia is common characteristic radiologic finding of FMD seen in adult females.⁷ This suggests that it is not a common finding of FMD in children. Therefore the absence of this finding also does not exclude FMD in our case.

In the 1960's, in the condition that renovascular hypertension had unilateral involvement, surgical nephrectomy was generally recommended.⁸ Recently, it was reported that the clinical findings of unilateral involvement were mostly mild and might be misleading, therefore bilateral involvement should be considered.^{7,8} Lawson et al have pointed out the need for revascularization especially in patients where angioplasty could not be performed.³ Small sizes of the renal vessels and the postoperative complications (re-stenosis, thrombosis and aneurismal dilatations) have contributed to the failure of such surgical treatment.^{3,6,8} In this

case, surgical aortarenal by-pass procedure was performed due to widely bilateral involvement as well as inappropriate vascular diameters.

Currently, some diseases and syndromes with diffuse vascular involvements have been reported in the etiopathogenesis of renovascular hypertension.⁹ One of these is Moyamoya disease, characterized by FMD of internal carotid and proximal cerebral arteries causing progressive obstruction and acute hemiplegia. The appearance of “cigarette smoke” may be observed in angiography that is pathognomonic for the disease.⁹ Moreover, Williams-Beuren syndrome was characterized by stenosis of large and small vessels due to elastin arthropathy and Takayasu arteritis, characterized by panarteritis involving aorta, large vessels and also proximal segments of renal arteries are the other diseases in the etiology.^{10,11} Vascular neurofibromatosis should also be considered in the differential diagnosis. It shows autosomal dominant transition and may be the cause of RAS due to renal artery compression by fibromas or intimal proliferation of renal artery.¹² Poliarteritis nodosa, the inflammatory disease of middle-sized or small vessels with clinically and angiographically typical findings, should be considered in the etiology of renovascular hypertension in children.¹³

In this case, normal cranial MR and angiography, no transient ischemic attack in hospitalization and follow-up period excluded Moyamoya disease from the possible diagnosis. The determination of no “café-au-lait spots”, negative familial history and no observation of hamarhoma in iris (lisch nodule) eliminated the diagnosis of neurofibromatosis. Takayasu arteritis is the segmental panarteritis of aorta and large vessels. Arterial uselessness of the affected extremity is diagnostic. There is no clinical or radiological evidence that supports the diagnosis of this disease in our case. William's syndrome is characterized by growth failure and typical face, supraaortic stenosis of aorta. The normal percentiles of the present case and nonspecific echocardiographic findings except for secondary left ventricular hypertrophy lead to elimination of William's syndrome. Poliarteritis nodosa was also excluded as no weakness, myalgia, mono-

polyneuropathy, and typical aneurysms in angiographic investigation was seen in this case.

In conclusion; renovascular hypertension is relatively common among juveniles but also may be seen in childhood and infancy. FMD and RAS are often accompanying pathological entities. The pathologic findings supported FMD in this case. It is recommended that angioplasty and surgery are the first choices of therapy in these patients. The goal of the therapy in patients with renovascular hypertension should be the prevention of end organ involvement. Our case was also very interesting in this respect even though the patient had severe renovascular hypertension with end organ damage she proceeded satisfactorily and had regulated blood pressure without additional drug therapy during the six month follow-up period. Finally; successful surgical management of renovascular hypertension is achievable in infancy with correct indications and proper technique.

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