Ege Journal of Medicine / *Ege Tıp Dergisi* 2017;56(1):37-39

A case of angiomyolipoma associated with severe hypertension

Şiddetli hipertansiyon ile birlikte olan anjiomiyolipom olgusu

Seçil Conkar Betül Sözeri Sevgi Mir

Ege University Faculty of Medicine, Department of Pediatric Nephrology, İzmir, Turkey

Abstract

Hypertension incidence in adolescent age has been increasing recently. One of the causes of this increase is the increase in obesity incidence. The main cause of hypertension in adolescent age group is primary essential hypertension. However other rare causes of hypertension should be discriminated prior to diagnosing primary essential hypertension. Renal masses are benign or malignant formations that are not commonly seen in childhood. Renal masses are a rare cause of hypertension in childhood. Angiomyolipoma (AML) is one of the rare renal mesenchymal tumors in childhood. AML usually coexists with tuberous sclerosis (TS) and TS cases are clinically in the foreground. Here we present a case of AML case emerging as malignant HT without TS. In adolescence, it sholud be kept in mind that renal AML may cause malignant hypertension.

Keywords: Angiomyolipoma, renovascular hypertension in adolescence, renal neoplasms.

Öz

Son zamanlarda adelosan yaşta hipertansiyon görülme sıklığı artmaktadır. Bu artış obezite sıklığında artış ile paralellik göstermektedir. Adelosan yaş grubunda hipertansiyonun ana nedeni esansiyel hipertansiyondur. Ancak hipertansiyonun diğer nadir nedenlerinin esansiyel hipertansiyon teşhisi öncesinde ayırıcı tanısı yapılmalıdır. Böbrek kitleleri çocuklarda sık görülmeyen iyi veya kötü huylu oluşumlardır. Böbrek kitleleri çocukluk çağında hipertansiyonun nadir nedenidir. Anjiyomiyolipom (AML), çocukluk çağında nadir böbrek mezenkimal tümörlerinden biridir. AML genellikle tubero skleroz (TS) ile bir arada olup TS klinik olarak ön plandadır. Burada TS'nin eşlik etmediği malign HT olarak ortaya çıkan bir AML olgusunu sunduk. Adelosan dönemde malign hipertansiyon nedeni olarak böbrek anjiyomiyolipomu olabileceği unutulmamalıdır.

Anahtar Sözcükler: *Anjiyomiyolipom, adölesan dönemdeki renovasküler hipertansiyon, böbrek tümörleri.*

Introduction

Primary hypertension prevalence is increasing owing to recent global epidemic childhood obesity (1). Renal masses are rare cause of hypertension in childhood. Tumors may cause compression of the renal vasculature, with renin increase, in which radiotherapy and postoperative fibrosis may result in renin–angiotensin system (RAS) or urethral stricture resulting hypertension (2). Angiomyolipomas (AML) is rare benign mesenchymal tumors composed of various amounts of fat tissue, smooth muscle fibres and thick wall blood vessels. The most common site for location is the kidneys (3,4). AML can rarely cause hypertension.

Corresponding Author: Seçil Conkar

Ege University Faculty of Medicine, Department of Pediatric Nephrology, İzmir, Turkey

Received: 12.11.2015 Accepted: 06.01.2016

Tuberosclerosis complex (TSC) is an inherited multisystem neurocutaneous disease with multiple benign hamartomas of the brain, eyes, heart, lung, liver, kidney, and skin. TSC commonly associated with renal angiomyolipomas. Many patients with TSC have epilepsy, cognitive deficits (5). Our case diagnosed having only AML without TSC. The physical evaluation did not reveal any clinical features of TSC.

Case Report

A 17-year-old male patient was admitted to Pediatric Emergency Department with blurred vision. Anthropomorphic measurements were as following: Weight 45 kg (25-50 p), height 160 cm (25-50 p) and body mass index (BMI) was 17.5 kg/m^2 (25-50 p). Blood pressure was measured at resting sitting position was 200/120 mmHg (stage 2). Retinal examination revealed bilateral papilla edema. Other systemic examinations were remarkably normal. There was no history of incipient drug use that could lead to hypertension.

Tests were performed towards hypertension causes and target organ injury. Renal function tests, blood gas analysis, electrolytes; urinary findings were detected to be normal. A significant obliteration was not detected in renal artery on renal doppler ultrasonography. His fundus examination findings were consistent with grade 2 hypertensive retinopathy. Echocardiographic increase was detected in left ventricle wall thickness as 40.9 g/m^2. Microalbuminuria level was found as 600 µg/min. Hypertensive end organ damage were at ocular and renal and cardiac system. Plasma renin (1.1 ng/mL/h) and aldosteron (120 pg/mL) levels were in normal levels. Endocrine causes of hypertension were excluded. Diurnal ACTH, cortisol, thyroid function tests, VMA were in normal range. Thorax computed tomography was performed in order to obtain a more detailed imaging of thoracic aorta, as gradient increase was detected at juxtaductal region on echocardiography. There was 4 mm of nonspecific nodule in anterior mediastinum and was interpreted as thymic hyperplasia. After consultation by an oncologist, thoracoscopic biopsy was performed. Cranial MRG and abdominopelvic dynamic tomography was performed for the causes of secondary hypertension. Kidneys and renal arteries were detected to be normal. On dimercaptosuccinic acid (DMSA) scan, bilateral activity uptake of kidneys was lower than normal and non-homogenous. Right kidney cortex was significantly thinned and had contour defects with hypoactive areas. Cortical functions were found as 64% in the left kidney and 36% in the right kidney. Vesicoureteral reflux was not detected at voiding cystourethrography.

Hypertension treatment was started with angiotensin converting enzyme blocker (ramipril, 6 mg/m^2/day) and angiotensin receptor blocker (losartan, 0.7 mg/kg/day). Ocular, cardiac and renal examinations were performed for target organ injury at every 6 months during clinical follow up. While micro albuminuria was regressed to 60 µg/min, left ventricle mass index was not decreased Hypertensive retinopathy was not progressed.

For three years his blood pressure was normal. Afterwards he admitted with stage 2 hypertension (170/100 mmHg), he was hospitalized. In abdominal computed tomography a solid nodular formation measuring 1.6x0.8 cm was detected in the left kidney. It was considered to be a benign soft tissue tumor. At MRG, a 23 mm angiomyolipoma in diameter was revealed at left kidney (Figure-1). Angiomyolipoma was small in size and blood pressures are under control with combination therapy of ACEI and ARB. We decided to follow up the patient with diagnosis of angiomyolipoma-related hypertension after discussing with oncologists.

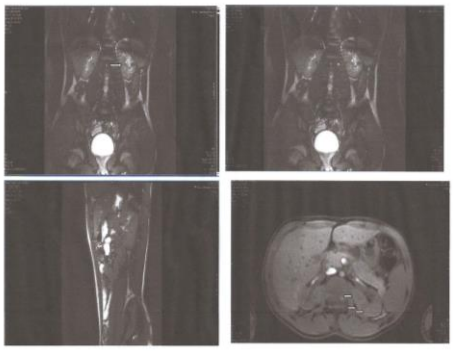

Figure-1. A magnetic resonance scan shows a 23-mm mass lesion (angiomyolipoma) at left kidney.

Written informed consent was obtained from the patient's legal custodian for publication the individual medical reports.

Discussion

Secondary hypertension is more common in early childhood (6). The prevalence of angiomyolipomas increases with age. While isolated renal AML are found in about half of cases, other half is together with TBC (3). Our case was diagnosed as isolated angiomyolipoma not accompanied by TBC.

Angiomyolipomas are rarely the cause of hypertension but may occasionally result in compression of renal tissue or damage from hemorrhage. Renal AMLs are usually benign tumors. Tchaprassian et al. (7) described three isolated AML cases without TSC aged 11, 13, 15 years. Malignant hypertension was reported only in a 15-month-old child in the literature (8).

AML enlarges in time and lead to end stage renal disease. Dialysis treatment is required especially in patients with bilateral renal involvement accompanied by TBC (9). Although most AMLs 4 cm and above in diameter are asymptomatic (82-94%) , there is a risk for spontaneous hemorrhage in 50-60% (10). In our case AML was under 4 cm in diameter. Angiomyolipoma-related hypertension was under control with medications. Hemorrhage and malignity were not developed. AML should also be kept in mind in secondary hypertension cases.

References

1. Lever AF, Harrap SB. Essential hypertension: a disorder of growth with origins in childhood? J Hypertens 1992;10(2):101-20.
2. Geller E, Smergel EM, Lowry PA Renal neoplasms of childhood. Radiol Clin North Am 1997;35(6):1391-413.
3. Tamboli P, Ro JY, Amin MB, Ligato S, Ayala AG Benign tumors and tumor-like lesions of the adult kidney. Part II: benign mesenchymal and mixed neoplasms, and tumor-like lesions. Adv Anat Pathol 2000;7(1):47-66.
4. Crino PB, Nathanson KL, Henske EP The tuberous sclerosis complex. N Engl J Med 2006;355(13):1345-56.
5. O'Hagan AR, Ellsworth R, Secic M, Rothner AD, Brouhard BH Renal manifestations of tuberous sclerosis complex. Clin Pediatr 1996;35(10):483-9.

6. Rimon U, Duvdevani M, Garniek A, et al. Ethanol and polyvinyl alcohol mixture for transcatheter embolization of renal angiomyolipoma. AJR Am J Roentgenol 2006;187(3):762-8.
7. Tchaprassian Z, Mognato G, Paradias G, D'Amore ES, Tregnaghi A, Cecchetto G Renal angiomyolipoma in children: Diagnostic difficulty in 3 patients. J Urol 1998;159(5):1654-6.
8. Springer AM, Saxena AK, Willital GH. Angiomyolipoma with hypertension mimicking a malignant renal tumor. Pediatr Surg Int 2002;18(5-6):526-8.
9. John J, Bissler J, Kingswood C. Renal angiomyolipomata. Kidney International 2004;66(3):924-34.
10. Halpenny D, Snow A, McNeill G, Torreggiani WC. The radiological diagnosis and treatment of renal angiomyolipoma current status. Clinical Radiology 2010;65(2):99-108.