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Case Report

SHOULD AN ACCESSORY RENAL ARTERY BE CONSIDERED AS A CAUSE OF HYPERTENSION IN ADOLESCENTS: A CASE REPORT

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SUMMARY – The role of an accessory renal artery in the pathogenesis of hypertension is still controversial. In this report, we describe a previously healthy 16-year-old girl with an accessory right renal artery who presented with hypertensive urgency (no progressive target organ dysfunction). Initial workup other than hypercholesterolemia and mild proteinuria was normal with no signs of other target organ damage. Further evaluation was aimed at determining the possible cause of secondary hypertension. High normal plasma renin with elevated plasma aldosterone led to a suspicion of renovascular hypertension. Magnetic resonance angiography and later computed tomography angiography showed two non-stenotic right renal arteries. Another diagnostic workup was normal. A satisfactory blood pressure control was eventually achieved with combination therapy including angiotensin-converting enzyme inhibitor. We conclude that although non-stenotic, an accessory renal artery should be considered as a possible cause of renovascular hypertension in children and adolescents even in the absence of hyperreninemia.

Key words: *Accessory renal artery; Renovascular hypertension; Children; Hypertensive crisis*

Introduction

In young adolescents, primary hypertension is more common compared to secondary hypertension[1]. Nevertheless, in the case of hypertensive crisis, investigation for a secondary cause should always be done. Renovascular hypertension is one of the potential causes of secondary hypertension but whether an accessory renal artery can be considered as a cause of hypertension is still a matter of debate.

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Case Report

A previously healthy 16-year-old girl presented at the Sestre milosrdnice University Hospital Center Emergency Room because of the sudden onset of nausea, severe headache and elevated blood pressure (BP) of 220/120 mm Hg on both arms. There was no record of elevated BP in her medical history. Family history revealed that both her parents were hypertensive with hypercholesterolemia while her grandfather had renal artery stenosis and was treated on hemodialysis. Her body mass index was normal (21 kg/m^2, 57th percentile) and physical examination unremarkable. A written consent was obtained from the patient and her parent.

Laboratory tests at presentation were normal including normal renal function (estimated glomerular

filtration 113 mL/min/1.73 m^2), normal serum electrolytes, uric acid and fasting plasma glucose. Urinalysis with sediment was also normal (pH 6, specific gravity 1.010 kg/L, protein, glucose and nitrate negative with 2 red blood cells, no leukocytes, no cellular casts, crystals and bacteria in sediment). Urine toxicology screen (tested for amphetamine, methamphetamine and 3,4-methylenedioxymethamphetamine (MDMA), barbiturates, benzodiazepines, cocaine, marijuana, methadone, morphine, tricyclic antidepressants, tramadol, fentanyl) was negative. Electrocardiogram (ECG) showed early repolarization pattern with intraventricular conduction disturbances while echocardiography was normal. Fundoscopy and computed tomography (CT) of the brain were also normal. Since there was no progressive target organ dysfunction, a diagnosis of hypertensive urgency was established.

Additional laboratory tests revealed significant hypercholesterolemia of 8.2 mmol/L with elevated LDL cholesterol of 5.4 mmol/L, which was considered as familial hypercholesterolemia according to her family history. Despite normal urinalysis, 24-hour urine revealed mild proteinuria 9.2 mg/m^2/h while her 24-hour sodium excretion was low (0.34 mmol/kg/24 h). Ambulatory blood pressure monitoring (ABPM) confirmed severe ambulatory hypertension. Initially she was treated with amlodipine achieving high normal BP values and gradual regression of symptoms.

Further investigation was focused on finding the etiology of hypertension. Urine metanephrines, urinary free cortisol, plasma cortisol, adrenocorticotropic hormone (ACTH) and thyroid function tests were within the normal limits. Her plasma renin was high normal 35.9 ng/L (reference range 5.1-38.7 ng/L) with elevated plasma aldosterone 1687 pmol/L (reference range 134-751 pmol/L). The plasma aldosterone (pmol/L)/plasma renin ratio (converted to mU/L) of 29.8 was within the normal limits. She had normal potassium and elevated bicarbonates. On kidney and renal artery ultrasonography with Doppler done by two independent examiners, there was no sign of renal artery stenosis (renal artery resistive index left 0.71 and right 0.8).

Magnetic resonance imaging (MRI) showed no pathology of the adrenal glands, therefore, along with the results of hormonal evaluation, the diagnosis of primary hyperaldosteronism was excluded. Further on, considering the possibility of renovascular hypertension, renal magnetic resonance angiography (MRA) was done with the finding of two non-stenotic right renal arteries arising from the normal abdominal aorta and solitary non-stenotic renal artery left (Fig. 1). The diameter of a single left renal artery was larger compared to the diameter of either right renal artery (left renal artery 6.0 mm *vs.* 4.1 mm and 3.9 mm for the upper and lower right renal artery). There was no significant difference in length (33.1 mm left *vs.* 38.2 mm and 42.3 mm for upper and lower right renal arteries). Other possible causes of secondary hyperaldosteronism (e.g., renal hypoperfusion, heart failure, edematous disorders) were ruled out. Her psychological assessment indicated neuroticism, low tolerance for frustration and low self-esteem while her reaction to stressful situations reached the level of panic attacks. It was concluded that some of her symptoms could be of psychogenic origin and she started group psychotherapy.

During the follow-up, mild proteinuria with albuminuria A2 persisted. The workup for glomerular disease was negative and a diagnosis of hypertensive nephropathy was considered. ABPM along with home BP measurements in the next two years showed significant visit-to-visit BP variability with 'white coat effect' on office BP measurements.

Since amlodipine failed to produce satisfactory BP control, and dietary measures had only minor effect on lipid profile, after exclusion of renal artery stenosis she was eventually switched to enalapril/hydrochlorothiazide and atorvastatin was introduced.

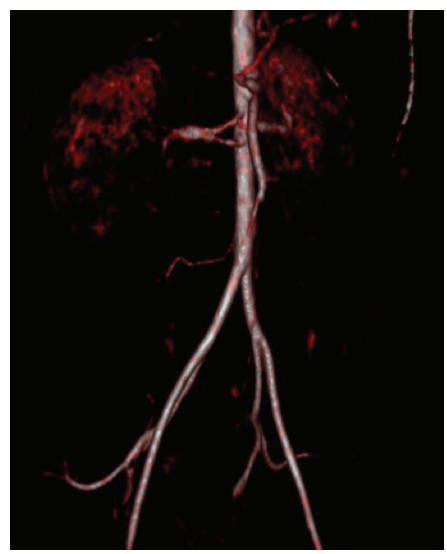

Fig. 1. Contrast-enhanced magnetic resonance angiography in a 16-year-old girl (volume rendering technique): the right kidney is supplied with two renal arteries.

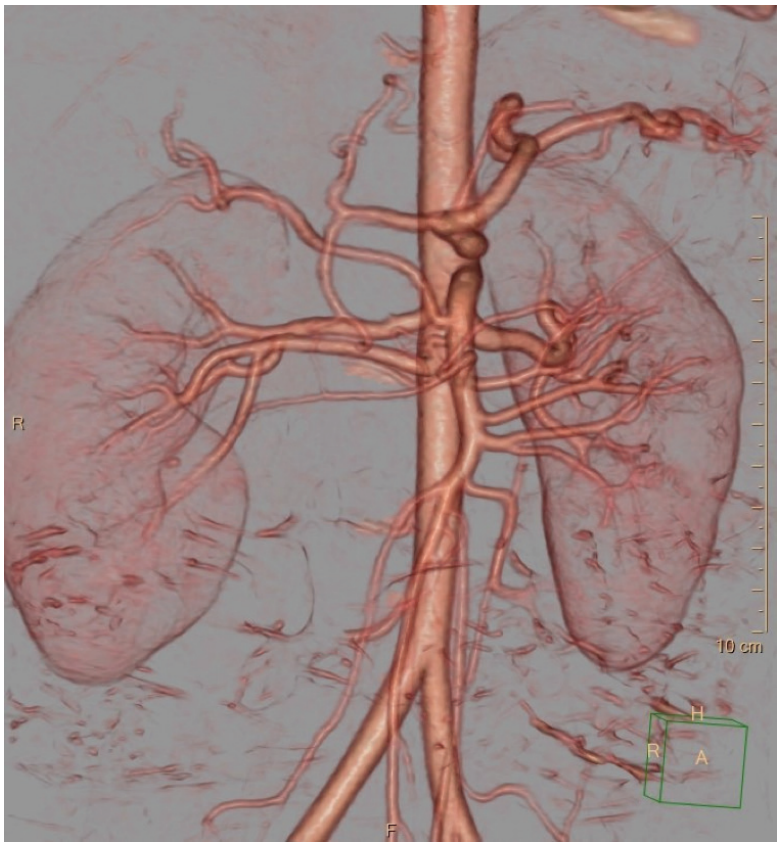

Fig. 2. The same patient at the age of 18 years. Computed tomography angiography (volume rendering technique) using 80-kVp protocol and moderate-concentration contrast medium: a solitary renal artery on the left and two symmetric renal arteries on the right.

At the age of 18, in order to provide better spatial resolution compared to MRA, CT angiography (CTA) was performed. It confirmed two symmetric right renal arteries with single left renal artery. The length of renal arteries according to CTA matched those on MRA. The diameter of left renal artery was again confirmed larger (5.92 mm) compared to the diameter of either upper (4.08-4.25 mm) or lower (3.8-3.9 mm) right renal artery. There was no sign of stenosis on either of the arteries. The morphology of abdominal aorta was also normal (Fig. 2).

Eventually, satisfactory BP control was achieved with the perindopril/indapamide/amlodipine combination therapy. Proteinuria gradually normalized and lipid profile improved (total cholesterol 5.1 mmol/L, LDL cholesterol 2.9 mmol/L). On recent ABPM, her average 24-hour BP was 127/76 mm Hg with 'white coat effect' on the office BP measurements. ECG was normal. There were no signs of target organ damage. Repeated plasma renin was elevated 105.9 ng/L and plasma aldosterone normal 787 pmol/L (reference range 103-1196) while on angiotensin-converting enzyme (ACE) inhibitor therapy.

Discussion

The prevalence of an accessory renal artery is reported to be around 25% in general population with a higher prevalence among hypertensives of up to 80%[2,3]. The role of an accessory renal artery in the pathogenesis of hypertension is still controversial. Such an association has been proposed by numerous studies, starting with Marshall *et al.* in 1951[4-7], whereas a few other studies did not confirm a correlation between hypertension and accessory renal arteries[8,9].

We report a case of a previously healthy adolescent girl who presented with hypertensive urgency (no progressive target organ dysfunction) in whom no secondary cause of hypertension other than an accessory

right renal artery was determined. Although we could not demonstrate a hyperreninemic state, a high normal plasma renin with elevated plasma aldosterone and no adrenal pathology on MRI led to a suspicion of renovascular hypertension.

Some authors confirmed bilateral multiple renal arteries with significant stenosis of the one extra-renal artery with normal plasma aldosterone and plasma renin[10]. Therefore, it can be concluded that normal plasma renin and aldosterone do not necessarily exclude the diagnosis of renovascular hypertension. Interestingly, some authors report on spontaneously resolving hyperreninemic hypertension caused by accessory renal artery stenosis in a child with contralateral localization of hyperreninemia[11].

On the other hand, the study by Glodny *et al*. demonstrated that patients with this relatively common vascular anatomic anomaly had a significantly higher blood pressure than patients with normal renal anatomy[5]. They also identified higher peripheral plasma renin activity in hypertensive patients with aberrant renal artery(ies) compared to a matched control group of hypertensive patients without aberrant renal arteries.

While repeat renal artery Doppler ultrasounds were within the normal limits for age in our patient, MRA showed two right renal arteries with no conclusive arteriographic evidence for stenosis in either the accessory or main renal artery although the diameter of a single left renal artery was larger than the diameter of either of the right renal arteries. These measurements were later confirmed on CTA. The smaller diameter of multiple arteries compared to the diameter of a single renal artery that caused impaired renal perfusion was also confirmed in other studies, and therefore, the diagnosis of renovascular hypertension remained[12]. However, primary hypertension in our patient could not be completely ruled out due to a strong family history of hypertension. Since hypertension urgency is unlikely without some trigger factor, while considering her psychological profile, it could be hypothesized that certain stressful event resulted in sudden elevation of blood pressure although it was not confirmed in her medical history.

Renovascular hypertension often presents a therapeutic challenge and usually requires a combination therapy. Patients with accessory renal artery(ies) might have less pronounced BP reduction achieved after renal denervation than patients with bilateral single renal artery, indicating the possible link between accessory renal artery and hypertension[13]. On the other hand, some reports indicate that accessory renal arteries do not influence the response to renal denervation demonstrating that the presence of small accessory arteries is not relevant mechanism in resistant hypertension pathogenesis[14].

In our patient, BP control was eventually achieved with the fixed-dose triple combination treatment including diuretic followed by resolution of proteinuria and no target organ damage on follow-up.

Conclusion

We conclude that accessory renal artery should be considered as a possible cause of renovascular hypertension in children and adolescents even in the absence of hyperreninemia or stenosis. Since this is not widely accepted, complete workup focused on finding a secondary cause of hypertension should be done as well. As in our case, hypertensive urgency could be the initial presentation of an accessory renal artery.

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Sažetak

MOGU LI VIŠESTRUKE BUBREŽNE ARTERIJE BITI UZROK HIPERTENZIJE U ADOLESCENATA: PRIKAZ SLUČAJA

B. Valent Morić, T. Krpan i *J. Josipović*

Uloga akcesornih bubrežnih arterija u patogenezi hipertenzije je kontroverzna. U ovom prikazu opisujemo slučaj do tada zdrave 16-godišnje djevojke s akcesornom desnom bubrežnom arterijom kod koje je prva manifestacija bolesti bila hipertenzivna urgencija. Nalazi osnovnih laboratorijskih pretraga pri inicijalnoj obradi bili su uredni osim prisutne hiperkolesterolemije i blage proteinurije. Nije bilo drugih znakova oštećenja ciljnih organa u sklopu hipertenzije. Daljnja obrada rađena je s ciljem otkrivanja potencijalnog uzroka sekundarne hipertenzije. Zbog visoko normalnog renina u plazmi uz povišen plazmatski aldosteron postavljena je sumnja na renovaskularnu hipertenziju. Na angiografiji magnetnom rezonancom opisane su dvije bubrežne arterije desno, što je kasnije potvrđeno kompjutoriziranom tomografijom angiografijom bez dokaza za stenozu bubrežnih arterija. Nalazi ostalih dijagnostičkih pretraga bili su uredni. Zadovoljavajuća regulacija arterijskoga tlaka postignuta je nakon uvođenja kombinirane antihipertenzivne terapije uključujući inhibitor angiotenzin konvertirajućeg enzima. Vjerujemo stoga da akcesorne bubrežne arterije, čak i bez dokazane stenoze, treba smatrati mogućim uzrocima renovaskularne hipertenzije u djece i adolescenata, čak i u odsustvu povišenih vrijednosti renina.

Ključne riječi: *Akcesorna bubrežna arterija; Renovaskularna hipertenzija; Djeca; Hipertenzivna kriza*