CASE REPORT

A successful therapy of renovascular hypertension

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Abstract

Authors describe a rare cause, diagnostic difficulties and successful therapy of renovascular hypertension in a 12 year-old girl caused by anular stenosis of the intrarenal arterial branch. Activation of the system renin-angiotensin (RAS) is found in all forms of renovascular hypertension at the beginning. Etiologically, stenosis in childhood is caused mostly by renal artery dysplasia, affecting mostly media, and fibromuscular dysplasia. Fibromuscular dysplasia affects middle and distal third of renal artery in 60 %, more frequently on right, only in 10 % of cases affects segmental branches; one quarter of patients are affected bilaterally. This disease is found predominantly in young women. During clinical course, typical signs include sudden onset of severe and poorly controlled hypertension, renal insufficiency, proteinuria and hypertensive retinopathy. From non-invasive diagnostic approaches, color duplex ultrasound, NMR and CT angiography are important, from invasive ones, digital subtractive angiography and the measurement of plasma rennin activity in renal veins. In therapy, it is percutaneous transluminal renal angioplasty, associated with low mortality and morbidity. The net result of angioplasty is dilation of stenosis, complete restoration of artery lumen and flow and decrease of blood pressure. The best results can be achieved in young patients with fibromuscular dysplasia, more then half can recover completely. Using this method, also our patient has recovered (Tab. 2, Fig. 2, Ref. 7).

Key words: renovascular hypertension, renin-angiotensin system, fibromuscular dysplasia, percutaneous transluminal angioplasty.

Systemic hypertension is found in both early and late childhood. In early childhood, we think more of secondary hypertension (1). In adolescence, we have to keep in mind also beginning hypertension. It is important to treat it, as it belongs to serious risk factors of many cardiovascular diseases like atherosclerosis, ischemic heart disease, stroke or nephrosclerosis in adulthood.

In this case report, we present an unusual hypertension of renovascular origin and its successful therapy.

Case report

A 13-year old girl, complaining on headache, was transmitted to our clinic from a regional hospital. She comes from a large family, has healthy parents a three healthy siblings. The family lives in a 4-room house with central heating in the country and has some pets. In the history, the patient has no problems, her development was normal, was sick only rarely, her results at school were good. Due to sustained headache, her pediatrician has measured her the blood pressure with the result of 160/110 torr. Following a consultation with cardiologist, she was admitted to the intensive care unit at the regional hospital. Her blood pressure was 180/120 torr. After furosemide, magnesium and trimepranol administration, her blood pressure has temporary decreased, but has increased during relatives visits.

During hospitalization, borderline hypernatremia and hypokalemia was found. Doppler flow measurement does not suggest left renal artery stenosis; right renal artery flow was not visualized, but segmental arteries flow was normal. In eye ground examination, retinal branches were markedly filled. When the patient was transmitted from intensive care unit to the children department, repeated blood pressure values up to 185/120 torr.
associated with headache were observed. On the fourth day of hospitalization, the patient was transmitted to our clinic.

In admission, the patient was hypertensive, with blood pressure 180/120 torr, associated with headache, tachypnoe, tachycardia, skin hyperemia and perioral and acral cyanosis during blood collection. Hypokalemia and metabolic alkalosis was found, what suggested the activation of renin-angiotensin-aldosterone. The results of both plasma renin activity (PRA) and aldosterone from periphery were so high during baseline and after stress, that the laboratory has suggested non-standard condition for blood collection. When repeatedly measured, the results were the same, extremely high values (6) (Tab. 1).

Hormonal profile has showed normal levels of suprarenal hormones and acid vanilla-almond. Complex evaluation of urine, renal function and renal arteries flow was within normal range. Cardiologic examination (ECG and echocardiography) has excluded the structural heart defect and myocardial ischemic changes. Examination of the eyes has excluded angiopathy. Ultrasound of the thyroid and suprarenal glands did not show pathological focus or expansion. Patient was subsequently administered norvasc, tensiomin and verospiron therapy and a decrease and stabilization of blood pressure was observed (7).

As we suspected renal arteries stenosis, digital subtractive angiography (DSA) was performed at the department of diagnostic and interventional radiology of NUSCH, and occlusions were found on peripheral parts of intrarenal arterial branches, at middle third of left kidney and distal half of right kidney. In both kidneys, rare small aneurysms up to 1–2 mm were found on arterial branches. We suggested polyarteritis nodosa, but it was not confirmed by laboratory values. We continued collecting blood for plasma renin activity from both renal veins, vena cava inferior and from site below the insertion of renal veins (Tab. 2).

These values have not helped us, so we continued in examinations.

| Tab. 1. Plasma renin activity and aldosterone levels before and after stress. |
|---------------------------------|-----------------|-----------------|
|                                | Actual values   | Reference values |
| PRA                             | baseline        | 2520 increased  | 50–190 ng/100ml |
|                                | after stress    | 14610 increased | 190–600 ng/100ml |
| Aldosterone                     | baseline        | 80.3 increased  | 0.75–16.0 ng/100ml |
|                                | after stress    | 144.0 increased | 3.5–31.0 ng/100ml |

| Tab. 2. Plasma renin activity in both renal veins, vena cava inferior (VCI) above and below the insertion of renal veins. |
|---------------------------------|-----------------|-----------------|
|                                | Actual values   | Reference values |
| PRA                             | Right renal artery | 754 increased | 50–190 ng/100 ml |
|                                | VCI above insertion | 722 increased |
|                                | VCI below insertion | 622 increased |
|                                | Left renal artery | 700 increased |

Fig. 1. Anular stenosis of cranial branch of renal artery.

Fig. 2. Dilatation caused by percutaneous transluminal renal angioplasty.

In time, due to increased and uncontrolled blood pressure, the patient was admitted to a control examination and therapy adjustment. An increased level of total cholesterol, LDL/C (Chol–HDL–TG/2.2) and proteinuria was found. Other laboratory parameters were within normal range. Kidney function was normal. Due to ischemic changes over left precardium echocardiography was performed, showing appropriate functional and metric parameters of left ventricle.

In all phases of exercise test patient denies subjective problems. Baseline values of blood pressure, pulse and breathing before an exercise and after a maximal exercise did not exceed
normal range. We have not found changes on eyeground. Calcium channel blocker (Norvasc) dose was increased and Tensimin was changed to Tritace.

Due to further increase of blood pressure and its unknown origin we decided to repeat digital subtractive angiography of abdominal aorta, both renal arteries and also mesenteric artery, suspected from polyarthritis nodosa. A marked anular stenosis with poststenotic dilatation was found intrarenally, on cranial branch of renal artery before segmental branching (Fig. 1).

Intrarenal aneurysms were not confirmed. During hospitalization we have to increase the dose of ACE inhibitors.

Based on this examination, patient has undergone percutaneous transluminal renal angioplasty (Fig. 2).

At the site of anular stenosis, dilatation was achieved without residual lumen reduction. The balloon had 3 mm in diameter. During intervention, spasms on both branches of main artery as well as on intrarenal peripheral part were observed. One day after intervention, blood pressure began to decrease, doses of antihypertensives have to be decreased and one week after intervention stopped. Patient was stabilized regarding blood pressure and pulse. In the period of blood pressure stabilization, plasma renin activity and aldosteron levels were within normal range. Therapy continues with only small dose of anopyrin.

Discussion

Renovascular hypertension is a chronic increase of systemic blood pressure caused by marked stenosis of renal artery or its branches. Patogenesis of renovascular hypertension is not known well. Regarding patogenesis (i.e. renal artery stenosis) besides renin-angiotensin-aldosteron system also renal production of vasorelaxans (prostaglandins, kalikrein-kinine system, neutral antihypertensive reno-medullar lipids and NO production) takes part. Decreased production of these substances could support the development of arterial hypertension. Etiologically, stenosis in childhood is caused mostly by renal artery dysplasia, affecting mostly media, and by fibromuscular dysplasia. This condition is suggested to be causative in our patient. Rare causes of stenosis are arteritis, atherosclerosis, trombosis, embolia, aorta aneurysm or pseudoaneurysm at site of renal artery insertion with subsequent compression, tumor, renal cyst, etc. Fibromuscular dysplasia affects middle and distal third of renal artery in 60 %, mostly on right; only in 10 % of cases affects segmental ranches, most patients have bilateral changes. This disease affects mostly young women. During clinical course, typical signs include sudden onset of poorly controlled hypertension, like in our patient. However, hypertensive retinopathy, severe proteinuria and increase of plasma creatinine during ACE inhibitors therapy were not observed. It is possible to suggest, that marked stenosis was developed recently. From diagnostic approaches, digital subtractive angiography and measurement of plasma renin activity in both venal veins is important. In therapy, percutaneous transluminal renal angioplasty is used (2, 5).

Since its beginning (Grunzig, 1978), percutaneous transluminal angioplasty of renal artery (PTRA) become an established basic endovascular method for stenosis treatment, representing less invasive alternative of surgical revascularization. One intervention, broadening the possibilities of endovascular correction of stenotic lesion, is stenting – implantation of a stent. Main indication of PTRA in case of stenting insufficiency is hypertension based on fibromuscular dysplasia (FMD) in children. Technical success of isolated PTRA is lower then in adults (60–80 %), because some stenosis resists even high dilution pressure. Technically well performed PTRA has almost 100 % clinical success.

For PTRA, short isolated non-ostial stenosis of main renal artery and its intrarenal branches are ideal. Ostial stenosis is indicated for stenting. The intervention is performed mostly from transfemoral approach, PTA balloon or stent ranges 4–7 mm in diameter in main renal artery lesion, and 3 mm or less in intrarenal branch correction. For insertion of endovascular devices, leading catheter technique is most frequently used. Incidence of serious complications ranges between 3–8 %. Less serious is hematoma in groins, more serious is retroperitoneal bleeding. Mortality 30 days after PTRA is maximally 1 % (3, 4).

Conclusion

Present diagnostic and mainly endovascular therapeutic methods can remove the cause of hypertension and prevent the development of further serious cardiovascular diseases even in childhood. Endovascular interventions (PTRA, stenting) decreasing blood pressure and improving renal function are indicated not only in adults but also in young patients with suggested fibromuscular dysplasia even in intrarenal localization, where the result of endovascular therapy is good predominantly in young women.

References


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