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Severe renal vascular hypertension caused by upper left branch renal artery stenosis complicated with multiple thrombosis and cardiac hypertrophy

Case Report

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Abstract: A severe hypertension was found in a 9 month old boy with signs and symptoms of severe arterial hypertension. An upper left branch renal artery stenosis complicated with multiple thrombosis and cardiac hypertrophy was diagnosed. Surgical heminephrectomy of the upper pole of the left kidney was successful in blood pressure normalization with significant improve of cardiac function and thrombosis recanalization.

Keywords: *Arterial hypertension • Left branch renal artery stenosis • Cardiac hypertrophy • Thrombosis • Children*

1. Introduction

Hypertension is not rare in children and can occur up to 2% of neonates [1]. The broad spectrum of causes includes potential life-threatening renal arterial stenosis [1,2]. In children it is mainly due to fibromuscular dysplasia and stenosis associated with syndromes involving single or multiple small branch vessels [3,4]. Prompt and thorough evaluation with a main focus on kidney disease is the key for appropriate therapy. The outcome of renal stenosis and heart failure (unrelated to indwelling umbilical catheters) with determined unilateral nephrectomy is favorable as seldom lethal outcome was reported [1,5].

2. Case report

After sudden loss of consciousness a 9 month old male child was admitted to Clinical intensive care unit. Except for severe hypertension (200/140 mmHg), elevated body temperature (39°C) and tachycardia, all other clinical examination, including neurological overview proved regular. A successful removal of a polipoid tumor in the terminal ileum was performed two months before, but restlessness, changes in behavior and polyuria persists. Elevated blood pressure during surgical procedure was not observed. Blood count, CRP, serum glucose level, bilirubin, creatinine, electrolyte, acid-base and coagulation tests were within normal limits. Brain EEG was normal.

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Laboratory tests revealed hypercalciuria (Ca/creatinine 3.75 mmol/mmol), initial proteinuria 3.56 g/24h (after successful treatment falls within normal limits) and elevated, urine RAP (63.5 µcg/L/h), serum aldosterone (3144 pmol/L) and urine aldosterone (373 nmol/24h urine).

Standard kidney and abdomen ultrasound examination was initially normal. Doppler kidney ultrasound discovered diverse renal artery RI, between 0.64 and 0.43 a piece with elevated diastolic pressure. Multislice CT renal angiography revealed a smaller size of the left kidney with 2 left renal arteries (one assumed accessory) and vena cava inferior thrombosis (Figure 1A). Abdominal aortal digital subtraction angiography found postbifurcational multiple stenosis for upper branch of left artery with delayed kidney upper pole contrast opacification (Figure 1B).

Repeated kidney scintigraphy (Tc-99m MAG3, Tc-99m DMSA) revealed small and inhomogenous left kidney with double renal supply for upper and lower poles. A slow radiopharmac activity arrival and clearance for the upper pole was noticed alongside with normal lower pole activity arrival. Repeated renal scintigraphy showed a decrease of separate renal function on the left kidney (27% left and 73% right differential function *VS* left 21% and 79% right respectively).

Cardiac ultrasound uncovers concentric cardiac left ventricle hypertrophy and low cardiac EF (43%). Thromboses of upper sagital and right transversal sinus on brain MRI were also found.

Following hypertensive crisis, immediate antihypertensive treatment (sodium nitroprusside) was applied until satisfactory blood pressure was achieved [1].

After left upper pole kidney heminephrectromy followed slow reduction of blood pressure, RAP and aldosterone. Treatment with anticoagulant (Clexane®) eventually resulted with full restitution of vena cava inferior and brain sinuses thrombosis with no notable consequences. After treatment considerable convalescence of cardiac EF (2L/min) is noticed and after 6 months follow-up antihypertensive treatment eventually ceased. The boy remained normotensive with both physically and cognitively normal development.

3. Discussion and conclusions

Hypertensive crisis in children are usually secondary to underlying disease [1,3,6]. It is prone to considerable complications, even lethal outcome [1]. When it is associated with signs of end-organ damage such as encephalopathy, congestive cardiac or renal failure, an emergency need for lowering blood pressure is needed [1]. As we were aware of a possible umbilical arterial catheterization consequences in certain cases of renovascular disease, such possibility was carefully ruled-out.

It is also essential to establish early proper diagnosis and precise location of arterial stenosis with angiography and radionuclide examination. We agree that digital

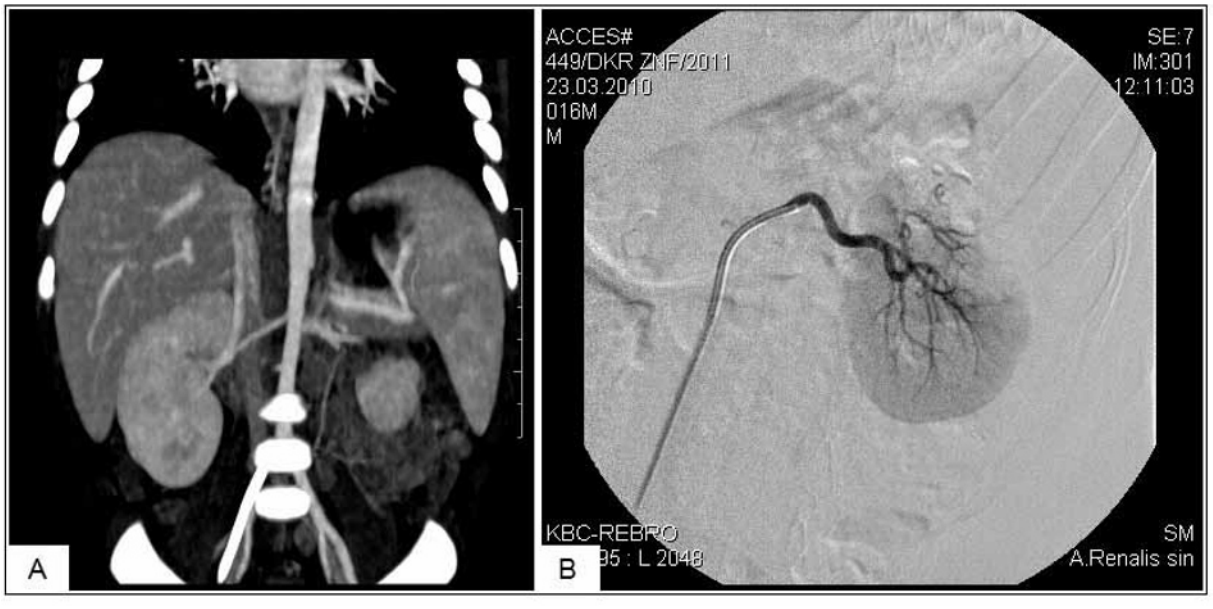

Figure 1. MSCT renal angiography with vascular anomaly (A) and abdominal aortal digital subtraction angiography (B) of the left kidney.

subtraction angiography remains the gold standard investigation [7,8]. Precise location of single arterial stenosis was the key support for our decision of surgical heminephrectomy and removal of just upper pole of the left kidney instead of total nephrectomy. Otherwise, a severe arterial hypertension would ensue with probable deterioration of cardiac function and thromboses (vena cava inferior and brain sinuses). A difficult decision between total nephrectomy and rational sparing *primum non nocere* approach has to be made in such children. Therefore, radiologic and clinical nuclear imaging is aimed to find exact and all renovascular stenosis position(s). Our successful clinical restitution serves as an encouragement for clinical decision of partial instead of total nephrectomy in carefully selected patients [9]. It is also important to acknowledge that persistent restlessness or changes in child's behavior is important for early diagnosis of neonate hypertensive crisis [10].

The written informed consent was obtained and signed by the parents of the child.

Conflict of interest statement

Authors state no conflict of interest.

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