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Case Report

Hyponatremic-hypertensive Syndrome in a 19-month-old Boy with Renovascular Hypertension

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ABSTRACT. Hyponatremic-hypertensive syndrome (HHS) is an uncommon disorder rarely seen in children. Herein, we report a 19-month-old boy with HHS. He had severe hypertension, polyuria, polydipsia, vomiting, and seizure at presentation. Laboratory findings revealed hyponatremia, hypokalemia, metabolic alkalosis, proteinuria, hypercalciuria, high levels of renin and aldosterone, and renal artery stenosis. All symptoms resolved after nephrectomy. Clinicians should be aware of this syndrome because prompt recognition can be lifesaving.

Introduction

The prevalence of hypertension (HTN) among children is reported to be 1%–3%.[1] Although the real prevalence of renovascular HTN in the general population is not known, it accounts for 5% and 25% of all cases with secondary HTN in children.[2,3] The combination of hyponatremia and renovascular HTN is called as hyponatremic-hypertensive syndrome (HHS).[4,5]

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Case Report

A 19-month-old boy was referred to our hospital because of polyuria, polydipsia, vomiting, and multiple episodes of generalized tonic-clonic seizures. He had a fever and vomiting for three days before admission to the hospital. The next day after the onset of his complaints, he was taken to a local hospital because of bronchitis. It was learned that at his follow-up, intravenous fluid and ceftriaxone were initiated. While he was continuing with these therapies, he had generalized tonic-clonic seizures. The pediatrician noticed that he had HTN, hyponatremia, and polyuria. Therefore, he was transferred from outside the hospital to our pediatric intensive care unit because of

HTN, hyponatremia, and seizure. On admission, his height was 80 cm (50–75th percentile in Turkey), his body weight: 10.5 kg (25–50th percentile in Turkey), body temperature: 36.7°C, heart rate: 114 beats/min, respiratory rate: 38 breaths/min, and his blood pressure (BP): 195/140 mm Hg. The patient was lethargic but responsive; signs of neither dehydration nor edema were obtained in his physical examination. The other systemic examinations were normal. He was the first child of his family, and there was no consanguinity between parents. In laboratory examination, sodium (Na) was 111 mmol/L, potassium (K): 2.8 mmol/L, chloride (Cl): 65 mmol/L, urea: 12 mg/dL, creatinine (Cr): 0.4 mg/dL, venous blood gas pH: 7.55, bicarbonate: 30.4 mmol/L. The plasma renin level was 5500 IU/mL (4.2–59.7), and the aldosterone level was 340 ng/dL (4–16). In urine analysis, density was 1006, protein +++ (urinary protein/Cr 40 mg/mg), and there was hypercalciuria (urinary calcium/Cr 3.2 mg/mg). He had polyuria (22 mL/kg/h), and the urinary Na level was 36 mmol/L (Table 1). He was urgently managed by intravenous antihypertensive drugs and Na replacement for hyponatremia. Renal sonogram showed a small left kidney. Renal Doppler ultrasound revealed severe left renal arterial stenosis. Computed tomography angiography showed that the renal arterial occlusion was 2–3 mm after the beginning of the artery and the branches of renal artery with collaterals (Figure 1). Fundoscopic examination was normal, and echocardiography showed left ventricular hypertrophy. Diethylenetriaminepentaacetic acid renoscan revealed that left renal function was low. The glomerular filtration level was 51 mL/min in the right and 8.1 mL/min in the left kidney. The patient was diagnosed as HHS with the clinical, laboratory, and radiologic features. After an uneventful left nephrectomy procedure, the patient was discharged without medication and without neurologic sequelae.

The authors obtained all appropriate consent forms from the patient's parents/guardians.

Discussion

The clinical picture characterized by hyponatremia, severe HTN, and hypokalemic alkalosis due to renal artery stenosis has been reported in adult patients since 1950.[6] This syndrome was named by Brown et al[7] in 1965, and it was originally reported, especially in adult patients. The majority of adult patients are elderly women with atherosclerosis, and this syndrome is not encountered frequently in pediatric cases.[8] Presenting symptoms of HHS include polyuria, polydipsia, weight loss, weakness, headache, enuresis, consciousness, seizures, and neurological and behavioral findings.[8]

Table 1. Laboratory results of the patient with hyponatremic-hypertensive syndrome.

Laboratory measurement	Measured value	Normal value
Blood		
Sodium (mmol/L)	111	136–146
Potassium (mmol/L)	2.8	3.5–5.1
Chloride (mmol/L)	65	101–109
Urea (mg/dL)	12	10.8–38.4
Creatinine (mg/dL)	0.4	0.26–0.77
pH	7.55	7.35–7.45
Bicarbonate (mmol/L)	30.4	20–22
Hemoglobin (g/dL)	13.7	13–15
Urine		
Sodium	36	
Potassium	9.5	
Protein	+++	
pH	7	
Density	1006	

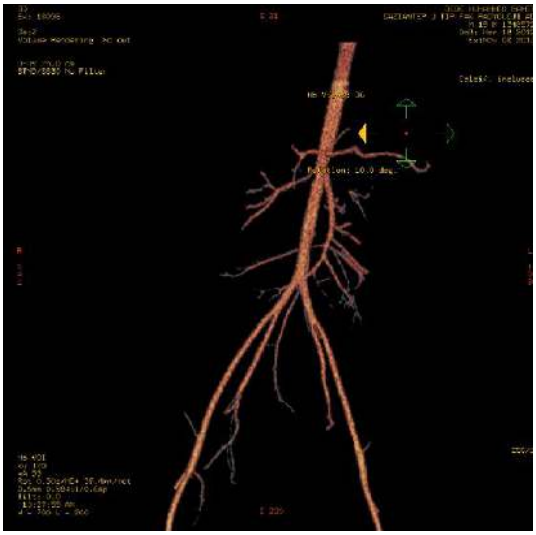

Figure 1. Left renal artery stenosis in computed tomography angiography.

HHS occurs due to complex mechanisms. Renal arterial thrombosis causes hypoperfusion of the kidney, this activates renin–angiotensin system, and it causes HTN. The contralateral nonstenotic kidney reacts to HTN by excreting salt and water. HTN also stimulates atrial natriuretic peptide (ANP) and brain natriuretic peptide (BNP) to excrete salt and protein. This causes polyuria and dehydration. Hypovolemia causes angiotensin II release, and it stimulates anti-diuretic hormone. Both of these aggravate hyponatremia. Aldosterone causes potassium loss which in turn stimulates renin secretion, and this causes a vicious circle.[9] The pressure-induced hyperfiltration and natriuresis might be responsible for glycosuria and hypercalciuria.[10-12] Plasma renin and aldosterone levels are elevated. Proteinuria can sometimes be in the nephrotic range because angiotensin II can alter glomerular hemodynamics and intrinsic selective properties of the glomerular membrane in rats.[13] In our patient, hyponatremia, hypokalemia, hypochloremic alkalosis, nephrotic range proteinuria, and hypercalciuria were obtained. Plasma renin and aldosterone levels were high, but no glycosuria in our patient.

Renovascular HTN accounts for up to 25% of cases of severe HTN in children, and fibromuscular dysplasia is the most common cause. HHS is a presenting cause of fibromuscular dysplasia or renal artery stenosis.[9] HHS can be described in different categories such as anatomic problems (fibromuscular dysplasia and extrinsic compression), vasculitis (Kawasaki disease, polyarteritis nodosa, and Takayasu's disease), other rare syndromes (neurofibromatosis 1, tuberous sclerosis, Marfan's syndrome, and Williams syndrome), localized tissue damage (trauma, radiation, and umbilical artery catheterization), and congenital reasons (congenital rubella).[14] In our patient, neither any syndromes nor vasculitis was shown, and in the neonatal period, HHS could be associated with umbilical arterial catheter insertion.[15] There was no history of umbilical catheterization in our patient. After suspicion of the disease, renal Doppler ultrasound, computed tomography, and renal scan could provide to reach diagnosis. We also performed these radiologic and scintigraphic examinations to reach the diagnosis.

Renal artery HTN causing HHS is difficult to treat with antihypertensives only. After initial control of BP and fluid therapy, either percutaneous transluminal angioplasty (PTA) or nephrectomy could be performed.[16] In 2006, Seracini et al[17] had performed successful PTA to a 15-month-old girl. In 2007, it has been suggested that nephrectomy may be an option both in ineffectual angioplasty procedure and the low addition of affected kidney to global renal function (<10%).[18] Ashida et al[2] also speculated that HHS becomes when the stenosis of the affected artery is so severe so that revascularization by PTA is impossible. Neeli[19] also performed nephrectomy in his patient because of complete renal artery occlusion with nonfunctioning kidney. Similarly, severe stenosis that cannot be intervened by low renal function in our patient and left nephrectomy was performed in our patient with satisfactory result.

Conclusion

Although HHS is rare in children, all clinicians should be aware of unilateral renal stenosis in cases presenting with hyponatremia

and severe HTN.

Conflict of interest: None declared.

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