Hyponatremic-Hypertensive Syndrome in an 18-Month-Old Male Child

Hiponatremik-Hipertansif Sendromlu 18 Aylık Erkek Çocuk

Hiponatremik-Hipertansif Sendrom / Hyponatremic-Hypertensive Syndrome

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Özet

Poliüri, polidipsi, hipertansiyon, ciddi hiponatremi, metabolik alkaloz ve nefrotik sınırda proteinürisi olan, 18 aylık erkek olgu sunuldu. Hipertansiyon ilaca dirençliydi. Angiografide, sağ renal arter tıkanıklığı saptandı. Böbreğin nükleer taramasında, afonksiyone sağ böbrek tespit edildi. Perkütan renal arter anjiyoplasti, hastamız için uygun olmadığından nefrektomi yapıldı. Tüm bulgular sağ nefrektomiden sonra düzeldi. Çocuklarda hiponatremik hipertansif sendromun nadir olması, onun yeterli bilinmediğini gösterir. Poliürik, hipertansif ve hiponatremik çocuklarda, renal hasarı ve yaşamı tehdit eden komplikasyonları önlemek amacıyla, hiponatremik hipertansif sendromun farkındalığının arttırılması amaçlandı.

Anahtar Kelimeler

Hiponatremi; Hipertansiyon; Renal Arter Darlığı; Poliüri; Hiponatremik-Hipertansif Sendrom

Abstract

An eighteen-month-old boy presented with polyuria, polydipsia, hypertension, severe hyponatremia, metabolic alkalosis and nephrotic-range proteinuria. Hypertension was drug resistant. Renal artery angiogram revealed right renal artery occlusion. Nonfunctional right kidney was also detected on the nuclear renal scan. As percutaneous transluminal renal artery angioplasty was not appropriate for our patient, nephrectomy was performed. Right nephrectomy resulted in the resolution of all the symptoms. Rarity of hyponatremic hypertensive syndrome in children may project its under-recognition. We aimed to increase awareness of early diagnosis of HHS among polyuric, hypertensive and hyponatremic children in order to prevent renal damage and life-threatening complications.

Keywords

Hyponatremia; Hypertension; Renal Artery Stenosis; Polyuria; Hyponatremic-Hypertensive Syndrome

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Introduction

Hyponatremic hypertensive syndrome (HHS) is a condition characterized by severe hypertension, hyponatremia or hypochloremia caused by unilateral renal artery stenosis or occlusion [1]. In children, 5-25% of the cases with secondary hypertension are due to renovascular hypertension. Although information about HHS in adults is well-known since 1950 [2], there are only a few cases available in the literature regarding this condition in children [1, 3-4]. However, some reports emphasized that rarity of this disease in children might be the result of the lack of its recognition [1, 5].

Activation of renin and angiotensin II system by the ischemic kidney causes hypertension and pressure diuresis from nonstenotic kidney. On the other hand, increase in aldosterone and antidiuretic hormone (ADH) secretion and thirst also cause hyponatremia and hyokalemia [1, 5].

HHS may be accompanied by weight loss, polyuria, polydipsia, hypertension, enuresis, fatigue, unconsciousness and personality changes [1, 3, 5]. Associated laboratory findings may include hyponatremia, hypokalemia, hypochloremic alkalosis, hyperreninemia, hyperaldosteronism, high sodium levels in the urine, and sometimes high nephrotic level of proteinuria, hypercalciuria and glycosuria [5-6]. The aim of this study was to report an infant who presented with polyuria and was diagnosed with hyponatremic hypertensive syndrome.

Case Report

An 18-month-old boy was referred to the Pediatrics department with symptoms of drinking large volumes of water, with frequent urination and weakness, as well as growth retardation for three months. It was reported that his fluid intake was 2000-4000 ml/day and daily urine output was 3000 ml/day since two months. The family history indicated that parents were third-degree relatives. The physical examination showed the infant's body weight to be 9800 g (<3 percentile), height 84 cm (50 percentile), and blood pressure of 180/100 mmHg (>99 percentile) (Table 1). He displayed symptoms of 10% dehydration, such as sunken eyes, dry mucus membranes and mild delay in skin turgor elasticity. All peripheral pulses were evident. Neurological, eye and all other system examinations were normal. No papilledema was found in the eye examination.

Complete blood count and renal function tests (blood urea 25 mg/dl, serum creatinine 0.34 mg/dl) were within normal ranges. Hyponatremia (125 mEq/L), hypochloremia (90 mEq/l), hypokalemia (2.9 mEq/L), hypoalbuminemia (2.9 gr/dl) and hypercholesterolemia (293 mg/dl) were detected. Analysis of blood gases howed metabolic alkalosis (pH: 7.50, bicarbonates 29.8 mEq/L). Urinalysis indicated Ph of 7, density:1005 and +3 proteins. The 24-hour urine protein test result showed 159 mg/m2/hr and the fractional excretion of sodium (FENa) was calculated to be 10% while the fractional excretion of potassium (FEK) was 14%. No glycosuria or hypercalciuria was found. The renin test performed in bed revealed 2500 pg/ml (2.77-61.80 pg/ml) and aldosterone was determined at 745 (30-350 pg/ml). The antidiuretic hormone was not evaluated.

Echocardiogram revealed left ventricular hypertrophy. Evidence of end-organ damages, secondary to hypertension was detected. The dimercaptosuccinic acid (DMSA) revealed a non-functional right kidney (<10% functioning). Renal Doppler ultrasound revealed left hydronephrosis and possible right renal artery stenosis. The left kidney showed normal vascular structure. Magnetic Resonance (MR) angiography revealed hypoplastic right kidney with occlusion in the right renal artery, 2 mm from the aorta outlet. The angiographic examination revealed that the right renal artery was almost completely constricted and the blood circulation in the kidney was via the collateral circulation (Fig. 1). The infant was diagnosed as having HHS with clinical findings of resistant hyponatremia and hypertension with unilateral renal artery stenosis.



Fig. 1. Very little filling or full stenosis of the right renal artery on the renal arteriograph (thin arrow) and normal filling of the left renal artery (bold arrow)

On admission rehydration and sodium chloride treatment were started. The serum sodium was corrected on the 7th day post treatment, with 20-30 mEq/kg/day of sodium chloride. Although maximum doses of nifedipine, enalapril and beta-blocker were administered, no response was observed. As percutaneous transluminal renal artery angioplasty (PTRA) could not be performed in this case, the only option available right nephrectomy, which was performed. One day post nephrectomy, diuresis decreased to 4 cc/kg/hour and blood pressure began to drop. Symptoms like hypertension, polyuria, polydipsia, hyponatremia, alkalosis and nephrotic proteinuria were all resolved by one week post nephrectomy (Table 1). During the follow up of 18 months after nephrectomy, no pathologic clinical or laboratory findings were observed.

Discussion

Hyponatremic hypertensive syndrome is a rare and special type of renovascular hypertension with hyponatremia, hypokalemia, polyuria, proteinuria and high renin activity [4-5]. It occurs more often in adults than in children, a few studies are available on children diagnosed with HHS [3, 6]. HHS could be underdiagnosed in children due to inadequate information and awareness regarding the disease [4-5].

Table 1. Clinical and laboratory parameters of a patient with hyponatremic hypertension syndrome

Variable	Preoperation	Postoperation
Blood Pressure (mmHg)	180/100	85/55
Daily urine volume (ml)	3000	650
Body weight (kg)	9.8	12
Serum parameters Na (meq/l)	125	136
K (meq/l)	2.9	3.7
CI (meq/l)	90	110
Bicarbonate (meq/l)	29.8	23
Albumin (g/dl)	2.9	3.8
Urinary parameters FENa (%)	10	2
FEK (%)	14	3.8
Urinary protein excretion (mg/m2/h)	159	8
Urine osmolarity (mOsm/kg)	169	350
Urinary Na (mmol/l)	90	27
Urinary K(mmol/l)	3	1

FENa=Fractional excretion of sodium; FEK=fractional excretion of potassium

Our patient revealed weight loss, polyuria, polydipsia, drug-resistant hypertension, resistant hyponatremia, hypokalemia, hypochloremic alkalosis, hyperreninemia, hyperaldosteronism, high sodium levels in the urine and nephrotic level of proteinuria. These symptoms are indicators of hyponatremic hypertensive syndrome. Although our patient had polyuria and growth retardation for three months, he was unrecognized as mentioned in the literature [4-5].

For a patient to develop HHS, the condition of unilateral renal artery stenosis is observed, with the other kidney displaying normal perfusion and function [1]. In the pathogenesis of the disease, the main problem is the release of high quantities of renin from the ischemic kidney due to renovascular stenosis [4]. Our patient had right renal artery occlusion. It was thought that renal ischemia in our patient caused hyperreninemia resulting with hypertension and pressure diuresis. Stimulation of thirst, causing polyuria, together with pressure diuresis was the reason of sodium loss in urine resulted with hyponatremia (Table 1). Direct renal action of angiotensin II and ADH secretion might also have role in hyponatremic renin angiotensin system activation caused hyperaldosteronism resulted with hypokalemia [1, 5].

Measurement of blood pressure in children with unexplained polydipsia and polyuria is important. Late diagnosis of hypertension in our patient caused end organ damage both in hearth as left ventricular hypertrophy and kidney as glomerular hyperfiltration. Nephrotic-range proteinuria in our patient similar to that reported by Trivelli [4] was probably due to the glomerular hyperfiltration, deriving from hyperreninemia and hypertension. Quick resolution of proteinuria after right nephrectomy demonstrated reversible nonstenotic kidney hyperfiltration.

Trivelli et al. [4] reported hypercalciuria in two children with HHS which lasted for several months after surgery. We supposed that in our patient, not having hypercalciuria, distal convoluted tubules were not involved.

The first choice for the antihypertensive treatment of cases with HHS should be the angiotensin-converting-enzyme inhibitors or angiotensin receptor blockers. Except potent diuretics, which may exacerbate the disorder, additional antihypertensive drugs can be used [5]. No response was observed to the antihypertensive agents used.

For the achievement of cure, surgical correction of the underlying renal ischemia is necessary Percutaneous transluminal renal angioplasty with or without stenting or uninephrectomy can be used [5, 7]. Nephrectomy is necessary if the ischemic kidney contributes less than 10% of the renal function or if PTRA fails [6, 8]. It is impossible to perform PTRA in our patient, because his right renal artery was almost completely constricted and DMSA revealed a non-functional (10 %) right kidney. Therefore, we performed to threat our patient with unilateral nephrectomy of the affected kidney.

In conclusion, clinicians should be careful in differantial diagnosis of the children with polyuria and unexplained electrolyte abnormalities. This case, with the other few reports, indicated that HHS may also be seen in renovascular hypertension in children. Our case report also emphasizes the importance of early diagnosis in preventing life-threatining complications and preserving renal function of the non-stenotic kidney.

Competing interests

The authors declare that they have no competing interests.

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