Hyponatremic Hypertensive Syndrome Complicating Unilateral Renal Artery Stenosis: A Rare Manifestation in Childhood

Sir,

Rarely adults with renovascular hypertension may present with neurological symptoms, polyuria, polydipsia and hyponatremia known as a hyponatremic hypertensive syndrome (HHS).[1] Similar occurrence in children is uncommon and unilateral renal artery stenosis (RAS) is reported to be the chief cause.[2] We report the case of an 11-year-old boy presenting with HHS secondary to unilateral RAS, complicated by posterior reversible leukoencephalopathy syndrome (PRES).

An 11-year-old boy presented with fever, headache, vomiting, polyuria and polydipsia for four days and multiple episodes of generalised tonic-clonic seizures for 1 day. He was responsive only to painful stimuli. His pulse rate was 110/min, palpable in all four limbs with no radio-radial or radio-femoral delay; the respiratory rate was 26/min. The blood pressure was 170/130 mm Hg. Pupils were bilaterally of normal size and reacting to light with no cranial nerve deficits. Tone and power were normal in all four limbs. Deep tendon reflexes were brisk with no meningeal signs. Fundus examination revealed disc oedema. A diagnosis of hypertensive encephalopathy was made. Blood investigations revealed urea of 88 mg/dL, creatinine of 3.2 mg/dL, sodium 126 mEq/L, potassium 3 mEq/L and metabolic alkalosis (pH = 7.48, HCO_3 = 28.8). Blood pressure was controlled over the next 48 h with labetalol infusion. The renal parameters and serum electrolytes gradually normalised over a period of 72 h. Besides the polyuria, polydipsia too disappeared with the control of hypertension. Abdominal ultrasonography revealed right-sided atrophic kidney (6.6 * 2.4 cm) with mildly raised echogenicity. The left kidney (9.6 * 4.5 cm) showed compensatory hypertrophy with normal cortico-medullary differentiation; ultrasound doppler showed a paucity of flow in the right renal artery. With these radiological findings, persistent hyponatremia and hypertension, a clinical diagnosis of HHS was made. MRI of the brain revealed features suggestive of PRES. CT Angiography revealed near-complete attenuation of the right main renal artery, hence confirming the diagnosis [Figure 1]. Diethylenetriaminepentaacetic acid (DTPA) scan showed nonfunctional right kidney with normally functioning left kidney. The child at discharge required five antihypertensive agents (labetalol, amlodipine, enalapril, furosemide and clonidine). In view of the nonfunctional right kidney and severe hypertension, a decision for right nephrectomy was taken along with the paediatric surgeon. The postoperative course was uneventful. Subsequently, he did not require any antihypertensive agent and is on follow-up for the last 6 months.

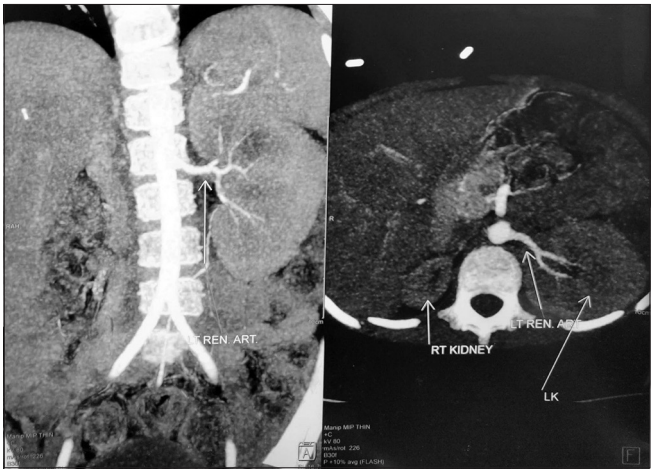

Figure 1: CT angiography of aorta suggestive of the small right kidney (5.1 cm *2.3 cm) with partial visualisation of the right renal artery (thread-like)

HHS is characterised biochemically by hyponatremia, hypokalemia, hypochloremia, metabolic alkalosis, hyperreninemia, hyperaldosteronemia and elevated urinary sodium and protein.[3] The central pathophysiology in HHS involves activation of the Renin-Angiotensin-Aldosterone system (RAAS) secondary to the unilateral renal ischemia. The RAS-mediated elevation in blood pressure causes pressure diuresis and natriuresis through the contralateral kidney. The resulting volume depletion stimulates the release of antidiuretic hormone, aggravating the hyponatremia and further release of renin. Besides the released aldosterone and angiotensin II also cause hypokalemia.[4] In a recent literature review, Ding *et al*. reported that hypertension, polyuria and polydipsia were the commonest presenting features of pediatric HHS.[2]

HHS requires prompt management with hydration to prevent further renal ischemic injury.[2] Calcium channel blockers are suggested as first-line agents for severe hypertension with acute kidney injury (AKI), while ACE inhibitors are required to suppress the overactive RAAS in unilateral RAS after AKI recovery.[2] Finally, the underlying cause must be treated. Revascularisation is preferred for unilateral RAS, usually by percutaneous transluminal angioplasty and authors have reported the normalisation of blood pressure after either angioplasty or nephrectomy.[2-5] Our patient had a nonfunctional right kidney, a source of persistent hypertension, hence, right nephrectomy was done.

Thus, the unique combination of hyponatremia and hypertension should raise suspicion of underlying renovascular disease, which is potentially curable with early diagnosis and treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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