Hypertensive Encephalopathy Caused by Fibromuscular Dysplasia of Renal Arteries in a Child

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Systemic hypertension is uncommon among infants and children. It is usually secondary to underlying diseases. Here, we present an 11-year-old boy with critical renal artery stenosis with unusual initial presentation of status epilepticus and hypertensive cerebellar encephalopathy. Angiography revealed bilateral renal arteries fibromuscular dysplasia. The encephalopathy improved dramatically after renal artery angioplasty. This patient reminds us renovascular disorder can be the cause of neurological diseases. Though he has bilateral renal arterial stenosis, we only dilated the focal critical stenosis of the right renal artery and his symptoms improved.

Key Words: Encephalopathy • Fibromuscular dysplasia • Renal artery stenosis • Status epilepticus

INTRODUCTION

Systemic hypertension is uncommon among infants and children. It is usually secondary to underlying diseases. In pediatric patients, the frequently mentioned causes are renal, cardiovascular, endocrine, and medication. Here, we present an 11-year-old boy with critical renal artery stenosis with unusual initial presentation of status epilepticus and hypertensive cerebellar encephalopathy.

CASE REPORT

An 11-year-old boy complained of dizziness and nausea since 5 days before admission. Progressive general malaise, poor activity, and unsteady gait were also found by his parents. Generalized tonic-clonic seizure suddenly happened on the day of admission and lasted more than 40 minutes. The blood pressure was 161/125 mmHg at presentation. Initial T2-weighted and fluid-attenuated inversion recovery (FLAIR) brain magnetic resonance imaging (MRI) (TE113/TR4020) showed multiple hyperintensities in the gray matter and subcortical white matter of bilateral parietal-occipital lobes, and in bilateral cerebellar hemispheres (Figure 1A), which was compatible with the diagnosis of hypertensive encephalopathy and explained the cause of ataxia. Magnetic resonance angiography (MRA) of cerebral vessels revealed non-specific findings. Initial survey including blood urea nitrogen, creatinine, and urine analysis was within normal limits. Normal architecture and echogenicity of bilateral kidneys were seen by renal sonography. No significant vascular stenosis could be identified by Doppler scan, either. However, significantly elevated blood renin (309 pg/ml) and aldosterone (951 pg/ml) were found at initial workup. Renal artery stenosis could not be ruled out. So, renal angiography was arranged for this boy. The angiography revealed multiple sites of stenosis over bilateral renal and iliac...
arteries (Figures 2A, B) with critical stenosis at one branch of the right renal artery (Figure 2A arrow). The celiac trunk and superior mesenteric artery were normal. The finding of angiography was compatible with the diagnosis of fibromuscular dysplasia (FMD). The boy received percutaneous transluminal renal angioplasty (PTRA) by a 5.0 mm balloon (NC Sprinter balloon, Medtronic, Inc., Minnesota, USA) (Figure 2C) with good results (Figure 2D). We did not perform angioplasty on the left renal artery because there was no critical stenosis within reachable regions. The blood pressure was 116/78 mmHg after PTRA. The patient was then discharged 2 days later after PTRA. Blood renin and aldosterone level decreased gradually to nadirs of 40 pg/ml and 129 pg/ml, respectively, 1 month after the procedure. The brain MRI follow-up (TE90/TR4500) 3 months later revealed no abnormal intensities (Figure 1B).

DISCUSSION

In this patient, the initial presentations of ataxia followed by status epilepticus were rare, and, to our best knowledge, the image of hypertensive cerebellar encephalopathy related to renal artery stenosis in children has never been reported before.

In this patient, hyperintensities in T2-weighted and FLAIR brain MRI without other abnormal signal intensity suggested cerebral edema. Significant hypertension was noted at presentation. So, hypertensive encephalopathy was a reasonable explanation for his cerebral edema. Infarction was less likely because there was no evidence of cerebral vessel anomalies on MRA. In general condition, cerebral blood flow was maintained by autoregulation of arterioles over a range of systemic blood pressure. When hypertension exceeds the autoregulation of cerebral flow, hyperperfusion will result in extravasation of fluids into the brain parenchyma and cause cerebral edema, so-called hypertensive encephalopathy.

In infants and younger children, systemic hypertension is uncommon, with a prevalence of 1~4.5%. Secondary cause can be found in a majority of young children. The most common etiology of pediatric hypertension is primary renal diseases (60-70%). The second most common is renovascular disorders, which account for 5-10% of patients with pediatric hypertension. Fibromuscular dysplasia (FMD) is an important cause of renal-vascular disorders. It is also occasionally associated with intracerebral vascular diseases. FMD is currently defined as an idiopathic, segmental, non-inflammatory and non-atherosclerotic disease of the musculature of arterial walls, leading to stenosis of small and medium-sized arteries. The pathogenesis of FMD is unknown, although various hormonal and mechanical...
factors have been suggested.\textsuperscript{5} It appears to be familial in 10\% of cases, and the renal arteries are the most common involved vessels.\textsuperscript{5} The symptoms of FMD with reno-vascular hypertension include headache, seizure, growth retardation, cognitive dysfunction, polyuria, palpitation, hemiplegia, and epistaxis.\textsuperscript{2,3} Angiography is the gold standard of diagnosis of FMD, and the treatment of hypertension related to renal artery stenosis includes antihypertensive agents, angioplasty, and surgery.\textsuperscript{5} Percutaneous transluminal renal angioplasty (PTRA) was reported to be effective and safe if the arterial lesions were in favorable locations.\textsuperscript{7-9} The success rate of PTRA was reported to be about 50\% in children.\textsuperscript{10} The prognosis of FMD is considered better than that of atherosclerotic renal artery stenosis due to less severe progression of arterial stenosis.\textsuperscript{5} There could be residual stenosis in this patient’s renal arteries because blood renin and aldosterone did not return to normal range after the procedure. The patient might need re-intervention in the future. However, good responses have been reported for patients undergoing re-intervention.\textsuperscript{8,11}

This patient reminds us clinicians should maintain a high index of suspicion for renovascular diseases when confronting patients with neurologic presentations. Aggressive treatment, if feasible, might improve the patient’s outcome.

REFERENCES