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A Case of Cyclic Vomiting Syndrome-Induced Hypertension Causing Posterior Reversible Encephalopathy Syndrome

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Abstract: Cyclic vomiting syndrome (CVS) is characterized by repeated episodes of vomiting in a stereotyped pattern and is a known cause of hypertension. Our patient is a 10-year-old female who presented with nonbilious, nonbloody vomiting, and constipation concerning for a flare of her known CVS. During the hospital course, she developed intermittent severe hypertensive episodes, leading to an acute episode of altered mental status and a tonic-clonic seizure. Magnetic resonance imaging confirmed diagnosis of posterior reversible encephalopathy syndrome (PRES) after eliminating other organic etiologies. This is one of the first documented cases of CVS-induced hypertension causing PRES.

Key Words: cyclic vomiting syndrome, posterior reversible encephalopathy syndrome, hypertension

INTRODUCTION

Cyclic vomiting syndrome (CVS) is characterized by repeated episodes of vomiting in a stereotyped pattern. Autonomic dysfunction and hypertension are known complications of CVS, likely due to increased sympathetic tone (1) and hypothalamic-pituitary-adrenal axis overactivity (2), respectively. Hypertension is a well-established risk factor for pediatric posterior reversible encephalopathy syndrome (PRES). We report a pediatric patient with CVS-induced hypertension leading to PRES.

CASE REPORT

A 10-year-old female with a history of reflux, CVS diagnosed within the first year of life, and status postgastrostomy tube presented with new onset nonbilious, nonbloody vomiting, and constipation concerning for an exacerbation of CVS. The patient's mother estimates that she has had 25–30 lifetime episodes, often requiring 1 week of hospitalization. The patient was not on prophylactic medications for CVS, with her last flare occurring 3 years before presentation. She was admitted to the inpatient pediatric gastroenterology (GI) service and given intravenous promethazine and fluids. Constipation was a known trigger for this patient's CVS episodes in the past. The patient was thus intended to initiate a bowel clean out with magnesium-citrate once able to tolerate enteral intake.

Overnight on hospital day 1, the patient developed intermittent episodes of hypertension, reaching systolic values in the 140s mm Hg (99th percentile for age/weight). The team decided to closely monitor the blood pressures (BP) and consider therapy with another hypertensive episode. On hospital day 2, the patient had an acute episode of waxing and waning altered mental status and left upper and lower extremity weakness. Vital signs during this episode were stable with a BP 116/81 mm Hg (95th percentile for age/weight), heart rate of 98 beats per minute, and glucose was 116 mg/dL. Electrolytes were within normal limits. The patient was transferred to the pediatric intensive care unit and underwent a tonic-clonic seizure which lasted 3 minutes and was aborted by 2 mg of intravenous lorazepam. Computed tomography of the head and magnetic resonance (MR) imaging of the brain showed a T2/FLAIR signal abnormality with diffusion restriction in the posteromedial right parietal lobe concerning for ischemic infarct versus PRES (Fig. 1). MR venous imaging was normal. She was started on levetiracetam, nicardipine, and made nil per os. After stabilization in the pediatric intensive care unit, the patient was transferred back to the pediatric GI service on hospital day 5. She was successfully weaned off of nicardipine with normal BP (Fig. 2). She underwent the magnesium-citrate clean out resulting in a bowel movement with no further episodes of emesis. On

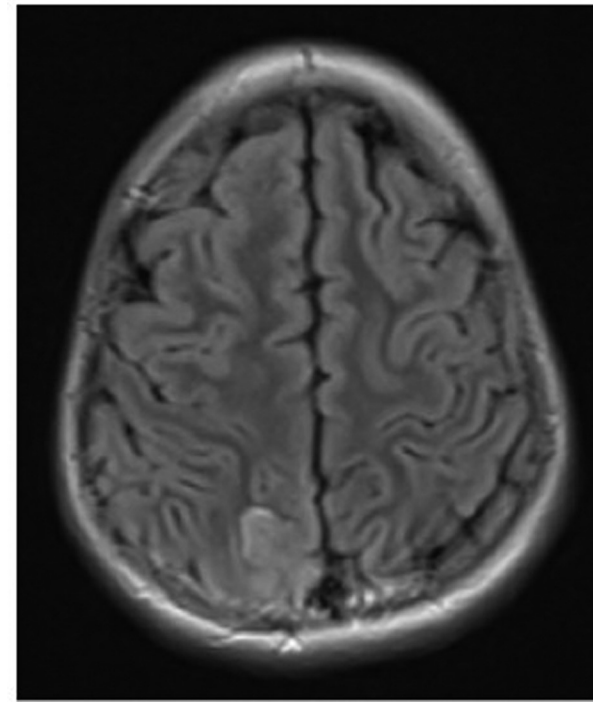

FIGURE 1. MR imaging T2/FLAIR status post tonic-clonic seizure. MR = magnetic resonance.

Received July 14, 2022; accepted January 17, 2023.
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J.M. is on the speaker's bureau for AbbVie Inc and the scientific advisory. The remaining authors report no conflicts of interest.

JPGN Reports (2023) 4:2(e294)
ISSN: 2691-171X
DOI: 10.1097/PG9.0000000000000294

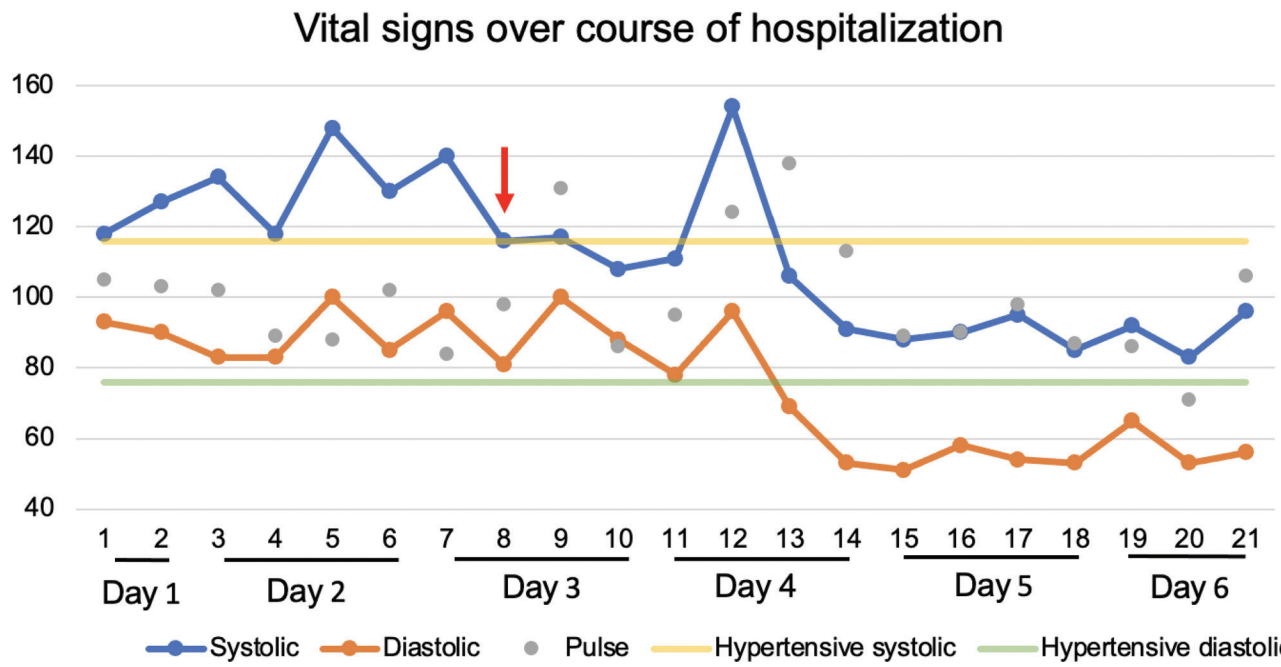

FIGURE 2. Vital signs charted over the course of the hospitalization. Vital sign points represent every 6 h since admission. The red arrow indicates when the episode of altered mental status started. The yellow and green lines represent the 95th percentile of systolic and diastolic blood pressure, respectively, for this patient's age and weight.

hospital day 6, repeat MR imaging of the brain showed complete resolution of diffusion abnormalities. The patient had undergone previous upper GI series, renal ultrasound, and brain MRI that were all normal. Extensive evaluation was unrevealing for genetic, metabolic, or nephrogenic etiologies, suggesting a likely diagnosis of PRES secondary to hypertension. The patient was discharged on CVS prophylactic medications including abortive rizatriptan and rescue lorazepam and promethazine in addition to a hypertension management plan with oral clonidine if systolic BP >140 mm Hg to take at home before presenting to the hospital for further care.

DISCUSSION

While hypertension is associated with CVS and an important risk factor for PRES, we do not find any reported case of hypertension secondary to CVS leading to PRES. One case report detailed a 56-year-old woman with CVS who presented in hypertensive crisis before the prodrome onset, suggesting that hypertension may also be an antecedent factor to acute episodes of CVS (3). Of note, the Sato-variant of CVS presents with hypertension and extreme lethargy due hypothalamic-pituitary-adrenal axis overactivity with associated laboratory abnormalities including elevated prostaglandin E2, cortisol, catecholamines, and antidiuretic hormone and an ACTH level often 5–6 times greater than the upper limit of normal. While 6% of patients with CVS are reported to have the Sato-variant, this patient did not have a full evaluation (4). The Sato-variant should be considered a possible cause for the patient's hypertension.

Common causes of PRES in the pediatric population include renal disease, hematologic disorders, and their associated medications such as corticosteroids and cytotoxic agents (5). Pediatric PRES has a nonspecific initial presentation ranging in acuity from hypertension and headaches to altered mental status, seizures, focal neurologic deficits, and visual disturbances (5). A broad differential exists for PRES, including ictal or postictal states, encephalopathies, osmotic demyelination syndrome, cerebral venous thrombosis, and mitochondrial myopathy encephalopathy lactacidosis and stroke-like syndrome (5). MR imaging is necessary for diagnosis. Despite its name, PRES can have significant morbidity associated with vasogenic edema and hemorrhage (6). Therefore, it is important to carefully monitor BP in CVS patients with active episodes, and to institute an aggressive management plan for those who develop significant hypertension during CVS episodes.

ACKNOWLEDGMENTS

Written informed consent was obtained from the patient's parents for publication of the case details.

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