

Posterior reversible encephalopathy syndrome in a child with cyclical vomiting and hypertension: a case report

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CASE REPORT

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Posterior reversible encephalopathy syndrome in a child with cyclical vomiting and hypertension: a case report

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Abstract

Introduction: Posterior reversible encephalopathy syndrome is characterized by headache, nausea and vomiting, seizures and visual disturbances. It has certain characteristic radiological features, which allow diagnosis in the appropriate clinical setting and enable appropriate clinical therapy to be instituted.

Case presentation: A 10-year-old Caucasian girl who was hospitalized due to recurrent vomiting was diagnosed as having posterior reversible encephalopathy syndrome after an initial diagnosis of cyclical vomiting and hypertension was made.

Conclusion: Posterior reversible encephalopathy syndrome is a rare disorder in children. Early recognition of characteristic radiological features is key to the diagnosis as clinical symptoms may be non-specific or mimic other neurological illnesses. To the best of our knowledge this is the first case to report an association between posterior reversible encephalopathy syndrome, cyclical vomiting and hypertension. Furthermore, in this case, the resolution of the abnormalities found on magnetic resonance imaging over time did not appear to equate with clinical recovery.

Introduction

Posterior reversible encephalopathy syndrome (PRES) is characterized by headache, nausea, vomiting, seizures and visual disturbances [1]. PRES is commonly associated with a sudden increase in blood pressure (BP) [1]. The MRI findings have been well characterized and include vasogenic edema in the white matter of the posterior regions of the cerebral hemispheres, particularly in the parieto-occipital regions [2]. PRES is more commonly reported in adults. The cause of PRES is thought to be multi-factorial, and it may develop in patients who have hypertension, renal disease, or who are immunosuppressed [1,3]. PRES is usually reversible and prompt recognition is important [1]. In the pediatric population, PRES has been associated with chronic renal disease [4], the administration of chemotherapeutic agents [5], adrenocortical disease and Cushing's syndrome [6].

We present the case of a 10-year-old girl found to have PRES in association with cyclical vomiting and hypertension.

Case presentation

A 10-year-old Caucasian girl presented to our department with ongoing symptoms of vomiting, non-specific abdominal pain and hypertension. She had been admitted about 15 times over a three year period with episodic attacks of frequent and severe vomiting lasting for a few days. During some of her admissions she demonstrated neurological and autonomic signs and symptoms such as confusion, disorientation, occipital headache, visual impairment, staring look, lack of response, head and eye turning to one side with nystagmus, non-reactive pupils, left arm and leg stiffening, and fluctuating and raised BP.

Investigations included abdominal and chest radiographs, and abdominal and renal ultrasonography, which all gave negative results. Gastroscopy and barium meal studies did not reveal any abnormalities, but a urease breath test revealed *Helicobacter pylori*, which was treated with standard triple therapy. A cranial

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computed tomography (CT) scan did not reveal any abnormalities; however, a magnetic resonance imaging (MRI) scan (Philips Intera 1.5T) demonstrated patchy areas of mainly subcortical high signal without mass effect, contrast enhancement or associated diffusion restriction. These abnormalities were bilateral but asymmetrical, affecting the right cerebral hemisphere more than the left side. The high signal lesions were mainly located in the posterior brain, particularly the parieto-occipital lobes. No abnormality was seen in the posterior fossa or the basal ganglia. The radiological features were consistent with a diagnosis of PRES (Figures 1 and 2). An abdominal MRI was unremarkable.

Electroencephalographic (EEG) studies initially demonstrated marked right hemisphere slow wave disturbances; however, repeat studies showed no definite epileptiform abnormality, with slow and asymmetrical, frequent theta and slow activity consistent with non-specific focal organic disturbance of cerebral activity. The results of 24-hour BP monitoring (38 readings in total) revealed 10 systolic readings >118 mmHg (95th centile). Electrocardiographic (ECG) studies were also unremarkable. Midnight and morning cortisol levels were within the normal ranges. Other investigations such as urinary catecholamines, serum amylase, lactate, ammonia, cholesterol, chloride, and bicarbonate were all within normal ranges.

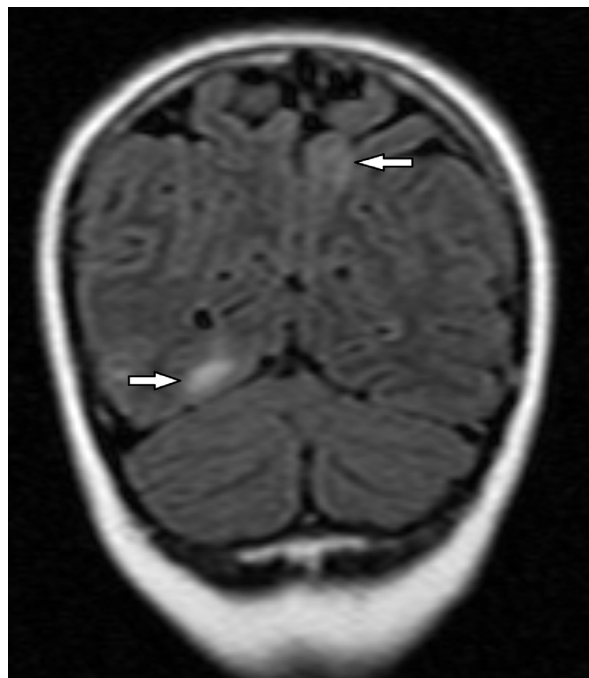


Figure 1 Coronal fluid attenuation inversion recovery (FLAIR) MRI through the posterior brain showing bilateral patchy areas of high signal within the subcortical white matter of right occipital lobe and left parietal lobe.

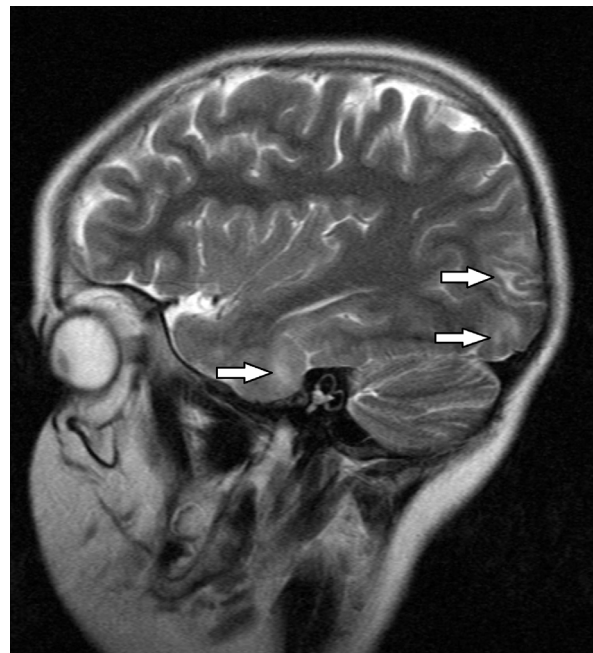


Figure 2 T2 sagittal MRI through a right paracentral position showing multiple subcortical white matter lesions in the right temporal lobe anteriorly, and the right occipital lobe posteriorly.

Other investigations with normal outcomes included tests for urine porphyrin, anti-mitochondrial antibodies (AMA), anti-nuclear antibodies (ANA), double-stranded DNA (dsDNA), anti-smooth muscle antibodies (ASA), liver-kidney microsomal (LKM) antibodies, gastric parietal cell antibodies, carnitine and acylcarnitines, and a coeliac screen. A screen for orotic acid was also normal. No abnormality was detected in the urinary organic acids, but there was a slight increase in the urine amino acids. Investigations for porphyria were also normal.

Our patient was treated with the antiemetics ondansetron and cyclizine, and a trial of lorazepam was also given to try and abort the vomiting cycle. Electrolyte abnormalities were treated using intravenous fluids. At five months after her initial MRI, a repeat scan was performed and all the abnormal features had resolved. Her seizure-like symptoms settled and the vomiting episodes became shorter and less frequent. She continued to have ongoing symptoms of acute episodes of vomiting associated with hypertension for a further three months. Treatment included ondansetron, atenolol and clarithromycin. Her symptoms eventually settled and she has remained symptom free for a period of about 6 months.

Discussion

PRES is a disorder of cerebrovascular autoregulation with multiple underlying etiologies and it is commonly

associated with increases in BP [1]. In the pediatric population, PRES has been associated with chronic renal disease [4], the administration of chemotherapeutic agents [5], adrenocortical disease and Cushing's syndrome [6]. It is thought that the sudden elevation in blood pressure leads to disruption of the autoregulatory mechanisms in the central nervous system, vasodilatation and vasoconstriction resulting in a breakdown of the blood-brain barrier [5]. However, it is documented in some cases, particularly in the pediatric population, that BP may be only minimally elevated or fluctuant during the development of PRES [7].

The diagnosis of PRES can be made via CT, but MRI is a more sensitive imaging modality. The radiological appearance of PRES does not seem to be influenced by the predisposing factor [2]. The most common abnormalities on CT and/or MRI scans are focal regions of vasogenic edema involving the white matter in the posterior cerebral hemispheres, often asymmetrically and most commonly involving the parieto-occipital lobes bilaterally, often in a watershed-type distribution. The medial occipital lobe structures are spared, which distinguishes PRES from bilateral posterior cerebral artery infarcts. The posterior predilection of this condition has been ascribed to the fact that these vascular territories are sparsely innervated with sympathetic nerves [7].

Lesions that are high signal on T2-weighted fluid attenuated inversion recovery (FLAIR) sequences can also be seen in the frontal lobes, the temporal-occipital lobe and the basal ganglia and cerebellum. Patchy grey matter involvement is also recognized. MRI diffusion-weighted imaging (DWI) demonstrates that the areas of abnormality represent vasogenic edema, which is usually completely reversible once therapy is instituted [7]. Rarely, contrast enhancement can occur, presumed to reflect disruption of the blood-brain barrier. In most patients who have repeat MRI scans after correction of hypertension, there is improvement or resolution of radiological abnormalities, although hemorrhages (seen in approximately 15% of cases) can cause permanent structural damage [7].

Manifestations of PRES in the adult population include seizures, visual disturbances and headache [1]. In children, studies have also found that seizures, headache and altered mental status can be the most common clinical features [5]. The other symptoms being nausea and vomiting, and blurring of vision [5]. Studies have also revealed an altered autonomic response in patients with cyclical vomiting [8,9] and there can be a heightened sympathetic cardiovascular tone [10] and symptoms such as pallor, flushing, lethargy and fever [11]. The stress response may induce episodes of cyclical vomiting with infectious, physical or psychological stressors potentially triggering an episode [12]. It has been

reported that an increased level of adrenocorticotrophic hormone (ACTH) and cortisol can be associated with lethargy and hypertension before the onset of vomiting [12], and it has been hypothesized that corticotropin-releasing factor (CRF) may be a brain-gut mediator that directly connects stress and vomiting [13].

In our patient, cyclical vomiting and hypertension coexisted and were associated with PRES. Our patient was extensively investigated for endocrine, renal, gastrointestinal, neurological, cardiac and metabolic causes with no conclusive pathology. Macrolides and ondansetron were effectively used to prevent episodes of vomiting or to decrease their frequency, and this has been previously reported in the literature [14,15]. A recent *in vitro* study has also highlighted the effectiveness of clarithromycin as a prokinetic agent [16], and in our patient it was better tolerated than erythromycin. However, more studies are required to compare the effectiveness of the two agents in patients with cyclical vomiting. Other modes of therapy include the use of tricyclic antidepressants, newer antiepileptic agents such as levetiracetam and topiramate, and the use of antimigraine medications such as sumatriptan [15]. Supportive care involves use of intravenous fluids, sedatives, analgesia and the avoidance of potential triggering factors.

Conclusion

PRES is a rare disorder in children. Early recognition of characteristic radiological features is key to the diagnosis as clinical symptoms may be non-specific or mimic other neurological illnesses. To the best of our knowledge this is the first case to report an association between PRES, cyclical vomiting and hypertension. Furthermore, in this case, the resolution of the abnormalities found on MRI over time did not appear to equate with clinical recovery.

Consent

Written informed consent was obtained from the patient's parents for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Authors' contributions

ZG reviewed the literature, wrote a first draft of the manuscript, corrected, finalized and submitted the manuscript. AR reviewed the literature and edited the manuscript. FGB analyzed the MRI images, reviewed the literature and edited the manuscript. MC reviewed the literature and edited the manuscript. MH was involved with the conception of the report and managed the case. All authors read and approved the final manuscript.

Competing interests

The authors declare that they have no competing interests.

Received: 5 September 2010 Accepted: 6 April 2011

Published: 6 April 2011

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doi:10.1186/1752-1947-5-137

Cite this article as: Gamie et al.: Posterior reversible encephalopathy syndrome in a child with cyclical vomiting and hypertension: a case report. *Journal of Medical Case Reports* 2011 **5**:137.

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