A 7 MONTH PREGNANT FEMALE PRESENTED WITH POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME WITHOUT ECLAMPSIA AND HYPERTENSION: A CASE REPORT

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ABSTRACT

A 7 MONTH PREGNANT FEMALE PRESENTED WITH POSTRIOR REVERSIBLE ENCEPHALOPATHY SYNDROME WITH OUT HYPERTENSION AND ECLAMPSIA

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 26 -year-old 7 month pregnant female guddi bai who was hospitalized due to recurrent altered sensorium, cyclic vomiting, visual disturbances, seizure and left hemi paresis was diagnosed as having posterior reversible encephalopathy syndrome without eclampsia after an initial diagnosis of cerebral venous thrombosis was made.

Conclusion: Posterior reversible encephalopathy syndrome is a rare disorder in pregnant female without eclampsia and hypertension. 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 syndromes, in pregnant female without eclampsia. 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].

We present the case of a 26-year-old pregnant female found to have PRES without eclampsia and hypertension.

CASE PRESENTATION

A 26-year old,7 month pregnant female guddi bai who presented to our department with ongoing symptoms of due to recurrent altered sensorium visual disturbances,cyclic vomiting, seizure and left hemi paresis with non specific abdominal pain. She had been admitted about 3 times over a 4 month period with episodic attacks of frequent and severe vomiting lasting for a few days and altered sensorium for few minute. During some of her admissions she demonstrated neurological 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. Vitals were normal.

Investigations included chest radiographs, and abdominal and renal ultrasonography, which all gave negative results. Gastroscopy and barium meal studies did not reveal any abnormalities, A cranial computed tomography (CT) scan did not reveal any abnormalities; however, a magnetic resnonace 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.

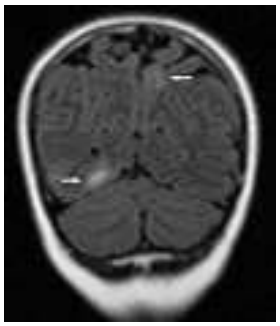

Figure 1. Coronal fluid attenuation inversion recovery (FLAIR) MRI through the posterior brain showing bilateral patchy areas of high signal within the subcortical white matter of right occipital lobe and left parietal lobe

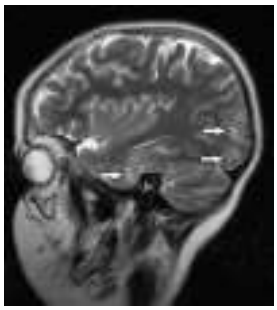

Figure 2. T2 sagittal MRI through a right paracentral position showing multiple subcortical white matter lesions in the right temporal lobe anteriorly, and the right occipital lobe posteriorly

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, cholesterol, chloride, and bicarbonate were all within normal ranges. Other investigations with normal outcomes included tests for urine porphyrin, , anti-nuclear antibodies (ANA), double-stranded DNA (dsDNA), anti-smooth muscle antibodies (ASA), liver-kidney microsomal (LKM) antibodies, gastric parietal cell antibodies. 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 neurologic symptoms for a further three months. Patient completed full term of pregnancy and delivered healthy child.Treatment included ondansetron, ppi 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. 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].

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[6].

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 [6]. 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 [6].

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]. In our patient, cyclical vomiting and neurologic symptoms coexisted and were associated with PRES. Our patient was extensively investigated for endocrine, renal, gastrointestinal, neurological, cardiac and metabolic causes with no conclusive pathology. Supportive care involves use of intravenous fluids, sedatives, analgesia and the avoidance of potential triggering factors.

CONCLUSION

PRES is a rare disorder in pregnant female without eclampsia and hypertension. 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 pregnant female without eclampsia 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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