Posterior Reversible Encephalopathy Syndrome: Case Report

Mahesh Babu Sodalagunta, Sreenivasa Rao Sudulagunta, Monica Kumbhat, Mona Sephrrad, Aravinda Settkere Nataraju, Shiva Kumar Bangalore Raja

Abstract

Posterior reversible encephalopathy syndrome (PRES) is a clinico-radiological syndrome characterized by headache, seizures, altered mental status, and visual loss, as well as white matter vasogenic edema affecting the posterior occipital and parietal lobes of the brain predominantly. This clinical syndrome is increasingly recognized due to improvement and availability of brain imaging, specifically magnetic resonance imaging (MRI). A 35-year-old female with history of unsafe abortion and massive blood transfusion 10 days ago was brought to the emergency room with three episodes of generalized tonic clonic seizures, urinary incontinence and altered sensorium since 3 hours. Brain MRI showed bilateral occipital, parietal, frontal cortex and subcortical white matter T2/fluid-attenuated inversion recovery (FLAIR) hyperintensities, suggestive of PRES. Patient improved after management with intravenous fluids, antibiotics, antiepileptics and monitoring of blood pressure. If recognized and treated early, the clinical syndrome commonly resolves within a week. PRES can be a major problem in rapid and massive blood transfusion. High index of suspicion and prompt treatment can reduce morbidity and mortality, and pave the path for early recovery.

Keywords: Posterior reversible encephalopathy syndrome; Blood transfusion; Brain MRI; Antiepileptics

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinico-radiological syndrome characterized by symptoms including headache, seizures, altered consciousness, and visual disturbances [1, 2]. PRES was first described in 1996 by Hinchey et al [3]. Shortly after the description in 1996, two other case series were published [4, 5]. This condition has been known by various names previously (reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral edema syndrome, and reversible occipital parietal encephalopathy). PRES is now the widely accepted term [6, 7]. It is commonly but not always associated with acute hypertension [1, 2]. This clinical syndrome is increasingly recognized, commonly because of improvement and availability of brain imaging.

The major clinical conditions associated with PRES are represented in Table 1 [6]. There is wide variation in the severity of clinical symptoms, i.e., the visual disturbance can vary from blurred vision, homonymous hemianopsia to cortical blindness. Altered consciousness may vary from mild confusion or agitation, to coma. Other symptoms include nausea, vomiting and brainstem deficits. Seizures and status epilepticus are common, while non-convulsive status epilepticus may be more common than generalized status epilepticus [8].

Non-convulsive status should be cautiously observed in patients with prolonged altered consciousness, which may be mistaken commonly for postictal confusion. Signs include stereotypic movements like staring, head turning or eye blinking. Postictal confusion usually lasts for hours, but PRES and non-convulsive status can last for many days and can be mistaken for drug intoxication, psychosis or psychogenic states [9]. If recognized and treated early, the clinical syndrome commonly resolves within a week.

Case Report

A 35-year-old female was brought to the emergency room with...
history of three episodes of generalized tonic clonic seizures, urinary incontinence and altered sensorium since 3 h. Patient was referred from a local hospital for further management. Patient had headache prior to the onset of seizures. Patient relatives revealed that patient had unsafe abortion 10 days ago and later developed severe fatigue, shortness of breath and pedal edema. Husband expired 3 years ago due to unknown chronic illness. Due to severe anemia (hemoglobin: 3.4 g/dL) and ongoing blood loss, patient was transfused 5 units of blood in a local hospital. No history of hypertension was found as per medical records. No other significant history was available. On examination, vital signs of patient were normal. Patient was drowsy, not obeying commands, but withdrew limbs to painful stimuli, deep tendon reflexes were sluggish and a withdrawal response was seen in plantar reflex. There were no signs of meningeal irritation.

Laboratory examination revealed normal hemoglobin (13.8 g/dL), neutrophilic leukocytosis (18,000/dL), urinary
tract infection (10 - 15 pus cells/hpf) and elevated C-reactive protein (4.7 mg/dL). Erythrocyte sedimentation rate, liver function tests and renal function tests were normal. Periph-ereral smear showed dimorphic anemia. Other blood tests, coagulation profile, autoantibodies, and neoplastic markers were normal. Cerebrospinal fluid analysis revealed an increase in protein level (50 mg/dL). Chest radiography and arterial blood gas analysis were normal. Brain magnetic resonance imaging (MRI) showed bilateral occipital, parietal, frontal cortex and subcortical white matter T2/FLAIR hyperintensities (Figs. 1-5), suggestive of PRES. Electroencephalography (EEG) showed bilateral temporal-occipital epileptiform discharges at times becoming general. Electrocardiography (ECG) showed incomplete right bundle branch block.

Patient was admitted in intensive care unit and was managed with intravenous fluids, antibiotics, antiepileptics and monitoring of blood pressure. Patient improved symptomatically in form of normal sensorium, leukocyte counts and vital signs. Patient was discharged after 7 days of admission at request. Follow-up after 1 week was uneventful.

Discussion

The term PRES has been used based on the similarity in appearance on imaging, the common location of the parietal-occipital lobe or “posterior” location of the lesions [10, 11]. The exact pathophysiological mechanism of PRES is still unclear [12]. Three hypotheses have been proposed till now, which include 1) cerebral vasoconstriction causing subsequent infarcts in the brain, 2) failure of cerebral auto-regulation with vasogenic edema, and 3) endothelial damage with blood-brain barrier disruption further leading to fluid and protein transudation in the brain [12-14]. The distinct imaging patterns in PRES are represented in Table 2 [15]. The reversible nature of PRES has been challenged recently based on new reports of permanent neurological impairment and mortality reaching 15% [16, 17].

No clinical studies are available till now regarding patients with PRES needing life-sustaining treatments. The improved knowledge and research about factors influencing the outcome of PRES will result in better early management, less morbid-

Table 2. Imaging Patterns in PRES [15]

<table>
<thead>
<tr>
<th>Pattern</th>
</tr>
</thead>
<tbody>
<tr>
<td>Holohemispheric watershed</td>
</tr>
<tr>
<td>Superior frontal sulcus</td>
</tr>
<tr>
<td>Dominant parietal/occipital</td>
</tr>
<tr>
<td>Partial and/or asymmetric PRES</td>
</tr>
</tbody>
</table>

Figure 4. Coronal view of magnetic resonance imaging of brain showing bilateral occipital, parietal, frontal cortex and subcortical white matter hyperintensities.

Figure 5. Magnetic resonance imaging of brain showing bilateral occipital, parietal, frontal cortex and subcortical white matter T2/FLAIR hyperintensities.
ity and mortality. According to studies, delayed diagnosis and treatment may lead to mortality or irreversible neurological deficit [18, 19]. In hypertension-associated or drug-induced PRES, the effective therapy includes withdrawal of offending agent, immediate control of blood pressure, anti-convulsive therapy and temporary renal replacement therapy (hemodialysis/peritoneal dialysis) if required. In systemic lupus erythematosus (SLE)-related PRES, aggressive treatment with corticosteroids and cyclophosphamide is effective. Corticosteroids may improve vasogenic edema, but there is no solid evidence for usage in PRES.

Blood transfusion may cause a rapid increase in total blood volume, which further leads to cerebral blood flow overload. The case reports of PRES associated with blood transfusion are represented in Table 3 [20-26]. Abrupt or acute cerebral hyper-perfusion exceeding the capacity of auto-regulation of cerebral capillary perfusion pressure might result in vasogenic edema found in PRES. The possibility of severe anemia as the predisposing factor, due to inadequate supply of oxygen to the brain may result in dysfunction of endothelial cells, further causing a functional loss or damage to the integrity of the blood-brain barrier in capillary circulation which cannot be ruled out [26]. Our patient had no hypertension even transiently during the whole episode. In conclusion, PRES can be a major problem in rapid and massive blood transfusion. High index of suspicion and prompt treatment can reduce morbidity and mortality, and pave the path for early recovery.

**Abbreviations**

PRES: posterior reversible encephalopathy syndrome; SLE: systemic lupus erythematosus; FLAIR: fluid-attenuated inversion recovery

**References**


13. Shin KC, Choi HJ, Bae YD, Lee JC, Lee EB, Song YW. Reversible posterior leukoencephalopathy syndrome in systemic lupus erythematosus with thrombo-


