Posterior Reversible Encephalopathy Syndrome: Is it Always Posterior and Reversible?

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Abstract

Posterior reversible encephalopathy syndrome is a clinical and imagological acute problem characterized by impairment of consciousness, headache, cortical blindness and, epileptic seizures associated with grey and white edema principally localized in temporal-parietal-occipital areas, frequently related hypertensive pregnancy disease. However, not all the cases are associated with pre-eclampsia and eclampsia, and not all of them are related to posterior cerebral edema changes. In this paper is showing ten cases from a Mexican hospital, not all related to pregnancy, neither to posterior vasogenic edema in the image. The immunological patterns were an holo-hemispheric watershed, superior frontal sulcus pattern, dominant parietal-occipital pattern, and a partial or asymmetric abnormality in occipital region bilaterally. Besides the hypertensive disease of pregnancy, we found and hypertensive emergency, renal disease, sepsis, autoimmune illness as etiology. The most had a satisfactory evolution however one was discharged with the minimally conscious state, and another died with a final diagnosis of vasculitis associated with tuberculosis. Therefore, the final result is not always reversible, and neither the image shows a posterior characteristic vasogenic edema.

Keywords: Posterior reversible encephalopathy syndrome; Vasogenic edema; Hypertension; Hypertensive disease of pregnancy

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical-radiological syndrome characterized by acute neurological symptoms (altered mental status, headache, seizures, visual disturbances) and by neuroimaging findings of vasogenic edema in magnetic resonance imaging (MRI) [1]. The are several conditions related to PRES such as hypertension, preeclampsia or eclampsia, kidney disease, immunosuppressive therapy [2]. All of them have in common an injury of the endothelium [3].

Epidemiology: The classic clinical and radiologic presentation of PRES is posterior (i.e., parieto-occipital) [4] and usually these manifestations are reversible, however, has been described other patterns besides the classics [5]. There is no specific treatment for PRES, and sometimes, it is a challenge to eliminate or treat the related condition [1]. Mostly, PRES has excellent short and long-term prognosis [6].

This study aims to describe the clinical and imaging findings and prognosis in the Mexican population.

Methods

Patients who were diagnosed with PRES at Neurology Service at Hospital Central “Dr. Ignacio Morones Prieto” in México were enrolled in PRES database. Data were collected from medical records from 2016 to 2018 on demographics, PRES-related conditions, clinical manifestations, blood pressure measurements at arrival and during hospitalization (defining a hypertensive emergency as an arterial pressure above 180/120), neuroimaging, and clinical recovery time.
Neuroimaging

The radiological PRES patterns were reported on basis patterns reported by Bartynski 2007. There is Holohemispheric watershed pattern (HWP) characterized by vasogenic edema located in the frontal, parietal, and occipital lobes, with lesser involvement of the temporal lobes. Superior frontal sulcus pattern (SFSP): vasogenic edema is located along the mid to posterior aspect of the superior frontal sulcus. Dominant Parietal-Occipital pattern (DPOP): vasogenic edema is located (involved) on the parietal and occipital cortex and white matter with variable involvement of the temporal lobes. Partial or Asymmetric expression (PE) of the primary patterns defined as the absence of signal intensity abnormality in either the parietal lobes bilaterally or the occipital lobes bilaterally [7].

Results

We identified ten diagnosed cases, nine female and one male. Patients age ranged from 13 to 66 years (mean = 29.7). None of these patients had previous episodes of PRES.

Conditions associated with PRES

Drugs (n=1 [10%]): which were tracolimus and mycophenolate mofetil, preeclampsia or eclampsia (n=4 [40%]); all women were in their first pregnancy, hypertensive emergency at arrival (n=3 [30%]), kidney disease (n=4 [40%]), sepsis (n=1 [10%]), autoimmune disease (n=1 [10%]) which was systemic lupus [8].

Clinical manifestations

The most frequent manifestation was altered consciousness on nine patients (90%) with somnolence or stupor. Headache was present by six patients (60%), the pain was reported bilateral and diffuse in all of them. Seizures were present in 7 patients (70%), generalized in 62.5% and focal in 37.5%. There was one patient with status epilepticus. Focal neurological deficit was present in three patients (30%), one patient with blurred vision and two with hemiparesis [9-12].

Neuroimaging

The four patterns of PRES were found. The holohemispheric watershed pattern was the most frequent on seven patients (70%) Figure 1; the other three patterns were found in one patient each (Figure 2). The neuroimaging pattern observed on those who developed preeclampsia was holohemispheric in all of them (Figure 1 & 2) [13-17].

Description of two cases

Case 1

A 25-year-old female with 30 weeks of her first pregnancy presented to the emergency department with generalized seizures, one day before the admission developed sudden-onset of severe headache with duration of about 12 hours. Initially the diagnosis was Eclampsia during the management the blood pressure received aggressive treatment, despite of it persisted with somnolence and the MRI showed PRES with holohemispheric pattern. The patient was discharged home asymptomatic.

Case 2

A 48-year-old female with a medical history of diabetes, hypertension, pulmonary tuberculosis, chronic kidney disease; who was admitted to the hospital for two weeks with fluctuant
somnolence and hallucinations. At the day of hospital admission, the patient presented generalized seizures. CT of the brain was not relevant. The lumbar puncture unremarkable, the high blood pressure was difficult to control, developed a refractory status epilepticus and finally died. Her MRI showed a PRES with a holohemispheric watershed pattern. The final postmortem diagnosis was meningitis tuberculosis, there was no evidence of vasculitis (Table 1).

Table 1: General characteristics of the population showed.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Gender</th>
<th>Age</th>
<th>Conditions Associated</th>
<th>Clinical Features</th>
<th>Neuroimaging Pattern</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>66</td>
<td>Sepsis</td>
<td>Altered mental status</td>
<td>HWP</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>48</td>
<td>Hypertensive emergency</td>
<td>Altered mental status, focal seizures, GTCS</td>
<td>Partial expression.</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>44</td>
<td>Hypertensive emergency</td>
<td>Headache, focal seizure, altered status, hemiparesis.</td>
<td>SFSP</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>27</td>
<td>Hypertensive emergency</td>
<td>Headache, focal seizure, decreased visual acuity.</td>
<td>HWP</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>25</td>
<td>Eclampsia</td>
<td>GTCS, headache, altered mental status</td>
<td>HWP</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>22</td>
<td>Preclampsia</td>
<td>Hemiparesis, headache, altered mental status, GTCS.</td>
<td>HWP</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>21</td>
<td>Kidney disease</td>
<td>Altered mental status, headache.</td>
<td>HWP</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>16</td>
<td>Eclampsia</td>
<td>GTCS, altered mental status</td>
<td>DPOP</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>15</td>
<td>Autoimmune and kidney disease + drugs.</td>
<td>Altered mental status, GTCS</td>
<td>HWP</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>13</td>
<td>Preclampsia</td>
<td>Altered mental status, headache.</td>
<td>HWP</td>
</tr>
</tbody>
</table>

GTCS: Generalized Tonic-Clonic Seizure; HWP: Holohemispheric Watershed Pattern; DPOP: Dominant Parietal-Occipital Pattern.

**Discussion**

PRES has many faces and is not always posterior neither reversible. The clinical characteristics associated with arterial hypertension and encephalic dysfunction do not necessarily correlate with the location of the vasogenic edema seen on MRI, which usually has four characteristic patterns.

The description of the two cases emphasized the alteration of the mental state and the presence of seizures were the main clinical presentation, and in both, it was necessary to make a myriad differential diagnosis. The neuroimaging pattern was not the classic parietal occipital, and the prognosis in one of the cases was the worst with irreversible symptoms.

Also, we found a possible association between preeclampsia/eclampsia and holohemispheric watershed pattern.

There are many questions about the association between the pathophysiology PRES and its neuroimaging and prognosis; it is essential to conduct more studies with a more significant population.

**Conclusion**

Since PRES is an evolving concept with an underestimated diagnosis, we have to study our patients deeply. A lack of an MRI posterior pattern does not discard this syndrome.

**Acknowledgement**

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**Conflict of Interest**

No conflict of interest.

**References**


