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# A rare case of fatal intractable posterior reversible encephalopathy syndrome in a male child

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## Abstract

Posterior reversible encephalopathy syndrome (PRES) is a neurological condition characterized by disruption of cerebrovascular autoregulation. Diverse etiologies with acute hypertension as common component can lead to PRES. The condition is usually but not always reversible. Here we present a rare case of fatal PRES which shows radiological features of different stages of PRES from normal to overt intracerebral hemorrhage (ICH) and emphasizes the importance of radiologist familiarity with neuroimaging of different stages of this entity.

## Introduction

Posterior reversible encephalopathy syndrome (PRES) is a neurological disorder characterized by disruption of cerebrovascular autoregulation. Various causes with acute hypertension as common component can be associated with PRES (1). Some of the etiologies are preeclampsia/eclampsia, chemotherapeutic agents, acute glomerulonephritis and sepsis. The clinical signs/symptoms are increased blood pressure, headache, seizure, decreased mental status and visual disturbance (2). This condition as its name implies is usually reversible. However, in rare circumstances it can be progressive and fatal (3).

The computed tomography (CT) scan and magnetic resonance imaging (MRI) may be normal in initial stage of the disease. However usually CT scan shows hypodense foci and MRI shows hyperintensities especially in parietooccipital lobes (4).

Rarely this condition can lead to overt intracerebral hemorrhage (ICH) which can be seen on both CT scan and MRI. It is of critical importance to make the correct diagnosis PRES on neuroimaging as it can change the treatment of these patients (5).

Here we represent a rare case of fatal PRES which highlights the importance of radiologist familiarity with neuroimaging of different stages of this disease.

**Key point**

It is important for a radiologist to be familiar with different stages of PRES and suggest this diagnosis in appropriate clinical setting as it can change treatment strategy.

## Case Presentation

A 2-year and 7-month-old baby known case of nephrotic syndrome which was diagnosed a few months ago was admitted to hospital due to periorbital and testicular edema. The patient was treated with prednisolone, amlodipine and atorvastatin and full remission never occurred during this period. The blood pressure was 140/70 mm Hg and the creatinine was 1.35 on first day of admission. On fifth day of admission the blood pressure raised to 170/100 mm Hg and serum creatinine raised to 2.4 mg/dL. Additionally, the patient had abnormal gaze with loss of consciousness which was diagnosed as seizure and then he was intubated. Due to high blood pressure labetalol infusion was started and anticonvulsant drugs was also administrated. The pediatric neurologist requested brain CT scan which was reported as normal (Figure 1). Five days later focal seizure in right limb was noted and the blood pressure was 170/120 mm Hg. The second CT scan revealed hypodense lesions in frontal and parietooccipital lobes which are typical findings in PRES (Figure 2). Six days later (16 day of admission) the blood pressure was still high (150/100) and due to refractory status epilepticus MRI was conducted. The

**Figure 1.** Axial CT scan is unremarkable on first day of the seizure.

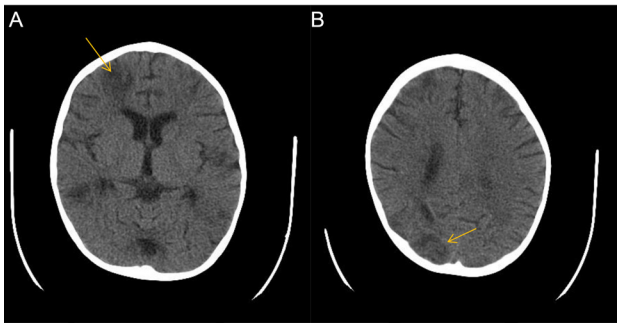


**Figure 2**. Patient's CT scan five days after the first seizure shows hypodense lesions in frontal lobe (arrow in A) and parietooccipital lobe (arrow in B) which are typical findings in PRES.

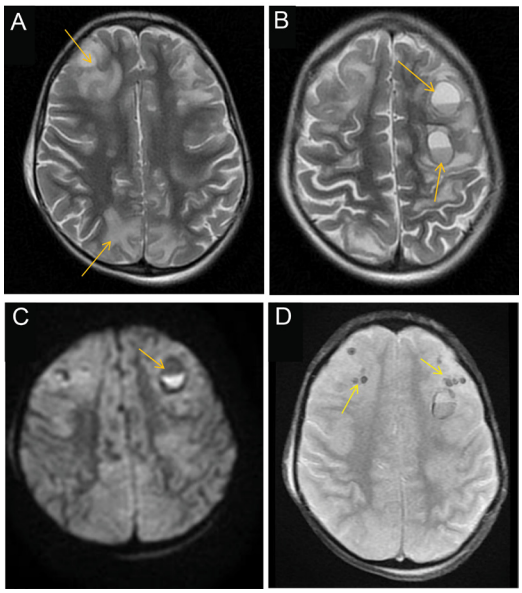


**Figure 3**. A: Axial T2 shows typical findings of PRES B: Axial T2 shows two lesions with fluid level (arrows) which are suspicious of abscess at first glance C: DWI shows the signal of the lesion to be high in dependent portion(arrow) which is suspicious but not typical for abscess as abscess shows more uniform increased signal. D: Axial T2 gradient shows multiple low signal foci (arrows) The overall findings are suggestive of hemorrhage rather than abscess.

MRI revealed multiple lesions with fluid level which could be mistaken for abscess. However careful evaluation of all sequences especially T2 gradient showed that these lesions are ICH with fluid formation (Figure 3). The last CT scan two days later confirmed that these lesions are ICH accompanied with intraventricular hemorrhage (IVH) and subarachnoid hemorrhage (SAH) (Figure 4). In the next day, due to severe bradycardia and decreased saturation, a cardiopulmonary resuscitation was conducted which was not successful and the patient expired.

**Figure 4.** Axial CT scan shows ICH, IVH and SAH (arrows).

## Discussion

Posterior reversible encephalopathy syndrome is a neurological disorder with disruption of cerebrovascular autoregulation. This condition is usually associated with increased blood pressure. Common etiologies are eclampsia/preeclampsia, chemo therapeutic agents, glomerulonephritis, and sepsis. The typical neuroimaging findings are low dense lesions on CT scan and high signal lesions on MRI especially in parietooccipital lobes. However, in early stages CT and MRI may be normal and rarely if the blood pressure is not controlled it can also lead to ICH. The diffusion-weighted imaging (DWI) sequence is usually normal in PRES unless it is complicated by hemorrhage or infarct (6).

In this case study, we introduced a rare case of a child with nephrotic syndrome and intractable increased blood pressure and status epilepticus. The first CT scan was normal however subsequent CT scan showed typical findings of PRES. As the disease could not be controlled MRI was performed and showed some confusing lesions with fluid level which could easily be mistaken for abscess however careful evaluation of all sequences depicted ICH as complication of PRES. These findings were confirmed on a subsequent CT scan. In this case, all stages of PRES were noted from normal to overt ICH. It is important for a radiologist to be familiar with different stages of PRES and suggest this diagnosis in appropriate clinical setting as it can change treatment strategy.

## Conclusion

It is essential for a radiologist to be familiar with different neuroimaging features of PRES as discussed in this case.

### Conflicts of interest

The author declares that he has no competing interests.

### Ethical issues

This case report was conducted in accord with the World Medical

Association Declaration of Helsinki. Patient has given us a written informed consent for publication as a case report. Ethical issues (including plagiarism, data fabrication, double publication) have been completely observed by the authors.

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