

## Recurrent and atypical posterior reversible encephalopathy syndrome in a child with hypertension

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### Abstract

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiologic entity with typical symptoms including headache, seizures, visual disturbance, altered mental status, vomiting, nausea and focal neurologic signs. In this article, we report recurrent and atypical PRES in a child with hypertension due to end-stage renal disease (ESRD) who was on a peritoneal dialysis program for 6 months. After the second hypertension attack, PRES findings did not recover and persisted as encephalomalacia. As far as we know, this case is the first child with ESRD who developed encephalomalacia after recurrent episodes of PRES. When a patient with a history of PRES presented with new clinical and neuroradiological findings, recurrent PRES should be considered.

**Keywords:** Child, end-stage renal disease, recurrent posterior reversible encephalopathy syndrome

### Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiological entity with typical symptoms including headache, seizures, visual disturbance, altered mental status, vomiting, nausea and focal neurologic signs, which was first described by Hinchey *et al.* in 1996.[1] The etiology of this condition includes sudden increase in blood pressure, renal failure, fluid retention, immunosuppressive drugs, eclampsia, sickle cell disease and intravenous immunoglobulin (IVIG) treatment.[1,2] Magnetic resonance imaging (MRI) findings are typically reversible and present as vasogenic edema in the parieto-occipital lobes. On the other hand, different regions of the brain, such as frontal lobes, cerebellum or brain stem, may be involved, and the lesions may persist in some cases.[3,4] Although knowledge about the pathogenesis of recurrent PRES is limited, infection and sudden increase in blood pressure are presumed to be the causes of the development of new lesions.[5]

In this case, we report recurrent and atypical PRES in a child with hypertension due to end-stage renal disease (ESRD) who was on a peritoneal dialysis program for 6 months. After the second hypertension attack, PRES findings did not recover, and the case evolved to encephalomalacia.

### Case Report

A 9-year-old girl with ESRD on a peritoneal dialysis program was admitted to our emergency department complaining of headache, vomiting, nausea, altered consciousness and generalized tonic-clonic seizure. She was on a continuous ambulatory peritoneal dialysis program for 6 months because of chronic renal failure due to chronic pyelonephritis. Her blood pressure was 170/110 mmHg (>95 p) and other vital signs were normal. In physical examination, she was sleepy and showed partial response to verbal stimulation. Her pupils were isochoric, and

direct-indirect light reflexes were normal. There was no limitation of her gaze. Deep tendon reflexes were hyperactive and Babinski sign was absent bilaterally. There were no findings of meningeal irritation. Her laboratory tests on admission were as follows: hemoglobin, 13.6 g/dL; hematocrit, 38.5%; white blood cells, 16300/mm<sup>3</sup>; platelets, 312000/mm<sup>3</sup>; blood urea nitrogen, 132 mg/dL; creatinine, 4.8 g/dL; Na, 133 meq/L; K, 5.3 meq/L; Cl, 109 meq/L; Ca, 8.8 mg/dL; and P, 5.9 mg/dL. Her liver function tests, total protein and albumin levels were all in normal ranges.

T2A-weighted and fluid-attenuated inversion recovery (FLAIR) MR images revealed hyperintense lesions on bilateral cerebellum and left temporale pole [Figure 1a and b]. Her electroencephalography (EEG) showed the slow rhythm of the background.

The patient was hospitalized in the pediatric intensive care unit and her blood pressure turned back to normal ranges with intravenous sodium nitroprusside infusion. Anticonvulsive and oral antihypertensive treatments were started and peritoneal dialysis cycles were increased as once per 2 h. Five days after admission, the patient recovered and was discharged without any deficiency. There was a significant recovery in the cranial MRI findings on the 3<sup>rd</sup> week of discharge [Figure 1c].

Five weeks after the first attack, the same patient was admitted to the emergency department with headache, altered consciousness and generalized tonic-clonic seizure. Her blood pressure was 210/130 mmHg, deep tendon reflexes were hyperactive and there was no focal neurologic deficit in physical examination. In the laboratory investigations, blood urea nitrogen was 54 mg/dL and creatinine was 10.1 mg/dL. Serum electrolytes and liver enzymes were in normal ranges. Oral antihypertensive agents (nifedipin, captopril, methyldopa) were started and peritoneal dialysis cycles were increased as once per 2 h. Because hypertension persisted despite oral antihypertensive medications, intravenous sodium nitroprusside infusion was administered. Although blood pressure was controlled with sodium nitroprusside infusion, hypertension attacks were observed during several attempts to cease sodium nitroprusside infusion. And, these attacks were accompanying seizures. Phenytoin, levetiracetam, valproic acid and midazolam were used for persistent convulsions. Ceftriaxon and acyclovir treatments were started on the second day of hospitalization because of fever. T2- and FLAIR-weighted images of cranial MR showed new hyperintense lesions in the bilateral parieto-occipital and frontal regions [Figure 1d and e].

Cranial MRI was repeated 2 months later, and it was shown that hyperintense lesions regressed significantly in the frontal and parieto-occipital lobes; however, a focal encephalomalacia evolved in the posterior part of the left parieto-occipital regions [Figure 1f]. The patient passed away from sudden cardiac event 6 months later.

## Discussion

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PRES usually presents with seizure, headaches, cortical blindness and other visual abnormalities, altered mental status and focal neurologic signs, and it is frequently observed in patients with sudden increased blood pressure due to renal failure.[1–3] Although the lesions are generally reversible with appropriate treatment, in unrecognized or not well-treated patients, lesions can progress to ischemia and infarction. This condition may cause neurological sequel such as epilepsy and permanent lesions on MR images.[6] Our patient had persistent hypertension attacks during the second admission, and MR images revealed encephalomalacia.

The etiology of PRES includes sudden increases in blood pressure and associated renal failure. Other reported causes of PRES are immunosuppressive (tacrolimus, cyclosporin A) and cytotoxic drugs, chemotherapeutic agents, fluid retention, eclampsia, IVIG treatment, sickle cell disease, systemic lupus erythematosus, polyarteritis nodosa, thrombotic thrombocytopenic purpura and hemolytic uremic syndrome.[1–3] Recurrent PRES is very rare in the literature, and ESRD is the underlying disease only in a few cases.[5,7] Among the patients with ESRD, our patient is the first case that had permanent encephalomalacia after recurrent PRES.

Although the exact mechanism is not well understood, pathophysiology of PRES involves two main mechanisms. Vasospasm due to acutely increased blood pressure and loss of autoregulation are supposed to be possible factors. In the first theory, it has been suggested that loss of autoregulation of cerebral blood flow contributes to ischemia and cytotoxic cerebral edema. In the second theory, because of high blood pressure, temporary failure of autoregulatory capabilities of the cerebral vessels leads to arteriolar vasodilatation, endothelial dysfunction and

hyperperfusion, and disrupted blood–brain barrier.[2,8,9]

In PRES, MRI findings are often very typical. The most common abnormality is edema involving the white matter in the bilateral parieto–occipital lobes of the cerebral hemispheres.[2,10] Although PRES typically affects the parietal and occipital lobes, in atypical PRES, lesions may develop in locations other than the parieto-occipital region. MRI lesions are hyperintense on T2-weighted and FLAIR sequences, and hypointense on T1-weighted sequences.[5–7] Although our patient had typical findings of PRES on MR images in her first attack, there were atypical MRI findings in the second attack.

In the management of PRES, early diagnosis and treatment is important. However, without rapid treatment, the functional vasogenic brain edema may lead to cytotoxic brain edema, which causes permanent brain injury and neurologic sequelae.[2,3,7] Treatment includes specific interventions to underlying etiology and supportive management of complications. In patients with hypertension, parenteral antihypertensive medication is very important. Sodium nitroprusside is the most preferred parenteral antihypertensive agent because of short activity and ease to titrate dose.[6–9,11] Our patient, with the typical findings of PRES, recovered with parenteral antihypertensive treatment (sodium nitroprusside) in her first attack. But, in the second attack, hypertension attacks were observed during the attempts to cease sodium nitroprusside infusion. We think that these hypertension attacks triggered convulsions so that we had to use multiple anti-convulsive agents.

Recurrent PRES have been rarely reported in the literature.[3,5–7] Onder *et al.* reported two recurrent PRES cases among 18 patients with kidney disease. In these patients, five of six attacks were due to high blood pressure and one attack was due to infection. After the recovery of lesions, factors that trigger the reoccurrence of new lesions are not clearly understood yet. Attacks may recur due to the same or different factors in a patient.[3] Sweany *et al.* and Girişgen *et al.* suggest that infections may be the factor behind the development of new lesions in patients with recurrent PRES.[5,6] Similarly, our patient had infection along with recurrent PRES attack.

As a result, our case is the first child with ESRD who developed encephalomalacia after recurrent episodes of PRES. We think that, in our patient, encephalomalacia had developed due to high blood pressure and seizures during recurrent PRES. When patients with a history of PRES present with new clinical and radiological findings, recurrent PRES should be considered.

## Footnotes

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**Source of Support:** Nil

**Conflict of Interest:** Nil

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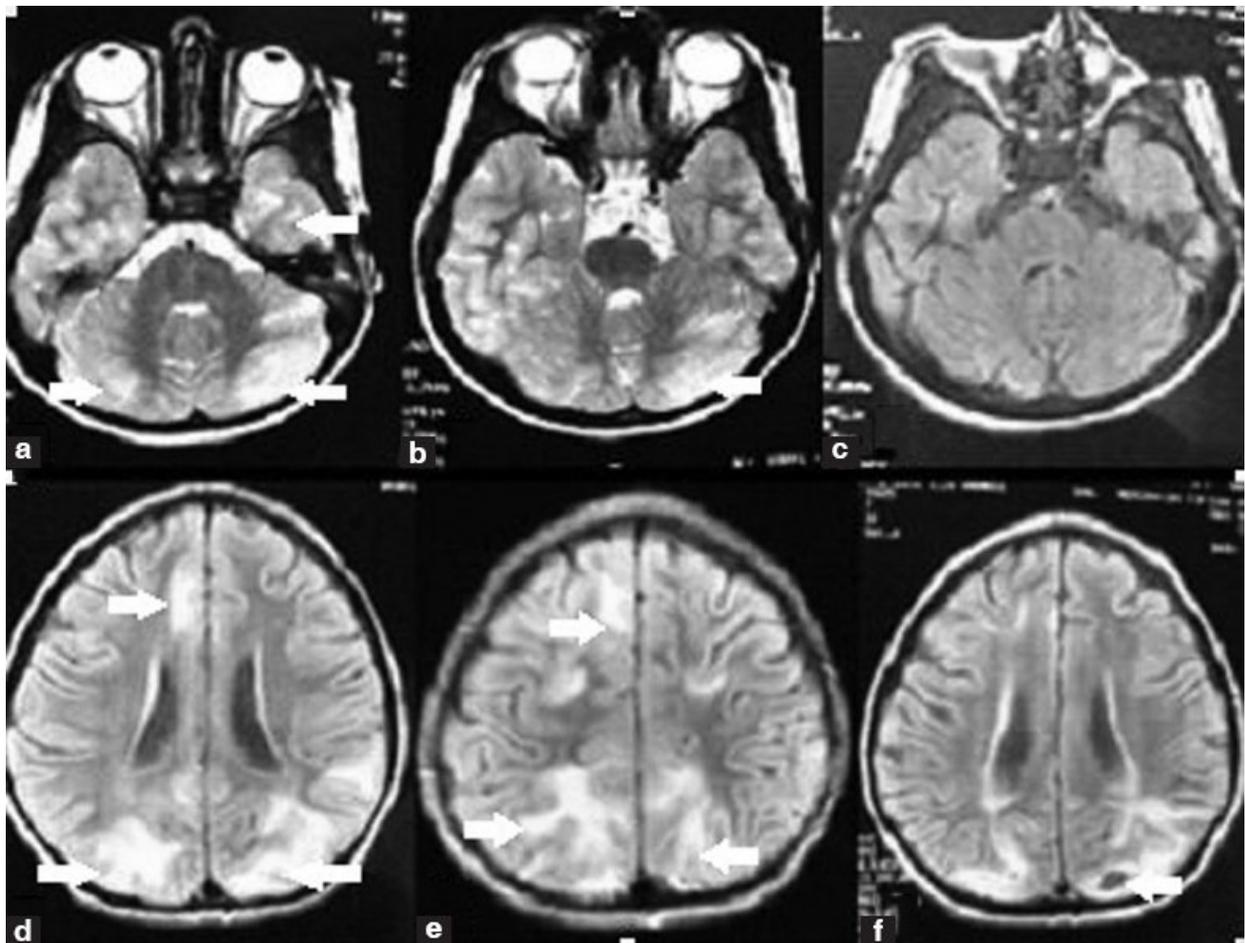
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## Figures and Tables

**Figure 1**



(a and b) T2A-weighted and fluid-attenuated inversion recovery (FLAIR) magnetic resonance (MR) images. Note the hyperintense lesions on bilateral cerebellum and left temporale pole. (c) Cranial MR images on the 3<sup>rd</sup> week of discharge are normal. (d and e) T2- and FLAIR-weighted images of cranial MR with new hyperintense lesions in the bilateral parieto-occipital and frontal regions. (f) Follow-up cranial MRI image with significantly regressed hyperintense lesions in the frontal and parieto-occipital regions, and a focal encephalomalacia in the posterior part of the left parieto-occipital region