

A case of reversible posterior leukoencephalopathy syndrome in a patient on peritoneal dialysis

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Abstract

Reversible posterior leukoencephalopathy syndrome (RPLS) is a recently identified clinical and radiologic entity. The characteristic radiologic findings are bilateral gray and white matter edema in the posterior regions of the cerebral hemispheres. The typical clinical syndrome includes headache, confusion, visual symptoms, and seizures. RPLS most often occurs in the setting of hypertensive crisis, preeclampsia, or with cytotoxic immunosuppressive therapy, but many other clinical settings are described, such as cryoglobulinemia, hemolytic uremic syndrome, SLE, and the use of erythropoietin. A 24-year-old man, diagnosed as having anaphylactoid purpura nephritis at 12 years of age and who started peritoneal dialysis (PD) at 23 years of age, was admitted to our hospital with a seizure and a consciousness disturbance. His blood pressure (BP) and body fluid volume had not been controlled well because of poor compliance with medication and PD. T2-weighted magnetic resonance imaging (MRI) revealed high signal intensity changes restricted to the cortex and subcortical white matter of the cerebellum. On the other hand, diffusion-weighted imaging (DWI) showed an isointense signal. From these findings, he was diagnosed as having RPLS. With appropriate control of BP and volume control by PD and hemodialysis (HD), his symptoms improved, and a follow-up cranial MRI 1 month later was almost normal. To the best of our knowledge, this is the first report of RPLS in an adult PD patient.

Introduction

The first series of patients with reversible posterior leukoencephalopathy syndrome (RPLS) was reported in 1996 by Hinchey et al.¹ The clinical syndrome of RPLS is characterized by headaches, altered consciousness, visual disturbances, and seizures. The visual disorders are frequently associated with neuroradiological findings, predominantly white matter abnormalities of the parieto-occipital lobes.¹⁻³ Reported comorbid conditions include hypertension (53%),² renal disease (45%), dialysis dependency (21%), malignancy (32%), and transplantation (24%).² In addition, untreated or undertreated chronic hypertension and renal failure have been implicated as causes of RPLS. Furthermore, the percent elevation of blood pressure (BP) over baseline is thought to be an important risk factor for RPLS.⁴ Neuroimaging is essential to the diagnosis of RPLS. Typical MRI findings are consistent with vasogenic edema and are predominantly localized to the posterior cerebral hemispheres. Diffusion-weighted imaging (DWI) can be particularly helpful in distinguishing RPLS from stroke.⁵

Patients with renal disease tend to have many risk factors for RPLS, such as hypertension, fluid overload, use of erythropoietin, and uremia. When a patient with renal disease has neurological symptoms, such as headache, visual symptoms, confusion, and seizures, clinicians should consider the possibility of RPLS in the differential diagnosis and order a brain MRI. DWI, if available, adds considerable diagnostic and prognostic information. Early diagnosis and

aggressive therapy are keys to a high chance of neurological recovery in patients with RPLS.

Here, we report the first case of RPLS in an adult peritoneal dialysis (PD) patient.

Case report

A 24-year-old man with end-stage renal failure secondary to Henoch-Schönlein purpura, who had been receiving maintenance PD for 6 months, presented with unconsciousness and a generalized seizure lasting 5 minutes. Though he should have been taking antihypertensive drugs and doing PD on a daily schedule of 4 × 2 L of 1.5% glucose dialysate, his BP and body fluid volume were not well controlled because of poor compliance with medication and PD. Although he had noticed a visual field disturbance 1 day before admission, he had ignored his symptom.

On the day of admission, he had respiratory failure, and oxygen 4 L/min with face mask was required to keep the SpO₂ at 99%. His BP was 170/108 mmHg, his pulse rate was 78/min, and his body temperature was 37.9°C. His level of consciousness was drowsy. Pupil size was normal, and the light reflex was prompt. There was no anemia of his palpebral conjunctiva. Since he had gained about 6 kg in weight over the past 6 months, his body weight was 60.3 kg. On chest examination, there were no rales or murmurs. The abdominal examination revealed no abnormal findings. There was no pretibial edema. There was no neck stiffness, pathological reflexes, or paralysis on neurological examination.

The laboratory tests showed hypoproteinemia, and the blood urea nitrogen and serum creatinine levels were 68 mg/dl and 13.02 mg/dl, respectively. Serum electrolytes were within

the normal ranges (Table 1). Chest X-ray showed cardiomegaly and pulmonary congestion, which suggested fluid overload (Fig. 1). To assess his altered level of consciousness, brain computed tomography (CT) was performed immediately, but there were no specific findings. Because the electroencephalogram revealed no specific changes, and there were no focal neurological symptoms, early cerebral infarction or cerebritis was considered. MRI was done for further investigation.

Fluid attenuated inversion recovery (FLAIR)-weighted and T2-weighted MRI showed increased signal intensity in the occipital lobes bilaterally, which was consistent with white matter vasogenic edema (Fig. 2). On the other hand, DWI showed an isointense signal. The findings on neuroimaging were characteristic of edema without infarction. From these findings, he was diagnosed as having RPLS.

He was started on aggressive BP management with intravenous administration of nicardipine. In order to improve volume status, an alternative osmotic agent, icodextrin, was added to the peritoneal prescription. As BP decreased, his consciousness improved and recovered to normal on the 4th day of admission. While he had lost 3 kg in body weight by the 4th day, his body fluid volume began to increase with improvement of his general condition and appetite, and his BP rose to over 150/80 mmHg. Although another oral antihypertensive drug was added, and the concentration of dialysate glucose was changed from 1.5% to 2.5% for high ultrafiltration

efficiency, his BP and body fluid volume could not be controlled. Thus, hemodialysis (HD) was added to PD to control body fluid volume and BP. In fact, transfer from PD to HD had been planned before this episode, because of poor compliance with PD and his low urine volume. Finally, PD catheter outflow failure was caused by omentum blocking the side holes of the catheter tubing, so he was transferred completely from PD to HD on the 27th day. His BP improved, and his fluid overload was gradually controlled by HD 3 times per week. On the 31st day, follow-up MRI showed complete resolution of abnormal findings (Fig. 2), and he was discharged from our hospital with no neurological deficit.

Discussion

We report here a case of RPLS with seizure and consciousness disturbance in an adult PD patient. To the best of our knowledge, this is the first report of RPLS in an adult PD patient. The previous case was a 12-year-old boy on PD, apparently triggered by erythropoietin-related hypertension.⁵

RPLS may develop in patients who have hypertension, renal failure, organ transplantation, autoimmune disease, or eclampsia, or those who are treated with cytotoxic anticancer drugs. An abrupt BP increase in patients with renal disease has been reported to be a particularly significant cause of RPLS.¹⁻³

Although the pathogenesis of RPLS remains unclear, it appears to be related to disordered cerebral autoregulation and endothelial dysfunction.⁴ Because of the heterogeneous nature of this syndrome, different mechanisms may be etiologically important in different clinical situations. With respect to autoregulation of cerebral blood flow, once the upper limit of cerebral autoregulation is exceeded, arterioles dilate and cerebral blood flow increases in a pressure-passive manner with rises in systemic BP. The resulting brain hyperperfusion, particularly in arterial border zones, may lead to breakdown of the blood-brain barrier, allowing extravasation of fluid and blood products into the brain parenchyma, which causes vasogenic edema.⁴ Endothelial dysfunction has also been implicated in the pathophysiology of RPLS,

especially in cases associated with preeclampsia, uremia, or cytotoxic therapies.^{6,7}

In the present case, renal failure, uncontrolled hypertension, and fluid overload due to inefficient PD were considered to have been involved in the onset of RPLS. Furthermore, the influence of recombinant human erythropoietin (rEPO) on the pathogenesis of RPLS could not be excluded in this case.

To date, it is not well understood why the lesions of RPLS tends to be found in the posterior brain region. One possibility is the regional heterogeneity of sympathetic innervation of the intracranial arterioles, which protects the brain from dramatic BP increases. Another is the lesser concentration of adrenergic nerves around pial and intracerebral vessels in the posterior region.⁸

Since the term 'RPLS' characterizes this disorder, RPLS is usually reversible within 2 weeks by immediate control of BP and discontinuation of immunosuppressive agents.³ When the diagnosis and treatment of RPLS are delayed, cerebral hemorrhage or cerebral infarction may occur, and the symptoms become irreversible.⁹ In this case, the patient was discharged with no neurological deficit because of early diagnosis and timely, appropriate therapy.

Generally, dialysis patients, including PD patients, have many risk factors for RPLS. Therefore, when dialysis patients with uncontrolled BP and fluid overload suddenly have a seizure, unconsciousness, headache, nausea, or visual disturbance, the possibility of RPLS should be considered in the differential diagnosis.

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Figure Legends



Fig. 1 Chest X-ray on admission. Chest X-ray shows cardiomegaly and pulmonary congestion, consistent with volume overload.

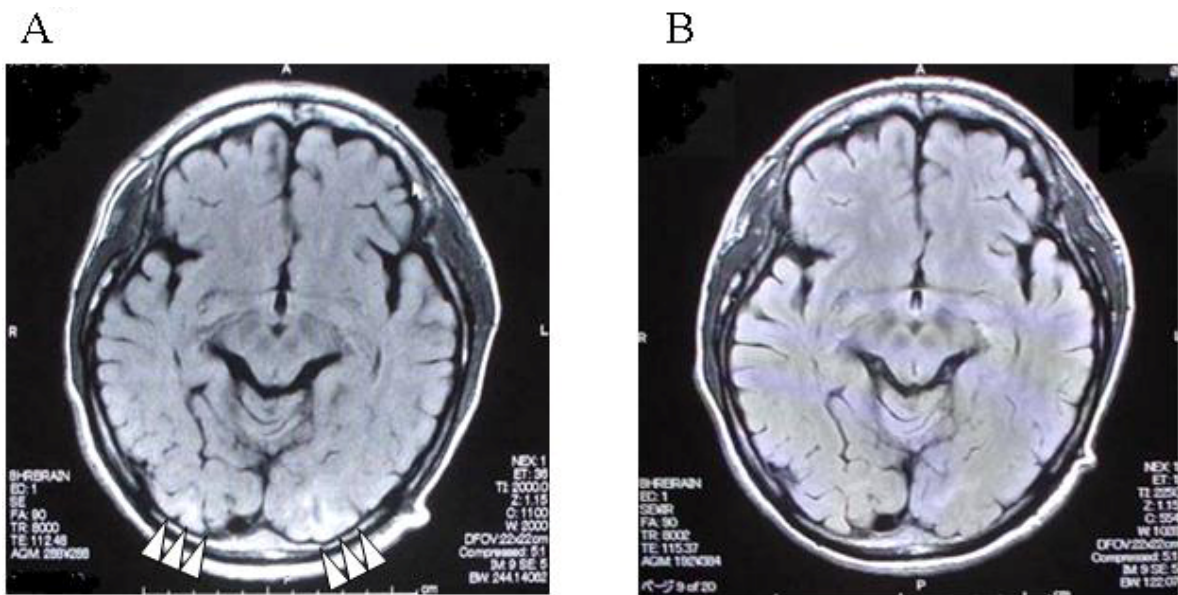


Fig. 2 Cerebral MRI (FLAIR)

A: MRI reveals abnormal increased signal intensity involving bilateral occipital lobes on admission (arrowheads).

B: MRI shows almost complete resolution of the bilateral occipital lobe lesions 1 month after admission.

Table 1

Laboratory data on admission

Hemogram		Others		Blood gas (face mask O₂ 4L/min)	
WBC	13.0 x10 ³ /mm ³	T.P.	6.3 g/dL	pH	7.401
Hb	10.8 g/dL	Alb.	3.2 g/dL	PaCO ₂	35.6 mmHg
RBC	4.15 x10 ⁶ /mm ³	BUN	68 mg/dL	PaO ₂	87.5 mmHg
Ht	34.4 %	Cre	13.02 mg/dL	HCO ₃ ⁻	21.6 mEq/L
Plt	165 x10 ³ /mm ³	Na	140 mEq/L		
		K	4.3 mEq/L		
		Cl	102 mEq/L		
		Ca	8.1 mg/dL		
		P	7.1 mg/dL		
		GLU	119 mg/dL		
		CRP	0.31 mg/dL		