Recurrent posterior reversible encephalopathy syndrome in a hypertensive patient with end-stage renal disease

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiologic entity characterized by headache, variable mental status, epilepsy, visual disturbances, and typical transient changes in the posterior cerebral perfusion. Recurrence of PRES is not common, but increasingly in recent years, studies demonstrate recurrence of this syndrome in populations with different diseases. In this report, we describe recurrent PRES in a hypertensive patient with end-stage renal disease, and discuss recurrence as the least-characterized feature of PRES. This condition can cause neurological sequelae such as persistent brain damage and epilepsy, arising from delays in diagnosis and therapy. To the best of our knowledge, this is the first report demonstrating recurrent PRES in a patient on hemodialysis for end-stage renal disease.

Key words: kidney failure, chronic • hypertension • recurrence • posterior reversible leukoencephalopathy syndrome

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiologic entity characterized by headache, variable mental status, epilepsy, visual disturbances, and typical transient changes in the posterior cerebral perfusion (1). Although reports of recurrence of this syndrome in different patient populations have increased recently in the literature, there is no information about recurrence of this syndrome in patients with end-stage renal disease undergoing hemodialysis (2). Here, we present a case of recurrent PRES in a patient with end-stage renal disease in a hemodialysis program, who responded well to treatment, and whose clinical and radiological (conventional magnetic resonance imaging [MRI]) findings improved.

Case report

A 23-year-old woman with end-stage renal disease was brought to our emergency department with complaints of headache, nausea, vomiting, loss of conscience, and tonic-clonic seizures involving her arms and legs. The patient had been undergoing hemodialysis three times per week. She had multiple congenital anomalies including agenesis of the left thumb, meningocele, ventricular septal defect, ectopic single kidney, and defective segmentation of the lumbosacral vertebrae. At the time of admission, she was in a state of post-ictal confusion, and she experienced seizures in the emergency department.

Following partial resolution of her confusion, she experienced headache and loss of vision. She was somnolent, and she showed partial response to verbal stimulation. She did not have findings of meningeal irritation. Her visual acuity was decreased, but she could perceive hand movements. Her pupils were isochoric, and direct and indirect light reflexes were normal. Ocular examination revealed that the borders of right disc were not clear. There was no limitation of her gaze. Motor and sensory examinations were normal. Deep tendon reflexes were equal and normoactive bilaterally, and Babinski sign was absent bilaterally. On admission, her blood pressure was 280/160 mmHg, which decreased gradually to 160/110 mmHg with antihypertensive medication.

The day after admission was the patient’s scheduled hemodialysis. Her laboratory tests on admission were as follows: hemoglobin, 11.2 g/dL; hematocrit, 33.4%; white blood cells, 8900/mm³, 62% segmented; platelets, 223,000/mm³; blood urea nitrogen, 54 mg/dL; creatinine, 15.1 g/dL; Na, 141 meq/L; K, 5.3 meq/L; Cl, 107 meq/L; P, 5.7 mg/dL (slightly elevated). Her liver function tests, total protein, albumin, and calcium levels were normal.

Computed tomography of the brain was normal, but there were slight impairment of baseline activity, and paroxysmal lateralizing epileptiform discharges on her electroencephalogram. MRI revealed edema in the posterior cerebral region, with preservation of the calcarine fissure...
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and paramedian lobe (Fig. 1). Diffusion MRI was not performed because this feature was not available at our hospital. Additionally, there were lesions in the subcortical white matter of both frontal lobes.

The patient was hospitalized in the intensive care unit, and her blood pressure was carefully monitored and controlled. With treatment for edema, anticonvulsive medication, hemodialysis every other day, and supportive care, her mean blood pressure was 120/80 mmHg, blood urea nitrogen 35 mg/dL, and creatinine 10.1 mg/dL (after hemodialysis). There was no electrolyte imbalance. Her vision had been restored completely by approximately one week after admission. Reevaluation with MRI three weeks later showed complete resolution of previously described abnormalities (Fig. 2). She did not experience any further seizures during her hospitalization.

The patient presented to the emergency department again six weeks after her previous admission, with the complaint of loss of vision. Her blood pressure was 240/130 mmHg, and her physical findings were normal other than a decrease in visual acuity. Her blood urea nitrogen was 54 mg/dL, and creatinine was 10.1 mg/dL. Her electrolytes and liver enzymes were normal. Cranial MRI revealed a new edematous lesion 2 cm in diameter at the right centrum semiovale (Fig. 3). The patient's visual disturbance improved in a short time with intravenous antihypertensive medications, hemodialysis, and supportive treatment. This lesion had resolved at follow-up MRI three months later. Cortical and subcortical changes attributable to chronic hemodialysis and end-stage renal disease were found on MRI two years later. Two additional years later, the patient died from a cardiovascular complication.

Discussion

PRES is characterized by mental impairment, headache, epilepsy, and visual disturbances, and is frequently seen in patients receiving chemotherapy. The lesions generally disappear with appropriate treatment, although reversibility of the lesions depends on the underlying disease and location of the lesions. In addition, different signal characteristics on MRI may affect the degree to which lesions appear to have resolved (3). Although there are reports in the literature concerning the development of new PRES lesions...
following complete disappearance of lesions from a previous episode, there is no information about this condition in patients with end-stage renal disease on chronic hemodialysis (2, 4, 5).

Hypertensive encephalopathy, eclampsia, immunosuppressive and cytotoxic medications, renal failure with hypertension, collagen vascular disease, thrombotic thrombocytopenic purpura, human immunodeficiency virus (HIV) infection, acute intermittent porphyria, and organ transplantation are among the known conditions associated with PRES (6). Of the limited cases of recurrent PRES in the literature, the underlying diseases were rheumatologic diseases in two cases (2, 5), bone marrow transplantation in one case, and sickle cell anemia in one case (2). Among patients undergoing chronic hemodialysis for end-stage renal disease, our patient is the first to have developed recurrent PRES.

In PRES, factors that trigger the development of new lesions following complete disappearance of the edematous lesions of an acute episode are not fully understood. Sweany et al. suggest that infections might trigger the development of new lesions in patients with recurrent PRES unrelated to inflammation (2); however, there was no infection in our patient at the time of development of the recurrent lesion.

Information about recurrent PRES is limited; however, the time period for the development of a new lesion is reported to be at least 30 days (5), and at most, two years (3). The time to recurrence was six weeks in our case.

Two possible mechanisms have been proposed in the pathophysiology of PRES. The first is vasospasm due to acutely increased blood pressure, and the second is loss of autoregulation. In the first hypothesis, it has been suggested that vasospasm contributes to ischemia and cytotoxic edema at regions of the arterial border zone (7). The second, more recent hypothesis is supported by diffusion images suggesting that dilation develops in cerebral arterioles due to autoregulatory failure. The objective of cerebral autoregulation is to keep blood flow constant, and to protect the brain during changes in blood pressure; however, sudden and severe increases in blood pressure can impair autoregulation, and such impairment can, in turn, lead to arteriolar vasodilation and endothelial dysfunc-

**Table.** Diagnostic criteria for posterior reversible encephalopathy syndrome (PRES) (12)

| 1. | The presence of neurological symptoms or findings such as epilepsy, weakness of an extremity, or mental status changes |
| 2. | Presence of risk factors for PRES such as cyclosporine treatment, eclampsia, or history of hypertension (diastolic blood pressure >100 mmHg) |
| 3. | Absence of other possible causes of encephalopathy |
| 4. | Reversible course, disappearance of signal changes on follow-up images, or complete resolution of clinical symptoms and signs following treatment |

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ebellar atrophy are seen frequently in patients with uremia, and may manifest as cortical or subcortical atrophy, or a combination of both. Histopathologic evaluation shows loss of neurons and nerve fibers. Cerebral atrophy in patients with uremia may be related to chronic anemia and a decrease in tissue oxygenation due to hypertensive heart failure. Vascular calcification due to hyperparathyroidism and/or dyslipidemia and systemic hypertension may also contribute to the development of atrophy via compromise of the vasculature of the brain.

Moreover, sudden pressure changes due to hemodialysis in hypertensive patients with volume overload may lead to brain injury (15). Although Al(OH)₃ is no longer a component of dialysate solution, previous intoxication from this compound remains one of the major causes of cerebral atrophy in patients undergoing hemodialysis.

First-line treatment of PRES is to regulate blood pressure. Mean arterial pressure should be reduced 20–25%, or diastolic pressure should be lowered to 100 mmHg within the first 1–2 hours. Rapid decrease in blood pressure should be avoided due to the risk of hypoperfusion and consequent cerebral infarction or impairment of organ function.

In this report, we presented the first case of recurrent PRES in a patient undergoing chronic hemodialysis for end-stage renal disease. Although PRES is a rare condition, in light of new clinical and radiological findings, the possibility of recurrence should be considered in various patient groups with a history of PRES.

References