CLINICAL CASE COMMENTARY

REVERSIBLE POSTERIOR LEUKOENCEPHALOPATHY SYNDROME IN A YOUNG ADULT PATIENT RECEIVING PERITONEAL DIALYSIS

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A 24-year-old Japanese woman who was receiving peritoneal dialysis (PD) was referred to our hospital because of generalized seizures and unconsciousness. She had been initiated on PD 3 years earlier, after uremia because of advanced reflux nephropathy had been diagnosed. Although the patient should have been following a day and night PD schedule of 4×1.2-L exchanges of 1.5% glucose dialysate and taking a phosphate binder, the active form of vitamin D, and so on, her uremia had not been well controlled for 2 months before admission because of poor compliance with PD and medication.

Before this admission, the patient's blood pressure had been 117/70 mmHg, 132/68 mmHg, and 153/88 mmHg (6 months, 3 months, and 1 month respectively). She showed mild hypertension 1 month before admission; however, her systolic blood pressure was elevated to around 200 mmHg on the morning of the reported episode. She had experienced severe headaches

and had taken analgesics several times on the day before admission.

On admission, the patient had a series of generalized seizures. Her level of consciousness was drowsy. Her blood pressure was 190/117 mmHg, her pulse was 144 bpm, and her body temperature was 36.8°C. There was no localized or generalized edema. Because administration of anticonvulsants had not stopped the seizures, artificial respiration was performed after intravenous anesthesia had been induced.

Laboratory tests revealed that her blood urea nitrogen was 80.4 mg/dL (28.7 mmol/L); serum creatinine, 13.6 mg/dL (1202.2 μmol/L); sodium, 140 mEq/L; potassium, 4.5 mEq/L; corrected calcium, 9.4 mg/dL (2.4 mmol/L); phosphorus, 8.7 mg/dL (2.8 mmol/L); magnesium, 3.2 mg/dL (1.3 mmol/L); total serum protein, 7.9 g/dL; serum albumin, 4.5 g/dL; and hemoglobin, 10.8 g/dL. Computed tomography imaging of the brain performed at this time did not reveal any major abnormalities. During further investigation by magnetic resonance imaging (MRI), fluid attenuated inversion recovery (FLAIR) and T2-weighted images revealed abnormalities in the posterior regions of the cerebral hemisphere, consistent with white-matter vasogenic edema [Figure 1(A)]. Diffusion-weighted imaging showed isointensity, also suggesting edema without infarction, and the patient was diagnosed with reversible posterior leukoencephalopathy syndrome (RPLS).

Aggressive antihypertensive therapy with intravenous administration of nicardipine was initiated, and the PD

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Figure 1 — Magnetic resonance imaging findings. (A) Axial fluid attenuated inversion recovery (FLAIR) images show multiple lesions in the posterior temporal and occipital lobes (arrows). (B) Follow-up FLAIR image taken 1 week after initiation of treatment shows complete resolution of the lesions.

dose was increased to control uremia. As the patient's blood pressure decreased, her consciousness improved, recovering to normal on day 2 after admission. On day 8 after admission, follow-up MRI showed complete resolution of the abnormal findings [Figure 1(B)].

The patient had mild abnormalities of the fundus because of hypertension (Scheie S1H1). Being free from seizures and neurologic deficits, she was discharged from our hospital on day 25 after admission.

DISCUSSION

Reversible posterior leukoencephalopathy syndrome was first described in 1996 in patients who had renal insufficiency or hypertension, or who were receiving immunosuppressants (1). Its clinical symptoms are characterized by severe headaches, visual disturbances, seizures, and altered consciousness. With regard to neuroimaging, MRI seems to be superior to computed tomography, with better sensitivity because of its high resolution and diffusion-weighted imaging. Patients with end-stage renal disease tend to have many risk factors, including hypertension, volume overload, use of recombinant human erythropoietin, and uremia. Indeed, RPLS is recognized in chronic hemodialysis patients both in the adult (2–4) and in the pediatric renal disease populations (5) and in children with nephrotic syndrome (6). Patients on hemodialysis with severe hypertension (7) and experiencing complications of erythropoietin therapy (2) were also described to have RPLS.

Although the pathogenesis remains unclear, hypertensive encephalopathy seems to be the cause of RPLS (1,2). A sudden elevation of systemic blood pressure exceeds the auto-regulatory capability of the brain vasculature, leading to vasoconstriction or vasodilation,

capillary leakage, and brain edema. The abnormalities on neuroimaging most likely reflect edema of the white matter in the parieto-occipital regions. The predilection for posterior circulation is unknown, but may be related the relatively poorer sympathetic innervation in that area, and thus poorer control of local cerebral auto-regulation. Endothelial dysfunction is another cause of RPLS, especially in cases associated with pre-eclampsia, uremia, and nephrotic states, and in patients receiving cytotoxic therapies (1,6).

In the present case, the sudden onset of hypertension and uremia because of inadequate PD were considered to be associated with the onset of RPLS. However, the effect of recombinant erythropoietin on the pathogenesis of RPLS could not be completely excluded, although this patient had received erythropoietin therapy for more than 3 years before the present admission.

The clinical reversibility of this syndrome has been extensively described (1,2,4–6,8). However, the functional vasogenic brain edema may convert to cytotoxic brain edema if proper intervention is not applied.

In adult patients on PD, RPLS was not recognized until recently (9). So far, RPLS seems to occur in PD patients in their teens and early twenties, as in the present case (9,10). However, all PD patients have many risk factors for RPLS. Therefore, PD patients—particularly young adults—presenting with severe headache and subsequent seizures and unconsciousness should undergo neuroimaging, including MRI, to rule out the possibility of RPLS and to ensure that an early and accurate diagnosis is made and that optimal treatment is administered.

DISCLOSURES

We declare that no financial conflicts of interest exist.

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