Recovery of severe dialysis disequilibrium syndrome with uncal herniation following therapy with mannitol, hyperventilation and hypertonic saline

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ABSTRACT

Dialysis disequilibrium syndrome (DDS) is a rare complication of dialysis, especially with the general application of preventive strategies. Severe DDS with brain herniation is believed to be fatal. We present a patient presenting with bilateral uncal herniation after receiving two dialysis sessions with low-efficiency settings. Serial brain magnetic resonance imaging studies showed the temporal evolution of DDS-induced cerebral edema. With aggressive treatment of hypertonic saline and mannitol, the patient made a remarkable recovery. This case highlights that we should be cautious about this severe complication of dialysis even with preventive strategies, and recovery is possible with prompt recognition and treatment.

Keywords: brain herniation, cerebral edema, dialysis disequilibrium syndrome, end-stage renal disease, hemodialysis, renal replacement therapy

BACKGROUND

Although the exact incidence of DDS is unknown, the incidence with low-efficiency setting seems to be declining, as preventive strategies are used during initiation of dialysis [3]. Here we present a case of severe DDS-induced brain herniation with unexpected recovery.

CASE REPORT

A 63-year-old woman presented to the emergency department with consciousness disturbance and involuntary movements a
few hours after her second hemodialysis course. Her long-term hemodialysis program was initiated electively 2 days before this episode. The indication for long-term hemodialysis was uremic symptoms of poor appetite and anorexia with a blood urea nitrogen level of 143 mg/dL and serum creatinine level of 10.7 mg/dL. The settings of the first and second hemodialysis are listed in Figure 1A. Her first hemodialysis course was uneventful. However, she experienced an episode of generalized convolution, which persisted for 1 min, ~2 h after the second dialysis session. Then, she became totally unarousable.

On physical examination, the patient was comatose with Glasgow Coma Scale, E1V1M1 and hypertension 174/94 mmHg. The height and weight were 160 cm and 44.4 kg. A neurological examination revealed bilateral loss of pupillary light reflex, flaccid muscle tone and bilateral pathological Babinski signs. Laboratory investigation showed hypernatremia (147 mmol/L); calcium level of 9.0 mg/dL with ionized calcium level of 1.19 mmol/L; and serum glucose level of 182 mg/dL. Arterial blood gas analysis revealed metabolic acidosis and CO₂ retention (pH, 6.995; HCO₃ level, 13.2 mEq/L; and partial pressure of CO₂, 55 mmHg).

Emergent computed tomography of the head revealed no intracranial hemorrhage. Magnetic resonance imaging (MRI) of the head showed profound cerebral edema with bilateral uncal herniations in T1-weighted images (Figure 1B, arrows). T2-weighted fluid-attenuated inversion recovery (FLAIR) images showed diffuse hyperintensity in the white matter (Figure 1C, upper). Based on clinical presentation and brain images, the tentative diagnosis was life-threatening DDS. The patient was intubated due to hypcapnic respiratory failure. Mannitol 20% (150 mL immediately + 75 mL every 4 h for 5 days) and phenytoin (750 mg immediately + 100 mg every 8 h) were administered, and hyperventilation was initiated for increased intracranial pressure. The third dialysis was performed ~60 h after the second dialysis with immediate intravenous 3% hypertonic saline 300 mL after the third dialysis for prevention of further deterioration of cerebral edema. The sequential change of serum biochemistry and osmolality are provided in Supplementary data (Figures S1–S3). On Day 21, the patient was successfully extubated. Two months later, she was responsive to verbal commands. Six months later, she was oriented and used a walker for ambulation. Serial MRI scans of the head were performed for follow-up, and they showed a dramatic resolution of brain lesions (Figure 1C-E).

**DISCUSSION**

Due to great variations of symptoms and severity, the prompt diagnosis of DDS is challenging. Although MRI of the head may be normal in mild DDS, it may be helpful in diagnosing severe...
DDS. In this case, MRI of the head revealed brain herniation, cerebral edema and hyper-intensity in white matter, which was compatible with the pattern of demyelination. The case highlights that, even with low-efficiency dialysis and mannitol infusion, severe DDS may happen and cause seizure, coma and brain herniation. In addition, severe DDS with brain herniation had been reported to be fatal [4]. Contrary to the previous report, our patient showed a remarkable recovery after treatment.

To our knowledge, this is the second report in the literature of a patient with DDS-induced brain herniation who survived and recovered [5] and the first case with brain imaging to document the temporal evolution of the DDS. Although previous literature reported that most treatments of DDS were futile, we believe that these patients should receive timely treatment since the build-up of osmotic gradient in the CNS system is transient and self-limiting once the dialysis is discontinued. Administration of osmotic active agents with hypertonic saline and mannitol will raise the serum osmolality and prevent further osmotic shifts. Due to the increasing number of patients receiving hemodialysis, the recognition and treatment of hemodialysis-related complications are paramount for patient safety.

PATIENT CONSENT
Informed consent was obtained from the patient’s family to publish this case.

SUPPLEMENTARY DATA
Supplementary data are available at ckj online.

CONFLICT OF INTEREST STATEMENT
The authors have no conflict of interest to declare.

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