CASE REPORT

A peritoneal dialysis patient with osmotic demyelination syndrome

Hing Ming Cheng*, Siu Hung Li

Department of Medicine, North District Hospital, Hong Kong

Available online 18 December 2015

KEYWORDS

osmotic
demyelination
syndrome;
peritoneal dialysis

Abstract

A peritoneal dialysis patient with cirrhosis presented with drowsiness, vomiting, and mild hyponatremia. Despite no active correction of hyponatremia, she developed convulsion and quadriplegia. Magnetic resonance imaging of the brain showed changes of osmotic demyelination syndrome. This case illustrates that osmotic demyelination syndrome may occur in peritoneal dialysis without rapid correction of hyponatremia.

Introduction

Osmotic demyelination syndrome (ODS) was most commonly reported after over-rapid correction of hyponatremia. It is uncommon in peritoneal dialysis patients. We report a case of peritoneal dialysis with ODS not associated with rapid correction of plasma sodium.

Case report

Madam C was a 50-year-old diabetic nephropathy patient treated with continuous ambulatory peritoneal dialysis. She had early cirrhosis due to hepatitis B but was a teetotaler. She was admitted for dizziness on July 5, 2013 and vomited undigested food once. There was no fever, diarrhea, nor abdominal pain. Her Glasgow Coma Score (GCS) was 5. No obvious focal neurological abnormality was found. Fundal examination was unremarkable despite a blood pressure of 210/120 mmHg. She had mild ankle edema but her jugular venous pressure was not elevated.

Three hours after admission, she developed generalized tonic–clonic convulsion which was aborted with intravenous diazepam after 3 minutes. Her temperature rose to 37.9°C. Her white cell count was 5.8 × 10^9/L. Her plasma sodium was 130 mmol/L. Her plasma potassium, calcium, phosphate, glucose, urea, and creatinine were 4.9 mmol/L, 2.28 mmol/L, 2.46 mmol/L, 5.6 mmol/L, 23 mmol/L, and 895 mmol/L, respectively. She had normal liver function tests and plasma ammonia level. Chest X-ray showed bilateral lower zone infiltration. Plain-computed tomography (CT) brain scan was unremarkable. Cerebrospinal fluid (CSF) protein was 0.5 g/L. CSF white cell count, glucose, Gram stain, Ziehl–Neelsen stain, and Indian ink stain were unremarkable. She was given intravenous phenytoin, acyclovir, ceftriaxone, sulperazon, and labetalol. Her hyponatremia was mild, and hypertonic saline was not administered. On Day 2, her GCS improved to 14. Her blood pressure was 187/108 mmHg. On Day 3, bedside electroencephalogram showed slow background waves with a
triphasic pattern compatible with metabolic or toxic encephalopathy. Intravenous valproate was added.

On Day 4, her GCS dropped to 8. She had no spontaneous limb movement but had bilateral brisk jerk, bilateral upgoing plantar reflexes, and bilateral ankle clonus. She was intubated for suspected status epilepticus. Intravenous propofol and levetiracetam were administered. Plain CT brain scan was repeated but was unrevealing. Electroencephalogram on Day 5 again showed triphasic pattern without ictal discharge. Cytology and polymerase chain reaction (PCR) of CSF for mycobacteria, herpes simplex virus, herpes zoster virus, and enterovirus were negative. Syphilis and Japanese B encephalitis serology were negative. Lead, manganese, and mercury levels in the blood were not elevated.

Magnetic resonance imaging (MRI) of the brain on Day 14 revealed T2 weighted hyperintensities and T1 weighted hypointensities in the central pons with sparing of peripheral tracts (Figure 1). The diagnosis was osmotic demyelination syndrome. She remained severely disabled and succumbed to sepsis 1 year later.

Discussion

We described a peritoneal dialysis patient with encephalopathy. Encephalitis may present similarly but was excluded by the CSF findings and negative serology and PCR tests for common infectious etiologies. Lead, mercury, and manganese poisoning were not supported by the blood levels. The MRI appearance, particularly the sparing of the peripheral tracts in the central pons, made osmotic demyelination syndrome more likely than pontine infarction.

The syndrome was previously named central pontine demyelinolysis. Adams et al reported four cases with alcoholism and malnutrition developing quadriplegia and pseudobulbar palsy. Autopsy showed symmetrical loss of myelin in the central pons. They postulated that the cause was a toxin or nutritional deficiency. Within a number of years, extrapontine lesions involving cerebellar peduncles, lateral geniculate nuclei, thalamus, basal ganglia, and midbrain were found, either alone or with the pontine lesions. With advances in neuroimaging, the diagnosis was not as rare as once thought. An incidence of 3/1000 of unselected urban hospital population was reported.

Tomlinson et al discovered the association of rapid correction of hyponatremia and ODS. Laureno rapidly corrected severe hyponatremia in dogs and reproduced the lesions. The syndrome did not occur when hyponatremia was corrected in a rate less than 10 mmol/L/day. It was postulated that abrupt osmotic changes lead to apoptosis of astrocytes, disrupting the blood brain barrier and allowing entry of myelinotoxic components like complements to cause demyelination. As the myelinolytic factors originate from the more vascular gray matter, areas where white and gray matter intermix (e.g. central pons) are more prone to the lesions. Tomlinson et al discovered the association of rapid correction of hyponatremia and ODS. Laureno rapidly corrected severe hyponatremia in dogs and reproduced the lesions. The syndrome did not occur when hyponatremia was corrected in a rate less than 10 mmol/L/day. It was postulated that abrupt osmotic changes lead to apoptosis of astrocytes, disrupting the blood brain barrier and allowing entry of myelinotoxic components like complements to cause demyelination. As the myelinolytic factors originate from the more vascular gray matter, areas where white and gray matter intermix (e.g. central pons) are more prone to the lesions.5,6 Our patient had plasma sodium of 127–130 mmol/L. However, her sodium level was not rapidly corrected. She had early hepatitis B cirrhosis, another established predisposing factor for ODS. Uremia was thought to protect against ODS. Nevertheless, Tarhan et al reported 17 renal failure patients with ODS. Hyponatremia and dialysis disequilibrium syndrome were present in 47% and 24% of this series. The initial encephalopathy of our patient improved but later deteriorated again. This biphasic pattern is also typical of ODS.

Though CT brain may reveal hypodense pontine lesions, the sensitivity is low because of beam hardening artifact. Diffusion weighted imaging (DWI) is more sensitive. Evidence of diffusion restriction like increased DWI signals and decreased apparent coefficient of diffusion values may appear in the central pons in a “trident pattern” within 24 hours of clinical onset. The changes improve significantly within the 1st week and revert to baseline in 3–4 weeks. T2 weighted hyperintensities in a “trident pattern” take 2 weeks to appear in the central pons. The volume and distribution of T2 signal abnormalities does not correlate with clinical severity and outcome. As symptoms resolve, the size and intensity of T2 abnormalities decrease. The classical "trident pattern" in DWI and T2 images is due to sparing of peripheral fibers and the axons of corticospinal tracts. T1 signal hypointensities may occur in acute settings but are unreliable. Gadolinium enhancement is
atypical but may occur in the periphery in a minority of cases. MRI abnormalities of ODS may occur in other regions like the cerebellum, lateral geniculate body, external capsule, thalamus, basal ganglia, the grey—white junction of the cerebral cortex, and hippocampi.

ODS can be prevented by avoiding over-rapid correction of hyponatremia. Relowering of plasma sodium prevented ODS in rat models. Treatment after the onset of ODS has not been established. Relowering of plasma sodium was effective in rat models and one case report. Plasmapharesis to remove the myelinotoxic components was useful in a small number of cases. Intravenous immunoglobulin, steroid, or thyrotrophin were used in isolated case reports but remain unproven therapies.

The outcome of our patient was poor but recent series showed better prognosis than previously thought. Singh et al. reported a mortality of approximately 25%, and showed better prognosis than previously thought. Singh et al. isolated case reports but remain unproven therapies.

The outcome may be favorable in survivors. MRI is useful for diagnosis. The outcome of our patient was poor but recent series showed better prognosis than previously thought. Singh et al. reported a mortality of approximately 25%, and there was a trend of improvement of outcome in recent years. More importantly, 52% of the survivors did not have significant disability. Louis et al. reported similar good functional outcome, even among the most severely affected individuals.

Osmotic demyelination syndrome may occur in peritoneal dialysis patients with only mild hyponatremia and without rapid correction. MRI is useful for diagnosis. The outcome may be favorable in survivors.

Conflicts of interest

All contributing authors declare no conflicts of interest.

Funding/support

None was received.

References

