Case Report

An Atypical Case of Uremic Encephalopathy with Bilateral Basal Ganglia and Cortical Involvement

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Abstract

Uremic encephalopathy (UE) is a metabolic disorder that occurs in patients with acute or chronic kidney disease. We report a case of 52 year-old-female with diabetic nephropathy for the past 5 years on maintenance hemodialysis (HD) presented with multiple episodes of seizures followed by altered sensorium. Magnetic Resonance Imaging (MRI) of the brain revealed T2 and Fluid Attenuated Inversion Recovery (FLAIR) hyperintensities with diffusion restriction in bilateral basal ganglia and cortex. Cerebrospinal Fluid (CSF) cytological, the biochemical analysis was normal. A provisional diagnosis of uremic encephalopathy was made. The patient underwent High flux hemodialysis. After 4 sessions of HD, she became conscious and oriented. Repeat MRI was done which revealed the disappearance of the diffusion-weighted lesion and improvement in T2 and FLAIR hyperintensities. UE is a toxic metabolic encephalopathy with typical neuroimaging features including vasogenic or cytotoxic edema in the cerebral cortex or basal ganglia region, producing clinical abnormalities, ranging from mild attention loss to coma. UE with predominant involvement of basal ganglia along with cortical involvement is uncommon and generally occurs in Asian patients with Diabetes Mellitus. Recognizing this syndrome is particularly important, considering its prognostic implications.

Introduction:

Uremic encephalopathy (UE) is a metabolic disorder that occurs in patients with acute or chronic kidney disease. **(**1, 2**)** This toxic encephalopathy is a result of endogenous uremic toxins, which may present as mild impairment of attention to unconsciousness. It may present as headache, alteration in sleep architecture and extrapyramidal disorders. **(**3, 4**)** The pattern of brain imaging can be a cortical, subcortical, white matter or basal ganglia involvement. Uremic encephalopathy specific imaging feature is also known as the lentiform fork sign. The lentiform fork sign although classically described in uremic encephalopathy, has also been reported in other metabolic disorders such as metabolic acidosis, methanol intoxication and dialysis disequilibrium syndrome. **(**2, 5**)** Involvement of basal ganglia is less common. Asian and diabetic patients are particularly susceptible to such involvement. **(**6, 7**)**

Here, we report a case of a patient with diabetic nephropathy patient with UE and acute bilateral basal ganglia and cortical lesions.

Case Report:

A 52 year-old-female with diabetic nephropathy for the past 5 years on maintenance hemodialysis (HD) presented with multiple episodes of seizures followed by altered sensorium. The patient was admitted and given antiepileptic medication. On examination, the patient was having an altered sensorium with a Glasgow Coma Scale (GCS) score of 7/15. She was intubated because of poor GCS. Her vitals were stable and routine blood investigations revealed urea of 335 mg/dl, creatinine of 7.2 mg/dl, calcium 7.2 mg/dl, sodium 135 mEq/L, potassium of 4.5 mEq/L. Magnetic Resonance Imaging (MRI) of the brain revealed T2 and Fluid Attenuated Inversion Recovery (FLAIR) hyperintensities with diffusion restriction in bilateral basal ganglia and cortex (Fig 1,2). Cerebrospinal Fluid (CSF) cytological, the biochemical analysis was normal. CSF neuroviral (Herpes Simplex, Japanese Encephalitis, Cytomegalovirus, Enterovirus) and the autoimmune panel were unremarkable.

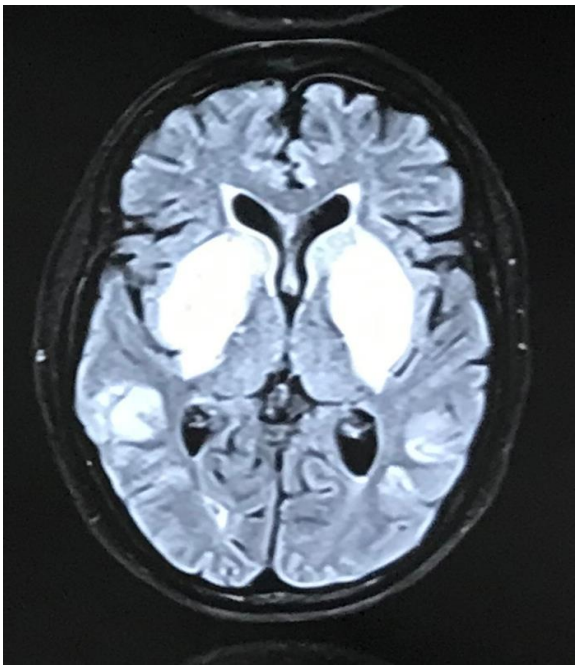

Figure 1 MRI brain FLAIR/T2 sequence showing bilateral basal ganglia and cortical hyperintensities.

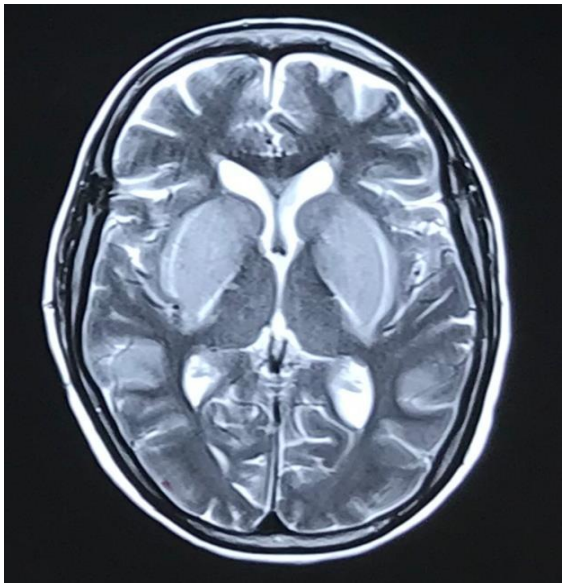

Figure 2 – MRI brain FLAIR/T2 sequence showing bilateral basal ganglia and cortical hyperintensities.

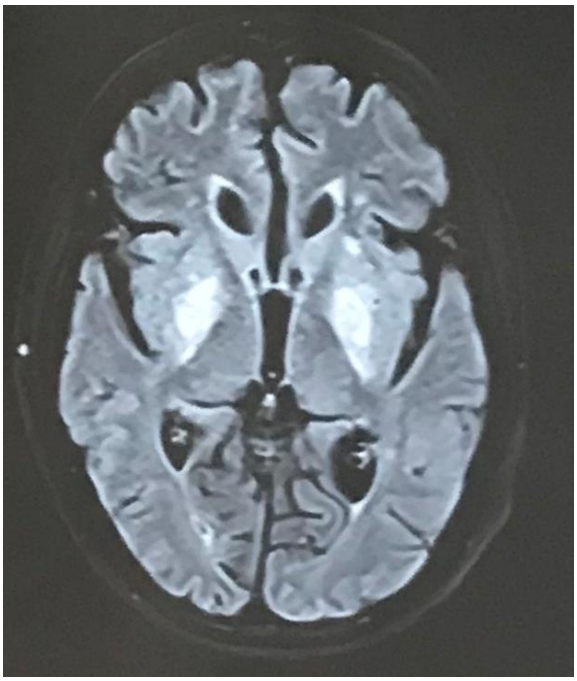

Fig 3 – MRI brain FLAIR sequence showing resolution of basal ganglia hyperintensities post hemodialysis.

A provisional diagnosis of uremic encephalopathy was made. The patient underwent High flux hemodialysis. Her sensorium improved slightly after the first session and she was weaned off mechanical ventilation. After 4 sessions of HD, she became conscious and oriented. Repeat renal function tests revealed a blood urea of 105 mg/dl and serum creatinine of 2.3 mg/dl. Repeat MRI was done which revealed the disappearance of the diffusion-weighted lesion and improvement in T2 and FLAIR hyperintensities (Fig 3). The patient was discharged to follow up in Nephrology OPD.

Discussion:

Uremic encephalopathy, a well-known complication of renal failure, constitutes a brain syndrome that may present either acutely or subacutely manifesting as various symptom complex. This complication usually results from brain edema. **(**8, 9**)**

The brain edema may be vasogenic or cytotoxic. The vasogenic edema involves the cortex and the clinicopathological findings are reversible with dialysis. Cytotoxic edema usually involves more metabolically active basal ganglia and is not fully reversible. **(**9**)**

The pathogenesis is incompletely understood. Various factors have been implicated including uremia, methyl guanidine, aluminum, changes in brain osmolality, hypo- and hyper-glycemia, fluctuations in blood pressure and metabolic acidosis. Possible mechanisms attributed are impairment of cerebral vascular autoregulation, changes in blood barrier permeability and cerebral oxygen consumption. **(**10**)**

UE is a toxic metabolic encephalopathy with typical neuroimaging features including vasogenic or cytotoxic edema in the cerebral cortex or basal ganglia region, producing clinical abnormalities, ranging from mild attention loss to coma. UE with predominant involvement of basal ganglia along with cortical involvement is uncommon and generally occurs in Asian patients with DM. Recognizing this syndrome is particularly important, considering its prognostic implications.

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