

A Case of Creutzfeldt–Jakob Disease in a Hemodialysis Patient

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Background: Creutzfeldt–Jakob disease (CJD) is characterized by encephalopathy with rapidly progressive cognitive dysfunction, myoclonus, and akinetic mutism. Although some cases with Creutzfeldt–Jakob–like syndrome due to drug intoxication has been reported, CJD itself has not been reported in hemodialysis patients with yet. Here, we report a hemodialysis patient with altered mental status, of which the cause is CJD.

Case: A 59–year old woman with a past history of hypertension and end–stage renal disease (ESRD) presented to our facility with stuporous mental status accompanying progressive motor aphasia for 2 months, myoclonus for 1 month, and akinetic mutism which began 3 days before hospitalization. Initially, the cause of altered mental status was assumed to be uremic or hypertensive encephalopathy combined with fever. But the proper managements did not improve the neurologic symptoms. Diffusion–weighted MRI demonstrated bilaterally asymmetric increased signal intensity lesions in both basal ganglia (putamen, caudate nucleus) and cortical portion of both cerebral hemispheres, suggesting Creutzfeldt–Jakob disease. An electroencephalogram (EEG) showed diffuse generalized theta to delta range slow wave on both hemispheres. There were also medium–to–high voltage complexes with a characteristic triphasic pattern. Cerebrospinal fluid (CSF) tapping for 14–3–3 protein was done as a confirmatory test.

Conclusion: Patients with ESRD often present to medical facilities with alteration of mental status. While CJD is incurable and shows poor prognosis, alternative diagnoses which may exhibit similar symptoms, but most of which are reversible with proper treatments should be excluded first. Through review of medical history including drug history, routine laboratory studies, EEG, brain imaging studies, especially brain MRI, and if necessary, CSF examination should be considered for differential diagnosis.

Key Words: Creutzfeldt–Jakob 질환, 투석, 의식장애

Creutzfeldt–Jakob disease, Hemodialysis, Mental status