Diffusion-weighted MRI for Differentiating Creutzfeldt-Jacob Disease Mimics

Masatsugu Takano, Keisuke Suzuki, Naoki Izawa and Koichi Hirata

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Sporadic Creutzfeldt-Jacob disease (CJD) is a rare neurodegenerative, prion disease characterized by rapidly progressive dementia, visual disturbances, myoclonus and extrapyramidal and pyramidal involvement. An 80-year-old woman presented with progressive dementia and visual and auditory hallucinations over a two-month period. Neurological examination revealed cognitive impairment and right hemiparkinsonism. Diffusion-weighted MR imaging (DWI) revealed high intensity lesions in the left putamen, caudate and cerebral cortex (Picture 1A). $^{99m}$Tc-ECD SPECT images showed hypoperfusion in the left fronto-temporal cortex and basal ganglia (Picture 1B). Cerebrospinal fluid (CSF) examination revealed no pleocytosis but showed increased neuron-specific enolase levels and positive 14-3-3 protein. Electroencephalography results showed diffuse slow waves with periodic discharges lateralized to the left hemisphere. The diagnosis of probable CJD was made based on clinical manifestations and CSF and imaging findings (1). Because CJD can present with various neurologic signs owing to differences in brain involvement and disease progression, nonconvulsive status epilepticus, encephalopathy, and other neurodegenerative diseases, especially in the early phase, can mimic CJD (2). The presence of hallucination with asymmetric parkinsonism initially suggested dementia with Lewy bodies or corticobasal degeneration. In the study using DWI, the value of apparent diffusion coefficient maps can be useful for the diagnosis of atypical parkinson syndrome (3); however, the findings that high signal intensity lesions observed in striatal and caudate in addition to diffuse cortical cortex on DWI strongly suggested CJD. Moreover,
there has been a report of a case of CJD in whom only DWI but not fluid-attenuated inversion recovery detected high-intensity lesions (4). We suggest that applying DWI is useful for the clinical diagnosis of dementia disorders.

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References