A CASE OF CREUTZFELDT-JAKOB DISEASE MISDIAGNOSED AS STROKE

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Creutzfeldt-Jakob disease (CJD) is a degenerative disease induced by prion and characterized by a rapidly progressive dementia but different patterns of initial presentation have been identified. We describe a case of probable CJD presenting as stroke-like episode and misdiagnosed as acute ischemic stroke. A 77-year-old man was admitted to our hospital for a 4-week history of progressive hemiparesis. One month before the admission, the patient developed acute onset of dysarthria and left hemiparesis (MRC grade 4). Initially he had been diagnosed with acute ischemic stroke in other hospital based on the clinical presentation and finding of diffusion-weighted MRI which suggested an acute ischemic lesion in the right insular area. He had a history of arteriosclerosis obliterans of the lower extremities and PCI for multi-vessel coronary disease. The neurological symptoms were stable for two weeks but the hemiparesis aggravated and the spontaneity and responsiveness decreased progressively over the next two weeks. He became mute and developed myoclonic jerk of the extremities within 6 weeks after the onset. Serial diffusion-weighted MRIs showed that high intensity lesions, which initially limited to the right insular cortex, spread to the bilateral cerebral cortices. The CSF 14-3-3 protein was found to be positive. This case demonstrates that CJD can present with acute focal neurological deficit mimicking stroke by selective involvement of brain initially.