P-52 One sporadic CJD with rapidly progressive mental deterioration as the first presentation

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[Introduction]

Creutzfeldt Jacob disease (CJD) is a rare neurodegenerative disease that has rapidly progressive mental deterioration and myoclonus. About only 100 cases are reported in Japan every year. We experience one sporadic CJD with rapidly progressive mental deterioration as the first presentation

[Case presentation]

The case is a 72 year-old man, who had aseptic meningitis one year before the admission day, with rapidly progressive mental deterioration within one and half month. On the admission day, he had difficulty in walking and pathological reflection was seen. Deep tendon reflex was exaggerated. Scattered areas of T2 and FLAIR signal hyper-intensity are seen in the cerebral cortex in MRI imaging. We suspected CJD from the clinical features and the MRI image. 14-3-3 protein was detected in his spinal fluid and abnormal prion was detected with RT-QUIC methods. The PRNP genotype was homozygous for methionine at codon 129 and we diagnosed him as sporadic CJD. After the admission, myoclonus and startle phenomenon was observed and periodic synchronous sharp wave complexes could be seen in the second electroencephalogram (EEG). The tonic-clonic convulsions appeared and he could not eat anything. He was transferred to a different hospital with the intravenous drip.

[Discussion]

We experienced one sporadic CJD with rapidly progressive mental deterioration as the first presentation. Our hospital had four CJD cases including it in seven years and the symptoms progressed inexorably once they appear. We could not diagnose it properly at their first visits in every case. Once we see rapidly progressive mental deterioration or involuntary movements, we should suspect CJD and take MRI images and EEG.