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Short Neuropathology Report

Immunohistochemistry of abnormal prion protein PrP^{Sc} in an autopsy case of sporadic type MM1 + 2 Creutzfeldt-Jakob disease (CJD)

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Abstract

We report an autopsy case of an 80-year-old man, clinically diagnosed as a probable CJD. The clinical course of the disease was 3 years and 8 months with the acute progression of dementia during the first 8 months. The clinical characteristics were closer to those observed in type MM2 rather than type MM1. Genetic and Western blot analyses of the abnormal prion protein (scrapie prion protein, PrP^{Sc}) confirmed this case as type MM1 + 2. With the exception of the cerebellar lesions, the immunohistochemical characteristics of the PrP^{Sc} were almost consistent with type MM1 + 2. In this report, we discuss particularly whether the cerebellar lesions in the present case are characteristic of type MM1 + 2. Tottori J. Clin. Res. 9(2), 176-189, 2017

Key words: sporadic Creutzfeldt-Jakob disease (sCJD), type MM1 + 2, scrapie prion protein (PrP^{Sc}), synaptic pattern, perivacuolar pattern, coarse granular deposition, plaque-like pattern, infectious CJD

Clinical course

We report an autopsy case of sporadic CJD (sCJD) in a male patient over a clinical course of 3 years and 8 months with the acute progression of dementia during the first 8 months, and death at 80 years old.

The patient's history revealed an operation for retinal detachment, but no other operations, endoscopies, blood transfusions, or acupuncture were reported. He had visited the United States twice but not Europe. He first noticed memory impairment at the age of 76 years. Three months later, he visited a Neurology Clinic, where he scored 23/30 by using the revised Hasegawa Dementia Scale (HDS-R). Thereafter, his HDS-R score rapidly decreased to 14/30 and 3/30 at 5 and 8 months after the onset, respectively. Thus, despite a lack of ataxia, dementia progressed rapidly during the first 8 months. The patient experienced hallucinations, optical illusions, and emotional and behavioral abnormalities 8 months after the onset, and visual agnosia, visuospatial agnosia, and sensory aphasia after 9 months. He exhibited akinetic mutism after 1 year and 3 months. At 1 year and 6 months, myoclonus began to be observed, but periodic synchronous discharges (PSDs) were always unclear in electroencephalo- grams (Fig. 1). However, both diffusion-weighted images (DWIs) and fluid-attenuated inversion recovery (FLAIR) images of magnetic resonance imaging (MRI) studies performed 3 and 9 months after the onset had already revealed ribbon-like-shaped, high signal intensity areas from the bilateral temporal cortical regions to the cortices of the bilateral occipital lobes, thus suggesting CJD (Fig. 2A). In DWIs on MRI studies performed 2 years and 5 months after the onset, the caudate nuclei and putamen were visualized as high signal intensity areas, in addition to the areas in the cerebral cortices. At that time, T1-weighted images (T1WIs) on MRI studies revealed marked cortic-