

脳外科手術歴を有するCreutzfeldt-Jakob病の特徴

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Table. Patients with history of neurosurgery who had no periodic sharp-wave complexes during whole disease duration of Creutzfeldt-Jakob disease.

Pt.	Age at onset	Sex	Disease of neurosurgery	Period of neurosurgery, years	Incubation period*, years	Initial symptom	Disease duration**, months	Codon 129 of PrP	Hyperintensity lesions on DWI	Pathological findings
1	75	Male	Head trauma	1977	30	Dementia	11	Met/Met	CC, BG	ND
2	49	Female	Subarachinoid hemorrhage	1985	9	Insomnia	28	Met/Met	ND	MM2-thalamic type sCJD
3	63	Female	Tumor	1985	27	Gait disturbance	19	Met/Met	BG, Th	ND
4	75	Female	Tumor	1985	14	Drowsiness, gait disturbance	6	Met/Met	BG, Th	CJD-MMiK
5	64	Female	Subdural hematoma	1993	10	Visual impairment	21	Met/Met	CC	MM2-cortical type sCJD

*Incubation period: period between dura mater grafting and onset of Creutzfeldt-Jakob disease, **Disease duration: duration between the onset of CJD and the appearance of the akinetic mutism or death in the patients who died without akinetic mutism.

Pt.: patient, PrP: prion protein gene, PSWCs: periodic sharp-wave complexes, EEG: electroencephalogram, DWI: diffusion-weighted images, ND: not done, CC: cerebral cortex, BG: basal ganglia, Th: thalamus

Figure 1

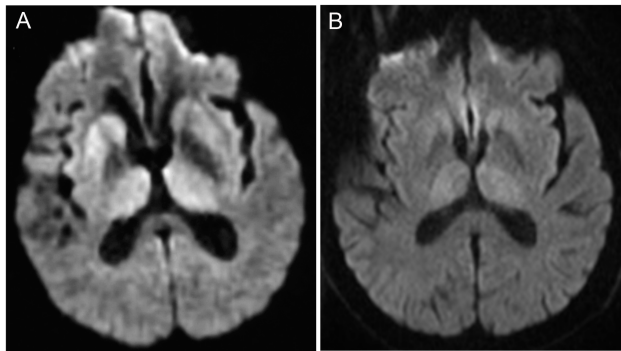


Figure 1. Findings of diffusion-weighted images (DWI) of the brain. A: DWI of Patient 4 at 8 months after the onset of Creutzfeldt-Jakob disease (CJD), who were diagnosed as acquired CJD-MMiK by autopsy, revealed hyperintensity lesions in bilateral thalamus in addition to basal ganglia lesions. B: DWI of Patient 3 at 9 months after the onset of CJD revealed hyperintensity lesions in bilateral thalamus in addition to basal ganglia.

解 説

1. 孤発性CJDまたは分類不能のCJDと診断されている症例の中には、CJD-MMiKと似た臨床・病理所見の特徴を有する症例がある。
2. 頭部MRI DWIでの視床高信号はCJD-MMiKの臨床診断マーカーとなるかもしれない。