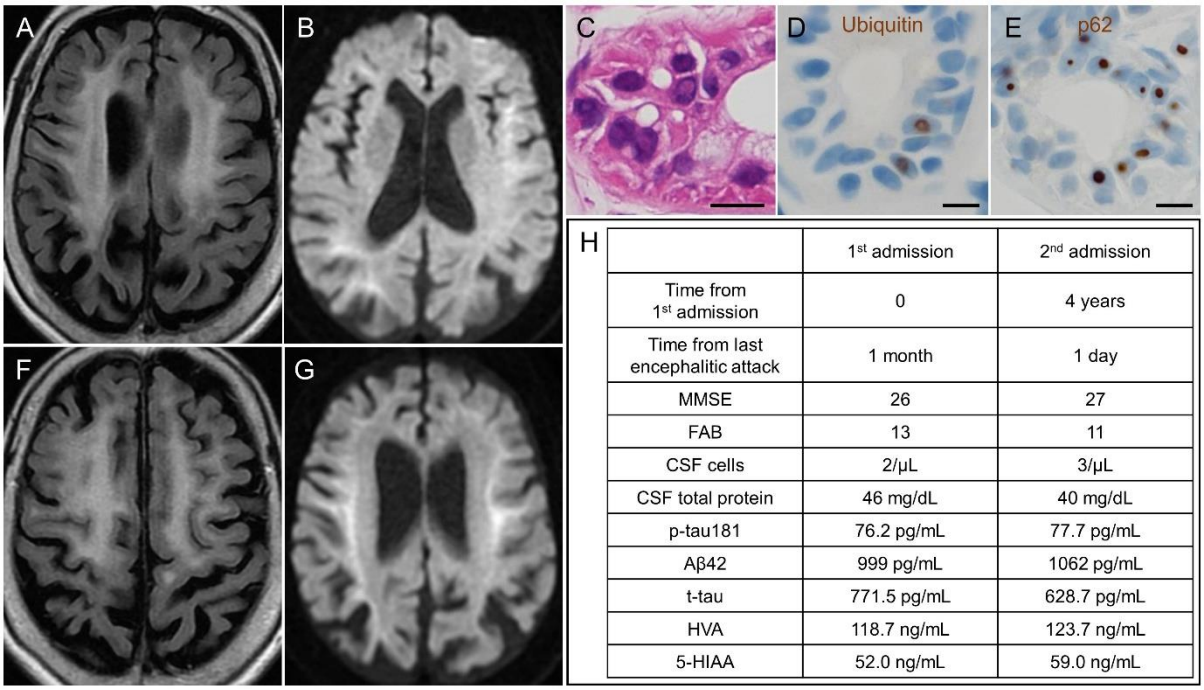

H

	1st admission	2nd admission
Time from 1st admission	0	4 years
Time from last encephalitic attack	1 month	1 day
MMSE	26	27
FAB	13	11
CSF cells	2/μL	3/μL
CSF total protein	46 mg/dL	40 mg/dL
p-tau181	76.2 pg/mL	77.7 pg/mL
Aβ42	999 pg/mL	1062 pg/mL
t-tau	771.5 pg/mL	628.7 pg/mL
HVA	118.7 ng/mL	123.7 ng/mL
5-HIAA	52.0 ng/mL	59.0 ng/mL

eFigure 1. Representative test results of Patient 7.

The patient started to experience transient fever, dizziness, and vomitting once a year in her 50s. At the age of 70, the patient experienced fever, impaired consciousness, and abnormal behavior. The fever resolved the next day, and her consciousness gradually improved to baseline in the following weeks. The patient presented to our clinic one month after this "encephalitic" attack and was admitted for evaluation. Brain MRI showed right-dominant diffuse FLAIR high-intensity signals in the white matter (A). Diffusion-weighted imaging (DWI) showed high-intensity signals along the corticomedullary junction (B). Hematoxylin and eosin staining of a skin biopsy specimen showed eosinophilic nuclear inclusions (C). These intranuclear inclusions were stained by antibodies against ubiquitin (D) and p62 (E). Scale bars = 10 μm. The patient continued to experience transient episodes of dizziness, vomitting, or mild impaired consciousness. The patient experienced mild cognitive decline and began to require assistance in preparing dinners, but remained independent in other daily activities during remission. Four years later, the patient was re-admitted because of fever, impaired consciousness, abnormal behavior, and motor aphasia. Brain MRI continued to show leukoencephalopathy with DWI high signals along the corticomedullary junction (F, G). Symptoms improved the next day. CSF was obtained during these two admissions as a workup, and biomarker analyses were conducted using the remaining CSF as a research. Baseline, cognitive, and CSF parameters are summarized (H). All the CSF parameters were similar despite the difference in the timing of CSF obtainment (H).