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RESEARCH PAPER

Specific early electroencephalogram for the diagnosis of sporadic Creutzfeldt-Jakob disease

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ABSTRACT

An early diagnosis is required for intervention in prion disease cases. To elucidate the specificity of early electroencephalography discharges in cases of sporadic Creutzfeldt-Jakob disease, we analysed epileptiform discharges through electroencephalography. Nine patients with methionine/methionine type 1/classic sporadic Creutzfeldt-Jakob disease and 20 patients with status epilepticus were included. Generalized periodic discharges, lateralized periodic discharges, and central sagittal sporadic epileptiform discharges were evaluated. Central sagittal sporadic epileptiform discharges were defined as nonrhythmic and nonperiodic waveforms showing generalized spike-and-wave complexes and/or sharp waves predominantly in the central sagittal region. In the sporadic Creutzfeldt-Jakob disease group, central sagittal sporadic epileptiform discharges, lateralized periodic discharges, and generalized periodic discharges were observed in five (55.6%), one (11.1%), and eight (88.9%) patients, respectively, with an average duration from onset to the appearance of the discharges of 1.6, 1.0, and 2.44 months, respectively. In the status epilepticus group, these discharges were detected in one (5.0%), six (30.0%), and six (30.0%) patients, respectively. The incorporation of central sagittal sporadic epileptiform discharges and lateralized periodic discharges into the World Health Organization diagnostic criteria, alongside generalized periodic discharges, significantly shortened the average lapse from symptom onset to sporadic Creutzfeldt-Jakob disease diagnosis (2.06 months vs. 2.44 months; $p = 0.02$). Central sagittal sporadic epileptiform discharges emerge as promising biomarkers for distinguishing sporadic Creutzfeldt-Jakob disease from status epilepticus, and together with lateralized periodic discharges provide an opportunity for early diagnosis of sporadic Creutzfeldt-Jakob disease.

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Introduction

Prion disease, a fatal and transmissible neurodegenerative disorder, is caused by conformational changes of normal prion protein (PrP^{C}) into misfolded prion protein (PrP^{Sc}), which accumulates in the central nervous system [1]. Sporadic Creutzfeldt-Jakob disease (sCJD), the most common human prion disease, is idiopathic and has been increasing in incidence worldwide [2,3]. sCJD is classified into six subtypes based on a polymorphism at codon 129 of the prion protein (PrP) gene (*PRNP*) and the proteinase K-resistant core size of PrP^{Sc} (types 1 and 2): MM1, MM2, MV1, MV2, VV1, and VV2. The codon 129 polymorphism comprises two alleles: methionine (M) and valine (V) [4]. Among these subtypes, MM1 and MV1 sCJD, also known as classic sCJD, causes rapidly progressive dementia with some neurological and psychiatric symptoms, leading to akinetic mutism within only a few months [5].

Regarding diagnostic biomarkers for sCJD, electroencephalography (EEG) is a non-invasive and convenient tool. The most characteristic EEG finding in sCJD is generalized periodic discharges (GPDs), also known as periodic sharp wave complexes. GPDs appear as symmetrical generalized triphasic, biphasic, or mixed complexes occurring approximately every second on the EEG [6]. GPDs, previously reported to have high diagnostic specificities for sCJD (86% and 91%) [7,8], were included in the most widely used diagnostic criteria for the disease

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[9]. However, the diagnostic sensitivity of GPDs for sCJD is relatively low, reported to be around 67% and 64% [7,8], probably because it is difficult to distinguish GPDs from other periodic discharges such as spikes and sharp waves in the early stages of the disease [10]. Several patients with early-stage sCJD have been misdiagnosed as having nonconvulsive status epilepticus (NCSE) [11,12]. Furthermore, patients with status epilepticus (SE), including NCSE and convulsive status epilepticus (CSE), often display GPDs on EEG [13–15], which complicates the differentiation between sCJD and SE. In a recent analysis of EEGs in patients with MM1/classic sCJD, we observed the presence of central sagittal sporadic epileptiform discharges (CSSEDs) or lateralized periodic discharges (LPDs) occurring prior to the emergence of GPDs [16]. Therefore, the primary aim of this study was to evaluate the potential use of epileptiform discharges, including LPDs, CSSEDs, and GPDs, in differentiating sCJD from convulsive and nonconvulsive SE. Additionally, we analysed whether CSSEDs and LPDs can be used to improve early diagnosis of sCJD.

Results

One patient with possible classic sCJD, one with MM1 + 2 sCJD, and eight with MM2c sCJD were excluded from the analysis, resulting in the inclusion of nine patients with MM1/classic sCJD (MM1/classic sCJD group). Twenty patients with SE (11 with CSE and 9 with NCSE) were selected from a total of 118 patients with epilepsy for the analysis (SE group). The clinical features, and laboratory and neuroimaging findings of both groups are summarized in Table 1.

The clinical details of the patients in the MM1/classic sCJD group are presented in Supplementary Table S1 and S2. Briefly, cortical hyperintensity of diffusion-weighted magnetic resonance imaging (DWI-MRI) were detected in all nine patients, and real-time quaking-induced conversion (RT-QuIC) analyses in the cerebrospinal fluid (CSF) revealed positive results in six of the patients. Confirmation via autopsy was obtained for one patient (11.1%) with MM1-sCJD, with the autopsy results revealing spongiform changes with small vacuoles and synaptic-type PrP^{Sc} deposition in the cerebral cortices.

Table 1. Clinical features and laboratory and neuroimaging findings of the MM1/classic sCJD group and the SE group.

Group	MM1/classic sCJD ($n = 9$)	SE ($n = 20$)	p
Male sex	2 (22.2%)	13 (65%)	
Onset (Age; median, range)	66 (51–78)	56.5 (21–85)	0.19
Clinical symptoms and signs			
Rapid progressive dementia	9 (100%)	1 (5.0%)	
Myoclonus	7 (77.8%)	5 (25.0%)	
Visual or cerebellar signs	6 (66.7%)	3 (15.0%)	
Pyramidal/extrapyramidal signs	7 (77.8%)	6 (30.0%)	
Akinetic mutism	6 (66.7%)	0 (0.0%)	
Diagnosis of CJD			
Definite	1 (11.1%)		
Probable	8 (88.9%)		
Possible	0 (0%)		
EEG			
CSSEDs*	5/9 (55.6%)	1/20 (5.0%)	
Abundant	0/5 (0%)	0/1 (0%)	
Frequent	1/5(20%)	0/1 (0%)	
Occasional	4/5(80%)	1/1 (100%)	
Rare	0/5 (0%)	0/1 (0%)	
LPDs	1/9 (11.1%)	6/20 (30.0%)	
GPDs	8/9 (88.9%)	6/20 (30.0%)	
Hyperintensity on DWI-MRI			
CC	9/9 (100%)	5/17 (29.4%)	
GP	0/9 (0%)	0/17 (0%)	
PU	8/9 (88.9%)	0/17 (0%)	
CN	8/9 (88.9%)	1/17 (5.9%)	
TH	0/9 (0%)	3/17 (17.6%)	
Cerebrospinal fluid			
T-tau protein positivity	9/9 (100%)	1/1 (100%)	
14–3–3 protein positivity	8/9 (88.9%)	3/6 (50%)	
RT-QuIC positivity	6/6 (100%)	0/6 (0%)	

*The frequency of the appearance of CSSEDs on EEGs were classified into abundant (≥1 per 10 s, but not periodic), frequent (≥1/min but less than 1 per 10 s), occasional (≥1/h but less than 1/min), or rare (less than 1/h). CC: cerebral cortex, CN: caudate nucleus, CSSEDs: central sagittal sporadic epileptiform discharges, DWI-MRI: diffusion-weighted magnetic resonance imaging, EEG: electroencephalogram, GP: globus pallidus, GPDs: generalized periodic discharges, LPDs: lateralized periodic discharges, PU: putamen, RT-QuIC: real-time quaking-induced conversion, SE: status epilepticus, TH: thalamus.

The clinical features, along with the laboratory and neuroimaging findings of the patients in the SE group are presented in Supplementary Table 3. Rapidly progressive dementia was observed in one patient with NCSE (5%). Cortical hyperintensity on DWI-MRI was detected in one of the eight patients with CSE (12.5%), and in four of the nine patients with NCSE (44.4%). The CSF analysis revealed positive results of 14-3-3 proteins in one of two patients with CSE (50%), and in two of four patients with NCSE (50%). None of the patients in the SE group had positive results for the RT-QuIC analysis.

A total of 30 EEG recordings from nine patients in the MM1/classic sCJD group (mean 3.3 ± 2.8; range 1–10) and 177 EEG recordings in the SE group (mean 8.9 ± 9.8; range 1–36) were analysed (Figure 1, Supplementary Figure S1). In the MM1/classic sCJD group, CSSEDs, LPDs, and GPDs were detected on the EEGs of five, one, and eight patients, corresponding to sensitivities of 55.6%, 11.1%, and 88.9%, respectively (Table 1). These three types of EEG changes were observed in one (5.0%), six (30.0%), and six (30.0%) patients, respectively, in the SE group. Therefore, the specificities for CSSEDs, LPDs, and GPDs in differentiating sCJD from SE were 95%, 70%, and 70%, respectively. In one patient with classic sCJD, the CSSEDs on EEG disappeared immediately after intravenous benzodiazepine administration.

The average intervals between disease onset and the appearance of CSSEDs, LPDs, and GPDs on EEGs in the MM1/classic sCJD group were 1.6 (range, 1.0–2.0), 1.0 (range, 1.0–1.0), and 2.44 (range, 1.5–4.0) months, respectively (Figure 2). The average number of EEG recordings before GPD detection in the MM1/classic sCJD group was 2.0 ± 2.5 (range, 0–8). The average interval between disease onset and diagnosis of probable sCJD using the original 1998 criteria was 2.44 months (range, 1.5–3.0). In contrast, substitution of the typical periodic complexes (0.5–2 hz generalized bi/triphasic periodic complexes) with CSSEDs or LPDs, along with GPDs (the typical periodic complexes) resulted in a significantly earlier ($p = 0.02$) diagnosis of probable sCJD. The average difference was 2.06 months (range: 1.0–3.0 months) (Figure 3a). The substitution of the typical periodic complexes in the diagnostic criteria for sCJD proposed by Hermann et al. (2021 criteria) [17] did not result in a significant difference ($p = 0.17$) in the interval from disease onset to diagnosis (2.06 months; range 1.0–3.0 vs. 2.17 months; range 1.0–3.0, respectively; Figure 3b).

CSSEDs, LPDs, and GPDs were detected on the initial EEG in 80% (4/5), 100% (1/1), and 37.5% (3/8) of the patients in the MM1/classic sCJD group, respectively. One of the patients with classic sCJD (case 6) did not exhibit CSSEDs on the initial EEG. However, CSSEDs were observed on the EEG during the 1.5-month follow-up, followed by the detection of GPDs on the 2-month follow-up EEG (Supplementary Figure S2).

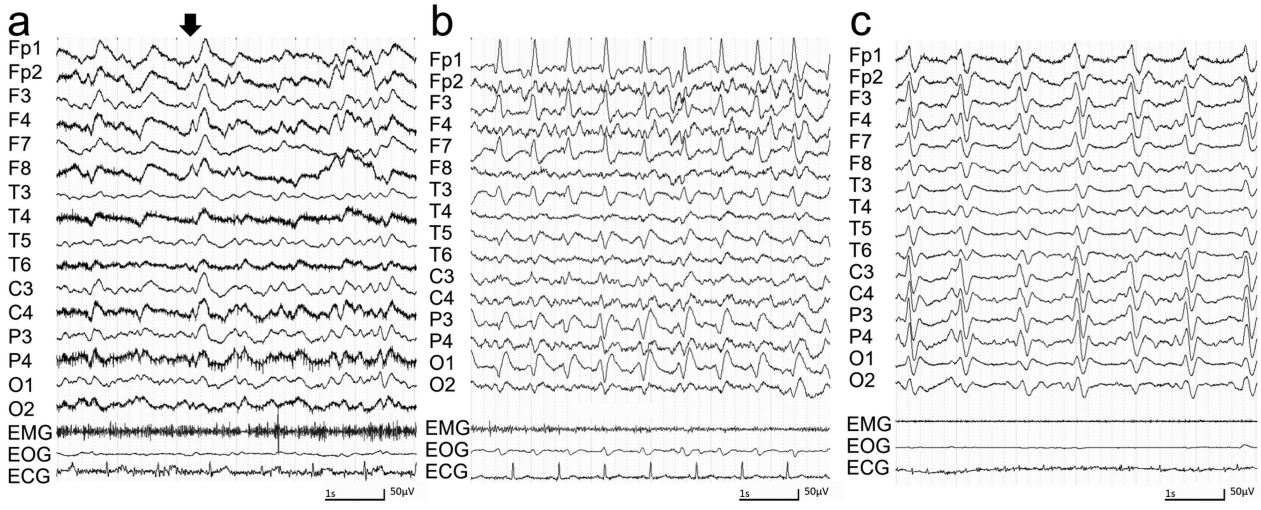

Figure 1. Representative EEG findings in the MM1/classic sCJD group. (a) EEG recordings in the early stage (2 months after disease onset) show background activity of 5–7 hz and 40–60 μV and CSSEDs (arrows). (b) EEG recordings in the early stage (1 month after disease onset) show background activity at 8–9 hz and 30–50 μV and LPDs in the left hemisphere. (c) EEG recordings in the middle stage (2.5 months after disease onset) show low-amplitude background activity at 5–7 hz and GPDs. CSSEDs: central sagittal sporadic epileptiform discharges, EEG: electroencephalography, GPDs: generalized periodic discharges, LPDs: lateralized periodic discharges, sCJD: sporadic Creutzfeldt-Jakob disease.

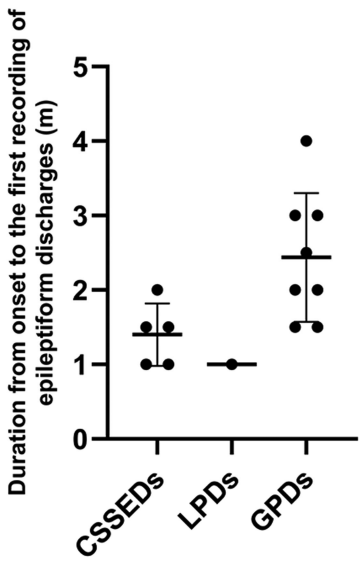

Figure 2. EEG findings in cases with MM1/classic sCJD. The interval from disease onset to the initial recording of CSSEDs, LPDs, and GPDs on EEG was plotted for each patient with MM1/classic sCJD. CSSEDs: central sagittal sporadic epileptiform discharges, EEG: electroencephalography, GPDs: generalized periodic discharges, LPDs: lateralized periodic discharges, sCJD: sporadic Creutzfeldt-Jakob disease.

Discussion

In this study, we evaluated epileptiform discharges (CSSEDs and LPDs) detected prior to the appearance of GPDs (the typical periodic complexes) in MM1/classic sCJD. CSSEDs exhibited a higher specificity compared to GPDs in distinguishing sCJD from the convulsive and non-convulsive varieties of SE. To the best of our knowledge, this is the first study to assess the utility of each epileptiform discharge in diagnosing sCJD, using a control group consisting solely of patients with SE. Including CSSEDs and LPDs as additional components of EEG findings significantly reduced the interval from disease onset to sCJD diagnosis. Since early diagnosis is associated with more effective therapeutic interventions, it would be reasonable to include these components in future diagnostic criteria for the disease.

Differentiating sCJD from SE (and from NCSE in particular) can occasionally be challenging owing to their shared clinical symptoms and laboratory biomarkers. Rapidly progressive dementia and cortical hyperintensity on DWI-MRI, which are essential findings in sCJD [16,18], are also frequently observed in NCSE

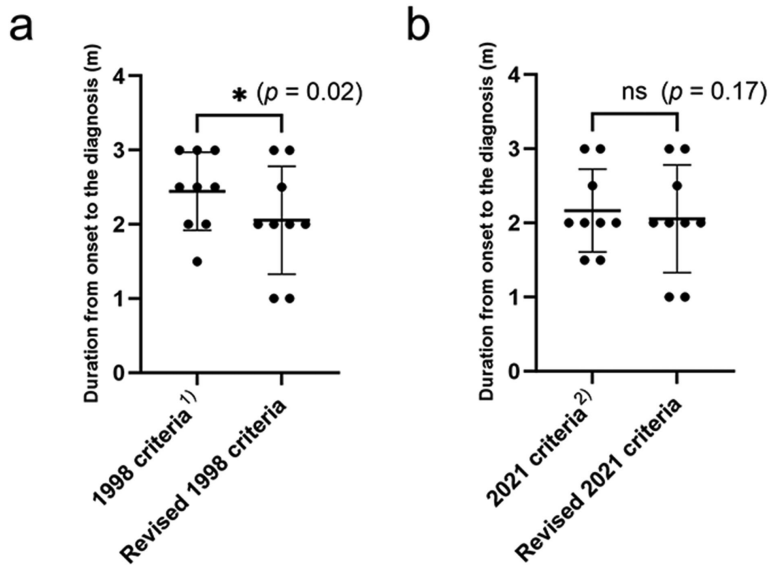

Figure 3. Comparison of interval between disease onset and diagnosis of probable sCJD using diagnostic criteria with or without the EEG changes.(a) intervals between disease onset and diagnosis of probable sCJD were compared using the original WHO diagnostic criteria (1998 criteria) [9] and the revised 1998 criteria, which included EEG findings (CSSEDs, LPDs, and GPDs) (b) the comparison was also conducted using the original diagnostic criteria for sCJD proposed by Hermann et al. (2021 criteria) [17] and the revised 2021 criteria, which included the same EEG findings. The data are presented as the mean ± standard error of the mean. *: $p < 0.05$ (paired t-test). CSSEDs: central sagittal sporadic epileptiform discharges, EEG: electroencephalography, GPDs: generalized periodic discharges, LPDs: lateralized periodic discharges, m: months, ns: not significant, sCJD: sporadic Creutzfeldt-Jakob disease.

[17–21]. This is consistent with the results of this study, including those for 14-3-3 proteins in the CSF. Besides sharing similar EEG findings, epileptiform discharges in sCJD also demonstrate benzodiazepine responsiveness [12], which complicates the distinction between sCJD and SE even after antiepileptic drug treatment. However, distinguishing sCJD from SE is crucial, considering the substantial differences in treatment approaches and prognostic outcomes. Notably, CSSEDs exhibited greater specificity than did GPDs in distinguishing sCJD from SE in this study, which might be the result of CSSEDs potentially reflecting pathological changes in sCJD, a premise supported by our previous study [16]. On the other hand, the pathophysiology of GPDs is presumed to be diverse and to involve selective synaptic failure as a common underlying mechanism [22]. Consequently, GPDs are common in both SE and sCJD, as was evident in the present study. Notably, the specificity of GPDs in diagnosing sCJD in this study was lower than that reported in previous studies [7,8]. This difference may have resulted from the difference in the composition of the control group, which only included patients with SE in the present study. In contrast, the control group was composed of patients with several different non-CJD diseases in previous studies. Regarding the sensitivity of epileptiform discharges in diagnosing sCJD in this study, the lower sensitivity of LPDs and CSSEDs compared to GPDs could stem from the inclusion of patients whose initial EEG recordings were evaluated more than 2 months after onset (as in cases 8 and 9; Figure 2). This delay might have affected the detection of CSSEDs and LPDs, which typically appear at earlier stages.

In a previous large cohort study on sCJD, the mean interval from onset to diagnosis using the 1998 criteria was 2.9 months [23], which is similar to the outcome of this study. Such an extended interval is not necessarily appropriate for effective therapeutic interventions in sCJD, considering the rapid progression of the disease [24]. To date, no drug has been successful in extending the survival period of patients with sCJD in clinical trials [25]. Therefore, establishing effective diagnostic criteria for sCJD to facilitate early intervention is of paramount importance. The incorporation of CSSEDs and LPDs to the diagnostic criteria shortened the average interval from disease onset to diagnosis of probable sCJD to approximately two months in the present study. Moreover, CSSEDs and LPDs were evident at early stages, consistently followed by GPDs in the later stages [16]. Thus, we would like to propose that epileptiform discharges before GPDs should be routinely evaluated as part of the diagnosis criteria for sCJD, as this can open opportunities for early intervention and treatment. Recent diagnostic criteria for sCJD have introduced disease-specific biomarkers such as MRI and RT-QuIC [17,26], but GPDs remain the sole EEG criterion. This study revealed that incorporating CSSEDs and LPDs as EEG findings into the 2021 criteria led to earlier diagnoses in some cases, despite no significant overall difference. Therefore, a promising approach for sCJD diagnosis in the future could involve a combination of early-stage epileptiform discharges on EEG that include CSSEDs and LPDs, and other biomarkers such as MRI and RT-QuIC.

In sCJD, repeated EEG monitoring is often essential to detect GPDs. Regarding the detection of GPDs on EEG during the disease course in MM1/classic sCJD, the initial EEG did not identify GDPs in more than half of the patients in this study (5/8) as well as in the previously reported study (7/7) [16]. However, the optimal interval for evaluating EEGs in patients with sCJD remain unclear. In this study, one patient with sCJD exhibited a transition in EEG findings from slow waves to CSSEDs and subsequently to GPDs over a period of two weeks. Additionally, CSSEDs appeared in this patient 1.5 months after onset, which closely aligned with the average duration from disease onset to the appearance of CSSEDs in this study. Consequently, the recommended practice for the monitoring of patients with sCJD could involve repeated EEG evaluations every 0.5 months.

In this study, LPDs in one patient (100%) and CSSEDs in four patients (80%) observed during the early stages transitioned to GPDs over the course of the disease, which is consistent with previous reports [16,27]. Notably, a previous study using MRI-DWI to estimate prion propagation in MM1-sCJD found that lesions spread from the parietal to the frontal cortex early in the course of the disease, with involvement of the temporal cortex occurring relatively later [28]. The regions identified as lesioned on MRI-DWI in the early stages of MM1-sCJD corresponded to the central sagittal region with CSSEDs on EEGs in this study, suggesting that the shift in EEG findings from CSSEDs to GPDs may reflect prion propagation. Furthermore, the presence of LPDs in the early stages of the disease in some cases suggests that there may be two patterns for this propagation: unilateral to bilateral spread, and spread from the central sagittal region to the lateral regions.

This study has several limitations. First, only one patient with sCJD underwent pathological analysis, which precluded the exclusion of sCJD subtypes such as MM1 + 2 from consideration. To address the low rate of pathologically-confirmed disease, we evaluated

PrP^{Sc} in the CSF using RT-QuIC. Second, we analysed a limited number of patients. Future prospective studies including a larger number of patients with sCJD are required to obtain more precise sensitivity and specificity values for differentiating sCJD from SE using CSSEDs and LPDs. Additionally, future studies are required to validate the monitoring approach for sCJD proposed in this study, which involves EEG evaluation every 0.5 months from the early stage.

In conclusion, CSSEDs on EEGs exhibited higher specificity in distinguishing sCJD from SE compared to GPDs. Furthermore, the incorporation of CSSEDs and LPDs as EEG findings into the 1998 criteria significantly shortened the interval from disease onset to the diagnosis of probable sCJD. These findings suggest that CSSEDs and LPDs on EEGs are valuable tools for the early diagnosis of sCJD and could facilitate early interventions. Additionally, they may serve as crucial factors in differentiating sCJD from SE.

Materials and methods

We conducted a retrospective cohort study, enrolling 19 patients with sCJD and 118 patients with epilepsy who were admitted to the Institute of Science Tokyo Hospital between April 2009 and March 2023. Polymorphisms at codon 129 of *PRNP* in the sCJD group were analysed in 15 patients, all of whom had methionine homozygosity (MM). None of the patients had pathological mutations in *PRNP*. Brain pathology analysis was conducted on the autopsied patients. Types 1 and 2 PrP^{Sc} were detected by western blot analysis of the protease K-resistant PrP [29]. *PRNP* and PrP^{Sc} typing were conducted as previously described [30]. MM1/classic and MM1 + 2 were diagnosed according to the World Health Organization (WHO) diagnostic criteria (1998) [9], whereas MM2c was diagnosed using the criteria proposed by Hamaguchi et al [31]. The electroencephalographic criteria for NCSE were derived from the modified Salzburg Consensus Criteria for NCSE [32].

All electrodes were configured according to the standard international 10–20 system [33]. EEG findings from each patient were reviewed by three different board-certified neurologists (T.M, H.N, and N.S). Additionally, we evaluated epileptiform discharges (including CSSEDs, LPDs, and GPDs) in both the MM1/classic sCJD and SE groups. Each epileptiform discharge was defined according to the EEG terminology from the American Clinical Neurophysiology Society, and the detailed findings of these epileptiform discharges have been reported elsewhere [16]. Briefly, CSSEDs were defined as nonrhythmic and nonperiodic waveforms showing midline-predominant generalized (bilaterally synchronous and symmetric) spike-and-wave complexes and/or sharp waves in the central sagittal region. LPDs were defined as repetitive lateralized (unilateral or bilateral asymmetric) waveforms lasting < 0.5 s with a frequency of 0.5–2 hz. GPDs were defined as repetitive generalized (bilaterally synchronous and symmetric pattern) waveforms lasting < 0.5 s with a frequency of 0.5–2 hz. The clinical features and laboratory and neuroimaging findings of the MM1/classic sCJD and SE groups were evaluated and compared.

The Mann – Whitney U test and paired t-test were used to compare differences between groups. Statistical analyses were conducted using the GraphPad Prism 9 software (GraphPad Software, San Diego, CA, USA). This study was conducted in accordance with the Declaration of Helsinki (2013).

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Disclosure statement

No potential conflict of interest was reported by the author(s).

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Author contributions

Data were collected by TM, HN, and NS and analysed by TM, HN, and NS. Pathological analyses were performed using TK software. The CSF samples were analysed using KS. The manuscript was drafted by TM and HN, and revised by TK, KS, TY, and NS. The study was supervised by TY and NS. This study was designed and conceptualized by NS. All authors have read and approved the final work in the author contribution statement of the manuscript

Data availability statement

The authors state that the anonymized data on which the article is based will be shared upon reasonable request from any qualified investigator.

Ethical statement

The study was conducted in accordance with the principles of the Declaration of Helsinki (2013). The study protocol was approved by the Institutional Ethics Committee of Institute of Science Tokyo (G2000–141), and written informed consent was obtained from each patient's family.

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References

[1] Prusiner SB. Prions. Proc Natl Acad Sci U S A. 1998;95(23):13363–13383. doi: 10.1073/pnas.95.23.13363
[2] Gambetti P, Kong Q, Zou W, et al. Sporadic and familial CJD: classification and characterisation. Br Med Bull. 2003;66(1):213–239. doi: 10.1093/bmb/66.1.213
[3] Uttley L, Carroll C, Wong R, et al. Creutzfeldt-Jakob disease: a systematic review of global incidence, prevalence, infectivity, and incubation. Lancet Infect Dis. 2020;20(1):e2–e10. doi: 10.1016/S1473-3099(19)30615-2
[4] Parchi P, Giese A, Capellari S, et al. Classification of sporadic Creutzfeldt-Jakob disease based on molecular and phenotypic analysis of 300 subjects. Ann Neurol. 1999;46:224–233. https://doi.org/10.1002/1531-8249(199908)46:2<224::AID-ANA12>3.0.CO;2-W
[5] Iwasaki Y, Yoshida M, Hashizume Y, et al. Clinicopathologic characteristics of sporadic Japanese Creutzfeldt–Jakob disease classified according to prion protein gene polymorphism and prion protein type. Acta Neuropathol. 2006;112(5):561–571. doi: 10.1007/s00401-006-0111-7
[6] Hirsch LJ, Fong MWK, Leitinger M, et al. American clinical neurophysiology Society's standardized critical care EEG terminology: 2021 version. J Clin Neurophysiol. 2021;38(1):1–29. doi: 10.1097/wnp.0000000000000806
[7] Steinhoff BJ, Räcker S, Herrendorf G, et al. Accuracy and reliability of periodic sharp wave complexes in Creutzfeldt-Jakob disease. Arch Neurol. 1996;53(2):162–166. doi: 10.1001/archneur.1996.00550020074017
[8] Steinhoff BJ, Zerr I, Glatting M, et al. Diagnostic value of periodic complexes in Creutzfeldt–Jakob disease. Ann Neurol. 2004;56(5):702–708. doi: 10.1002/ana.20261
[9] World Health Organization. WHO manual for strengthening diagnosis and surveillance of Creutzfeldt-Jakob disease/by Martin Zeidler, Clarence J. Gibbs, François Meslin. 1998 [cited 2024 Oct 1]. Available from: http://apps.who.int/iris/handle/10665/66394
[10] Wang PS, Wu YT, Hung CI, et al. Early detection of periodic sharp wave complexes on EEG by independent component analysis in patients with Creutzfeldt-Jakob disease. J Clin Neurophysiol. 2008;25(1):25–31. doi: 10.1097/WNP.0b013e318163a7d5
[11] Espinosa PS, Bensalem-Owen MK, Fee DB. Sporadic Creutzfeldt–Jakob disease presenting as nonconvulsive status epilepticus case report and review of the literature. Clin Neurol Neurosurg. 2010;112(6):537–540. doi: 10.1016/j.clineuro.2010.03.025
[12] Marquetand J, Knake S, Strzelczyk A, et al. Periodic EEG patterns in sporadic Creutzfeld-Jakob-disease can be benzodiazepine-responsive and be difficult to distinguish from non-convulsive status epilepticus. Seizure. 2017;53:47–50. doi: 10.1016/j.seizure.2017.10.023
[13] Foreman B, Claassen J, Abou Khaled K, et al. Generalized periodic discharges in the critically ill: a case-control study of 200 patients. Neurology. 2012;79(19):1951–1960. doi: 10.1212/WNL.0b013e3182735cd7
[14] Husain AM, Mebust KA, Radtke RA. Generalized periodic epileptiform discharges: etiologies, relationship to status epilepticus, and prognosis. J Clin Neurophysiol. 1999;16(1):51–58. doi: 10.1097/00004691-199901000-00005
[15] Orta DS, Chiappa KH, Quiroz AZ, et al. Prognostic implications of periodic epileptiform discharges. Arch Neurol. 2009;66(8):985–991. doi: 10.1001/archneurol.2009.137
[16] Matsubayashi T, Akaza M, Hayashi Y, et al. Specific electroencephalogram features in the very early phases of sporadic Creutzfeldt-Jakob disease. J Neurol Sci. 2022;437:120265. doi: 10.1016/j.jns.2022.120265
[17] Hermann P, Appleby B, Brandel J-P, et al. Biomarkers and diagnostic guidelines for sporadic Creutzfeldt-Jakob disease. Lancet Neurol. 2021;20(3):235–246. doi: 10.1016/S1474-4422(20)30477-4
[18] Qi C, Zhang JT, Zhao W, et al. Sporadic Creutzfeldt-Jakob disease: a retrospective analysis of 104 cases. Eur Neurol. 2020;83(1):65–72. doi: 10.1159/000507189
[19] Mutis JA, Rodriguez JH, Nava-Mesa MO. Rapidly progressive cognitive impairment with neuropsychiatric symptoms as the initial manifestation of status epilepticus. Epilepsy Behav Case Rep. 2017;7:20–23. doi: 10.1016/j.ebcr.2016.11.002
[20] Zhao Q, Sun L, Hu B, et al. Nonconvulsive status epilepticus manifesting as rapidly progressive dementia and infarction in the splenium of the corpus callosum: a case report. Medicine (Baltimore). 2021;100(15):e25263. doi: 10.1097/MD.0000000000025263
[21] Sarria-Estrada S, Santamarina E, Quintana M, et al. Magnetic resonance imaging findings in focal-onset status epilepticus. Eur J Neurol. 2022;29(1):3–11. doi: 10.1111/ene.15065
[22] van Putten MJ, Hofmeijer J. Generalized periodic discharges: pathophysiology and clinical considerations. Epilepsy Behav. 2015;49:228–233. doi: 10.1016/j.yebeh.2015.04.007
[23] Nakatani E, Kanatani Y, Kaneda H, et al. Specific clinical signs and symptoms are predictive of clinical course in sporadic Creutzfeldt–Jakob disease. Eur J Neurol. 2016;23(9):1455–1462. doi: 10.1111/ene.13057
[24] Nozaki I, Hamaguchi T, Sanjo N, et al. Prospective 10-year surveillance of human prion diseases in

Japan. Brain. 2010;133(10):3043–3057. doi: 10.1093/brain/awq216

[25] Miranda LHL, Oliveira A, Carvalho DM, et al. Systematic review of pharmacological management in Creutzfeldt-Jakob disease: no options so far? Arq Neuropsiquiatr. 2022;80(8):837–844. doi: 10.1055/s-0042-1755341

[26] Hermann P, Laux M, Glatzel M, et al. Validation and utilization of amended diagnostic criteria in Creutzfeldt-Jakob disease surveillance. Neurology. 2018;91(4):e331–e338. doi: 10.1212/WNL.0000000000005860

[27] Wieser HG, Schindler K, Zumsteg D. EEG in Creutzfeldt–Jakob disease. Clin Neurophysiol. 2006;117(5):935–951. doi: 10.1016/j.clinph.2005.12.007

[28] Pascuzzo R, Oxtoby NP, Young AL, et al. Prion propagation estimated from brain diffusion MRI is subtype dependent in sporadic Creutzfeldt–Jakob disease. Acta Neuropathol. 2020;140(2):169–181. doi: 10.1007/s00401-020-02168-0

[29] Parchi P, Notari S, Weber P, et al. Inter-laboratory assessment of PrP Sc typing in Creutzfeldt–Jakob disease: a Western blot study within the NeuroPrion consortium. Brain Pathol. 2009;19(3):384–391. doi: 10.1111/j.1750-3639.2008.00187.x

[30] Kitamoto T, Ohta M, Doh-Ura K, et al. Novel missense variants of prion protein in Creutzfeldt-Jakob disease or Gerstmann-Sträussler syndrome. Biochem Biophys Res Commun. 1993;191(2):709–714. doi: 10.1006/bbrc.1993.1275

[31] Hamaguchi T, Sanjo N, Ae R, et al. MM2-type sporadic Creutzfeldt-Jakob disease: new diagnostic criteria for MM2-cortical type. J Neurol Neurosurg Psychiatry. 2020;91(11):1158–1165. doi: 10.1136/jnnp-2020-323231

[32] Leitinger M, Beniczky S, Rohracher A, et al. Salzburg consensus criteria for non-convulsive status Epilepticus–approach to clinical application. Epilepsy Behav. 2015;49:158–163. doi: 10.1016/j.yebeh.2015.05.007

[33] Klem GH, Lüders HO, Jasper HH, et al. The ten-twenty electrode system of the international federation: the international federation of clinical neurophysiology. Electroencephalogr Clin Neurophysiol Suppl. 1999;52:3–6.