学位論文

Clinical Characteristics of Japanese Patients With Corticobasal Degeneration

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Clinical characteristics of Japanese patients with corticobasal degeneration

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Abbreviations
CBD, corticobasal degeneration
CBS, corticobasal syndrome
DLB, dementia with Lewy body
FBS, frontal behavioral-spatial syndrome
naPPA, nonfluent/agrammatic variant of primary progressive aphasia
PD, Parkinson's disease
PSPS, progressive supranuclear palsy syndrome
TF, tube feeding
TS, tracheostomy

Abstract

Introduction: Corticobasal degeneration (CBD) is a clinically heterogeneous neurodegenerative disorder, for which pathological investigations are essential for a definitive diagnosis. This study explored the clinical characteristics of Japanese patients with pathologically confirmed CBD.

Methods: We reviewed the data of Japanese patients with pathologically confirmed CBD who were consecutively autopsied at our institute. Clinical data were obtained from medical records and clinicopathological conferences.

Results: Of the 34 patients initially reviewed, three were excluded because of a lack of detailed clinical data. Of the remaining 31 patients, 16 were men and 15 were women. The mean ages at onset and death were 63.3 ± 6.7 (51–79) years and 69.1 ± 6.9 (54–86), respectively. The median disease duration was 6.0 (2.5–12) years. The clinical phenotypes were as follows: progressive supranuclear palsy syndrome (PSPS; n = 20, 64.5%), probable or possible corticobasal syndrome (n = 6, 19.4%), frontal behavioral-spatial syndrome (n = 4, 12.9%), nonfluent/agrammatic variant of primary progressive aphasia (n = 1, 3.2%). Furthermore, 28 (90.3%) patients exhibited dysphagia with a median latency of 3.5 (1.0–10.0) years, and 22 (71.0%) patients who underwent tube feeding survived longer than those who did not (P = 0.013).

Conclusions: Compared with Western populations, a high prevalence of PSPS may be a clinical characteristic of Japanese patients with CBD. Additionally, dysphagia occurs in many patients with early latency and may shorten survival. Tube feeding contributes to the prolonged survival of patients with CBD.

Key words: corticobasal degeneration, corticobasal syndrome, dysphagia, progressive supranuclear palsy syndrome, tube feeding

1. Introduction

Corticobasal degeneration (CBD) is a neurodegenerative disorder pathologically classified as a four-repeat tauopathy in which phosphorylated tau accumulates in astroglia and neurons [1]. Classically, CBD presents with asymmetric extrapyramidal and cortical symptoms [2-4], now known as "corticobasal syndrome (CBS)" [5]. However, phenotypes other than CBS have been reported, including aphasia, frontotemporal dementia, and posterior cortical atrophy [5]. CBD is clinically heterogeneous, making clinical diagnosis challenging and necessitating pathological investigations for an accurate diagnosis [6].

Several studies have described the clinical characteristics of patients with pathologically proven CBD [7-22]. However, most of these studies are from Western countries. To elucidate the general clinical characteristics of CBD, it is necessary to investigate them in patients of multiple ethnicities. Therefore, in this study, we examined the clinical characteristics of Japanese patients with pathologically confirmed CBD.

2. Methods

2.1. Subjects

This single-center retrospective study investigated Japanese patients with pathologically confirmed CBD who underwent consecutive autopsies at our institute. Patients with insufficient clinical data were excluded. Written informed consent was obtained from patients' relatives before the autopsy. All investigations were approved by the Research Ethics Committee of Aichi Medical University (approval number: 15-017).

2.2. Pathological diagnosis of CBD

The Pathological diagnosis of CBD was based on the presence of Gallyas/tau-positive lesions in the cerebral cortex and basal ganglia, including astrocytic plaques, neuropil threads, coiled bodies, and pretangles [23].

2.3. Clinical analysis

Clinical data were obtained from medical records and clinicopathological conferences. We retrospectively examined the information regarding sex, age at onset, age at death, disease duration, initial symptoms, cause of death, final clinical diagnosis, and clinical phenotype. Additionally, we examined the implementation of tube feeding (TF) or tracheostomy (TS) and the presence of dysphagia and central respiratory disturbances. Disease duration was defined as the time from the onset of CBD-related symptoms to death, expressed in years. Bradykinesia, muscle rigidity, and resting tremors were regarded as parkinsonism. Gait impairment was defined as difficulty in walking with easy falling. Behavioral abnormalities or cognitive impairment included disinhibition, personality and behavioral changes, memory loss, and general cognitive impairment. Language disorders included aphasia or verbal apraxia but not dysarthria. Apraxia included all types of apraxia except for verbal apraxia. Herein, we adopted the four clinical phenotypes proposed by Armstrong *et al*.: possible or probable CBS, frontal behavioral-spatial syndrome (FBS), nonfluent/agrammatic variant of primary progressive aphasia (naPPA), and progressive supranuclear palsy syndrome

(PSPS) [20]. Based on their criteria [20] and patients' clinical courses, we determined one clinical phenotype per patient. TF was achieved with a percutaneous endoscopic gastrostomy or nasogastric tube. We did not consider the temporary use of TF as its implementation. We determined the presence of dysphagia and central respiratory disturbances based on the description of diagnoses by the clinicians. Additionally, catching an aspiration pneumonia and the increase of choking on foods or drinks were regarded as the presence of dysphagia. We treated missing data as nonexistent.

2.4. Statistical analysis

Data are expressed as mean ± standard deviation (range) or median (range). The Holm method was used to compare nominal scales. The Mann–Whitney U-test and Steel–Dwass tests were used to compare continuous variables. Pearson's correlation coefficients were calculated between the disease duration and latency to dysphagia. Statistical significance was set at $P < 0.05$. Analyses were performed using EZR (Saitama Medical Center, Jichi Medical University, Saitama, Japan), which is a graphical user interface for R (The R Foundation for Statistical Computing, Vienna, Austria, version 4.2.2) [24].

3. Results

Of the 34 patients with pathologically confirmed CBD, three were excluded owing to a lack of detailed clinical data. The clinical phenotypes of the remaining 31 patients were as follows: PSPS (n = 20, 64.5%); probable or possible CBS (n = 6, 19.4%); FBS (n = 4, 12.9%); and naPPA (n = 1, 3.2%). The clinical characteristics of the patients are presented in Table 1. The initial symptoms were significantly different between the CBS and FBS groups ($P = 0.029$). The final clinical diagnosis differed significantly among the phenotypes: CBS vs. FBS ($P = 0.019$), CBS vs. PSPS ($P = 0.016$), and FBS vs. PSPS ($P = 0.016$). Other characteristics were not significantly different among the clinical phenotypes.

Table 1. Clinical characteristics of the included patients with pathologically confirmed CBD.

Characteristics	All patients, n = 31	PSPS, n = 20	Probable or possible CBS, n = 6	FBS, n = 4	naPPA, n = 1
Sex, male	16 (51.6)	14 (70.0)	2 (33.3)	0 (0)	0 (0)
Age at onset, years	63.3 ± 6.7 (51–79)	63.4 ± 6.2 (53–79)	67.3 ± 6.9 (58–77)	59.3 ± 6.2 (51–66)	53
Age at death, years	69.1 ± 6.9 (54–86)	69.0 ± 6.0 (60–86)	73.5 ± 8.4 (61–83)	65.0 ± 7.4 (54–70)	62
Disease duration, years	6.0 (2.5−12)	5.0 (3–12)	6.0 (3.5–10.5)	4.8 (2.5–11)	9
Initial symptom					
parkinsonism	10 (32.3)	5 (25.0)	5 (83.3)	0 (0)	0 (0)
gait impairment	8 (25.8)	8 (40.0)	0 (0)	0 (0)	0 (0)
behavioral abnormalities or cognitive impairment	8 (25.8)	4 (20.0)	0 (0)	4 (100)	0 (0)
apraxia	3 (9.7)	2 (10.0)	1 (16.7)	0 (0)	0 (0)
dystonia	1 (3.2)	1 (5.0)	0 (0)	0 (0)	0 (0)
language impairment	1 (3.2)	0 (0)	0 (0)	0 (0)	1 (100)
Cause of death					
pneumonia	19 (61.3)	11 (55.0)	6 (100)	1 (25.0)	1 (100)
choking	5 (16.1)	5 (25.0)	0 (0)	0 (0)	0 (0)
urinary tract infections	2 (6.5)	2 (10.0)	0 (0)	0 (0)	0 (0)
cancer	2 (6.5)	1 (5.0)	0 (0)	1 (25.0)	0 (0)
gastrointestinal hemorrhage	1 (3.2)	0 (0)	0 (0)	1 (25.0)	0 (0)
arrythmia	1 (3.2)	1 (5.0)	0 (0)	0 (0)	0 (0)
unknown (sudden death)	1 (3.2)	0 (0)	0 (0)	1 (25.0)	0 (0)
Final clinical diagnosis					
PSP	11 (35.5)	11 (55.0)	0 (0)	0 (0)	0 (0)
CBD or CBS	10 (32.3)	4 (20.0)	6 (100)	0 (0)	0 (0)
Parkinson's disease or dementia with Lewy bodies	5 (16.1)	4 (20.0)	0 (0)	1 (25.0)	0 (0)
frontotemporal lobar degeneration or frontotemporal dementia	3 (9.7)	0 (0)	0 (0)	2 (50.0)	1 (100)
Alzheimer's disease	1 (3.2)	0 (0)	0 (0)	1 (25.0)	0 (0)
multiple system atrophy	1 (3.2)	1 (5.0)	0 (0)	0 (0)	0 (0)

Data are expressed as mean ± standard deviation (range), median (range), or number (%).

Abbreviations: CBD, corticobasal degeneration; CBS, corticobasal syndrome; FBS, frontal behavioral–spatial syndrome; naPPA, nonfluent/agrammatic variant of primary progressive aphasia; PSP, progressive supranuclear palsy; PSPS, progressive supranuclear palsy syndrome.

Table 2 shows the proportion of patients who underwent TF or TS and a comparison of disease duration between the received and non-received groups. The nutritional statuses of patients who received TF were regularly checked by nutrition support teems, and the required amount of calories were administered. After the implementation of TF, the patients' general conditions had been well, and none of the patients died of malnutrition.

Table 2. The proportion of patients who received tube feeding or tracheostomy and comparison of the disease duration between the received and non-received groups.

	Received group	Non-received group	P value
Tube feeding			
patients	22 (71.0)	9 (29.0)	
latency to tube feeding, years	4.0 (1.5–10.5)	-	
disease duration, years	6.5 (3.0–12.0)	3.5 (2.5–6.0)	0.013 [a]
Tracheostomy			
patients	3 (9.7)	28 (90.3)	
latency to tracheostomy, years	6.0 (3.0–6.5)	-	
disease duration, years	4.5 (3.0–8.0)	3.5 (2.5–12.0)	0.973 [a]

Data are expressed as number (%) or median (range).

[a] Mann–Whitney U-test.

Twenty-eight (90.3%) patients experienced dysphagia during the disease course. The median time of onset was 3.5 (1.0–10.0) years (the onset time was unknown in one case). The latency to dysphagia significantly correlated with the disease duration (Pearson's correlation coefficient 0.596, 95% CI 0.279–0.796, P = 0.001; Figure 1). All patients who died of pneumonia or choking, except for one, had dysphagia. The patient without dysphagia was bedridden for 4 years before death. Two (6.5%) patients had central respiratory disturbances. The time of the symptom onset was unknown in these patients. Two patients with central respiratory disturbances died of pneumonia at 8 and 10 years after disease onset.

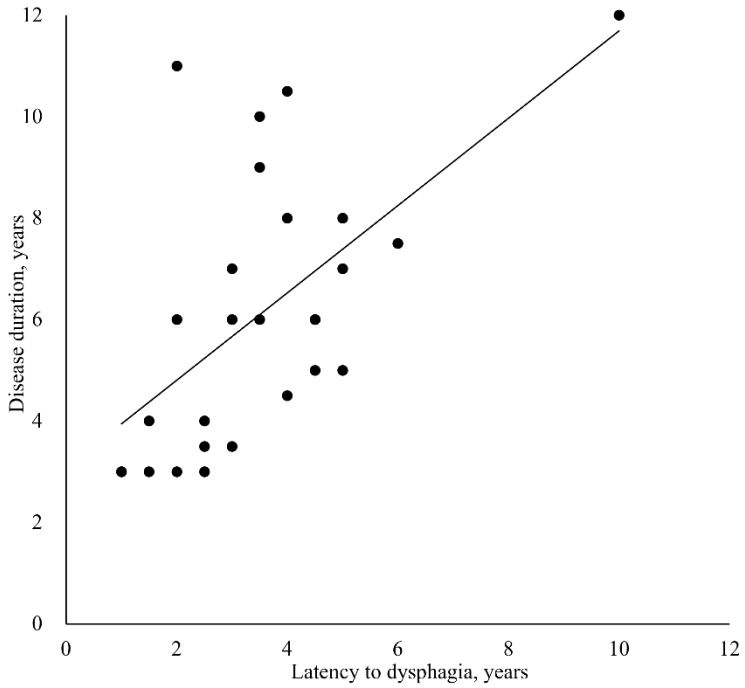

Figure 1. The correlation between the disease duration and latency to dysphagia in patients with corticobasal degeneration (Pearson's correlation coefficient 0.596, 95% CI 0.279–0.796, P = 0.001).

4. Discussion

This study investigated the clinical characteristics of Japanese patients with pathologically confirmed CBD at a single institute.

One of the characteristics of this cohort was the high prevalence of PSPS (64.5%). Four clinical phenotypes proposed by Armstrong *et al*. have based on the final clinical diagnosis of patients with pathologically proven CBD: including CBS (37.1%), PSPS (23.3%), and frontotemporal dementia (13.8%) [20]. Most patients investigated in their study were ethnically Western. Among the studies based on the Armstrong criteria [20], one study in the United Kingdom reported a low prevalence of PSPS [21], whereas another in Japan reported a high prevalence of this phenotype [22]. The method used to enumerate the phenotypes in the present study differed from those used in previous studies [21, 22]. In addition, the background of the present cohort was dominated by movement disorders. The method and background may have influenced the results. However, we believe that the high prevalence of PSPS is a clinical characteristic of Japanese patients with CBD.

The sex distribution of patients with CBD differs among studies, ranging from female predominance [7, 11, 19] to almost equal [9, 10, 15, 16, 18, 21, 25] to completely equal [17]. One Western study reported no significant differences in the sex distribution among clinical phenotypes; however, the method adopted for classifying clinical phenotypes differed from that adopted in the present study [19]. Regarding the sex distribution of CBD, Japanese patients exhibit equality [14, 22] or slight female predominance [8]. In the present cohort, the number of men was slightly higher, and no significant differences were detected among the clinical phenotypes. Revealing sex distribution differences specific to Japanese patients was challenging.

In previous studies, the mean ages at disease onset and death ranged from 63.1 to 67.0 years [7, 14, 17, 19-22] and from 69.3 to 73.0 years [7-11, 17-19, 22], respectively. The median disease duration has been reported to range from 5.2 to 8.2 years [7, 10, 11, 14, 22, 25]. Our results were within these ranges, and no characteristics specific to Japanese patients were observed.

In this study, parkinsonism was the most common initial symptom (32.3%), followed by gait impairment (25.8%) and behavioral abnormalities or cognitive impairment (25.8%). One study reported that the initial presentations were parkinsonism (47%), memory decline (37%), and aphasia (16%) [18]. The high proportion of patients with gait impairment in our study may have resulted from the high frequency of PSPS. Aiba *et al*. also reported a high frequency of gait disturbance as an initial symptom, and PSPS was the most common phenotype in that Japanese cohort [22]. Another Japanese study reported non-motor symptoms as initial signs of CBD, including limb apraxia (30%), aphasia (20%), memory disturbance (20%), abnormal behavior (20%), and delusion of persecution (10%) [14]. Six of the ten included patients were recruited from psychiatric institutions, which may have influenced their results.

Pneumonia is the main cause of death in patients with CBD, with reported proportions of 100% [9] and 66% [22]. This study showed a proportion similar to the latter study. Dysphagia and immobility are considered as causes of pneumonia in CBD [9]. In the present study, all patients who died of pneumonia had dysphagia or became bedridden before death, which supports that report [9]. To our knowledge, no studies reported a cause of death other than pneumonia in patients with pathologically proven CBD. Choking was the second most common cause of death in this study. Dysphagia may result in choking as well as pneumonia. The two patients who died of urinary tract infections were bedridden for 1 year before death. A bedridden status is one of risk factors for treatment failure of complicated urinary tract infections [26]. Other causes of death would simply be complications.

Despite exhibiting phenotypes different from CBS, four patients were clinically diagnosed as CBD. The clinical diagnoses of these patients have based on asymmetrical symptoms and brain imaging findings. Asymmetricity would be an important indicator of CBD. Parkinson's disease (PD) or dementia with Lewy bodies (DLB) were the third most common final clinical diagnosis in this study. However, the basis of the clinical diagnosis was ambiguous or vulnerable. In addition, patients diagnosed with PD or DLB had an atypical clinical course as those conditions. It is difficult to consider the presence of PD/DLB phenotype in patients with CBD from the present study.

Reports on TF in patients with CBD are limited. No studies have reported on the proportion of Western patients with CBD receiving TF. Regarding TF in other neurodegenerative diseases, approximately one-third of US nursing home residents with advanced dementia receive TF [27], while the use of TF in patients with dementia or PD in the UK is uncommon [28]. Thus, we speculated that the proportion of patients receiving TF in the present study was high. Another Japanese study reported a high proportion of patients with CBD who underwent TF [22]. The Japanese healthcare system, social environment, and traditional ethical values may encourage patients with advanced neurological disorders to receive life-sustaining treatments [29].

The efficacy of TF in patients with advanced neurological disorders remains controversial. One

study reported that TF after the akinetic mutism state has been reached was the most crucial factor for prolonged survival in Japanese patients with sporadic Creutzfeldt–Jakob disease [30]. Another study reported that TF did not prevent aspiration and decrease mortality in advanced dementia [27]. In the present study, the total disease duration was significantly longer in tube-fed patients than in non-tube-fed patients. Thus, TF may contribute to prolonged survival in patients with CBD. The nutritional statuses of all patients who received TF were regularly checked; however, comprehensively analyzable numerical data accounting for their nutritional statuses were unavailable in this study, which is a limitation in discussing the efficacy of TF.

In one study, most patients with CBD died of pneumonia due to dysphagia and immobility [9]. Dysphagia has been reported in 31% of patients with CBD, with a median latency from disease onset of 64 months [25]. A Japanese study reported a median latency to dysphagia of 5 years [22]. In this study, more patients with CBD had dysphagia with an earlier latency than that previously reported. Wenning *et al*. reported that the early onset of dysphagia had no significant effect on survival in patients with CBD, whereas dysphagia at the last visit was a predictor of shorter survival, suggesting that the lack of predictive power might be related to a later onset of dysphagia in their study [9]. Another study showed that the latency to dysphagia was significantly correlated with disease duration in patients with parkinsonian disorders, including CBD [25]. According to our results, dysphagia with early latency may shorten the disease duration despite the high proportion of patients undergoing TF.

To our knowledge, no previous studies have investigated TS or central respiratory disturbances in patients with CBD. In this study, disease duration was slightly longer in patients who received TS than in those who did not; however, no significant difference was observed. Two of the three patients who received TS had severe pneumonia at the time of receiving TS and died 2 months later after TS. TS may not influence the disease course in patients with CBD. However, we consider it difficult to determine the effect of TS on the disease course. Although clinical data on central respiratory disturbances are limited, this condition would be rare in patients with CBD and may not influence the disease duration.

5. Conclusion

Herein, we investigated the clinical characteristics of Japanese patients with pathologically confirmed CBD at a single institute. Compared with Western patients, a high prevalence of PSPS may be a characteristic of Japanese patients with this condition. Disease duration in this cohort was not different from that reported for Western populations. However, dysphagia occurred more often with early latency and might be correlated with shorter survival. TF was a significant clinical factor in the prolonged survival of patients with CBD.

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Declarations of Competing Interest

None.

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References

[1] L. Buée, A. Delacourte, Comparative biochemistry of tau in progressive supranuclear palsy, corticobasal degeneration, FTDP-17 and Pick's disease, Brain Pathol. 9 (1999) 681–693. https://doi.org/10.1111/j.1750-3639.1999.tb00550.x.

[2] J. J. Rebeiz, E. H. Kolodny, E. P. Richardson, Jr., Corticodentatonigral degeneration with neuronal achromasia: a progressive disorder of late adult life, Trans Am Neurol Assoc. 92 (1967) 23–26.

[3] J. J. Rebeiz, E. H. Kolodny, E. P. Richardson, Jr., Corticodentatonigral degeneration with neuronal achromasia, Arch Neurol. 18 (1968) 20–33. https://doi.org/10.1001/archneur.1968.00470310034003.

[4] W. R. Gibb, P. J. Luthert, C. D. Marsden, Corticobasal degeneration, Brain. 112 (1989) 1171–1192. https://doi.org/10.1093/brain/112.5.1171.

[5] B. F. Boeve, A. E. Lang, I. Litvan, Corticobasal degeneration and its relationship to progressive supranuclear palsy and frontotemporal dementia, Ann Neurol. 54 Suppl 5 (2003) S15–19. https://doi.org/10.1002/ana.10570.

[6] B. F. Boeve, D. M. Maraganore, J. E. Parisi*, et al.*, Pathologic heterogeneity in clinically diagnosed corticobasal degeneration, Neurology. 53 (1999) 795–800. https://doi.org/10.1212/wnl.53.4.795.

[7] J. A. Schneider, R. L. Watts, M. Gearing*, et al.*, Corticobasal degeneration: neuropathologic and clinical heterogeneity, Neurology. 48 (1997) 959–969. https://doi.org/10.1212/wnl.48.4.959.

[8] T. Komori, N. Arai, M. Oda*, et al.*, Astrocytic plaques and tufts of abnormal fibers do not coexist in corticobasal degeneration and progressive supranuclear palsy, Acta Neuropathol. 96 (1998) 401–408. https://doi.org/10.1007/s004010050911.

[9] G. K. Wenning, I. Litvan, J. Jankovic*, et al.*, Natural history and survival of 14 patients with corticobasal degeneration confirmed at postmortem examination, J Neurol Neurosurg Psychiatry. 64 (1998) 184–189. https://doi.org/10.1136/jnnp.64.2.184.

[10] D. A. Grimes, A. E. Lang, C. B. Bergeron, Dementia as the most common presentation of

cortical-basal ganglionic degeneration, Neurology. 53 (1999) 1969–1974. https://doi.org/10.1212/wnl.53.9.1969.
[11] M. S. Forman, V. Zhukareva, C. Bergeron, *et al.*, Signature tau neuropathology in gray and white matter of corticobasal degeneration, Am J Pathol. 160 (2002) 2045–2053. https://doi.org/10.1016/s0002-9440(10)61154-6.
[12] N. L. Graham, T. Bak, K. Patterson, *et al.*, Language function and dysfunction in corticobasal degeneration, Neurology. 61 (2003) 493–499. https://doi.org/10.1212/01.wnl.0000081230.09863.ed.
[13] J. R. Hodges, R. R. Davies, J. H. Xuereb, *et al.*, Clinicopathological correlates in frontotemporal dementia, Ann Neurol. 56 (2004) 399–406. https://doi.org/10.1002/ana.20203.
[14] K. Tsuchiya, S. Murayama, K. Mitani, *et al.*, Constant and severe involvement of Betz cells in corticobasal degeneration is not consistent with pyramidal signs: a clinicopathological study of ten autopsy cases, Acta Neuropathol. 109 (2005) 353–366. https://doi.org/10.1007/s00401-004-0966-4.
[15] K. A. Josephs, R. C. Petersen, D. S. Knopman, *et al.*, Clinicopathologic analysis of frontotemporal and corticobasal degenerations and PSP, Neurology. 66 (2006) 41–48. https://doi.org/10.1212/01.wnl.0000191307.69661.c3.
[16] R. Murray, M. Neumann, M. S. Forman, *et al.*, Cognitive and motor assessment in autopsy-proven corticobasal degeneration, Neurology. 68 (2007) 1274–1283. https://doi.org/10.1212/01.wnl.0000259519.78480.c3.
[17] A. Lladó, R. Sánchez-Valle, M. J. Rey, *et al.*, Clinicopathological and genetic correlates of frontotemporal lobar degeneration and corticobasal degeneration, J Neurol. 255 (2008) 488–494. https://doi.org/10.1007/s00415-008-0565-8.
[18] H. Ling, S. S. O'Sullivan, J. L. Holton, *et al.*, Does corticobasal degeneration exist? A clinicopathological re-evaluation, Brain. 133 (2010) 2045–2057. https://doi.org/10.1093/brain/awq123.
[19] S. E. Lee, G. D. Rabinovici, M. C. Mayo, *et al.*, Clinicopathological correlations in corticobasal degeneration, Ann Neurol. 70 (2011) 327–340. https://doi.org/10.1002/ana.22424.
[20] M. J. Armstrong, I. Litvan, A. E. Lang, *et al.*, Criteria for the diagnosis of corticobasal degeneration, Neurology. 80 (2013) 496–503. https://doi.org/10.1212/WNL.0b013e31827f0fd1.
[21] S. K. Alexander, T. Rittman, J. H. Xuereb, *et al.*, Validation of the new consensus criteria for the diagnosis of corticobasal degeneration, J Neurol Neurosurg Psychiatry. 85 (2014) 925–929. https://doi.org/10.1136/jnnp-2013-307035.
[22] I. Aiba, Y. Hayashi, T. Shimohata, *et al.*, Clinical course of pathologically confirmed corticobasal degeneration and corticobasal syndrome, Brain Commun. 5 (2023) fcad296. https://doi.org/10.1093/braincomms/fcad296.
[23] D. W. Dickson, C. Bergeron, S. S. Chin, *et al.*, Office of Rare Diseases neuropathologic criteria for corticobasal degeneration, J Neuropathol Exp Neurol. 61 (2002) 935–946. https://doi.org/10.1093/jnen/61.11.935.
[24] Y. Kanda, Investigation of the freely available easy-to-use software 'EZR' for medical statistics, Bone Marrow Transplant. 48 (2013) 452–458. https://doi.org/10.1038/bmt.2012.244.

[25] J. Müller, G. K. Wenning, M. Verny, *et al.*, Progression of dysarthria and dysphagia in postmortem-confirmed parkinsonian disorders, Arch Neurol. 58 (2001) 259–264. https://doi.org/10.1001/archneur.58.2.259.
[26] N. Eliakim-Raz, T. Babitch, E. Shaw, *et al.*, Risk factors for treatment failure and mortality among hospitalized patients with complicated urinary tract infection: A multicenter retrospective cohort study (RESCUING Study Group), Clin Infect Dis. 68 (2019) 29–36. https://doi.org/10.1093/cid/ciy418.
[27] American Geriatrics Society Ethics Committee and Clinical Practice and Models of Care Committee, American Geriatrics Society feeding tubes in advanced dementia position statement, J Am Geriatr Soc. 62 (2014) 1590–1593. https://doi.org/10.1111/jgs.12924.
[28] T. Stavroulakis, C. J. McDermott, Enteral feeding in neurological disorders, Pract Neurol. 16 (2016) 352–361. https://doi.org/10.1136/practneurol-2016-001408.
[29] Y. Iwasaki, K. Mori, M. Ito, *et al.*, Gastrostomy in patients with prion disease, Prion. 11 (2017) 186–194. https://doi.org/10.1080/19336896.2017.1306164.
[30] Y. Iwasaki, A. Akagi, M. Mimuro, *et al.*, Factors influencing the survival period in Japanese patients with sporadic Creutzfeldt-Jakob disease, J Neurol Sci. 357 (2015) 63–68. https://doi.org/10.1016/j.jns.2015.06.065.