

硬膜移植や脳外科手術歴を有するプリオン病と若年発症脳アミロイドアンギオパチー関連脳出血症例の検討

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Table 1. Distribution of the countries where the patients with cerebral amyloid angiopathy-related cerebral hemorrhage and Creutzfeldt-Jakob disease with histories of neurosurgery or dura mater graft were reported.

Country	CAA-CH	CJD	p
Japan	4 (16.7%)	69 (60.5%)	< 0.001
UK	7 (29.2%)	9 (7.9%)	
Germany	2 (8.3%)	6 (5.3%)	
Spain	0 (0%)	7 (6.1%)	
Belgium	4 (16.7%)	1 (0.9%)	
Austria	1 (4.2%)	4 (3.5%)	
Italy	2 (8.3%)	2 (1.8%)	
France	2 (8.3%)	1 (0.9%)	
Australia	0 (0%)	3 (2.6%)	
Switzerland	0 (0%)	2 (1.8%)	
Netherlands	0 (0%)	2 (1.8%)	
Portugal	1 (4.2%)	0 (0%)	
Croatia	0 (0%)	1 (0.9%)	
New Zealand	0 (0%)	1 (0.9%)	
Korea	0 (0%)	1 (0.9%)	
South Africa	0 (0%)	1 (0.9%)	
Slovenia	1 (4.2%)	0 (0%)	

CAA-CH: cerebral amyloid angiopathy-related cerebral hemorrhage, CJD: Creutzfeldt-Jakob disease, UK: the United Kingdom

Table 2. Comparison of the patients with cerebral amyloid angiopathy-related cerebral hemorrhage and Creutzfeldt-Jakob disease with histories of neurosurgery and dura mater graft.

	CAA-CH	CJD	p
N	24	114	
Sex, female (%)	5 (20.8%)	50 (44.6%)	0.039
Age at the onset of diseases (y)	37.46 ± 7.18 (27-51)	39.79 ± 11.38 (15-68)	0.236
Age at the surgeries (y)	2.95 ± 4.13 (0-17)	28.14 ± 13.07 (1-67)	< 0.001
Incubation period (y)	34.50 ± 5.60 (25-49)	11.56 ± 7.21 (1-30)	< 0.001

CAA-CH: cerebral amyloid angiopathy-related cerebral hemorrhage, CJD: Creutzfeldt-Jakob disease, N: number, y: years

Table 3. Comparison of medical conditions leading to neurosurgeries or dura mater grafts between the patients with cerebral amyloid angiopathy-related cerebral hemorrhage and Creutzfeldt-Jakob disease with histories of neurosurgery or dura mater graft.

	CAA-CH	CJD	p
Brain tumor	3 (12.5%)	51 (45.1%)	< 0.001
Head trauma	13 (54.2%)	20 (17.7%)	
Cerebrovascular disease	0 (0%)	12 (10.6%)	
Arnold-Chiari malformation	1 (4.3%)	9 (8.0%)	
Facial spasm/trigeminal neuralgia	0 (0%)	8 (7.1%)	
Hemangioma	4 (16.7%)	1 (0.9%)	
Others	3 (12.5%)	12 (10.6%)	

CAA-CH: cerebral amyloid angiopathy-related hemorrhage, CJD: Creutzfeldt-Jakob disease

解説

1. 硬膜移植や脳外科手術歴を有するプリオン病は日本に多くその割合は60%を超えるが、硬膜移植や脳外科手術歴を有するCAA関連脳出血症例は英国(29.2%)、日本(16.7%)、ベルギー(16.7%)とその国別の分布に有意な差を認めた。
2. 硬膜移植や脳外科手術歴を有するCAA関連脳出血症例は、硬膜移植や脳外科手術歴を有するプリオン病と比較して、硬膜移植や脳外科手術を受けた年齢が若く、潜伏期間が長かった。
3. 硬膜移植や脳外科手術歴を有するCAA関連脳出血症例は、硬膜移植や脳外科手術歴を有するプリオン病と比較して、硬膜移植や脳外科手術の原因として頭部外傷の頻度が高かった。