

硬膜移植後Creutzfeldt-Jakob病の臨床的特徴の検討

研究代表者: 金沢大学大学院脳老化・神経病態学(脳神経内科学) 山田正仁

Clinical features of pathologically confirmed dCJD

	Plaque type	Non-plaque type	p Value
Patients (n)	17	24	—
Male/Female	8/9	12/12	NS
Site of dura mater graft: supra-/infratentorial	9/6	13/9	NS
The year of dura mater graft (range)	1982-1988	1981-1993	
Age at dura mater grafting, y, mean ± SD (range)	39.6 ± 15.5 (9-58)	41.3 ± 16.3 (16-63)	NS
Incubation period, y, mean ± SD(range)	16.4 ± 4.7 (9-26)	17.8 ± 5.1 (11-27)	NS
Age at onset, y, mean ± SD (range)	56.6 ± 14.1 (26-71)	59.4 ± 15.7 (30-79)	NS
Disease duration, m, mean ± SD (range)	12.4 ± 7.6 (5-38)	3.1 ± 2.4 (1-13)	<0.0001

Manifestations of pathologically confirmed dCJD

	Plaque type (n=17)		Non-plaque type (n=24)		p Value	
	Occurrence (%)	Duration	Occurrence (%)	Duration	Occurrence	Duration
Myoclonus	70.6	7.5 ± 3.2	95.8	2.0 ± 1.1	0.024	<0.0001
Dementia	100	4.5 ± 3.1	95.8	0.7 ± 0.8	NS	<0.0001
Pyramidal signs	41.2	7.8 ± 3.0	87.5	1.9 ± 1.3	0.002	0.001
Extrapyramidal signs	76.5	7.8 ± 4.1	54.2	1.8 ± 0.9	NS	0.001
Cerebellar signs	88.2	1.9 ± 2.6	58.3	0.7 ± 0.9	0.038	NS
Visual disturbance	58.8	4.8 ± 3.4	41.7	1.1 ± 1.3	NS	0.024
Psychiatric symptoms	58.8	3.4 ± 3.2	70.8	0.7 ± 0.9	NS	0.043
Akinetic mutism	70.6	10.9 ± 2.9	95.8	2.7 ± 1.2	0.024	<0.0001
Death	100	14.8 ± 8.0	100	17.7 ± 11.4	—	NS

Laboratory and MRI findings of pathologically confirmed dCJD

	Plaque type(n=17)		Non-plaque type(n=24)		p Value
	No of examined patients	No (%) of patients with positive	No of examined patients	No (%) of patients with positive	
EEG					
PSWCs	17	1 (5.9)	24	24 (100)	<0.0001
CSF					
Positive 14-3-3 protein	8	5 (62.5)	14	11 (78.5)	NS
Positive tau protein	3	2 (66.7)	4	4 (100)	NS
MRI					
T2WI, FLAIR images, or DWI	10	8 (80)	22	19 (86.4)	NS
DWI	9	7 (77.8)	18	15 (83.3)	NS
PrP genotyping					
Codon 129 polymorphism	14	MM 14 (100)	22	MM 22 (100)	—
Codon 219 polymorphism	13	EE 13 (100)	22	EE 20 (91) EK 2 (9)	NS

Comparison of clinical features between codon 219 polymorphism

Codon 219 polymorphism	EE	EK	p Value
Patients (n)	60	5	—
Men/women	24/36	4/1	NS
Site of dura mater graft: supra-/infratentorial	27/28	4/0	NS
The year of dura mater graft (range)	1978-1993	1975-1993	
Age at dura mater grafting, y, mean ± SD (range)	39.8 ± 16.1 (7-64)	35.6 ± 16.7 (16-53)	NS
Incubation period, y, mean ± SD(range)	17.5 ± 5.4 (6-30)	25.4 ± 4.5 (19-30)	0.006
Age at onset, y, mean ± SD (range)	57.6 ± 15.7 (24-81)	61.4 ± 17.9 (35-79)	NS
Disease duration, m, mean ± SD (range)	6.7 ± 6.8 (1-38)	8.8 ± 14.2 (1-34)	NS

解説

1. 硬膜移植後Creutzfeldt-Jakob病(dCJD)の非プラーク型は典型的孤発性CJDに類似した臨床病型を呈するのに対し、プラーク型は緩徐な進行で、早期から小脳失調がみられ、PSWCsの出現頻度が低かった。
2. コドン219多型EK群(グルタミン酸[E]とリジン[K]のヘテロ)の硬膜移植から発症までの潜伏期間はEE群(グルタミン酸[E]のホモ例)と比較して有意に長く、EKヘテロ接合はdCJDの発症を遅らせる可能性が考えられた。