

Supplementary Online Content

Regenhardt RW, Thon JM, Das AS, et al. Association between immunosuppressive treatment and outcomes of cerebral amyloid angiopathy–related inflammation. *JAMA Neurol*. Published online June 22, 2020. doi:10.1001/jamaneurol.2020.1782

eTable 1. Clinical Presentation, Cerebrospinal Fluid, and Immunosuppressive Treatments After all Episodes Combined

eTable 2. Associations With Receiving Immunosuppressive Treatment for First Episode of Cerebral Amyloid Angiopathy–Related Inflammation

eTable 3. Sensitivity Analyses for Recurrence After the First Episode of Cerebral Amyloid Angiopathy–Related Inflammation According to Treatment With Immunosuppressive Agent

eTable 4. Associations of Clinical and Imaging Variables With Pathologic Vessel Wall Inflammation After First Episode of Cerebral Amyloid Angiopathy–Related Inflammation

This supplementary material has been provided by the authors to give readers additional information about their work.

eTable 1. Clinical Presentation, Cerebrospinal Fluid, and Immunosuppressive Treatments After all Episodes Combined

11 of 12 patients treated with cyclophosphamide were also treated with steroids. Other symptoms were atypical for CAA-ri, but additional symptoms or imaging was consistent with diagnostic criteria. Ct count, N total number with available information.

	Total (Ct/N)	Total (%)
Clinical Presentation, All Episodes		
Headache	26/67	39
Cognitive/Behavioral Change	40/67	60
Focal Deficit	11/67	16
Seizure	23/67	34
Other	9/67	13
Cerebrospinal Fluid, All Episodes		
Nucleated Cells >5/ μ L	9/28	32
Protein >45mg/dL	29/31	94
Normal	1/28	3

eTable 2. Associations With Receiving Immunosuppressive Treatment for First Episode of Cerebral Amyloid Angiopathy–Related Inflammation

5 of 6 patients treated with cyclophosphamide were also treated with steroids. *A priori* selected variables with $P < 0.10$ in univariate analyses were included in multivariate analyses. CSF cerebrospinal fluid, OR odds ratio, CI confidence interval.

	OR (95% CI)	P
Univariate		
Age (per year)	1.01 (0.95-1.08)	>0.2
APOE ε4 (per allele)	2.53 (0.95-6.76)	0.064
Any Vessel Wall Inflammation	3.00 (0.28-32.2)	>0.2
CSF Nucleated Cells >5	0.27 (0.03-2.53)	>0.2
T1 Contrast Enhancement	0.83 (0.15-4.54)	>0.2
Fazekas Score	0.91 (0.32-2.59)	>0.2
Acute Infarcts	0.97 (0.15-6.23)	>0.2
Lacunae	0.35 (0.04-2.98)	>0.2
Microbleeds>50	3.50 (0.59-20.7)	0.168
Headache	1.42 (0.39-5.14)	>0.2
Cognitive/Behavioral Change	3.76 (1.02-13.9)	0.047
Focal Deficit	0.13 (0.03-0.63)	0.012
Seizure	2.22 (0.58-8.49)	>0.2
Multivariate		
APOE ε4	3.62 (0.94-14.0)	0.061
Cognitive/Behavioral	10.3 (1.19-88.6)	0.034
Focal Deficit	0.06 (0.00-1.63)	0.096

eTable 3. Sensitivity Analyses for Recurrence After the First Episode of Cerebral Amyloid Angiopathy–
Related Inflammation According to Treatment With Immunosuppressive Agent

Ct count, N total number with available information, HR hazard ratio.

	Total		No Immunosuppression		Any Immunosuppression		HR (95% CI)	P
	Ct/N	%	Ct/N	%	Ct/N	%		
Including All Recurrences	19/48	40	10/14	71	9/34	26	0.19 (0.07-0.48)	<0.0001
Excluding Recurrences <1 Mo	17/46	37	8/12	67	9/34	26	0.22 (0.08-0.59)	0.003
Excluding Recurrences <2 Mo	13/42	31	5/9	56	8/33	24	0.27 (0.09-0.86)	0.026

eTable 4. Associations of Clinical and Imaging Variables With Pathologic Vessel Wall Inflammation After First Episode of Cerebral Amyloid Angiopathy–Related Inflammation

CSF cerebrospinal fluid, OR odds ratio, CI confidence interval.

	OR (95% CI)	P
Age	1.06 (0.95-1.18)	>0.2
APOE ε4	0.67 (0.15-3.08)	>0.2
CSF Nucleated Cells >5	4.00 (0.21-75.7)	>0.2
T1 Contrast Enhancement	2.00 (0.18-22.1)	>0.2
Fazekas Total	0.73 (0.19-2.80)	>0.2
Acute Infarcts	13.5 (0.88-208)	0.062
Lacunae	6.00 (0.39-92.3)	0.199
Microbleeds>50	0.50 (0.04-6.68)	>0.2