Supplemental Table 1 – Demographics and imaging characteristics of CAA patients according to cognition

	Cognitively Normal (n=23)	**MCI (n=10)**	**Dementia (n=25)**	**p-value**
Mean age (years ± std)	67.17 (±10.65)	70.8 (±8.82)	75.88 (±7.49)	***F*(2) = 5.537; p-value = 0.006** [TukeyHSD: Dementia vs Cognitively Normal: p-value = 0.004]
Male(%)	15 (65%)	4 (40%)	10 (40%)	*X*(2) = 3.53; p-value = 0.1711
Hypertension (%)	18 (78%)	8 (80%)	19 (76%)	Fisher's exact test; p-value = 1
Dyslipidemia (%)	16 (70%)	5 (50%)	13 (52%)	Fisher's exact test; p-value = 0.4141
Diabetes (%)	8 (35%)	1 (10%)	4 (16%)	Fisher's exact test; p-value = 0.2162
Lobar hemorrhage	17 (74%)	7 (70%)	18 (72%)	Fisher's exact test; p-value = 1
Cortical superficial siderosis	11 (48%)	7 (70%)	20 (80%)	Fisher's exact test; p-value = 0.063
Lobar Microbleeds				
0	4 (17.4%)	1 (10%)	0 (0%)	Fisher's exact test; p-value = 0.1563
1-5	11 (47.8%)	5 (50%)	10 (40%)	
>5	8 (34.8%)	4 (40%)	15 (60%)	

Supplemental Table 1 – Summarized demographic information for 58 cerebral amyloid angiopathy (CAA) patients. Mean age is represented with standard deviation and male as number and percentage. Cognition as evaluated by a senior neurologist was divided into normal, mild cognitive impairment (MCI) and dementia and frequencies are shown as number and percentage.

Supplemental Table 2 – Intra-class correlation coefficients

Neuroimaging Feature	ICC	95% CI
AC	0.57	[0.33; 0.74]
OF	0.58	[0.34; 0.75]
AT	0.74	[0.57; 0.85]
FI	0.78	[0.63; 0.88]
MTA	0.82	[0.70; 0.90]
Post	0.69	[0.48; 0.82]
ARWMC BG	0.69	[0.50; 0.82]
ARWMC CS	0.79	[0.65; 0.88]
Lobar Microbleeds	0.89	[0.80; 0.94]
EPVS CS	0.77	[0.61; 0.87]
EPVS BG	0.65	[0.44; 0.79]

Supplemental Table 2 – Intra-class correlation coefficients for two raters based on a two-way mixed effects model for consistency. ARWMC – age-related white matter changes; EPVS – enlarged perivascular spaces; BG – basal ganglia and CS – centrum semiovale.

Supplemental Table 3 – Characteristics of CAA with dementia vs CAA without dementia patients

	CAA with dementia (n=25)	**CAA without dementia (n=33)**	**p-value**
Mean age (± std)	75.88 (±7.49)	68.27 (±10.14)	***t*(55.978) = -3.28; p-value = 0.002**
Male(%)	10 (40%)	19 (57%)	*X*(1) = 1.1248; p-value = 0.2889
Hypertension (%)	19 (76%)	26 (79%)	*X*(1) = 2.8592e-31; p-value = 1
Dyslipidemia (%)	13 (52%)	21 (64%)	*X*(1) = 0.387; p-value = 0.534
Diabetes (%)	4 (16%)	9 (27%)	*X*(1) = 0.387; p-value = 0.534
Lobar hemorrhage	18 (72%)	24 (73%)	*X*(1) = 8.0416e-31; p-value = 1
Cortical superficial siderosis	20 (80%)	18 (55%)	*X*(1) = 3.0305; p-value = 0.082
Lobar Microbleeds			
0 **1-5** **>5**	0 (0%) 10 (40%) 15 (60%)	5 (15.2%) 16 (48.5%) 12 (36.4%)	Fisher's exact test; p-value = 0.057
Basal ganglia ARWMC (± std)	2 [IQR: 1]	1.5 [IQR: 1]	***W*(573); p-value = 0.01**
Centrum Semiovale ARWMC (± std)	2 [IQR: 1.5]	1.5 [IQR: 1]	***W*(542); p-value = 0.04**

Supplemental Table 3 – Demographical and imaging characteristics of cerebral amyloid angiopathy (CAA) patients without dementia (normal cognition and MCI) and with dementia. ARWMC – age-related white matter changes.

Supplemental Table 4 – Characteristics of CAA with dementia vs CAA without dementia patients according to sex

	Female without dementia (n=14)	**Male without dementia (n=19)**	**Female with dementia (n=15)**	**Male with dementia (n=10)**	**p-value**
Mean age (years ± std)	66.07 (±10.92)	69.89 (±8.82)	75.67 (±9.49)	76.2 (±8.08)	***F*(3) = 3.758; p-value = 0.016** [TukeyHSD: Male with Dementia vs Female without Dementia; p-value = 0.047; Women with Dementia versus Women without Dementia: p-value = 0.032]
Hypertension (%)	11 (79%)	15 (79%)	12 (80%)	7 (70%)	Fisher's exact test; p-value = 0.9199
Dyslipidemia (%)	9 (64%)	12 (63%)	11 (73%)	2 (20%)	Fisher's exact test; p-value = 0.057
Diabetes (%)	4 (29%)	5 (26%)	2 (13%)	2 (20%)	Fisher's exact test; p-value = 0.7828
Lobar hemorrhage	12 (86%)	12 (63%)	11 (73%)	7 (70%)	Fisher's exact test; p-value = 0.6067
Cortical superficial siderosis	8 (57%)	10 (53%)	12 (80%)	8 (80%)	Fisher's exact test; p-value = 0.2806

Lobar Microbleeds					
0	2 (14.3%)	3 (15.8%)	0 (0%)	0 (0%)	Fisher's exact test; p-value = 0.4602
1-5	6 (42.9%)	10 (52.6%)	6 (40%)	4 (40%)	
>5	6 (42.9%)	6 (31.6%)	9 (60%)	6 (60%)	

Supplemental Table 4 – Demographical and imaging characteristics of cerebral amyloid angiopathy (CAA) patients without dementia (normal cognition and MCI) and with dementia by sex.

Supplemental Table 5 – Medial temporal lobe atrophy: Multiple regression coefficients and p-values

Dependent Variable	Intercept	Age	Sex	cSS	Dementia	Dementia* Sex
MTA (%)	-31.559	**0.971****	-0.843	0.823	0.802	**21.369***

Supplemental Table 5 – Multiple linear regression with medial temporal atrophy (%), as measured by the visual rating score, as the dependent variable and age, sex (male = 1), presence of cortical superficial siderosis, presence of dementia and a dementia*sex interaction term as independent variables. Values represent coefficient estimates. Statistical significance considered at p-values < 0.05. p-values: *< 0.05, ** < 0.01 and *** < 0.001.

Supplemental Table 6 – Contrast tests between groups for medial temporal lobe atrophy

Contrast Tests	Estimates	Standard Error	p-values (Bonferroni correction)
Male Dem vs Female Dem	20.526	7.10162	**0.0246**
Male Dem vs Male no Dem	22.171	7.28875	**0.0118**
Female Dem vs Female no Dem	0.802	7.005	1.0000
Male no Dem vs Female no Dem	-0.843	6.323	1.0000

Supplemental Table 6 – Contrast coefficients based on multiple linear regression in Supplemental Table 5. Coefficient estimates are shown with standard error and p-values after Bonferroni correction for multiple comparisons. Statistical significance considered at p-values < 0.05 after bonferroni correction. Male no Dem: Male, no dementia; Female no Dem: Female, no dementia; Male Dem: Male, dementia; Female Dem: Female, dementia.

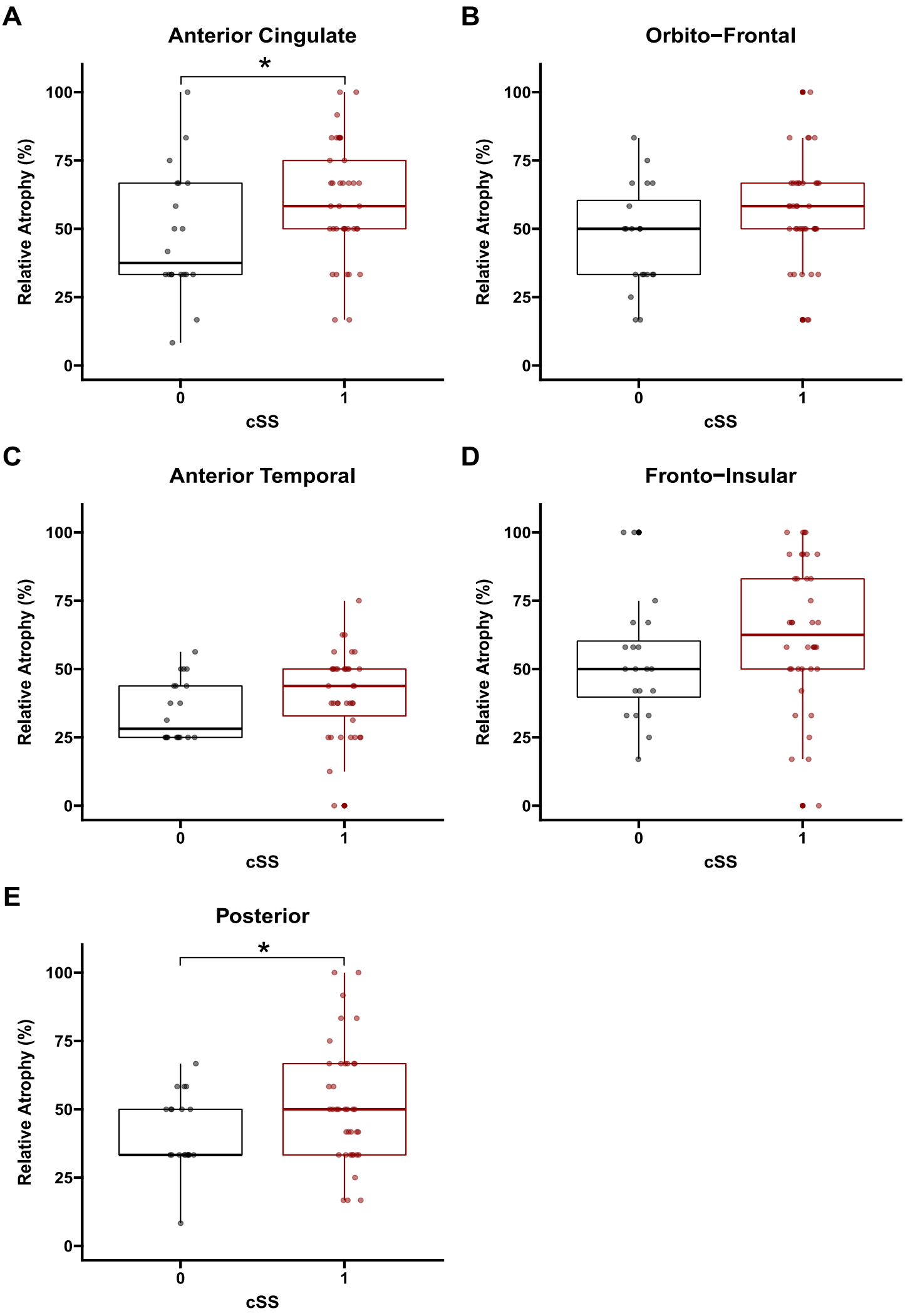
A
Anterior Cingulate
*
Relative Atrophy (%)
100
75
50
25
0
0
1
cSS
B
Orbito−Frontal
Relative Atrophy (%)
100
75
50
25
0
0
1
cSS
C
Anterior Temporal
Relative Atrophy (%)
100
75
50
25
0
0
1
cSS
D
Fronto−Insular
Relative Atrophy (%)
100
75
50
25
0
0
1
cSS
E
Posterior
*
Relative Atrophy (%)
100
75
50
25
0
0
1
cSS

Figure S1

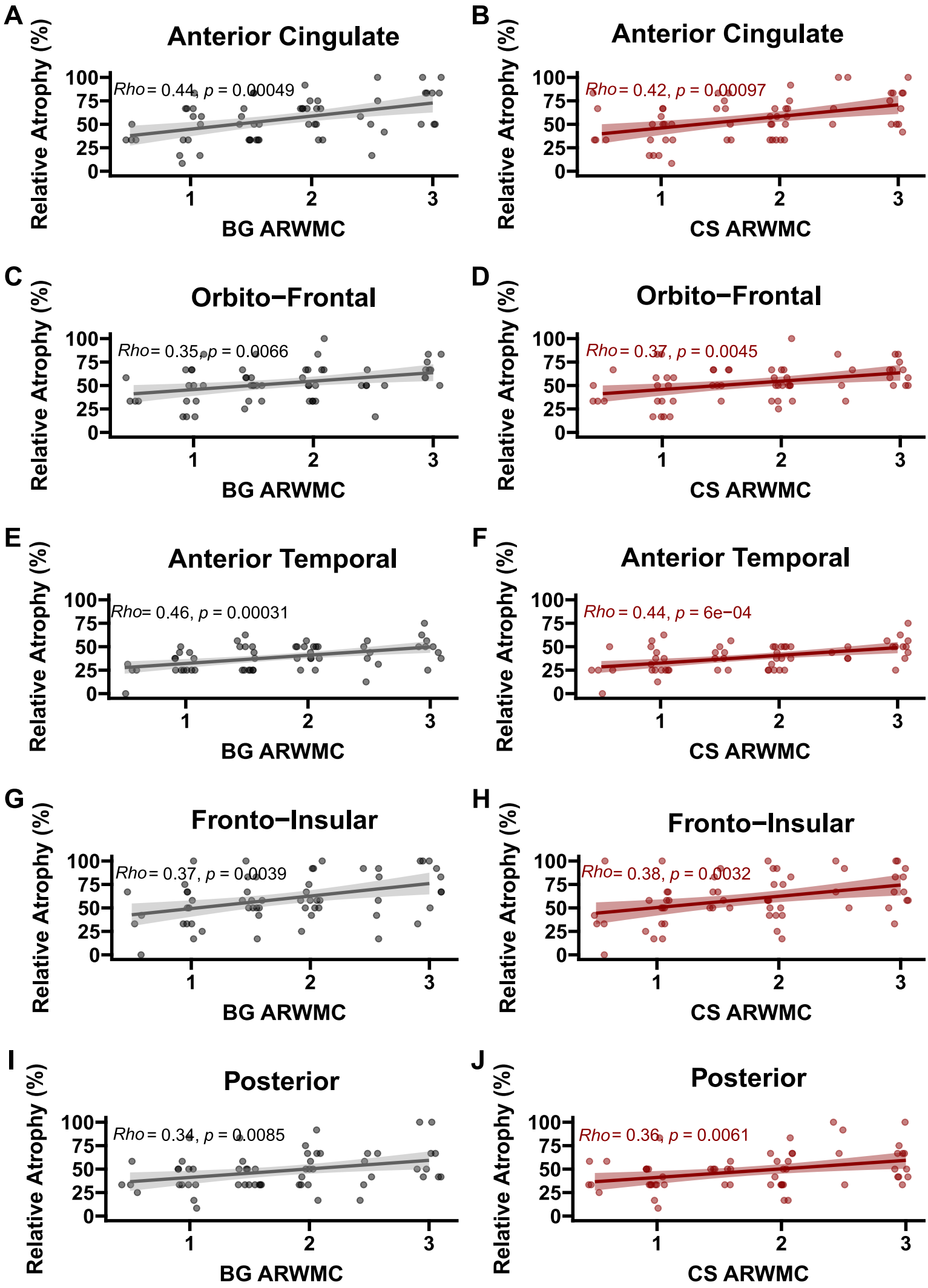
A
Anterior Cingulate
Rho = 0.44, p = 0.00049
Relative Atrophy (%)
BG ARWMC
B
Anterior Cingulate
Rho = 0.42, p = 0.00097
Relative Atrophy (%)
CS ARWMC
C
Orbito−Frontal
Rho = 0.35, p = 0.0066
Relative Atrophy (%)
BG ARWMC
D
Orbito−Frontal
Rho = 0.37, p = 0.0045
Relative Atrophy (%)
CS ARWMC
E
Anterior Temporal
Rho = 0.46, p = 0.00031
Relative Atrophy (%)
BG ARWMC
F
Anterior Temporal
Rho = 0.44, p = 6e−04
Relative Atrophy (%)
CS ARWMC
G
Fronto−Insular
Rho = 0.37, p = 0.0039
Relative Atrophy (%)
BG ARWMC
H
Fronto−Insular
Rho = 0.38, p = 0.0032
Relative Atrophy (%)
CS ARWMC
I
Posterior
Rho = 0.34, p = 0.0085
Relative Atrophy (%)
BG ARWMC
J
Posterior
Rho = 0.36, p = 0.0061
Relative Atrophy (%)
CS ARWMC

Figure S2

Supplemental Figure 1

Visual Rating Scores of all brain regions according to the presence or absence of Cortical Superficial Siderosis (cSS). Statistical significance considered at p-values < 0.05.

Supplemental Figure 2

Spearman correlation analysis of Visual Rating Scores of all brain regions with Basal Ganglia ARWMC Score (BG ARWMC); Centrum Semiovale ARWMC Score (CS ARWMC). Spearman correlation coefficient (Rho). Statistical significance considered at p-values < 0.05.