

Supplemental material

Title

Isolated anterior cerebral artery occlusion: an atypical form of moyamoya disease

Journal name

Stroke and Vascular Neurology

Table S1. Comparison of the primary symptoms and genotype in adult and pediatric patients

First admission	Age < 18 years old (n=27)	Age ≥ 18 years old (n=112)	P value
Female	14(51.9%)	65(58.0%)	0.560
Primary symptom at onset			
Asymptomatic	0	1(0.9%)	1.000
Transient ischemic attack	19 (70.4%)	68(60.7%)	0.352
Infarction	3(11.1%)	12(10.7%)	1.000
Haemorrhage	4(14.8%)	20(18.3%)	0.881
Headache	1(3.7%)	5(4.6%)	1.000
Others	0	6(5.5%)	0.599
Mutation †			0.066
GG	16/25(64.0%)	74/91(81.3%)	
GA	9/25(36.0%)	17/91(18.7%)	

GG, wildtype *RNF213* p.R4810K; GA, heterozygous *RNF213* p.R4810K; ACA, anterior cerebral artery; PCA, posterior cerebral artery; mRS, modified Rankin scale; MMD, moyamoya disease.

†*RNF 213* p.R4810K (rs112735431) gene mutation analysis was performed in 116 of the 139 patients

Table S2. Neuropsychological test performances of patients with ACA-occlusive MMD

	Number (n=65)	%
MoCA (Mean±SD)	21.82±4.21	
<10	0	0
10–17	10	15.4
18–26	37	56.9
≥26	18	27.7
IADL (Mean±SD)	14.48±1.89	
<16	59	90.8
16–22	5	7.7
≥22	1	1.5
HAMD-17 (Mean±SD)	4.37±3.80	
<7	49	75.4
7–17	16	24.6
18–24	0	0
>24	0	0

MoCA, Montreal Cognitive Assessment; IADL, Instrumental Activity of Daily Living Scale; HAMD, 17-item Hamilton Rating Scale for Depression.

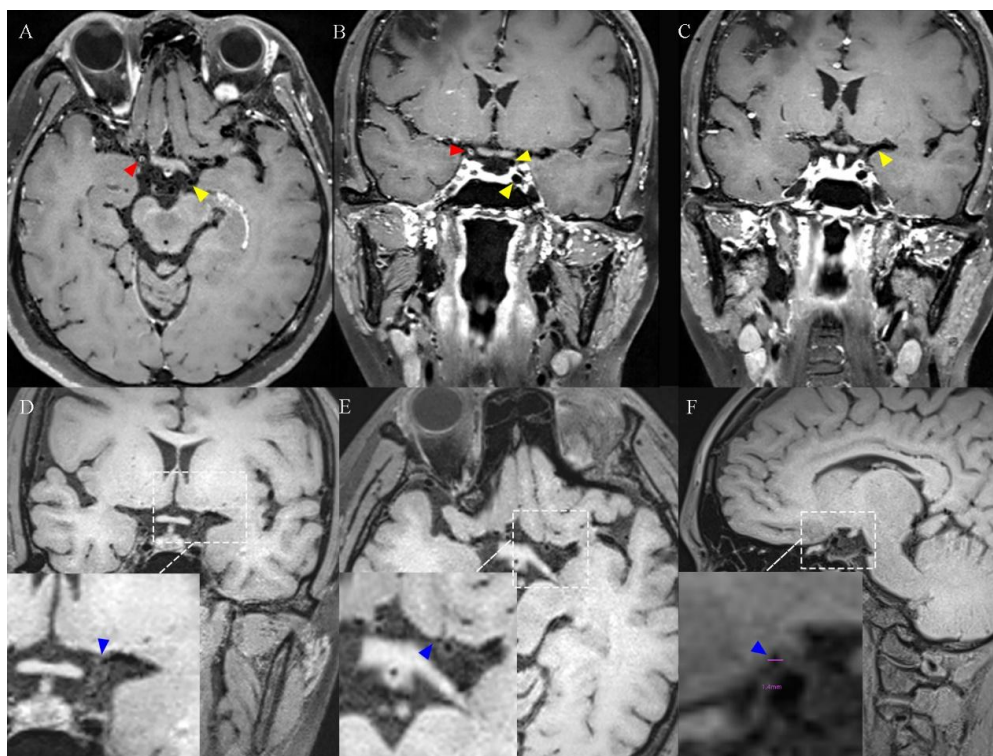


Figure S1. The radiological characteristics of high-resolution MRI in ACA-occlusive MMD patients. a-c shows the HR-MRI diagram of the ICA in ACA-occlusive MMD. The red arrows indicate a centripetal constriction of the distal segment of the internal carotid artery in the contralateral hemisphere, while the yellow arrows indicate that there is usually no significant lesion in the ipsilateral ICA and MCA. d-f shows the HR-MRI image of ACA occlusion. The blue arrows indicates a centripetal constriction at the proximal section of the ACA. The outer diameter of the ACA here is 1.40 mm. MRI, magnetic resonance imaging; HR, high resolution; ACA, anterior cerebral artery; MMD, moyamoya disease; ICA, internal carotid artery.