

Background and objectives

Intracranial arterial dolichoectasia (IADE) is defined as an increase in the length and diameter of at least one intracranial artery. IADE more often involves the posterior than the anterior circulation, being the basilar artery affected in 80% of cases. Some evidence suggests a possible association with cerebral small vessel disease (SVD) [1] and extracranial arterial abnormalities [2]. Our aims were to identify patients with IADE and describe: 1) clinical and neuroimaging SVD-related features; 2) the possible coexistence of extracranial arteriopathy.

Materials and methods

Starting from December 2014, we prospectively identified IADE patients among those attending the Florence VAS-COG Clinic and Stroke Unit of our Hospital. If IADE was identified, we assessed: familiar and clinical history, with particular focus on stroke and vascular risk factors profile; neuropsychological performances; brain MRI, with visual rating of SVD features, including white matter hyperintensities (WMH), count of lacunar infarcts, perivascular spaces, and, if T2* gradient-echo sequences were available, count of microbleeds. Each patient underwent a neck, thoraco-abdominal aorta, and lower limbs CT angiography, according to a predefined protocol.

Results

	♂, 68 yrs	♂, 68 yrs	♂, 75 yrs	♂, 65 yrs	♂, 77 yrs
Familiar history of stroke	No	Yes	Yes	No	No
Current smokers	No	No	No	No	No
Hypertension	No	Yes	Yes	Yes	Yes
Diabetes mellitus	No	No	Yes	No	No
Hypercholesterolemia	No	Yes	No	Yes	No
Previous myocardial infarction	No	No	No	Yes	Yes
Previous stroke	No	Yes*	Yes*	Yes*	Yes*
Cognitive decline	Yes (dementia)*	No	Yes (mild cognitive impairment)	Yes (dementia)	Yes (mild cognitive impairment)

*reason for referral

Neuroimaging

WMH (Fazekas scale, FS)	Severe (FS=3)	Mild (FS=1)	Moderate (FS=2)	Severe (FS=3)	Moderate (FS=2)
Lacunar infarcts	4	>10	1	9	>10
Perivascular spaces	>20	>10	>10	>10	>20
Microbleeds	10	Not available	>50	10	Not available

Intracranial vessels

IADE	Basilar artery dolichoectasia	Basilar artery dolichoectasia Carotid siphons ectasia Middle cerebral artery ectasia	Basilar artery dolichoectasia	Basilar artery dolichoectasia Middle cerebral arteries ectasia	Basilar artery dolichoectasia

Extracranial vessels

Arterial abnormalities	No enlargement	Thoracic aortic ectasia Abdominal aortic ectasia	Thoracic aortic ectasia Abdominal aortic ectasia	Abdominal aortic ectasia Iliac arteries ectasia	Abdominal aortic ectasia Iliac arteries ectasia

Discussion and conclusion

Preliminary data from this small group of IADE patients confirm the possible association with cerebral SVD. Systemic arterial abnormalities may coexist and deserve consideration by neurologists. These data are in line with the current hypothesis that IADE should be considered as a disease entity, different from atherosclerosis, in which the involvement of the brain-supplying arteries may be only part of a systemic arteriopathy [3]. The relationship with cerebral SVD needs further evaluation.

References

- [1] Pico F, Labreuche J, Touboul PJ, Leys D, Amarenco P; GENIC investigators. Intracranial arterial dolichoectasia and small-vessel disease in stroke patients. *Ann Neurol* 2005;57:472-479.
 [2] Pico F, Labreuche J, Cohen A, Touboul PJ, Amarenco P; GENIC investigators. Intracranial arterial dolichoectasia is associated with enlarged descending thoracic aorta. *Neurology* 2004;63:2016-2021.
 [3] Pico F, Labreuche J, Amarenco P. Pathophysiology, presentation, prognosis, and management of intracranial arterial dolichoectasia. *Lancet Neurol* 2015;14:833-845.