Epidemiology, pathophysiology, diagnosis, and management of intracranial artery dissection

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Lancet Neurol 2015; 14: 640–54

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Spontaneous intracranial artery dissection is an uncommon and probably underdiagnosed cause of stroke that is defined by the occurrence of a haematoma in the wall of an intracranial artery. Patients can present with headache, ischaemic stroke, subarachnoid haemorrhage, or symptoms associated with mass effect, mostly on the brainstem. Although intracranial artery dissection is less common than cervical artery dissection in adults of European ethnic origin, intracranial artery dissection is reportedly more common in children and in Asian populations. Risk factors and mechanisms are poorly understood, and diagnosis is challenging because characteristic imaging features can be difficult to detect in view of the small size of intracranial arteries. Therefore, multimodal follow-up imaging is often needed to confirm the diagnosis. Treatment of intracranial artery dissections is empirical in the absence of data from randomised controlled trials. Most patients with subarachnoid haemorrhage undergo surgical or endovascular treatment to prevent rebleeding, whereas patients with intracranial artery dissection and cerebral ischaemia are treated with antithrombotics. Prognosis seems worse in patients with subarachnoid haemorrhage than in those without.

Introduction

Cervicocephalic artery dissection, which corresponds with a haematoma in the wall of a cervical or an intracranial artery, is an important cause of stroke in children and young and middle-aged adults.[1–3] Although dissection of the extracranial cervical arteries has been extensively studied and described,[4–12] less information is available about pure intracranial artery dissection (ie, not including the cervical portion of the artery).[4] Early reports were exclusively based on autopsy series, hence biased towards the most severe cases of intracranial artery dissection.[13,14] Several possible reasons are available for the absence of information about intracranial artery dissections. First, intracranial artery dissection happens less frequently than cervical artery dissection in non-Asian countries, where the largest series of patients who had cervical artery dissection have been reported so far.[9–12] Second, patients who have cervical artery dissection and mainly present with headache, cervical pain, and ischaemic stroke are mostly seen by neurologists, whereas patients with intracranial artery dissection can also develop a subarachnoid haemorrhage and are therefore managed not only by neurologists, but also by neurosurgeons and interventional neuroradiologists, all of whom might have an incomplete picture of the disease. As a result, no consensus is agreed on for the diagnostic criteria and optimum treatment of patients with intracranial artery dissections.

In this Review we provide a comprehensive overview of reported studies into the epidemiology, pathophysiology, diagnosis, management, and outcome of spontaneous intracranial artery dissections, in addition to proposing a consensus statement by a group of international experts from various specialties and countries about the diagnosis and management of intracranial artery dissections.

Epidemiology

The incidence of intracranial artery dissections is unknown, but is probably lower than that of symptomatic cervical artery dissection (2·6–3·0 per 100 000 people per year[15,16]) in populations of European ethnic origin. The proportion of intracranial artery dissections in all cervicocephalic dissections substantially varies between ethnic origin and age groups, and also depends on study recruitment strategies and ascertainment methods used. Recruitment of patients for studies through neurology departments is biased towards those with cervical artery dissection or intracranial artery dissection without subarachnoid haemorrhage, whereas patient recruitment through departments of neurosurgery or interventional neuroradiology is biased towards intracranial artery dissection with subarachnoid haemorrhage. In a series of 195 patients with vertebral artery dissections who were recruited in neurology departments in France and Switzerland, only 11% of dissections were located exclusively in the intracranial portion of the artery.[17] In a Mexican study[18] of 100 patients admitted to a neurology department for vertebral artery dissection with ischaemic stroke and without subarachnoid haemorrhage, 27 (27%) patients had intracranial artery dissection. In studies undertaken in east Asia,[19–23] in which patients were mostly recruited through neurosurgery and interventional neuroradiology departments, intracranial artery dissection

accounted for up to 67–78% of all cervicocephalic artery dissections.[19,20] Most reported series of patients with intracranial artery dissection are from Asia (95% of studies including >40 patients with intracranial artery dissection, and 61% of studies including 20–39 patients with this disorder). Whether this suggests publication bias, differences in disease prevalence across ethnic origin groups, or both, is unclear.

Intracranial artery dissections can also affect children, but a dearth of scientific literature exists about this problem in children, given the rarity of childhood stroke. In a North American single-centre series[24] of

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	N	Country origin (department)	Imaging method	Mean age (range)	Sex	Anatomical location	Presenting symptoms
All types							
Yamaura et al (2000)[23]	357	Japan (neurosurgery survey)	DSA	51 (8–86) years (SAH 53 years, non-SAH 49 years)	Ratio of men to women 2:1; with non-SAH 2·6:1	3% anterior circulation (SAH 2%, non-SAH 5%); 97% posterior circulation (SAH 98%, non-SAH 95%), in VA (261 patients), BA (22 patients), ICA (ten patients), or other artery (29 patients)*	SAH (206 patients [58%]), cerebral ischaemia (112 patients [31%]), headache alone (26 patients [7%]), other (13 patients [4%])
Mizutani (2011)[29]	190	Japan (neurosurgery, radiology)	MRA, DSA, or CTA	49 (0–74) years (SAH 52 [0–65] years, non-SAH 45 [22–47] years)	69% men (SAH 62%, non-SAH 77%)†	14% anterior circulation (SAH 11%, non-SAH 14%); 88% posterior circulation (SAH 89%, non-SAH 86%), VA (155 dissections), PICA (11 dissections), ACA (11 dissections), BA (ten dissections), MCA (eight dissections)†	SAH (108 dissections [52%]), headache, or cerebral ischaemia (98 dissections [48%])†
Ono et al (2013)[30]	143	Japan (neurosurgery)	DSA and CT in all patients, MRI in some patients	51 (7–82) years (SAH 53 [31–83] years, non-SAH 48 [10–74] years)	59% men (SAH 58%, non-SAH 61%)	22% anterior circulation (SAH 15%, non-SAH 32%); 78% posterior circulation (SAH 85%, non-SAH 68%), VA (99 patients; 16 [11%] IADs in the VA were bilateral), ACA (11 patients), MCA (11 patients), ICA (eight patients), BA (seven patients), PICA (five patients), PCoA (one patient), PCA (one patient)	SAH (86 patients [60%]), headache, or cerebral ischaemia (57 patients [40%])
Kwak et al (2011)‡[19]	92	South Korea (radiology)	DSA	51 years§	58% men§	24% anterior circulation (SAH 7%, non-SAH 44%); 76% posterior circulation (SAH 93%, non-SAH 57%)	SAH (25 patients [27%]), SAH and ischaemia (three patients [3%]), infarction (20 patients [22%]), other cerebrovascular symptoms (44 patients [48%])
Metso et al (2007)[31]	45¶	Finland (neurology, neurosurgery)	SAH: DSA (50%) or CTA (50%); non-SAH: MRA (100%), MRI (96%), US (39%), or CTA (9%)	46 (21–67) years (SAH 51 [32–67] years, non-SAH 42 [21–56] years)	58% men (SAH 50%, non-SAH 65%)	16% anterior circulation (SAH 14%, non-SAH 22%); 84% posterior circulation (SAH 86%, non-SAH 78%), VA (28 patients; one [2%] IAD in the VA was bilateral), ICA (five patients), BA (four patients), PICA (three patients), ACA (two patients), SCA (one patient), PCA (one patient), pericallosal artery (one patient)	SAH (22 patients [49%]), headache, or cerebral ischaemia (23 patients [51%])¶
Vertebrobasilar IAD							
Ahn et al (2012)‡[32]	210	South Korea (neurosurgery, radiology)	DSA in all; CTA, MRI, or MRA in some	Median 47 (21–80) years (SAH 45 years, non-SAH 48 years)	61% men	Vertebrobasilar IAD included: 20 (10%) IAD in the VA were bilateral	SAH (48 patients [21%]), non-SAH (182 patients [79%]; ischaemia frequency unknown)
Kim et al (2011)‡[33]	111	South Korea (neurosurgery, radiology)	DSA	45 (24–78) years	63% men	Vertebrobasilar IAD included: BA involved (ten), PICA involved (47), eight (7%) IADs were bilateral	SAH (73 patients [66%]), ischaemia, or headache (38 patients [34%])
Matsukawa et al (2012)‡[34]	103	Japan (neurosurgery)	MRI, MRA, CTA, DSA	53 (IQR 45–66) years (SAH 50 [46–59] years, non-SAH 54 [45–69] years)	69% men (SAH 77%, non-SAH 67%)	Vertebral IAD included: three (3%) IADs were bilateral	SAH (22 patients [21%]), ischaemia, or headache (81 patients [79%])
Kashiwasaki et al (2013)[35]	73	Japan (neurosurgery)	Not specified	52 (SD 9) years	55% men	Vertebral IAD without PICA involvement	SAH (45 patients [62%]), non-SAH (28 patients [38%], asymptomatic, or headache)

(Table 1 continues on next page)