Spontaneous Anterior Intracranial Artery Dissection: An Important Cause of Stroke in Young People

Intracranial artery dissection (IAD) is rarely reported and possibly underdiagnosed. We present a case of spontaneous right middle cerebral artery dissection causing repeated small ischaemic lesions in the right hemisphere, presenting with frequent, mild intermittent left-sided neurological symptoms and right-sided headache in a 28-year-old female. The presentation was subtle and diagnosis unusual, highlighting the importance of considering dissection as a cause of neurological deficits with associated headache in young people. Contrast-enhanced magnetic resonance angiography (CE-MRA) is fast and effective and is the recommended imaging modality for detecting vascular pathology.

Introduction

IAD causing ischaemic stroke or subarachnoid haemhorrhage is rare but possibly under-reported, with only 10-20 documented cases to date.[1-3] The mean reported age of IAD is 25 years,[4] however IAD has also been reported in children.[5,6] Predisposing risk factors include; preceding trauma and collagen disorders such as fibromuscular dysplasia , cystic medial necrosis,[7] moya moya disease, Marfan's syndrome and Ehlers-Danlos syndrome type IV. Common vascular risk factors (hypertension, smoking, diabetes mellitus, hyperlipidaemia and oral contraceptives) have also been implicated in the pathogenesis of arterial dissection. There are individual case reports of intracranial dissection in the context of orgasmic headache,[8] post-coitus[9] and post partum,[10] but in the majority of cases no cause is found.

Anterior intracranial dissections typically present with ipsilateral headache and a contralateral neurological deficit with altered consciousness. Presentation with subarachnoid bleeding secondary to intracranial dissection is more common in the posterior circulation. Pseudoaneurysm formation is another complication resulting from blood tracking through the media to the subadventitial layer and causing dilatation of the outer wall of the vessel, which tends to occur more commonly in the posterior circulation but has also been known to occur in the anterior circulation.[11] Previously, those reported in the literature have usually presented with significant morbidity and the diagnosis in some has only been realised at post-mortem. Those presenting with mild transient ischaemic attacks are vanishingly rare, or perhaps go undiagnosed.

The diagnostic imaging modalities for intracranial artery dissection include formal catheter digital subtraction angiographic techniques (DSA). Less invasive techniques include conventional MRA revealing a 'rat's tail' or 'string sign' or T1-weighted axial MRI revealing a double lumen or intramural thrombus.[12] Contrast-enhanced MRA can be performed efficiently in a single breath hold and is comparable to DSA in providing diagnostic information of body arteries[13] and provides more extensive and accurate information than conventional MRA. Computerised tomography angiogram (CTA) can also be an adequate fast screening modality for cerebral artery pathology, especially with modern 3-D digital subtraction techniques.

We present a case of spontaneous right middle cerebral artery dissection, which resulted in repeated small cerebral ischaemic insults in a young female. This presented with relatively subtle symptoms and signs, which could have been easily missed if not considered in the differential diagnosis. See Table 1 for summary characteristics of intracranial dissections.

Table 1: Key points in intracranial artery dissection.

1. *An important cause of TIA/stroke in young people (mean age 25 years old).*
2. *Pain (unilateral headache) is a predominant presenting symptom.*
3. *Intracranial artery dissection is less common than extracranial artery dissection.*
4. *Posterior circulation (vertebrobasilar artery) dissection is more common and more likely to be associated with subarachnoid bleeding than anterior circulation dissection.*
5. *Risk factors for cerebral artery dissection include trauma, collagen disorders (fibromuscular dysplasia, cystic medial necrosis, Marfan's syndrome, Ehlers-Danlos syndrome type IV), and common vascular risk factors (hypertension, smoking, diabetes mellitus, hyperlipidaemia and oral contraceptives).*
6. *Contrast-enhanced MRA is the most efficient imaging modality of choice with comparable diagnositic yield to formal angiography.*
7. *Treatment options include surgery, stenting, anticoagulation and antiplatelets, although evidence for favouring one option over another is not yet available.*

Case report

A 28-year-old female presented with a one month history of progressive left sided sensory symptoms. This started with intermittent numbness in her left hand, involving the thumb and first two fingers, followed by left facial numbness and then left leg numbness. These sensory symptoms would last a few minutes at a time over a period of several weeks. At the time of initial consultation, she had mild weakness of her left leg and face. The patient also described a continuous right frontal throbbing headache. There was no previous history of headache but her mother was known to suffer migraines. Her only medication was the oral combined contraceptive pill. Blood pressure was 135/90 with no other vascular risk factors.

An initial MRI brain revealed some high signal abnormalities, with one predominant lesion in the right corona radiata suggestive of inflammation or ischaemia (Figure 1a). A lumbar puncture was acellular with a normal protein level and an absence of intrathecal oligoclonal band synthesis. An interval brain scan three months later showed similar findings with the addition of a further new lesion in the right peritrigonal area. Unusual radiological features on the second scan included restriction of the lesions to the territory of the right middle cerebral artery and a cavitating appearance of some of the lesions which was more in keeping with ischaemia than inflammation (Figure 1(a) and 1(b)).

To investigate further, a contrast-enhanced MRA (CE-MRA) was performed. This provided views from the aortic arch to the circle of Willis and other intracranial vessels, not routinely included on the normal field of view when assessing the neck vessels in patients with ischaemic

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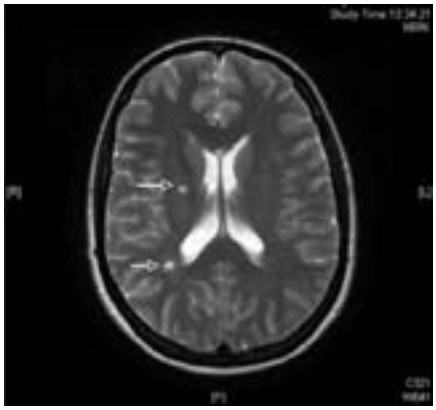

Figure 1(a): T2-weighted axial MRI brain showing high signal lesions in the right hemisphere (white arrows).

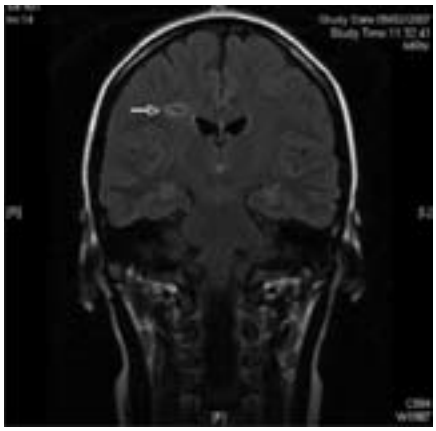

Figure 1(b): T1-weighted MRI brain coronal showing cavitating lesion in right hemisphere (white arrow).

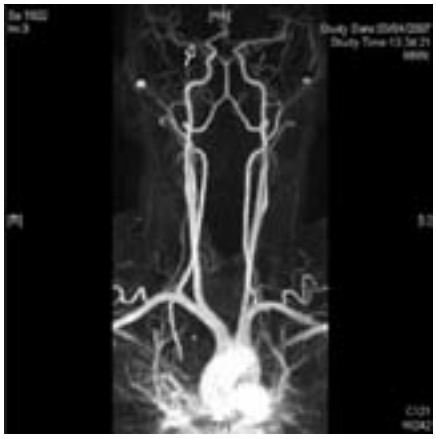

Figure 1(c): CEMRA reconstructed frontal view demonstrates normal carotid bifurcations and internal carotid arteries and a right M1 stenosis with reduced signal in the middle cerebral circulation (white arrow).

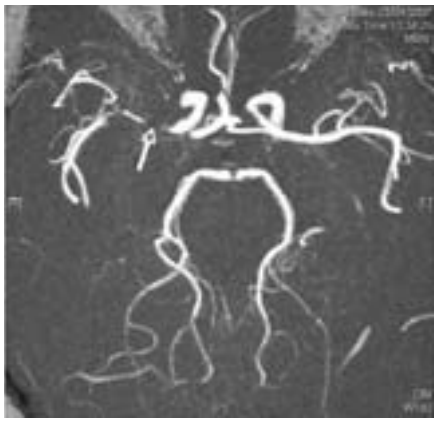

Figure 1(d): A maximum intensity submentovertical projection reconstruction 3D TOF MR angiogram centred on the Circle of Willis confirms the presence of a right M1 stenosis, likely to be the result of a dissection. (white arrow).

stroke. The CEMRA demonstrated a significant M1 stenosis with reduced signal indicating reduced flow in the more middle cerebral circulation (Figure 1c). 3D time of flight (TOF) MR angiography confirmed the M1 stenosis (a 'string sign') consistent with an arterial dissection (Figure 1d), and assumed to be responsible for repeated ischaemic insults to the right cerebral hemisphere.

Aspirin was started (75mg once a day) and the patient was advised to stop the oral combined contraceptive pill and attend regular review. At the latest review, she continued to complain of headache and residual mild left-sided sensory symptoms but the initial left facial and left leg weakness had fully recovered.

Discussion

Intracranial artery dissections can cause subarachnoid haemorrhage and ischaemic infarcts. These sequalae have often only been diagnosed at post-mortem in the past but with improved imaging techniques and awareness of this condition, intracranial artery dissection is likely to become a more familiar diagnosis. As thrombolysis is a treatment option for presumed thrombotic stroke, but not necessarily advisable with dissection, the aetiology of stroke or transient ischaemic attacks in a young person should always be carefully considered.

The mechanism of ischaemia in intracranial artery dissection is thought to be hypoperfusion due to artery occlusion rather than embolic events, which are more typical of extracranial artery dissections. The anatomy of the intracranial arterial wall differs from cervical arteries in that the media is thinner and there is no external elastic lamina. Haemorrhage as a result of dissection can occur between the arterial layers or directly across the lumen wall and as intracranial arterial walls are thinner the risk of subarachnoid bleeding is thought to be higher.[4]

Due to the rare occurrence of intracranial dissection, there is currently no evidence base to guide treatment. Options include surgery, stenting, anticoagulation and antiplatelet therapy. We favoured antiplatelet therapy over anticoagulation given the belief that there is a greater risk of intracranial bleeding. There is an obvious need for a randomised, controlled study of treatments in intracranial artery dissection.

The evidence base for treatment of extracranial dissection is larger but still inadequate.[14,15] In order to try to answer whether we should be using antiplatelets or anticoagulation for the treatment of extracranial artery dissection, there is currently a national pilot study underway to determine whether it would be possible to perform a large-scale multi-centre, international randomised study which would aim to recruit enough patients to provide statistical power to be able to reliably compare antiplatelet with anticoagulation treatment[16] (Cervical Artery Dissection in Stroke Study: CADISS - personal communication, Professor John Norris, St George's Hospital, London www.dissection.co.uk).

As imaging availability and techniques advance, contrast-enhanced MRA is seriously challenging DSA as the primary vascular imaging modality. It enables fast and reliable diagnosis of intra and extracranial artery dissection, revealing the true prevalence, course and outcome of this condition and is the recommended imaging modality of choice in suspected arterial dissection.

References

1. Chaves C, Estol C, Esaola MM, et al. *Spontaneous intracranial internal carotid artery dissection.* Arch Neurol. 2002;59:977-81.
2. Sharif AA, Remley KB, Clark HB. *Middle cerebral artery dissection: a clinicopathological study.* Neurology. 1995;45:1929-31.
3. Lee JS, Bang OY, Lee PH, et al. *Two cases of spontaneous middle cerebral arterial dissection causing ischemic stroke.* Journal of the Neurological Sciences. 2006;250(1-2):162-6.
4. Farrell M, Gilbert J, Kaufmann J. *Fatal intracranial arterial dissection: clinical pathological correlation.* Journal of Neurology, Neurosurgery and Psychiatry. 1985;48:111-21.
5. Duncan IC. *Clinically occult chronic dissecting aneurysm of the superior cerebellar artery in a child.* Pediat Radiol. 2005;35:1118-20.
6. Danchaivijitr N, Cox T, Saunders DE, et al. *Evolution of cerebral arteriopathies in childhood arterial ischemic stroke.* Ann Neurol. 2006;59:620-6.
7. Bahr M, Postler E, Meyermann R. *Fatal stroke in a patient with carotid and middle cerebral artery dissection associated with cystic medial necrosis.* J Neurol. 1996;243(10):722-3.
8. Szatmary Z, Boukobza M, Vahedi K, et al. *Orgasmic headache and middle cerebral artery dissection.* Journal of Neurology, Neurosurgery and Psychiatry. 2006;77:693-4.
9. Prabhakaran S, Krakauer JW. *Multiple reversible episodes of subcortical ischemia following postcoital middle cerebral artery dissection.* Arch Neurol. 2006;63(6):891-3.
10. Inoue T, Nishimura S, Hayashi N, et al. *Postpartum dissecting aneurysm of the posterior cerebral artery.* J Clin Neurosci. 2007;14(6):576-81.
11. Kurino M, Yoshioka S, Ushio Y. *Spontaneous dissecting aneurysms of anterior and middle cerebral artery associated with brain infarction:a case report and review of the literature.* Surg Neurol. 2002;57(6):428-36.
12. Hosoya T, Adachi M, Yamaguchi K, et al. *Clinical and neuroradiological features of intracranial vertebrobasilar artery dissection.* Stroke. 1999;30:1083-90.
13. Schaefer FK, Schaefer PJ, Altjohann C, et al. *A multicenter, site-independent, blinded study to compare the diagnostic accuracy of contrast-enhanced magnetic resonance angiography using 1.0M gadabutrol (Gadovist) to intraarterial digital subtraction angiography in body arteries.* Eur J Radiol. 2007;61(2):315-23.
14. Lyrer P, Engelter S. *Antithrombotic drugs for cervical artery dissection.* Cochrane Review, Oxford, UK. 2002(1).
15. Beletsky V, Nadareishvili Z, Lynch J, et al. *Cervical artery dissection: Time for a therapeutic trial?* Stroke. 2003;24:2856-60.
16. The CADISS Trial Investigators. *Antiplatelet therapy vs. anticoagulation in cervical artery dissection: rationale and design of the Cervical Artery Dissection in Stroke Study (CADISS).* International Journal of Stroke. 2007;2:292-6.

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