

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 haemorrhage 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-coitus⁹ 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 subdural 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 arteries¹³ 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 diagnostic 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



Gina Kennedy BSc, MBBCh, MRCP, PhD, is a Neurology SPR at Frenchay Hospital, Bristol. Her first degree in the neurosciences and a PhD in human vision founded her interest in neurology. After graduating from Oxford Medical School, she completed her SHO training in Bristol and is currently developing an interest in stroke.

Dr Patrick Ruane, BSc, MBChB, MRCP, is a GP registrar in Bristol having completed SHO training and having attained MRCP at North Bristol NHS trust. A degree in physiology with an emphasis on neurophysiology developed an interest in neurology which he now plans to pursue as a GP with special interest.

Dr Shelley Renowden, BSc, MRCP, FRCR, is a consultant neuroradiologist based at Frenchay Hospital, Bristol and has been in post since January 1996, having trained previously at the Radcliffe Infirmary, Oxford. Her main interest is neurointervention and neurovascular disease. She currently works in a tertiary referral centre for complex neurovascular disorders and one of her main objectives, with her clinical colleagues, is to enhance and expand stroke services/treatment in North Bristol.



David Cottrell, MBChB, BSc, MRCP, PhD, was appointed as a consultant neurologist and senior clinical lecturer at Frenchay Hospital, Bristol in 2005. He specialises in multiple sclerosis and in particular primary progressive multiple sclerosis.

