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lesson of the month (1)

A small left pupil and a headache

Carotid artery dissection is an important cause of stroke in young adults. When focal neurological deficit or classic traumatic history is absent, the diagnosis can be challenging. This lesson reports an interesting case of a patient in whom pupillary dysfunction was the presenting sign of acute dissection of carotid artery.

Lesson

A 24-year-old previously healthy student presented with a two-day history of a small left pupil. He first noticed unequal pupil sizes when looking at the mirror, after being alarmed by a sudden onset but brief-lasting 'funny' sensation affecting his face. He described two further similar episodes, which seem to have involved left-sided facial paraesthesia associated with an element of asymmetry. Of note, the patient had experienced persistent cephalgia, worse behind his left orbit and associated with visual aura. There was no associated neck pain, although on closed questioning he described a transient sensation of 'stiffness'. His medical history was unremarkable and he was not on any regular medications. The patient's family history was notable for 'stroke' at a young age affecting his father and paternal grandfather. He was a non-smoker with minimal alcohol intake and denied any illicit drug use.

An urgent magnetic resonance (MR) angiogram was arranged which showed a decrease in the internal calibre of the left internal carotid artery in its extracranial course, consistent with dissection (Fig 1). The right internal carotid artery and the circle of Willis were unremarkable and there was no evidence of aneurysmal lesions. The patient was subsequently commenced on unfractionated heparin intravenously. Within 72 hours, his symptoms had resolved. A thorough search for clinical features suggesting conditions commonly associated with primary inherent arteriopathy was unrevealing. Having been established on oral anticoagulant therapy, he was discharged. Genetics follow-up was arranged, in view of the strong family history and absence of risk factors.

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Discussion

Internal carotid artery dissections are classified according to the aetiology as either traumatic or spontaneous. Spontaneous dissection may be associated with an inherent arteriopathy caused by genetic factors and connective tissue disorders such as Marfan syndrome, Ehlers-Danlos syndrome type IV, pseudoxanthoma elasticum, osteogenesis imperfecta type I, fibromuscular dysplasia, and autosomal dominant polycystic kidney disease. Additionally, spontaneous dissections are related to intracranial aneurysms, bicuspid aortic valve, coarctation of the aorta, a widened aortic root, arterial redundancy and distensibility. Traumatic dissections are predominantly due to a definite head or neck injury, and most documented cases involve motor vehicle accidents.[1] The extracranial cervical arteries are more vulnerable to traumatic injury because of the tethering at the skull base and their mobility within the neck soft tissues along the cervical spine.[2] Two cases of internal carotid artery dissection in the context of vigorous exercise have recently been reported that occurred during sport game practice using the Wii computer system.[3]

Internal carotid artery dissection typically presents with focal cerebral ischaemic symptoms, unilateral head or neck pain, and partial Horner syndrome. Unilateral headache on the side of the dissection is the most common symptom, occurring in 68–92% of patients.[1] Classically, the headache is most often described as a constant ache in the frontal or periorbital

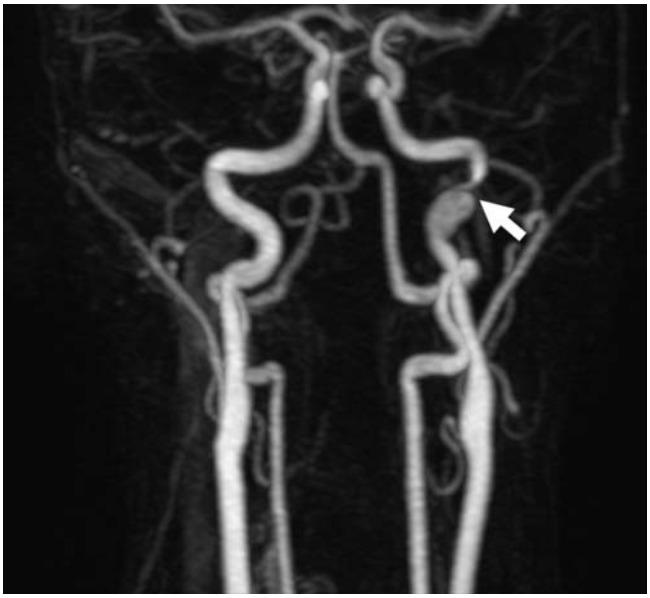

Fig 1. Magnetic resonance angiography of aortic arch and carotids. A dissecting aneurysm arising from the left internal carotid artery is demonstrated measuring 13 mm in length with a narrow neck of 2 mm.

region.[4] Besides head or neck pain, most patients will develop symptoms of contralateral focal cerebral ischaemia, such as aphasia, visual symptoms and hemiparesis.

A partial or incomplete Horner syndrome consisting of miosis and ptosis is present in 36–58% of patients with internal carotid artery dissection.[1,5]

Even though history and physical examinations are indicative of carotid artery dissection, diagnostic imaging studies are essential for definitive diagnosis. Computed tomography (CT) angiography is rapidly emerging as a highly sensitive and specific modality for large and medium vessel pathologies of the head and neck.[6] In a recent study comparing CT and magnetic resonance (MR) angiography, it was demonstrated that CT angiography is the ideal cross-sectional modality to outline the imaging features of cervical dissections.[7]

Almost 85% of extracranial dissections heal spontaneously.[8] The purpose of early diagnosis and timely start of treatment is to prevent cerebral infarction or any development of neurological symptoms. It is estimated that more than 90% of dissection-related infarcts are due to thromboembolic, rather than haemodynamic, causes with transcranial Doppler confirming course of microemboli.[9] Treatment options consist of anticoagulation, antiplatelet therapy, surgery and endovascular stent reconstruction.

Interval treatment with anticoagulation therapy is advocated to prevent thromboembolic complications, despite the lack of randomised prospective studies. Anticoagulation therapy with intravenous heparin followed by oral warfarin intake at a target international normalised ratio of 2.0 to 3.0 is the standard treatment for three to six months. Follow-up MR angiography imaging is carried out at three months and six months with a predicted high rate of dissection healing and recanalisation at three months.[8] If a dissection-related abnormality continues at six months, warfarin treatment is frequently discontinued in preference for antiplatelet therapy.[8] Even though aspirin is most frequently used in this context, at variable doses, no study has directly compared other antiplatelet or anticoagulation agents, such as clopidogrel or low molecular weight heparin.

Early surgical or endovascular treatment is recommended in symptomatic patients refractory to medical treatment to prevent further thromboembolic complications, or in patients with contraindications to anticoagulation. Such contraindications include pseudoaneurysm, enlarging or ruptured dissecting aneurysm, intracranial or subarachnoid haemorrhage.[10] Because of high complication rates of ischaemic injury or cranial nerve deficits, in particular the pharyngeal and superior laryngeal branches of the vagus nerve, endovascular stent reconstruction is the primary interventional option over surgical vessel deconstruction or surgical bypass. It is an effective and rather safe technique, but studies have been restricted to a few case reports and retrospective small case series mainly due to the low incidence of carotid dissections.[11–13]

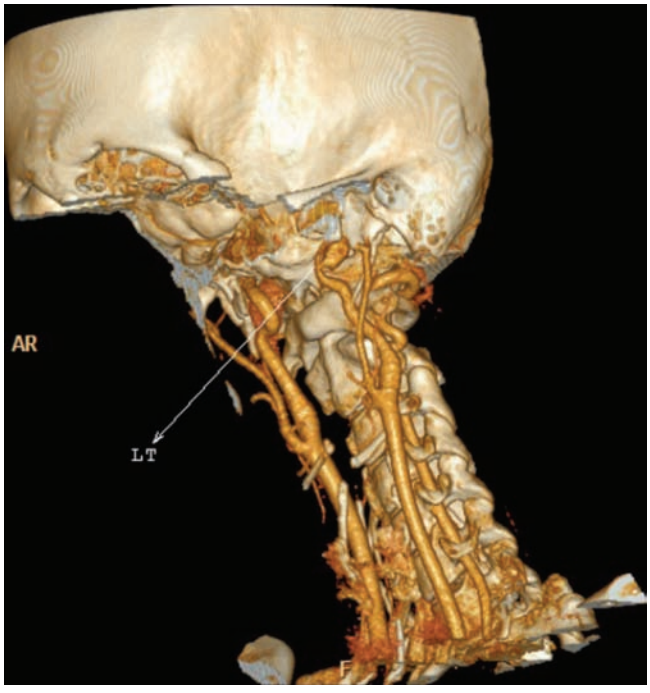

Fig 2. Volume rendered image from follow up CT angiography examination showing pseudoaneurysm of left internal carotid artery. The study was undertaken a month after his initial presentation and shows evolution of the dissection to pseudoaneurysm. Resolution was achieved three months later on anti-coagulation.

Conclusion

Taking an accurate history and conducting a careful physical examination is crucial to diagnosing, sometimes clinically occult, physical signs of carotid dissection. Early diagnosis and initiation of appropriate therapy may prevent or improve neurological deficits and avoid the potentially devastating complications of carotid artery dissection. This case highlights the significance of considering internal carotid artery dissection in patients presenting with restricted clinical manifestations.

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LESSON OF THE MONTH

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lesson of the month (2)

Cushing's syndrome with low levels of serum cortisol: the role of inhaled steroids

With the introduction of new drugs and new devices believed to have less potential for systemic effects, the propensity for potent inhaled glucocorticoids to cause potent hypothalamic–pituitary–adrenal axis suppression is still under recognised.

Lesson

In January 2010, a 22-year-old man presented to Barts and the London School of Medicine for investigation of apparent adrenal insufficiency but with a cushingoid habitus. Around one year earlier, he had complained to his GP of central weight gain and the appearance of purple striae on his upper body; the GP found, to his surprise, that the patient's random serum cortisol was <50 nmol/l on three occasions (normal range 200–600 nmol/l); on a short synacthen test (adrenocorticotropic hormone (ACTH) (1–24) 0.25 mg, intravenous (iv)) the basal cortisol of 28 nmol/l and only rose to 133 nmol/l 30 minutes after stimulation (normal, >550 nmol/l). His 0900 plasma ACTH was 22 ng/l (normal, 10–60 ng/l). Other markers of pituitary function were normal. A pituitary magnetic resonance image was reported as normal. In view of these results, interpreted as adrenal insufficiency, the patient was initiated on hydrocortisone replacement treatment 30 mg daily in divided doses. Over the following 11 months he continued to gain weight centrally and noticed the further development of his striae, and thus sought medical assistance.

On clinical examination the patient was markedly cushingoid, with a body mass index (BMI) of 26.4 kg/m^2, centrally distributed fat and broad purple striae over his shoulders and upper arms. His blood pressure was normal. He denied inadvertently taking oral corticosteroid medication, but on direct questioning admitted to be taking inhaled fluticasone propionate 250 micrograms plus salmeterol 50 micrograms per blister (Seretide250 Accuhaler®), two blisters twice daily, for four years, for asthma. It was hypothesised that he had iatrogenic Cushing's syndrome and adrenocortical suppression due to inhaled glucocorticoids (IGC), and he was advised to stop hydrocortisone and given a Steroid Card, a hydrocortisone emergency pack and a supply of hydrocortisone with education as to corticosteroid replacement during episodes of severe intercurrent illness, trauma or surgery.

Four months after stopping hydrocortisone the patient had noticed a drop in weight to a BMI of 25.4 kg/m^2 and a marked reduction of his purple striae. Twenty-four hours off inhaled glucocorticoids, basal and dynamic tests were performed. All baseline pituitary function tests were normal other than a suppressed serum cortisol of <20 nmol/l.

The patient had a long corticotropin (ACTH-Synacthen) test (1 mg intramuscular with sampling for 24 hours; Table 1). This showed a suppressed 0900 serum cortisol with an impaired response at 30 and 60 mins, confirming adrenocortical insufficiency; however, cortisol levels showed a delayed

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