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Dissection of the internal carotid artery causing transient ischaemic attack and Horner's syndrome

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ABSTRACT Dissections of the internal carotid artery continue to be one of the underrecognised causes of transient ischaemic attack and stroke, especially in the young. The prompt identification of the dissection as a cause of stroke or transient ischaemic attack is essential for the initiation of appropriate treatment. We report a case of spontaneous extracranial carotid dissection presenting as a transient ischaemic attack followed by the development of Horner's syndrome in a 57-year-old male patient. The diagnosis was confirmed by magnetic resonance angiography. Prompt anticoagulation was initiated, and the patient remained stable and well at the end of three months.

KEYWORDS Carotid artery dissection, Horner's syndrome, miosis, transient ischaemic attack

DECLARATION OF INTERESTS No conflict of interests declared.

Cervicocephalic dissections (dissections of carotid and vertebral arteries) were once considered uncommon as a cause of stroke or transient ischaemic attacks (TIA).[1] However, with the advent of non-invasive methods of magnetic resonance angiography (MRA) and computerised tomographic (CT) angiography, carotid and vertebral dissections are being recognised more often. In the following article we present a case of extracranial carotid dissection causing a transient ischaemic attack followed by the development of Horner's syndrome. The case is being reported to highlight the importance of carotid dissection as a cause of stroke and transient ischaemic attacks.

CASE REPORT

A 57-year-old male patient presented to the emergency department of our hospital with a history of weakness of the right side of the body, along with an inability to speak. He did not have any significant medical illness in the past. Clinical examination at the time of presentation revealed motor aphasia and grade 0 power in the right upper and lower limbs. The patient had a gradual improvement in his weakness over the next three hours and by the end of the day the power and speech had returned to normal. He was admitted for further evaluation with a provisional diagnosis of TIA in the left middle cerebral artery territory causing transient weakness and motor aphasia.

The patient underwent further evaluation for his condition including a CT scan of the brain, which was reported as normal. A carotid Doppler did not reveal any evidence of dissection or significant stenosis. An echocardiogram done as part of the work-up did not

FIGURE 1 A picture of the patient on the third day of his admission when he developed ptosis and miosis suggestive of partial Horner's syndrome. The 57-year-old male patient presented with a history of right-sided weakness with motor aphasia and spontaneous improvement.

reveal any cardiac abnormalities. All other routine investigations were normal, and the patient continued to be stable over the next three days. On the third day of his admission, he reported that his left eye had become smaller compared with the right. He also complained of temporal headache on the left side. As shown in Figure 1, clinical examination revealed that he had developed an incomplete Horner's syndrome on the left side with miosis and partial ptosis. There was no evidence of anhydrosis. A detailed neurological examination did not reveal any other focal neurological deficits.

In view of the recent TIA followed by the development of a partial Horner's syndrome and left temporal headache, the possibility of carotid dissection was entertained and an urgent magnetic resonance imaging (MRI) scan of the brain with MRA was arranged. Multiplanar views were obtained, including intra- and extracranial MRA, which confirmed a left internal carotid abnormality just below the base of the skull of approximately 25 mm in length with a stenosis of 80% suggestive of a dissection of the left internal carotid artery (Figure 2). An acute left frontal infarct of 18–19 mm was also identified.
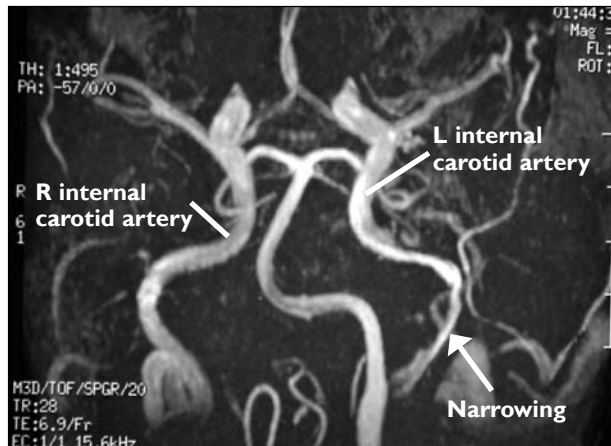
FIGURE 2 Magnetic resonance angiogram showing narrowing and stenosis of the extracranial segment of the left internal carotid artery taken three days after the patient developed neurological signs of a stroke.

Anticoagulation was initiated with a heparin infusion followed by oral warfarin, with a target international normalised ratio of 2–3. The patient remained stable and was discharged on the fifth day with a plan to continue anticoagulation and repeat the MRA or CT angiogram after three months. The left Horner's syndrome had shown partial improvement, and the headache had completely resolved at the time of discharge. Final diagnosis at discharge was spontaneous left internal carotid artery dissection causing partial Horner's syndrome and TIA. At the end of three months the patient continued to remain stable, and a repeat CT angiogram was performed which showed complete recanalisation of the dissection (Figure 3). After a discussion with the neurologist it was decided to continue the anticoagulation for three more months and then to maintain the patient on antiplatelet agents.

DISCUSSION

Once considered uncommon, cervicocephalic arterial dissections are now being recognised more frequently as one of the causes of stroke and TIA, especially in the young. Dissection is a cause of stroke in 0.4–4% of the general population but 5–20% in younger age groups.[2,3] The first report of spontaneous internal carotid artery dissection was made by Jentzer in 1954.[4] Dissection can occur either spontaneously or as result of trauma and can affect either the internal carotid artery or vertebral artery. Dissection of the internal carotid artery can be intra- or extracranial, with the latter being more frequent.

The exact cause for spontaneous cervical arterial dissections is still not clear. Mechanical forces such as blunt injury and stretching, as well as underlying arteriopathies, could account for the majority of the cases. Young women, especially in their third and fourth decades, appear to be particularly susceptible to this condition.[5] In a study by Brandt et al. ultrastructural abnormalities
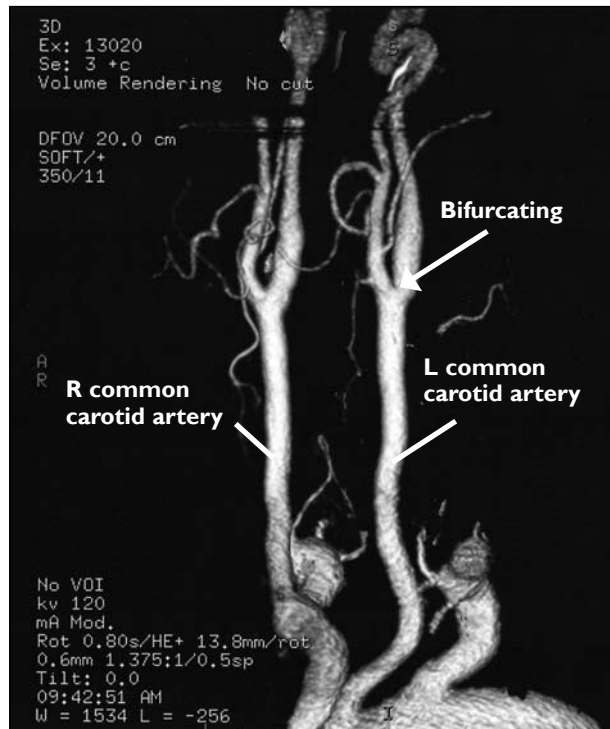

FIGURE 3 This volume-rendered CT angiogram, taken three months after the initial event, shows the recanalisation of the dissection.

were shown in 68% of 25 cases of proven spontaneous carotid dissections, revealing a correlation of dissection with connective tissue abnormalities.[6] Reduction in type 3 collagen has been found to be a contributing factor for dissection in some cases.[7] Fibromuscular dysplasia and Ehlers-Danlos syndrome type 4 have been implicated as important causes for cervical arterial dissections, especially in the young.[8] Other arteriopathies and connective tissue diseases such as Behçet's syndrome, Marfan's syndrome, osteogenesis imperfecta and other causes of vasculitis could account for a significant number of spontaneous carotid dissections.[8,9]

Traumatic arterial dissections can be a result of severe whiplash injuries or minor direct trauma to the head and neck. Dissections have been reported after violent bouts of coughing, scuba diving, chiropractic manipulation and bronchoscopy, during pregnancy and post partum and even after a visit to the hairdresser. Rubinstein and colleagues conducted a systematic review of the risk factors for carotid artery dissection and found strong associations for the following: aortic root diameter >34 mm, migraine, relative diameter change during the cardiac cycle of the common carotid artery and trivial trauma in the form of manipulative therapy of the neck.[10]

Multiple cervical arterial dissections are quite rare. In a study by Arnold et al. 15 out of 740 patients with spontaneous cervical arterial dissections showed involvement of multiple arteries. This was more common in females, and the majority of these multiple arterial

dissections were preceded by minor trauma or infections. However, clinical symptoms in all these patients were confined to a single arterial territory.[11]

The clinical picture associated with carotid dissections is extremely variable and can make the diagnosis difficult, even for experienced physicians. Patients may present with trivial neurological signs or TIAs and occasionally with fully evolved stroke. The majority of the patients have had warning attacks of unilateral cranial or facial pain, followed within minutes or days by signs of ischaemia in the internal carotid territory.[12–14] The pain is non-throbbing and centred most often in and around the eye; less often it is in the temporal or frontal regions, angle of the jaw or high neck. It can be often mistaken for migraine, causing delay in diagnosis.[13] Hypoageusia or decreased taste sensation can also be a presenting symptom of carotid dissection as can transient monocular blindness (amaurosis fugax), pulsatile tinnitus and transient visual field disturbances.[14]

The ischaemic manifestations consist of transient attacks in the territory of the internal carotid, followed frequently by signs of hemispherical stroke, which may evolve over a period of a few minutes to days. Cranial nerve palsies can also be an uncommon manifestation of carotid dissection. In a review of 190 patients with carotid dissection by Mokri and colleagues, 5.2% had lower cranial nerve palsies, 3.6% had fifth cranial nerve involvement and 2.6% had oculomotor palsies.[14] A partial Horner's syndrome occurs in less than 50% of cases of carotid dissection. The term partial is used since anhydrosis is absent as the sympathetic fibres innervating the facial sweat glands are located on the external carotid artery rather than the internal carotid artery. Rarely a mydriatic pupil is the first sign of carotid dissection.[15,16] Occasionally a cervical bruit may be heard at the site of dissection as a result of the stenosis.

Conventional angiography has been considered as the gold standard for the diagnosis of carotid dissection. However, the widespread use of MRI and MRA may soon supersede angiography as the investigation of choice for this condition. Internal carotid artery dissection can be identified by irregular vessel margins, filling defects, vascular occlusion, calibre changes of the vessel, the string sign, double lumen or internal flaps. The improved resolution, speed, non-invasiveness, good negative predictive value and the fact that it does not require the use of iodinated contrast dye makes MRA an excellent screening tool for carotid dissection. Stringaris et al. reported 12 cases of carotid dissection in which MRI in combination with MRA was superior to conventional angiography for the diagnosis.[17] However, it should be noted that at present MRA has a lower rate of detection for vertebral artery dissections and intracranial carotid artery dissections.

Ultrasound techniques are also a valuable tool for the diagnosis of carotid dissections. In a series by Sturzenegger and colleagues, who analysed the value of different sonographic techniques for carotid dissection in 43 patients, it was found that the combined sensitivity was 95%, with 93% for extracranial Doppler, 86% for transcranial Doppler and 79% for duplex ultrasonography.[18]

Helical CT angiogram is another screening modality of promise and may be the only option in many centres with limited facilities. The suspicion of carotid dissection may be raised from indirect findings of soft tissue swelling, haematoma around the internal carotid artery and infiltration of the perivascular fat planes. The direct evidence comes from the decrease in the calibre of the vessel. Computerised tomographic angiograms have a comparable efficacy to MRA for the detection of extra-cranial carotid artery dissections. A minor drawback of this modality is the necessity for contrast enhancement. In a study by Vertinsky et al. on 18 patients with 25 dissected vessels (15 internal carotid and 10 vertebral arteries) CT/CT angiography identified more intimal flaps, pseudo-aneurysms and high-grade stenoses than MRI/MRA.[19] Prior discussion with the radiologist regarding the clinical suspicion can increase the chances of picking up the dissection.

The exact mechanism for cerebral ischaemia in dissection remains unclear. The possibilities suggested include an embolic injury and/or a haemodynamic injury as the cause of acute ischaemic symptoms. An analysis of stroke patterns by Lucas et al. in carotid dissection was suggestive of an embolic origin rather than an occlusive haemodynamic cause.[20] This justifies the use of anti-coagulation in the treatment of dissection in order to prevent embolisation. However, at present there is no consensus on treatment. A systemic review by Lyrer and Engelter found no randomised trials comparing anticoagulants with antiplatelet agents for carotid arterial dissections and concluded that there was no evidence to support the use of anticoagulation in this clinical situation.[21] A systematic review and meta-analysis of the treatment of cervical arterial dissections by Menon and colleagues found no data to support the therapeutic superiority of anticoagulants over antiplatelet agents. They also commented that thrombolysis in carotid dissection appears safe, but more data on efficacy are required.[22]

Risk versus benefit of antithrombotic therapy has to be weighed in individual cases, especially those due to high impact trauma. Most reports in spontaneous carotid dissection suggest immediate heparinisation after diagnosis, followed by oral anticoagulation.[23] Most neurologists take the approach that anticoagulation, if used, should be discontinued after several months or a year, when angiography or MRA shows the lumen of the carotid artery to be patent to at least 50% of the normal diameter and smooth-walled. The ongoing Carotid Artery

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Dissection in Stroke Study (CADISS) is a prospective multi-centre randomised controlled trial in acute carotid and vertebral arterial dissections looking at the benefits of anticoagulation over standard antiplatelet therapy, and should provide guidance on the management of these patients.[24] The surgical therapy of dissections is considered when neurological symptoms progress despite maximal medical therapy or when an accompanying pseudo-aneurysm is present. Surgical options include ligation, resection with revascularisation and bypass. Endovascular intervention is a relatively new treatment option in patients in whom the location of dissection precludes safe vascular bypass.[25]

The prognosis of carotid dissection is highly variable, with excellent outcomes in cases with limited neurological deficits. In a study by Mokri and colleagues, complete resolution of the dissection occurred in 85% of cases, the majority without a major stroke.[26] The other 15% of cases had poor outcomes, with death or major disability as a result of dissection. The outcome is more favourable with medical treatment. The risk of stroke after a dissection is highest in the first month and the risk of recurrence of dissection is about 1% per year after the first year.

CONCLUSION

Carotid dissections should be considered as an aetiological factor in cerebrovascular diseases, unexplained headaches, cranial nerve palsies and visual symptoms, especially in the younger population. A high index of suspicion can enhance the chances of diagnosing this comparatively rare cause of these conditions. Prompt diagnosis and rapid institution of treatment can lead to excellent outcomes in the majority of cases and prevent devastating cerebral ischaemic damage.

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