Extracranial cerebrovascular disease – a management dilemma

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A 71-year-old man presented to the vascular clinic with a 5-year history of amaurosis fugax in his right eye. He described this as a blanket descending over his visual field, which would come on slowly, lasting approximately 5 minutes before rapidly resolving, and was associated with a feeling of dizziness. Initially, attacks had occurred once every 2 months but recently the frequency had increased to twice a week. The patient was known to suffer from hypertension and hypercholesterolaemia, both of which were controlled medically. Ten years earlier, the patient had undergone an aorto-iliac graft for peripheral vascular disease.

Examination revealed a regular pulse of 76 beats/min and a blood pressure of 136/84 mmHg; a late systolic murmur was detected in the mitral region. The right carotid artery pulse was absent, with a low pitched bruit. A left carotid thrill was detected with a high-pitched murmur. An ophthalmic examination confirmed embolic events in the right eye.

Duplex scan showed a diseased innominate artery with total occlusion of the right common carotid, 75% stenosis of the left internal carotid and occlusion of the left subclavian artery with reversal of flow through the vertebral artery. An aortic arch angiogram (figures 1 and 2) was performed.

Figure 1 Aortic arch angiogram

Figure 2 Aortic arch angiogram

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Questions

- 1 What is the cause of the visual symptoms?
- 2 What does the arteriogram show?
- 3 What is subclavian steal and how would you treat it?
- 4 How can the patient's symptoms be explained?

Answers

QUESTION 1

The patient is suffering from amaurosis fugax or temporary blindness secondary to embolisation within the retinal artery. Of significance in this patient was the fact that he was exhibiting evidence of hypoperfusion (dizziness) which is only seen in 5% of cases of carotid disease.

QUESTION 2

Currently, duplex scan remains the first choice of investigation for anatomical and functional assessment of the arterial tree, as it is non-invasive, reliable and can be repeated, thus it has almost replaced arteriogram in most situations.^{1,2} However, arteriogram is often obtained to confirm the duplex scan findings in complex situations, such as in this case. The arteriogram (figures 1 and 2) in this case confirmed the findings of the duplex scan accurately and demonstrated that the right common carotid is occluded and there is no filling of the right internal or external carotid artery. There is 75% stenosis of the left internal carotid artery and occlusion of the left subclavian artery at its origin. There is distal filling of the left subclavian by reverse flow from the left vertebral artery.

QUESTION 3

Subclavian steal (figure 2) is a phenomenon where the distal blood supply in the subclavian (usually on the left) is derived from a reversal of blood flow from the vertebral to the subclavian artery distal to the occlusion. This occurs because the proximal segment of the subclavian artery has been occluded and therefore the arterial pressure gradient favours blood flow in the reverse direction. It is usually only treated if the patient is symptomatic (clumsiness, dizziness or drop attacks), and is usually brought on by exertion. The main form of treatment is arterial reconstruction of the occluded segment of the subclavian artery or angioplasty or stenting of the occluded segment.

QUESTION 4

The patient's symptoms are the result of embolisation from one of his carotid arteries. The most likely source of the emboli was felt to

be the left internal carotid with emboli traversing the circle of Willis to impact in the right retinal artery. The other possibility was that emboli were being released by the occluded right internal carotid artery, a phenomena known as carotid stump syndrome. Thus, the surgical options available were to either tie off the occluded stump on the right, perform a carotid endarterectomy on the left, or do both.^{3,4}

A carotid endarterectomy was performed on the left, with no complications in the postoperative period. Out-patient follow-up has revealed that the patient's symptoms of embolisation and hypoperfusion have resolved since discharge.

Discussion

This case illustrates the extensive and complex nature of atheromatous disease and that management of these patients often can be difficult. In this category of patient the risk of not operating was that these transient episodes of blindness could lead to an event causing permanent neurological damage. It has been estimated that 20% of patients with amaurosis fugax will suffer from a stroke if untreated.⁵ This has to be weighed against the risk of mortality and stroke from performing a carotid endarterectomy. Mortality from this procedure has been estimated at 1.3% after 30 days for those with asymptomatic disease and 1.8% for those with symptomatic disease with a risk of fatal stroke of 0.47% and 0.91%, respectively.⁶

Attempts have been made to evaluate the effectiveness of anticoagulating this cohort of patients. The general consensus appears that, although anticoagulation reduces the risk of a cerebral event, it does not do this to a significant enough degree.

Final diagnosis

Amaurosis fugax secondary to embolisation within the retinal artery.

Keywords: carotid artery; subclavian steal; amaurosis fugax

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