Reminder of important clinical lesson

Sudden loss of vision in a patient with significant ipsilateral internal carotid disease

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A 74-year-old man developed sudden onset of painless persistent partial loss of vision in the right eye 3 days after right total knee replacement. The initial investigation was for amaurosis fugax with a normal CT scan of the brain but the carotid Doppler confirmed >90% stenosis of the right internal carotid artery. Urgent ophthalmological review confirmed anterior ischaemic optic neuropathy. Subsequently, he lost his vision completely in the right eye. He had a previous high risk hemispheric transient ischaemic attack with carotid Doppler reported as normal.

BACKGROUND

Visual manifestations are commonly seen in transient ischaemic attack (TIA)/stroke clinics. Many of these clinics are run by physicians trained in Geriatric medicine without specific training in ophthalmology or neurology. This case also underlines the need for considering a second carotid imaging modality in a patient with high risk hemispheric TIA without atrial fibrillation or features of systemic embolisation.

CASE PRESENTATION

A 74-year-old former lorry driver was admitted to Basildon Hospital for an elective right total knee replacement for severe osteoarthritis. The medical history included hypertension, type 2 diabetes mellitus and a quadruple coronary bypass graft operation. Six months earlier he had two TIA with left arm weakness resolving within few hours. An urgent CT scan of the head showed no focal lesion and carotid Doppler scan on the same day was reported as normal. During these TIAs, he was in sinus rhythm and a transthoracic echocardiogram showed dilated left ventricle but with preserved systolic function. He was treated medically with metformin, aspirin, lisinopril and modified release dipyridamole, but he was intolerant to statins. He underwent a right knee replacement on 28th February 2011 which was uneventful until the third postoperative day when he experienced a sudden blurring of vision in the right eye which was rapidly followed by loss of vision in the lower half of the right eye. At that time he had no pain in the eye, headache, double vision, slurring of speech or motor weakness. The patient was urgently referred to the stroke team. Review by the stroke team confirmed the sudden onset of blurring of vision complicated by the loss of vision affecting lower and outer aspects of right eye without, facial droop, motor or sensory deficit. The stroke team's initial assessment included F.A.S.T¹ which was negative and the patient scored only three points on the ABCD2 score (one point for age above 60 and another two points for hypertension and diabetes,

respectively) and thereby classifying him as low risk.^{2 3} A provisional diagnosis of amaurosis fugax or ophthalmic TIA complicated by partial visual loss was made for which an urgent CT scan of the head and a carotid Doppler were made. Clinically he was in sinus rhythm confirmed with an ECG. The CT scan of the head showed only mild age related cerebral involution. The carotid Doppler ultrasound scan revealed extensive, mainly calcified plaque in the right internal carotid artery resulting in >90% stenosis (figure 1A,B). In the left proximal common carotid artery, a 3 mm soft, thick plaque was noted in keeping with 60% stenosis which was confirmed with CT angiogram of the carotids and vertebral arteries. He was urgently referred to the ophthalmology clinic. Review on the 4th March in the ophthalmology clinic confirmed right anterior ischaemic optic neuropathy. Unfortunately 3 days after the initial visual symptoms, he reported sudden onset of painless, complete loss of vision in the right eye. On examination, the patient was unable to detect hand movements with his right eye and had a right relative afferent pupillary defect without other neurological deficits. Examination of the optic fundi remained unchanged. He underwent an urgent right carotid endarterectomy as an inpatient on 14th March. The operation was uneventful. The left internal carotid disease (60%) is currently asymptomatic and it is planned that we review his left carotid artery disease within the next few weeks. He was discharged home next day on dual antiplatelet therapy (aspirin, dipyridamole retard), lisinopril, metformin and life style advice. He was intolerant to statins. He is planned for further postoperative assessment and follow-up of the left common carotid disease by the vascular surgeons. Unfortunately, the right eye remains blind permanently. A review of the fundi on the 11th October 2011 confirmed atrophy of the right optic nerve as pale distinct optic disc with background hypertensive changes; silver wiring and AV nipping where as the right optic disc was normal in May 2008 when it was screened as a part of the diabetic screening program (figure 2).

Figure 1 (A) Patient's right internal carotid artery showing a maximum peak systolic velocity (PSV) reaching 471 cm/s indicating over 90% stenosis (normal <125 cm/s). (B) Right internal carotid artery in a normal person showing a maximum peak systolic velocity (PSV) under 100 cm/s.

DISCUSSION

The case history of this gentleman raises many issues in the management of high risk TIAs, ophthalmic TIAs and the limitation of some investigations with consequent missed opportunities. First, the initial presentation with a high risk right hemispheric TIA was assessed with a carotid Doppler scan reported as normal while he was in sinus rhythm without evidence of systemic embolisation. A small embolic source off a fissured athermanous plaque from the aortic arch or intracranial right internal carotid

artery has been accepted as the most likely explanation for which medical treatment was continued. However, a significant disease of the right internal carotid artery 6 months later indicates that the initial carotid Doppler scan was a false negative result. This is a source of concern as our current local guidelines would usually accept a secondary carotid imaging if the carotid Doppler scan is significantly abnormal as this is not a part of a national guideline. The experience with this gentleman who had an unexplained high risk TIA with a normal carotid Doppler raises

Figure 2 (A) shows normal right optic disc and it was recorded as part of diabetic screening program. (B) shows pale right optic disc with distinct margins due to optic atrophy complicating AION months after the acute illness. There is background hypertensive changes of silver wiring and AV nipping.

Box 1 Amaurosis fugax: the main features

- ▶ Amaurosis fugax is a sudden painless monocular transient blindness (seconds to min) and often described as feeling like the pulling of a curtain or a shade over one eye.
- ▶ Amaurosis fugax is considered as an ocular transient ischemic attack.
- ▶ The commonest causes are embolisation from atherosclerotic carotid arteries and/or aorta, cardiac emboli and migraine due to spasms and narrowing in arteries leading to the eye.
- ▶ Should be regarded as a harbinger of stroke, and monocular blindness.

the need for either a repeat the carotid Doppler or arrange another carotid imaging modality such as MR angiography or CT angiography to avoid a missed opportunity of surgical treatment if needed.

Second, the sudden partial loss of vision in the right eye was assessed by the stroke team with a provisional diagnosis of amaurosis fugax. The latter is a sudden painless transient blindness in one eye that goes away within seconds or min. It is often described as feeling like the pulling of a curtain or a shade over one eye^{4 5} (box 1). However, this patient's symptoms were originally partial loss of the lower half of vision of the right eye which was persistent for three days after which it deteriorated to total loss of vision. The accurate clinical diagnosis of anterior ischaemic optic neuropathy (AION) was made by the ophthalmologist.

AION is characterised by acute loss of vision, optic oedema, an afferent pupillary defect and monocular visual field abnormalities due to an infarction of the optic nerve head. It is caused by compromised blood flow in one or more of the posterior ciliary arteries⁶ which later on will be seen as optic atrophy (figure 2). The main reasons for missing the diagnosis by the stroke team were the combination of inadequate awareness of the duration of the visual loss and deficient examination of the pupillary reflexes and optic fundi.

The blood supply of the optic nerve could be compromised at two levels namely; the anterior part (optic nerve head) supplied primarily by the posterior ciliary artery

Box 2 Causes and patterns of monocular blindness

Transient painless visual loss

- ▶ Amaurosis fugax
- ▶ Acute ischaemic optic neuropathy (AION)
- ▶ Papilloedema which typically causes greyouts rather than blackouts and are often brought on by change in posture
- ▶ Retinal migraine which might occur without headache
- ▶ Acute angle closure glaucoma may be painless*

Acute sudden visual loss with fundoscopic abnormalities

- ▶ Central retinal/branch artery occlusion (CRAO)
- ▶ Central retinal/branch venous occlusion (CRVO)
- ▶ AION
- ▶ Retinal detachment
- ▶ Vitreous hemorrhage

Subacute visual loss**

- ▶ Papillitis (exclude multiple sclerosis)
- ▶ Papilloedema
- ▶ Diabetic papillopathy
- ▶ Psychogenic

*Abe A et al Headache. 2007;47:1098–9.

**Visual loss develops over days to weeks but in some the loss can progress rapidly over several hours.

which if compromised will lead to AION. The posterior part of the optic nerve is supplied by multiple sources other than posterior ciliary artery circulation which if compromised results in posterior ischemic optic neuropathy.^{7 8} The anterior ischemic optic neuropathy is further subdivided into Arteritic AION (A-AION) due to giant cell arteritis, and the non-arteritic AION (NA-AION) which is an ischaemic infarction of the optic nerve not due to giant cell arteritis. The commonest type of AION is the ischaemic type that is, NA-AION and they are commonly found in middle aged and older patients. The usual predisposing factors are systemic arterial hypertension, nocturnal arterial hypotension, diabetes mellitus, ischemic heart disease, hyperlipidemia and atherosclerotic disease.^{7 8} Other associated systemic diseases have also been reported, including sleep apnoea, arterial hypotension due to a

variety of causes, malignant arterial hypertension and migraine.

The aetiology and the pathogenesis of non-arteritic AION are well described by Hayreh^{7,8} and are mainly due to either transient non-perfusion or embolic lesions of the arteries/arterioles feeding the optic nerve head. Hypoperfusion of the optic nerve head occurs most likely due to a transient fall of blood pressure such as during sleep or a nap during the day, or shock leading to fall in blood pressure below the critical auto-regulatory range of the optic nerve capillaries in susceptible persons. Ischaemic damage to the optic nerve head due to hypoperfusion is more common than damage caused by embolisation. There is evidence from correlative studies between 24 hourly ambulatory blood pressure monitoring and intraocular pressure measurements that nocturnal hypotension, by reducing the optic nerve head blood flow below a crucial level during sleep in a vulnerable optic nerve head may play an important role in the pathogenesis of AION and glaucomatous optic neuropathy which may be the final insult in a multifactorial situation.⁹ Furthermore, patients with 'idiopathic' AION and patients with systemic hypertension who developed non-arteritic AION had a statistically significant increased risk of experiencing cerebrovascular events and myocardial infarctions compared with appropriately matched control groups.¹⁰ In addition, patients with NA-AION have a cumulative probability of the fellow eye developing NA-AION in up to 25% of the patients within 3 years¹¹ while recurrence in the same eye is reported in up to 6% at 2 years.¹²

An awareness of the patterns of acute and subacute loss of vision in one eye (monocular visual loss) remains an important aspect of training of junior staff in the TIA/stroke clinics. This is because transient or permanent monocular visual loss as a manifestation of neurovascular disease is becoming an increasing challenge for many of the stroke teams even though they are supported by emergency ophthalmology services. An element of training program in this area for future trainees will be much needed. This is because of the complexity of the ischaemic presentations of the eye and the differential diagnosis. The complexity is illustrated by the various ways the ischaemia of the eye may lead to visual disturbance or loss. Hedges¹² suggested the following clinical categories based on the duration of the ischaemic visual impairment:

- ▶ Transient visual obscuration—Episodes lasting seconds that are associated with papilloedema and increased intracranial pressure.
- ▶ Amaurosis fugax—Brief, fleeting attack of monocular partial or total blindness that lasts seconds to min.
- ▶ Transient monocular visual loss or transient monocular blindness – A more persistent vision loss that lasts min or longer.
- ▶ Transient bilateral visual loss—Episodes affecting one or both eyes or both cerebral hemispheres and causing visual loss.
- ▶ Ocular infarction—Persistent ischemic damage to the eye, resulting in permanent vision loss.

However for practical reasons, Lueck¹³ suggested four groups of presentations with some expected overlap depending on the time for the loss of vision to develop as follows:

1. Transient: patients have returned to normal by the time they are seen

2. Acute: coming on instantaneously, or over seconds to min
3. Subacute: coming on over days to weeks
4. Chronic: coming on over months to years.

The causes of a painful loss of vision with a red eye are usually an ophthalmological condition such as acute close angle glaucoma, scleritis, anterior uveitis (iritis), keratitis, corneal abrasion, or ulcer and they are all medical emergencies.¹³ Optic neuritis is another cause of painful monocular visual loss but without a red eye.¹⁴ Occasionally, acute close angle glaucoma might present without pain.¹⁵

Finally this patient's clinical course shows many lessons including a missed opportunity to diagnose a significant right internal carotid disease presenting with a high risk hemispheric TIA in the absence of cardioembolic disease and a negative carotid Doppler. In these circumstances, another modality of carotid imaging needs to be arranged. Furthermore, a sudden loss of vision in one eye due to amaurosis fugax is transient. Whereas persistent partial or complete monocular visual loss has a wide differential diagnosis (box 2) and urgent ophthalmic opinion should be available to exclude AION.

In all the cases of monocular visual loss, pupillary reflexes, visual fields and funduscopy should be integral parts of the examination to exclude AION, central/branch retinal artery or vein occlusion, vitreous haemorrhage or retinal detachment. An ischaemic optic neuropathy in patients with previous TIA or known carotid disease is more likely cause of the monocular vision loss when the symptoms persist while the symptoms of amaurosis fugax are usually transient

Learning points

- ▶ A negative carotid Doppler in a patient with high risk TIA without evidence of cardiac source of emboli should raise the suspicion of a false negative carotid Doppler scan.
- ▶ If false negative carotid Doppler scan is suspected, either a repeat scan or another imaging modality of carotid imaging is necessary.
- ▶ A sudden monocular visual loss referred to the TIA clinics or stroke teams is not always due to amaurosis fugax.
- ▶ In all the cases of monocular visual loss, pupillary reflexes, visual fields and funduscopy should be integral parts of the examination.

Competing interests None.

Patient consent Obtained.

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