

# Amaurosis fugax: a warning symptom not to miss

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**Amaurosis fugax is the sudden, severe, temporary loss of vision in one eye, with sight returning within seconds or minutes. It often represents an embolic transient ischaemic attack of the retinal artery. This warning symptom of impending major cerebral stroke or permanent blindness must always be urgently investigated.**

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## CASE PRESENTATIONS

### Case 1. Amaurosis fugax: a warning symptom of impending stroke

A 23-year-old woman presented to her GP complaining of a one-minute episode of sudden-onset complete blindness of her left eye that morning, which had rapidly spontaneously resolved. Her vision was now back to normal.

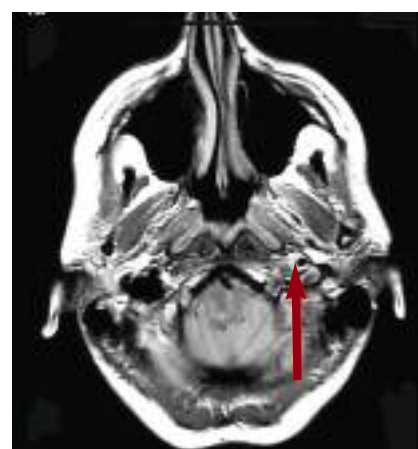
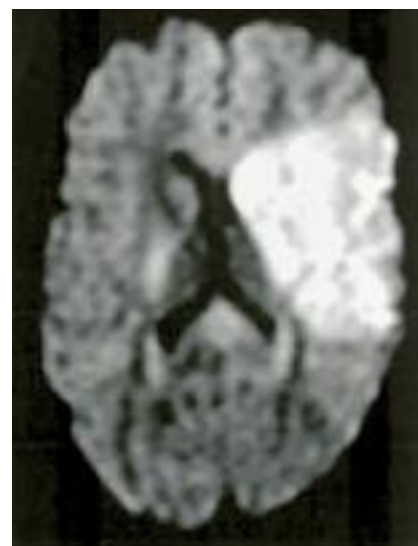
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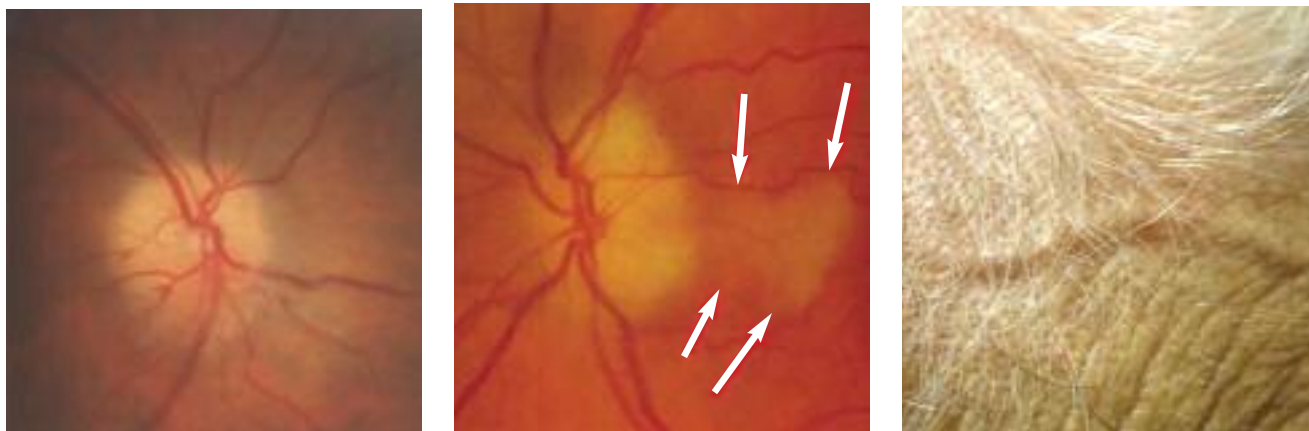
The patient had been in a car accident four days previously and had no injuries other than a sore neck. This had been diagnosed by an emergency department doctor as 'whiplash' and treated with simple analgesics.

On examination her visual acuity was 6/6 in each eye, pupils were equal and reactive to light, visual fields full to confrontation and ophthalmoscopy was normal (Figure 1a). The GP reassured the patient that he could not find anything wrong with her eyes, but to see an optometrist if the blurred vision recurred.

The following day the patient suffered a major left middle cerebral artery stroke (Figure 1b). Investigations including MRI



Figures 1a to c. Case 1. a (top). Ophthalmoscopy was normal. b (middle). Left middle cerebral artery infarct on MRI (diffusion sequence). c (bottom). Left internal carotid artery dissection (arrow) on MRI of the neck. The dissection is seen as a bright ring of blood in the wall of the artery.



Figures 2a to c. Case 2. a (left) and b (middle). Optic disc appearance at presentation to the emergency department, demonstrating bilateral optic nerve head infarction and cilioretinal artery infarction (arrows). c (right). Temporal artery biopsy revealed giant cell arteritis.

and magnetic resonance angiography (MRA) of the neck identified the cause to be a left internal carotid artery (ICA) dissection (Figure 1c), secondary to her recent neck trauma. A small embolus from the ICA dissection had temporarily blocked the left retinal artery before dissolving, causing the amaurosis fugax. The next day a larger embolus from the ICA had lodged in the middle cerebral artery, causing the stroke.

The stroke caused a dense right hemiplegia, which left the patient unable to work or care for her young children. If the episode of amaurosis fugax had been urgently and appropriately investigated, the dissection identified and the patient anticoagulated, the stroke may have been prevented.

### Case 2. Amaurosis fugax: a warning symptom of impending blindness

A 69-year-old man presented to his GP complaining of a two-minute episode of loss of vision in the top half of his right visual field earlier that day. He described it as 'like someone has pulled a black curtain down over the top part of my vision'. The bottom half of the eye's vision remained normal. The patient's sight spontaneously returned to normal and had remained normal since.

There were no other neurological symptoms during the episode. The patient also complained of intermittent headaches and feeling tired. Medical history included treated hypertension and gout.

On examination his visual acuity was 6/6 in each eye, pupils were equal and reactive to light, visual fields full to confrontation and ophthalmoscopy was normal. The GP sent the patient to the local optometrist who reported a normal eye examination but suggested that the GP send the patient for a carotid ultrasound to exclude ICA stenosis. This was booked for three days later.

The next day the patient woke blind in both eyes. On admission to hospital his visual acuity was 'no perception of light' in the right eye and 'hand movements' only in the left eye. Both optic discs were pale and swollen (Figures 2a and b). Urgent blood tests revealed an erythrocyte sedimentation rate (ESR) of 182 mm/h (normal <10 mm/h) and C-reactive protein (CRP) level of 85 mg/L (normal <6 mg/L). A temporal artery biopsy showed giant cell arteritis (GCA; also known as temporal arteritis; Figure 2c).

Despite immediate high-dose intravenous methylprednisolone treatment, the patient remained bilaterally blind. If the episode of amaurosis fugax had been urgently and appropriately investi-

gated, GCA suspected (based on the very high ESR and CRP level, headaches and fatigue) and corticosteroid treatment initiated, the patient's sight may have been saved.

### DISCUSSION

'Eye' problems often take a very low priority in general practice, 'temporary' eye problems that have already resolved even more so. However, in both of the patients described, their trivial 'eye' complaints actually presented an opportunity for urgent intervention that could have prevented severe, permanent disability.

Just as a cerebral transient ischaemic attack (TIA) represents a chance for GPs to prevent a major stroke, amaurosis fugax ('a TIA of the eye') may also alert GPs that emboli are moving through the retinal artery of affected patients.

The challenge is then to identify the source of the emboli and institute urgent management. Successful intervention may prevent blindness (from a central retinal artery occlusion or arteritic ischaemic optic neuropathy), permanent neurological disability or death (from a cerebral stroke).

### What are the symptoms and signs of amaurosis fugax?

The cardinal symptom of amaurosis fugax is sudden, severe, painless visual

**COMMON CAUSES OF AMAUROSIS FUGAX**

- Internal carotid artery stenosis from atheroma
- Internal carotid artery dissection
- Giant cell arteritis (temporal arteritis)
- Other vasculitis
- Cardiac embolic source (e.g. bacterial endocarditis)
- Retinal vasospasm

loss in one eye only. The vision loss may be all over the visual field (with transient complete 'blacking out' or 'greying out' of the vision), or only in the top or bottom half of the vision. Some patients describe the episode as 'like someone pulling a black curtain down in front of my vision'. After a variable period of seconds to minutes, the vision clears and returns to normal. There are usually no other neurological symptoms at the time, and the patient feels otherwise well during the episode.

Usually an eye examination after an episode of amaurosis fugax will be completely normal. Dilated examination of the retinal blood vessels may occasionally show a cholesterol or platelet embolus in one of the branch retinal arteries, but this is difficult to see with a direct ophthalmoscope.

**What diseases can cause amaurosis fugax?**

Emboli to the retinal artery can arise anywhere between the heart and the eye. Common causes of amaurosis fugax are summarised in the box on this page and described further below.

*Internal carotid artery stenosis from atheroma*

ICA stenosis from atheroma is the most common cause of amaurosis fugax and is normally seen in older patients or those

with significant vasculopathic risk factors. ICA stenosis is readily identified on duplex Doppler ultrasound of the carotid arteries. Treatment of a high-grade symptomatic ICA stenosis often involves carotid endarterectomy by a vascular surgeon whereas low-grade stenosis may be managed with antiplatelet agents.

*Internal carotid artery dissection*

ICA dissection is a less common but serious cause of amaurosis fugax in patients of any age, including children or teenagers who have experienced neck trauma. 'Spontaneous' dissections without trauma are also possible. The ICA wall splits and blood 'dissects' up the wall, causing narrowing and roughening of the lumen, which can cause emboli and/or obstruction of blood flow. Patients often, but not always, complain of neck pain on the side of the dissection. Horner's syndrome, a drooping upper eyelid (ptosis) and a smaller pupil (miosis) may or may not be present on the side of the dissection, due to damage to the cervical sympathetic chain. ICA dissection may be missed on ultrasound of the carotid artery and is better detected by MRI plus MRA of the neck.

*Giant cell arteritis (temporal arteritis)*

GCA is a potentially blinding or fatal vasculitis, which only affects patients over the age of 50 years. In patients with GCA, amaurosis fugax often presages permanent blindness from anterior ischaemic optic neuropathy or central retinal artery occlusion. Some patients with GCA also have systemic symptoms including headache, scalp tenderness, jaw muscle pain on chewing, myalgias and fatigue. Any patient with a history of polymyalgia rheumatica (PMR) who develops amaurosis fugax should be strongly suspected of having developed GCA because PMR and GCA are closely linked; up to 15% of patients with PMR will develop GCA. In some, but not all, patients with GCA the temporal

**KEY POINTS FOR GPs: MANAGEMENT OF AMAUROSIS FUGAX**

The following must all be performed urgently (ideally on the same day as presentation).

- Physical examination including blood pressure, temperature, and carotid and cardiac auscultation.
- Urgent examination by an ophthalmologist.
- Blood test (full blood count, ESR, CRP level, electrolytes and liver function tests) to look for giant cell arteritis, other vasculitis, bacterial endocarditis and polycythaemia.
- ICA ultrasound to look for ICA stenosis.
- If ICA ultrasound is normal then echocardiography should be performed, looking for a cardiac embolic source.
- If the patient is young, has neck pain or has Horner's syndrome, MRI plus magnetic resonance angiography of the neck should be carried out, looking for ICA dissection.

ABBREVIATIONS: CRP = C-reactive protein;  
ESR = erythrocyte sedimentation rate;  
ICA = internal carotid artery.

arteries are tender and/or pulseless on palpation. Most patients with GCA have an elevated ESR and CRP level on blood tests. Temporal artery biopsy is diagnostic and treatment is with high-dose corticosteroids.

*Other vasculitides*

Rarely, patients with other vasculitides such as systemic lupus erythematosus can present with amaurosis fugax. Patients with hyperviscosity syndromes (e.g. polycythaemia, paraproteinaemias and hyperlipidaemias) may also rarely present first with amaurosis fugax.

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### *Cardiac embolic source*

Patients with a cardiac emboli source, including previously undiagnosed cardiac disease such as bacterial or fungal endocarditis, can present with amaurosis fugax if an embolus from the diseased heart valve pass through the retinal artery. Diagnosis is with clinical examination, echocardiogram and blood tests.

### *Retinal vasospasm*

Some otherwise well young adults present with repeated stereotypical episodes of transient painless visual loss in one eye due to vasospasm. Sometimes there are other vasospastic phenomena (e.g. Raynaud's phenomenon) or sometimes the spasm is purely retinal. This is a diagnosis of exclusion after the other causes of transient visual loss have been ruled out with thorough investigations.

## **SUGGESTED CLINICAL APPROACH**

Amaurosis fugax signifies an impending ophthalmic or neurological emergency and must be immediately investigated. It is recommended that the following evaluation (summarised in the box on page 82) be pursued urgently, preferably on the same day that the patient first presents with amaurosis fugax.

- Physical examination, including blood pressure, temperature, carotid and cardiac auscultations, and temporal artery palpation.
- Urgent examination by an ophthalmologist, looking specifically for emboli in the retinal arteries or other ophthalmic clues to the cause (e.g. retinal cotton wool spots in GCA or Horner's syndrome in ICA dissection).
- Urgent blood tests (full blood count, ESR, CRP level, electrolytes and liver function tests) to look for GCA, other vasculitis or bacterial endocarditis.
- ICA ultrasound to look for ICA stenosis.
- If the ICA ultrasound is normal, or if there is a cardiac history or cardiac signs, echocardiography should be carried out to look for an embolic source such as valve disease.
- If the patient is young, has a history of neck trauma, has neck pain or a Horner's syndrome is seen on eye examination, then MRI plus MRA of the neck should be requested, specifically looking for ICA dissection.

## **CONCLUSION**

Amaurosis fugax (transient partial or complete blindness in one eye) may be a critical warning symptom that the affected patient is about to progress to blindness or a major cerebral stroke. Urgent referral and investigation may save the patient's sight or life. MT

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COMPETING INTERESTS: None.