Visual loss – is it serious?

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Transient visual loss or obscuration can be a symptom of minor ocular surface disorder or it may be the harbinger of permanent and total visual loss or even stroke. The key to determining which of these afflicts your patient is in a targeted history, comprehensive examination, judicious investigation and appropriate treatment or referral.

Case presentation

A 78-year-old woman presented to her GP describing the sudden onset of right-sided visual loss. The episode initially resolved after five minutes but then progressed rapidly and painlessly to total loss of vision in her right eye several days later. The woman also described six weeks of difficulty with swallowing, which was later associated with jaw claudication and pain behind her right eye. These symptoms occurred on a background of polymyalgia rheumatica that had been well controlled with oral prednisolone 2.5 mg per day.

On ophthalmological review, her visual acuity was measured to be count fingers in her right eye and 6/9 in her left eye. The right eye had a relative afferent pupillary defect, and her right visual field to confrontation demonstrated global field loss. Examination of the anterior segment of the eye was otherwise normal for both eyes. Fundoscopy revealed a pale and swollen optic nerve head in the right eye (Figure 1). The left fundus was normal.

Further investigations revealed an elevated erythrocyte sedimentation rate (ESR) of 91 mm/hour (normal <45 mm/hour). The C-reactive protein (CRP) level was also elevated at 100 mg/L (normal <10 mg/L). These results and the clinical findings led to the provisional diagnosis of giant cell arteritis (GCA) and further blood tests and a temporal artery biopsy were arranged. Platelets were elevated at 421 x 10^9/L (normal 150 to 400 x 10^9/L) and there was a normocytic anaemia (haemoglobin level 108 g/L; mean corpuscular volume 87.7 fL).

The patient was admitted to hospital and commenced on a three-day course of high-dose intravenous methylprednisolone (1 g/day) in an effort to prevent bilateral visual loss. Oral prednisolone (55 mg/day, i.e. 1 mg/kg/day) was then commenced prior to discharge. The temporal artery biopsy, which was performed two days into admission, revealed histological features consistent with GCA.

The patient was seen three weeks later in an outpatient clinic to assess her ongoing corticosteroid therapy. Although her ESR had lowered further to 26 mm/hour, visual acuity in her right eye remained poor at hand movements only. Her other symptoms of arteritic ischaemia had resolved.

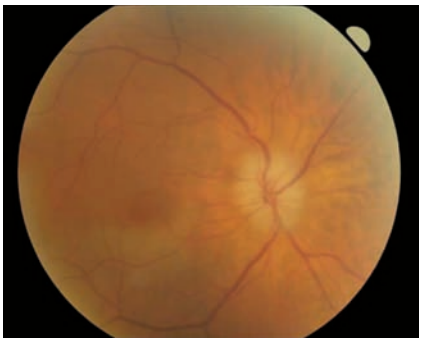

Figure 1. Colour fundus photograph showing pallid disc swelling in giant cell arteritis anterior ischaemic optic neuropathy.

Case discussion

GCA (or temporal arteritis) is a medical emergency with the potential to cause blindness and rarely death.[1,2] The prevalence varies from 0.5 to 25.4 people per 100,000, and it is most common within Caucasian populations.[3] GCA can involve any medium or large artery in the body,[3] but it often affects the superficial temporal artery and as a result the hallmark symptoms are jaw claudication and headache (occurring in less than 30 to 80% of people with this condition). Visual symptoms occur when the ophthalmic artery or its branches are affected (occurring in less than 20% of people with this condition).[4] A provisional diagnosis can be made based on these symptoms, as well as an abnormal temporal artery examination and a rising ESR. Definitive diagnosis is made with a temporal artery biopsy.

Up to 15% of patients with polymyalgia rheumatica may develop coincident GCA; however, standard treatment for polymyalgia rheumatica is with low-dose prednisolone, which unfortunately is inadequate as prophylaxis against GCA.

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Table 1. Causes of monocular and binocular visual obscuration

Visual loss duration association	Causes of monocular visual obscuration	Causes of binocular visual obscuration
Seconds duration	Tear film abnormalities Papilloedema Orbital tumour Optic disc drusen	Tear film abnormalities Papilloedema Optic disc drusen
Less than 10 minutes duration	Carotid occlusive disease Cardiac emboli Giant cell arteritis	Vertebral or basilar artery disease Cardiac emboli Postural hypotension Syncope Giant cell arteritis
Minutes to hours duration	Ocular migraine Recurrent hyphaema Vitreous debris Transient angle closure glaucoma	Cerebral migraine Pituitary tumour Transient ischaemic attack
Associated with exercise or elevated body temperature (Uhthoff's phenomenon)	Optic neuritis	Optic neuritis
Associated with pain on eye movement	Orbital disease/tumour Optic neuritis	Orbital disease/tumour Optic neuritis
Associated with changes in head position	Papilloedema – especially idiopathic intracranial hypertension Optic disc drusen	Papilloedema – especially idiopathic intracranial hypertension Optic disc drusen

Therefore, patients with polymyalgia rheumatica and a rising ESR should alert the medical practitioner to GCA. An increase in ESR, even in the absence of typical symptoms such as jaw claudication and headache, must prompt an urgent referral of the patient for either rheumatological or ophthalmological assessment. Anaemia of chronic disease, rising platelets or CRP levels and an abnormal temporal artery on examination (i.e. tender, enlarged, relatively nonpulsatile) are also predictors that the patient may develop a severe ischaemic manifestation such as loss of vision.[5,6]

Once GCA (with visual loss) has been diagnosed, initial treatment involves high-dose intravenous methylprednisolone (Depo-Medrol, Depo-Nisolone) for three days followed by 1 mg/kg/day of oral prednisolone (Panafcortelone, Solone). This should be tapered down after one month according to ESR response, and then continued for one to two years on a lower dose. Even if one eye is irreversibly damaged, careful management is required to avoid subsequent vision loss in the other eye. Rheumatological review is often required and corticosteroid-sparing agents (e.g. methotrexate) have been used with varying success. Regular GP review is also required to monitor for adverse effects of corticosteroids such as diabetes mellitus, peptic ulceration and osteoporosis.

History

The key to determining the significance of transient visual obscuration lies in the history. This will help to identify the anatomical site of the cause and lead to a more targeted examination. The first important historical feature is to determine whether the visual loss involves one or both eyes (Table 1). Patients must be 'pinned down' on this issue as some may, for example, confuse a right homonymous field loss for loss of vision in the right eye. Visual loss in one eye almost always suggests a lesion anterior to the chiasm. This may be caused by disease of the carotid artery, retinal circulation or optic nerve. Bilateral visual disturbance might indicate bilateral ocular or optic nerve disease or more commonly diseases of the chiasm, optic tract or cerebral cortex, such as impaired posterior circulation or migraine.

The duration of the visual loss is also important (Table 1), as is the degree to which it recovers and the associated symptoms (as demonstrated by this case). Visual loss lasting for seconds to minutes may be caused by embolic phenomena,



Figures 2a to c. A young woman with afferent pupillary defect. She had acute onset of painless visual loss in the left eye (6/60) associated with pain on eye movement. a (left). In dim light, the pupils were equal size. b (centre). When a bright light was shone on the right pupil there was equal constriction of both pupils due to the direct and consensual light reflex. c (right). After the light was quickly swung across to the left pupil, both pupils dilated due to a relative reduction in the afferent impulse from the left optic nerve.

whereas visual loss occurring over days to weeks may indicate either an inflammatory or compressive pathology. A visual obscuration that recovers with blinking is likely to be caused by tear film abnormalities, whereas one that occurs in the setting of headache, photophobia and sonophobia may be caused by migraine.

It is also important to elicit associated symptoms such as double vision, uncomfortable eyes, red eyes, haloes and flashing lights. These symptoms may be associated with other cranial nerve lesions, ocular surface disorders, angle closure glaucoma and migraine. A past history of migraine, childhood motion sickness, family history of migraine, transient ischaemic attack or cerebrovascular accident, atherosclerotic risk factors including cigarette smoking or recent trauma (e.g. neck manipulation, motor vehicle accident or trauma from a roller coaster ride) may also provide clues to the aetiology.

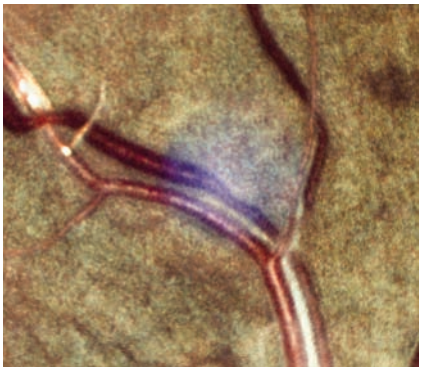
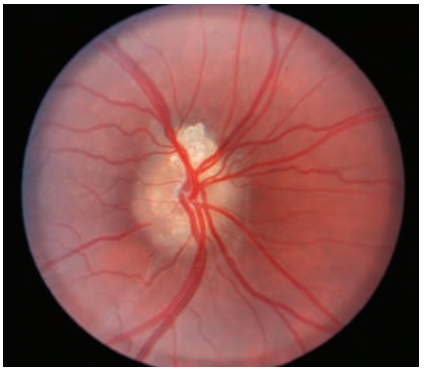
Examination

Patients presenting with transient visual loss require full ophthalmological examination. This includes examining visual acuity, pupils (especially checking for a relative afferent pupillary defect [see Table 2 and Figures 2a to c]), fields to confrontation, extraocular movement, slit lamp examination and fundoscopy (Figures 3a and b). Inspection of the face and eyes should also be directed towards looking for ptosis, proptosis or chemosis (oedema of the conjunctiva).

Historical features and examination findings may also direct the clinician to check the blood pressure or blood sugar levels, and to auscultate the carotid arteries for bruits or the heart for murmurs. It may also be necessary to check cranial nerve function or to conduct a more thorough neurological examination, especially in the setting of an homonymous field defect. If GCA is suspected, it

Table 2. Steps in examining a patient for a relative afferent pupillary defect

- Dim the lights
- Ask the patient to fix on a distant target (e.g. the top letter on the Snellen chart)
- Use a bright light source
- Alternate the light rapidly (less than one second) between the two eyes, spending two seconds on each eye
- Compare the initial constriction and the initial dilatation of the pupils
- Shining the light on the normal eye will result in bilateral pupillary constriction
- Swinging across to the abnormal eye will result in bilateral pupillary dilatation

Figures 3a and b. Colour fundus photography. a (left). An embolus within a branch retinal artery. b (right). Optic nerve head drusen.

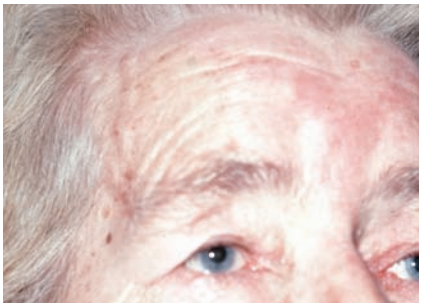


Figure 4. An elderly woman with giant cell arteritis. The temporal artery is hardened and tortuous. On palpation it was tender and nonpulsatile.

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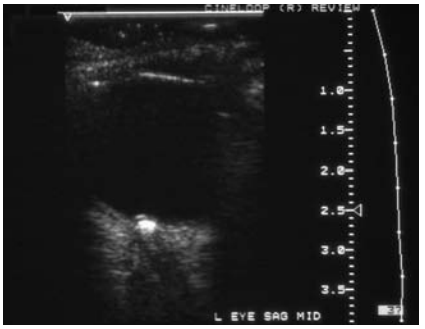

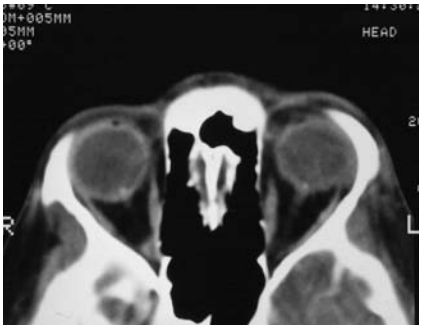

Figures 5a and b. Optic disc drusen. a (left). Optic disc drusen ultrasound of the eye demonstrating the high echoes being returned from calcified drusen in the left optic nerve head. b (right). An axial CT scan through the orbits at the level of the optic nerves demonstrating calcified drusen within the optic nerve heads of both eyes. These were responsible for bilateral transient visual loss with changes in head position.

is also imperative to inspect the temporal arteries for tortuosity and to palpate them, looking for hardening, tenderness or reduced pulsatility (Figure 4).

Investigation

Investigation should be determined depending on the findings from the history and examination of the patient with transient visual loss. Several of the more common and potentially sight- or life-threatening aetiologies will be discussed here.

If amaurosis fugax is suspected to be caused by a transient embolic phenomenon, the patient would require carotid duplex scanning and often an echocardiogram, commencing with a transthoracic echo. If these investigations prove unfruitful but the suspicion is high (e.g. an embolus can be seen in a retinal arteriole), one should proceed to a transoesophageal echocardiogram and CT angiography looking for more proximal carotid artery disease, including of the aortic arch. Screening for cardiovascular risk factors should also be undertaken.

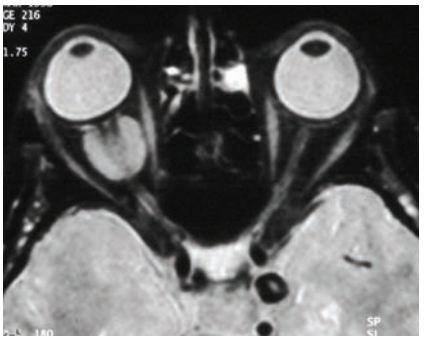

Figure 6. An axial CT scan through the orbits at the level of the optic nerves demonstrating an optic nerve sheath meningioma encasing the right optic nerve. In this case it was resp - onsible for gaze evoked transient visual loss.

Suspicion of GCA in the setting of visual loss should prompt immediate referral of the patient for ophthalmological review. Patients should have their ESR and CRP level checked and a complete blood examination (CBE); if the diagnosis is suspected, a temporal artery biopsy can be performed.

If examination findings are suspicious for raised intracranial pressure, proptosis, chemosis, homonymous field defect or eye movement disorder, CT imaging of the brain and orbits will be required, although for definitive diagnosis an MRI is often necessary. If idiopathic intra - cranial hypertension is suspected, a lumbar puncture will be needed. Optic disc drusen can be demonstrated on ocular ultrasound (Figure 5a) and may be confirmed on CT scan (Figure 5b). Computed tomography will also demonstrate any intraorbital pathology that may be contributing to transient visual loss (Figure 6).

Conclusion

Patients often present with transient changes in vision for which no cause can be found. These changes may be attributed to tear film abnormalities and, less commonly, migraine. However, it is important to remember that more sinister causes for visual obscuration exist, including GCA, carotid or cardiac emboli, orbital tumours and cerebrovascular events. A careful history, complete examination, astute investigation and correct referral will aid in early diagnosis and management of the patient and prevent further loss of vision, or even life. MT

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Further reading

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COMPETING INTERESTS: None.