An elderly man with persistent headaches and visual loss

MEDDUMA B. KAPPAGODA
MB BS, DO, FRCS(Ed), FRACS, FRACO, FRCOphth

A history of persistent headaches and visual loss in an elderly patient may be a sign of impending irreversible bilateral blindness. A relevant history and careful clinical assessment are essential in making a diagnosis.

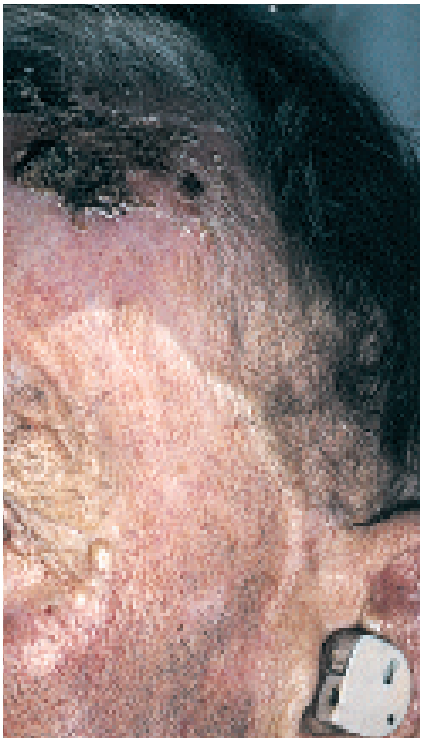

Figure 1. Prominent temporal artery and ischaemic necrosis of the scalp.

Case report

Presentation

A 78-year-old man presented with sudden onset of blindness in his left eye. He had been unwell for four weeks, with headaches as well as aches and constant pains in his limbs. He had not had much sleep during this period.

The patient wore a hearing aid but enjoyed relatively good health. He gave no history of hypertension or diabetes, was a nonsmoker and was not taking any regular medications. He was philosophical about his blindness but wondered whether it could occur in the right eye.

Examination

The patient appeared to be in pain, and pointed to the left side of his head as the site of the pain. The left superficial temporal artery was prominent, nonpulsatile and acutely tender to touch (Figure 1). He did not experience pain on jaw movement but admitted to tiredness in the jaw muscles while eating. Although he complained of weakness and pain in his shoulder muscles, no tenderness could be elicited.

His visual acuity was 6/9 in the right eye with correcting spectacles but he was unable to read 6/60 with the left. The right pupil reacted briskly and promptly to direct light but there was an afferent pupillary defect in the left.

Ocular fundus examination showed a slightly swollen left optic disc with a peripapillary linear haemorrhage (Figure 2). Visual field testing showed a dense field defect inferiorly in the left eye, which corresponded with the location of the ischaemic area.

Differential diagnosis

The patient's uniocular loss of vision and afferent pupillary defect indicated an optic nerve lesion, and the acute onset of blindness suggested vascular pathology. Pallid oedema of the disc associated with a peripapillary linear haemorrhage is typical of anterior ischaemic optic neuropathy, which is associated with systemic diseases:

- giant cell arteritis
- systemic hypertension
- diabetes
- connective tissue disease
- carotid artery disease
- migraine.

The prodromal symptoms of myalgia followed by temporal headaches and acute visual loss strongly suggested giant cell arteritis and led to prompt estimation of the patient's erythrocyte sedimentation rate (ESR, 110 mm/h). Nonarteritic anterior ischaemic optic neuropathy (which is distinct from giant cell arteritis) occurs in younger patients; visual loss tends to be less severe and ESR is not raised.

Dr Kappagoda is former Head of the Department of Ophthalmology, Concord Hospital, Concord, NSW.

Progress

Treatment was commenced immediately with 80 mg prednisolone daily, and the patient's headache and muscular aches were much relieved the following day. High dose steroids were maintained for four weeks, then tapered off. The ESR was used as an inflammatory marker and the symptomatic improvement was used to monitor dosage.

The ESR became normal and stable after three months. Examination of the fundus showed that the left optic disc had become pale from optic atrophy. Apart from poor vision in the left eye, the patient has been free from symptoms of giant cell arteritis and his vision in the right eye has remained unaltered.

Discussion

Giant cell arteritis is a polysymptomatic disease resulting from disseminated granulomatous arteritis involving elastic arteries of large to medium size. It is a disease of the elderly, generally occurring in patients between 60 and 80 years of age. Women are affected three times as often as men.

Classic histology shows fragmentation of the internal elastic lamina with a granulomatous inflammatory reaction involving the media (Figure 3), proceeding to panarteritis and eventual fibrosis with occlusion of the lumen.

The anterior part of the optic nerve is susceptible to segmental infarction from occlusion of the centripetal end vessels derived from the posterior ciliary system. The central retinal artery lacks internal elastic lamina and is therefore not involved in the inflammatory process.

Signs and symptoms

A prodromal stage of systemic symptoms, which may include weight loss, malaise, weakness or low grade fever, is followed in one or two weeks by a continuous disabling headache with episodic exacerbations. The abrupt onset of severe pain is often localised to the frontotemporal or occipital region.

Myalgia

Polymyalgia rheumatica may occur in the prodromal stage of giant cell arteritis and is then considered to be part of the disease spectrum. The myalgia affects the shoulders, neck, thighs and buttocks – there is stiffness and pain but no associated tenderness. Symptoms are worse in mornings. There is pain in jaw muscles and pain on chewing and (usually) pain and tenderness of the scalp, temple and forehead. Polymyalgia rheumatica is usually followed by headache, then by visual loss or disturbance of ocular movement.

Isolated polymyalgia will need diagnostic work up to exclude other conditions, such as malignancy and connective tissue disorders.

Preliminary visual symptoms

In 10% of patients, the initial symptom is visual rather than systemic:

- amaurosis fugax (grey patches or 'steam rising', lasting minutes)
- visual hallucinations (lights and 'stars' just before onset of blindness)
- transient diplopia.

Visual loss

Visual loss is the first complaint in nearly 50% of all cases. Ophthalmoplegia from ischaemia to the third and sixth cranial nerves occurs in 12% of patients. Visual disturbances of cerebral origin (such as hemianopia or cortical blindness) are rare.

Assessment

Blindness caused by giant cell arteritis may occur without classic symptoms and signs. The cause may then need to be differentiated, particularly from the nonarteritic type of ischaemic optic neuropathy (in which ESR is not raised and steroids are of no benefit). Medical evaluation is required to rule out other vasculitidies, such as systemic lupus erythematosus (SLE). Other ocular causes of visual loss (such as central retinal vascular occlusions or retinal detachment) can be excluded by ophthalmic examination.

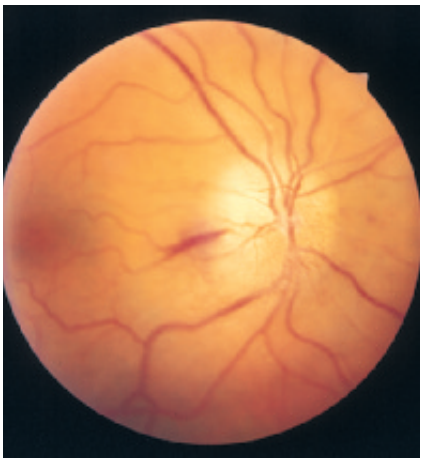

Figure 2. The blurred disc margins, segmental pallor of the disc, peripapillary linear haemorrhage and normal appearance of retinal vessels.

Ocular findings

A palpable, nonpulsatile temporal artery may be evident. Tenderness over the artery is extremely acute.

Visual field

The visual field defect may be:

- altitudinal (commonly, loss of the lower half of the field)
- segmental or sector-shaped
- arcuate or central scotoma
- peripheral contraction.

Pupils

An afferent pupillary defect of the light reflex accompanies the loss of vision. Pupil paralysis may occur from oculomotor nerve palsy.

Ocular fundus

A day or two after the onset of blindness, the optic disc may show slight swelling. The disc may show pallid

swelling, appear hyperaemic or even normal. The appearance of the disc is similar to that seen in nonarteritic anterior ischaemic optic neuropathy.

Fundal changes may include:

- pallid oedema of the optic disc with small peripapillary linear haemorrhages (90% of cases)
- diffuse ischaemic retinopathy
- cotton wool spots.

After the ischaemic episode, optic atrophy develops early. The optic disc swelling subsides (usually in two or three weeks) to produce a pale disc with well defined margins. The atrophy may be segmental or show excavation resulting in cupping of the disc.

In the absence of optic disc changes, an intracranial or orbital lesion should be excluded. Optic nerve sheath infiltrations and rapidly expanding intracranial or orbital masses can lead to similar loss of vision with acute onset.

Investigations

The ESR should be measured immediately. An elevated ESR is the diagnostic feature of giant cell arteritis – a value of more than 70 mm is shown in 80% of patients – but arteritis does occur (rarely) with a normal ESR. C reactive protein is often elevated and normocytic, normochromic anaemia may be found in 50% of patients.

If the ESR is normal in the presence of a relevant history and suggestive features, temporal artery biopsy may become necessary. Pathological changes can be confined to short segments; therefore, a segment of at least 3 cm should be obtained. However, a negative result does not invalidate the diagnosis, so the biopsy does not necessarily change diagnosis or treatment.

Management

Treatment is urgent. If the patient has suffered visual loss in one eye, early treatment is essential to prevent blindness in the fellow eye.

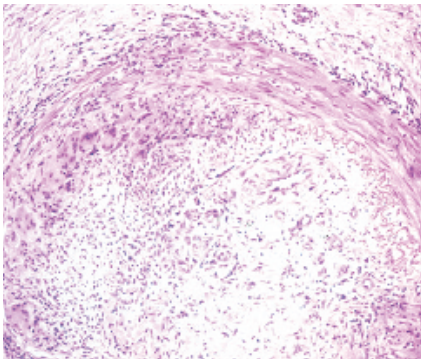

Figure 3. A temporal artery biopsy showing fragmentation of internal elastic lamina and giant cells.

Systemic treatment should be commenced with prednisolone ([Panafcortelone, Solone] 80 to 100 mg daily). Corticosteroids provide early symptomatic relief, normalise the ESR and offer protection against loss of vision.

The high dosage should be maintained for four weeks and then tapered off. The daily dose is tapered off by 10 mg per week, provided that the ESR is maintained at 40 mm. Treatment is monitored by the ESR, a clinical indicator of disease suppression.

Treatment is usually required for about one year, with the minimum dose needed to keep the ESR normal and the patient asymptomatic.

It should be remembered that long term corticosteroid therapy is attended with complications, particularly in elderly patients, and that one should treat the patient, not the ESR. ESR escape may occur after withdrawal of corticosteroid therapy.

Myalgia alone may be controlled symptomatically with nonsteroidal analgesics, but these agents cannot protect against blindness.

Key points

- Giant cell arteritis is a clinical diagnosis. A high index of suspicion is necessary.
- Estimation of the ESR is an essential investigation.
- Urgent treatment with high dose corticosteroids is necessary to prevent loss of vision. Without treatment, there is a significant risk of bilateral blindness.

Course

Giant cell arteritis is a self-limiting disease with a natural history of about 12 months' duration. Clinical indicators and ESR evidence are used to suggest that the disease has run its course.

Blindness is the serious complication: visual loss is abrupt, severe and maximal at onset; subsequent improvement is rare although it can occur with treatment. Without corticosteroid therapy, the fellow eye may become affected in 30 to 40% of those presenting with blindness in one eye.

Loss of vision usually occurs within two weeks and up to four weeks after the onset of headache. The chance of losing vision after six to eight weeks is slight. Ocular complications are rare 10 months after the onset of systemic symptoms.

Involvement of the second eye occurs usually within three weeks after visual loss in the first eye. After six weeks the chance of losing vision in the second eye becomes negligible.

Conclusion

Giant cell arteritis is a medical emergency. Commonly, a patient is symptomatic before the onset of blindness. The diagnosis should be suspected in an elderly patient presenting with vague ill health, myalgia and headache.

Sudden loss of vision in an elderly patient with a history of persistent headaches should alert the clinician to a provisional diagnosis.

Urgent treatment with corticosteroids is essential. MT