Lesson of the Week

Preventable blindness in giant cell arteritis

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Giant cell arteritis is an important cause of blindness in the elderly. Often vision may be lost because of a delay in diagnosis. The commonest ophthalmic presentation is anterior ischaemic optic neuropathy with optic disc pallor and swelling; also recognised are diplopia, occlusion of the central retinal artery, and amaurosis fugax.¹ These, in the presence of an increased erythrocyte sedimentation rate or plasma viscosity, require prompt steroid treatment. We report on two patients with unusual presentations in whom the diagnosis of giant cell arteritis was delayed, resulting in blindness.

Elderly patients with acute visual symptoms should be assumed to have giant cell arteritis until proved otherwise; in this way blindness may be averted

Case reports

CASE 1

A 72 year old woman who had a short history of flashing lights in both temporal peripheral fields presented to the eye casualty department. Her acuity, visual fields, and eye movements were normal. The left pupil was larger than the right, reacting to light and accommodation without afferent pupillary defects. The results of an external eye examination were normal, showing clear corneas and quiet anterior chambers with open angles. The intraocular pressures were 15 mm Hg in the right eye and 6 mm Hg in the left. The results of dilated funduscopy and three mirror biomicroscopy were normal. She was reassured and discharged.

Three days later she returned suffering from a painless loss of vision that had occurred at home 24 hours previously. Her range of vision was reduced to bilateral finger counting only. Both eyes were white with severe corneal oedema, appreciable striate keratopathy, and flare in both anterior chambers; there was no cellular activity. The intraocular pressures were 8 mm Hg in the right eye and 10 mm Hg in the left. The fundi could not be seen clearly because of the corneal oedema. She was thought to have developed a toxic endothelial reaction, with corneal decompensation, to the solutions used during her previous examination. She was admitted and given prednisolone eye drops that did not contain any preservatives. Forty eight hours later her range of vision was reduced to the perception of light by the right eye and to hand movements by the left. She seemed to be inappropriately unconcerned about her loss of vision. On examination poor limbal perfusion and sludging of the blood in the episcleral vessels were noted. When questioned directly she admitted to general malaise and anorexia, brow ache, and jaw claudication: giant cell arteritis was then suspected, and she was given intravenous and oral steroids. Her plasma viscosity was 2.18 mPa.s (normal range 1.50-1.72 mPa.s) and her erythrocyte sedimentation rate 43 mm in the first hour; the results of temporal artery biopsy confirmed the diagnosis.

The next morning her right eye could not perceive light, and the visual acuity of her left eye remained limited to discerning hand movements. Both of her pupils were unreactive, and it was now possible to observe anterior ischaemic optic neuropathy on the right pupil and optic disc pallor on the left. The results of fluorescein angiography confirmed the presence of anterior segment ischaemia. The corneal oedema cleared slowly, and she was

discharged. When subsequently seen at another hospital the corneas were clear, and keratic precipitates were noted. A few weeks later she suffered a fatal myocardial infarction. Necropsy was not performed.

CASE 2

A 64 year old man with a two month history of non-specific headaches, depression, anorexia, and weight loss was seen as a medical outpatient. The headaches were eased by simple analgesics and diazepam. He smoked and drank heavily but had been in good health. He had a longstanding right divergent squint with amblyopia and had palpable pulsations in the temporal artery and normal ocular fundi. He had bilateral carotid and femoral bruits but no other abnormalities. The results of investigations were normal apart from a γ -glutamyl transferase activity of 58 U/l (normal range 0-50 U/l), alkaline phosphatase activity of 155 U/l (40-130 U/l), and plasma viscosity of 1.77 mPa.s (1.50-1.72 mPa.s), equivalent to a Westergren erythrocyte sedimentation rate of 20 mm in the first hour. He was advised to stop smoking and moderate his drinking, after which his headaches improved.

Six weeks later he was admitted to the ophthalmic ward suffering from a sudden painless loss of central vision in his right eye. The previous day he had experienced three episodes of obscured vision, each lasting for 20 seconds. Two weeks earlier he had developed a tender erythematous crusted rash, thought to be herpes zoster, on his scalp. Examination of the right eye showed a pale swollen disc; the left eye was normal. Treatment with oral steroids was started, but because of repeated plasma viscosities of 1.74-1.77 mPa.s and a single erythrocyte sedimentation rate of 19 mm in the first hour it was thought that atheromatous anterior ischaemic optic neuropathy was the most likely diagnosis. Treatment with steroids was stopped the next day and treatment with aspirin and dipyridamole started. After a review of his outpatient history, however, which showed a plasma viscosity of 1.93 mPa.s (equivalent to an erythrocyte sedimentation rate of 30 mm in the first hour) recorded by his general practitioner, a temporal artery biopsy was performed and treatment with steroids restarted on his fifth day in hospital. The next day the patient lost all vision in both eyes; fundal examination showed bilateral, pale, swollen discs. Carotid ultrasound showed that atheroma was present but that there was no turbulent flow. Cranial and orbital computed tomograms were normal. Biopsies confirmed the diagnosis of giant cell arteritis. With treatment plasma viscosity decreased to 1.63 mPa.s within one week, and liver function returned to normal. The concentrations of albumin, globulins, and acute phase proteins were all normal before and after treatment. His scalp ulcers healed, but his vision did not return.

Discussion

The patient in case 1 had giant cell arteritis causing anterior segment ischaemia, presumably caused by disease of the long posterior ciliary artery.^{2,3} This rare presentation has not to our

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knowledge been reported in Britain. The short posterior ciliary arteries supply the optic nerve head, and the long posterior ciliary arteries pass forwards to supply the anterior segment (cornea, iris, and ciliary body), where they anastomose with the anterior ciliary vessels. Acute corneal oedema with hypotony may result from infarction of the ciliary body, which causes changes in the aqueous humour and decompensation in the endothelial cells. These changes may be produced experimentally by occluding these vessels.⁴

The patient in case 2 presented with loss of vision in the right eye and ulcers on his scalp. His erythrocyte sedimentation rate was normal. The evidence for atherosclerosis was strong, and time was wasted pursuing a diagnosis of embolic amaurosis fugax and anterior ischaemic optic neuropathy with an atheromatous cause.

Anterior ischaemic optic neuropathy is the commonest cause of loss of vision in giant cell arteritis⁵ and is caused by occlusion of the short posterior ciliary artery.⁶ The Westergren erythrocyte sedimentation rate usually distinguishes well between arteritic and atheromatous aetiologies and correlates with the findings on biopsy, though patients with giant cell arteritis (confirmed histologically) and a normal erythrocyte sedimentation rate have been reported on.⁷

The vision of about half of the patients suffering from classic temporal arteritis is affected, and blindness may occur while headache and malaise are improving.¹ Occult temporal arteritis, in which the onset of blindness is the first or only manifestation of the disorder, is well recognised.⁵ It affects the skin only rarely, producing conditions ranging from painful erythema to ulceration of the scalp, which mimics herpes zoster.⁸ Depression and apathy are prodromal symptoms in their own right.⁹ About one third of patients yield abnormal results of liver function tests that return to normal with steroid treatment.¹⁰ Abnormal serum protein concentrations are almost universal, with increased erythrocyte sedimentation rate, plasma viscosity, globulin concentration, and C reactive protein concentrations.¹¹ In our second patient all of these values were normal apart from plasma viscosity. Plasma viscosity correlates well with erythrocyte sedimentation rate and is at least as sensitive an indicator of disease in the elderly,¹² but the range of plasma viscosity from 1.73 to 1.80 mPa.s (erythrocyte sedimentation rate 18-22 mm in the first hour) does not indicate disease reliably at any age: such viscosity has been recorded in one fifth of apparently healthy people attending an outpatient department.¹³

Oral corticosteroids are effective at preventing blindness in the

unaffected eye, but established blindness is irreversible.¹¹ Delaying treatment is not justifiable: the second eye may be affected in a few hours or days, even if it was normal at presentation. Results obtained by temporal artery biopsy are not greatly influenced during the first week of steroid treatment, and in any case a negative biopsy result should not inhibit the giving of steroid treatment.^{14 15}

In both of these patients earlier consideration of giant cell arteritis would have resulted in prompt steroid treatment in suitable doses. This disease should always be considered in the differential diagnosis of unexplained visual symptoms in the elderly. We think that these cases illustrate well the observation made by Paulley and Hughes: "When elderly people begin to fail, mentally or physically, this should be one of the first disorders considered and not one of the last."⁹

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Medicine and the Media

TOTAL HIP REPLACEMENT is one of the most rewarding treatments for relieving symptoms and improving the patient's quality of life. At a cost of around £2500 it is excellent value for money, but demand is going to increase as the proportion of the elderly in the population rises and the need for revision operations increases as more younger patients are given the operation.

Last June Tony Van den Bergh, presenter of many medical television programmes, thought that he should be his "own guinea pig" and asked specifically that his fourth operation should be done under both a spinal anaesthetic and television cameras. He hoped that by so doing some of the mystery surrounding operations would be expelled and patients would see that it was acceptable to be frightened. *Treat Me Gently* (ITV, 17 February) will follow his progress from his initial visit to his general practitioner through the operation to his progress after discharge.

Patients admitted to the Royal Free Hospital, where Mr Van den Bergh had his operation, receive their identity bracelet at reception, reminding one of an immediate handcuffing ruling out any hope of escape. The staff throughout were helpful and pleasant and did not

seem overawed by the cameras. There were some amusing incidents and comments: the surgeon described himself as "half blacksmith, half carpenter," and the long term friend, a retired hospital sister, who nobly held the patient's hand throughout his operation, was obviously a great source of strength.

The only occasions when the programme could possibly upset a non-medical audience were when the anaesthetist had difficulty with the spinal anaesthetic because of the patient's arthritis and when the gauze wrapped, dislocated leg looked a bit like a roll of bacon hanging from a butcher's pulley. The patient described himself as "perfectly happy" throughout the procedure even during drilling and hammering. He appeared in excellent shape at the end but described himself as a "bit tipsy" the next day.

For me as a doctor more medical detail would have been interesting. I would have liked to study the radiographs and the special information booklet given to patients. Having had two hips replaced myself, I appreciated the neat fit of the new, differently shaped prosthetic stem. I think that the programme achieved what it set out to do—future patients will be less frightened after