Syndrome of ischaemic ocular inflammation: six cases and a review

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SUMMARY Patients with carotid occlusive disease may develop a variety of symptoms and signs secondary to chronic ocular ischaemia. We report six cases affecting nine eyes and review the findings of teichopsia, anterior segment ischaemia, venous stasis retinopathy, and ocular neovascularisation. The assessment and surgical management of carotid obstruction are discussed. Some comments on the relevance of coexisting diabetes are made.

Although much attention has been paid to the acute ocular manifestations of carotid arterial disease, the chronic aspects have been less well reported. Retinal ischaemia is a more familiar concept than ischaemia of the eye as a whole.

The syndrome of ischaemic ocular inflammation described by Knox2 consists of amaurosis fugax, pain without photophobia, and marked reduction in visual acuity disproportionate to the degree of inflammation. The pupil is dilated, and the fundus shows midperipheral haemorrhages and arterial narrowing.

We discuss the manifestations of chronic ocular ischaemia in six patients with underlying carotid occlusive disease. Successful management may depend on expeditious identification and referral of these patients.

Case reports

CASE 1 A woman aged 39, a smoker and a hypertensive with a history of right-sided transient ischaemic attacks and angina, presented with a two-year history of blurred left vision. This followed an episode of amaurosis fugax. She also complained of pain and persisting after-images in this eye for two months. Visual acuity (VA) was 6/12, with mild anterior chamber activity and early formation of peripheral anterior synechiae (PAS). The fundus showed marked venous stasis with beading, haemorrhages, and microaneurysms (Fig. 1). Fine disc new vessels were present.

We saw her one month after the successful saphenous by-pass graft for a left common carotid artery occlusion which had been demonstrated on angiography (Fig. 2).

CASE 2 A man aged 55, a smoker, presented with a five-day history of a 'fierce pain' over the right eye associated with blurred vision. VA was 6/36, with an unreactive dilated pupil, ruberosis of the angle, and an intraocular pressure (IOP) of 30 mmHg. Iris angiography confirmed ischaemia. There was also venous stasis retinopathy (Fig. 3) associated with cotton-wool spots. On review three months later episcleral injection, uveitis, and new vessels over the disc were also observed.

A left carotid bruit was heard. Angiography revealed occlusion of the right internal carotid artery and left internal carotid stenosis (Fig. 4). A left carotid endarterectomy was performed.

One year after the patient's initial presentation the right VA was perception of light (PL), with formation of a complicated cataract.

CASE 3 A man aged 67, a smoker and a hypertensive recovering from a right hemiparesis, presented with a six-month history of gradually deteriorating left vision. He also complained of pain over the left eye and a persisting after-image described as a 'jagged gold light'. His VA was counting fingers (CF), with episcleral injection, anterior chamber flare, early
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Fig. 1 Left fundus fluorescein angiogram (case 1) showing microaneurysms and venous beading.

Fig. 2 Arch aortogram (case 1) showing occluded left common carotid artery (arrow).

Fig. 3 Venous stasis retinopathy (case 2) well marked in nasal quadrant.

Fig. 4 Carotid angiography (case 2) shows right internal carotid occlusion (black arrow) with left internal carotid stenosis (white arrow).

rubeosis iridis, and an unreactive dilated pupil. Fundal examination showed venous stasis retinopathy and cotton-wool spots.

Carotid auscultation was hindered by an aortic valve murmur, but intravenous digital subtraction angiography showed arch atheroma with left common carotid artery occlusion.

Eight months after the patient’s initial presentation the left VA was PL, with a complicated cataract and rubeotic glaucoma. The IOP was 25 mmHg.

CASE 4

A man aged 66, a hypertensive recovering from a right hemiparesis, was found to be diabetic on
presentation. He complained of left-sided floaters following an episode of amaurosis fugax. His VA was 6/18 right, 6/12 left, with bilateral rubeosis iridis and IOPs of 20 mmHg right, 24 mmHg left. Both fundi showed only mild background diabetic change. In addition there was a left vitreous haemorrhage emanating from the disc.

Two months later he complained of pain over the left eye. His VA was CF, with anterior chamber flare and rubeotic glaucoma, with an IOP of 35 mmHg. A full retinal laser photocoagulation was carried out in this eye. After a further two months the VA on the right had fallen to 6/36, and a right macular branch vein occlusion was noted. The left fundus showed the appearance of an old central retinal artery occlusion associated with a pale disc. Rubeosis iridis was marked bilaterally.

He was experiencing episodes of disorientation. Bilateral carotid bruits were noted, but intravenous digital subtraction angiography was not helpful.

Eighteen months after initial presentation his VA was hand movements right, PL left, with complicated cataracts and rubeotic glaucoma bilaterally (Fig. 5).

Case 5
A man aged 55, a diabetic with hypertension and angina pectoris, presented with episodes of upper field amaurosis in his amblyopic right eye. His VA was 6/60, with an upper macular branch artery occlusion. Six months later he complained of 'coloured lights' affecting central vision, precipitated by straining. Soon afterwards he returned with a right vitreous haemorrhage and disc new vessels. Episcleral injection, anterior uveitis with posterior synechiae, and early PAS were noted. Despite partial retinal laser photocoagulation, gross proliferative retinopathy with traction detachment ensued (Fig. 6). At around this time he suffered two left-sided hemipareses.

On review two years after his initial presentation he gave a history of persisting positive scotomata in the left eye, particularly on waking. His VA was 6/9, and examination of this previously normal eye showed the appearance of venous stasis retinopathy.

Bilateral carotid bruits were detected. Angiography revealed occlusion of the right internal carotid artery, with tight stenoses of the left internal and external carotid arteries.

Case 6
A man aged 77, a hypertensive and ex-smoker, presented with episodes of left upper field amaurosis fugax and blurring of vision. He also complained of seeing bright flashes of light and persisting after-images likened to 'a bright map of Australia'. His VA was 6/5 and no ocular abnormality was found. Fifteen months later he returned with a VA of 6/24 in this eye. Fine rubeosis of the pupil margin, gross disc new vessels (Figs. 7A, B) and peripheral blot haemorrhages on the retina were apparent. A full retinal laser photocoagulation caused only a temporary regression of the disc new vessels. After a further three months he complained of blurred right vision, and this eye also showed venous stasis retinopathy with disc new vessels.

A right carotid bruit was heard. Carotid angiography showed bilateral internal carotid stenoses, more marked on the right, and in addition bilateral
external carotid stenoses were present, more marked on the right (Fig. 8).

The case reports are summarised in Table 1.

Discussion

Because the time sequence of events is relatively short prompt investigation of patients presenting with symptoms and signs of chronic ocular ischaemia is desirable, as early intervention may offer hope of therapeutic effect. For this reason a high index of suspicion is necessary.

The visual symptom of note was the description of positive after-images on exposure to bright light, described variously by our patients as 'jagged gold lights', 'coloured lights', and 'a bright map of Australia'. Ross-Russell and Page1 have previously drawn attention to positive scotomata in patients with carotid artery obstruction. Furlan et al.3 reported five such patients who complained of blurring of vision after exposure to sunlight. Brindley,4 investigating positive after-images, concluded that after the first few seconds these were due to photochemical effects. Possibly choroidal vascular insufficiency leads to compromise of photoreceptor metabolism.

The signs of chronic anterior segment ischaemia1 have been well documented in the context of the changes seen after extensive extraocular muscle surgery.6 Our patients showed episcleral injection, anterior uveitis, and iris atrophy. Iris angiography confirmed ischaemia (case 2). Of six eyes with inflammatory signs four had rubeotic glaucoma, a sign which has not been reported after acute anterior segment ischaemia.5 Neovascularisation of the iris may follow release of vasoproliferative agents, anteriorly from chronically ischaemic iris and posteriorly from ischaemic retina and choroid.7 The eyes with rubeosis had relatively low IOPs (25–35 mmHg), which we think are explained by low perfusion pressures1 causing reduced ciliary body blood flow. Pain in this syndrome189 is probably related to ischaemia rather than to increase of IOP.

Venous stasis retinopathy, described by Kearns and Hollenhorst1 and consisting of venous dilatation, microaneurysms (Fig. 1), and blossom haemorrhages (Fig. 3), is the commonest ocular sign of chronic ischaemia in carotid obstruction, reported in up to 20%180 of cases.

The haemorrhages are larger and darker than those of diabetic retinopathy, are found in the midperiphery, and may be limited to one quadrant. In addition cotton-wool spots may be seen. Disc neovascularisation (Fig. 7) has been described more recently6 as a response to chronic ischaemia of the retina and choroid (Fig. 7B). Diabetic patients present special diagnostic difficulty, for early stasis retinopathy is difficult to recognise. Important clues are marked asymmetry of retinopathy6 (Fig. 6) and gross disproportion in the extent of anterior and posterior segment pathology (case 4).

Earlier reports of chronic ocular ischaemia showed...
most patients to have aortic arch disease, with loss of peripheral pulses due to atheroma, Takayasu’s pulseless disease, or syphilitic aortitis. In our patients the major factor was atheroma affecting the carotid systems. Four of them had central nervous system involvement consistent with carotid artery disease, three of them having had a completed stroke. Amaurosis fugax is the commonest ocular symptom of carotid occlusive disease, and was described by four patients. The manifestations of chronic ocular ischaemia are far less common, and, although they may coexist, patients presenting with amaurosis fugax are not particularly likely to develop chronic ischaemic signs.

Many diagnostic techniques are available for carotid investigation. Ophthalmodynamometry and ocular plethysmography assess carotid artery patency by measurement of the ophthalmic artery pulse pressure. The latter is the more useful test if bilateral lesions are present. The dynamic response to carotid artery compression may be monitored tonographically. Continuous wave Doppler is helpful in establishing whether a significant stenosis is present at the carotid bifurcation, and has the advantage of being non-invasive. Real-time ultrasound can reveal the presence of atheromatous plaques even if they do not have a significant haemodynamic effect. If no carotid artery lesions are present, or if the patients have cardiac arrhythmias, cardiac echography should be carried out to exclude emboli of cardiac origin. Arteriography is the most reliable method of demonstrating carotid artery lesions, but this carries a complication rate of up to 3.7%. Intravenous digital subtraction angiography is safer and has been reported in as many as 96% of selected cases to show a lesion at the carotid bifurcation.

Our findings were similar to those of Brown et al. There was either common carotid occlusion on the affected side (Fig. 2) or bilateral internal carotid obstruction (Fig. 4). The patients with bilateral ocular involvement showed a combination of internal and external carotid stenoses (Fig. 8). The pattern of carotid disease seems to be aetiologically important in the development of chronic ocular ischaemia. Huckman and Haas have reported two cases in which there was reversal of flow in the ophthalmic artery, and have suggested there may be a ‘steal’ syndrome. However, this reversal of flow is a common finding in internal carotid obstruction, and one of our patients (case 6) has orthograde flow in the left and retrograde flow in the right ophthalmic artery, yet shows bilateral ocular ischaemia.

Stenosis of the origin of the internal carotid artery can be relieved by endarterectomy. However, internal carotid occlusion cannot be dealt with in this way, as retrograde thrombosis up to the level of the ophthalmic artery occurs. By taking advantage of the external carotid supply it may instead be by-passed with an anastomosis of the superficial temporal artery to a carotid branch of the middle cerebral artery.

<table>
<thead>
<tr>
<th>Case no.</th>
<th>General health</th>
<th>CNS involvement</th>
<th>Angiography results</th>
<th>Eye</th>
<th>Amaurosis</th>
<th>Ischaemic symptoms</th>
<th>Episcleral injection and/or anterior chamber activity</th>
<th>Rubeotic glaucoma</th>
<th>Low IOP</th>
<th>Ischaemic pain</th>
<th>Venous stasis</th>
<th>Disc new vessels</th>
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<td>1</td>
<td>Hypertension, angina, smoker, 39 yr</td>
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<td>Light-headed, R IC occluded</td>
<td>IC + EC</td>
<td>RE</td>
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<td>-</td>
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*This patient’s left eye showed fine rubeosis of the pupil margin. CNS = central nervous system. CVA = cerebrovascular accident. CC = common carotid artery. IC = internal carotid artery. EC = external carotid artery.
more difficult because it causes a plethora of signs and has to be differentiated from other causes of decreased visual acuity, ocular pain, teichopsia, anterior uveitis, rubeotic glaucoma, retinal venous dilatation with haemorrhages, and disc new vessel formation. Early recognition of the syndrome of chronic ocular ischaemia is important because there is now hope of useful improvement by reconstructive vascular surgery in the initial stages, while treatment later on fails to halt progression of the disease.

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References


Fig. 8 Subtraction angiogram (case 6) showing right internal (straight arrow) and external carotid stenoses (curved arrow).
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