LESSON OF THE MONTH

Transient monocular visual loss due to uveitis-glaucoma-hyphaema (UGH) syndrome

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Abstract

Uveitis-glaucoma-hyphaema (UGH) syndrome is an unusual cause of transient monocular visual loss which may follow cataract extraction and intraocular lens implantation. If misdiagnosed as amaurosis fugax, patients may undergo unnecessary investigations and inappropriate treatment with aspirin.

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Keywords: transient visual loss, cataract extraction, intraocular lens, hyphaema

We present two illustrative cases of uveitis-glaucoma-hyphaema (UGH) syndrome in which misdiagnosis was made.

Case reports

CASE 1

A 67 year old man presented to his general practitioner with transient loss of vision in the left eye. He had experienced 12 episodes over the preceding 3 months. These episodes lasted up to half an hour varying between a slight impairment of vision and a dense mistiness which cleared slowly. He had stopped smoking three months earlier, but had no other risk factors for cerebrovascular disease. Medical history was unremarkable apart from a left extracapsular cataract extraction and posterior chamber intraocular lens implant performed four years previously. A diagnosis of amaurosis fugax was made and the patient was started on aspirin before referral for a neurological opinion. The neurologist found no abnormality on examination and all investigations proved normal (full blood count, erythrocyte sedimentation rate, serum glucose, serum lipids, ECG, chest radiography, echocardiogram, and magnetic resonance angiography of the carotid arteries). Treatment with aspirin was continued, but the episodes of transient visual loss now occurred more often.

Sixteen months after his initial presentation, the patient developed reduced vision in the left eye associated with pain and photophobia. He was duly referred to an ophthalmologist who found his vision to be reduced to hand movements due to a dense microscopic hyphaema. This cleared spontaneously, but recurred a few days later. It was apparent that

his previous episodes of transient visual loss were, in fact, due to UGH syndrome. The intraocular lens was decentred superiorly with the haptics located in the ciliary sulcus. There were no other ocular or systemic conditions to account for the recurrent hyphaemas. Aspirin was withdrawn with a subsequent reduction in the frequency of his transient visual disturbances. He had already developed some visual field loss due to associated glaucoma, which was subsequently controlled by a topical β blocker.

CASE 2

A 72 year old man was referred to an ophthalmologist after his optician found raised intraocular pressure in his left eye. On referral, he complained of transient visual loss affecting the same eye over the preceding nine months. Each episode was characterised by a rapid deterioration of vision over several minutes followed by gradual recovery over a period of 10 minutes to 2 hours. Occasionally this was accompanied by a slight left sided headache. He had had bilateral extracapsular cataract extractions with posterior chamber intraocular lens implants performed 8 years previously. His only risk factor for cerebrovascular disease was a positive family history. The examining ophthalmologist made a diagnosis of primary open angle glaucoma and commenced treatment with a topical β blocker. The episodes of transient visual loss were attributed to amaurosis fugax and the patient was referred for a neurological opinion. The neurologist found no abnormality on examination and all investigations proved normal (full blood count, erythrocyte sedimentation rate, serum glucose, serum lipids, ECG, chest radiography, echocardiogram, and magnetic resonance angiogram of the carotid arteries). Although the visual symptoms were considered atypical for amaurosis fugax, treatment with aspirin was started.

Table 1 Causes of transient monocular visual loss

Amaurosis fugax (retinal microembolisation)
Papilloedema of raised intracranial pressure
Giant cell arteritis
Retinal migraine
Hypotension (orthostatic, arrhythmia)
Haematological causes (hyperviscosity syndromes, coagulopathies, anaemia)
Uveitis-glaucoma-hyphaema syndrome
Intermittent angle closure glaucoma
Optic disc drusen

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Table 2 Comparison between the classic symptoms of visual loss in amaurosis fugax and UGH syndrome

	<i>Amaurosis fugax</i>	<i>UGH syndrome</i>
Speed of onset	Sudden (seconds)	Gradual (minutes)
Character of onset	Dark curtain passing across visual field	Gradual misting of vision $\pm$ erythropsia
Recovery	Rapid (seconds to minutes)	Slow (hours to days)
Complete loss of light perception	Present in at least one sector of visual field	Never present
Associated pain	None	$\pm$ Ache in affected eye

Over the next 9 months, the episodes of visual disturbance occurred more often. The intraocular pressure also remained raised despite treatment. Eventually, the patient was seen during an attack when he was found to have a microscopic hyphaema and a diagnosis of UGH syndrome was made. The intraocular lens was decentred superiorly with the haptics located in the ciliary sulcus. There were no other ocular or systemic conditions to account for the recurrent hyphaemas. Aspirin was discontinued, after which the episodes of transient visual loss persisted but at a reduced frequency. His glaucoma remained poorly controlled by topical β blockers and a trabeculectomy was performed which proved successful.

Discussion

UGH Syndrome is an uncommon complication of cataract extraction with intraocular lens implantation.^{1,2} The condition may present as the classic triad of anterior uveitis, glaucoma, and hyphaema, or as its individual elements. It generally develops at an interval—often several years—after cataract surgery. The syndrome was originally described in association with poorly manufactured anterior chamber intraocular lenses which abraded the iris.³ It may, however, also occur with posterior chamber intraocular lenses which are supported in the ciliary sulcus, as in the two patients described. Current methods of cataract surgery by phacoemulsification allow stable fixation of the intraocular lens within the capsular bag which should reduce the occurrence of UGH syndrome.

There are many important causes of transient monocular visual loss (table 1). These can generally be distinguished by a careful history and examination, supported by appropriate investigations. UGH Syndrome is characterised by a rapid reduction in vision over several minutes followed by a more gradual resolution over several hours to days. There is never complete loss of light perception. Other features which are suggestive of the syndrome include erythropsia (reddening of vision) and an ache in the affected eye due to associated anterior uveitis or raised intraocular pressure. Ophthalmic examination during an attack will confirm the diagnosis by demonstrating a microscopic hyphaema. This may be accompanied by anterior uveitis (generally mild) or raised intraocular pressure. Occasionally the intraocular bleed is sufficient to produce a macroscopic hyphaema which is visible without a slit-lamp. Diagnosis of UGH syndrome may be difficult because a microscopic hyphaema can be cleared from the anterior chamber within hours. Gonioscopy is therefore valuable in sus-

pected cases because it may disclose blood in the trabecular meshwork between attacks. Very rarely, spontaneous hyphaemas may occur in eyes which have not undergone cataract surgery due to other causes such as vascular anomalies of the iris, iris neoplasms, rubeosis iridis, and blood dyscrasias.

Whereas amaurosis fugax is the most common cause of transient monocular visual loss, it is important to be aware of the possibility of UGH syndrome in any patient who has previously undergone cataract surgery. Table 2 shows a comparison between the classic symptoms of visual loss in amaurosis fugax and UGH syndrome. Misdiagnosis of the syndrome as amaurosis fugax may result in the patient undergoing unnecessary investigations and being inappropriately treated with aspirin. In both of the patients described, the frequency of visual disturbance increased after aspirin was started and subsequently decreased on its discontinuation. There is evidence that aspirin can exacerbate the tendency to rebleed in traumatic hyphaemas.⁴ It seems likely that its antiplatelet effect would also exacerbate the tendency to intraocular bleeding in UGH syndrome.

Treatment is required for UGH syndrome if the repeated episodes of visual disturbance are disabling or glaucoma develops. Definitive treatment involves intraocular lens rotation, exchange, or removal.⁵ Intraocular lens manipulation may, however, be a complicated procedure and is generally reserved for the more severe cases. Occasionally, an iris blood vessel can be identified as the source of bleeding which can then be destroyed by argon or Nd:YAG laser therapy. The anterior uveitis is generally mild and responds to topical corticosteroids (if required). Untreated glaucoma can, however, result in permanent visual loss and it is important that all patients with UGH syndrome are followed up for its development. The intraocular pressure may be raised only intermittently due to temporary obstruction of the trabecular meshwork by erythrocytes, but chronic elevation in intraocular pressure can develop in some patients. Glaucoma is treated with topical medication or drainage surgery.

1 Percival SPB, Das SK. UGH syndrome after posterior chamber lens implantation. *Am Intraocul Implant Soc J* 1983;9:200-1.

2 Van Liefferinge T, Van Oye R, Kestelyn P. Uveitis-glaucoma-hyphaema syndrome: a late complication of posterior chamber lenses. *Bull Soc Belge Ophthalmol* 1994;252:61-6.

3 Ellingson FT. The uveitis-glaucoma-hyphaema syndrome associated with the Mark VIII anterior chamber lens implant. *Am Intraocul Implant Soc J* 1978;4:50-3.

4 Ganley JP, Geiger JM, Clement JR, et al. Aspirin and recurrent hyphaema after blunt ocular trauma. *Am J Ophthalmol* 1983;96:797-801.

5 John GR, Stark WJ. Rotation of posterior chamber intraocular lenses for management of lens-associated recurring hyphaemas. *Arch Ophthalmol* 1992;110:963-4.

NEUROLOGICAL PICTURE

A syndrome of lower cranial nerve palsies

A 42 year old man presented acutely with left sided head and neck pain. The next day his symptoms had progressed to involve his tongue, which he considered "swollen". Examination disclosed a transient Horner's syndrome and lower cranial nerve palsies involving IX,X,XI, and XII nerves (figs A-C) on the symptomatic side. T2 weighted MR images of the jugular foramen disclosed intramural haematoma (arrow) in the ipsilateral internal carotid artery (fig D) but without compromise of the true lumen of the artery, as confirmed by a normal MR angiogram.

Lower cranial nerve palsies caused by internal carotid dissection are rare. Most ICA dissections occur in the subintimal space and compromise the vessel lumen. However, a small proportion occur in the subadventitial layer, causing haematoma formation with vessel wall expansion into the carotid space. This results in compression of adjacent structures such as the lower cranial nerves without vessel narrowing. As in this case, MR or conventional angiography may be normal.

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