CASE REPORT

Spontaneous hyphaemas requiring a closer look

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SUMMARY

We present four cases of acute blurred vision that presented to the Ophthalmology Emergency Department of University Hospital Waterford. All four patients were found to have a spontaneous hyphaema with neither neovascular cause nor traumatic history. The pathophysiology was initially uncertain. On closer investigation, these patients were found to have rare iris microhaemangiomas (IMs) or Cobb's haemangiomas. They were all treated conservatively and made full recoveries. Though some reports recommend treating IM prior to intraocular surgery, one of these patients proceeded to have uncomplicated cataract surgery at a later date with no need for prior intervention.

BACKGROUND

These cases demonstrate how a rare condition can easily be missed if the clinician is not thinking outside the normal scope of differential diagnoses. Very often, spontaneous hyphaemas are treated without any further investigation as to why they occurred. Owing to the very rare nature of iris microhaemangiomas (IMs), many ophthalmologists may not even know of this disease entity or may mistake the IM as rubeosis, which then leads to unnecessary fundal examinations, fundus fluorescein angiograms (which have their own risks including anaphylaxis) and costly systemic work up investigations (for systemic diseases related to rubeosis). Also, of the very few cases reported in the literature, there has been speculation that performing intraocular surgery on these patients carries an increased risk of intraoperative hyphaema. Although it is a reasonable assumption, one of these cases went on to have uncomplicated cataract surgery without the need for prior targeted treatment of their IMs.

CASE PRESENTATION**Case 1**

A 60-year-old woman presented to her optician, with a sudden onset of blurred vision in her left eye for 4 h that she first noticed while washing

dishes. Her optician noted blood in the anterior chamber (AC) of her left eye and referred her to the ophthalmology emergency department of University Hospital Waterford, Ireland. Her background medical history included peptic ulcer disease and fibromyalgia. She had no previous ophthalmological problems and was otherwise well.

Her vision was reduced in her left eye at 0.22 and 0.1 in her right eye. On examination, she was noted to have a 1 mm hyphaema in the left eye with a raised intraocular pressure (IOP) of 25 in that eye. She was also noted to have several fine tufts of blood vessels in both eyes at the pupillary margin as seen in [figures 1–3](#). Gonioscopy revealed no neovascularisation. Fundal examination was unremarkable. Her hyphaema was treated conservatively with dexamethasone drops and topical IOP lowering agents, and she was referred to the outpatients department (OPD) for anterior segment photos and an iris fluorescein angiogram, shown in [figure 4](#). She subsequently made a full visual recovery. Below are her images.

Case 2

A 72-year-old woman presented to the ophthalmology emergency department after noticing blurred vision in her left eye on awakening. She was a known non-insulin dependent diabetic and also suffered from hyperlipidaemia and hypertension. On examination, her visual acuity was reduced at 0.52 in her left eye and 1.0 in the right eye. She had a visible hyphaema of 1.5 mm and red blood cells in the AC of the left eye, as seen in [figure 5](#). Her IOP was normal at 15 in the left eye. Gonioscopy did not show any neo-vascularisation and her fundal examination did not show any diabetic retinopathy or any signs of ischaemia. She was noted to have

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Figure 1 Photo of the right and left eyes showing small tufts of vessels at the pupillary margins.

Figure 2 Transillumination used to demonstrate IM. IM, iris microhaemangioma.

Figure 3 High magnification slit lamp photo of fine vascular tufts that may easily be missed on clinical examination.

bilateral tufts of iris blood vessels and was sent for anterior segment photos that day. She was also referred to OPD for further anterior segment photos and iris fluorescein angiography, shown in figures 6 and 7. Her hyphaema was treated with dexamethasone drops and cycloplegic agents and she made a full visual recovery.

Case 3

A 53-year-old woman presented to the ophthalmology emergency department of University Hospital Waterford reporting of sudden blurred vision in her left eye ongoing for the last 24 h. Her medical history included Crohn's disease and vertigo. She had been started on g. Latanoprost for primary open angle glaucoma 1 week prior to this.

Her visual acuity was 0.1 in both eyes. Clinical examination revealed red blood cells in the AC with a small blood clot at the 7 o'clock position, along the pupillary margin in her left eye. Her IOP was 45 in this eye. Gonioscopy did not reveal any neovascularisation but showed blood in the angle inferiorly. Dilated fundal examination was unremarkable.

The patient was given a stat dose of g. Azarga (Brinzolamide 10 mg and Timolol 5 mg), g. Iopidine (Apraclonidine), oral Diamox 250 mg SR (Acetazolamide) and g. Maxidex (dexamethasone). Her IOP responded well and on examination the following day the clot at 7 o'clock had resolved. She was referred into clinic where she was noted to have small tuft-like vessels along the pupillary margin in both eyes. Anterior segment photos and an anterior segment fluorescein angiogram were performed, shown in figures 8 and 9.

Case 4

A 79-year-old woman presented to the eye casualty department with a 5-h history of blurring of vision in her right eye. Her medical history was significant for hyperlipidaemia. She was

Figure 4 Iris fluorescein angiogram of the right eye showing IM as hyper-fluorescent spots along the pupillary margin and late leakage on the left. IM, iris microhaemangioma.

Figure 5 Initial photo of left eye showing a stream of blood originating from IM. IM, iris microhaemangioma.

known to be amblyopic in her left eye and had a previous diagnosis of non-neovascular age-related macular degeneration in both eyes.

The patient's visual acuity in her left eye was count fingers and 0.6 in her right eye. The IOP in her right eye was raised at 37. She was noted to have a 1 mm hyphaema in her right eye. Note was also made of nuclear sclerotic cataracts bilaterally. Gonioscopy did not show any neovascularisation and funduscopy showed bilateral retinal pigment epithelium (RPE) changes at the macula. She was treated with dexamethasone drops, apraclonidine drops and cycloplegia. Her hyphaema and IOP settled well on this treatment. One month later, she represented to the eye casualty department, with a further episode of sudden blurring of her right eye, and was noted to have another spontaneous hyphaema. She was again treated conservatively and made a full recovery. No reason was found for her recurrent hyphaema at the time and 3 months later she was brought in for right phacoemulsification. This was performed without complication and the vision in her right eye improved to 0.1. On a routine clinic examination for her macular degeneration, she was noted to have bilateral iris vascular tufts along the pupillary margin. These were so small that they had initially been missed. Below are her anterior segment photos and iris fluorescein angiography, shown in figures 10–12. At her last follow-up, which was 8 years from the initial event, it was noted that she had not experienced any further episodes of spontaneous hyphaema.

INVESTIGATIONS

All four patients had a full ophthalmic clinical investigation including IOP assessment, gonioscopy and dilated fundal

Figure 6 Slit lamp photos showing fine vascular tufts.

Figure 7 Iris fluorescein angiogram of both eyes showing IM. IM, iris microhaemangioma.

examination. They all had anterior segment photographs taken as well as anterior segment fluorescein angiography. After reading the limited literature on IM, it was decided that an anterior segment fluorescein angiogram would be the most appropriate investigation for exposing the extent of the disease. Importantly, in some cases, the pupillary border appeared almost normal and without performing an anterior segment fluorescein angiogram, the diagnosis would not have been confirmed. Although not performed, it would be interesting to see if anterior segment OCT can demonstrate the small excrescences at the pupillary border to aid with diagnosis.

DIFFERENTIAL DIAGNOSIS

1. Traumatic hyphaema: no history of significant trauma was noted.
2. Hyphaema secondary to rubeosis: no history of previous diabetic retinopathy or relevant fundal pathology that would cause rubeosis. Clinical examination did not reveal any signs of disease normally associated with rubeosis.
3. Iris melanoma and other intraocular tumours: ruled out on clinical examination.
4. Juvenile xanthogranuloma: ruled out on clinical examination.
5. Medication related: no patient was on anticoagulants.
6. Valsalva related: the patients denied performing any Valsalva-like manoeuvres prior to their presenting episode.
7. Uveitis related: there was no evidence of anterior uveitis in any of the four cases, only red blood cells were noted in the AC on examination.
8. Idiopathic spontaneous hyphaema: the finding of fine, tuft-like vessels at the pupillary border ruled this out.

TREATMENT

Treatment was chosen based on the standard departmental protocol for treating hyphaemas and for treating acutely raised IOP.

Figure 8 Photo of both eyes, appearing unremarkable (possibly due to the diffuse lighting used). Ectropion uveae just visible at the 6 o'clock position in left eye.

Figure 9 Early fluorescein angiography of left and right eyes showing hyper-fluorescent spots at the pupillary margin, representing IM. IM, iris microhaemangioma.

OUTCOME AND FOLLOW-UP

The patients treated conservatively had full visual recovery and none of the patients required directed laser or surgical treatment.

DISCUSSION

IM, also referred to as Iris Vascular Tufts or Cobb's haemangiomas, are very rare hamartomas of the stromal vasculature of the iris, first described in 1958 by Fechner.^{1 2} They are typically found in older patients (>50 years of age) except for cases associated with myotonic dystrophy, where they may occur in much younger patients.³ Most cases of IM are bilateral though a few unilateral cases have been described.⁴ Owing to their very subtle clinical appearance, they pose diagnostic difficulties and may easily be missed.

The exact aetiology of IM has yet to be elucidated, but associations have been noted with Sturge-Weber syndrome, myotonic dystrophy, diabetes mellitus, juxtafoveal retinal telangiectasis, hereditary haemorrhagic telangiectasia, hypertension and chronic obstructive pulmonary disease.⁵ Owing to the higher incidence of IM in patients in their fifth decade and above, it is thought to be an acquired as opposed to a congenital anomaly.⁴ We did not note any systemic disease association in our four cases presented here.

The most frequent symptomatic presentation of these microhaemangiomas is related to spontaneous hyphaema, which all four of our cases had. Patients may complain of transient blurring of vision, pain associated with increased IOP or erythropsia,

Figure 10 Photo of an amblyopic left eye, using a green filter, showing vascular tufts at the pupillary margin as well as cataract.

Figure 11 Photo of the right eye. One can make out IM along the pupillary border inferotemporally. IM, iris microhaemangioma.

and may therefore be misdiagnosed as having amaurosis fugax, uveitis, Posner-Schlossman syndrome or ocular hypertension.⁵ Clinically, there may be a variety of findings including visible bleeding from a tuft, a visible hyphaema, red blood cells in the AC, raised IOP or visible tufts of thin, tightly coiled vessels seen at the pupillary margin up to 150 microns in size.² Conversely, there may also hardly be any clinical findings, depending on when the patient presents, and this may lead to an inaccurate diagnosis.

It is important to consider IM in all patients with spontaneous hyphaema alongside other differentials including neovascular tufts, iris melanoma, traumatic hyphaema, Valsalva-related hyphaema, medication-related hyphaema, uveitis-related hyphaema and juvenile xanthogranuloma in younger patients. Spontaneous hyphaema associated with uveitis is a well-documented phenomenon,⁶ and it is particularly important to distinguish this entity from IM-related hyphaema as both conditions may have similar clinical features such as cells in the AC. Unnecessary systemic investigations and expensive blood work can thus be avoided if IM is kept in the differential.

In IM, iris fluorescein angiography is perhaps the most beneficial investigation as it will likely reveal much more extensive

Figure 12 Iris fluorescein angiogram of the left eye showing iris vascular tufts.

involvement than is clinically apparent, and will often also reveal involvement of the second eye. Gonioscopy may reveal blood in the angle, but is otherwise normal. Fundal examination is also often normal,⁵ though there have been reports of disc hyperaemia and tortuous retinal vessels associated with IM.⁷ Anterior segment fluorescein angiography is particularly important if treatment is to be considered, in order to localise tufts. Of note, these vascular tufts will often demonstrate leakage of dye in late frames of angiography, however, this has been documented to occur in normal eyes with increasing age as well.²

Currently, treatment is mainly directed against raised IOP and the hyphaema, and, in general, IMs themselves do not require specific treatment. Conservative treatment should follow departmental protocol and may include oral acetazolamide, topical IOP-lowering agents such as β -blockers, topical cycloplegia and topical steroids. However, laser and surgical treatment have previously been employed to directly target IM, and Dharmasena and Wallis⁵ have proposed a management protocol for these lesions. Proposed indications for direct treatment include recurrent spontaneous hyphaema and/or as prophylaxis prior to cataract surgery due to the presumed increased risk of intraoperative hyphaema. The laser settings vary from a spot size of 50–250 μ m, power 100–400 mW and duration 0.05–0.2 s, and several sessions may be required before treatment is complete.⁵ Laser treatment should be directed by a pretreatment iris fluorescein angiogram and the settings should be adjusted according to the therapeutic response. Surgical iridectomy, though not frequently indicated, has also previously been performed to treat lesions resistant to laser treatment or when a tissue diagnosis is needed to rule out other pathology such as neoplasm.²

It should be noted that IM can present as an isolated case of spontaneous hyphaema and laser treatment carries a risk of corneal decompensation, thus active treatment should be reserved for recurrent cases or prophylactic treatment prior to intraocular surgery, if deemed necessary. Winnick *et al*⁸ successfully treated IM with Argon laser prior to cataract surgery, using 50 shots of 200 μ m spot size, 200 mW power and 0.1 s duration. Owing to the high total energy that may be required to fully laser IM, treatment should be broken up into multiple sessions over an extended time period to avoid corneal complications.⁹ Of note, a case is presented in this series of successful cataract extraction without prior laser treatment for IM. In this case, there was neither an intraoperative nor a secondary complication such as hyphaema. Such a case has not been reported previously, and demonstrates that uncomplicated phacoemulsification may be carried out in cases of IM.

In conclusion, it is important to carefully examine all eyes that present with spontaneous hyphaema, particularly at the pupillary border. IMs themselves tend to be more numerous than clinically apparent, and fluorescein angiography can prove invaluable in delineating the extent of the condition. Many of the vascular tufts present are not visible clinically, and without an anterior segment fluorescein angiogram, the diagnosis can easily be missed and the presenting hyphaema may be attributed to other speculated pathophysiology. Recurrent hyphaemas may result from IM and thus it is important to always consider IM in the differential of spontaneous hyphaemas even in the presence of other ocular findings. Treatment, if deemed appropriate, may be successfully carried out with laser photocoagulation, as previously reported, and surgery can also be employed in select cases. The majority of cases, however, can be managed conservatively.

Learning points

- ▶ Anterior segment fluorescein angiography is useful in delineating the extent of iris microhaemangioma (IM), which is crucial if treatment is to be carried out, but may also help make the diagnosis if there is no apparent cause for hyphaema.
- ▶ Do not diagnose idiopathic spontaneous hyphaema without very carefully examining the pupillary margin for microhaemangiomas.
- ▶ The course of this condition is variable; some patients may have recurrent hyphaemas that may require treatment but others may only have a single event.
- ▶ The extent of the disease may be masked clinically.
- ▶ Patients with IM may have successful intraocular surgery without the complication of a recurrent hyphaema.

Contributors RKG saw the patients in the Ophthalmology Emergency Department, arranged initial investigations, started initial treatment and wrote up the case reports. EC aided by performing further investigation including diagnostics and continuing medical treatment, and contributed to the case report write up.

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