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Haemolytic glaucoma occurring in phakic eyes

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SUMMARY The occurrence of haemolytic glaucoma in phakic eyes due to ghost cells has not previously been reported. Three cases of haemolytic glaucoma occurring over two years after massive vitreous haemorrhage in patients with an intact lens iris diaphragm are described. In one case there was histological confirmation of the presence of ghost cells in the anterior chamber. The mechanism proposed for the passage of ghost cells to the anterior chamber is through a defect in the anterior hyaloid face, created as the vitreous liquefies and degenerates. The reported cases were satisfactorily treated by trabeculectomy with anterior chamber washout, which management has not previously been reported in haemolytic glaucoma.

The term haemolytic glaucoma was introduced by Fenton and Zimmerman[1] to describe a case of secondary open-angle glaucoma occurring in a phakic eye shortly after a vitreous haemorrhage. They believed that the condition was due to haemorrhagic debris and pigment-containing macrophages blocking the angle recess and the trabecular meshwork. Fenton and Hunter[2] and Hunter[3] described the condition again in two phakic eyes. Phelps and Watzke[4] later ascribed five cases to the same mechanism, three of their cases after vitreous haemorrhage in phakic eyes, one case followed vitreous haemorrhage in an aphakic eye, and one followed a traumatic anterior chamber haemorrhage. Jaffe[5] mentioned several cases which he considered followed vitreous haemorrhage after cataract extraction.

Campbell *et al.*[6] in a classic paper described two cases of haemolytic glaucoma occurring in aphakic eyes, and they established that the cells blocking the trabecular meshwork were in fact degenerate ghost cells—rigid spherical degenerate red blood cells containing Heinz bodies, which had previously been described in the eye by Fuchs.[7] They presumed that these cells entered the anterior chamber through a rent in the anterior hyaloid face. Campbell *et al.*[8] then described six more cases complicating vitrectomy, and Campbell and Essigmann[9] further described 19 patients with ghost cell glaucoma in aphakic eyes which were clinically of the same pattern, and the diagnosis was confirmed by examination of the aspirate in seven eyes. They considered that the classical haemolytic glaucoma due to macrophage and haemoglobin blockage of the angle must be very rare, and they had not encountered a case. Campbell[10] later described 14 cases of ghost cell glaucoma following trauma, all with forward passage of ghost cells into the anterior chamber via an opening in the anterior hyaloid face. Recently Summers *et al.*[11] reported a case of ghost cell glaucoma following vitreous haemorrhage following cataract extraction, where the ghost cells were confirmed by phase contrast microscopy.

There remains some doubt whether haemolytic glaucoma can occur after vitreous haemorrhage, in the absence of trauma, in an eye with a clinically intact lens iris diaphragm. The earlier cases described include five with vitreous haemorrhage and an intact lens iris diaphragm, though the presence of ghost cells was not established.

We describe three cases with a typical clinical picture of haemolytic ghost cell glaucoma and an intact lens iris diaphragm with histological proof of ghost cell glaucoma in one case.

Case reports

CASE 1

A 73-year-old white female suffered a spontaneous right total vitreous haemorrhage on 11 March 1973 reducing her vision to perception of light, with a poor direct pupillary reaction to light and no fundus details

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visible. The left visual acuity corrected to 6/5, with a normal left fundus. Applanation tonometry was 12 mmHg in each eye. Her general health was said to be good, with neither diabetes nor hypertension.

Over the next two years the vitreous haemorrhage did not clear and she developed a right posterior subcapsular cataract. On 20 August 1976 she presented with right corneal oedema, a pressure of 42 mmHg, and brownish cells (2+) in the anterior chamber. This did not respond to pilocarpine 2% eyedrops. The anterior chamber depths were right 2·7 and left 2·6 mm, with no rubeosis iridis. On gonioscopy the right angle was open all around, with a raised ridge of light fawn coloured deposit overlying the posterior third of the trabecular meshwork, compared with only moderate pigment in the left angle. The right intraocular pressure rose to 70 mmHg in spite of intensive medical treatment.

On 12 January 1977 right trabeculectomy with anterior chamber washout was performed. Postoperatively the right intraocular pressure was low normal, but it then rose to 34 mmHg over the following month and was controlled to 19 mmHg by adrenaline 1% drops. Seven months after operation the vision was hand movements. By electronic tonography Po/C for the right eye was 616 and for the left eye 82. After two years the light fawn coloured layer on the trabecular mesh had disappeared, and the pressure was 12 mmHg on adrenaline 1% eyedrops.

Five years postoperatively the patient's health was failing, pressures were 16 mmHg in the right and 10 mmHg in the left eye on no treatment, and the vitreous haemorrhage could be seen to be clearing through a dense brown cataract.

CASE 2.

A 58-year-old obese white female having poorly controlled hypertension presented on 3 May 1974 with a decreased vision in the left eye of 6/36 due to a haemorrhagic central retinal vein occlusion. The right corrected visual acuity was 6/5, but there was tortuosity of the veins of the right optic disc. The intraocular pressures were right 20 and left 17 mmHg. The vitreous haemorrhage did not clear.

On 27 April 1977 she presented with a blind painful left eye, though the pupil reacted directly to light. There was corneal oedema, with numerous brownish red cells in the anterior chamber and in the vitreous. The intraocular pressures were right 24 and left 75 mmHg, and the anterior chamber depths were right 3·1 and left 3·3 mm, with no rubeosis iridis and no view of the fundus through the dense vitreous haemorrhage. Gonioscopy showed that both angles were wide and open with a line of dark red blood cells along the posterior trabecular meshwork below. The diagnosis of haemolytic glaucoma was made, and she was treated with pilocarpine 2% eyedrops, prednisolone disodium phosphate 0·5% eyedrops, and oral acetazolamide, which controlled the intraocular pressure to 20 mmHg. Electronic tonography recordings were Po/C right 90, left 266.

Over the ensuing eight years the pressure remained controlled on timolol maleate 0·5% eyedrops, with some clearing of the vitreous blood and improvement of vision to hand movements. There is now a posterior subcapsular cataract and pseudoexfoliation of the lens capsule. There is a posterosuperior detachment of the degenerate syneretic vitreous.

CASE 3

A 42-year-old hypertensive white female gave a history of a right vitreous haemorrhage in 1971. It recurred two years later after a minor blow to the right side of her face, but there was no ocular evidence of contusion injury. The vision was right eye, counting fingers, left 6/6; the intraocular pressure was 14 mmHg in right and left eyes. A large right vitreous haemorrhage cleared in five months to reveal a superotemporal retinal vein occlusion with neovascularisation; this was treated with photocoagulation, the vision improving to 6/6.

On 17 December 1974 she suffered a further right vitreous haemorrhage, reducing her vision to counting fingers. It failed to clear in spite of further photocoagulation, remaining as a dense vitreous haemorrhage.

On 7 April 1978 she presented with a painful right eye. The right vision was perception of light, the left vision 6/5, and there was right corneal oedema. The pressure was not controlled by treatment and rose to 50 mmHg, with brownish cells in the anterior chamber, heterochromia iridis (right brown, left grey iris) and a 'hyphaema' which settled with a khaki layer above and a darker layer below (Fig. 1). It was not possible to see the right angle on gonioscopy while the left was wide and open.

A diagnosis of haemolytic glaucoma was made, and a trabeculectomy was carried out with washout of the anterior chamber and broad iridectomy. Histologically, after vital staining with methyl violet, the anterior chamber aspirate was reported to show 'great numbers of degenerate red blood cells or ghost cells as described by Campbell *et al.*[6] together with a small number of red blood cells and a very occasional leucocyte. The ghost cells contain Heinz bodies consisting of precipitated denatured haemoglobin attached to the cell membrane' (C H Greer). Sections of the trabecular meshwork showed 'ciliary muscle and the scleral spur together with the trabecular meshwork and Schlemm's canal, in both of which there are abundant normal appearing red blood cells

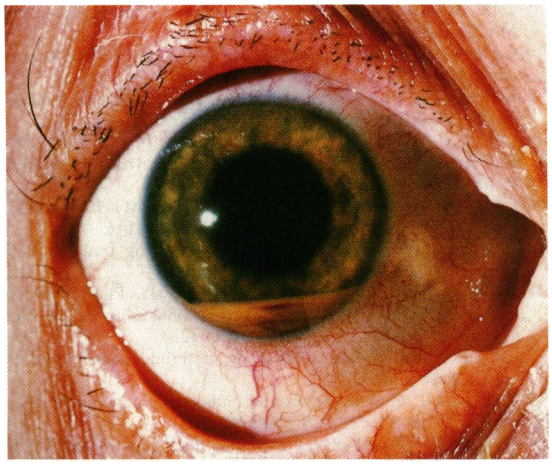

Fig. 1. *'Hyphaema' in case 3 showing the characteristic khaki colour.*

and scanty melanin granules. In addition there are deposits both in the meshwork and on its cameral surface of a light tan-coloured amorphous substance. None of this substance appears to have reached Schlemm's canal.' Sections of the iris showed 'stromal haemorrhages and a minor degree of new vessel formation on the iris surface.' In addition there was 'on the anterior surface of the iris a thin layer composed of red blood cells, leucocytes and some tan-coloured amorphous substance as was present in the meshwork' (C H Greer).

Postoperatively the pressure was controlled, though pilocarpine 2% eyedrops and adrenaline 1% eyedrops were necessary after one week, later being replaced by timolol maleate 0·25% eyedrops.

Over eight years her vision improved to 6/36 with clearing of the vitreous blood, leaving a degenerate liquid vitreous. Her intraocular pressure remained controlled at 20 mmHg with timolol maleate 0·25% eyedrops to the right eye, and the optic disc appears healthy. There is a massive vitreous detachment, with degeneration and syneresis of the vitreous, which superiorly appears shrunken almost to the back of the lens.

Discussion

Of the five phakic patients described prior to the paper of Campbell *et al.*[6] in 1976 four had short intervals of some weeks between the onset of the vitreous haemorrhage and the development of intractable glaucoma, while one case of Phelps and Watzke[4] had a much longer interval of over one year. It remains doubtful whether these earlier cases were due to ghost cells. If they were, the mechanism by which ghost cells migrate from the vitreous cavity to the anterior chamber through an intact vitreous face and lens iris diaphragm is difficult to explain.

In all three of our cases the onset of clinical haemolytic ghost cell glaucoma occurred more than two years after dense vitreous haemorrhage. Clinically the vitreous cavity was loaded with red blood cells and degenerative products, the vitreous itself appearing degenerate. In two cases deepening of the anterior chamber in the affected eye was noted. Campbell and his coworkers[6 9] concluded that ghost cells could not pass through an intact hyaloid face and demonstrated experimentally that such cells layered on the anterior hyaloid face, which appeared to form a barrier to their diffusion from the vitreous into the anterior chamber.

In the patients described here the long-standing vitreous haemorrhage and associated vitreous degeneration may be associated with a functional defect in the anterior hyaloid face and so explain the appearance of ghost cells in the anterior chamber. Cibis[12] considered that degenerate red blood cells might pass into the anterior chamber in patients with ciliary detachment of the vitreous.

These cases are important in their demonstration that haemolytic glaucoma does occur in the presence of an intact lens iris diaphragm, as was confirmed histologically in case 3. Early examination of aqueous aspirates in this condition is valuable. Moreover, the specimens must be filtered with the greatest care to avoid rupturing the cells, as occured in case 1.

The successful treatment of haemolytic glaucoma with anterior chamber washouts has been reported with vitrectomy in recurrent refractory cases.[4 9 13 14] However, treatment by trabeculectomy and anterior chamber washout has not previously been reported; this has the added advantage of long-term control, although some medical therapy was needed for a time postoperatively. In view of the severity of the condition surgery should be used soon after medical treatment has failed, since retention of useful vision is possible.

Haemolytic glaucoma must be differentiated clinically from both haemosiderotic and neovascular (haemorrhagic) glaucoma. Haemosiderotic glaucoma has a more chronic course, with recurrent vitreous bleeding and other signs of haemosiderosis, including retinal degeneration, cataract, iris discolouration, and iron staining of the cornea. However, both conditions may be present in the same eye as in case 3. Neovascular glaucoma is characterised by rubeosis iridis and new vessel formation in the angle.

It may be that a condition of haemolytic glaucoma does occur with an intact lens iris diaphragm and not due to ghost cell obstruction of the drainage angle. However, we believe that true haemolytic ghost cell

glaucoma may occur in phakic patients with long-standing massive vitreous haemorrhage and intact lens iris diaphragm as exemplified by these three patients.

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