Another feature of CGD involves poor wound healing.¹⁰ Therefore, subretinal granulation tissue mass in our case may represent an abnormal reparative response to previous chorioretinal injury. The absence of infection or granuloma, intravitreal pro-inflammatory cytokines, and improvement with immunosuppression suggests that his ocular disease is probably the result of aberrant inflammatory responses. Routine ocular biopsy is not recommended as part of standard ophthalmic evaluation.

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Ocular decompression retinopathy following trabeculectomy with mitomycin C associated with familial amyloidotic polyneuropathy

Familial amyloidotic polyneuropathy (FAP), a disorder inherited in autosomal dominant fashion, is characterised by systemic accumulation of polymerised mutated amyloidogenic TTR (ATTR) in peripheral nerves and in organs.¹ We report two patients with FAP ATTR Y114C (a point mutation, from tyrosine to cysteine, at codon 114) who developed diffuse retinal haemorrhages immediately after uncomplicated trabeculectomy with mitomycin C (MMC).

Case reports

A woman underwent vitrectomy for vitreous opacity associated with FAP ATTR Y114C in the left eye when she was 34 years old and in the right eye when she was 35 years old. Neurological examination revealed that she had polyneuropathy and autonomic dysfunction, and she underwent liver transplantation at 34 years old. Thereafter, intraocular pressure (IOP) in the left eye gradually increased and visual field loss progressed. Trabeculectomy with MMC in the left eye was performed when she was 39 years old. Preoperative IOP was 43 mm Hg despite maximal medical therapy. On the next postoperative day, IOP was 5 mm Hg, and fundus examination revealed scatter retinal haemorrhages in the periphery and posterior pole (fig 1). The haemorrhages had completely resolved and visual acuity was 20/20 in the left eye.

A woman underwent vitrectomy for vitreous opacity associated with FAP ATTR

Y114C when she was 45 and trabeculectomy in the right eye at another hospital when she was 46, respectively. At 48, vitrectomy in the left eye was performed. She underwent liver transplantation at 50. Thereafter, IOP in the left eye gradually increased and non-penetrating trabeculectomy with MMC was performed when she was 51. However, IOP increased postoperatively, and trabeculectomy with MMC was added. IOP before the additional trabeculectomy was 37 mm Hg despite maximal medical therapy. On the next postoperative day, IOP was 5 mm Hg, and fundus examination revealed scatter retinal haemorrhages in the periphery and posterior pole involving the fovea. Although haemorrhages had almost resolved during the follow up period, visual acuity decreased from 20/20 to 20/50.

Comment

Retinal haemorrhages are a rare complication after filtration surgery.^{2–6} Fechtner *et al* reported this condition for the first time under the descriptive term of ocular decompression retinopathy.² Affected patients are typically young, but not exclusively, with relatively high preoperative IOP, a very low postoperative IOP, and advanced cupping. Elevated IOP levels are generally significantly high in cases of secondary glaucoma related to FAP,⁷ and diffuse retinal haemorrhages immediately after uncomplicated trabeculectomy with MMC are noted in both cases. One possible mechanism is hypothesised to be caused by a loss of autoregulation of retinal vessels, which overwhelms their capacity to respond to changes in IOP, resulting in retinal haemorrhages.² Our cases had an autonomic dysfunction as a systemic symptom, further increasing the susceptibility to such a phenomenon. Also, although both

Figure 1 Scatter retinal haemorrhages in the periphery and posterior pole on the first postoperative day.

cases underwent trabeculectomy after vitrectomy, there were no complications, such as the collapse of eye, caused by a vitreous condition. Visual prognosis is usually benign after the resolution of haemorrhages; however, cases with poor visual acuity as shown in case 2 have also been described. Taken together, because clinical features in glaucoma secondary to FAP are related to the pathogenesis of this rare complication after trabeculectomy, early postoperative hypotony should be avoided to prevent it.

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Dapsone induced haemolytic anaemia in patients treated for ocular cicatricial pemphigoid

Ocular cicatricial pemphigoid (OCP) is a systemic autoimmune disease of unknown aetiology. It causes a chronic, scarring conjunctivitis and frequently affects other mucous membranes. Definitive diagnosis is made by immunofluorescent staining of conjunctival tissue demonstrating IgG, IgM, and or IgA in the basement membrane.¹ Dapsone is an immunomodulating sulphonamide and has widely been used in the treatment of mild to moderate OCP.²⁻⁴ All patients treated with dapsone show varying degrees of haemolysis.⁵ Haemolytic anaemia, requiring withdrawal of therapy, has been shown to occur in approximately 10% of patients.²⁻⁵

The medical records of 12 patients treated with dapsone for ocular cicatricial disease were reviewed. Eleven of these patients were treated with dapsone as first line therapy and one as second line therapy; 11 patients had ocular cicatricial pemphigoid (OCP) and one had idiopathic cicatrizing disease. There were an equal number of male and females in this group with a mean age of 72 years (range 55-89 years). The daily dose of dapsone was consistent at 50 mg twice daily taken orally. Mean follow up time while on dapsone therapy was 19 months with a range of 1-60 months.

Six (50%) patients had reticulocytosis including four (33%) with clinically significant haemolytic anaemia with a raised mean cell volume and a steady fall in haemoglobin from baseline. The development of the haemolytic anaemia was not dose dependent and all the patients had a diagnosis of OCP (table 1).

Our cohort of patients had a much higher rate of haemolytic anaemia than previously published reports.²⁻⁵ Although our study sample is small, the proportion of patients with haemolytic anaemia is striking. Patients who are glucose-6-phosphate hydrogenase (G6PD) deficient are known to be more at risk of developing haemolytic anaemia.⁴ We do not routinely check for this rare deficiency in our department. Clinically significant dapsone induced haemolytic anaemia may occur more frequently than previously expected and clinicians should be acutely aware of any downward trend in haemoglobin from baseline. We no longer use dapsone as a first line agent in the management of OCP.

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Bilateral juxtapapillary choroidal neovascularisation associated with interferon alfa treatment of a metastatic cutaneous melanoma

Interferon alfa (IFN α) is commonly used in the treatment of many neoplastic diseases owing to its antiproliferative and immunomodulatory effects. IFN α is used in adjuvant therapy of melanoma stage IIa/b or higher.¹ A wide variety of ocular adverse events related to IFN therapy have been reported during the past decades.²⁻³ A case of bilateral juxtapapillary choroidal neovascularisation is described here.

Case report

A 48 year old woman reported acute vision loss in her left eye (LE) 1 week after starting treatment with IFN α for a cutaneous metastatic melanoma. She had been receiving IFN α , 5 million international units (MIU), subcutaneously three times a week. On examination, visual acuity (VA) was right eye (RE) 20/50 and LE 20/60. Funduscopy showed bilateral optic disc oedema and subretinal haemorrhages in inferior temporal and nasal arcades. To rule out any cause of papilloedema a brain computed tomography was performed, which was normal. One month later, IFN α doses were increased to 8 MIU; VA decreased to RE 20/100 and LE counting fingers at 9 feet. Funduscopy showed bilateral optic disc oedema, bilateral juxtapapillary serous retinal detachment, and subretinal haemorrhages (fig 1A, B).

Table 1 Patients with haematological complications from dapsone

Patient	Age (years)	Sex	Diagnosis	Dapsone dose	Complication
1	77	M	OCP	50 mg twice daily	Reticulocytosis (with normal Hb)
2	78	M	OCP	50 mg twice daily	Haemolytic anaemia*
3	67	M	OCP	50 mg twice daily	Reticulocytosis (with normal Hb)
4	60	F	OCP	50 mg twice daily	Haemolytic anaemia*
5	89	F	OCP	50 mg twice daily	Haemolytic anaemia*
6	85	F	OCP	50 mg twice daily	Haemolytic anaemia*

OCP, ocular cicatricial pemphigoid, Hb, haemoglobin, *dapsone withdrawn.