nerve fibres occur more commonly in association with neurofibromatosis, Down's syndrome and craniofacial dysostosis.[1,8] There are, however, very few reports of familial myelination of retinal nerve fibres.[9,10] The findings of myelinated fibres, in both mother and daughter, suggest that a genetic factor could play a role.

Normal myelination of optic nerve axons is thought to occur in three phases. Firstly, the oligodendrocyte lineage disseminates, via migratory oligodendrocyte progenitors, along large axons. The progenitors are thought to end migration when a local axonal signal falls below a critical level. Secondly, progenitors generate oligodendrocytes responsible for early myelination. The third phase involves consolidation of myelin, via mature progenitors. These migrate only short distances, slowly producing oligodendrocytes.[4]

The formation of aberrant myelinated retinal nerve fibres is thought to be due to failure to prevent oligodendrocyte lineage cells from passing through the lamina cribrosa or optic nerve head. It has been postulated that astrocytes in the lamina cribrosa are specialised to act as a barrier via the orientation and number of their glial filaments.[4] The mechanism by which an inheritable defect could affect this process is still unknown.

It is possible that familial myelinated retinal nerve fibres are more common than stated as they are generally asymptomatic and family members are not regularly screened.

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Sir,

Post-chemotherapy premacular subhyaloid haemorrhage

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We read with interest Rennie's series of premacular subhyaloid haemorrhage treated successfully with Nd:YAG laser in the August 2001 issue.[1] The causes of the haemorrhage in their series included Valslava retinopathy, macroaneurysm, branch vein occlusion, proliferative diabetic retinopathy and idiopathic. We describe here a case of bilateral premacular subhyaloid haemorrhage as a result of chemotherapy-induced pancytopenia.

Case report

A 59-year-old Asian man was referred to the Eye Department 2 weeks after developing sudden deterioration of vision in both eyes. A diagnosis of Stage IV B Mixed Cellularity Hodgkin's lymphoma had been made 6 months previously and chemotherapy had been commenced using the ChlVPP/PABLOE regime (chlorambucil, vinblastine, carbazine, prednisoline/prednisolone, adriamycin, bleomycin, vincristine, etoposide). The second chemotherapy treatment was given 10 days before the onset of his ophthalmic symptoms. This was complicated

by post-chemotherapy pancytopenia, resulting in a haemoglobin of 6.6 g/dl, a white cell count of 0.4×10^9/l, and a platelet count of 17×10^9/l. Five units of packed cells and one unit of platelets were transfused, which returned the haemoglobin count to 12.2 g/dl, white cell count to 3.5×10^9/l, and platelet count to 58×10^9/l.

On examination, his visual acuity was hand movements in the right eye and 6/60 in the left, with no pinhole improvement. The anterior segments and intraocular pressures were normal. Fundoscopy reviewed multiple areas of dense round intraretinal haemorrhages, measuring one-quarter to two disc diameters in size in both eyes. There were also bilateral premacular subhyaloid haemorrhages (Figures 1 and 2).

Nd:YAG laser membranotomy was performed in the right eye at 3 weeks after the initial symptoms. The internal limiting membrane was visibly punctured but no dispersion of blood was seen during the procedure. At 1 week follow-up, there was no change in the visual acuity or fundal appearance. Urgent vitrectomy was arranged, but the patient died from Hodgkin's lymphoma before the operation.

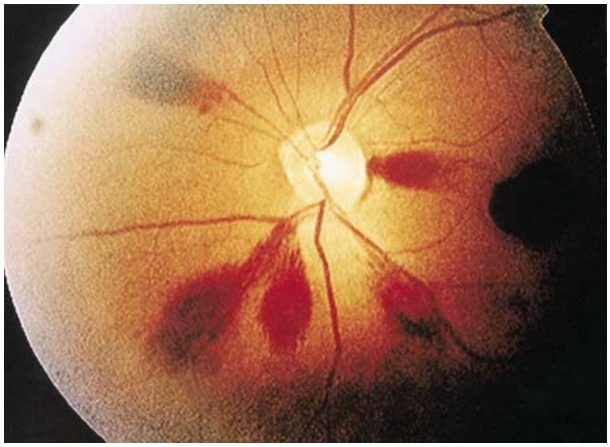

Figure 1 Fundus photograph of the left eye showing well-circumscribed sub-internal limiting membrane haemorrhages, including a large premacular haemorrhage.

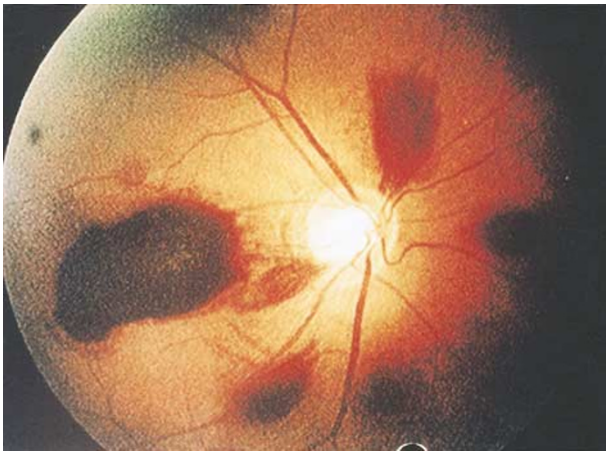

Figure 2 Fundus photograph of the right eye showing similar haemorrhages to the left eye, also including a large premacular haemorrhage.

Comment

This case reports an unusual cause of premacular haemorrhage. The patient suffered from Hodgkin's lymphoma and the onset of symptoms coincided with the development of post-chemotherapy pancytopenia.

Although Nd:YAG laser membranotomy has been well described as an effective mode of treatment, the success rate varies, depending on the cause and the timing of the treatment.[1–4] Complications including macular hole and retinal detachment have been described.[3–5] There has been no study into the time frame for such laser treatment in the literature. We speculate that the failure of the laser treatment in this case is due to the 3-week delay between symptom onset and treatment, during which time the blood had clotted under the internal limiting membrane.

We conclude that premacular haemorrhage should be considered as a complication of chemotherapy-induced cytopenia. Patients should be assessed urgently by an ophthalmologist and Nd:YAG membranotomy should be considered as there is a small window of time in which laser treatment has the highest chance of success. Delay in treatment can significantly reduce the chance of restoring vision in patients who have limited life expectancy.

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Sir,

Uveitis in a patient with common variable immunodeficiency

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Common variable immunodeficiency (CVID) is the most common primary immunodeficiency. It can occur at any age, and is characterised by recurrent bacterial infections, hypogammaglobulinaemia, and deficient antibody responses. It is thought to be acquired, and auto-immune disorders can complicate the clinical picture. Chronic uveitis is often thought to have an auto-immune basis, and we report here a CVID patient who developed multifocal choroiditis with pan-uveitis.

Case report

In August 1997, a 55-year-old emmetropic man who worked as a printer presented with a 3-month history of deteriorating right vision. In 1981 he had a splenectomy for a presumed lymphoma. Subsequent histology and immunoglobulin levels, and a retrospective history of recurrent infections, revised the diagnosis to granulomatous antibody deficiency (GAD), a variant of common variable immunodeficiency (CVID). Since then he has been receiving intravenous immunoglobulin and continuous penicillin prophylaxis.

Unaided visual acuity was 6/18 right and 6/6 left. Inflammatory flare and cells were noted in the anterior chamber and vitreous cavity on the right, and fundal examination revealed numerous peripheral chorio-retinal scars, as well as cystoid macular oedema (Figure 1). There was no evidence of retinal vasculitis, and the left eye was normal except for fine macular drusen.

He was treated with an orbital floor injection of depomedrone 40 mg, and topical steroid eye drops were also started. One month later there had been little clinical response, and the steroid injection was repeated. The uveitis and macular oedema persisted however, and a small inflammatory vitreous 'snowball' was also noted at this time. Further (systemic) immunosuppression was not felt to be justified because of his hypogammaglobulinaemia and previous splenectomy, and also because his other eye was normal.

He was seen again 18 months later when the right acuity had dropped to counting fingers level, and a prominent submacular scar had developed (Figure 2). Moderate intra-ocular inflammatory activity persisted, and similar though milder inflammatory change was now also documented on the left. Vision on that side was, however, well maintained at 6/6, and the clinical picture has remained unchanged since then.

Routine full blood count and plasma viscosity measurements have been repeatedly normal, and serum angiotensin converting enzyme levels have been in the normal range. Thoracic CT scanning had been carried out in 1999 and had revealed bronchiectatic change consistent with his known CVID. It is not possible to test for serology to toxoplasma and toxocara as this would only reflect the serology of the immunoglobulins administered.

Comment

CVID is the most common primary immunodeficiency, and is characterised by recurrent bacterial infections, hypogammaglobulinaemia, and deficient antibody responses. It can occur at any age, and is thought to be acquired: auto-immune disorders and auto-antibodies can complicate the clinical picture. Non-caseating granulomata mimicking sarcoidosis have also been described affecting the lungs, liver, spleen and skin, and appear to be responsive to steroid therapy.

Uveitis can be caused by infections such as syphilis, tuberculosis or toxoplasmosis, but is more commonly 'idiopathic' or associated with systemic diseases that are thought to have an auto-immune basis such as Behcets, systemic lupus erythematosus or Vogt–Koyanagi–Harada syndrome. In our patient the presence of scattered peripheral chorio-retinal scarring in addition to the uveitis suggests a diagnosis of multifocal choroiditis with pan-uveitis (MCP), a descriptive term used to characterise an ocular inflammatory disorder that is thought to have an auto-immune basis and that can be