On detailed questioning, the patient now gave a history of visual deterioration prior to menstruation, and improvement with the onset of menstruation, for several years prior to admission. This history, which correlates with the normal premenstrual progesterone rise, together with the progesterone receptor content of the tumour, strongly suggested that she has a progesterone-responsive tumour. Since the meningioma(s) in this case was considered surgically unresectable, and the history of visual deterioration and resolution corresponding with the peak and trough of progesterone levels during the menstrual cycle, together with the high progesterone receptor content of the tumour, suggested a possible hormonal effect, there was a strong possibility that this tumour could be hormonally sensitive and thus susceptible to endocrine manipulation. Meningiomas have long been known to grow more rapidly during the menstrual cycle or during pregnancy.[4] There is also a higher incidence of meningiomas in individuals who have hormonally mediated tumours such as breast cancer, endometrial or ovarian cancer.[5] An association with uterine leiomyomata has not previously been reported.

High titres of non-oestrogen/oestrogen receptor regulated progesterone receptors (PR) are present in over 70% of meningioma cytosols,[6] and meningiomas of meningothelial type are known to have the highest PR values.[7] By contrast, oestrogen receptors are positive in only 31%. Grunberg *et al.*[8] recently reported an objective response of partial tumour regression in 5 of 14 patients who had unresectable intracranial meningioma and were treated with the progesterone antagonist Mifepristone. We have used the synthetic antiprogesterone hormone gestrinone (ethylnorgestrienone or $C_{21}H_{24}O_2$) to treat this patient. The patient has been taking 2.5 mg of this drug twice weekly for 3 years with no side effects, and she has no further redness or discomfort in either eye. Repeat MRI scan shows no change in tumour size after 3 years (however, these tumours grow extremely slowly), and field analysis shows no significant deterioration. An antiprogesterone agent is still necessary in this patient despite bilateral salpingo-oophorectomy, as progesterone is produced in the subcutaneous tissues and adrenal glands. She will remain on gestrinone, and will be reviewed at 6 monthly intervals. Surgery will only be performed should the tumour extend intracranially, as at the present time tumour resection would render her totally blind. This case demonstrates stabilisation of bilateral optic nerve meningioma with hormonal manipulation.

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Sir,

Cytomegalovirus Retinitis in the Presence of a Normal Helper T-cell Count

Cytomegalovirus (CMV) infection is seen in severely immunodeficient patients with carcinomatosis, following organ transplant, or on cytotoxic or immunosuppressant therapy. CMV retinitis now occurs most often in patients with the Acquired Immune Deficiency Syndrome (AIDS), usually in the presence of a helper T-cell ($CD4^+$) count of less than 0.05×10^9/l.

We describe a relatively fit patient with no risk factors for Human Immunodeficiency Virus (HIV) infection and a normal $CD4^+$ count who presented with CMV retinitis.

Case Report

A well looking 67-year-old woman presented with a 1 week history of painless blurred vision in the right eye. She had suffered from polymyalgia rheumatica (PMR) for several years, but was asymptomatic on prednisolone 5 mg, azathioprine 150 mg and cyclophosphamide 50 mg, daily. She had no risk factors for HIV infection.

On examination her visual acuity was 6/18 right and 6/6 left. There was a moderate right anterior uveitis with 2+ cells and fine keratitic precipitates, and minimal vitreous activity. Right fundus examination showed extensive areas of pale retinal necrosis and haemorrhage in association with the retinal blood vessels, typical of CMV retinitis (Fig. 1). There was no abnormality of the left eye. Her haematological profile was normal; specifically the $CD4^+$ count was 1.2×10^9/l (normal range $0.5\text{-}1.8 \times 10^9$/l). An HIV test was offered but this was declined. Cytomegalovirus was isolated from the patient's urine.

Azathioprine and cyclophosphamide were discontinued and a course of intravenous ganciclovir (10 mg/kg per day) was commenced. Within 20 days the retinitis resolved completely and ganciclovir was discontinued after 6 weeks. Follow-up at 9 months shows no recurrence. An inferior hemi-field defect remains in the left eye.

Discussion

CMV retinitis most frequently presents in AIDS patients, occurring in up to 34% of these individuals.[1] A normal T-cell population seems to be necessary to prevent retinal infection with CMV, and patients with AIDS are particularly at risk of developing CMV retinitis when their $CD4^+$ count drops below 0.05×10^9/l.[2] CMV retinitis in the absence of a low $CD4^+$ cell count is extremely rare in AIDS patients[3] and other immunocompromised patients.[4] Azathioprine is known to depress the action of T cells without decreasing the population, and the effect is reversible under experimental conditions.[5] The patient we present is unusual, being a relatively fit, elderly woman with no risk factors for HIV infection and a normal $CD4^+$ count. It is likely that CMV retinitis occurred following the inhibition of T-cell function by azathioprine, a theory supported by the failure of infection to recur after cessation of this immunosuppressant.

This case highlights the need for prompt ophthalmic referral in immunosuppressed patients with the onset of even mild visual symptoms such as floaters or blurring, as CMV retinitis may be quite advanced before giving rise to subjective visual deficit. The diagnosis of CMV retinitis is largely a clinical one, based on the characteristic clinical appearance, supported by a thorough history seeking risk factors and medications. The $CD4^+$ count should not be relied upon to exclude the possibility of CMV retinitis.

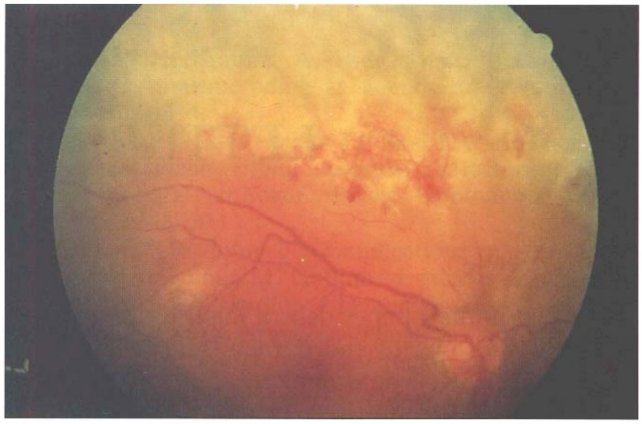

Fig. 1. *Fundus photograph of the right eye showing extensive areas of pale retinal necrosis and haemorrhage in association with the retinal blood vessels, typical of CMV retinitis.*

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