Figure 3 Arteriovenous phase fluorescein angiography. Typical angiographic pattern of a cavernous haemangioma of the optic disc is seen, with numerous small aneurysms showing plasma/erythrocyte sedimentation.

while the posterior third was highly reflective and without spontaneous movement. Axial tomography did not reveal evidence of tumour infiltration of the optic nerve, and a thorough general examination did not disclose any distant metastases. Because of these negative findings it was decided to observe the spontaneous evolution of the tumour and to wait for resorption of the vitreous haemorrhage.

One month later the size of the tumour had decreased and its height measured 4.2 mm; tumour thickness decreased to 1.5 mm 4 months later. This apparent decrease in the tumour size was attributed to possible dissolution of a blood clot located at the top of the tumour. Because the vitreous haemorrhage did not resorb, a pars plana vitrectomy was performed; this restored visual acuity to 20/20. At this point, cavernous haemangioma which had been suspected clinically, was confirmed by fluorescein angiography (Fig 3).

Comment

When ocular melanocytosis is present, the risk of the eye harbouring a uveal melanoma is multiplied by a factor of 30–60.^{3–5} For that reason, in a patient with melanocytosis, it is justifiable to consider any intraocular tumour as a possible malignant melanoma, especially if the media are opaque. In our case, the echographically documented decrease in the size of the tumour made the diagnosis of malignant melanoma unlikely. Moreover, the 7-year evolution of the tumour suggested a stationary or very slowly growing tumour. Diagnosis was made only after a vitrectomy was performed.

The occurrence of a cavernous haemangioma of the optic disc in an eye with ocular melanocytosis may be considered as an extremely rare, albeit fortuitous association. This case illustrates the diagnostic problems presented by intraocular tumours in eyes with opaque media, and also emphasises the need for a long period of observation of any non-progressive intraocular tumour when the diagnosis is not clear.

The known association of melanocytosis with malignant melanoma must not rule out the possibility of a less common intraocular tumour in a patient with melanocytosis.

- 1 Gass JDM. Cavernous hemangioma of the retina: a neuro-oculo-cutaneous syndrome. *Am J Ophthalmol* 1971; 71: 799–814.
- 2 Lewis RA, Cohen MH, Wise GN. Cavernous haemangioma of the retina and optic disc: a report of three cases and a review of the literature. *Br J Ophthalmol* 1975; 59: 422–34.
- 3 Gonder JR, Shields JA, Albert DM, Augsburger JJ, Lavin PT. Uveal malignant melanoma associated with ocular and oculodermal melanocytosis. *Ophthalmology* 1982; 89: 953–60.
- 4 Velazquez N, Jones IS. Ocular and oculodermal melanocytosis associated with uveal melanoma. *Ophthalmology* 1983; 90: 1472–6.
- 5 Bercher L, Zografos L, Egger E, Chamot L, Uffer S, Gailloud C. Mélanocytose oculaire, mélanocytose oculodermique et mélanomes de la chorode. *Klin Monatsbl Augenheilkd* 1991; 198: 361–4.

British Journal of Ophthalmology 1994; 78: 74–76

Atypical cytomegalovirus retinitis: a clinicopathological correlation

Hunter Maclean, James W Ironside, Baljean Dhillon, Sheila M Burns

Department of
Ophthalmology, Princess
Alexandra Eye Pavilion,
Edinburgh
H Maclean
B Dhillon

Department of
Neuropathy, Western
General Hospital,
Edinburgh
J W Ironside

Regional Virus
Laboratory, City
Hospital, Edinburgh
S M Burns

Correspondence to:
Dr Hunter Maclean, Princess
Alexandra Eye Pavilion,
Chalmers Street, Edinburgh
EH3 9HA.

Accepted for publication
17 August 1993

Cytomegalovirus (CMV) retinitis occurs in 15%–20% of patients with AIDS¹ and untreated is a progressive and potentially blinding disease.² Treatment of CMV retinitis with intravenous ganciclovir or foscarnet is well established³ but both of these drugs are only virostatic³ and therefore once CMV retinitis is in remission a maintenance dose is required for the rest of the patient's life to prevent reactivation. We present a case of CMV retinitis which remained in

remission for 20 months without ganciclovir or foscarnet treatment.

Case report

A 26-year-old homosexual man was diagnosed HIV antibody positive in 1988. In February 1990, routine ophthalmic screening revealed HIV retinopathy of the right eye while the appearance of the left eye was of an uncertain

Figure 1 Fundus photographs of the left eye. (a) A large peripapillary lesion of an uncertain nature is seen. Temporally two cotton wool spots typical of HIV retinopathy are present. (b) Two weeks later gross disc swelling with haemorrhage and oedema are present. The vasculitic process spreading along the vessels is typical of CMV retinitis. (c) Four weeks later showing regression of CMV retinitis. (d) One year later complete resolution of CMV retinitis with mild retinal scarring is seen.

nature (Fig 1a). He was managed conservatively but after 2 weeks the fundal picture of the left eye had dramatically changed (Fig 1b). Investigations revealed a CD4 count of 0×10^6 cells per litre and negative CMV serology. Despite this, a clinical diagnosis of CMV retinitis was made and induction therapy of 500 mg ganciclovir twice a day was started. He also began oral therapy with zidovudine 250 mg twice a day. Four weeks later the lesion in his fundus had considerably receded (Fig 1c). By May 1990 his vision had recovered to 6/4 with mild retinal scarring and slight optic disc pallor.

The patient then refused to continue with ganciclovir but did agree to continue with zidovudine. It was explained in detail that this action would be expected to lead to a recurrence of CMV retinitis. Follow up was weekly for the first 2 months and then monthly until his death of a brain stem lymphoma in January 1992. One year after the initial retinitis he showed no signs of recurrence (Fig 1d) and remained free of disease with the same fundal appearance noted 1 week before his death. Autopsy sections of the left retina were normal except in the peripapillary area where evidence of CMV infection was seen

(Fig 2a) and this was confirmed using immunocytochemical stains for CMV proteins (Fig 2b). No evidence of CMV infection was noted in the right eye.

Comment

Without specific anti-CMV treatment resolution or continued remission of CMV retinitis is unusual but has been reported in two similar cases.¹⁴ In both reports CMV retinitis resolved while the patients were receiving zidovudine therapy alone but in the first case CMV was not demonstrated on pathological examination of the affected eye¹ and in the second case it is not known whether the remission was maintained.⁴

In both articles it was postulated that zidovudine, an antiretroviral agent which has no demonstrable effect against CMV in vitro (K Biron, Burroughs Wellcome, oral communication, 1980) may have had an influence in suppressing CMV retinitis. One theory suggested that the action of zidovudine on HIV would lead to an improved immune function thereby suppressing CMV replication. However, in neither of the cases did the CD4 count show any

Figure 2
Photomicrographs of the peripapillary area of the left retina. (a) Destruction of the retinal architecture is seen with many 'owl's eye' inclusions (arrows) typical of CMV retinitis (haematoxylin and eosin, $\times 80$). (b) Immunostain for early and late CMV proteins ($\times 160$). Positive areas show brown staining.

sustained improvement with zidovudine therapy.

In the case presented zidovudine was again the only treatment used while the patient remained in remission from CMV retinitis. However, as in the other cases, no sustained improvement of immune function was seen. Regular CD4 counts fluctuated between 0 and 10×10^6 cells per litre, revealing poor absolute numbers of T helper cells. B cells were also severely disturbed as shown by the negative CMV serology at the time of infection. Subsequently, very low levels of CMV antibody were detected when the patient's stored sera were retested using the sensitive enzyme linked immunosorbent assay (ELISA) method.

Since the laboratory results do not show a convincing rise in immune competence after treatment with zidovudine there is no direct evidence that zidovudine was responsible for the

continued suppression of CMV infection. It therefore remains unclear by what mechanism remission of CMV retinitis was maintained especially in view of the fact that this patient contracted other HIV related diseases during this period, including cerebral toxoplasmosis and HIV myelopathy.

This research was supported by the W H Ross Foundation.

Tissues in this study were provided by the Edinburgh MRC HIV Brain Bank.

- 1 Guyer DR, Jabs DA, Brant AM, Beschoner WE, Green WR. Regression of cytomegalovirus retinitis with zidovudine. *Arch Ophthalmol* 1989; **107**: 868-74.
- 2 Palestine AG, Rodrigues MM, Macher AM, Chan CC, Lane HC, Fauci AS, *et al.* Ophthalmic involvement in acquired immunodeficiency syndrome. *Ophthalmology* 1984; **91**: 1092-9.
- 3 Teich SA, Cheung TW, Freidman AH. Systemic antiviral drugs used in ophthalmology. *Surv Ophthalmol* 1992; **37**: 19-53.
- 4 D'Amico DJ, Scolnick PR, Kosloff BR, Pinkston P, Hirsch MS, Schooley RT. Resolution of cytomegalovirus retinitis with zidovudine therapy. *Arch Ophthalmol* 1988; **106**: 1168-7.