

mas, and inflammatory granulomas. Ferry questioned the existence of iris haemangioma.³

A review of the literature suggests that there are some well documented cases of both cavernous and capillary haemangioma of the iris. The cavernous haemangioma, or microhaemangioma, which is more common, appears as a venous tuft at the pupillary margin. It can bleed and produce a spontaneous hyphaema.⁴ Capillary haemangioma has also been reported to occur in the iris. This rare tumour can be associated with a cutaneous capillary haemangioma and it can show spontaneous resolution coincidental with the natural regression of associated cutaneous lesions.⁵

To our knowledge, racemose haemangioma has not been previously reported to affect the iris. The lesion that we report here appears to be entirely compatible with a racemose haemangioma as seen in the retina.⁶ In contrast with a cavernous haemangioma, a racemose haemangioma looks like a small cluster of grapes and fills slowly with fluorescein. The racemose haemangioma of the retina is actually a complex arteriovenous communication that fills rapidly with fluorescein but does not usually show appreciable leakage of fluorescein. Our case in the iris showed similar angiographic features.

Racemose haemangioma of the retina can be associated with the Wyburn-Mason syndrome, characterised by arteriovenous communications in the midbrain, mandible, maxilla, and other adjacent structures. However, our patient, with an identical vascular lesion in the iris, did not have any clinical manifestation

of that entity. Since our patient is entirely asymptomatic, he has not been inclined to undergo additional studies to detect the presence of other arteriovenous communications.

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- 1 Shields JA, Shields CL. *Intraocular tumors. A text and atlas.* Philadelphia: WB Saunders, 1993:393-419.
- 2 Shields JA, Shields CL. Vascular tumors of the uvea. In: Shields JA, Shields CL, eds. *Intraocular tumors. A text and atlas.* Philadelphia: WB Saunders, 1993: 239-59.
- 3 Ferry AP. Hemangiomas of the iris and ciliary body. Do they exist? A search for a histologically proved case. *Int Ophthalmol Clin* 1972;12:177-94.
- 4 Coleman SL, Green WR, Patz A. Vascular tufts of pupillary margin of iris. *Am J Ophthalmol* 1977; 83:881-3.
- 5 Rutum MS, Mittelman D, Singh P. Iris hemangiomas in infants with periorbital capillary hemangiomas. *J Pediatr Ophthalmol Strabismus* 1993;30:331-3.
- 6 Archer DB, Deutman A, Ernest JT, Krill AK. Arteriovenous communications of the retina. *Am J Ophthalmol* 1973;75:224-41.

'Creeping' cytomegalovirus retinitis in AIDS

EDITOR.—Cytomegalovirus retinitis (CMVR) is the most common sight threatening infection in AIDS. Despite antiviral therapy, recurrent infection is common and leads to progressive visual loss. The classic signs of uncontrolled CMVR include extension of existing lesions, the occurrence of new lesions, fresh haemorrhage or necrosis at the borders of atrophic lesions, or new perivascular infiltrates—early recognition of these signs should prompt increased antiviral therapy. 'Smoldering' CMVR has been described as slow but definite progression of the borders of previously inactive retinitis, and the only sign of activity may be a thin white or grey line between atrophic and unaffected retina.¹ Conversely, border opacification may persist on the edge of healed CMVR without



Figure 1 Retinal vascular closure in the left fundus of patient 1 which progressed over a 3 month period without evidence of retinal opacification.

advancement,² and this should not be confused with active infection. Close attention to the border site, serial retinal drawings, and fundus photographs will confirm movement of the border; however, this may be difficult if the retinal vascular landmarks become obliterated or if border activity is absent.

We report on two patients with CMVR as the AIDS defining diagnosis who exhibited recurrent infection over a prolonged period characterised by progressive retinal vascular closure and retinal atrophy with very minimal sign of border activity.

CASE REPORTS

A 39-year-old HIV positive woman complained of blurred vision in her right eye. The ophthalmoscopic appearance of CMVR at the right posterior pole prompted induction intravenous therapy with ganciclovir at 10 mg/kg for 2 weeks. Severe neutropenia interrupted maintenance therapy allowing CMVR progression. Eighteen months later CMVR was diagnosed in the left eye. Despite intravenous foscarnet therapy, progression continued over a 9 month period with little clinical evidence of border activity (Fig 1). She died more than 2 years after diagnosis of CMVR.

A 30-year-old HIV positive heterosexual man complained of floaters in the left eye. Ophthalmoscopy revealed foci of CMVR in the right peripheral nasal retina and left inferior hemiretina. Intravenous induction ganciclovir therapy failed to prevent progression; therefore, intravenous induction foscarnet was used. Progressive retinal scarring and enlarging bilateral scotoma were documented over an 18 month period with minimal clinical evidence of border activity (Fig 2). Foscarnet toxicity during the maintenance phase led to periods of subtherapeutic dosing during this

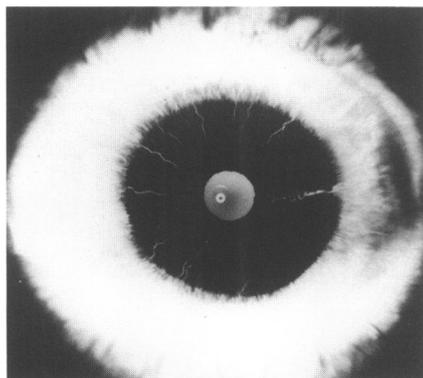


Figure 1 Fluorescein angiogram at 26.4 seconds after injection, showing filling of the prominent artery at the 3 o'clock position temporally. Note that the area adjacent to the artery is relatively void of blood vessels.

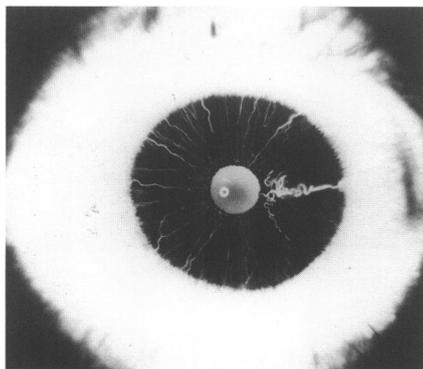


Figure 2 Fluorescein angiogram at 30.9 seconds showing filling of the vascular complex temporal to the pupillary margin and beginning filling of the two draining veins. There was no leakage of dye from the blood vessels in the late angiograms.

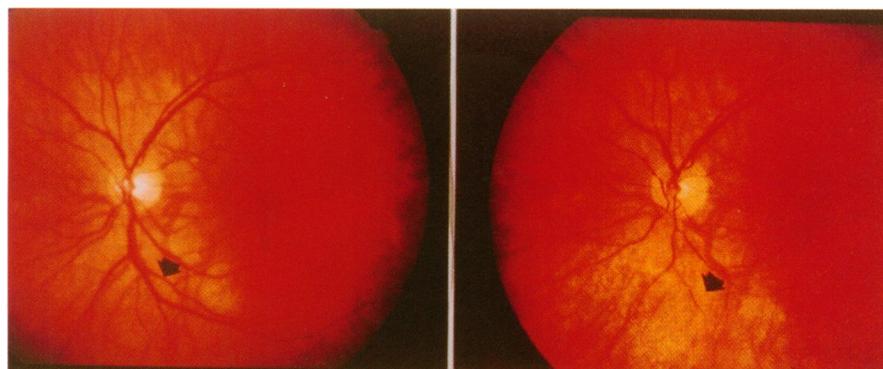


Figure 2 Progressive retinal vascular closure in the left fundus over a 6 month period. Minimal retinal opacification was only observed after creeping disease had been present for a further 8 months. The arrowhead indicates the position of a major arteriole for comparison.

period. The addition of local therapy halted further progression. He died 19 months after developing CMVR.

COMMENT

The clinical appearance of the initial retinal lesions and early clinical response to therapy supported the diagnosis of CMVR. Both patients experienced interruption in maintenance therapy due to systemic toxicity leading to subtherapeutic treatment; therefore, progression was not unexpected. Both patients showed progressive CMVR with little clinical sign of border activity, though retinal vascular attenuation and closure with retinal pigment epithelial reaction at the site of retinal atrophy were observed.

It is assumed that slowly progressive indolent CMVR allows clearing of the necrotic debris compared with the more rapid spread

of more fulminant CMVR.³ The clinical appearance of the CMVR border may be modified by haemorrhological alterations, atypical host response to CMVR, CMV strain differences or a combination of these factors. Histopathological studies of globes with CMVR have failed to demonstrate the presence of virus in the retinal vascular endothelium and the vascular closure observed in our patients is likely to be the consequence of widespread retinal atrophy. It is possible that border activity may have developed between ophthalmic assessments which were arranged at 1–3 week intervals. Creeping disease should be recognised as one form of progression and prompt recognition will allow the treating physician to institute more aggressive therapy before additional normal retina is lost. It is not clear which regimen of systemic and/or local therapy is most efficacious in arresting creeping type disease.

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- 1 Holland GN, Sidikaro Y, Kreiger AK, Hardy D, Sakamoto MS, Frenkel LM, *et al.* Treatment of cytomegalovirus retinopathy with ganciclovir. *Ophthalmology* 1987;94:815–23.
- 2 Keefe KS, Freeman WR, Peterson TJ, Wiley CA, Crapotta J, Quicento JL, *et al.* Atypical healing of cytomegalovirus retinitis. Significance of persistent border opacification. *Ophthalmology* 1992; 99:1377–84.
- 3 Holland GN, Shuler JD. Progression rates of cytomegalovirus retinopathy in ganciclovir-treated and untreated patients. *Arch Ophthalmol* 1992;110:1435–42.