CASE REPORTS

Retinal dysfunction as an initial ophthalmic sign in AIDS

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Abstract

Three cases of patients with AIDS in whom severe retinal dysfunction preceded the development of widespread funduscopy abnormalities are presented. The disparity between the minimal extent of visible retinal lesions and the severe loss of retinal function was confirmed by visual field and electroretinographic studies. This pattern of early visual loss in AIDS patients may represent infection of the retina by the HIV virus.

Previous descriptions of retinopathies associated with AIDS, such as cytomegalovirus, herpes simplex virus, and toxoplasmosis, have emphasised their ophthalmoscopic and histopathological

manifestations.¹ We report here on three patients in whom visual loss and electroretinographic (ERG) abnormalities preceded the development of visible exudative or haemorrhagic lesions of the retina and pigment epithelium. In one patient visual loss was the initial presenting symptom of AIDS.

CASE 1

A homosexual male aged 28 presented with a three-month history of progressive loss of vision in both eyes. A concurrent history of fatigue, weight loss, intermittent diarrhoea, and generalised lymphadenopathy was noted. Visual acuity fell to light perception in the right eye, and barely light perception in the left eye. Examination of the anterior segments gave normal results, but a diffuse vitritis without clumps or skeins was seen in both eyes (Fig 1). No focal lesions could be seen in the retina in either eye. Visual fields were markedly constricted bilaterally (Fig 2). The ERG was extinguished under photopic and scotopic conditions in both eyes (Fig 3).

The patient was admitted to hospital and underwent a diagnostic vitreous aspiration, which revealed only a sparse infiltrate of lymphocytes, histiocytes, neutrophils, and a few pigmented cells. Cultures for bacteria, fungi, and viruses were negative. Silver strains for fungi and *Pneumocystis carinii* were also negative. While in hospital the patient developed *Pneumocystis carinii* pneumonia with pleural effusion. The diagnosis of AIDS was confirmed by positive serology for HIV by enzyme linked immunosorbent assay (ELISA) and western blot tests and inversion of the T-cell helper/suppressor ratio.

Figure 1: Fundus photograph of left eye of patient 1, showing vitreous haze without abnormalities of the retinal architecture.

Figure 2: Goldmann visual fields of patient 1, showing marked, irregular constriction in both eyes.

Figure 2 left

Figure 2 right

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Accepted for publication
31 August 1989

Figure 3: ERG recordings from left eye of patient 1. Upper tracings show response to single flash; lower tracings show average of responses to 10 repetitions of the flash stimulus. ERGs are extinguished under photopic conditions (left) and scotopic conditions (right).

Figure 3 left

Figure 3 right

Figure 4: Goldmann visual field, left eye of patient 2. (The patient was unable to respond consistently to the test targets with the right eye.)

Figure 5: Fundus photograph of right eye of patient 2. There is a mild vitreous haze and a white intraretinal lesion between the disc and macula.

Figure 6: Fluorescein angiogram of right eye of patient 2. During the arterial phase the intraretinal lesion seen in Fig 5 blocks the background choroidal flush (left: 45° photograph). During the late phase of the study the lesion shows persistent fluorescein staining (right: 30° photograph).

Figure 6 left

Figure 6 right

The patient was treated with pentamidine and co-trimoxazole. The pneumonia resolved, and his vision showed a small improvement to 20/100 in the right eye and 20/400 in the left eye. On discharge his vision had returned to the level of light perception. Subsequently the patient's general medical condition deteriorated further, and he ultimately died of *Pneumocystis carinii* pneumonia.

CASE 2

A 35-year-old bisexual male developed *Pneumocystis carinii* pneumonia. A diagnosis of AIDS was confirmed by positive serology (ELISA and western blot tests) and inverted T-cell helper/suppressor ratios. One year later he presented with a six-week history of asymmetrical bilateral loss of visual acuity. On examination his visual acuity was 20/200 in the right eye and 20/30 in the left eye. Visual fields were severely constricted (Fig 4). The anterior segments were normal, but a diffuse vitreous infiltrate was present in both eyes. One white intraretinal lesion, about one-half disc diameter in size, was noted between the macula and the optic disc in the right eye (Fig 5). On fluorescein angiography this lesion blocked fluorescence early but stained homogeneously late in the study (Fig 6). A sector of depigmentation of the retinal pigment epithelium was seen inferonasally in the right eye (Fig 7). No other retinal lesions were seen in either eye.

ERG recordings were significantly reduced in amplitude bilaterally, particularly under scotopic conditions (Fig 8). The maximal B-wave amplitudes elicited from the dark-adapted eyes by an unattenuated white flash stimulus measured 60% and 80% of the lower limit of normal in the right and left eyes, respectively.

Figure 7: Localised retinal depigmentation, nasal to optic disc in right eye of patient 2.

Figure 8: ERG recordings from the right eye of patient 2. Upper tracings, single flash responses; lower tracings, average of 10 responses. Photopic ERGs show blunted waveforms, with B-wave amplitude at lower limit of normal range (left). Scotopic responses are reduced to 60% of normal (right).

Figure 8 left

Figure 8 right

Tests for opportunistic infections (toxoplasmosis, cytomegalovirus, fungi) were negative. A course of ganciclovir produced no effect on visual acuity or on the appearance of the intraretinal lesion. The area of depigmentation of the retinal pigment epithelium appeared to enlarge slightly. The patient remained clinically stable, with reduced vision in the right eye, for several months, but ultimately succumbed to disseminated cytomegalovirus infection.

CASE 3

A 35-year-old black male, employed as a postal worker, was referred for evaluation of rapid visual loss in both eyes over an eight-week period. He was married to a former intravenous drug abuser who had contracted AIDS. Their 9-month old son was born with AIDS. Serological tests, including ELISA and western blot, were positive for HIV infection. The recent medical history was notable for progressive weight loss, anorexia, fatigue, and scattered lymph node enlargement.

An ophthalmological examination showed best corrected visual acuity of finger counting at 1 m in the right eye, and 20/400 in the left eye. The anterior segments were notable for a trace of flare of the aqueous humour and occasional cells in the anterior chambers bilaterally. There was a diffuse vitritis, with a uniform distribution of cells throughout the vitreous cavity, without clumps or skeins. The optic nerves appeared pale. The retinal arterioles appeared narrowed in calibre, but there were no focal retinal or choroidal lesions.

Fundus photographs through the vitreous haze were unsatisfactory. Visual fields, obtained on the Goldmann perimeter, were markedly constricted bilaterally. The ERG was substantially reduced under photopic and scotopic conditions in each eye.

The patient refused admission to hospital. Laboratory studies revealed lymphopenia and immune suppression. A thorough examination for occult bacterial, viral, or fungal infection gave negative results. The patient has remained medically stable, but his vision has continued to deteriorate.

Discussion

The mechanism of the visual loss in these patients is not known. All three patients showed evidence of vitritis clinically, but there was no sign of ophthalmoscopically visible retinal or choroidal inflammation in two patients and only

a very small retinal lesion in the third. Nonetheless the constricted visual fields and reduced or extinguished electroretinograms strongly suggest the presence of widespread retinal dysfunction. In particular, this type of retinal dysfunction may precede the development of the cotton-wool spots, retinal haemorrhages, exudates, and retinal oedema generally understood to comprise the microvascular retinopathy of AIDS. It is conceivable that this dysfunction may represent the ophthalmic equivalent of HIV encephalitis,² particularly in view of the electron microscopic demonstration of particles resembling the HIV virion in retinal tissue obtained at necropsy from patients with AIDS,¹ and the isolation of HIV-1³ and the demonstration of HIV-1 p24 antigen in the aqueous humour of AIDS patients. Recently, Farrell and coworkers reported successful treatment of an HIV-1 positive patient who had an iridocyclitis and retinitis refractory to conventional treatment, with oral zidovudine.⁴ They observed resolution of the ocular inflammatory disease, lending further support to our thesis that direct intra-ocular infection with HIV-1 may be the source of ocular pathology.

Presented in part at the 1987 annual meeting of the Association for Research in Vision and Ophthalmology, Sarasota, Florida.

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