Conjunctival tumour as the primary manifestation of infectious mononucleosis in a 12 year old girl

EDITOR,—Infectious mononucleosis (IM), a common disease in childhood, is an acute lymphoproliferative disease caused by Epstein-Barr virus (EBV), 

a member of the herpes virus family. The association of EBV infection with benign and malignant lymphoproliferative diseases, as well as its role in the pathogenesis of nasopharyngeal carcinoma, is well known. The course of IM is usually self limiting but can be extremely variable, ranging from asymptomatic to a fatal outcome. Fever, pharyngitis, lymphadenopathy, and hepatosplenomegaly are typical systemic manifestations. Essential for the diagnosis is an increase in relative and absolute numbers of lymphocytes and monocytes, including 20–40% atypical forms. Ocular involvement of IM is unusual and occurs often as a mild unilateral follicular conjunctivitis simultaneously with these other symptoms. We describe a girl with a conjunctival tumour as the first manifestation of IM; systemic symptoms of the disease occurred some days later.

CASE REPORT

A previously healthy 12 year old girl with a 10 day history of a painless red left eye was treated with antibiotic eye drops. Despite these, a unilateral conjunctival swelling developed, and the girl was admitted for further ophthalmological consultation. On examination, a 5 × 5 mm red coloured mass in the upper nasal conjunctiva of the left eye was observed. The globe was not displaced, the ocular motility and visual acuity were normal, and funduscopy was unremarkable. At this time, there were no palpable cervical lymph nodes. An excisional biopsy of the conjunctival tumour was performed. The initial histological diagnosis was “high grade malignant B cell lymphoma” and, consequently, the patient was referred to the department of oncology. On examination some days later, the girl was referred to the department of oncology.

A 5 mm red coloured mass in the upper nasal conjunctiva of the left eye was observed. The infiltrate mainly consisted of large polyclonal B immuno blasts, smaller mixed lymphocytes and polytypical plasma cells. Occasional immuno blasts, particularly those close to necrotic areas, expressed the latent membrane protein of the EBV (CS1–4) and were also demonstrated in situ hybridisation for EBV-encoded early nuclear RNA (EBER). In addition, in situ hybridisation for EBV encoded early nuclear RNA (EBER) showed positive reaction in some immuno blasts. The growth fraction of the infiltrating cells was 60–80%, using the antibody Ki-67. On the basis of the conventional histology and immunohistochemistry, in particular of the polyclonal nature of the immunoblasts and plasma cells as well as the demonstration of EBV infection, a diagnosis of IM involvement of the conjunctiva could be made; a malignant lymphoma was excluded. During the subsequent several weeks, all symptoms and signs of the IM resolved spontaneously.

Figure 1 Conjunctival tumour of the left eye (after incisional biopsy).

Figure 2 Epstein–Barr virus infection detected by antibodies against the latent membrane protein (CS1–4) (APAAP, original magnification×40).

shown mildly enlarged para-saorl lymph nodes. The peripheral blood count revealed a relative and absolute lymphocytosis (WBC 13 ×109/L, 76% lymphocytes) with 33% atypical lymphocytes. Elevation of LDH and alkaline phosphatase was noted. The girl was admitted for further examination.

Re-evaluation of the pathological findings by a lymphoma referral centre was undertaken. An extensive lymphocytic infiltrate and large necrotic areas were seen below an ulcerated conjunctival epithelium. The infiltrate mainly consisted of large polyclonal B immunoblasts, smaller mixed lymphocytes and polytypical plasma cells. Occasional immunoblasts, particularly those close to necrotic areas, expressed the latent membrane protein of the EBV (CS1–4) (Fig 2) and were also positive for the EBV related proteins P52 and BZ1. In addition, in situ hybridization for EBV encoded early nuclear RNA (EBER) showed positive reaction in some immunoblasts. The growth fraction of the infiltrating cells was 60–80%, using the antibody Ki-67. On the basis of the conventional histology and immunohistochemistry, in particular of the polyclonal nature of the immunoblasts and plasma cells as well as the demonstration of EBV infection, a diagnosis of IM involvement of the conjunctiva could be made; a malignant lymphoma was excluded. During the subsequent several weeks, all symptoms and signs of the IM resolved spontaneously.

COMMENT

Manifestations of IM in ocular tissues are rare, with the conjunctiva being the commonest tissue involved and usually in the form of a follicular conjunctivitis. In general, the conjunctivitis occurs in parallel with the systemic symptoms of IM described above. Conjunctival involvement in the form of a swelling or a tumour has been reported previously. These occurred in 11, 38, and 14 year old patients, respectively, with the latter age group showing no clinical signs of nasopharyngeal carcinoma. The coexistence of, or subsequent development of, a malignant lymphoma following infectious mononucleosis has been reported. In the present case, the initial histological diagnosis was made of malignant lymphoma. This diagnosis could be subsequently refuted on the basis of the immunohistochemistry and in situ hybridisation which demonstrated a polyclonal lymphocytic infiltrate and the detection of EBV associated antigens on the surface and within the nucleus of the infiltrating cells. The subsequent clinical progression with a self limiting course and the positive serology for EBV support the diagnosis of a lymphoproliferative process associated with an acute EBV infection and infectious mononucleosis. In conclusion, infectious mononucleosis should be considered in the differential diagnosis in patients with conjunctival lymphoid tumours.

Ultrason biomicroscopy in the diagnosis of a foreign body simulating iris melanoma

EDITOR,—Several lesions of the iris can clinically resemble melanoma. Failure to recognise a simulating condition could lead to misdirected therapy. The most common iris pseudomelanomas include primary iris cyst,
Letters

Diagnosis of a foreign body may sometimes be difficult in the absence of a history of injury, especially when slit lamp biomicroscopy of the anterior segment does not reveal any sign of ocular trauma. Imaging modalities such as computed tomography and ultrasonography may help in such situations. However, magnetic resonance imaging is contraindicated in cases where a ferromagnetic foreign body is suspected. Standard B-scan ultrasonography has been replaced at many centres by UBM for the evaluation of anterior segment foreign bodies.  

We used UBM to confirm our clinical impression of an iris foreign body in this case. In addition to characterising the foreign body, UBM detected a subtle irregularity in Descemet’s membrane.

It is of interest that a seemingly metallic foreign body could remain inert for 50 years without causing ocular inflammation, siderosis or secondary glaucoma, which prompted us to choose to observe this patient rather than to intervene surgically.

When the ophthalmologist is confronted with a dark iris lesion, an intraocular foreign body is a possibility, irrespective of a negative history of trauma. Slit lamp biomicroscopy coupled with gonioscopy provides excellent diagnostic clues. We believe that UBM is a sensitive imaging technique for confirmation of the diagnosis.

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Optical coherence tomography findings in early solar retinopathy

EDITOR,—Visual deterioration caused by watching a solar eclipse has been recognised from the time of Plato. The aetiology of solar retinopathy has been attributed to photochemical effects, which may be enhanced by elevated tissue temperature. Typically, a small yellow spot may be noted in the foveolar area immediately after exposure. Histopathological studies of solar retinal lesions revealed retinal pigment epithelium (RPE) and photoreceptor damage. We present two patients with acute, severe solar retinopathy after observation of the total eclipse on 11 August 1999. Fundusoscopic findings were accompanied by optical coherence tomography (OCT) investigation of the macula.

CASE REPORTS

Case 1

An 18-year-old man presented 24 hours after watching the total eclipse without appropriate protection. Total exposure time was approximately 20 minutes. During direct viewing the patient kept his right eye closed. One hour after observation he noted blurred vision and a central scotoma on the left eye. Best corrected visual acuity was 0.8 in the right and 0.1 in the left eye. Fundus examination of the right eye did not show any pathology. Funduscopy of the left eye revealed a yellow lesion in the fovea, surrounded by a circular red area.
A 26 year old woman complained of blurred vision and central scotoma after watching the eclipse without eye protection. Total exposure time was 5 minutes. Forty eight hours after exposure visual acuity was 1.0 in the right and 0.8 in the left eye. Fundus examination showed a small yellow round lesion in the fovea of both eyes. On OCT, a hyperreflective area in the centre of the fovea was demonstrated in both eyes. Similar to case 1, all layers of the fovea were affected. Retinal thickness (110 µm/118 µm) was within the normal range.

**COMMENT**

Solar retinopathy is characterised by a yellow foveolar dot and a central scotoma. For the first time we describe OCT findings of patients suffering from solar retinopathy. The main finding was a hyperreflective area involving all foveal retinal layers without showing any sign of retinal oedema. It correlated in size and location with the characteristic funduscopy visible yellow dot. At present, the origin of this well delineated area of hyperreflectivity is unclear. Moreover, histopathological analysis of solar retinopathy has demonstrated that mainly the pigment epithelium and outer segments of the photoreceptor layer are damaged. The OCT scan however showed pathological appearance of all retinal layers. In one patient we have been able to repeat OCT investigation 9 days after solar exposure, revealing that the retinal changes were reversible, increasing pathological appearance of the RPE and choriocapillaris layer.

In addition, macular oedema, which has been described in the literature before, could not be demonstrated by OCT. On OCT examination, no significant increase in retinal thickness could be observed when comparing the affected and unaffacted eye (case 1) or average values of both affected eyes (case 2). To the best of our knowledge, none of these OCT findings have been published before by other investigators.

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**Granulomatous uveitis, CREST syndrome, and primary biliary cirrhosis**

Eisentraut—There is a well known link between different autoimmune diseases. Up to 84% of patients with primary biliary cirrhosis (PBC) may have at least one other autoimmune disease, such as thyroiditis, scleroderma, rheumatoid arthritis, or Sjögren’s syndrome. The association between uveitis and scleroderma is well recognised, but we failed to find a single report of the association of uveitis and CREST syndrome (calcinosis cutis, Raynaud’s phenomenon, oesophageal dismotility, sclerodactyly, and telangiectasia), a more benign variant of scleroderma in the form of limited cutaneous sclerosis. The simultaneous occurrence of PBC and uveitis is probably also very unusual as we could only find one such case report in our literature search. We report the case of a patient with uveitis whose investigation disclosed PBC and CREST.

**CASE REPORT**

A 48 year old woman with a longstanding history of Raynaud’s phenomenon complained of blurred vision in the right eye over the past 3 months for which she had not sought medical attention. Ophthalmological examination revealed diminished visual acuity (VA) in the right eye to counting fingers. VA in the left eye was normal. Pupillary light reflexes were normal. Normal OCT test was 66.2 µm (normal 5 mm in the right eye and 12 mm in the left). Slit lamp examination showed mutton fat keratic precipitates, flare, and cells (1+ in the anterior chamber and vitreous (anterior and posterior) of both eyes, but differing increasing subcapsular cataract accounting for the visual loss. The fundus were normal without any signs of vasculitis or pars planitis. General clinical examination revealed microstomia, facial telangiectasias, sclerodactyly, and multiple finger pulp cutaneous infaracts.

Laboratory investigations showed raised serum aspartate aminotransferase (131 U/l, normal 0–40 U/l), alanine aminotransferase (154 U/l, normal 7–56 U/l) and γ-glutamyl transferase (274 U/l, normal 8–78 U/l), and alkaline phosphatase (471 U/l, normal 38–126 U/l). Serum bilirubin, calcium, and angiotensin converting enzyme levels were normal. The serum albumin was also normal (38.6 g/l) as was the prothrombin time (12.4 seconds). The serum IgM was 3.66 g/l (normal 0.60–2.63 g/l) whereas IgG and IgA levels were normal. RA test was 662 U/ml (normal 0–20 U/ml), Waaler-Rose was negative, and circulating immune complexes were detected at a level of 2.80 mg/l (normal <1.5 mg/l). Complement levels were normal. Antinuclear antibodies were positive (1:1280) with a speckled pattern. Anticentromere, antimitochondrial (>1:100), and anti-M2 antibodies were also positive. There was no evidence of past or present hepatitis B infection. The VDRL was non-reactive and Lyme, toxoplasma, CMV, and HIV serologies were negative. The HLA phenotype was A1, A9, B17, Bw6, Cw4, Cw6, DR5, DR52, DR8, DQ4, DQ7.

A chest x ray was normal and a radiograph of the hands revealed subcutaneous calcinosis. The Mantoux test (2 units) was anergic. There was no evidence of pathergy. The liver biopsy showed stage III PBC.

This patient was treated with ursodeoxycholic acid, calcium, vitamin A and D supplements. Topical corticosteroids and oral prednisolone 60 mg/day were also started. Systemic steroids were tapered to 10 mg/day after 2 months, and then slowly tapered over the next 4 months until they were discontinued.

There was a temporary increase in liver enzymes (AST, 167 U/l; ALT, 313 U/l; GGT, 774 U/l) which returned to baseline values soon after steroids were stopped. The uveitis resolved and 5 months later, on eye medic, she underwent cataract surgery successfully, significantly regaining vision in that eye (best corrected visual acuity = 1.0).
COMMENT

In PBC, CREST, and uveitis abnormalities of the humoral and cellular immune systems are common. These include increased levels of serum immunoglobulins, the presence of circulating autoantibodies, increased turnover of complement, the presence of activated T and B cells in the peripheral blood, and impaired T cell regulation. 1 2 Despite the high frequency of these immunological abnormalities, a definite immunopathogenetic mechanism has not yet been clearly established for any of these disorders. Furthermore, no single mechanism seems to provide a link between these clinical entities. A mouse model for the development of liver damage through the intrahepatic inoculation of mycoplasma-like organisms, which cause human chronic uveitis, has recently been developed, 3 but it remains a hypothetical concept. Our case report illustrates an association between oculocutaneous lupus and hepatic diseases, which may be entirely coincidental but could, nevertheless, encourage further investigation for a common immunological pathway.

PATIENT HISTORY

The patient was a 65 year old white woman with a painful traumatic blind eye and a large cataract extraction suspected to be choroidal melammas later. There are several ultrasonographic signs cited as characteristic of dislocated lenses: a hyperechogenic mass with an oval shape, which can become highly reflective as cataract develops, and vitreal strands adherent to the mass which is mobile and changes location with eye movements.

We believe this particular ultrasonographic sign can be ascribed to the following process. The lens detaches, encounters the anterior vitreous face, and pushes it posteriorly by its weight towards the retina. The anterior vitreous face, vitreous gel, and collagen fibres within the vitreous are pushed back by the lens creating the wall of an aqueous filled tunnel with the lens at its bottom (Fig 1B).

In most cases this sign is easy to demonstrate, and we recommend it as a useful new clue to the ultrasonographic diagnosis of dislocation of the lens.

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Combined central retinal vein occlusion and cilioretinal artery occlusion in a patient on hormone replacement therapy

EDITOR,—Combined occlusion of the central retinal vein and the central retinal or a cilioretinal artery is unusual. There have been many previously reported systemic and ocular associations. The pathogenesis of this condition, however, is not established and remains controversial. Whether the venous obstruction or the arterial blockage acts as the initiating event is unknown. We describe a patient with combined central retinal vein occlusion (RVO) and cilioretinal artery occlusion occurring 6 weeks after starting hormone replacement therapy (HRT). We observed the evolution of the arterial occlusion. This suggests that the initiating event was the RVO.

CASE REPORT

A 46 year old patient presented with a 2 day history of decreased vision in the left eye noticed on waking. She reported no previous visual disturbances. She had been started on cyclical combined HRT 6 weeks before presentation. She was taking no other medication and had previously used the combined oral contraceptive pill with no adverse effects. She was a non-smoker. There was no past or family history of venous thromboembolism.

Visual acuities were right eye 6/5 and left eye 6/60. Right intraocular pressure (IOP) was elevated (22 mm Hg). There was a left relative afferent pupillary defect. Gonioscopy showed open angles. Blood pressure was 140/90 mm Hg.
Hg. Pulse rate was 72 beats per minute and regular. There was no cardiac murmur and no carotid bruit. On the right there was pathological cupping of the optic disc and venous dilatation (Fig 1). Examination of the left fundus showed dilated, tortuous retinal veins in all four quadrants, with scattered retinal haemorrhages and a swollen, haemorrhagic optic disc. There was retinal whitening in the territory of a cilioretinal artery. No intra-arterial embolus was seen (Figs 1 and 2). She was diagnosed, on clinical grounds, with left combined central retinal vein and cilioretinal artery occlusion and right primary open angle glaucoma.

One week later, the cilioretinal artery had become narrow and irregular in calibre, with fragmentation of the blood column (Fig 2). Two months after presentation, the perrivenous haemorrhages and optic disc swelling had resolved, the calibre of the cilioretinal artery had returned to normal (arrow) and most retinal haemorrhages have resolved.

Hormonal profile confirmed the patient’s postmenopausal status. She was managed with timolol 0.25% drops and various causes of retinal vascular disease caused by a patent central retinal artery and ultimately cilioretinal artery occlusion secondary to the high intravascular pressure. There have been many reported systemic and ocular associations of combined retinal arterial and venous occlusion including temporal arteritis,2 migrane, systemic lupus erythematosus,3 lupus anticoagulant, syphilis,4 optic neuritis,5 and various causes of retinal vasculitis. Relevant investigations excluded any of these as an underlying cause. Elevated IOP probably plays some part in the development of some central RVOs.6 Left IOP was normal at presentation but elevated IOP may have preceded the RVO, with subsequent stagnation of blood flow, elevation of intraluminal capillary pressure caused by a patent central retinal artery and ultimately cilioretinal artery occlusion secondary to the high intravascular pressure.

COMMENT

The pathogenesis of combined retinal venous and arterial occlusion is not established. Whether the venous obstruction or the arterial blockage acts as the initiating event is unknown.7,8 Carotid artery disease has never been described as a cause of simultaneous retinal arterial and venous occlusion. Evolution of the arterial occlusion has been documented in this case. This suggests that the initiating event was the RVO, with subsequent stagnation of blood flow. Elevation of intraluminal capillary pressure caused by a patent central retinal artery and ultimately cilioretinal artery occlusion secondary to the high intravascular pressure.

Histological examination of the pigment epithelium-Bruch membrane-chorio-capillaris complex after macular translocation

EDITOR.—Macular degeneration is the leading cause of blindness in the older population, and it is becoming more and more prevalent.1 At present there is no treatment for the dry type of macular degeneration; for treatment of the wet form, several medical and surgical therapies have been tried, with varying results.2,3 Photodynamic therapy with verteporfin had a significant treatment benefit in predominantly classic choroidal neovascularisation (CNV) lesions.4

Surgical removal of the subretinal membrane is a promising method, but in many cases the visual acuity does not improve. A new technique has been proposed by Machemer and Steinhorst, where after surgical removal of the subretinal membrane the fovea is translocated to an area with healthier pigment epithelium, Bruch’s membrane, and choroid.2,5

Variants have been described where only the temporal part of the retina was detached, or the retinal detachment was created in the temporal side through the sclera, without retinotomy. Here the scleral resection will ultimately shift the relation of the pigment epithelium to the fovea.

CASE REPORT

In this case we performed a macular translocation using the Machemer and Steinhorst technique, with some modifications from Eckardt (personal communication, 24 October 1997), and from our experience. After surgery usually we see a retinal fold starting from the optic disc and extending to the periphery, but this fold usually disappears after 5–7 days.

Our patient died (from intracranial haemorrhage) on the fifth day after the operation, and it was possible to examine the operated eye pathologically.

At gross examination we found that rotation had been achieved through an angle of 25–30 degrees. The next step was to perform...
sections of the translocated macula and in the region of the former subretinal membrane, and our findings are discussed below.

COMMENT
As we expected we found a thickened Bruch’s membrane, few points of calcification, some part of membrane remains in place even after surgical “removal” and from the membrane situated only above the pigment epithelium this will not occur. We think that more research is needed to discover if there is any connection between the type of membrane, related to the condition of the pigment epithelium, and the apparent stronger adhesion between the pigment epithelium and retina (or relatively weaker adhesion to the Bruch’s membrane).

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1 Machemer R. Macular translocation. (Editorial.)  

Optic neuropathy as the presenting feature of HIV infection: recovery of vision with highly active antiretroviral therapy

EDITOR.—We describe a rare case of bilateral optic neuropathy caused by HIV which responded to highly active antiretroviral therapy (HAART).

CASE REPORT
In March 1996, a 52 year old man presented with a 6 month history of slowly deteriorating vision in both eyes. By the time he sought medical help he had difficulty reading the newspaper and watching television. He was otherwise well except for a chronic productive cough.

He had worked for many years as a teacher in Botswana. He was heterosexual. His only risk factor for HIV transmission was a single encounter with an African woman in the mid 1980s. In 1988 he had dengue fever and malaria. The latter was treated in the UK with quinine and fansidar. In 1995 he was treated in Iwawa for a pneumothorax. He was known to be hypertensive and was taking...
Evoked responses were seen on the ERG. The remainder of the neurological examination was normal.

Full blood count, electrolytes, glucose, B12, folate and folate levels were all normal. Autoantibody screen and syphilis serology were negative. Haemophilus influenzae was grown from his sputum but neither acid fast bacilli nor Pneumocystis carinii were detected. Toxoplasma serology showed no significant titre. Other viral serology including cytomegalovirus (CMV) antibody, hepatitis B surface antigen, and hepatitis C antibody were negative.

A chest radiograph showed bilateral basal shadowing. A high resolution computed tomograph scan confirmed bronchiectasis. A magnetic resonance imaging scan of head and orbits was normal. His CSF contained 28 white cells all of which were lymphocytes. The CSF protein was 0.73 g/l (normal <0.4 g/l) and glucose of 2.5 mmol/l (plasma 5.5 mmol/l). No organisms were detected. CSF cryptococcal antigen test (latex agglutination) was negative.

The lymphocyte subsets showed a T helper cell count (CD4) of 100 cells ×10⁶/l (normal 700–1100) and T suppressor cell count of 2332 cells ×10⁶/l (normal 500–900). In view of his history he was counselled and consented to HIV testing. HIV-1 antibody test was positive.

He was commenced on triple therapy zidovudine 200 mg three times daily, lamivudine 150 mg twice daily, indinavir 800 mg three times daily (HAART or highly active antiretroviral therapy) with co-trimoxazole prophylaxis for Pneumocystis carinii. He responded well clinically and on review at 6 months his vision had improved to 6/9 and 6/6 in his right and left eyes respectively with a concomitant improvement in his visual fields at 24 months (Fig 1B). His CD4 count rose to 170 cells ×10⁶/l and HIV viral load (Roche Amplisor) was below 400 copies/ml 6 months after initiating HAART. Unfortunately, the HIV viral load at presentation was not available.

Figure 2 (A) Right and (B) left colour fundus photographs showing bilateral optic disc pallor.

COMMENT

Visual deterioration in HIV is a common problem. Ocular manifestations of HIV include retinal microangiopathy, opportunistic retinal infections, Kaposi’s sarcoma of the conjunctiva and eyelids, and herpes zoster ophthalmicus. Retinal disease occurs frequently, sometimes in association with optic nerve disease. Occasionally the optic nerve is selectively involved. The commonest aetiology causes include toxoplasmosis and tuberculous retinal infarcts.

A chest radiograph showed bilateral basal shadowing. A high resolution computed tomograph scan confirmed bronchiectasis. A magnetic resonance imaging scan of head and orbits was normal. His CSF contained 28 white cells all of which were lymphocytes. The CSF protein was 0.73 g/l (normal <0.4 g/l) and glucose of 2.5 mmol/l (plasma 5.5 mmol/l). No organisms were detected. CSF cryptococcal antigen test (latex agglutination) was negative.

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In our case funduscopry excluded CMV and toxoplasma retinitis. The other infective aetiologies were excluded by serological testing. Magnetic resonance imaging excluded a compressive lesion. It is possible that our patient had idiopathic bilateral optic neuropathy with spontaneous improvement. However, it is exceedingly rare for simultaneous, rather than stepwise, bilateral visual deterioration to occur with progressive deterioration for months and then a slow delayed recovery. One of the best series of bilateral optic neuropathy remains that by Hierons and Lyle in 1959. They reported 47 cases of bilateral optic neuritis, seven of which developed simultaneous bilateral visual disturbance. Two of these seven patients regained their vision over 6–12 months. Therefore, given the rarity of such an idiopathic presentation and the temporal relation of the improvement to HAART it is very likely that our patient had a primary HIV related optic neuropathy.

Primary HIV infection is well known to cause neurological disease. However, there has only been one case report of bilateral retrobulbar optic neuropathy in whom the presumed aetiology was the HIV virus itself. Newman and Lessell reported two patients who were already known to be HIV positive. The first patient was a 39 year old man who awoke with bilateral blurred vision and slight retrobulbar pain. His vision continued to deteriorate to 5/200 right eye and hand movement perception in the left eye. Funduscopry was normal. Four weeks into his illness he was commenced on AZT. Within 10 days his vision was beginning to improve. Eight months later his acuity was 20/70 right eye and 20/40 left eye. He was left with bilateral optic atrophy. The second patient did not improve with AZT but did show improvement with prednisolone. The only other case of optic neuropathy due to primary HIV infection was of a man with sudden monocular blindness. This was clinically due to an anterior ischaemic optic neuropathy, presumed to be secondary to an optic nerve microangiopathy.

What is the mechanism of the optic neuropathy? There is a significant loss of cortical neurons and optic nerve axons in patients with AIDS. Also far fewer AIDS patients have ocular signs than have ocular lesions discovered on post mortem. Therefore, it is not surprising that changes were noted in the optic nerves of eight AIDS patients who did not have visual signs or symptoms before death as well as those who did. Patchy axonal degeneration, oligodendrocyte, and myelin degeneration were noted in association with mononuclear cell infiltration, suggesting that optic nerve degeneration may be mediated by HIV infected macrophages. The HIV virus infects mononuclear phagocytic cells rather than neurons or oligodendrocytes so the neuronal losses must occur through a secondary mechanism. A more recent study has shown that optic nerve astrocytes in four patients with primary HIV related optic neuropathy stain strongly for tumour necrosis factor α (TNFα). No comparable staining was seen in control optic nerves. TNFα, an immunomodulatory agent, thus appears to be a major player in HIV induced neuronal apoptosis.

In conclusion, HIV may directly cause an optic neuropathy. The improvement of our patient’s vision with HAART is suggestive that much of the optic nerve failure is due to a reversible dysfunction of the optic neurons rather than their death.

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