found to be CD2, CD3, CD4, CD25, and HLA-DR positive but CD8 negative. It was revealed, with the particle agglutination method, that the serum titre for HTLV-I was 25 600 times dilution on repeated tests.

Also, physical examination showed hepatomegaly and hypercalcemia. Based on these data, the diagnosis of ATL was established. He was admitted to the department of internal medicine of Yokohama City University Hospital and treated with chemotherapy. Initially he appeared to respond well to the therapy, but gradually showed severe complications such as opportunistic respiratory infection and acute renal failure. Hypoxaemia and hypercalcemia progressively worsened in spite of intensive therapy. On 22 November 1991, he died in the hospital, and necropsy was carried out.

Histopathological examination of the eyes revealed the slight infiltration of mononuclear cells in the episclera, but ATL cells were not found in the eyes (Fig 3). Interestingly enough, on viral examination the pX region of HTLV-I proviral genome was detected from the episclera and the sclera with polymerase chain reaction (Fig 4).

Comment

Adult T cell leukaemia is a haematological malignancy of high mortality with an increase of atypical lymphocytes characterised by multiple irregularly lobulated nuclei. Its major ocular manifestations are opportunistic infectious lesions and infiltrative lesions by leukaemic cells.1–5 There has been only one report of episcleritis associated with ATL,6 but there has been no report of ATL that began with recurrent episcleritis. It has been reported that HTLV-I infects fibroblasts or endothelial cells and transforms these membrane antigens. In this case episcleritis recurred many times before ATL developed, therefore it seems likely that this episcleritis may be related to an immunomediated reaction to HTLV-I infected cells. Further immunological and molecular biological studies are needed to clarify the exact mechanism of HTLV-I associated ocular diseases. Recognition by ophthalmologists of the various ocular inflammatory lesions is important in diagnosing ATL.


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Development of retinal vascular leakage and cystoid macular oedema secondary to central serous chorioretinopathy

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We present what we believe to be the first case of central serous chorioretinopathy to be diagnosed, documented angiographically, and followed and which then developed retinal vascular leakage and cystoid macular oedema. We have documented this development with serial fundus photography and fluorescein angiography.

Case report

A 43-year-old man had reduced vision in the right eye for 2 months before being evaluated. On examination, his vision measured 20/30 right eye and 20/15 left eye. The patient had a sensory retinal detachment of the right macula. There was a thin layer of fibrin under the retina just nasal to the fovea, and a small retina pigment epithelial (RPE) detachment below that (Fig 1A). There was no sign of intraocular inflammation, nor any sign of subretinal neovascularisation. The left macula showed a few RPE changes. A fluorescein angiogram confirmed these findings and showed that there was an RPE leak into the subretinal space from the shallow RPE detachment in the nasal fovea (Figs 1B, 1C). A careful stereo examination of the retinal vessels of both fundus photographs and fluorescein angiograms confirmed that the retinal vessels and capillaries were entirely normal and there was no cystoid macular oedema. A diagnosis of central serous chorioretinopathy was made and the patient was followed.

The patient returned several months later; the vision was 20/50 right eye. Examination showed that cystoid macular oedema had developed in the right macula. The subretinal fibrin was still present. Fluorescein angiography demonstrated the presence of leaking retinal vessels and con-
Development of retinal vascular leakage and cystoid macular oedema secondary to central serous chorioretinopathy

Figure 1A  Right macula, red free photography. Note the turbid sensory retinal detachment of the macula. There is also a layer of fibrin (pale round area with dark spot in centre) under the retina.

Figure 1B  Right macula, early phase of the fluorescein angiogram. Note the small RPE detachment inferonasal to the macula. Faint hyperfluorescence just nasal to the central fovea was a shallow RPE detachment inferonasal to the macula. Faint hyperfluorescence just nasal to the central fovea was a shallow RPE detachment that had a small focal leaking spot.

Figure 1C  Right macula, later arteriovenous phase of the fluorescein angiogram. The small RPE detachment inferonasal to the macula remains well demarcated and hyperfluorescent. The RPE leakage just nasal to the fovea shows fuzzy hyperfluorescence indicating fluorescein leakage under the sensory retina.

Figure 2A  Right macula, red free photograph. Note the pale appearance of the central macula. The tiny dark dots that are seen in this area are small cystic spaces in the retina.

Figure 2B  Right macula, early arteriovenous phase of the fluorescein angiogram. Note the retinal capillaries which are beginning to leak (giving the false appearance of dilatation and telangiectasis). The small RPE detachment inferonasal to the fovea appears the same.

Figure 2C  Right macula, late phase of the cystic macular oedema from the leaking retinal capillaries. The RPE detachment inferonasal to the fovea remains the same.

firmed cystoid macular oedema. Because of the intense leakage into the retinal vasculature, it was not possible to discern the presence of an RPE leak.

Three months later, the vision was still 20/50, but the sensory retinal detachment was slightly greater, and the cystoid macular oedema appeared more pronounced. A repeat angiogram confirmed the retinal vascular leakage and marked cystoid macular oedema (Figs 2A–C). The left macula had remained the same.

Argon laser photocoagulation was directed to
the leaking retinal vessels around the right fovea in a very light, minimal manner. Over the course of the next year, the sensory retinal detachment flattened, the retinal vessels stopped leaking, and the cystoid oedema resorbed. Twelve years later, the patient had 20/15 vision in the right eye. There were small RPE laser scars, but the sensory retina was flat and there was no retinal vascular leakage.

Comment
We carefully re-examined all fundus photographs and fluorescein angiograms of this patient. At no time was there any evidence of subretinal neovascularisation. Although the fluorescein angiogram appeared to show retinal vascular telangiectatic changes, this was only in appearance (false impression), caused by the rapidly leaking fluorescein through the capillaries. There were no retinal vascular telangiectatic channels or dilated retinal vessels seen on slit-lamp contact lens examination, stereo fundus photography, or red free photography. The fine, tiny, dark spots seen in the macula were small cystic spaces.

Yannuzzi et al.1 reported that retinal telangiectases and cystoid macular oedema were associated with the sensory retinal detachment of central serous chorioretinopathy. They did not show, however, that retinal telangiectases (and cystoid macular oedema) developed after central serous chorioretinopathy had occurred and had been diagnosed. They noted the retinal telangiectases only apparent on fluorescein angiography; no telangiectatic vessels were seen on slit-lamp biomicroscopic examination.

Three mechanisms are possible causes of the breakdown of the inner blood-retinal barrier that lead to the development of retinal vascular leakage in our case: (1) subretinal fluid damages the outer retinal layers causing toxic breakdown products in the retina that result in retinal vascular leakage; (2) the subretinal fluid is directly toxic to the retinal capillaries; (3) the detached retina is hypoxic, and this leads to retinal vascular leakage in a compensatory effect to provide more oxygen to the outer retinal layers. The fibrin that appeared attached to the detached photoreceptors might also have led to hypoxia of the retina.

Malbran and colleagues1 reviewed 3260 retinal detachments and found a 10-7% incidence of retinal vascular changes in retinas that had been detached for 6 months or more. This supports the idea that prolonged separation of the sensory retina from the RPE could cause the subsequent development of retinal vascular changes, perhaps by prolonged hypoxia and deprivation of nutrients to the detached retina.

In our patient the separation of the outer retina from the RPE may have caused retinal hypoxia of the outer retinal layers by separating the outer retina from its normal oxygen supply, the chorioid.14

The subretinal fluid may be directly toxic to the outer retinal layers apart from any hypoxic effect. The breakdown of the photoreceptors and their toxic byproducts may alter the retinal capillaries.

The type of subretinal fluid does not appear to be significant, because fluid of chorioidal origin, as in central serous chorioretinopathy,1 or vitreous fluid, as in rhegmatogenous retinal detachment,1 both appear to be associated with retinal vascular changes. The common factor is detachment. This tends to support the theory that direct hypoxia to the outer retinal layers causes their breakdown, since the two different subretinal fluids produce the same effect.

It may be that the subretinal fluid is directly toxic to the retinal capillaries, and any leakage of fluid causes retinal vascular leakage. Experimental studies have shown that large molecules like horseradish peroxidase can diffuse easily through all layers of the retina.1

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