Recurrent episcleritis associated with adult T cell leukaemia

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Adult T cell leukaemia (ATL) is an aggressive adult leukaemia caused by human T lymphotropic virus type 1 (HTLV-I). It shows various clinical features such as lymphadenopathy, hepatosplenomegaly, skin eruptions, and ocular lesions. Among ocular lesions we can often find direct infiltrations of ATL cells into the conjunctiva and opportunistic infectious lesions. In this paper we present an unusual finding, episcleritis, and propose the relation between episcleritis and HTLV-I.

Case report
A 52-year-old man was referred to Yokohama City University Hospital in January 1991 because of a recurrent ocular hyperaemia of both eyes. He originally came from Kumamoto prefecture located in south west Japan, where HTLV-I infection is highly endemic. He had undergone appendectomy in 1956, but he had had no other disease.

On initial ophthalmic examination, his corrected visual acuity was 1.2 in the right eye and 1.0 in the left eye. Applanation tonometry showed 20 mmHg in the right eye and 28 mmHg in the left eye. On both eyes the bulbar conjunctivae showed intense congestion and the anterior chambers showed 1+ cells (Fig 1). On fundoscopy the retinas appeared normal. The routine blood film was normal and the biochemistry, in particular urea and uric acid, was normal. Physical examination showed no lymphadenopathy, hepatosplenomegaly, or skin eruption.

The patient was treated with topical corticosteroids and β blocking agents. Inflammation of the anterior chambers and the ocular hyperaemia were controlled initially with topical treatment, but the ocular hyperaemia of both eyes recurred many times as the therapy was tapered. In August 1991, he complained of fatigue and blood examination showed a white blood cell count of 13 × 10^9/l with 22.5% atypical lymphocytes (Fig 2). These lymphocytes were

Figure 1 Photograph of the right eye showing intense hyperaemia with a moderate amount of serous secretion.

Figure 2 Atypical lymphocytes were found in the peripheral blood that are characterized by multiple lobulated nuclei, named ATL cells or flower cells (Giemsa stained, magnified ×530).

Figure 3 Haematoxylin and eosin stain of a section of the episclera showing the slight infiltration of mononuclear cells around the blood vessels, but there were no ATL cells in the eyes (magnified ×210).

Figure 4 The pX region of HTLV-I proviral genome was detected from the episclera and the sclera with polymerase chain reaction (PCR). As his death the white blood cell count had dropped under 1 × 10^9/l with no ATL cells because of the severe chemotherapy, but the PCR band was as dense as the positive control. Therefore it is impossible that the PCR product was from ATL cells in the peripheral blood. This indicates the focal infection of HTLV-I in the episclera and the sclera.

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Accepted for publication 31 May 1993
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 hemoptysis 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.1-3 There has been only one report of episceritis associated with ATL, but there has been no report of ATL that began with recurrent episceritis. It has been reported that HTLV-I infects fibroblasts or endothelial cells and transforms these membrane antigens.1 In this case episceritis recurred many times before ATL developed, therefore it seems likely that this episceritis may be related to an immunologically 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.