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A Case of Uveal, Palpebral, and Orbital Invasions in Adult T-Cell Leukemia

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Background: Patients with adult T-cell leukemia (ATL) may have eyelid lymphoma, uveitis, or cytomegalovirus retinitis due to being immunocompromised. However, there have been few reports on the invasion of multiple ocular lesions. We treated 1 unusual ATL patient with uveitis in whom multiple ocular invasions were suspected.

Case: A woman in whom ATL was diagnosed 10 years previously complained of blurred vision and decreased visual acuity in the right eye. Anterior uveitis of the right eye was suspected. One week later the cells increased in the anterior chamber, and fibrin exudates and hyphema appeared. She was admitted to our hospital.

Observations: The visual acuity was 0.04 in the right eye and finger-counting from 30 cm in the left. She was treated with systemic steroid therapy. Inflammation disappeared, but both eyelids became swollen and multiple ocular lesions appeared. She was given carcinostatic therapy once more and the mass lesions decreased. Mass lesions appeared in the iris and in the bulbar conjunctiva. Computed tomography and magnetic resonance imagining (MRI) showed that the mass lesions extended to the right orbit and both nasal cavities. MRI also demonstrated choroidal thickening in the left eye.

Conclusion: This case documents that ATL cells may cause severe uveitis and invade multiple ocular tissues such as the iris, eyelid, choroids, and orbit. **Jpn J Ophthalmol 2003;47:599–602**

Key Words: Adult T-cell leukemia, multiple ocular lesions, tumor formation, uveal invasion, uveitis.

Introduction

Adult T-cell leukemia (ATL) is caused by the human T-lymphotropic virus type 1 (HTLV-1). Patients with ATL may have eyelid lymphoma, opportunistic uveal and retinal infections, and direct ATL cell infiltration.[1–11] To our knowledge, there have been no reports of multiple ocular invasions. We treated 1 unusual ATL patient with uveitis in whom invasion of multiple ocular lesions was suspected.

Case Report

In September 1990, a 33-year-old Japanese woman noted the swelling of her right cervical lymph node. One year later, a tumor was found in the left ventricle, and she underwent heart surgery at Nagasaki University Hospital. A tumor biopsy and Southern blot analysis led to the diagnosis of ATL (chronic type). She was treated with vincristin, cyclophosphamide, adriamycin, and prednisone the first week; adriamycin, cymerin, and prednisone the second week; and vindesine, etoposide, and carboplatin the third week. Thereafter, she had a constant fever every year in winter. A physician followed her at Nagasaki University Hospital and the ATL became chronic.

Ten years after the onset of ATL, she complained of blurred vision in the right eye and visited a local eye

Received: May 2, 2002
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Published by Elsevier Science Inc.

0021-5155/03/$–see front matter
doi:10.1016/j.jjo.2003.08.008

Jpn J Ophthalmol
Vol 47: 599–602, 2003

doctor. The visual acuity was 0.5 in the right eye and finger-counting from 30 cm in the left. The ocular pressure was 15 mm Hg in both eyes. Due to a childhood infection, the patient had macular corneal opacities in the left cornea. A few cells were found in the anterior chamber of the right eye, but the fundi of both eyes looked normal. Anterior uveitis was suspected and she was treated with a topical steroid, Rinderon A (β-methozone + frasiomycin), three times a day. However, 1 week later the cells were very numerous in the anterior chamber of her right eye, and fibrin membrane and hyphema appeared. When the right fundus became invisible, in September 2000, she was admitted to our hospital. The visual acuity in her right eye was 0.04, and in the left, it was only finger-counting from 30 cm. The ocular pressure was 15 mm Hg in both eyes. In the right eye, the cornea was edematous, and in the left, opaque. In the right anterior chamber, there were cells, fibrin, and hemorrhages, and the right fundus was invisible. Fluorescein angiography was unclear because of anterior chamber opacity due to inflammation in the right eye and corneal opacity in the left. The red blood cell count was 4,580,000/mm^3 and the white blood cell count was 16,200/mm^3, with 62% abnormal lymphocytes. She had lymph node swelling in the neck, axilla, inguinal region, hands, feet, and chest.

Because her general condition was stable, we considered that eye treatment was necessary first. She was treated with systemic steroid at an initial dose of 60 mg with a gradual decrease, for a total of 730 mg of prednisolone. The cells, fibrin, and hemorrhages in the anterior chamber decreased and the right fundus became visible and looked normal. However, on the 15th day of hospitalization, both eyelids began to swell (Figure 1), and mass lesions appeared on the right iris (Figure 2).

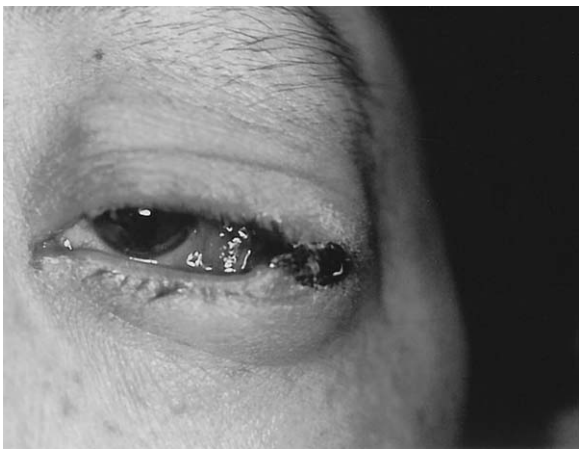

Figure 1. Front view showing proptosis and protrusion of the left eye and hemorrhage at the temporal side of the conjunctiva in a patient found to have adult T-cell leukemia.

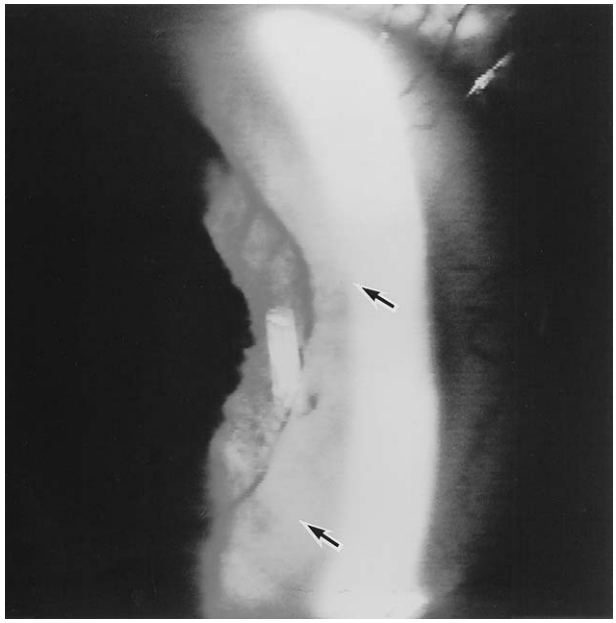

Figure 2. The arrow indicates temporal protrusion in the iris of the left eye.

The mass lesions increased daily. Computed tomography (CT) and magnetic resonance imaging (MRI) showed masses extending to the orbits and the nasal cavities, right parapharyngeal space, and both parotid spaces. The MRI showed left choroidal thickening (Figure 3). There were tumor invasions in the liver and kidneys. Skin tumors of the extremities and axillary lesions increased day by day.

We consulted with surgeons, who biopsied an axillary tumor. The conjunctiva was not biopsied because of her poor general condition. Biopsy specimens showed diffuse lymphoma, pleomorphic type ATL (Figure 4). She was transferred to the Department of Internal Medicine at Nagasaki University Hospital for carcinostatic therapy. She was treated with etoposide 140 mg, vindesine 3.36mg, carboplastin 350 mg for 1 week and then had a 1-week intermission. This treatment was administered three times.

After this therapy, visual acuity in her right eye improved to 0.8 and eyelid swelling decreased. The mass in the iris also decreased. Visual acuity in her left eye was unchanged. CT scan showed that the mass lesions in the orbits, nasal cavities, liver, and kidneys had also decreased. As she was transferred to another hospital, we have not been able to follow her ocular and general condition since the initial therapy.

Discussion

Lymphocytes infected with HTLV-1 flow through the peripheral circulation and infiltrate many organs such

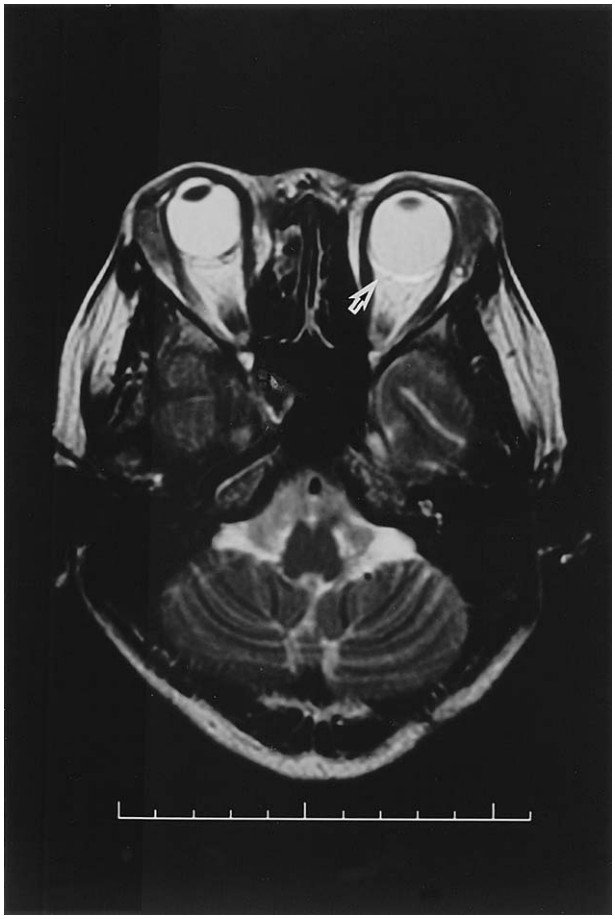

Figure 3. T_1-weighted magnetic resonance image showing extraconal mass in both orbits. These masses extend to the nasal cavity, right parapharyngeal space, and bilateral parotid spaces. The arrow indicates thickening of the choroid in the left eye.

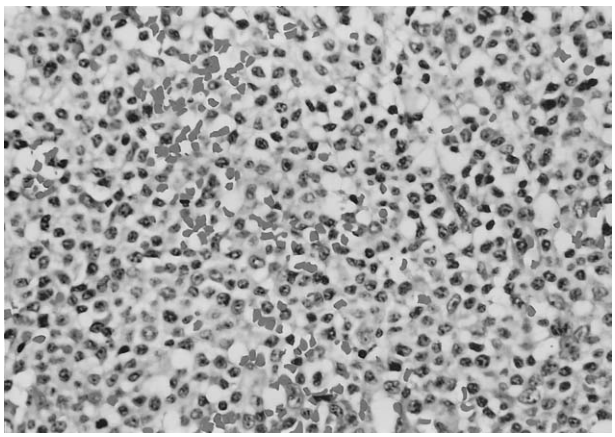

Figure 4. A biopsy of axillary tumor shows medium-to-large atypical pleomorphic lymphoid cells (×100).

as the lungs[12] and the central nervous system.[13] Ocular manifestations of ATL are rare, and most of them involve retinitis or uveitis, which are opportunistic and of the direct infiltration type.[1–7] The uvea may have been invaded because of the abundant blood flow in this patient. However, it is rare for ATL cells to invade the uvea and form a mass.[1–11]

Lauer et al[8] first reported ATL tumor formation in the orbit. Their patient had proptosis of the left eye and demonstrated a large tumor mass on CT. They did a biopsy of the orbital and sinus masses and diagnosed lymphoma, but their patient died in spite of local radiation therapy. Ohba et al[9] reported that 5 of 10 patients with ATL had inflammatory or opportunistic infectious ocular lesions and 1 patient had an eyelid tumor. Kohno et al[10] reported conjunctival tumors in 3 of 17 ATL patients. Ichibe et al[11] reported a case of ATL with a protruding lesion of the ocular fundus that was demonstrated with CT. Our patient had tumors in the eyelid, iris, and orbit complicating anterior uveitis. To our knowledge, there have been no other reports of such multiple ocular tumors.

Diagnostic criteria have been proposed to classify four clinical subtypes of ATL.[14] Our patient had been diagnosed with the chronic type for 10 years, but upon admission to the hospital turned out to have the relatively rare acute type of ATL. The acute type is the most aggressive type of ATL. Thus, in our case, we assume that infected lymphocytes moved more rapidly than usual and invaded multiple ocular lesions.

In conclusion, we treated 1 unusual ATL patient with uveitis and multiple ocular tumors. Carcinostatic therapy was considered to be effective in this case.

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Jpn J Ophthalmol
Vol 47: 599–602, 2003

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