Successfully Treated Optic Nerve Infiltration with Adult T-Cell Lymphoma

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A 45-year-old woman with adult T-cell leukemia had a sudden onset of decreased vision with central scotoma in the right eye. Right optic disc edema and thickening of the optic nerve were observed by magnetic resonance imaging. A diagnosis of leukemic infiltration to the optic nerve was made. A course of oral corticosteroid therapy was prescribed and 40 Gy of radiation was administered to the whole brain and right orbit. Her visual acuity recovered from finger counting to 20/20 within 3 weeks. We believe that this patient represents the first report of successfully treated optic nerve infiltration with adult T-cell leukemia caused by human T-lymphotropic virus type-I infection.

Key Words: Adult T-cell lymphoma—Virus—Optic nerve.

Human T-lymphotropic virus type-I causes adult T-cell leukemia (1). Few Japanese reports have described optic nerve involvement in this disease (2), and, to our knowledge, no case has been successfully treated. We treated a woman who had optic nerve infiltration in adult T-cell leukemia with oral corticosteroid therapy and irradiation to the optic nerve and whole brain.

CASE REPORT

A 45-year-old woman who complained of easy fatigability was found to have leukocytosis in 1990. A small number of atypical lymphocytes were detected in the peripheral blood. Increased titers to human T-lymphotropic virus type I were found in the serum (1:128) and in the cerebrospinal fluid (1:2). In 1991, the patient developed a gait disturbance. A radiograph of the patient's femur showed osteolytic lesions. A chest radiograph disclosed disseminated shadows. Splenomegaly was found by computed tomography. Adult T-cell lymphoma infiltration in the spinal cord was noted by magnetic resonance imaging.

The patient was hospitalized and treated with two courses of cyclophosphamide, hydroxydaunomycin, oncovin, and prednisolone therapy. Gait disturbance improved thereafter. In late April 1992, she noticed slight ocular pain in the right eye. Sudden loss of vision with ocular pain in the right eye occurred on May 28. On June 3, she was referred to the ophthalmology service. Corrected visual acuity was finger counting in the right eye and 20/20 in the left eye. She showed right afferent pupillary defect. Goldmann perimetry revealed dense central scotoma in the right eye. The right optic disc was swollen (Fig. 1). Sheathing around retinal veins and vitreous floaters close to the peripheral retina lesion were noted in both eyes.

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A tentative diagnosis of optic nerve infiltration with lymphoma cells was made. Oral prednisolone, 40 mg/day, was initiated on that day. Magnetic resonance imaging disclosed thickening of the right optic nerve on June 5 (Fig. 2). Her visual acuity was 20/400 on June 10, and it continued to improve. On June 14, 40 Gy of irradiation to the right orbit and whole brain were initiated. Corrected visual acuity returned to 20/20 in 3 weeks. Visual fields were almost full. She had clear media, but retinal vasculitis was only slightly improved. Fluorescein angiography disclosed hyperfluorescent leakage around the retinal vasculitis lesion, but the optic nerve was normal (Fig. 3). The patient has maintained good visual function after more than 13 months’ follow-up.

DISCUSSION

We treated a patient with human T-lymphotropic virus type I related adult T-cell leukemia. At the onset of the disease, raised titers to human T-lymphotropic virus type I were positive in serum and in cerebrospinal fluid. Atypical lymphocytes in the peripheral blood were diagnostic of adult T-cell leukemia (1). An infiltration of leukemic cells to the spinal cord rather than virus-associated myelopathy was suspected from the magnetic resonance imaging (3). The patient’s condition was improved by two courses of corticosteroid therapy, nevertheless leukemic infiltration of the central nervous system involved the optic nerve and caused acute visual loss in the right eye.

Reported ocular manifestations in patients with adult T-cell leukemia include retinal vasculitis (4-7), anterior uveitis (5-8), orbital tumor (9), and optic neuropathy (2). To our knowledge, no detailed report of optic nerve infiltration with human T-lymphotropic virus type I has been made.

We believe that the loss of visual acuity in our patient was due to optic nerve infiltration with adult T-cell leukemia. Disc edema was observed, and optic nerve thickening was shown by magnetic resonance imaging. Past history of spinal cord infiltration suggests optic nerve infiltration of adult T-cell leukemia cells, rather than optic neuropathy associated with this type of leukemia. Intracranial pressure appeared normal. Although ocular pain preceding the visual loss is common in optic neuropathy, it also has been reported in optic nerve infiltration with leukemia by some authors (10-14). High affinity of T-lymphocytes to endothelial cells in myelopathy associated with human
T-lymphotropic virus type-I was reported (15). Loging of infected T-cells on the endothelium of vessels may be the initial step not only for virus-related myelopathy but also the occurrence of optic nerve infiltration with adult T-cell leukemia.

Only a few cases have been reported of optic nerve involvement in adult T-cell leukemia (2,16). In general, good clinical results in patients with leukemic involvement of the optic nerve with radiation have been described (13,17,18). To date, good clinical results in optic nerve infiltration with adult T-cell leukemia have not been reported. A combination of rapid corticosteroid administration and optic nerve radiation may be desirable for such cases because the effect of oral steroid therapy may be quick and transient. Also, planning the irradiation takes at least several days before it can actually be initiated.

Our patient also had retinal vasculitis. Funduscopy examination revealed sheathing of retinal vessels and whitish opacities in the vitreous. Fluorescein angiography disclosed fluorescein leakage from retinal lesions. These findings were similar to the retinal changes reported in patients with adult T-cell leukemia (5,6,8) or a uveitis patient seropositive for human T-lymphotropic virus type I (7). These retinal lesions reportedly do not affect visual acuity severely (6). Retinal vasculitis responded little to corticosteroid treatment. Takatori and associates (19) reported spontaneous resolution of vasculitis a long time after the termination of steroid treatment. These authors suspect that retinal vasculitis with adult T-cell leukemia may react only slightly to corticosteroid treatment.

Severe optic nerve involvement occurs with adult T-cell leukemia. Ophthalmologists should be aware of this complication and should initiate steroid therapy immediately. Thereafter, irradiation may be suggested.

REFERENCES


