Primary Intraocular Lymphoma

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Intraocular non-Hodgkin’s lymphoma is a relatively uncommon condition that has been divided into 2 types: (1) systemic lymphoma metastatic to the eye and (2) lymphoma (formerly reticulum cell sarcoma) involving the central nervous system and eye.\(^1\)\(^2\) Intraocular lymphoma alone or ocular involvement of the second type of lymphoma sometimes masquerades as chronic uveitis.\(^1\)\(^2\)

We herein report a case of primary intraocular lymphoma in which vitreous opacities initially were found. We also reviewed the literature for additional reports of Japanese patients who had a diagnosis of primary intraocular lymphoma.

Case Report

At examination on April 14, 1998, a 65-year-old woman complained of blurred vision in her left eye. She had a past medical history of Meniere disease. Her family history was noncontributory.

On examination, her visual acuity was 0.6 in the right eye and 0.5 in the left. Her intraocular pressures were 16 mm Hg, right eye, and 17 mm Hg, left eye. The corneas were clear bilaterally. Nuclear opacities were noted in both lenses. Cellular floaters were found in the anterior chamber and vitreous in the left eye. Ophthalmoscopically, the right fundus appeared normal. A moderately diffuse vitreous opacity was found in the left eye, but the fundus was normal.

Fluorescein angiography showed no leakage from either fundus. Results of the following laboratory studies were negative or within the normal range:

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blood cell count; C-reactive protein; angiotensin-converting enzyme; serum titers for herpes simplex virus, varicella-zoster virus, and Epstein-Barr virus; blood chemistry panel; Treponema pallidum hemagglutinin; antinuclear antibody; serum titer for Toxoplasma gondii; and urinalysis. A chest X-ray also was normal. No oral aphthous or genital ulceration or arthritis was found.

On April 27, 1998, the patient was treated with systemic prednisolone, 40 mg/day for 5 days and 20 mg/day for 5 days, and topical instillation of 0.1% betamethasone four times a day. In early May, intracameral cells and the vitreous opacity were slightly diminished. Thereafter, the dosage of systemic corticosteroid was tapered.

In October 1998, the patient again complained of blurred vision in her left eye. Vitreous opacities had increased in both eyes. No other ocular abnormalities were noted. Systemic prednisolone, 20 mg/day for 7 days, failed to reduce the vitreous opacities.

On January 4, 1999, vitreous opacity further increased in both eyes (Fig 1). On February 8, 1999, vitreous aspiration biopsy was performed in the right eye. Cytologic study of the biopsy specimen showed many large atypical lymphocytes (Fig 2), indicating malignant lymphoma. Anti-CD 20 monoclonal antibody staining was positive on the cells in the vitreous aspirate, demonstrating B-cell lymphoma. Computed tomography and magnetic resonance imaging showed no abnormalities in the brain or orbit. No other systemic lesions were noted on gallium scintigraphy.

On March 25, the patient was begun on a regimen of external beam radiation therapy to both eyes. A total of 40 Gy was delivered in 1.6-Gy fractions. Twenty-five fractions were given over 37 days. One month after the radiotherapy was begun, the vitreous cleared (Fig 3). No recurrence of vitreous opacity or brain abnormality developed during the 5-month follow-up period.

Discussion

Primary intraocular lymphoma is a relatively uncommon condition. We reviewed articles published from 1986 to 1998 for reports of Japanese cases of primary intraocular lymphoma verified cytologically or histopathologically (Table 1).3-16 A total of 19 patients (6 men and 13 women), including our present case, were identified. Their ages ranged from 38 to 84 years, similar to those of patients described in the United States.17-19 Of the 19 Japanese patients, 7 had unilateral lesions and 12 had bilateral involvement. All 19 patients had vitreous opacities, as also reported by other investigators17-20; 15 of the 19 patients had retinochoroidal lesions, and 12 had central nervous system involvement. The diagnosis of primary intraocular lymphoma was made by enucleation (in 1 case), choroidal biopsy (4 cases), brain biopsy (4 cases), aqueous sampling (2 cases), autopsy (2 cases), vitreous cytology (5 cases), and orbital biopsy (1 case). Radiotherapy to the eyes or brain was performed in 17 patients. During the follow-up period, 14 patients were alive and 5 had died.

Unlike typical uveitis, vitreous opacities in these 19 patients were unresponsive to steroidal treatment. It has been recommended that diagnosis of intraocular lymphoma be based on cytopathologic examination of vitreous cells17-19 and choroidal biopsy.21 Of 19 Japanese patients, 5 underwent vitreous cytologic examination, and 4 had choroidal biopsy.

In our patient, vitreous cytologic findings were able to show B-cell lymphoma as the cause of steroid-resistant vitreous opacities mimicking uveitis. We believe