A 59-year-old man with a history of mycosis fungoides developed loss of visual acuity and visual field in the left eye. Epiretinal lesions were present in the right eye and multifocal choroidal lesions, optic disc edema, and vitritis were present in the left eye. A diagnostic vitrectomy was performed and cytologic examination of the vitreous confirmed the diagnosis of T-cell lymphoproliferative disorder. Systemic and intrathecal chemotherapy resulted in marked improvement in ocular signs and symptoms. At last follow-up, the patient was found to have improved visual acuity in the left eye; however, significant worsening of his systemic condition developed and he died shortly thereafter.

Mycosis fungoides (MF) is a cutaneous T-cell lymphoma characterized by erythematous lesions that progress into reddish-brown infiltrated plaques and tumors and may involve extracutaneous tissues, including lymph nodes, lungs, liver, and spleen.1 Ocular involvement is seen in up to one third of advanced cases, usually involving the eyelids, cornea, conjunctiva, caruncle, sclera, and orbit.2 Intraocular involvement, however, is uncommon3 and choroidal lesions4 and optic nerve involvement are rare.5 The diagnosis of intraocular involvement can be confirmed by flow cytometry of the vitrectomy specimen.6 Mycosis fungoides may also be associated with subcutaneous, visceral, and orbital involvement.7

We describe a patient with intraocular T-cell lymphoproliferative disorder associated with MF involving the vitreous, choroid, and optic nerve. The diagnosis of this unique case was confirmed by vitrectomy and flow cytometry analysis.

REPORT OF A CASE

A 59-year-old man had a 2-month history of painless visual loss in the left eye. He saw an ophthalmologist who noted a swollen optic nerve and began treatment with a 4-day course of intravenous steroids for presumed optic neuritis.

His medical history was significant for skin biopsy–proven mycosis fungoides in 1995 (Figure 1) that was treated with chemotherapy and psoralen–UV-A. Medications included trimetrexate, omeprazole, amoxicillin-clavulanate, and prednisone (100 mg/d and tapering).

Ophthalmologic examination revealed a visual acuity of 20/25 OD and 20/100 OS. The pupils were isocoric and reactive with an afferent pupillary defect in the left eye. Superior and temporal visual field loss was noted in the left eye. Slit-lamp biomicroscopy results were normal. Ophthalmoscopic examination disclosed a clear vitreous and discrete white epiretinal plaques in the inferior periphery of the right eye. A dense vitreous cellular infiltrate was present in the left eye with optic disc swelling and multifocal irregular choroidal lesions (Figure 2). A magnetic resonance scan of the head demonstrated subtle enhancement of the left subcerebellar folia consistent with possible leptomeningeal seeding with lymphoma. Lumbar puncture revealed atypical lymphocytes, confirming central nervous system involvement.

Cytologic examination of the vitrectomy (cell block technique) of the left eye revealed numerous atypical large neoplastic cells (average 12-14 µm), most of which...
showed prominent nucleoli and scanty cytoplasm (Figure 3). Immunohistochemical staining for CD3 (pan-T-cell marker) was positive in 65% to 70% of the cells (Figure 4). Flow cytometry of the vitrectomy specimen showed that all T-cell markers (CD2, CD3, and CD5) as well as CD25 (interleukin 2 receptor) were above 90%. The lack of CD4 positivity (only 1%) precluded an immunotypic diagnosis of MF. Levels of CD8 were also markedly decreased (21%).

The patient underwent placement of an Ommaya reservoir, intrathecal chemotherapy with methotrexate, and systemic chemotherapy with arabinosylcytosine and methotrexate. Three weeks after initiation of chemotherapy, the left eye showed significant resolution of the choroidal infiltrates and disc swelling, and the visual acuity had improved to 20/60 OS.

Two months after initiation of chemotherapy, the patient developed high fevers and neutropenia and was admitted to the hospital with lower extremity weakness and urinary retention. He subsequently developed paraplegia, loss of bladder and bowel control, and severe bilateral sensorineural hearing loss. He was discharged approximately 1 month later to a skilled nursing facility for palliative care. The patient died shortly thereafter.

We believe that both conditions in this patient must have been closely related, since the vitrectomy specimen contained numerous large malignant lymphoma cells expressing T-cell markers by immunohistochemistry and flow cytometry and MF is typically a cutaneous T-cell disorder.

COMMENT

Although lymphoma is known to affect the eye and ocular adnexae, the vast majority of cases are of the B-cell type. Intraocular involvement by T-cell lymphoma (including MF) is rare. Stenson and Ramsay described 30 patients with MF and 11 were found to have ocular abnormalities. Of these 11 cases, 3 had optic atrophy (27%) and 1 had uveitis (9%). In their review of the literature, only 4 reports of retinal pigment epithelium or retinal involvement were noted. Choroidal involvement of MF has been reported previously only once. Our patient had bilateral intraocular involvement with epiretinal, optic nerve, vitreous, and choroidal lesions. The vitreous biopsy specimen from the patient described herein showed cell surface markers that were consistent with a T-cell lymphoproliferative disorder.

Mycosis fungoides and Sézary syndrome cells are CD4 positive and also express CD2, CD3, and CD5. The lack of expression of CD4-positive cells militates against the diagnosis of MF. In our case, CD4 was virtually absent (1%), and CD8 was markedly reduced (21%). We prefer the term T-cell lymphoproliferative disorder, since the results of flow cytometry of the vitrectomy specimen are not those commonly observed in other cases of MF.

The course of intraocular lymphoma may mimic infectious endophthalmitis (eg, fungal) and therefore a correct diagnosis is essential.
Confirmation of intraocular tumor by vitreous biopsy is indicative of extracutaneous involvement and necessitates aggressive systemic treatment. The role of radiotherapy for retinal and choroidal lesions is unclear. Intrathecal and systemic chemotherapy without radiation therapy in our patient resulted in improvement in ocular signs and symptoms. Although uncommon, T-cell lymphoma (as opposed to B-cell lymphoma) may have intraocular involvement.

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