macular edema, macular pucker, diop-
lopia, and cyclotropia. Full visual re-
habilitation after MT360 also re-
quires silicone oil removal and
strabismus surgery.

Because of photoreceptor and
retinal pigment epithelial degenera-
tion, retinal translocation would not
be expected to improve visual func-
tion in long-standing cases of GA. In
contrast, 50% of eyes with early GA
and good vision have reduced con-
trast sensitivity and reading rates sec-
dondary to parafoveal scotomata. Eyes
with GA and recent visual loss, or
reduced visual function with good cen-
tral vision, may benefit from MT360
before severe foveal photoreceptor de-
generation occurs. However, the po-
tential for visual recovery must be
weighed against the sight-threaten-
ing complications associated with this
surgery. A case-control study may be
warranted to investigate the treat-
ment of these selected cases of end-
stage nonneovascular AMD with
MT360.

Corresponding author: Cynthia A.
Toth, MD, Duke University Eye Cen-
ter, Room 107, Erwin Road, PO Box
3802, Durham, NC 27710 (e-mail:
info@dukeeye.org).

Mark T. Cahill, FRCOphth
Sharon F. Freedman, MD
Cynthia A. Toth, MD
Durham, NC

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* Presumed Teratoma-
Associated Paraneoplastic
Retinopathy

Paraneoplastic retinopathies are char-
acterized by retinal dysfunction ow-
ing to the remote effects of systemic
malignancy. The mechanism of reti-
nal degeneration in these syn-
dromes is believed to be related to the
presence of autoantibodies against tu-
mor-related antigens that cross-
react with molecularly similar reti-
nal antigens. The two most frequently
described paraneoplastic retinopa-
thies are cancer-associated retino-
pathy and melanoma-associated
retinopathy (MAR), each of which has
well-described clinical and electrophysiologic features. Cancer-
associated retinopathy is most com-
monly associated with small-cell lung
carcinoma but has also been associ-
ated with breast, cervical, and uter-
ine cancer. Recoverin, one of the an-
tigens implicated in cancer-associated
retinopathy, has been detected in a pa-
tient with autoimmune retinopathy in
the absence of demonstrable malig-
nancy. We report a case of pre-
sumed paraneoplastic retinopathy as-
sociated clinicopathologically with a
mature teratoma.

Report of a Case. A 39-year-old Gambia-
woman was referred for evalua-
tion of bilateral vitritis. Three
months before her initial visit, she
noted the sudden onset of blurred
vision, nyctalopia, shimmering pho-
topsias, and a migrainous head ache.
Her ocular symptoms gradually
worsened during the following 3
months, despite resolution of head-
ache symptoms. Her medical history was remarkable only for childhood malaria treated with quinine. Her ophthalmic history was unremarkable.

Ophthalmic evaluation documented visual acuity of 20/16 OD and 20/25 OS with intraocular pressures of 15 mm Hg OD and 13 mm Hg OS. Results of anterior segment biomicroscopy were normal in both eyes. Funduscopic examination revealed 1+ vitreous cells in the right eye and 2+ vitreous cells in the left eye, with no vitreous haze in either eye. The optic nerve cup-disc ratio was 0.9 OU without pallor or edema (Figure 1A and B). The maculae were normal. Mild arteriolar attenuation and peripheral vascular sheathing were present in both eyes. Distinct areas of peripheral retinal pigment epithelial depigmentation were present bi-

Figure 1. Photographs illustrate optic nerve cupping and retinovascular attenuation in the right eye (A) and left eye (B). C, The demarcation line is between normal-appearing retinal pigment epithelium in the posterior pole of the right eye and hypopigmented retinal pigment epithelium in the periphery.

Figure 2. Full-threshold Humphrey 30-2 visual fields, stimulus size V in the right (A) and left (B) eyes, demonstrate profoundly constricted visual fields recorded at the initial visit. There were no fixation losses or false-positive errors in either study (false-negative errors, 5/19 in the right eye and 4/11 in the left eye).

lateral (Figure 1C). Results of a comprehensive laboratory evaluation, including serum chemistry, liver functions, complete blood cell count, syphilis serologic testing, and angiotensin-converting enzyme levels, were unremarkable. Humphrey visual fields were profoundly constricted bilaterally (Figure 2). In electroretinograms (ERGs) recorded according to the international standard technique, combined (rod-and-cone-mediated) maximal responses had an “electro-negative” configuration (Figure 3A). Cone-mediated ERGs elicited by 100-millisecond stimuli showed absence of the “on” response with relative preservation of the “off” response (Figure 3B).

The characteristic ERG and clinical history raised the possibility of MAR. Results of comprehensive dermatologic and systemic metastatic evaluations were negative, except for a computed tomographic scan of the chest, which revealed a cystic anterior mediastinal mass consistent with teratoma (Figure 4). Two fine-needle aspirations of the mass were performed and were nondiagnostic. Serum analyses for α-fetoprotein and β-human chorionic gonadotropin, tumor markers for teratoma, were negative. During the following 6 months, the patient received 3 periocular injections of 40 mg of triamcinolone acetonide (Kenalog; Bristol-Myers Squibb, New York, NY) in both eyes. There was a subsequent decrease in the vitreous cell count and documented improvement in the visual fields bilaterally (Figure 5). During this time, serial computed tomographic scans demonstrated progressive growth of the mediastinal lesion, threatening the great vessels of the chest. Median sternotomy and excisional resection of the tumor was subsequently performed.

Gross inspection of the tumor revealed a multicystic mass with hair projecting into the central cavity. Histopathologic analysis confirmed the diagnosis of a mature teratoma (Figure 6A). Staining of the tumor with HMB45, an immunoreagent specific for melanocytic elements, was negative (Figure 6B). In contrast, immunostaining with antibodies against retinal S antigen produced avid staining of multiple tis-
sues within the teratoma specimen, demonstrating tumor expression of retinal 5 antigen (Figure 6C). The patient’s serum was also autoreactive against normal rhesus monkey retina (Figure 6D). The patient’s vision and visual fields remained stable for 3 months after surgery without any further treatment.

Comment. Teratomas are part of the larger family of dysgerminomas or germ cell tumors. Teratomas are composed of primitive cells that represent more than one germ layer and usually all 3. As the teratoma grows, these cells can differentiate along various germ lines, producing skin appendages, cartilage, bone, thyroid tissue, and any other tissue in the body. Teratomas usually originate in the hypothalamus or gonad but uncommonly arise in ectopic locations, including the orbit and mediastinum.

This is the first report of paraneoplastic retinopathy associated with a mature teratoma. A previous report of retinal periphlebitis mimicking sarcoidosis in a child with pineal germinoma proposed a paraneoplastic mechanism. Electronegative ERGs with abnormal on-responses are typically described in MAR and congenital stationary night blindness. The shimmering photopias, nyctalopia, visual field constriction, and clinical appearance of the patient all were typical of MAR; however, metastatic workup failed to reveal evidence of melanoma or other occult malignancy, except for the mediastinal teratoma. Malignant melanomas may arise de novo in teratomas. However, histopathological and immunochemical analyses did not demonstrate the presence of melanocytic elements. Expression of
retinal S antigen in the teratoma and staining of the normal primate photoreceptors with the patient's serum support an association between the teratoma and the patient's vision loss in this case of presumed paraneoplastic retinopathy.

Although similar clinically and electrophysiologically to MAR, this case differs from the typical MAR presentation in a few notable ways. This patient had bilateral vitritis as an initial sign, which has been reported in MAR; however, her objective and subjective visual field improvement with periocular corticosteroids are distinctly unusual for MAR. In addition, in MAR antiretinal antibodies are typically localized to bipolar cells, explaining the typical electronegative ERG. The most avid immunostaining in this case was in the photoreceptor layer, consistent with the presumed pathophysiologic mechanism of an immune response directed against retinal S antigen, which is localized to photoreceptor outer segments. However, the electrophysiologic data are contradictory, suggesting that the site of the major functional defect is proximal to the photoreceptor inner segments, as in MAR. Immunostaining was also present in the inner nuclear layer, albeit less conspicuously than in the photoreceptors. The peripheral retinal pigment epithelial depigmentation and visual field changes that occurred bilaterally may represent sequelae of autoimmune phenomena occurring in the overlying retina.

We described a case of presumed paraneoplastic teratoma-associated retinopathy. This case suggests that benign tumors such as teratomas possibly may be associated with paraneoplastic retinopathy, which has previously been reported only as a sequelae of malignant neoplasms. Additional case reports and/or clinicopathologic investigations may assist in determining the mechanisms of visual loss in such cases.

Eric B. Suhler, MD
Chi-Chao Chan, MD
Rafael C. Caruso, MD
David S. Schrump, MD
Bethesda, Md
Charles Thirkill, PhD
Davis, Calif
Janine A. Smith, MD
Robert B. Nussenblatt, MD
Ronald R. Buggage, MD
Bethesda

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Corresponding author: Eric B. Suhler, MD. Ophthalmology Service, Portland Veterans’ Administration Medical Center, 3710 SW US Veterans’ Hospital Rd, Portland, OR 97207 (e-mail: eric.suhler@med.va.gov).

Isolated Anterior Uveitis as the Initial Sign of Systemic Candidemia

Endogenous fungal endophthalmitis may occur in both healthy and immunocompromised patients. Commonly, such patients have choroidetinal infiltration and a variable degree of posterior uveitis at the initial examination. Treatment is based on the severity of the disease and the response to treatment. We report a case of isolated iridocyclitis with a hypopyon secondary to Candida albicans infection. The atypical manifestation, diagnostic procedures, treatment, and outcome of this case are discussed.

Report of a Case. A 14-year-old girl who underwent cardiac transplantation 4 months earlier was admitted with a 1-week history of fever of unknown origin. Her immunosuppressive medications at the time included tacrolimus and prednisone. In addition, she had been receiving intravenous immunoglobulins monthly through an indwelling catheter for hypogammaglobulinemia. A workup included blood and urine cultures that were negative except for cytomegalovirus antigenemia. Ganciclovir treatment was started, and the patient was referred for an ophthalmic examination. Her corrected visual acuity was 20/50 OU. Low-grade anterior uveitis was noted in the left eye without evidence of vitritis or chorrioretinitis. The patient denied any history of ocular trauma. Additional tests, including rapid plasma reagin, the hemagglutination treponemal test for syphilis, rheumatoid factor, antinuclear antibody levels, angiotensin-converting enzyme levels, HLA-B27 antigen, and chest radiography, were ordered, and the results of all tests were unrevealing.

Six weeks later, the patient’s visual acuity had dropped to 20/200 OS, and she developed a hypopyon. Because of the progression of anterior uveitis despite topical steroid treatment and persistent fever, an anterior chamber tap and a lumbar puncture were recommended. Stains of the aqueous humor revealed 3+ polymorphonuclear neutrophil leukocytes with no organisms. Aqueous cultures grew C albicans. The cerebral spinal fluid showed a marked lymphocytic pleocytosis with increased protein but no growth. The patient was started on a regimen of intravenous amphotericin B, and topical fluconazole and ganciclovir treatments were discontinued.

Postoperatively, fungal invasion of the iris and lens with formation of a fungal ball at the pupillary margin were noted (Figure 1). Another anterior chamber tap was performed and amphotericin B (5.0 µg) was injected into both the anterior chamber and vitreous, but the iridocylenticular opacity persisted. Subsequently, because of lack of clinical response, pars plana vitrectomy, lensectomy, and excision of the involved iris with intravitreal amphotericin B (5.0 µg) were performed in the left eye. Intraoperatively, white fluffy material within the body of the crystalline lens and over the pars plana was noted and cultured. These findings prompted complete capsulectomy and aggressive vitreous base shaving with scleral depression. Intraoperative examination of the fundus did not show any involvement of the retina or choroid. Cultures obtained from the lens material and vitreous cassette were both positive for C albicans. The patient then underwent 2 additional administrations of intravitreal amphotericin B (5.0 µg) every 72 hours. The results of cultures from these procedures remained negative for organisms.

After initiation of systemic treatment, the patient’s fever ceased, and...