**Visual Loss Due to Progressive Multifocal Leukoencephalopathy in a Congenital Immunodeficiency Disorder**

A 20-year-old man with Wiskott-Aldrich syndrome (WAS) initially developed a mild visual disturbance that progressed to blindness, increasing neurological deficits, and death within 4 months. Wiskott-Aldrich syndrome is an X-linked immunodeficiency disorder characterized by thrombocytopenia, eczema, and susceptibility to infection.1 This case illustrates the difficulties in reaching the final diagnosis of progressive multifocal leukoencephalopathy (PML) in this individual and its unusual histopathologic features.

**Report of a Case.** A 20-year-old white man with WAS had a 1-month history of decline in vision. Nine months previously, he had omitted 3 consecutive doses of immunoglobulin, which he had been receiving every 3 weeks since the age of 12 years. This was restarted when he developed lethargy and malaise. His mother had been diagnosed with multiple sclerosis at age 26 years.

An ophthalmological examination revealed a visual acuity of 20/17 OD and 20/20 OS with normal pupillary reactions. Color vision was abnormal, and visual field testing showed bilateral enlarged blind spots with paracentral scotomas. Fundi appeared grossly normal. Results of fundus fluorescein angiography were unremarkable. Electrodagnostic evaluation was normal apart from delayed visual evoked potentials, which were suggestive of demyelination.

The patient's biochemistry and complete blood cell count were normal except for a low platelet count (12 × 10^3/µL). Mutational analysis for Leber hereditary optic neuropathy and for the WAS protein gene demonstrated no Leber mutation, but there was a single nucleotide substitution (C155T) in exon 1 of the WAS gene.

Six weeks later his vision was bilateral finger counting. A magnetic resonance imaging (MRI) scan showed scattered areas of white-matter foci, predominantly peripheral and not periventricular, on proton density and T2-weighted scans. He was given intravenous methylprednisolone sodium succinate because of the possibility of demyelination. No improvement occurred, and he was registered as blind. Reduced sensation and generalized mild weakness (level 4-5) developed in his right-hand side and face. Reflexes were normal and he had no dysphasia.

A repeat MRI scan showed an increase in the number of small signal foci together with more confluent areas of high signal intensity and a scalloped edge in the left occipital region (Figure 1). Rapid deterioration occurred with protracted focal seizures, total blindness, loss of consciousness, and death. A postmortem histopathologic examination of the brain revealed the papovavirus particles associated with PML.

The fresh brain was examined and then prepared in slices. Tissue underwent light microscopic examination and was embedded in paraffin wax, sectioned, and stained with hematoxylin-eosin. The brain weighed 1570 g. External examination showed mild diffuse vascular congestion but no focal lesions. Brain slices had large numbers of small, gray, circular lesions in the white matter that were 1 to 3 mm in diameter, many of which were close to the cortico–white-matter junction. In some cases the lesions were also visible within the cortical ribbon. The lesions had the appearance of small regions of demyelination and resembled the individual lesions of PML. In addition to the spherical lesions, there were also irregular linear lesions along the cortico–white-matter junction that in some instances were accompanied by visible, fine-linear white scarring in this area (Figure 2). In the most severely affected gyri, the cortex was also involved and had a darker color, with apparent expansion and softening of its affected parts.

Although the individual lesions were present throughout the cerebral hemispheres, they were most frequent bilaterally in the occipital poles, which were also the major site of linear cortico–white-matter junction lesions and scarring as well as the regions of diffuse cortical involvement. A microscopic examination of the brain sections showed small, spherical, white matter lesions that were atypical of PML because they were associated with a considerable perivascular and parenchymal inflammatory infiltrate (Figure 3). The inflammatory infiltrate was mostly lymphocytic in character but also contained plasma cells that were occasionally binuclear. However, further examination indicated the presence of diagnostically enlarged magenta-colored oligodendrocyte nuclei around the periphery of the foci of demyelination, containing the papovavirus particles associated with PML (Figure 4). Other differences from typical examples of PML were that the pleomorphic astrocytic reaction within the lesions was less florid and that foamy macrophages were not as conspicuous, although there were numerous mononuclear cells within the larger lesions. In a few cases the isolated lesions had the character of “burnt-out” lesions, with no enlarged oligodendrocytes, no significant inflammatory reaction, and a predominantly fibrous gliosis. The linear lesions in the cortico–white-matter junction had the same histological characteristics as the spherical lesions. The diffuse cortical lesions showed demyelination within the cortex and reactive astrocytosis extending through the neuropil and around the neurons, although no enlarged oligodendrocyte nuclei were identi-
fied within the affected regions of the cortical ribbon.

Comment. Delay in the diagnosis of PML was probably due to visual symptoms dominating this case, late development of neurological signs, and the outstanding longevity and relative good health of the patient. Inherited conditions including Leber hereditary optic neuropathy or the possibility of a contiguous deletion in the same region as the WAS gene, leading to a phenotypic complex of WAS and perhaps an X-linked cone dystrophy, were explored and excluded.

Although the lesions that appeared on the initial MRI scan were not completely typical of multiple sclerosis,2 electrodiagnostic test results were suggestive of optic nerve demyelination. Visual disturbance in multiple sclerosis is almost always due to optic nerve rather than cortical involvement. The reverse is true for PML. Despite increasing central scotomata, the preserved pupillary reflexes should have directed us earlier to a cortical etiology. Typical

Figure 1. Second magnetic resonance imaging scan (17 weeks after initial symptoms) showing an increase in the number of small high signal foci with more confluent areas of high signal intensity, a scalloped edge in the left occipital region, and a serpiginous scalloped appearance at the gray matter/white matter interface involving the arcuate fibers.

Figure 2. Fresh brain slices showing large numbers of small, gray, circular lesions in the white matter close to the cortico–white-matter junction (white arrows). Also seen are irregular linear lesions along the cortico–white-matter junction (black arrowheads).

Figure 3. Microscopic picture showing small, spherical, white matter lesions with marked vascular prominence and parenchymal infiltrate (hematoxylin-eosin, original magnification ×20).

Figure 4. Microscopic section showing the presence of diagnostic enlarged homogenous oligodendrocyte nuclei, which contain the papovavirus particles associated with progressive multifocal leukoencephalopathy (arrows) at the periphery of the focus of demyelination (hematoxylin-eosin, original magnification ×100).
PML features became more identifiable by the second MRI scan (Figure 1). The histopathologic characteristics of this case include an unusual variant of PML: there was a marked inflammatory response, and the demyelinating lesions had an unusual distribution. The inflammatory response was presumably a reflection of the immune status of this patient. Whereas most cases of PML are associated with immune suppression, no matter what the cause, this case is unusual because a marked inflammatory response was noted.3,4 A study of ophthalmic signs in patients with acquired immunodeficiency syndrome and PML stated that bilateral occipital lobe PML may lead to cortical blindness, which appears to be a relatively common event but has received inadequate recognition.3 The initial features of this case offer a useful model for other immunodeficiencies in which patients experience loss of vision and neurological signs.

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Complication From Use of Alcohol to Treat Epithelial Ingrowth After Laser-Assisted In Situ Keratomileusis

Epithelial ingrowth occurs in up to 4.3% of patients following laser-assisted in situ keratomileusis.1 Treatment varies from observation to lifting the flap and scraping away the epithelium. With recurrence, additional treatment options include the use of an excimer laser, cocaine, proparacaine hydrochloride, or alcohol on the stromal bed and flap and suturing the abnormal flap edge. Studies on the efficacy of these interventions are lacking. We report a case of total flap melting following alcohol application to the interface to treat recurrent epithelial ingrowth.

Report of a Case. A 52-year-old woman underwent bilateral laser-assisted in situ keratomileusis for moderate myopic astigmatism. Postoperatively the patient had irritation and focal flap edema in her left eye that persisted for 6 months before epithelial ingrowth was noted. The patient’s flap was lifted, scraped, and irrigated 8 months postoperatively. Twelve days later, recurrent epithelial ingrowth was observed. The retreatment consisted of elevating the flap, scraping the stromal bed and flap undersurface, applying absolute alcohol on a 6-mm sponge to the stromal bed and flap undersurface twice for 10 seconds, and irrigating the interface.

At the time of referral 6 days later, the patient’s visual acuity was 20/200 with pinhole approximation, and she had a large central epithelial defect. The edematous flap made the interface difficult to examine. Her epithelial defect improved during the next week with conservative treatment, and her visual activity improved to 20/80 with pinhole approximation. Four days later, the patient returned with 80% melting of the flap (Figure 1). The remaining nasal and temporal pieces of the flap were removed and sent to the pathology department (Figure 2).

Comment. Many theories exist regarding the source of the epithelial cells in epithelial ingrowth, but most researchers believe that the cells grow under the flap from the keratomileusis incision. The factors that stimulate or allow this growth are not known, but poor adhesion of the flap may be an important factor. When treating epithelial ingrowth,
complete removal or death of all of the cells is important to prevent recurrence. When initial scraping and irrigating fails, additional measures such as the use of an excimer laser or alcohol on the stromal bed and flap have been attempted. Alcohol has been used in photorefractive keratectomy for many years to remove the epithelium. However, some reports show that alcohol can induce keratocyte death and increase inflammation compared with mechanical debridement.

Use of alcohol in this case may have influenced flap adhesion in several ways, allowing aggressive epithelial ingrowth to occur with subsequent flap melting. First, the alcohol may have caused keratocyte death on both the flap and stroma, which can affect the interaction and wound healing of these 2 surfaces. Second, the alcohol may have inadvertently touched the surface epithelium, leading to the epithelial defect, flap edema, and poor flap adhesion. Because there is a lack of basic and clinical research evaluating alcohol in the treatment of epithelial ingrowth, use of this toxic agent in the lamellar interface should be approached with caution.

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Serous Retinal and Choroidal Detachment After Macular Hole Surgery

One of the most serious complications of macular hole surgery is retinal detachment, which is usually treated promptly with a second surgery. Recently, 4 cases of spontaneous resolution of retinal detachment following macular hole surgery were reported. The authors proposed several possible mechanisms for postsurgical retinal detachment in the absence of a clinically detectable retinal break. We present the first report, to our knowledge, of a patient with concurrent peripheral annular choroidal and inferior bullous retinal detachment noticed 1 week after macular hole surgery. After observation for 7 weeks, both resolved spontaneously. This case provides evidence that the cause of retinal detachment after macular hole surgery may be exudative.

Macular holes in stage 2, 3, or 4 are now widely managed with pars plana vitrectomy, posterior hyaloid peeling, and intravitreal perfluorocarbon gas tamponade. Complications of macular hole surgery include cataract, retinal pigment epithelial changes, visual field loss, endophthalmitis, choroidal neovascularization, cystoid macular edema, and retinal detachment. The latter has been recognized to occur secondary to iatrogenic or postoperative peripheral retinal breaks. Recently, retinal detachment following macular hole surgery has been reported to resolve spontaneously in the absence of any clinically detectable retinal tear. Several hypotheses were proposed: (1) small occult breaks that ultimately close, (2) a postsurgical temporary increase in fluid flow through the macular hole, or (3) subretinal exudation caused by surgery.

Report of a Case. A 69-year-old African American man with hypertension and hyperlipidemia was seen for a 1-year history of central distortion in the right eye. Best-corrected visual acuity was 20/400 OD and 20/30 OS. Intraocular pressure was 14 mm Hg in both eyes. Findings from slitlamp examination revealed bilateral nuclear sclerotic cataracts with cortical spurs. Fundus examination of the right eye revealed a stage 3 macular hole with surrounding neurosensory detachment and a mild epiretinal membrane. The patient underwent pars plana vitrectomy, peeling of the posterior hyaloid from the disc to the periphery with a soft-tip silicone cannula, perifoveal epiretinal membrane removal using a soft-tip diamond-dusted silicone cannula, and intravitreal 14% perfluoropropane gas tamponade. The posterior hyaloid membrane was extremely adherent to the disc, and multiple attempts were required to separate it in this location. No adjuvant for hole closure was used. Careful examination of the peripheral retina was performed immediately prior to the fluid-air exchange, identifying no disease. Intraocular pressure 2 and 4 hours postoperatively was 19 and 21 mm Hg, respectively. On postoperative day 1 the vitreous cavity was filled 80% with gas, the retina appeared flat, and intraocular pressure was 21 mm Hg.

One week later, the patient reported no new symptoms and was noticed to have a 360° peripheral annular choroidal detachment of moderate height accompanied by an inferior bullous retinal detachment extending from the 4- to the 8-o’clock position with shifting subretinal fluid. The vitreous cavity was filled 60% with gas. No peripheral retinal tears were seen. The patient continued receiving topical 1% prednisolone acetate 4 times a day. Two weeks later the choroidal detachment resolved. On postoperative week 6, the retinal detachment decreased in extent, and 1 week later it resolved completely. The macular hole was closed, with visual acuity of 20/200 OD. Three months later, the patient underwent phacoemulsification with posterior chamber intraocular lens implantation, and visual acuity improved to 20/50 OD.

Comment. This is the first report, to our knowledge, of choroidal detachment after macular hole surgery, concurrent with a bullous retinal detachment that showed spontaneous resolution. Akduman et al reported several cases of retinal detachment occurring after macular hole surgery that spontaneously resolved. The authors proposed that retinal detachment may be secondary to (1) small occult breaks that ultimately close when traction on the vitreous base from the intraocular gas bubble decreases as the gas reabsorbs; (2) a temporary increase in flow of fluid from the vitreous cavity through the macular hole after the cortical vitreous is surgically removed; or (3) subretinal exudation from tissue stress caused by the surgery.
This patient provides strong evidence that the cause of retinal detachment in some cases is exudative. Although this is supported by its bullous appearance, inferior localization, and shifting subretinal fluid, the coexistence of the retinal detachment with a choroidal detachment, both showing spontaneous resolution, reaffirms exudation as the most likely underlying mechanism. The cause for postsurgical choroidal and subretinal exudation may be related to intraoperative and/or perioperative transient hypotony and/or surgical tissue trauma with consequent release of chemomodulators. The only feature in this patient’s intervention that sets him apart from the usual macular hole surgery case is the considerable effort required to separate the posterior hyaloid from the disc. It is possible that this played a role in causing subretinal and choroidal exudation from the optic disc and peripapillary region. This case stresses the need for close patient observation instead of an immediate reoperation when retinal detachment following macular hole surgery does not show a clear rhegmatogenous origin.

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Intravitreal Triamcinolone for Refractory Cystoid Macular Edema Secondary to Birdshot Retinochoroidopathy

Birdshot retinochoroidopathy is a chronic, bilateral uveitic disorder. Originally described by Ryan and Maumence, it is characterized by posterior segment inflammation in the presence of multiple depigmented choroidal lesions symmetrically scattered throughout the post-equatorial retina. The cause is presumed to be autoimmune and...
more than 90% of patients test positive for the HLA-A29 serotype. Approximately half of affected eyes develop cystoid macular edema (CME), and this represents a major cause of considerable visual loss from this condition. A rationale for treatment with corticosteroids has been established based on the inflammatory nature of the disease. However, systemic and periocular corticosteroids have failed to produce significant improvement in most treated patients. We report 2 cases of refractory CME secondary to birdshot retinochoroidopathy that were successfully treated with intravitreal injections of triamcinolone acetonide.

**Report of Cases.** 

**Case 1.** A 60-year-old woman was diagnosed with birdshot retinochoroidopathy 3 years prior to initial examination. She was positive for HLA-A29 and had a fundus appearance consistent with this condition (Figure 1). This included multiple creamy yellow choroidal lesions posterior to the equator bilaterally. The anterior vitreous showed mild cells with some vitreous debris.

At initial examination, she complained of chronic floaters with occasional photopsia. However, she noted acute blurring of vision and distortion in the left eye during the prior 6 weeks. Her best-corrected visual acuity was 20/20 OD and 20/60 OS. Intraocular pressures were 14 mm Hg bilaterally. Anterior segment examination findings were normal with the exception of mild nuclear sclerotic cataracts. Indirect ophthalmoscopy and slitlamp biomicroscopy showed a normal optic disk and retinal vasculature. The right macula was normal, and the left macula showed intraretinal thickening involving the fovea with a cystoid appearance. The retinal periphery showed symmetric birdshot lesions as described.

Fluorescein angiography was obtained and showed leakage in a petalloid pattern involving the left fovea. Optical coherence tomography (OCT) confirmed CME with intraretinal thickening measured at 540 µm.

The patient was given ketorolac topical drops 4 times a day. At 3 months, her visual acuity remained 20/60 OS and OCT showed no improvement in CME (Figure 2). The patient was offered an intravitreal triamcinolone injection to treat residual edema. Informed consent was obtained, and the patient underwent injection of 4 mg of triamcinolone acetonide (Kenalog 40; Apothecan, Princeton, NJ) in 0.1 mL. The injection was performed under topical anesthesia through the pars plana inferiorly using a 27-gauge needle. Immediately after the injection, the patient described a transient visual perturbation owing to the opaque corticosteroid compound suspended in the vitreous cavity. This had resolved over the next 2 days. Within 10 days, OCT showed reduction of macular thickness to 240 µm with improvement of visual acuity to 20/50 OS. At 2 months, CME resolved completely with a return of OCT macular thickness to 190 µm (Figure 3). Her visual acuity improved to 20/25 OS at this interval. After 6 months of follow-up, the patient maintains this level of acuity and has shown no recurrence of CME. Macular thickness remains normal at 190 µm as measured by OCT. The greatest intraocular pressure measured during the follow-up period was 18 mm Hg. The patient showed no progression of cataract during this interval.

**Case 2.** A 38-year-old woman was diagnosed with birdshot retinochoroidopathy on initial presentation based on characteristic fundus findings. Findings from HLA-A29 testing were positive. Slitlamp biomicroscopy showed considerable neovascularization of the right optic disc along with venous sheathing. Both maculas showed trace CME. The peripheries were notable for symmetric, creamy yellow birdshot lesions scattered throughout the postequatorial region. The
vitreous showed 1+ cells bilaterally. Panretinal photocoagulation was performed on the right eye, and the patient was followed clinically for 3 years with stable visual acuity at the 20/40 level.

On an emergency visit, the patient reported acute visual loss in the right eye accompanied by central distortion during the prior week. Her best-corrected visual acuity measured 20/400 OD and 20/60 OS. Intraocular pressures were 12 mm Hg and 15 mm Hg, respectively. The anterior segments were normal with the exception of rare cells in each anterior chamber. Examination of the anterior vitreous revealed 2+ cells on the right and 1+ cells on the left. Fundus examination of the right eye showed persistent disc neovascularization with a band of new preretinal hemorrhage beneath the inferotemporal arcade. The right macula showed considerable CME. Although the left macula showed mild CME, she had remained essentially stable in this eye and was not symptomatic until visual loss occurred on the right.

Fluorescein angiography confirmed leakage from disc neovascularization as well as CME in a petalloid pattern. The OCT measured the intraretinal thickening at 290 µm. The patient was treated with further panretinal photocoagulation and a sub-Tenon injection of triamcinolone acetonide (40 mg/mL). At 1-month follow-up, her visual acuity remained at 20/200 OD. The CME showed no response to therapy and actually increased to 370 µm on OCT (Figure 4). The patient was followed up for 1 additional month without a clinical response. At this time, she was offered an intravitreal triamcinolone injection to treat residual edema. After obtaining informed consent, she was injected with 0.1 mL of triamcinolone acetonide (40 mg/mL) through the pars plana inferiorly. She experienced a transient visual perturbation from the opaque intravitreal corticosteroid suspension lasting 2 days. At 1 month, her visual acuity improved to 20/100 OD. The macular thickness was reduced to 220 µm on OCT. Her visual acuity gradually recovered to 20/50 OD at 3 months with a corresponding macular thickness of 140 µm on OCT (Figure 5). At 6 months following intravitreal corticosteroid injection, she maintained a stable macular thickness of 140 µm and visual acuity measures 20/30 OD without correction. Of note, neovascularization of the disc has shown regression at 6 months. It is unclear whether this is an effect of prior photocoagulation and/or antiangiogenic effect from the corticosteroid. Intraocular pressure never exceeded 16 mm Hg during the follow-up interval, and there was no evidence of cataract formation during this time.

**Comment.** Birdshot retinochoroidopathy often is seen with CME, a common cause of visual loss in this uveitic condition. Current treatments target the inflammatory nature of the disorder. Corticosteroids represent the mainstay of therapy, but systemic and periocular routes of administration have produced disappointing results in controlling inflammation and preserving visual acuity. Cyclosporine A has shown promise owing to its potent immunosuppressive effect, but its role has not been fully established in birdshot retinochoroidopathy.4,5 Two patients are described who had CME that responded promptly to intravitreal administration of triamcinolone acetonide, an injectable corticosteroid suspension. Both showed marked improvement in macular thickness with a corresponding dramatic increase in visual acuity maintained for 6 months of follow-up.

The rationale for intravitreal corticosteroids parallels that established for other routes of corticosteroid administration, specifically the anti-inflammatory effect. However, the intravitreal route alleviates the pharmacologic issues of penetra-
tion and bioavailability. A potent dose of medication is delivered directly to its site of action with a rapid onset. With this more aggressive approach, concerns arise regarding adverse events associated with the corticosteroid medication and the injection procedure.

Specifically, corticosteroids have been associated with a rise in intraocular pressure as well as the development of cataracts. All routes of corticosteroid administration share these risks, although the risk may be theoretically amplified with injection into the eye. The injection procedure itself introduces unique risks of endophthalmitis, retinal detachment, and hemorrhage. Larger studies of corticosteroid injections for other conditions have not shown significant morbidity associated with the intravitreal injection procedure. However, this intervention may best be reserved for those truly refractory cases that have failed standard topical, regional, and oral routes of delivery. Of note, neither treated patient experienced any adverse effects related to the drug or the injection procedure. Both experienced transient visual disturbance lasting a few days owing to the opaque nature of triamcinolone suspended in the vitreous cavity. The risks seem justified based on the failure of more conservative approaches in the presence of progressive visual loss.

Intravitreal corticosteroid injection seems to be a viable option for the treatment of refractory CME owing to birdshot retinochoroidopathy. Preliminary results show prompt resolution of edema with corresponding improved visual acuity. Improvement in visual acuity lags resolution of macular edema temporally, with recovery of retinal function after restoration of structural integrity. The duration of effect exceeds 6 months in both treated patients. Further study is warranted to evaluate the long-term risks and benefits associated with this promising treatment modality for CME complicating birdshot retinochoroidopathy.

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Exudative Retinal Detachment in Behçet Disease

Behçet disease is a systemic vasculitis of uncertain cause. Ocular inflammation is 1 of 4 clinical criteria on which the diagnosis is based. Anterior uveitis and occlusive retinal vasculitis with or without retinitis are characteristic. Rhegmatogenous retinal detachment has been reported. An unspecified type of retinal detachment was observed subsequent to severe necrotizing retinitis. This report describes 2 patients who developed exudative reti-
nal detachment (ERD) during the course of Behçet disease. One had an unusual pseudohypopyon in the subretinal space. Both ERDs were recurrent and associated with hemorrhagic retinal vasculitis.

Report of a Case. Case 1. A 24-year-old Albanian man developed acute, painful, severe visual loss in his right eye. Similar episodes had occurred in both eyes during the preceding 6 months. Pain and redness resolved spontaneously within a few weeks, and visual acuity improved over several months. Previous treatment included topical steroids for uveitis with subretinal fluid. His medical history was unremarkable. Positive findings on systems review included painful oral and genital ulcers and nodular rashes on the lower extremities. He denied tinnitus, headache, stiff neck, viral illness, vertigo, ocular trauma, and arthritis.

Visual acuity was counting fingers at 1 foot OD and 20/400 OS. Biomicroscopic examination revealed 4+ anterior chamber cells, microhypopyon visible with goniophotography, and posterior synechiae in the right eye. The vitreous was hazy and cellular. Exudative retinal detachment with sheets of yellowish subretinal material extended into the macula (Figure 1). There was a sheathed inferonasal vessel with associated hemorrhage. Left eye anterior segment and vitreous were quiet. Yellow subretinal linear deposits were in the fovea. The retinal vessels and optic nerve were normal. Intravenous fluorescein angiography of both eyes showed no serious vascular perfusion defects.

The patient received a subtenon steroid injection and oral prednisone (60 mg). He was admitted the following morning to receive intravenous methylprednisolone, 1 g daily. Azathioprine, 150 mg daily, was also started. Three days later, there was considerable vitreous clearing and subretinal fluid resolution. Hemorrhagic vasculitis was more visible (Figure 2). Indocyanine green angiography findings were normal. Human leukocyte antigen testing was positive for HLA B51 antigen. Findings from chest radiography, PPD (purified protein derivative), fluorescent titer antibody-antibody screen, angiotensin-converting enzyme, complete blood cell count, chemistry panel, and urinalysis were normal. Prescriptions at discharge included prednisone, 60 mg daily, and azathioprine, 150 mg daily. Panuveitis without ERD recurred in the right eye 4 months later owing to noncompliance with administration of medication. Resumption of medications controlled the recurrence. No additional ocular recurrences developed. Prednisone was slowly tapered to 10 mg, azathioprine was increased to 200 mg, and cochicine, alendronate sodium, and calcium were added. Twenty-three months later, visual acuity was 20/400 OD and 20/30 OS. Foveal retinal pigment epithelial clumping was present more prominently in the right eye. There were no residual exudates.

Case 2. A 60-year-old African American woman was seen for retinal consultation. She had a 12-year history of multiple emergency department visits and hospital admissions for uveitis diagnosed previously as toxoplasm retinitis, possible Behçet disease, and sarcoidosis. Treatment included 3 admissions for intravenous steroids, multiple courses of topical steroid drops, and several 3- to 6-month courses of oral prednisone. Each recurrence caused additional visual loss. Her medical history was notable for a Ghon complex on chest radiography, isoniazid-
induced hepatitis, and osteoporosis. Systems review revealed painful genital and oral ulcers but no arthritis, headache or central nervous system disease, tinnitus, vitiligo, or skin rashes. Abnormal test results included positive PPD without anergy, positive toxoplasma enzyme immunosorbent assay, occlusive vasculitis on intravenous fluorescein angiography, and reduced flow in the left posterior ciliary artery and both ophthalmic arteries on color Doppler ultrasonography. Angiotensin-converting enzyme, gallium scan, and cerebrospinal fluid examination findings were normal. Human leukocyte antigens included HLA A28, B70, and Bw6.

Medical records revealed that ERDs were recognized on 2 occasions in the left eye. In December 1987, 1 month after a 10-month oral prednisone tapering treatment for papillitis and panuveitis in the right eye, visual loss, redness, pain, hypopyon iritis, and vitreitis developed in the left eye. B-scan ultrasonography demonstrated ERD (Figure 3). Inflammation was controlled, and the ERD resolved with intravenous steroids, but visual acuity was counting fingers.

In January 1992, pain, redness, and visual loss developed in the left eye after a 2-year period marked by bilateral cataract extraction and recurrent asymptomatic inflammatory branch vein occlusions treated with intermittent oral, topical, and subtenon steroids. Clinical examination revealed panuveitis and recurrent ERD with an unusual hypopyon in the left eye (Figure 4). Ocular inflammation and ERD resolved during a 4-month course of oral prednisone. Visual acuity was hand motions. Multiple relapses at 2- to 6-month intervals were managed with intermittent subtenon injections, steroid drops, and short courses of oral prednisone. In November 1994, Behçet disease was diagnosed, and systemic immunosuppression was initiated. The patient has remained relapse free with visual acuity of 20/60 OD and hand motions OS.

Comment. The differential diagnosis of ERD should include Behçet disease. Exudative retinal detachment may complicate posterior uveitides, including Vogt-Koyanagi-Harada (VKH) disease, sympathetic ophthalmia, posterior scleritis, and sarcoidosis, but has not been described in Behçet disease. Unlike most other entities, ERDs in Behçet diseases were recurrent and associated with hemorrhagic retinal vasculitis.

The visual outcome in Behçet ERDs ranged from prolonged, gradual recovery to no recovery after each relapse. One important goal of treatment therefore must be to prevent relapses with long-term immunosuppression rather than to respond to ERDs after they develop. Single or recurrent exacerbations may cause irreversible visual acuity or visual field loss despite treatment.

Spontaneous remissions following sudden and severe exacerbations characterize Behçet uveitis. Spontaneous remissions may be misinterpreted as indications of a self-limited or benign process or that subtherapeutic amounts of steroid have controlled the disease. Inaccurate or delayed diagnosis may impede timely, appropriately aggressive, long-term administration of immunosuppressive agents needed to prevent recurrences and blindness.4

Behçet ERD may be differentiated from other posterior uveitides. Recurrent ERDs are unusual in VKH disease in which ERD typically occurs during an initial attack that is followed by anterior segment inflammation. Hemorrhagic vasculitis is not
a characteristic feature of VKH disease, sympathetic ophthalmia, or posterior scleritis, although it may complicate sarcoidosis. Also, ocular inflammation often is not the presenting manifestation of Behçet disease, occurring initially in only 8.6% to 34% of patients.1-3 Thorough systems review is of utmost importance when Behçet disease is suspected. Human leukocyte antigen testing may support the diagnosis. Behçet disease should be considered in the differential diagnosis of ERD, especially if it is recurrent or associated with hemorrhagic retinal vasculitis.

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