

Grand Rounds

7-29-09

Introduction: Howard Tessler, MD and Debra Goldstein, MD
(Attendings)

This week's Cornea Grand Rounds presents several interesting cases encountered by our uveitis faculty.

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Serpiginous Choroiditis: Javaneh Abbasian, MD (Resident)

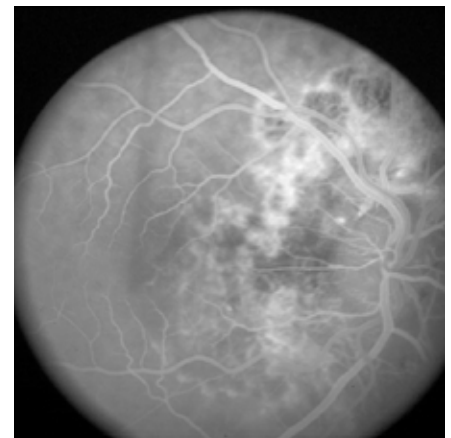


A 71 year-old male patient presented with complaints of a central scotoma in the right eye starting in December 2006. The patient was treated by the retina service with ranibizumab and was evaluated by the uveitis service. He denied any other ocular symptoms; no flashes, floaters, pain, or diplopia. His review of systems was negative for rashes, neurologic symptoms, consumption of raw meats, genital or oral ulcers, tick bites, TB exposure, shortness of breath and digestive symptoms. He did have a history of cat exposure. His past medical was non-contributory, his surgical history only significant for pars plana vitrectomy with membranectomy in the right eye for an epiretinal membrane in February 2007. He did admit to excessive alcohol consumption but denied cigarette and illicit drug use.

The physical exam was significant for a best corrected visual acuity of 20/25 OD and 20/20 OS. His extraocular motions, confrontation visual fields, intraocular pressure and anterior segment exam were all within normal limits. His posterior exam is shown below with fundus photography and fluorescein angiography (Fig 1, 2). He was diagnosed with serpiginous choroiditis based on the clinical exam findings and the patient was treated with high dose oral prednisone while in the active stage. His disease course was significant for two recurrences in the left eye with new activity at the edge of previous scars, as well as an active subretinal neovascular membrane. He required immunosuppression with chlorambucil and a second injection of ranibizumab. The patient was maintained on chlorambucil with weekly monitoring of his white blood cell count.

TREATMENT: The dose of chlorambucil was increased based on a known protocol published in Ophthalmology in 2002 by Goldstein et al. Dosing begins at 2 mg daily with increases of 2 mg per week if the white blood count remains above 3000. If the white blood count is between 2800 and 3000, the dose is decreased between by 2-4 mg/day. Therapy is ultimately discontinued when the white blood count is below 2400 or if platelets drop below 125,000. This patient showed rapid remission of disease and the alkylating agent was subsequently discontinued.

REFERENCE: Goldstein DA, Fontanilla FA, Kaul S, Sahin O, Tessler HH. Long-term follow-up of patients treated with short-term high-dose chlorambucil for sight-threatening ocular inflammation. Ophthalmology. 2002 Feb;109(2):370-7.



FIGURES 1, 2
Color and fluorescein angiographic fundus photos of the right and left eyes, respectively.

Acute Macular Neuroretinopathy: Clement Chow, MD (Resident)



Three days after having a flu-like illness, a 27 year-old Caucasian healthy female complained of scotomas in both eyes. Her symptoms included fever, chills, headache, enlarged lymph nodes, oral and genital ulcers, and a rash involving her scalp, chest, and thighs. The initial work-up included a head CT, brain MRI, lumbar puncture, chest x-ray, labs (ASO, HIV, CMV, ANA, toxoplasma, influenza), vaginal examination, and skin biopsy. All tests were noncontributory. The patient was treated with intravenous antibiotics and oral valacyclovir for 10 days. Her symptoms slowly resolved over the next 4-5 weeks, however the scotomas persisted.

Past ocular history included LASIK surgery 4-5 years ago. She was previously healthy and was on no medications other than oral contraceptive pills. She owned a cat but denied smoking, drinking, drug use, travel history, tick bites, eating raw meat, or history of any STDs. On exam, the vision was 20/20 in both eyes, there were 4-5 small keratic precipitates and central corneal subepithelial haze in both eyes. The anterior chambers had rare cell in the right eye and was quiet in the left. The anterior segment examination was otherwise normal in both eyes, and pressures were 6 in the right, 11 in the left. Dilated fundus examination showed multiple sharply defined, reddish brown, non-elevated, tear-drop lesions in a flower-petal arrangement around the foveas of both eyes (fig 3). Heidelberg OCT showed focal loss at the IS/OS junction and thinning of outer nuclear layer (fig 4). Fluorescein angiography was normal.

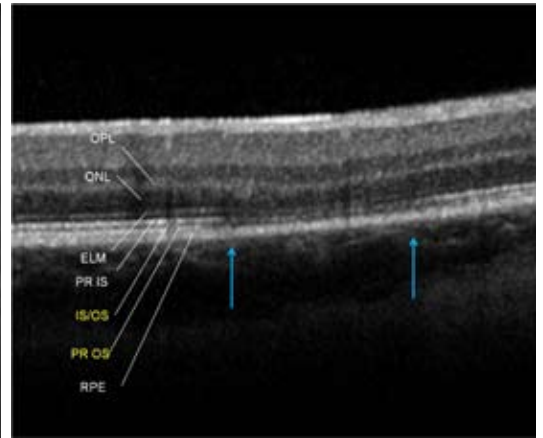
The patient was treated with oral prednisone, however had no improvement at 3 months follow-up.



FIGURE 3
Fundus photo with corresponding red-free photo of the left eye showing the characteristic lesions in AMNR. Similar findings were present in the right eye.



FIGURE 4.
Heidelberg OCT showing the AMNR lesions (between blue arrows); affected inner/outer segment junction as well as the photoreceptor outer segment. The outer nuclear layer was also thinned.



BACKGROUND The patient was diagnosed with acute macular neuroretinopathy (AMNR). It is a rare disease first described by Bot and Deutmann in 1975 and is characterized by lesions as described above which correspond with the shapes and locations of scotomas. Fluorescein angiography is usually normal. Multifocal ERG typically shows localized depression of retinal function. Earlier OCT demonstrates thinning of the inner retina, the junction between the OS and RPE, or RPE itself. In this case, high-resolution OCT demonstrated focal loss at IS/OS junction and thinning of the outer nuclear layer. Turberville SD, et al. (2003) reviewed 41 case reports of AMNR from 1975 - 2002 and showed that 83% of cases were female, mean age being 27 (range 20-53), and can be unilateral or bilateral. 54% showed no improvement whereas 29% showed some degree of improvement.

There is no specific treatment, however oral steroids may help during active disease. Risk factors include oral contraceptives (50%), flu-like syndrome (44%), epinephrine (10%), migraine headaches, contrast media, trauma, postpartum hypotension, and shock. The proposed mechanism includes acute immune damage to the photoreceptors, increased retinal pressure from intense coughing, and possible vasoconstrictive effects on the retinal vasculature.

REFERENCES: Turberville SD, Cowan LD, Gass DM. Acute Macular Neuroretinopathy: A Review of the Literature. *Surv Ophthalmol.* 2003 Jan-Feb;48(1):1-11.

Sarcoidosis: Sing Your Li, MD (Resident)



A 40 year-old female with skin biopsy-proven sarcoidosis presented to the uveitis service in 1995 with bilateral uveitis. Her course has been chronic with frequent exacerbations, requiring the long term use of oral prednisone. She has either failed or been non-compliant with several steroid-sparing agents. She has also had several anterior subconjunctival and posterior subtenons steroid injections. Early on, she developed a steroid response secondary to topical prednisolone drops as well as dense PSC cataracts bilaterally, eventually requiring combined trabeculectomies/cataract extractions with posterior chamber intraocular lenses in both eyes. Her course was also notable for a tractional retinal detachment in the left eye that was repaired and CME in both eyes. Recently, she presented to the uveitis clinic with a recurrence in eye pain and blurry vision bilaterally for three weeks.

Her exam at that time was remarkable for BVCA of 20/40 in the right eye and 20/200 in the left (the left eye vision was at baseline). Her intraocular pressures were normal. The anterior segment exam was significant for conjunctival blebs bilaterally, significant anterior chamber reaction (3+ cell OD and 1+ cell OS), and 1+ cell in the anterior vitreous OU. She did not have any keratic precipitates or iris nodules.

Her dilated fundusoscopic exam (fig 5, 6) was significant for multiple small, white intraretinal and preretinal granulomas in both eyes. She also had an epiretinal membrane (OS>OD) and CME in both eyes. OCT demonstrated that the granulomas were in fact located in the intraretinal space with extension into the preretinal space (fig 7).



FIGURE 5
Color fundus photograph of the right eye demonstrating creamy yellow lesions most notably along the inferior arcade. On clinical exam, these lesions were noted to extend into the preretinal space.

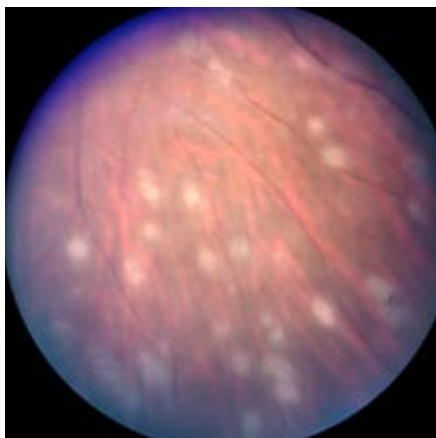


FIGURE 6
Color fundus photograph of the periphery in the left eye demonstrating creamy yellow lesions. On clinical exam, these lesions were noted to extend into the preretinal space.

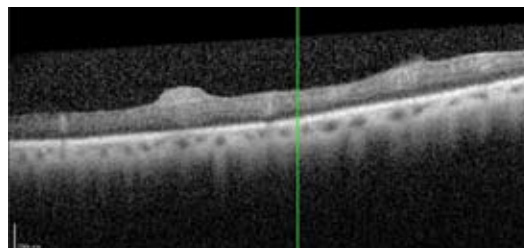


FIGURE 7
Heidelberg OCT image of the right eye. The image slice was taken through an area of the granulomas, demonstrating extension into the preretinal space.

BACKGROUND Sarcoidosis is a systemic granulomatous disease, with 15-50% of patients demonstrating ocular involvement. The main pathologic lesion is the non-caseating granuloma comprised of epithelioid cells, multinucleated giant cells, and a rim of lymphocytes, which can be found in the skin, conjunctiva, iris, or posterior segment. The most common ocular manifestation is anterior granulomatous uveitis. Posterior manifestations include optic neuritis, periphlebitis, retinal vascular disease, CME, and granulomas in choroid, retina, vitreous, and optic nerve.

Previously, histopathologic data demonstrated sarcoid granulomas intraretinally with extension into the pre-retinal space. However, this is the first in vivo documentation of retinal granulomas, showing that they can break into the pre-retinal space.

REFERENCES: 1) Moorthy, Ramana S. (editor) Basic and Clinical Science Course (BCSC): Intraocular Inflammation and Uveitis Section 9 (Basic and Clinical Science Course 2008-2009). American Academy of Ophthalmology, 2008.

2) Frank KW, Weiss H. Unusual clinical and histopathological findings in ocular sarcoidosis. *Br J Ophthalmol.* 1983 Jan;67(1):8-16.

3) Gass JD, Olson CL. Sarcoidosis with optic nerve and retinal involvement. A clinicopathologic case report. *Trans Am Acad Ophthalmol Otolaryngol.* 1973 Nov-Dec;77(6):OP739-50.

4) Frohman L, Grigorian R, Slamovits T. Evolution of sarcoid granulomas of the retina. *Am J Ophthalmol.* 2001 May;131(5):661-2.

Discussion:

Serpiginous choroiditis can be a difficult disease to manage, due to its relentlessly recurrent nature. Early treatment is of the utmost importance, as this can often prevent involvement of the central macula and fovea, making a tremendous difference in an individual's visual function and quality of life. Patients with sight-threatening ocular inflammatory disease may require treatment beyond the use of oral steroids. Although their use may be unfamiliar to most ophthalmologists, systemic immunosuppressants such as chlorambucil are a powerful and important tool in the armamentarium of the uveitis specialist.

The second case is that of a rare condition, acute macular neuroretinopathy. Although AMNR may not be at the top of any practitioner's differential diagnosis, it is important to note for residents and students that non-specific visual symptoms such as scotomas, floaters, photopsias should not be ignored, especially in the context of recent viral or flu-like illness. Such patients must undergo a complete dilated fundus exam, even with normal central visual acuity, as in our patient.

The final case is an interesting presentation of ocular sarcoidosis. Sarcoidosis can present with a wide range of findings in the eye, affecting any structure from anterior to posterior. Although intraretinal sarcoid granulomas are not uncommon, this case was interesting because granulomas extending from the intraretinal to preretinal space have not previously been documented on OCT.

UPCOMING CME COURSES

March 13-19, 2010	Illinois Eye Review
April 4, 2010	Retina Symposium
April 16, 2010	Uveitis Symposium
May 21-22, 2010	Oculoplastics Symposium
June 25, 2010	34 th Annual Alumni Day
September 25, 2010	Pediatric Ophthalmology/Adult Strabismus

Upcoming Grand Rounds

Illinois Eye and Ear Infirmary Ophthalmology Grand Rounds are held Wednesdays at 5:00 pm on the UIC campus at 909 S. Wolcott in the College of Medicine Research Building. For a complete schedule go to www.uic.edu/com/eye and click on Grand Rounds under the Education drop down menu. Or, call 312-996-6590.