Retinal and Optic Nerve Grand Rounds:
Challenging Cases You Don’t See Everyday
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Disclosures: Nothing to Disclose

A. Central Serous Choroidopathy
   a. Two Clinical Presentations
      i. One or more discrete isolated leaks at the level of the RPE
      ii. Diffuse RPE dysfunction: broad areas of hyperfluorescence on FA that contain one or many leaks.
   b. Signs of an OLD Central Serous Choroidopathy
      i. Decreased VA in affected eye
      ii. Window defects on fluorescein angiography
      iii. No leakage
      iv. A U-Shaped area of retinal pigmentary changes
      v. CSC “Drip”
      vi. No fluid
   c. Signs of a Recurrent Central Serous Choroidopathy
      i. Sudden decrease in VA
      ii. Ring of fluid in or near the macula
      iii. VA may improve with more “plus”
      iv. Late stage leakage noted on fluorescein angiography
   d. Treatment of Recurrent CSC
      i. Tincture of time
      ii. Focal laser
      iii. PDT
      iv. Bevacizumab (Avastin)
      v. Why not steroids?
      vi. Kenalog injection

B. Idiopathic CNV After LASIK
   a. Reported to occur in patients who have had LASIK and who also have a history of AMD
   b. POHS
   c. Degenerative myopia
   d. Rare (0.06% in one study) in patients with no history of any ocular pathology
   e. Differential Diagnosis of Serous Detachment in a Young Patient
i. No CNV
   1. Central serous choroidopathy
   2. Fluorescein angiography is the key
   3. No nets in acute CSC
ii. CNV
   1. POHS
   2. High myopia
   3. Trauma
   4. Angioid Streaks
   5. Idiopathic
   6. Best’s disease
   7. Laser photocoagulation scars
f. Etiology after CNV
   i. Flap creation
   ii. Temporary increase in IOP
   iii. Can stress retina
   iv. Photoablation
      1. Is a “punch to the eye”
      2. Contracoup waves that stress the retina
      3. Cause breaks in Bruch’s membrane
C. Coat Disease
   a. Vascular response in the peripheral retina
   b. Unilateral (95%); Males (76%)
   c. Telangiectatic microaneurysms
   d. Exudation
   e. Capillary drop-out
   f. Neovascularization
   g. Vitreal Hemorrhage
   h. Retinal Detachment
i. Differentials
   i. Sickle Cell Disease
   ii. Familial Exudative Vitrereinal Response (FEVR)
   iii. Hemangioma
   iv. Retinopathy of Prematurity
   v. Norrie’s Disease
   vi. Eale’s Disease
   vii. Incontinentia Pigmenti
j. Molecular Genetics
   i. Fascioscapulamuscular dystrophy (FSHD) Genetic Marker
      1. Genetic marker on chromosome 4q35
2. Gene not yet identified
3. Marker consists of deletions or short fragments in this region (band pattern)
4. Almost all patients with FSHD exhibit this pattern

ii. Molecular Genetic Results
   1. A normal band is 36-38 kb
   2. This patient’s bands were 24-27 kb.

k. Diagnosis: FSHD

D. Diffuse Unilateral Subacute Neuroretinitis (DUSN)
   a. Term coined by Don Gass, MD
   b. Unilateral loss of vision in youngsters
   c. Fundus looks like unilateral retinitis pigmentosa, but etiology is inflammatory not hereditary
   d. ERG is flat in affected eye
   e. Etiology believed to be similar to a nematode parasite related to but different than toxocara

E. IRVAN: Idiopathic Retinal Vasculitis, Aneurysms and Neuroretinitis
   a. Bilateral retinal vasculitis
   b. Unknown etiology (idiopathic)
   c. Characterized by
      i. Multiple aneurysms at arterial bifurcations
      ii. Neuroretinitis
         1. Optic disc leakage on fluorescein angiography
      iii. Capillary non-perfusion
      iv. Progressive ischemia leading to
         1. Neovascularization
         2. Vitreal hemorrhage
         3. Tractional retinal detachment
      v. Vascular changes are believed to be due to VEGF over-expression
      vi. IRVAN is a diagnosis of exclusion after other infectious and inflammatory etiologies of vasculitis and aneurysms have been ruled out
         1. Requires referral to PCP for extensive blood work-up and evaluation, for example:
            a. Diabetes
            b. Sickle cell disease
            c. Blood dyscrasias
            d. Anticardiolipid antibodies; anticoagulant testing
            e. Sarcoidosis
            f. Infectious diseases, e.g.
               i. Lyme disease
ii. TB
iii. Syphilis
iv. Rubella
v. CMV
vi. HIV
vii. IRVAN responds favorably to PRP and anti-VEGF injections
viii. Long-term prognosis is unknown

F. Retinal Toxicity
   a. Psychotropic drug use in the past caused degenerative retinal changes in the posterior pole
   b. Differential diagnosis of numular (scalloped) lesions in retina
      i. High Myopia
         1. Also affects the posterior pole greater than the periphery
      ii. Gyrate Atrophy
         1. Disease affecting the production of ornithine transferase which affects the metabolism of ornithine in the body
         2. Causes nummular lesions in the periphery of the retina initially not the posterior pole
         3. Toxicity
            a. Poor excretion of the toxic agent causes continued retinal destruction
            b. Affects posterior pole

G. Vitreomacular Traction Syndrome (VMT)
   a. A vitreoretinal disorder producing traction on the macula
   b. Can cause macular pucker or folds or the macula can look normal
   c. Can mimic a juvenile foveal retinoschisis
   d. SD-OCT
      i. very valuable in demonstrating vitreal traction
   e. Management
      i. Monitor
         1. Spontaneous resolution into a PVD can occur
         ii. If VA drops to 20/60 refer for vitrectomy
      iii. Jetrea (Ocriplasmin)
         1. Intravitreal injected substance that dissolves the adhesion
         2. Works best on small adhesions
            a. 1500 microns or less (one disc diameter)
            b. No associated ERM

H. Retinoschisis (Degenerative)
   a. A split in the retina at the outer plexiform layer
b. Usually bilateral (38-82% of cases)
c. Differentiated from retinal detachment because of visibility of underlying choroid
d. Can be elevated with blister-like borders or flat with pigmentary changes
e. Associated with absolute field loss in affected area
f. Monitor-can progress to RD (10%)
g. Most frequently occur in the inferotemporal quadrant followed by superior temporal quadrant

I. Accutane Toxicity
   a. Differential Diagnosis of Bull’s Eye Retinopathy
      i. Stargardt disease
      ii. Cone dystrophy
      iii. Benign concentric macular dystrophy
      iv. Fenestrated sheen maculopathy
      v. Toxic maculopathy
         1. Plaquinil (Hydrochloroquine)
         2. Isotretinoid-related maculopathy
   b. Importance of a comprehensive drug history
   c. Important tests to perform
      i. Structure
         1. SD-OCT
         2. Fundus autofluorescence
      ii. Function
         1. Full flash ERG
         2. Multifocal ERG
         3. 10 degree visual fields

J. Chronic Pigment Epithelial Detachment (PED)
   a. Separation of the RPE from Bruch’s membrane
   b. Macular edema
   c. Reduced VA
   d. Can be associated with
      i. CSC
      ii. AMD
   e. Monitor or Treat?
      i. Make sure pt. is not using:
         1. Steroids
         2. Erectile Dysfunction (ED) medications
      ii. Treatment?
      iii. Lucentis for Age-Related Macular Degeneration Pigment Epithelial Detachment Study
         1. 3 or 6 0.5 mg injections for 1 year
2. Resolution of edema vs. edema and PED
3. Results
   a. 42% decrease in volume after 1 month but was not maintained over one year
   iv. Some success with PDT and anti-VEGF has been reported
K. Presumed Ocular Histoplasmosis (POHS)
   a. Inflammatory disease causing a chorioretinitis caused by exposure to ocular histoplasmin spores in the air
      i. Associated with residence in river valleys
      ii. Chickens
      iii. Triad of Findings
         1. Punched out chorioretinal lesions
         2. Peripapillary atrophy
         3. Macular CNV
   b. Differentials
      i. Punctate Inner Choroidopathy (PIC)
         1. More vitritis than POHS
      ii. Multifocal Choroiditis
   c. Treatment of macular CNV
      i. Anti-VEGF
L. Astrocytoma (Astrocytic Hamartoma)
   a. Mass composed of astrocytes
   b. Can appear on the optic nerve, in the retina or in the brain
   c. Is associated with Tuberous Sclerosis
      i. Can be an isolated finding
   d. Look for ash-leaf shaped skin lesions
   e. Look for tuberous growths on the fingers
   f. Differentials
      i. Disc drusen
   g. Management
      i. MRI to r/o intracranial astrocytomas
M. Diabetic Papilopatthy
   a. Sudden ischemic event causes swelling of the optic nerve head
   b. Usually unilateral
   c. Occurs in uncontrolled diabetes
   d. Usually in young Type I diabetics
   e. VA can be normal or reduced
   f. VF loss-usually arcuate-can occur
   g. Is not a “papillitis”—not inflammatory
   h. Is not a “papilledema”—no increase in intracranial pressure
i. Monitor
   i. Condition usually resolves on its own in a few weeks
   ii. Unlike AION where VA resolution is more rare
   iii. Includes resolution of VA loss and visual field loss
   iv. Perform other tests to rule out infectious/inflammatory etiology of disc swelling

N. Cerebellar Hemangioblastoma
   a. Cerebellar Mass (Cyst)
      i. Made up of vascular tissue
      ii. Cyctic nature causes compression on the cerebellum and chiasm
      iii. Herniated ventricles and increased intracranial pressure cause papilledema
      iv. Most common CNS lesion seen associated with Von-Hippel Lindau disease
         1. Cystic lesions are found elsewhere in the body
            a. Kidney
         2. Genetic testing available
      v. Can be an isolated finding as seen in this patient
      vi. Look for signs of cerebellar dysfunction and pituitary compression
         1. Balance problems
         2. Hormonal problems

O. Decreased Visual Acuity in Early Glaucoma
   a. Supported by small C/Ds
   b. Supported by well-preserved visual field OD except for central loss
   c. It is rare but has been reported
   d. Will occur in early papillomacular bundle involvement also affecting the fovea
   e. When you can’t explain central VA loss, do a 10 degree visual field test.
   f. This is a case of glaucoma without cupping