Posterior Segment Disease: Case Challenges

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Macular Hole

- Full thickness macular hole OS
- Consult with retinal specialist
  - Felt that due to duration of situation, unlikely that any surgery would have meaningful benefit on vision
  - RTC q 6 mos
  - Monocular precautions including polycarbonate RX

Macular Hole

- Present as a circular to oval depression of varying degrees in the avascular area of the macula
  - May have surrounding cuff of edema
- Most common cause is idiopathic
  - other causes include blunt trauma, severe myopia, solar retinopathy, CME
- Highest incidence in 7th decade of life
- Women 2x as often as men
- Bilateral in 10% of cases

Macular Hole

- Stage I: Foveal detachment, aka Impending hole
- Stage II: Partial thickness holes
- Stage III: Full thickness hole
-Stage IV: full thickness hole with vitreous separation

Macular Holes

- Less than 50% of stage I will progress to stage II w/o treatment
- About 75% of stage II progress to stage III w/o intervention.
  - If pt has macular hole in one eye, 28 to 44% chance of macular hole in other eye w/o a PVD
    - If PVD already, very little chance
### Macular Hole
- Stage I and Stage II holes should be sent to retina specialist to see if vitrectomy may be of use
  - Vitrectomy may help in Stage I to prevent progression to stage II
  - Vitrectomy may be of help in stage II to preserve vision
- Stage III may require macular hole surgery with growth factors

### Epi-retinal Membrane
- AKA macular pucker, cellophane maculopathy
- Can be secondary to peripheral retinal disease, such as detachment or tear; a retinal vascular disease such as BRVO; inflammation; trauma or idiopathic
- Idiopathic tend to be more mild and non-progressive vs. those after retinal tear

### Epi-retinal Membrane
- VA can range from 20/20 to 20/200 or worse
  - Studies show > 5% have worse than 20/200
- Often metamorphopsia is only complaint with idiopathic ERM
- Fewer than 20% of cases are bilateral
- Surgical removal is considered if severe vision loss or distortion

### Epi-retinal Membrane
- Consider surgery if:
  - VA 20/40 or worse
  - Symptomatic
  - Visual need of patient
- 30 minute procedure
- Face down compliance after surgery for up to 2 weeks
- Make sure you have an experienced surgeon!!

### ERM with Pseudohole
- Can get pseudohole with ERM
  - This represents area of normal retinal tissue within abnormal ERM
  - No surrounding halo or rim of subretinal fluid as with hole
  - VA generally good
  - Negative Watske-Allen sign

### Central Serous Retinopathy
- Common disorder of unknown etiology which typically affects men between age 20 and 45
  - Males to females 10:1
- Serous detachment of neurosensory retina due to leakage from small defect in RPE
Central Serous Retinopathy

- Pt typically presents with fairly recent onset of blurred VA in one eye with a scotoma, micropsia, or metamorphopsia
  - VA typically 20/30-20/70
  - Often correctable with low hyperopic RX
  - Unilateral in 70% of cases
- Risk factors
  - Type A personality
  - Stress
  - Use of systemic cortico-steroids

- Appears as a shallow round or oval elevation of the sensory retina often outlined by a glistening reflex
- FA is helpful in providing definitive diagnosis
  - Classic Smoke stack appearance (occasionally)
  - Ink-blot appearance
- OCT shows marked elevation

- 80-90% of pts will undergo spontaneous resolution and return to normal (or near normal) VA within 1-6 mos.
  - >60% resolve back to 20/20
  - Rare to have vision remain < 20/40
- Approx 40% will get recurrence
- CNVM is VERY rare occurrence, but possible

Central Serous Retinopathy

- No known medical therapy has been proven effective
  - Steroids, NSAIDs etc
- Localized photocoagulation may be of some benefit, but only if
  - Duration at least 4 months
  - VA in other eye is reduced from other attacks
  - Recurrent CSR has already reduced VA in that eye
  - Pt is intolerant of vision and willing to take risk
- PDT suggested in some cases
- Behavior modification?

CSR and PDT

- Several small studies have evaluated role of PDT alone, PDT with IVT, and PDT with Avastin for PDT
- Most show that these agents appear to improve VA, but most studies do not include controls
  - Hard to weigh against normal progression

CSR and Avastin

- 6 pts treated with IV Avastin
  - Initial VA: 20/38
  - VA after 3 mos: 20/20
  - CMT decreased from 331.5 um to 164 um
  - 4/6 had recurrence
- Bottom line: Avastin and PDT should probably be reserved for chronic cases, pts that need immediate VA improvement, or in other select cases
Cystoid Macular Edema

- Accumulation of fluid in the outer plexiform and inner nuclear layers of the retina, centered at the fovea
- Very common finding with several etiologies
  - Irving-Gass Syndrome; following cataract sx
  - Retinal Vein Occlusions
  - Intermediate or posterior uveitis
  - Retinitis Pigmentosa

Cystoid Macular Edema

- VA can range from 20/25 to 20/400, depending upon level of edema
- Appears as macular thickening and swelling, with loss of foveal reflex
- May appreciate fluid filled cysts with red-free light, sometimes in a honey-comb pattern
- FA demonstrates early leakage with a petaloid or flower like pattern

Cystoid Macular Edema

- Most cases of Irving-Gass resolve spontaneously with in 6-12 mos.
  - Pred Forte and Acular or Voltaren qid is standard treatment for days to weeks
  - Subtenons steroid injection if no response
  - Avoid prostaglandin analogs
- If from other condition, such as BRVO, laser photocoagulation is often beneficial
- Intra-vitreal Steroid Injections

CME

- CME is a complication even in uncomplicated surgery (12%)
  - The more complicated the surgery, the more likely and more severe the CME
- If a pt develops CME in one eye following uncomplicated cataract sx, about a 50% chance it will develop in the second eye
- Caution with PGs and CME!!

Stargardt’s Disease

- AKA Fundus Flavimaculatus
- Most Common hereditary macular dystrophy
- Associated with bilateral pigmented maculopathy, with or w/o deep yellowish-white flecks
- Inheritance thought to be AR, although some dominant cases are described
- Both sexes affected equally

Stargardt’s Disease

- Typically presents in 1st to 2nd decade of life with impaired vision
- Initial finding is non-specific mottling at macula
- With time, a 1.5 DD oval lesion develops at the macula with a “beaten-bronze” appearance
- May or may not be surrounded by yellow-white flecks.
Stargardt’s Disease
- FA may show absence or decrease in normal background choroidal flush, with easy visualization of retinal capillaries
- Peripheral vision and night vision normal
- ERG and EOG only abnormal in advanced cases
- Typical VA 20/200-20/400

Stargardt’s Disease
- No treatment needed
- Management includes:
  - Pedigree analysis
  - Genetic counseling
  - Low vision rehabilitation

Best’s Vitelliform Macular Dystrophy
- Second most common macular dystrophy
- Age of onset very variable, with range from shortly after birth to sixth decade
- Vision varies as well, from 20/20 to 20/400.
- AD with variable penetrance
- Divided into stages based on appearance
- Classically associated with “egg-yolk lesion”, but not always.

Best’s Vitelliform Macular Dystrophy
- Stage 1 (pre-vitelliform): Abnormal EOG with no symptoms or lesion
- Stage 2 (vitelliform): Classic “egg-yolk” stage with lesion 0.50 to 3 DD in size
- Stage 3(Psuedo-hypopyon stage): Lesion can become either fully or partially absorbed
- Stage 4 (vitelliruptive stage): Egg yolk becomes scrambled egg appearance, VA typically reduced at this time
- Stage 5 (end stage) Lesion can scar down or become atrophic

Best’s Vitelliform Macular Dystrophy
- EOG is severely subnormal
- EOG also affected in carriers as well with normal appearing fundi
- VF normal
- VA very variable, 20/20 to 20/400

Best’s Vitelliform Macular Dystrophy
- No treatment needed
- Management includes:
  - Pedigree analysis
  - Genetic counseling
  - Low vision rehabilitation
Branch Retinal Vein Occlusion
- Second most common retinal vascular disorder
- Often associated with systemic HTN
- Peak incidence in 5th to 6th decades, with no sex predilection
- Seen bilaterally 4-5% of the time

BRVO
- Clinically, varies depending upon severity of presentation
- Classic presentation is dilated tortuous veins and dot-blot hemes from site of compression to periphery in sector normally drained by that vein
  - Can also see flame-shaped hemes and cotton wool spots as hypoxia develops
  - Lipid can also develop leading to macula edema
- Literature states temporal area is affected most often

BRVO
- Management is somewhat controversial, due to good prognosis if left untreated
  - Studies show 20/40 or better VA at one year in 53-60% of untreated BRVO patients
- Each patient should be investigated for presence of underlying systemic disease
- Complications such as ME and NV should be treated if arise

Macula Edema
- Macula edema is reason for vision reduction in these patients
  - Often resolves on its own (50-60%) with no treatment needed
  - If does not resolve, then treatment may be indicated
- Branch Vein Occlusion Study Group concluded that grid laser improves visual outcome in eyes with BRVO and vision 20/40 or worse from macular edema
  - BRVO at least 3 months old
  - VA 20/40 or worse
  - FA within 1 month, demonstrating macula edema and absence of foveal ischemia

BRVO
- Other sequelae of BRVO include neovascularization, either of the retina, disk, or iris
  - NV can lead to vitreous hemorrhage or retinal traction in rare cases
  - Only in 25-30% of BRVO cases
- Typically collateral vessels form to drain the affected areas, preventing NV

Macula Edema
- After focal macular laser is performed, RTC 4 mos, as resolution generally slow
- If persistent macula edema and decreased VA, repeat FA
- Several treatments may be needed
- RTC every 6 to 12 months once edema stabilized, unless metamorphopsia or decreased vision
• BRVO study group also concluded that PRP should be administered in eyes with BRVO and Neovascularization
• Results suggest that there may be no advantage to treat to prevent development of NV

• BRVO
  • If no NV or ME, pts with BRVO can be followed clinically, with repeat DFE and FA if changes occur, every 6-12 mos
    – Home Amsler grid may be useful to monitor for edema

• Macular branch retinal vein occlusion may also occur
• Occlusion of small tributary branch near macula
• Visually disruptive, as macular edema presents up to 85% of the time

• Management includes diagnosis and management of underlying etiology
• At minimum, should have
  – BP evaluated
  – Fasting Blood sugars (FBS)
  – CBC
  – Lipid profile

• Additional tests might include
  – Carotid artery evaluation
  – Cardiac evaluation
  – Additional blood tests
    • ANA
    • RF
    • FTA/ABS
    • ESR

• Most often associated with DM and HTN
• However many other possible etiologies
  – Carotid artery disease
  – Hyperlipidemia/hypercholesterolemia
  – Altered platelet function
  – Coats disease
  – Von-Hippel Lindau
  – Eales’ disease
  – Trauma
### CRVO
- Very visually destructive disease with strong systemic association
- Typically occurs in men, over the age of 50
- Vision is typically compromised, ranging from moderate to total vision loss

### CRVO
- Typically caused by thrombus formation in the central retinal vein at the lamina cribosa
- Thrombus formation from various etiologies
  - Arteriosclerosis
  - Vasculitis

### CRVO
- Presents two different ways, ischemic vs. non-ischemic, although not always so clear cut
  - Non-ischemic characterized by dot/blot hemes, intra-retinal hemes, and possible macula edema
    - 30% of non-ischemic convert to ischemic
    - >10 dd of non-perfusion
  - Ischemic CRVO presents with dot/blot hemes, flame-shaped hemes, CWS, and gross intra-retinal and macula edema. Also, papillidema commonly present

### CRVO
- Visual prognosis is not good, but better in non-ischemic vs. ischemic.
- Biggest concern is risk for neovascular glaucoma, as end result is often total loss of vision
  - NVG in 14-20% of all CRVO
  - NVG almost 60% of the time in ischemic CRVO

### Ischemic vs non-ischemic
- IOP often reduced more with ischemic vs. non-ischemic CRVO
- APD often present with ischemic
- VA generally reduced more with ischemic
  - Rule of thumb: if VA < 20/200 then ischemic.
  - In order to know for certain, FA needed
    - Helps to stratify risks, prognosis

### CRVO
- Management of acute event is controversial
  - Anti-coagulation
  - Oral pred
  - Triclopidine (platelet aggregation inhibitor)
- Pan retinal photocoagulation may be indicated to reduce the risk of NVG
  - Especially in ischemic
  - Little to no effect on visual outcome
  - NVG can occur even after PRP
CRVO

- Patients with macular edema from CRVO typically do not respond well to FML at all
- CVOS Study: Improvement on appearance, but no gain in acuity
- Newer Treatments:
  - Intravitreal Trimancinolone (SCORE)
  - Intravitreal Lucentis (CRUISE, BRAVO)
  - Others

Steroids

- CRVO SCORE
  - ¼ patients receiving IVT had a 15 letter or better improvement in VA a 12 months
  - Pts 5x as likely to have VA improvement vs. observation alone
- BRVO SCORE
  - Almost equal number of patients in laser or steroid group had > 15 letter improvement
  - More complications in IVT group

VEGF

- CRUISE (CRVO) Study:
  - Vision improved > 15 letters in almost 50% of patients vs. 17% with sham at 6 mos
  - Mean VA gain of almost 15 letters
- BRAVO (BRVO) Study:
  - Vision improved > 15 letters in over 60% of patients vs. 28% with sham
  - Mean VA gain of approx 18 letters
  - Few side effects in either group

VEGF

- Based on these results, Lucentis FDA approved late June 2010 for treatment of macula edema due to retinal vein occlusion
- Two new studies, RABAMES and RELATE, looking at laser vs. Lucentis vs. combo for treatment of macular edema secondary to RVO

Ozurdex

- Ozurdex by Allergan
  - Biodegradable Dexamethasone Intravitreal implant (0.7 mg) indicated for treatment of macula edema from BRVO and CRVO
  - FDA approved June 18, 2009
    - Available 3rd Q of 2009
  - 20-30% of pts with macula edema had 3-line improvement of visual acuity
  - Stable for 1-3 mos
  - Administered as in-office procedure

CRVO

- Management also focuses on diagnosis and management of underlying systemic disease
- At minimum, should have
  - BP evaluated
  - Fasting Blood sugars
  - CBC
  - Lipid profile
### CRVO

- Additional tests might include
  - Carotid artery evaluation
  - Cardiac evaluation
  - Additional blood tests
    - ANA
    - RF
    - FTA/ABS
    - ESR

### CRVO

- Most common etiologies varied with age at presentation
  - Under age 50
    - Head injury
    - Hyperlipidemia
    - Estrogen, esp oral contraceptives
  - Over age 50
    - HTN
    - DM
    - Chronic lung disease

### CRVO

- FA should/may be performed after blood has cleared to determine non-ischemic vs. ischemic
  - Helps evaluate for risk of NVG
- Pt should be seen monthly till resolution, esp for first 3-6 months, esp if ischemic
  - Look for NVI/NVG
- Then periodically, q 4-6 months after resolution to monitor for changes
  - Again NVI/NVG

### CRAO

- Mechanism similar to BRAO, but larger embolus causes obstruction prior to laminar cribosa, so entire central retinal artery is obstructed
- Pts typically present with sudden painless loss of vision in an eye that was previously thought to be healthy
- Typically pts from 50-80 years of age

### CRAO

- Vision typically in the hand motion to counting fingers range
- Most often present with an APD as well
- If a cilioretinal artery is present, there may be a small island of vision that correlates to the area of vascular supply
  - Present in about 10% of eyes
- Can see an embolus in 20-40% of cases

### CRAO

- Early appearance is that of retinal narrowing and haziness of retinal tissue
- After 1-2 hours, retina appears white and edematous, with a "cherry red" macula, representing the choroidal blood supply to the macula
- With time, the arteries may assume a more normal appearance, with irregular narrowing often the only clue
- Optic atrophy may occur, but NVG is very rare
CRAO

- Management often includes attempts to dislodge embolus if pt presents within first 1-2 hours
  - Digital massage, paracentis to lower IOP, carbogen, anti-thrombotic agents, etc have little to no value
- Management lies in diagnosis and management of underlying systemic disease

BRAO

- Result of emboli dislodged from elsewhere which travels through the system until a vessel too small for passage is reached
- Arterial occlusion causes anoxia due to lack of oxygenated blood
  - Anoxia causes loss of retinal layers, including NFL through inner nuclear layer

BRAO

- Occurs most frequently in superior temporal region of the retina
- Visual acuity and field loss dependent on location and extent of blockage
  - VF loss is classically a sharp edged defect stopping abruptly at the horizontal raphe

BRAO

- Appearance varies as time progresses
  - Initially, affected arteries narrow and retina becomes hazy
  - Over a few hours, the retinal tissues whitens and appears edematous
  - Segmental optic atrophy may also develop in the affected area

BRAO/CRAO

- Prognosis depends upon area affected as well as extent of blockage
- Also depends upon prompt therapy, to lesser extent
  - Some studies indicate that if emboli can be dislodged within 1-2 hours, recovery can be complete
  - After this period, initial acuity is not likely to improve
- Immediate ESR needed to r/o GCA if pt over 55
  - Only 2%-5% secondary to GCA in one study
- Most often associated with DM, HTN, and carotid artery disease
  - Many other etiologies including: sickle cell, oral contraceptives, Lupus, Bechets disease, Lyme disease, etc
### BRAO/CRAO
- Blood pressure
- Lab tests
  - FBS
  - CBC
  - ESR
  - Lipid profile
  - PT/PTT
  - ANA/RF
- Carotid Artery Evaluation
- Cardiac Evaluation
  - Echocardiogram and possible Holter monitor

### BRAO/CRAO
- Follow-up
  - BRAO: 3-6 mos after ruling out underlying etiology
  - CRAO: follow closely for first 1-3 mos for NVI, then periodically after
    - If NV, then PRP indicated to prevent NVG

### Retinal Plaques
- Several different types of plaques can often be visualized in the retinal vasculature
- Pt is typically elderly, has HTN, CAD, hypercholesterolemia/hyperlipidemia, and/or atherosclerotic disease
- Often totally asymptomatic and found on routine exam

### Retinal Plaques
- May present with amarosis fugax, transient episodes of monocular blindness
- Rarely, may report transient ischemic attack (TIA), which is above with hemiparesis, parasthesia or aphasia
- Three different types of plaques, but all share strong association to significant cardiovascular disease

### Retinal Plaques
- Cholesterol (Hollenhorst) plaque
  - shiny yellow-orange in appearance
  - typically from the ipsilateral carotid artery
  - Rarely causes occlusion, unless multiple
  - Typically occurs at bifurcations
  - Mobile in nature

### Retinal Plaques
- Calcific
  - Appears more whitish than HH
  - Classically within arteriole, not at bifurcation
  - Typically immobile
  - Often causes BRAO
  - Often from cardiac arethromas of heart valves
Retinal Plaques

• Fibrino-platelet
  – Appear as dull white to gray, long plugs
  – Typically within arterioles, not at bifurcations
  – May break-up and dissolve with time
  – May lead to BRAO or CRAO
  – Often associated with carotid disease or mitral valve insufficiency

Retinal plaques

• Talc retinopathy
  – Represents an exogenous plaques as opposed to others
  – Appears typically as multiple shiny yellow plaques within capillaries in posterior pole
  – Typically smaller than other plaques
  – Typically seen in IV drug users
  – Rarely cause complications, but reported cases of associated NV and occlusions

Retinal Plaques

• No direct management of plaques is needed
• Management is aimed at discovering source of embolus to decrease risk of other emboli, occlusion, or stroke
• Pts need referral to internist for complete physical

Retinal Plaques

• Examination should include
  – Complete physical, including cardiac risk factors and BP evaluation
  – Carotid ultrasound
  – Stress echocardiogram
  – Fasting BS
  – Lipid profiles
  – Cardiac enzymes

Retinal Plaques

• After ruling out underlying etiology, see patient regularly, q 6 -12 mos, to evaluate for additional plaques or other disease associated with vascular disease
  – BRVO/CRVO
  – BRAO/CRAO
  – NTG

Retinal Plaques

• If carotid stenosis or coronary artery disease is found treatment may include
  – Carotid endarterectomy
  – Angioplasty
  – Aspirin therapy
  – Other anti-coagulation therapy, such as coumadin
• Pts with cholesterol HH emboli have 15% mortality at 1 yr, 29% by year 3, and 54% by 7 years
  – Mostly from cardiac disease
Retinal Breaks

• Occur in 3 to 7% of adult population
• Usually asymptomatic
• 1-2% with breaks progress to detachment
• Risk factors include lattice degeneration, high myopia, atrophic holes, aphakia/pseudophakia, and trauma

Procedure

• Laser treatment is used to seal the break by creating adhesion between the retinal tissue and underlying RPE
• Provides barrier to continued enlargement from vitreo-retinal traction and prevents accumulation of subretinal fluid
• Adhesion present 24 hours after surgery, and strengthens over several days

Procedure

• Topical or retrobulbar anesthesia
• Entire lesion should be enclosed by at least 3 rows in a honeycomb pattern

Follow-up

• RTC 1-2 weeks after laser for symptomatic tears
• 3-4 weeks for asymptomatic
• If large or superior, RTC even sooner
• If enlargement or new subretinal fluid, retreat with 1 week follow-up
• RTC 6-8 weeks after initial follow-up
• Yearly thereafter

Complications

• Few complications
  – Inadequate burn intensity, causing ineffective adhesion
  – Possible CNVM
  – Intraretinal hemorrhage
  – Vitreous hemorrhage
  – ERM formation