CASE 1

Case History

• 38 black male, complaining that the vision in his right eye is blurry.
  — Got the current Rx 3 weeks previously, and started out good but in last couple of days OD vision has become blurry
• Medical Hx: no current health concerns and no medications
Entrance Skills

• Va’s: OD: 20/25, OS: 20/20
• Pupils: PERRL
• CVF: full to finger count
• EOM’s: FROM
• Amsler: central metamorphopsia OD
• HVF: 10-2 (see VF)
CASE 2

Lid Nevi

- Lid nevi:
  - congenital or acquired
  - occur in the anterior lamella of the eyelid and can be visualized at the eyelid margin.
- The **congenital eyelid nevus** is a special category with implications for malignant transformation.
- With time, slow increased pigmentation and slight enlargement can occur.
- An **acquired nevus** generally becomes apparent between the ages of 5 and 10 years as a small, flat, lightly pigmented lesion.

Congenital Nevus

- The nevus is generally well circumscribed and not associated with ulceration.
- The congenital nevus of the eyelids may present as a "kissing nevus" in which the melanocytes are present symmetrically on the upper and lower eyelids.
  - Presumably this nevus was present prior to eyelid separation.
Congenital Nevus

- Most nevi of the skin are not considered to be at increased risk of malignancy.
  - However, the large congenital melanocytic nevus appears to have an increased risk of malignant transformation of 4.6% during a 30 year period

Acquired Lid Nevi

- Acquired nevi are classified as:
  - junctional (involving the basal epidermis/dermis junction), typically flat in appearance
  - intradermal (involving only the dermis), tend to be dome shaped or pedunculated
  - compound (involving both dermis and epidermis) tend to be dome shaped

CHRPE vs Nevus
Nevi Trivia

• 31% of choroidal nevi show slight enlargement over time without the transformation to a melanoma (Ophthalmology 2011)

• The prevalence of choroidal nevi in the white U.S. population ranges from 4.6% to 7.9%
  – If it is assumed that all choroidal melanomas arise from preexisting nevi, then the published data suggest a low rate (1/8845) of malignant transformation of a choroidal nevus in the U.S. white population. (Ophthalmology 2005)

• Choroidal melanoma risk for metastasis, ranging from 16% to 53% (at 5 years of follow-up) depending on the size of the tumor at the time of diagnosis. (Arch Ophthalmol 1992)

TFSOM—“To Find Small Ocular Melanoma”

**Thickness:** lesions >2mm

**Fluid:** any subretinal fluid (suggestive of serous retinal detachment)

**Symptoms:** photopsia, vision loss

**Orange pigment overlying the lesion**

**Margin touching optic nerve head (<3mm)**

• None of these factors = 3% risk of a nevus converting to melanoma in five years.
  One of these factors = 8% risk of conversion in five years.
  Two or more factors = 50% risk of conversion in five years.
  For any changes noted during the course of follow-up, refer the patient to a retinal practice or an ocular oncology service.

TFSOM-UHHD:

“To Find Small Ocular Melanoma Using Helpful Hints Daily”

**Thickness:** lesions >2mm

**Fluid:** subretinal fluid

**Symptoms:** photopsia, vision loss

**Orange pigment overlying the lesion**

**Margin touching optic nerve head (<3mm)**

**Ultrasound Hollowness**

**Halo absence**

**Drusen absence**

• Choroidal nevi showing no features should be initially monitored twice yearly and followed up annually
  
  • 1 or 2 features should be monitored every 4 to 6 months.

  • Nevus with 3 or more features should be evaluated at an experienced center for management alternatives and possible treatment owing to the high risk of ultimate growth.
Case

- 65 yr old white male
  - Notices spot in vision in his left eye
  - Diabetes for 15 years
- Vision: 20/20 (6/6) and 20/40 (6/12)
- Dilated exam:
  - Large lesion noted in left eye (not noted in exam 6 months previously
  - See photo and B-scan
Ocular Tumors

Astrocytic Hamartoma  Amelanotic Melanoma

Retinoblastoma  Metastatic Choroidal Tumor

Choroidal Melanoma Metastases

• 80 to 90% of metastases from uveal melanoma occurred in the liver, less common sites being the skin and lung.

Melanoma and Mortality

• Tumor Size:
  – 5-year mortality after enucleation:
    • 16% for small melanoma,
    • 32% for medium melanoma, and
    • 53% for large melanoma.
  – the prognostic importance of tumor size:
    • each 1-mm increase in melanoma thickness adds approximately 5% increased risk for metastatic disease at 10 years

• Tumor genetics:
  – Chromosome monosomy 3 (approx 50% of patients)
    • 50% of them develop metastasis within 5 years of diagnosis
    • 70% mortality within 4 years of ocular treatment
    • one of the most important independent risk factors of poor survival
CASE 3

30 YR WM

• Patient calls from his PCP office asking if we can see him today because he has had red/painful eyes for over a week and has not resolved
• Medical history:
  – Past week has been experiencing painful urination and discharge
  – New sexual partner approx 10 days ago, who also had developed a red eye
  – Chlamydia and gonorrhea testing were negative
  – Has tested positive for HSV2 but no current flare up

30 YO WM

• Medications:
  – In the past week patient:
    • 2 courses of azithromycin (1 gram each)
    • Injection of rocephin
    • Injection of penicillin G
    • Currently taking doxycycline 100 mg bid
    • Valtrex 1 gram 3 times per day for 7 days (d/c 1 day ago)
    • Was on Vigamox qid for 7 days (d/c 1 day ago)
• VA: 6/7.5 (20/25) OD, OS
• Entrance skills unremarkable though some pain on eye movement
30 YO WM

• SLE:
  – 2+ injection conjunctival both eyes
  – 1-2+ lid edema
  – Mixed papillary and follicular response
  – 1-2+ diffuse SPK (no staining noted above infiltrates)
  – No cells or flare noted

30 YO WM

• AdenoPlus:
  – Performed on the right eye (patient felt that was the worst eye)
  – Negative

30 YO WM

• Started patient on the miracle drop
  – Tobradex 4 times per day and scheduled patient to come back the next day
• 1 day f/u
  – Patient was feeling better
  – Less redness and much reduced photophobia and discomfort
  – No improvement on painful urination or discharge and is now seeing blood in his urine
  – Continue tobradex 4 times per day and RTC in 4 days for f/u with dilation and told to contact PCP to update on the blood in the urine
30 YO WM

- 4 day f/u:
  - Patient says his eyes are doing great and that all of his urogenital problems abruptly stopped on Saturday
  - Discussion with PCP: Kidney stone
  - What was going on with the eye?
    - Viral conjunctivitis likely EKC

What did we learn from this?

CASE 4

Case

- 50 YR WM
- POHx: had cataract surgery in his left eye at age 25 secondary to trauma to the eye,
  - Has a mid-dilated pupil post trauma
- PMHx: no known health problems and no medications
- VA: 6/6 (20/20) OD, OS
Health Assessment

• SLE:
  – OD unremarkable
  – OS: mid-dilated pupil with sluggish response to light
    • PCIOL well centered and no haze
• IOP: OD 12 and OS 26 mm Hg (TAG)
  • NCT OS (31 and 23)
  • Second visit: OD: 13 and OS: 27

Health Assessment

• Gonioscopy:
  – OD: unremarkable
  – OS: see photo

Optic Nerves

OS

OD
Visual Fields

Ganglion Cell Analysis

RNFL and ONH Analysis
CASE 5

Case

- 65 year old Caucasian patient presents with sudden onset loss/blurring of vision in the right eye
- PMHx: HTN for 15 years, takes “water pill”
- VA’s: 20/60 OD, 20/25 OS
- Pupils: PERRL – APD
- CVF: Inferior defect right eye, no defects noted in the left eye

Vision Loss Without Pain: Diabetes/Diabetic Retinopathy

- Microvascular complications resulting in capillary closure & abnormal permeability
- S&S include:
  - blurring of vision (maculopathy and refractive error shifts),
  - sudden drop in vision (vitreous heme),
  - dot and blot hemes,
  - exudate,
  - cotton wool spots,
  - neovascularization (iris, retina and disc)
Diabetic Retinopathy

CSME (DME)

CSME (DME) OCTA

VEGF and DME
Aug. 10, 2012: FDA approves Lucentis to treat diabetic macular edema

- The drug’s safety and effectiveness to treat DME were established in two clinical studies involving 759 patients who were treated and followed for three years.
  - patients were randomly assigned to receive monthly injections of Lucentis at 0.3 milligrams (mg) or 0.5 mg, or no injections during the first 24 months of the studies
  - after 24 months, all patients received monthly Lucentis either at 0.3 mg or 0.5 mg
- Results:
  - 34.45% of those treated with monthly Lucentis 0.3 mg gained at least three lines of vision compared with 12-18% of those who did not receive an injection.

Vision Loss Without Pain: Vein Occlusion

- Associated with:
  - hypertension,
  - coronary artery disease,
  - DM and
  - peripheral vascular disease.
- Usually seen in elderly patients (60-70), slight male and hyperopic predilection.
- Second most common vascular disease after diabetic retinopathy.

Branch Retinal Vein Occlusion: Signs/Symptoms

- BRVO: sudden, painless, visual field defect.
  - patients may have normal vision.
  - quadrantic VF defect,
  - dilated tortuous retinal veins with superficial hemes and CWS
  - typically occurs at A/V crossing (sup/temp)
BRVO

- BRVO more common than CRVO and has more favorable prognosis
  - Overall 50-60% of BRVO patients will maintain VA of 20/40 or better
- Visual loss results from:
  - Macular edema
  - Foveal hemorrhage
  - Vitreous heme
  - Epiretinal membrane
  - RD
  - Macular ischemia
  - Neovascularization complications

http://www.healio.com/ophthalmology/journals/osli/

Study Design (n=397) BRVO

- BRVO retinal Vein Occlusion study safety/efficacy
- 1:1:1 Randomization
- Ranibizumab 0.3 mg
- Ranibizumab 0.5 mg
- Ranibizumab 0.3 mg
- Rescue Laser (if eligible beginning at Month 3)
- PNR ranibizumab for all patients Rescue Laser (if eligible beginning at Month 3)
- Month 6 Primary Endpoint
Mean Change from Baseline BCVA

**BRVO**

The gain of additional 3 lines occurred at a rate of 61% of 0.5 mg AVT grp, 55% for 0.3 mg AVT & 29% placebo.

Central Retinal Vein Occlusion: Signs/Symptoms

- CRVO: thrombus occurring at lamina is classical theory but new evidence indicates that the occlusion is typically in the optic nerve posterior to the lamina cribrosa.
  - decreased VA ranging from near normal to hand motion with majority 20/200 range
  - dilated tortuous vessels, with numerous retinal hemes and CWS

Central Retinal Vein Occlusion

- Visual morbidity and blindness are primarily from:
  - persistent macular edema,
  - macular ischemia and
  - neovascular glaucoma
Central Retinal Vein Occlusion

- CRVO's can be ischemic or non.
  - Classical definition of ischemic is 10-disc area of non-perfusion found on angiography
  - RAPD and ERG maybe better predictor
  - VA's typically worse in ischemic
  - Increased number of cotton wool spots with decreased VA maybe predictive

Central Retinal Vein Occlusion

- Ischemic CRVO may lead to iris neovascularization and neovascular glaucoma
  - Estimated apprx 20% of CRVO's are ischemic with 45% of those developing neo
- Regular examinations (1-2 wks) to monitor for ischemia or neo development
  - should include gonio as angle neo can precede iris rubecosis

Study Design CRUISE (n=392)

- 1:1 Randomization
- Sham (n=130)
- Ranibizumab 0.3 mg (n=130)
- Ranibizumab 0.5 mg (n=130)

Monthly Injections (Last at 114M) M to Period

RUNOUTS available for all patients: 1M to period

- 0.5 mg
- Ranibizumab 0.3 mg
- Ranibizumab 0.5 mg

Month & Primary Endpoint
Vision Loss Without Pain: Artery Occlusion

- Primarily embolic in nature from cholesterol, calcifications, plaques.
- Usually occurs in elderly associated with:
  - Hypertension (67%),
  - Carotid occlusive disease (25%),
  - DM (33%) and
  - Cardiac valvular disease.
- Sudden loss of unilateral, painless vision
  - Defect dependent upon location of occlusion

Vision Loss Without Pain: Artery Occlusion

- BRAO typically located in temporal retinal bifurcations.
CRAO

- CRAO has profound vision loss with history of amaurosis fugax.
  - Vision is usually CF (count fingers) to LP (light perception) with positive APD.
  - Diffuse retinal whitening with arteriole constriction, cherry red macula.

Ophthalmic Emergency

- Treatment is controversial due to poor prognosis and questionable benefit.
- Treat immediately before workup, if patient presents within 24 hours of visual loss:
  - Digital ocular massage,
  - systemic acetozolamide (500 mg IV or po),
  - topical ocular hypertensive drops (lopidine, B-blocker),
  - anterior chamber paracentesis,
  - consider admission to hospital for carbogen Tx (high carbon dioxide)

Macular hole

- Unilateral, decreased vision
  - Often in 60-80 year old women
  - Anyone w/ a history of trauma
- Symptoms:
  - Decreased vision, metamorphopsia
    - 20/200 for full thickness holes
- Signs:
  - Red hole in the macula
  - (+) Watzke-Allen sign
Macular hole

- Stages
  - Stage 1a -> impending hole. Normal foveal depression with yellow spot/dot in fovea.
  - Stage 1b -> Abnormal foveal depression with yellow ring.

Stage 1b macular hole

Macular hole

- Stages
  - Stage 2 -> Small full-thickness hole. 20/80 - 20/400.
  - Stage 3 -> Full-thickness hole w/ cuff of SRF. No PVD
  - Stage 4 -> Full-thickness hole with cuff of SRF, with complete PVD.

Stage 2 macular hole

Macular hole

- Stages
  - Stage 2 -> Small full-thickness hole. 20/80 - 20/400.
  - Stage 3 -> Full-thickness hole w/ cuff of SRF. No PVD
  - Stage 4 -> Full-thickness hole with cuff of SRF, with complete PVD.

Stage 3 Macular hole

Stage 4 macular hole
New Macular Hole Staging

Table 2: Correlation between Commonly Used Clinical Macular Hole Staging and the International Vitreomacular Traction Study Classification System for Vitreomacular Adhesion, Traction, and Macular Hole

<table>
<thead>
<tr>
<th>Full-Thickness Macular Hole Stages in Common Use</th>
<th>International Vitreomacular Traction Study Classification System</th>
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<tbody>
<tr>
<td>Step 1: Intact macular hole</td>
<td>VMA</td>
</tr>
<tr>
<td>Step 2: Small hole</td>
<td>VMT</td>
</tr>
<tr>
<td>Step 3: Large hole</td>
<td>Small or medium FTMH with VMT</td>
</tr>
<tr>
<td>Step 4: FTMH with PVD</td>
<td>Medium or large FTMH with VMT</td>
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Small FTMH w/o traction

B

154 microns

237 microns
### New Macular Hole Staging

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<td>VH/MV</td>
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<tr>
<td>Step 2: small hole</td>
<td>Small or medium FTMH w/o VAT</td>
</tr>
<tr>
<td>Step 3: large hole</td>
<td>Medium or large FTMH w/ VAT</td>
</tr>
<tr>
<td>Step 4: FTMH with PVD</td>
<td>Small, medium, or large FTMH without VAT</td>
</tr>
</tbody>
</table>

**Image C:** Medium FTMH w/o traction

**250-400 microns**

**Image D:** Large FTMH with traction

**> 400 microns**

### Case 6
13 YR Female

CC: noticed that her left eye became blurry and objects were “wavy” a couple of days ago. Sudden onset and she had experienced a headache over the left eye just prior to the vision going blurry.

Ocular Hx: she currently wear glasses for distance

Medical Hx: she is currently not diagnosed with any health problems and is not taking any medications

Entrance Skills

VA with current Rx: 20/30 OD and 20/30 OS

Entrance skills unremarkable

Amsler: metamorphopsia OS

BCVA: 20/20 OD with increased minus, no improvement possible in the left eye

IOP's: 13 mm Hg OD and OS

Fundus Photos
Retina Consult

- Referred patient to retina and they confirmed the diagnosis of VKH.
- She was begun on oral prednisone 60 mg per day and she was re-evaluated in 1 week.
- At the follow up, there was reduction in her serous retinopathy and vision was improved.

From the Experts

- Vogt-Koyanagi-Harada (VKH) disease is a multisystemic disorder characterized by granulomatous panuveitis with exudative retinal detachments that is often associated with neurologic and cutaneous manifestations.
- VKH disease occurs more commonly in patients with a genetic predisposition to the disease, including those from Asian, Middle Eastern, Hispanic, and Native American populations.
From the Experts

• VKH:
  – Patients have no prior history of ocular trauma or surgery
  – Patients have no evidence of another ocular disease based on clinical or laboratory evidence
  – Patients have bilateral ocular involvement.

From the Experts

• VKH:
  – The neurologic and auditory signs include the following:
    • Malaise, fever, headache, nausea, abdominal pain, stiffness of the neck and back, or a combination of these factors; headache alone is not sufficient to meet the definition of meningitis
    • Tinnitus
    • Cerebrospinal fluid pleocytosis
  – Integumentary signs include the following:
    • Alopecia: loss of body hair
    • Poliosis: loss of pigment in hair
    • Vitiligo: loss of skin pigmentation in blotchy pattern

VKH Treatment

• For most patients with bilateral serous detachments and severe visual loss, begin therapy with systemic prednisone (1-2 mg/kg/day).
• The length of treatment and subsequent taper must be individualized for each patient.
  – Most patients require therapy for 6 months and occasionally up to 1 year before successful tapering of systemic corticosteroids.
  – Systemic therapy should not be discontinued during the 3 months following the onset of the disease because of the risk for recurrence.