Retinal Cases: Not rare if it’s in your chair

KELLEN KASHIWA, OD
BENNERT EYE INSTITUTE

Goals
- Review cases of uncommon retinal diseases
- Discuss differential diagnoses
- Evaluate retinal diagnostic test and how they can help with diagnosis

Case 1
- 19 yo AF – CC: Blurry vision at near, annual visit
- VA: 20/20 OD, OS, OU
- IOP: 12, 13 mmHg
- Anterior segment: Unremarkable
- Posterior segment: Next Slide

Fundus Photo

Differential Diagnosis
- Diabetic Retinopathy
- Hypertensive Retinopathy
- Vein occlusion
- Vasculitis
- HIV Retinopathy
- Leukemia
- Anemia
- Preeclampsia

Roth Spots
- White centered round, flame-shaped retinal heme
- First seen in patient with subacute bacterial endocarditis
- Lab testing if suspicion for endocarditis includes
  - CBC with differential
  - Erythrocyte sedimentation rate
  - C-reactive protein
  - Blood cultures
### Lab Results

<table>
<thead>
<tr>
<th>Test</th>
<th>Patient 1</th>
<th>Normal Female</th>
<th>Normal Male</th>
</tr>
</thead>
<tbody>
<tr>
<td>ESR</td>
<td>4 mL per hour</td>
<td>0–20 mL per hr</td>
<td>0–15 mL per hr</td>
</tr>
<tr>
<td>CRP</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CBC</td>
<td>RBC: 65g/L</td>
<td>WBC: 109 x 10^9</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Platelet: 40.2 x 10^9/L</td>
<td>&lt;250 U/L</td>
<td></td>
</tr>
<tr>
<td>Serum lactate dehydrogenase</td>
<td>950 U/L</td>
<td>&lt;250 U/L</td>
<td></td>
</tr>
</tbody>
</table>

### Leukemia Retinopathy

- **Leukemia Retinopathy**
  - Most case diagnosed prior to eye exam
  - In a 2020 review, 25% of leukemia cases were diagnosed after eye exam
- **Posterior segment manifestations**
  - Secondary to direct invasion of leukemic cells
  - Retinal grayish white nodules surrounded by hemorrhage
  - Roth spots – retinal hemorrhage with white-center
  - Pale gray swelling of the optic nerve head

### Case 2

- 32 yoAF
- Sudden vision loss OD
- Vision: OD: 20/40; OS: 20/30
- IOP: OD: 13; OS: 13mmHg OU
- Anterior segment: Unremarkable, NSC OU
- Posterior segment: Next Slide

### Fundus Photos

### Differentials

- Diabetic Retinopathy
- Hypertensive Retinopathy
- BRVO
- Purtscher retinopathy

### Purtscher Retinopathy

- **Similar findings to DR**
  - Retinal hemorrhage (65%)
  - Retinal whitening
  - Cotton wool spots (93%)
  - Optic disc edema
  - Purtscher flecken (areas of inner retinal whitening)
- **Mostly associated with trauma**
- **Typically bilaterally seen**
In absence of trauma other causes:
- Acute pancreatitis
- Renal failure
- Preeclampsia and childbirth
- Connective tissue disorder
- Valsalva maneuver

Case 3
- 43 year old Hispanic female
- CC: sudden vision loss OU
- Hx: Hypertension
- BCVA: OD: 20/400 OS: CF @1'
- IOP: 12, 13mmHg
- Anterior segment: MGD, s/p LASIK, NSC trace OU
- Posterior segment: Next Slide

Findings
- Bilateral optic nerve head edema grade 4+
- Scattered Exudates
- Subretinal fluid

Differential Diagnosis
- Papilledema
- CRVO
- Optic neuritis
- Diabetic papillopathy
- Hypertensive disc edema
- NAION
- AION
- Compressive optic neuropathy
- Posterior scleritis
Additional Testing

- Blood pressure
- Hemoglobin A1C/Serum glucose
- Fasting lipid profile
- If secondary hypertension suspected:
  - Urinary cortisol – Cushing’s
  - Plasma renin to aldosterone ration – hyperaldosteronism
  - Angiography – renal vascular disease

Results

- In office blood pressure 210/104 mmHg
- Sent to Emergency room for immediate hypertensive crisis treatment

Posterior Reversible Encephalopathy Syndrome

- Rapid onset of symptoms
- Headaches
- Seizures
- Vision loss/disturbance
- Often due to acute hypertension
- Chronic/Acute kidney disease/injury
- Pre-eclampsia

Treatment

- Hypertensive control -> Stent to circumflex artery
  - Blood pressure next visit 128/82mmHg
- Optic nerve head neuroprotection
  - Alphagan BID OU for optic nerve head protection
- Macular edema
  - Subtenon kenalog for macular edema
  - VA responded
    - 2 wks post from 20/400 -> 20/60
    - 4 wks post 20/60 -> 20/40
    - 8 wks post 20/40 ->20/20!

Case 3

- 54 year old Brazilian female
- CC: blurred reading vision
- 20/20 OD, OS, OU
- IOP: 13, 14
- Recent diagnosed with pneumonia and recovered
- Anterior Segment: Pterygium, mild bleph
- Posterior segment:
Cotton Wool Spots

- Not a normal finding
- In a series of patients with CWS
  - Diastolic BP was >90mmHg
  - Elevated blood sugar in 20%
- Secondary to ischemia from retinal arteriole obstruction
- Eosinophilic segments of ganglion cell axons that are swollen – defective axoplasmic flow

Etiology

- Ischemic: HTN, Diabetes, RVO
- Infectious: HIV retinopathy, Cat-Scratch
- Idiopathic
- Trauma
- Medication: Interferon Retinopathy
- Inflammatory: Lupus, GCA

COVID

- COVID-19 caused by SARS-CoV-2 uses the ACE2 receptor to gain entry into cells
- ACE2 receptor is widely expressed in multiple organs – including the retina
- Coronavirus have been shown to cause optic neuritis and retinitis

Conjunctivits

- Hyperreflectivity at the Ganglion cell layer and Inner plexiform layers
- Cotton-wool spots and microhemorrhages seen
- Animal model suggest retinitis and optic neuritis can be seen

- 7% of COVID-19 patients had viral RNA in the conjunctival secretions
- Autoimmune response
- Possible macrophage activation syndrome
Case 4

- 18 yoAM complains of sudden decreased vision and photophobia.
- BCVA
  - OD: 20/50
  - OS: 20/400
- IOPs
  - OD: 11mmHg
  - OS: 12mmHg
- Ant Seg: 3+ cells/flare in the anterior chamber and vitreous OU. Keratic precipitates OU and Koepppe nodules OU.
- Post Seg: See next slides.
- General Health is excellent. No recent illness/malaise, systemic changes or medications.

Findings:
- Panuveitis
- Bilateral serous RDs
- Multiple hyperfluorescent dots at RPE

Differentials???
Differential Diagnosis

- Sympathetic ophthalmia
- Vogt-Koyanagi-Harada Syndrome
- Posterior scleritis
- Acute Posterior Multifocal Placoid Epitheliopathy (APMPPE)
- Syphilis
- Tuberculosis
- Behcet’s disease
- Multiple White Dot Syndrome
- Lupus
- Sarcoidosis
- Ocular Lyme Disease

Vogt-koyanagi-Harada

- Autoimmune Disease
- Higher Rate in Asian, Latin, Mediterranean descent
- Chronic, bilateral granulomatous uveitis
- Disease vs. Syndrome

Triad
- Alopecia
- Poliosis
- Vitiligo

Treatment
- Systemic Corticosteroids
- InVit / SubTenon Corticosteroids
- Surgery

Case 5

- 39 year old Filipino male
- CC: Cloudy, FB sensation OS, chef – rubbed his eyes with gloves and may have got something in it
- VA: OD: 20/20 OS: 20/60
- Anterior segment:
  - OS: Arcus, 3+ KPs, 3+ cells / 2+flare
- Posterior segment:
  - OS: Vitreal cells

OPTOS

Findings

- Panuveitis
- RPE tracks with white lesions
- Peripheral subretinal FB

Further questioning reveals
- Often consumes raw meats and poultry!

Differential Diagnosis

- Sarcoidosis
- Focal chorioretinitis
- Toxoplasmosis
- Histoplasmosis
- Multifocal choroiditis
- Acute multifocal placoid pigment epitheliopathy
- MEWDs
- DUSN
- Ophthalmomyiasis
Ophthalmomyiasis Interna

- Parasitic disease affecting the outer retinal and RPE
- Nematode
  - Strongyloides stercoralis

Treatment
- Immediate photocoagulation
- Patient is at risk for a significant inflammatory response once larva dies
- Careful monitoring and prompt treatment of inflammation is essential
- Albendazole 400mg
- Pred forte QID

Outcome
- Followed closely and treated with oral antihelminthic for months
- Vision returned to 20/20 OS!

CASE 6
- 72 YOWF CC: Sudden, painless vision loss OD
- BCVA
  - OD: 20/200
  - OS: 20/30
- IOP
  - OD: 14mmHg
  - OS: 15mmHg
- Ant Seg: Unremarkable other than 2+ NSC OU
- Post Seg: See following slides
- Medical Hx: Thyroid cancer 20+; Treated 1980s
- Medication: None

Optos and RNFL OD
Optos and RNFL OS

Findings

<table>
<thead>
<tr>
<th>Findings</th>
<th>Differentials??</th>
</tr>
</thead>
<tbody>
<tr>
<td>Optic nerve head edema OD</td>
<td>AION</td>
</tr>
<tr>
<td>Optic nerve pallor OS</td>
<td>NAION</td>
</tr>
<tr>
<td>Slight Attenuation</td>
<td>Papillitis</td>
</tr>
<tr>
<td>AION</td>
<td>Multiple Sclerosis</td>
</tr>
</tbody>
</table>

Differential Diagnosis?

<table>
<thead>
<tr>
<th>Differentials??</th>
<th>Additional Testing?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Foster Kennedy Syndrome</td>
<td>Imaging – CT/MRI</td>
</tr>
<tr>
<td>Papilledema</td>
<td>CSF</td>
</tr>
<tr>
<td>Pseudotumor</td>
<td>CRP vs Sed Rate</td>
</tr>
<tr>
<td>AION</td>
<td>Carotid Dissection</td>
</tr>
<tr>
<td>NAION</td>
<td></td>
</tr>
</tbody>
</table>

Diagnosis of Exclusion

**PSEUDO – FOSTER KENNEDY SYNDROME**

<table>
<thead>
<tr>
<th>Findings</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Optic nerve head edema in one eye</td>
<td>Similar to GCA</td>
</tr>
<tr>
<td>Optic nerve head pallor in the fellow eye</td>
<td>High level IV steroids</td>
</tr>
<tr>
<td>No mass / tumor underlying cause</td>
<td>Followed by oral steroids</td>
</tr>
<tr>
<td>Active NAION with old NAION in fellow eye</td>
<td>Carotid dissection</td>
</tr>
</tbody>
</table>

Case 7

- 65 year old male with reduced VAs
- OD 20/20→20/50
- OS 20/20→20/60
- Retinal exam and OCT show no intraocular fluid
- MP1 shows significant reduced central sensitivity

**Case 7**

- MP1 shows central loss, decreased sensitivity
- OCT shows RPE changes
- ERG shows reduced high contrast function (photopic)
What’s your differentials?
- Cone-Rod dystrophies
- Leber’s congenital amaurosis
- Stargardt disease
- Bardet-Biedl syndrome
- Refsum disease
- Batten disease
- Spinocerebellar axtaia type 7

Cone Dystrophy
- Vision loss ranges from 20/40 to CF
- Color vision loss
- RPE loss
- ERG best test for diagnosis
- Treatment
  - Increased anti-oxidant supplementation
  - Beta-carotene
  - Increase omega 3 fatty acid consumption
  - Electrical current stimulation?

Electrical Current Therapy
- Frequency specific microcurrent electrical stimulation
- RPE function

MiraGel Implant

Coat’s Disease
- Splitting of OPL
- Typically in the macula
- VA ranges 20/200 – CF
- Responds very well to magnification
- No Treatment

Juvenile Retinoschisis
Von Hippel Lindau

Choroidal Melanoma

Pars Planitis

Histoplasmosis

Histoplasmosis

Mahalo For Coming!

- Please feel free to contact me if you have any questions
- kkashiwa@retinahawaii.com
- 808-398-3766
- www.bennetteyeinstitute.com