Case Conference
April 17, 2009
Massachusetts Eye Research and Surgery Institution

Raafay Sophie
Case

• **CC:** ”My vision in the right eye has been bothering me for a month.”

• **HPI:** A 42 year old male with a history of increasing central vision loss OD that had now progressed to a persistent blind spot in the center with peripheral vision preserved.

• No pain, redness, floaters or flashes.
History

- Pulmonary complaints - coughing, difficulty breathing and wheezing episodes
- Diagnosed with bronchitis and treated
- Then diagnosed with sinus infection and treated with antibiotics
- Finally diagnosed with asthma and started on asthma treatment.
- Subsequently developed chest pain which was attributed to "a cracked rib" from coughing.
History

• For the past 3-4 months
  – Fevers
  – Night sweats
  – Fatigue
  – Poor appetite
  – Weight loss
  – Frequent nose bleeds
Family History

• Relatives had “problems” with
  – Eyes
  – Lungs

• History of “Cancer in the family”
Social History

- Had only lived in US
- Had owned cats and dogs
- No history of tobacco use
- No history of IV drug use
- Occupation: carpenter
Past Medical History

• Blood transfusion 24 years ago.
• Stiff lower back secondary to injury over the years.
• Chicken Pox
• Chlamydia infection
• No known allergies
Examination

Pt alert & oriented to person, place and time
Vitally stable except for heart rate of 108

Visual Acuity

<table>
<thead>
<tr>
<th></th>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acuity</td>
<td>Dva</td>
<td>Dva</td>
</tr>
<tr>
<td></td>
<td>sc 20/100</td>
<td>sc 20/25</td>
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</tbody>
</table>

Intraocular Pressure

<table>
<thead>
<tr>
<th></th>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pressure</td>
<td>10mmHg</td>
<td>10mmHg</td>
</tr>
</tbody>
</table>

Pupils

|       | OD: equal, round, reactive, no APD | OS: equal, round, reactive, no APD |

Conjunctiva:

|       | OD: normal | OS: normal |

Cornea:

|       | OD: clear and compact | OS: clear and compact |

Iris:

|       | OD: normal |

Anterior Chamber:

|       | OD: deep and quiet | OS: deep and quiet |

Lens:

<p>|       | OD: clear | OS: clear |</p>
<table>
<thead>
<tr>
<th>Examination</th>
<th></th>
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<tbody>
<tr>
<td><strong>Vitreous:</strong></td>
<td></td>
</tr>
<tr>
<td>OD: vitreous debris with trace cells</td>
<td></td>
</tr>
<tr>
<td>OS: trace cells</td>
<td></td>
</tr>
<tr>
<td><strong>Optic Nerve:</strong></td>
<td></td>
</tr>
<tr>
<td>OD: hyperemia/disc edema +1</td>
<td></td>
</tr>
<tr>
<td>OS: hyperemia/trace disc edema</td>
<td></td>
</tr>
<tr>
<td><strong>CD Ratio:</strong></td>
<td></td>
</tr>
<tr>
<td>OD: .3</td>
<td></td>
</tr>
<tr>
<td>OS: .3</td>
<td></td>
</tr>
<tr>
<td><strong>Macula:</strong></td>
<td></td>
</tr>
<tr>
<td>OD: edema/central hemorrhages</td>
<td></td>
</tr>
<tr>
<td>OS: focal hemorrhages</td>
<td></td>
</tr>
<tr>
<td><strong>Vessels:</strong></td>
<td></td>
</tr>
<tr>
<td>OD: increased tortuosity</td>
<td></td>
</tr>
<tr>
<td>OS: increased tortuosity</td>
<td></td>
</tr>
<tr>
<td><strong>Retina:</strong></td>
<td></td>
</tr>
<tr>
<td>OU: scattered Roth’s spots and hemorrhages with ghost vessels</td>
<td></td>
</tr>
</tbody>
</table>
Roth Spots!

• Subacute bacterial endocarditis (SBE)
• Leukemias
• Anemia
• HIV
• Anoxia
• Carbon monoxide poisoning
• Prolonged intubation during anaesthesia
• Hypertensive retinopathy
• Pre-eclampsia
• Diabetic retinopathy
• Neonatal birth trauma
• Mothers who have undergone traumatic deliveries
• Battered children / shaken baby syndrome
• Intracranial hemorrhage from arteriovenous malformation
• Ocular decompression following trabeculectomy
• Kala azar
History of Roth Spots

- In 1849, William Bowman noted “Ophthalmitis...accompanying extensive inflammation of the heart and brain.
- In 1856, Rudolf Virchow linked suppurative retinitis with emboli from cardiac valvular disease.
- In 1872, Moritz Roth described “retinitis septica” in patients who with bacteremia.
- In 1878, Litten assigned the name 'Roth spot' to these white-centered retinal hemorrhages.
Histopathology of Roth Spots

• Early investigators could not find any definite aggregations of bacteria and leukocytes in the white centre of the lesions in specimens of patients who died of sepsis.
• Recent investigators have consistently found fibrin deposits in these lesions.
Pathophysiology of Roth Spots

Rupture of retinal capillaries
  Extrusion of whole blood
  Platelet adhesion and activation
  Coagulation cascade
  Platelet-fibrin thrombus.
Pathophysiology of Roth Spots

• Thrombocytopenia- SBE and leukemia
• Ischemic insults- anemia, anoxia, carbon monoxide poisoning
• Increased capillary fragility- Hypertension, pre-eclampsia and diabetes
• Elevated venous pressure- neonatal birth trauma, traumatic deliveries in mothers, battered baby syndrome and intracranial hemorrhages
**Figure 1.** Behçet's disease with multiple systemic thrombotic manifestations (notice disk involvement).

**Figure 2.** Idiopathic aplastic anemia (white blood cell count, 200; hemoglobin, 5.9; platelet count, 6000).

**Figure 3.** Acute myeloblastic leukemia.

**Figure 4.** A 40-year-old woman with rheumatic heart disease and Streptococcus viridans endocarditis. Three lesions demonstrate, from left to right, evolution of a Roth spot.
Roth Spots

• Roth spot is a **morphological manifestation** of retinal capillary rupture and the ensuing reparative process.

• Can occur in a **variety of conditions**.
Assessment

• Posterior Uveitis OU - vision threatening OD>OS with significant vasculitis and hemorrhagic changes OU, macular edema OD>OS

• Immediate hospitalization at MGH
  – ID consult and empiric intravenous antibiotic therapy
  – Labs including blood cultures, HIV testing, thin film
Labs

• MERSI.
  – WBC so high could not be measured
  – ESR >130 mm/min

  – Smear
Labs

• CBC:
  – WBC 694,000 (4,800-10,800)
  – HB 10.4 (14-18)
  – PLT 525 (150-400)

• Glucose 118 (67-106)
• Electrolytes - normal
• Bleeding profile – normal
• Homocysteine 18.7 (0-11.3)
Labs

- LFTS - normal
- Hep B - normal
- Hep C - normal
- Cardiolipin - normal
- Uric acid
- Mg
- UCS negative
- UDR negative
- Blood cultures negative.
Labs

- C-reactive protein 1.2 (0-0.7)
- Total complement
- ANA
- ACE
- Immune complex c1q
- IL6 7.21 (0.31-5)
- Tnf alpha
- C3d immune complex
- FTA-ABS
- MPO-ABS
- Properdin factor b
- ANCA vasculitides
- C3
- C4
Further Management

- Admitted to MGH where he had an infectious diseases and hematological consult-diagnosed with CML
- Started on chemotherapy- Allopurinol 300mg daily, Gleevac 400mg daily.
- Came to clinic after 2 weeks of treatment stating “I feel great”
## Examination

### Visual Acuity
- **OD:** Dva sc 20/80
- **OS:** Dva sc 20/20

### Intraocular Pressure
- **OD:** 12mmHg
- **OS:** 10mmHg

### Pupils
- **OD:** equal, round, reactive, no APD
- **OS:** equal, round, reactive, no APD

### Conjunctiva:
- **OD:** normal
- **OS:** normal

### Cornea:
- **OD:** clear and compact
- **OS:** clear and compact

### Anterior Chamber:
- **OD:** deep and quiet
- **OS:** deep and quiet

### Iris:
- **OD:** normal

### Lens:
- **OD:** clear
- **OS:** clear
## Examination

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<tr>
<th>Vitreous:</th>
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<tr>
<td>OD: clear</td>
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<tr>
<td>OD Vertical/horizontal .4/.4</td>
<td>OD: improved hemorrhages</td>
</tr>
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<td>OD Vertical/horizontal .4/.4</td>
<td>OS: improved hemorrhages</td>
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Progress

• Called for Follow up in one month but did not return

• We spoke to him last week
  – Reports having visited his ophthalmologist who told him his hemorrhages have almost completely resolved
  – His VA is 20/25 OD and 20/20 OS.

• He is apologetic he could not return, but he is busy with his new job. He says he is very grateful to us here at MERSI and refers to us as “the people who saved my life”
References

• Roland Ling, Bruce James, White-centred retinal haemorrhages (Roth spots), Postgrad Med J. 1998; 74: 581-582.


• Mark E. Silverman et al., Extracardiac Manifestations of Infective Endocarditis and Their Historical Descriptions, Am J Cardiol. 2007 Dec 15; 100(12): 1802-7.
Acknowledgments

• Thanks to John Mauro and David Hinkle!