Review of Systems
Live and Unplugged!
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Financial Disclosure
• This course is based on a column that I co-author in Review of Optometry, for which I receive an honorarium.
• I have no proprietary interest in any products.

Course Goal
• To provide the participant with useful clinical information about caring for patients living with oculosystemic disease.

QUESTIONS AND COMMENTS?
Pizzi’s 4 Pillars of Wellness

Reach/maintain ideal weight ↔ Healthy Diet ↔ Physical Activity ↔ Supplementation

The eye does not exist in isolation. It is an extension of the brain/CNS.
The anatomy of the eye is structured to serve the functions of the retina.
Primary reason for dilation is to detect systemic disease.

The eye is the only part of the body where neurological and vascular tissues can be viewed directly.

- Inflammatory
- Infectious
- Vascular
- Endocrine
- Neurologic
- Collagen-vascular
- Neoplastic

The Eye in Systemic Disease

Ocular Blood Flow

Motor and Sensory Areas

Iris / Ciliary Body 15%
Choroid 80%
Sympathetic NS
Retina 5%
Auto-regulated

Efferent Ciliary Body 15%
**Diabetes**

- **M____S____** is characterized by central (abdominal) obesity, dyslipidemia, raised blood pressure, and insulin resistance.

- "Diabesity"
  - Up to 97% of type 2 caused by excessive weight
  - Obesity = Increased weight caused by excess accumulation of fat.
  - "Over-fat" = normal BMI w/large waist
  - Visceral fat

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**Obesity Trends* Among U.S. Adults**

BRFSS, 1994

<table>
<thead>
<tr>
<th>No Data</th>
<th>&lt;10%</th>
<th>10%–14%</th>
<th>15%–19%</th>
<th>≥20%–24%</th>
<th>≥25%</th>
</tr>
</thead>
</table>

* 3 or more are diagnostic of Metabolic Syndrome:

- **Waist circumference:**
  - Men — > 40 inches
  - Women — > 35 inches

- triglycerides ≥ 150 mg/dL

- HDL cholesterol:
  - Men — <40 mg/dL
  - Women — <50 mg/dL

- BP ≥ 130/85 mmHg

- FPG ≥ 100 mg/dL
Obesity Trends - 2012

Obesity Trends - 2014

“Diabesity Belt”

Medical Nutrition Therapy

"People are fed by the Food Industry, which pays no attention to health, and are treated by the health industry, which pays no attention to food." - Wendell Berry
Food Matters
Optimal nutrition always starts with food.

Eat
Diets that "starve" are seldom sustainable.

Real Food
Not refined, synthetic, food-like products.

Not too much.
Portion size

Mostly plants.
A plant-intensive diet provides most essential nutrients.
QUESTIONS AND COMMENTS?

DM + Smoking = Blindness

Cigarette Smoking, Ocular & Vascular Disease
- Increased arteriolar stiffness (sclerosis)
- Increased Vascular Endothelial Growth Factor (VEGF)
- Development/worsening of DR
- Development/worsening of AMD

Arteriosclerosis with calcification of vessel wall

AMD + Smoking = Blindness
The AMD Epidemic

AMD: a sick eye in a sick body?

Is AMD a Systemic Disease?

Johanna Seddon, MD (Tufts U)

"Don't smoke; follow a healthful diet rich in dark green leafy vegetables and low in fat; eat fish a few times a week; maintain a normal weight and waist size; exercise regularly; and control blood pressure and cholesterol."

"Anyone with signs of intermediate-level macular degeneration in both eyes or advanced macular degeneration in one eye should also take dietary supplements that contain lutein, zeaxanthin, vitamin C, vitamin E, and zinc."

The Eye in Connective Tissue Disease

What is connective tissue?

“Cellular glue” that gives tissues their shape and helps them do their work. Cartilage and fat are examples.

There are over 200 disorders that impact connective tissue.
Connective Tissue Disorders
- Ankylosing Spondylitis
- Sjogren Syndrome
- Pseudoxanthoma Elasticum
- Ehlers Danlos Syndrome
- Paget’s Disease
- Marfan Syndrome
- Systemic Lupus Erythematosus

Angioid streaks are present in 85% of patients with PXE.

Masqueraders of Angioid Streaks
- High Myopia
- Trauma
- Lacquer Cracks
- Chorioidal Rupture
Differential Dx. of Angioid Streaks: PEPSI

<table>
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<td><strong>Angioid Streaks</strong></td>
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<td><strong>PEPSI</strong></td>
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Angioid Streaks:
- Alterations breaks of the Retinal Pigment Epithelium (RPE), Bruch’s Membrane and Choroid plexus
- Patient is usually asymptomatic unless CNV develops
- Approximately 50% have associated systemic disease
- Decreased vision is secondary to CNV More a streak through the fovea

Etiology:
- Pseudoxanthoma elasticum (85%)
- Ehlers-Danlos syndrome
- Paget’s Disease
- Sickle Cell Anemia

Management: Angioid Streaks
- Observation if no CNVM
- Focal laser, PDT, Anti-VEGF if CNVM is present
- Management of underlying systemic disease

Follow up:
- Twice a year with a dilated fundus examination, OCT/OCTA
- Amsler Grid self-testing (~3 x week)

A Word About Uveitis

What is uveitis?
- Defined as inflammation of the uveal tract.
- For decades, considered a single disease.
- Fact: Uveitis entails a multitude of diseases.
  - Some uveitic diseases are local, ocular immune.
  - Many are systemic diseases with ocular manifestations.

What is uveitis?
- Because the spectrum of pathogenesis ranges from autoimmunity to neoplasia to viruses, management requires an understanding of:
  - Internal medicine
  - Infectious diseases
  - Rheumatology
  - Immunology
Uveitis is an Immunological Process

Immune Privilege
- The eye enjoys a special relationship with the immune system.
  - Ability to quench unwanted immune-mediated inflammation.
  - This ability is known as immune privilege.
  - Immune privilege enables ocular tissues to remain clear.

Common Etiologies of Anterior Uveitis
- In uveitis, immune privilege is overcome
- Idiopathic (post-viral syndrome)
- Human leukocyte antigen (HLA)-B27-positive or HLA-B27-associated
- Trauma or s/p intraocular surgery

HLA-B27
- HLA-B27 is present in 1.4-8% of the general population.
- However, it is present in 50-60% of patients with acute iritis.
- HLA-B27 diseases include:
  - Ankylosing spondylitis
  - Reiter syndrome
  - Inflammatory bowel disease
  - Psoriatic post-infectious arthritis

“"A patient with recurrent, acute, unilateral, alternating anterior uveitis is nearly 80% likely to be HLA-B27 positive.”
Zamecki and Jabs
Am J Ophthal, 2010

Review of Systems Quiz
- A granulomatous condition is characterized by an organized collection of:
  A. Macrophages.
  B. Eosinophils.
  C. Histamine.
  D. Tumor cells.
Review of Systems Quiz

• A granulomatous condition is characterized by an organized collection of:

A. Macrophages.
B. Eosinophils.
C. Histamine.
D. Tumor cells.

Find the Cells

• Dark adapt
• Sl. on max illum
• Low mag
• Optic section (long)
• Increase mag
• ID the cells
• Shorten to short optic section or conic beam
• Count the cells

Hypopyon with 4+ cell and 3+ flare

Hypopyon

• A collection of leukocytes that settle in the inferior anterior chamber angle.
• Related to amount of fibrin which allows the WBCs to clump and settle.
• Highly suggestive of HLA-B27 disease, Behçet disease, or endophthalmitis.

Hyphema

• Can occur in eyes with a chronic uveitis (UGH)
• Due to neovascularization of iris/angle
KPs and Iris Nodules

Serous/Exudative RD in Posterior Scleritis

Questions/Comments?

Review of Systems Quiz

What is the most common cause of death in the United States?

A. Stroke.
B. Myocardial infarction.
C. Cancer.
D. Pneumonia.

Review of Systems Quiz

What is the most common cause of death in the United States?

A. Stroke.
B. Myocardial infarction.
C. Cancer.
D. Pneumonia.
Key Points

- Myocardial Infarction is the most common cause of death in USA.
- 610,000 per year
- Cardiac valve disease is most common cause of cardiac emboli to the eye.

Hypertension

<table>
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<th>Category</th>
<th>Optimal*</th>
<th>Normal</th>
<th>High-normal</th>
<th>Hypertension*</th>
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<tr>
<td>Optimal</td>
<td>&lt;120 and &lt;80</td>
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<tr>
<td>Normal</td>
<td>&lt;130 and &lt;85</td>
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<tr>
<td>High-normal</td>
<td>130-139 or 85-89</td>
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<tr>
<td>Hypertension*</td>
<td></td>
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<tr>
<td>Stage 1</td>
<td>140-159 or 90-99</td>
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<td>160-179 or 100-109</td>
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<tr>
<td>Stage 3</td>
<td>&gt;180 or &gt;110</td>
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Grading of Hypertensive Retinopathy

- Grade 1: Retinal vessels narrowed
- Grade 2: Nodules of cotton-wool
- Grade 3: CWS, Hemorrhage, Lipid exudates
- Grade 4: Optic disc edema

- Grades 3 and 4 increase risk of central nervous system and kidney problems.
Hypertension Quiz

- What is the most frequently encountered and primary manifestation of hypertensive retinopathy?
  a. dot-blot hemorrhages
  b. arteriole sclerosis
  c. exudative macular star
  d. optic nerve swelling

Hypertension Quiz

- What is the most frequently encountered and primary manifestation of hypertensive retinopathy?
  a. dot-blot hemorrhages
  b. arteriole sclerosis-widening/whitening of ALR
  c. exudative macular star
  d. optic nerve swelling
Essential Hypertension – Long standing

Arteriosclerosis Grade 2-3

Arteriosclerosis with calcification of vessel wall

Atherosclerosis – Most common cause of thrombosis

- Diabetes
- Hypertension
- Hyperlipidemia
- Cigarette Smoking
- Alcohol consumption

Obesity

Genetics, Environmental (super-size), Psychological, Behavioral

Retinal Arterial Macroaneurysm

Classification of Hypertension

- Primary (“Essential”) Hypertension
  - Elevated BP without obvious “cause”
  - 90-95% of all cases

- Secondary Hypertension
  - Elevated BP with a specific cause
  - Kidney disease – both parenchymal and vascular
  - Coarctation of the Aorta
  - Endocrine
  - Neurologic
  - 5-10% of all cases
Risk Factors for Primary Hypertension
- Age (>55 for men; >65 for women)
- Excess dietary sodium
- Excess alcohol
- Cigarette Smoking
- Diabetes
- Hyperlipidemia
- Family history
- Obesity (BMI >30)
- Ethnicity
- Socioeconomic status

The Deadly Quarter
- Diabetes
- Insulin Resistance
- Obesity
- Dyslipidemia

Early Cardiovascular Disease
- Macrovascular
- Microvascular

Amputation
- Stroke
- Coronary Artery Disease

Strokes
- Coronary Artery Disease
- Blindness
- Renal Failure

Hypertension increases risk and progression of ocular disease in numerous situations:
- More advanced DM retinopathy in HTN/DM
- Risk factor for retinal venous & arterial occlusion, embolism, macro-aneurysm
- MAY be risk factor for macular degeneration and open-angle glaucoma.

Impact of Hypertension on Mortality due to End-Organ Damage
- Cardiac: CHF, CHD, Sudden Death
- Cerebrovascular: Stroke, TIA
- Renal Tissue/Vascular: Renal failure
- Vascular Disease: Peripheral and Aortic

Hypertension and Ocular Disease
- Hypertension increases risk and progression of ocular disease in numerous situations:
  - More advanced DM retinopathy in HTN/DM
  - Risk factor for retinal venous & arterial occlusion, embolism, macro-aneurysm
  - MAY be risk factor for macular degeneration and open-angle glaucoma.

Summary – Benefits of Lowering BP

<table>
<thead>
<tr>
<th>Condition</th>
<th>Average % Risk Reduction</th>
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<tbody>
<tr>
<td>Stroke Incidence</td>
<td>35-40%</td>
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<tr>
<td>Heart Attack</td>
<td>20-25%</td>
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<tr>
<td>Congestive Heart Failure</td>
<td>50%</td>
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Treatments
- Step 1: Lifestyle modifications
  - Diet and exercise
  - Limit alcohol and tobacco use
  - Reduce stress factors
- Step 2: If lifestyle changes are not enough, drug therapy will be introduced
- Step 3: If previous steps don’t work, drug dose or type will be changed or another drug is added
- Step 4: More medications are added until blood pressure is controlled
Goals in Hypertension Therapy
- Lower blood pressure
- Facilitate regression of LV hypertrophy
- Reduce risk of coronary athero and myocardial infarct
- Mitigate renal damage
- Avoid stroke and CNS hemorrhage
- Prevent peripheral vascular and carotid athero
- **PROTECT THE EYES!!!**

Questions and Comments?

Carotid Artery Occlusive Disease

Hypoperfusion Retinopathy
and the
Ocular Ischemic Syndrome

Carotid Artery Occlusive Disease

Carotid Occlusive Dx: Bruit

*Dot and Blot hemis in mid-peripheral retina*
**Key Point**

- **Q:** Bilateral involvement in patients with ocular ischemic syndrome may occur in up to approximately what percentage of cases?
- **A:** 20%
Key Point

- The most common etiology of ocular ischemic syndrome is severe unilateral or bilateral atherosclerotic disease of which artery?
  - **Internal carotid**

**OIS Work Up**

- Carotid artery evaluation (Carotid – Duplex Scanning) – ICA, ECA, CCA
- Color Trans-cranial Doppler (TCD) – ocular arteries
- Possible MRA (Magnetic Resonance Angiography)
- Computed Tomography (CT) Angiography
- Cardiology work up (Echocardiogram) – Transesophageal/Transthoracic
- HTN, DM, Lipid Panel, ESR, C-reactive protein

**Ophthalmic Signs of Carotid Occlusion: Ocular Ischemic Syndrome**

- Dilated (but not tortuous) retinal veins
- Retinal Hemorrhages in mid-periphery (80%) of patients
- Cotton Wool Spots (5%)
- Neovascularization of the Disc (3.5%)
- Neovascularization of the Retina (8%)
- Ruberosis iridis/NVA (65%)
- Uveitis – mild anterior (20%)
- Emboli (retinal)
- Lower IOP - initially, then NVG

**55 yo AA male**

- BRAO OD

**55 yo AA male OS**
Ocular Ischemic Syndrome

Treatment:
- Consider carotid surgery if warranted (Endarterectomy)
  - European Carotid Surgery Trial (ECST)
  - North American Symptomatic Carotid End. Trial (NASCET)
- Therapeutic approach: Aspirin (325 mg QD or BID), Plavix
- Control modifiable vascular risk factors (HTN, DM, dyslipidemia)
- Stop smoking
- Panretinal photocoagulation (PRP) if neovascularization

**Important Note:**
Leading cause of death in OIS = Ischemic heart disease
Second leading cause of death = Stroke

QUESTIONS AND COMMENTS?

Inflammatory Disease
History
• A 34 year-old black female presents symptoms of bilateral redness x 7 days
• Gradual onset, gradual worsening
• Mild pain, mild photophobia OU
• Ocular history positive for previous episodes OU

Clinical Findings
• Biomicroscopy
  – 2+ cells in AC OU
  – “Mutton fat” deposits on endothelium OU
  – Iris nodules OU
  – Areas of posterior synechia OU
• TAP: 9 mmHg OD/11 mmHg OS
• DFE
  – “Snowbanking”
  – Gray/white (old) vitreous “puff balls” inferior PP OU

Anterior Seg Findings

Posterior Seg “Puff-balls” and “Snowbanking”

What is your **ocular** diagnosis?

Assessment
• Bilateral anterior uveitis
  – Probably recurrent/chronic
• Granulomatous
  – Mutton-fat KPs
  – Iris nodules
• Prior posterior segment inflammation
What is your plan?

Ocular management?
Systemic testing?
Consultation?

A granulomatous uveitis has an increased likelihood of being part of a s________ disease process.

Actual Management

- Treated anterior uveitis using conventional topical meds.
  - Steroid
  - Cycloplegic
- Ordered targeted systemic “uveitis” work-up
  - ACE will be elevated in up to 80% of patients with active s________.
- Chest imaging

Corticosteroids

- Topical steroids are the mainstay to treat ocular inflammatory conditions
- Choosing which medication to use depends on the severity and location of the ocular inflammation

Bilateral Hilar Lymphadenopathy on Chest X-Ray in Pulmonary Sarcoid

Bilateral Hilar Lymphadenopathy on CT Scan of Chest
Outcome

- Sarcoidosis
  - Patient was also placed on Prednisone (short-term)
  - Good ocular response to medical therapy
- What imaging tests to order:
  - Chest X-ray
  - CT of chest and abdomen

Key Points: Sarcoidosis

- A multi-system disease.
- Most often occurs between 20 and 40 years of age, with women being diagnosed more frequently than men.
- 10 to 17 times more common in African-Americans than in Caucasians.

Questions/Comments?

Types of Hematologic Disorders

- Excess production of blood cells
- Impaired production of blood cells
- Destruction of blood cells
- Abnormal function of existing blood cells

Common Disorders

- Anemia
- Sickle cell hemoglobinopathy
- Hematologic malignancies
Anemia:
*A decrease in red blood cells and/or decrease in the level of hemoglobin.

Prevalence of Anemia:
approx 1 in 77 or 3.5 million people in USA (Mayo Clinic, CDC)

Anemia
- Microcytic- MCV<80fl
  - Iron Deficiency Anemia
- Macrocytic- MCV>100fl
  - Vitamin B12 Deficiency/Folate Deficiency
    - Pernicious Anemia**
  - Liver Disease
- Normocytic- MCV 80-100fl
  - Aplastic Anemia
  - Hemolytic Anemia
  - Anemia of Chronic disease

Vitamin Deficiency Anemia
- Folate and Vitamin B-12
- Pernicious Anemia (megaloblastic)
  ***lack intrinsic factor
  - needed to absorb vitamin B12 from GI*****
  - Neurological deficits

Iron Deficiency Anemia
- Inadequate Fe+ intake
- Blood loss
  - recycled when blood cells die….if you lose blood, you lose iron
- Malabsorption
  ***most common cause of anemia***
- Chronic Diseases
  - Cancers, Collagen Vascular, Kidney
Anemia
- Symptoms
  - Fatigue
  - Dizziness
  - Headaches
  - Parathesia in fingers & toes
- Signs
  - Pallor of skin
  - Edema
  - Tachycardia

Anemic Retinopathy
- Retinal Findings:
  - Hemorrhages, CWS
  - Dilated & tortuous vessels, exudates
  - Roth spots

Anemic Retinopathy
- DDX:
  - Hypertensive or diabetic retinopathy
- Pathophysiology:
  - Anoxia, venous stasis, angiospasm, increased capillary permeability, and thrombocytopenia
  - Severity of the anemia/increased blood viscosity
- Manifestation of systemic disease

Ocular Complications
- Conjunctival pallor/jaundice or hemorrhage
- Optic Nerve:
  - Pallor
  - Disc Swelling

Questions/Comments?

Case: 44 yo BM
- CC: Floaters OD X 6 months/ - flashes of light/ +frontal headaches
- PMHX: Positive Sickle Cell Trait
  - Uncontrolled HTN X 15 years- h/o poor compliance with medications
- Blood pressure was 170/124 RAS
- BCVA: 20/20 OD, 20/20 OS.
Retinal Evaluation

OD: Fibrotic scaffolding with venous tortuosity and hemorrhage

OS: Fibrotic scaffolding with venous tortuosity and areas of NV

**Hemoglobin: HbA 60%, HbS 40%**

Case Report: 45 yo BF

- Annual wellness
- PMHX: stroke one-month ago/ Hypertension/ hypercholesterolemia
- Aneurysmal dilation of the ascending thoracic aorta, and sickle cell trait.
- BCVA: 20/20 OD, 20/20 OS
- SLE: unremarkable

Retinal Evaluation

OD: Periferal dot-hemorrhage

OS: (BRAO) with seidel neovascularization

**Hemoglobin: HbA 80%, HbS 40%**

Red-free and FA of OS

**Hemoglobin: HbA 80%, HbS 40%**

Ddx. Of Peripheral NV

- Familial exudative vitreoretinopathy
- Hyperviscosity syndromes
- Radiation retinopathy
- Sarcoidosis
- Ocular ischemic syndrome
- Sickle cell retinopathy
- Chronic myelogenous leukemia

Sickle Hemoglobinopathies

- The most prevalent genetic disorders in US
  - 10-14% of African-Americans/Mediterranean ancestry

- **Autosomal recessive**

- Pathophysiology
  - Sickle shape of RBC’s
    - Response to decrease O2 tension
    - Hypoxia, acidosis, and ischemia
Sickling Crisis

- Deoxygenation
- Dehydration
- Acidity

Organ: Stasis
Hepatic: Acidosis/Dx

Sickling Crisis

- Morbidity and Mortality
  - Vaso-occlusive events + chronic hemolytic anemia = tissue damage
- Variants
  - Sickle cell anemia: Hb SS
  - SC disease: Hb SC
  - Sickle β-thalassemia
  - Sickle cell trait – A** (Most prevalent variant of sickle-cell dx)
    - 8-10% of the Black population
      - 35-40% Hb S and 55-60% Hb A

Additional Testing for Sickle cell:
- CBC with hematocrit
- Sickledex
  - Solubility test that detects the presence of Hemoglobin S
- Hemoglobin Electrophoresis
- DNA analysis

Ocular Complications

- Focal iris atrophy in a patient with sickle cell disease

Ocular Complications of Sickle Cell

- Sickling: micro-vascular occlusion, ocular ischemia, infarction, neovascularization, and fibrovascularization
- Retinopathy
  - Increased severity in SC and β-thal
  - Why???
    - May be due to higher blood viscosity.....

Non-proliferative SC Ret

- Venous tortuosity (peripheral)
- Salmon Patch Hemorrhages
  - intra-retinal heme
- Black Sunbursts - RPE hyperplasia
- Dark without pressure
- Iridescent spots
Non-proliferative SC Ret

Proliferative SC Ret

Review of Systems Quiz

Proliferative sickle cell retinopathy is characterized by ____________________?

a. Venous tortuosity of the peripheral vessels
b. Salmon Patch Hemorrhages
c. Black Sunbursts
d. Sea-fan neovascularization

Proliferative Retinopathy

Five stages:

- **Stage 1.** Peripheral Arteriolar Occlusion
- **Stage 2.** Peripheral Arteriovenous Anastomoses
- **Stage 3.** Neovascular and Fibrous Proliferations - Sea Fan formation
  - Auto-infarct or spontaneously regress (20-60%)
- **Stage 4.** Vitreous Hemorrhage
- **Stage 5.** Retinal Detachment

Other Ocular Complications

- CRAO/BRAO
- Sickling maculopathy
- Thin, atrophic macula
- Epiretinal membrane
- Optociliary shunt vessels
- Disc Sign
  - Segmentation of capillary vessels (rare)
  - NOT Neovascularization

Proliferative SCR. The peripheral retina (left) is completely nonperfused. The right side shows a partially perfused retina. The brighter areas are the junction where the NV is leaking.
Treatment

- Proliferative retinopathy:
  - Stage 1-2: follow-up in 3-6 months
  - Stage 3-5: Laser Photocoagulation or retinal surgery.
  - Peripheral circumferential retinal scattered photocoagulation (PCRP)

- New/future- anti-VEGF therapy
  - Study by Siqueira: regression of retinal neovascularization with intravitreal Avasitin injection

Take home message:

**Marked sickle cell retinopathy** can occur in the presence or absence of systemic diseases
- Hypertension, Diabetes, Collagen Vascular Diseases, Sarcoidosis, Ocular Trauma

Leukemia

- Ocular involvement occurs in ~25% of cases

- Ocular manifestations can be divided into three categories
  a. leukemic infiltrates
  b. secondary complications related to anemia, hyperviscosity
  c. opportunistic infections (CMV, fungal, etc.)

- The ocular manifestations resolve after chemotherapy or radiation

Treatment of Leukemia

- Chemotherapy
- Radiation
- Bone marrow transplantation
- Biological therapy - Interferon
Radiation Retinopathy

Radiation doses range from 11–35 Gy. Onset from 1-8 years.

Case: 54 WF
- CC: non-specific ocular irritation
- OD>OS
- PMHX: frequent bruising of extremities for the last three months

Pertinent Findings
- Best-corrected VA: 20/20 OD, 20/20 OS.
- Pupils: Equal & round - APD
- EOM/CVF: Unremarkable
- SLE: Unremarkable

WBC count of 112 (normal ranges: 4.5-11.5)

Review of Systems Quiz
- Enlarged lymph nodes are a clinical manifestation of ______________________?
  a. Acute myelogenous leukemia-AML
  b. Sickle cell trait
  c. Iron deficient Anemia
  d. Pernicious Anemia

Hematological Malignancies
- Leukemia
  □ Acute myelogenous leukemia-AML
  □ Chronic myelogenous leukemia-CML
  □ Acute lymphoblastic leukemia-ALL
  □ Chronic lymphocytic leukemia-CLL

- Lymphoma
  □ Hodgkin Lymphoma
  □ Non-Hodgkin Lymphoma
Review of Systems Quiz

- Enlarged lymph nodes are a clinical manifestation of ________________?
  
  a. Acute myelogenous leukemia (AML)
  b. Sickle cell trait
  c. Iron deficient Anemia
  d. Pernicious Anemia

Leukemia

- Acute Leukemia
  - Rapid/progressive course that ends in death within months
  - Without tx, avg survival rate is 4 months
  - Immature leukocytes
  - Enlarged lymph nodes, spleen
  - Bone pain (bone marrow)
  - CNS involvement

Leukemia

- Chronic
  - Non-specific symptoms/signs
    - Weakness, weight loss, fever
    - Unlike Acute—rarely causes pallor or bleeding
  - Three phase:
    - Chronic phase—respond to treatment
    - Accelerated phase—difficult to control
    - Blast phase—transform into acute leukemia

Leukemia: Ocular Manifestations

Ocular Complications of Leukemia

- Ocular Adnexa
  - Lacrimal gland infiltration
  - EOM infiltration
  - Eyelid swelling (infiltration)
  - Exophthalmos, chemosis, pain
- Conjunctiva
  - Infiltrates (leukemic plaques)
  - Subconjunctival hemorrhage
  - episcleritis
- Cornea
  - Peripheral ulcers with pannus

Leukemic Retinopathy

- Hemorrhages, exudates, CWS, neovascularization
- Retinal vein tortuosity and dilation
- Retinal leukemia infiltrates
  - gray-white sheathing
- Peripheral retinal microaneurysms and retinal neovascularization
- Sea fans
- Direct infiltration of the ONH, Papilledema

***Histopathological studies have shown the choroid to be the ocular structure most commonly involved by leukemia***
Leukemic Retinopathy

- Opportunistic Infections:
  - CMV
  - Toxoplasmosis
  - Herpes
  - Fungal
  - Bacterial

Management

- Leukemic retinopathy-usually is not treated directly
- Intraocular leukemic infiltrates-systemic chemotherapy or direct radiation therapy
- Anterior segment- radiation/ injection subconjunctival chemotherapeutic agents

Ocular S/E of Treatment

- Cytotoxic drugs
  - Cataracts, EOM palsy
  - Toxic optic neuropathy
- Bone Marrow transplantation with chemo
  - Graft-host diseases
    - Sjogren like illness- dry eye
    - Conjunctival keratinization
    - Uveitis

Conclusion

- The eye does not exist in isolation, but is a mirror of systemic health.

Thank you for spending your precious time with Mickey and me!

Joe

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