Ocular Manifestations of Systemic Diseases

Komal B. Desai, MD
Assistant Professor
Eye Foundation of Kansas City
Vision Research Center
UMKC, School of Medicine
Sabates Eye Centers

Overview: History Taking
- The ability to take a thorough and meaningful history requires knowledge of the basic methods and also the correlations between specific medical conditions and their associated signs and symptoms.....
- Discuss basic components:
  - CC/HPI
  - PMHx/PSHx
  - FHx
  - ALL/MEDS/SOC
  - ROS

The Chief Complaint [CC]
- What is the reason for today’s visit?
  - Maybe be 1 or more complaints
  - Duration of each?
- Use patients own words if possible
- Try to incorporate medical language

History of Present Illness [HPI]
- “Tell me about your problem”
  - If you say, “what’s the matter with you”
  - Patient says, “that’s what I came to find out”
- Focus your questions
  - Let the patient’s story guide related questions
    - Ex: “blurry vision”…..
    - do you use glasses?
    - is it blurry at near/distance?
    - double vision?

Past Medical History [PMH]
- Should include past ocular history
  - Eye surgeries, lasers
  - Glaucoma
  - Cataracts
  - Lazy eye
  - Diabetic eye disease
  - Macular degeneration
  - Traumatic eye injury
  - Retinal detachment

Past Medical History [PMH]…..
- Diabetes mellitus
- Thyroid disease
- Hypertension
- Asthma
- Cardiovascular disease
- Stroke (TIA or CVA)
- Hypercholesterolemia
- Migraines
- Arthritis
- Osteo
- Rheumatoid
- Degenerative
- Sarcoidosis
- Tuberculosis
Family History [FHx]

• Do any diseases run in the family?
• Always ask about:
  - Diabetes, Hypertension, Heart Disease
  - Glaucoma, Cataracts
  - Unusual Blindness
  - Cancer

Allergies

• Are you allergic to any medicine?
  - List the type of allergy
• Are you allergic to shellfish or have you had any problems with contrast dye (as in CT scans)?
  - Correlation with fluorescein dye (FA)
  - Vasovagal reaction at dentists/needles?

Medications

• Eyedrops
  - Including visine, tears
• Systemic Medications
• Vitamins
• Herbal supplements

Review of Systems [ROS]

• Main purpose:
  - Make sure that you have not missed any important symptoms that could be relevant
  - For instance:
    - Positive ROS with rash and shortness of breath may correlate with acne/sinus infections found on exam and guide the Doctor to test for sarcoidosis
    - Positive ROS with headache, dizziness may correlate with retinal findings of optic nerve edema and retinal hemorrhages……hypertensive retinopathy/PFO
  - A general question that introduces each system is helpful
  - More specific questions should be guided by PMHx, complaints, age, and general state of health
  - Examples:
    - How are your ears and hearing?
    - How about your lungs and breathing?
    - Any trouble with your heart?
    - How is your digestion and bowel?

• The ROS is not a list of medical conditions the patient has. You should not list HTN, DM, High Chol in this section!

ROS…..

• General-
  - weakness, fatigue, fever, weight change
• Skin-
  - rashes; color changes; brittle; dry
• Lymph Nodes-
  - enlarged; painful
• Musculoskeletal-
  - pain; arthralgia; weakness
• Endocrine-
  - polyphagia; polydipsia; goiter
• Head-
  - headache; migraine; trauma; syncope

ROS…..

• Eyes-
  - as previously mentioned
• Ears-
  - deafness; tinnitus; vertigo; pain
• Nose-
  - rhinitis; sinus symptoms; epistaxis
• Throat-
  - hoarse; sore; vocal changes
• Respiratory-
  - SOB; cough; pleurisy
• CV-
  - chest pain; tachycardia; palpitations; claudication
ROS….

- **GI**- dysphagia, nausea, vomiting, pain, hematemesis
- **GU**- polyuria, hematuria, pain, pregnant?
- **CNS**- paralysis, paresthesias, sweating, intolerance to hot or cold
- **Psych**- mood, hallucinations, sleep disturbances, depression

---

**Has you had any…?**

- Nausea
- Vomiting
- Diarrhea
- Fever
- Chills
- Headache
- Dizziness
- Wt loss
- Wt gain
- Chest pain
- SOB
- Blood in urine
- Blood in stool
- Coughing up blood
- Stomach pain
- Rashes
- Muscle aches
- Joint aches
- Swelling in arms/legs
- Problems chewing
- Problems swallowing
- Problems sleeping

***TAKES ABOUT 30 SECONDS***

---

Categories of Systemic Disease

- Congenital
- Traumatic
- Vascular
- Neoplastic
- Autoimmune
- Idiopathic
- Infectious
- Metabolic/Endocrine
- Drugs/Toxins

---

**Congenital Disorders**

- Down syndrome
- Marfan syndrome
- Myotonic dystrophy
- Neurofibromatosis **
- Congenital lysosomal storage disorders

---

**- Neurofibromatosis**

- One of the most common inherited disorders (3 in 10,000)
- Characterized by:
  - 6+ café-au-lait spots
  - 2+ cutaneous neurofibromas
  - Lisch nodules (95%)
  - Intertriginous freckles
  - Sphenoid dysplasia
  - Optic nerve glioma
  - Positive Family History
Traumatic Disorders
- Shaken baby syndrome
- Child abuse
- Dilated fundus
  - Preretinal heme
  - Intraretinal heme
  - Vitreous heme
- Legal implications

Vascular Disorders
- Hypertension
- Embolic disease
- Amaurosis fugax

Hypertension
- Hypertensive retinopathy
  - Vessel attenuation & arteriosclerosis
  - Flame-shaped hemorrhages
  - Cotton-wool spots
    - Precapillary occlusion of arterioles
    - Ischemic infarction of superficial retina
  - Papilledema

Intracranial Hypertension
- Papilledema
  - Increased ICP w/ optic nerve edema
- Symptoms
  - Visual obscurations
  - Loss of color vision
  - Visual field defects
- Causes
  - Pseudotumor cerebri
  - Infection/inflammation
  - Neoplasm
  - Thrombosis
  - Hematoma
  - Hydrocephalus

Embolic Disease
- Severe vision loss
  - Transient
  - Permanent
- Location
  - Ophthalmic artery
  - Central retinal artery
- Sources
  - Cholesterol: Carotids
  - Calcium: Cardiac
  - Talc: IVDA
  - Lipid/air: chest trauma

Amaurosis Fugax
- Monocular dimming of vision
  - Returns to normal once emboli passes
- Sudden, transient, painless loss of vision
  - “curtain coming down over eye”
- Temporary arterial obstruction
  - TIA of ophthalmic/central retinal artery
  - Lasts few minutes
- Eval: Cardiovascular, CNS, Ophthalmic
- **Central Retinal Artery Occlusion**
  - Severe vision loss
  - Painless
  - Retina is yellow-white
    - Swollen due to lack of blood supply
    - Cherry red spot in macula

- **Central Retinal Vein Occlusion**
  - Painless vision loss
    - Mild to profound
    - Macular edema
    - Rapid onset
  - Examination
    - Diffuse retinal hemorrhages
    - Cotton-wool spots
    - 50% have assoc. POAG or HTN
  - System evaluation
    - HTN, elevated lipids
    - Coagulopathies, collagen vascular diseases
    - Paraneoplastic syndromes

- **CRAO Treatments**
  - Rebreathing carbon dioxide
  - Topical beta blockers
  - IV acetazolamide 500 mg
  - Digital massage of globe
  - Anterior chamber paracentesis
  - Calcium channel blockers
  - Hyperbaric oxygen
  - Interventional radiology

- **Central Retinal Vein Occlusion**
  - Vision loss
    - Mild to profound
    - Macular edema
    - Rapid onset
  - Examination
    - Diffuse retinal hemorrhages
    - Cotton-wool spots
    - 50% have assoc. POAG or HTN
  - System evaluation
    - HTN, elevated lipids
    - Coagulopathies, collagen vascular diseases
    - Paraneoplastic syndromes

- **Migraines**
  - Visual symptoms
    - Scintillations
    - Amaurosis fugax
    - Transient homonymous hemifield loss
    - Transient cortical blindness
  - Types: classic, common, complicated, ocular
  - Treatment
    - Evaluate visual loss and possible CNS vascular disease
    - Prophylactic treatment
    - Stop oral contraceptives?

- **Blood Dyscrasias**
  - Hyperviscosity syndromes
    - PCV, MM, dysproteinemia, leukemia
    - Amaurosis fugax; permanent visual loss
    - Retinal heme, disc edema, retinal edema
  - Recommendations
    - Periodic eye exams if asymptomatic
    - Prompt referral for any ocular symptoms

- **Blood Dyscrasias**
  - Sickle Cell Retinopathy
    - HbSC disease is most common
    - Retinal occlusions in periphery
      - Ischemia: “sea fan” neovascularization
  - Asymptomatic with severe disease
Neoplastic Disorders

- Common neoplasms
  - Primary ocular melanoma
  - CNS lymphoma
  - Metastatic carcinoma
    - Most common in adults
      - Breast, lung (F)
      - Lung, prostate (M)
  - Treatment
    - XRT
    - Chemotherapy
    - Eye (resection/enucleation)
    - Eye exams at regular intervals

Autoimmune Disorders

- Connective tissue diseases
  - Dry eyes most common manifestation
  - Burning, foreign body sensation, photophobia
  - Treatment: artificial tears, night time lubrication, punctal occlusion
  - Sjogren’s syndrome
    - Dry eyes (keratoconjunctivitis sicca)
  - Rheumatoid arthritis
    - Dry eyes, episcleritis, scleritis, corneal ulcers
    - Uveitis

- Giant Cell Arteritis

  - Temporal arteritis
    - Systemic vasculitis in people over 60
  - Symptoms
    - Headache
    - Scalp tenderness
    - Jaw claudication
    - Polymyalgia rheumatica
  - Acute visual loss (ischemic optic neuropathy)
  - Diagnosis
    - History, stat ESR, fundus exam, temporal artery biopsy
  - Treatment
    - High-dose corticosteroid therapy
    - Start immediately if high degree of suspicion exists

- Thyroid Ophthalmopathy

  - Graves’ disease
  - Doesn’t always correlate with serum thyroid levels
  - Can progress after thyroid function is normalized
  - Treatment: lids, muscle, orbital decompression

- Myasthenia Gravis

  - Autoimmune disease affecting Ach Receptors
  - Conduction deficit at neuromuscular jxn
  - 75% present with ocular manifestations
  - Bilateral ptosis
  - Limitation of motility, diplopia
  - 20% will not have systemic MG
  - Eval in all patients with ptosis/diplopia of unclear etiology (especially if worsens with fatigue)
Idiopathic Disorders

• Sarcoidosis
  - 25% have ocular involvement
  - Granulomatous uveitis
    - steroids

• Multiple Sclerosis
  - Optic neuritis
    - Optic nerve edema
    - Acute, painful vision loss
    - May be initial sign
    - IV steroids
    - MRI, neurology eval

• Multiple Sclerosis
  - Optic neuritis
    - Optic nerve edema
    - Acute, painful vision loss
    - May be initial sign
    - IV steroids
    - MRI, neurology eval

Infectious: AIDS

• Ocular manifestations
  - Dry eye
  - Cotton wool spots
  - CMV retinitis
    - IV therapy is treatment of choice
    - Ganciclovir
    - Foscarnet
  - Kaposi’s sarcoma of lids/conjunctiva
  - CD4 counts
    - <100 cell/ml for infectious diseases (CMV)

Metabolic/Endocrine: DM

• Diabetic retinopathy is leading cause of new blindness in 20-40 year olds in the United States

• Classification:
  - No retinopathy - 1yr
  - Background (non-proliferative) - 6mos
    - Hemorrhages
    - Cotton wool spots
  - Proliferative - 3mos
    - Neovascularization of disc/retina
    - Macular edema

Drugs/Toxins

• Systemic medications cause significant ocular side effects
  - Toxic retinopathy
  - Thioridazine
  - Chloroquine
  - Hydroxychloroquine
  - Tamoxifen
  - Toxic optic neuropathy
  - Ethambutol
  - Isoniazid
  - Fluoroquinolone

• Require monitoring with fundus exams at regular intervals

Conclusions

Ophthalmologist  Primary Care Physician