Ophthalmic- Systemic Disease

Ophthalmic Manifestations of Selected Systemic Diseases

Categories of Systemic Diseases
- Congenital
- Genetic
- Trauma
- Vascular
- Neoplastic
- Autoimmune
- Idiopathic
- Infectious
- Metabolic/
- Endocrine
- Drugs/Toxins

Chronic Hypertensive Retinopathy
- Severity and duration of hypertension.
- AV nicking
- Arteriolar narrowing and irregularity
- Sclerotic vessels
- Blot/flame herne
- Cotton wool spots

Acute hypertensive retinopathy
- Malignant, accelerated, hypertensive crisis
- BP >200/120
- Renal disease, toxemia of pregnancy, vasculitis
- Usually associated with vision loss
- Severe exudative changes, disc edema, hemorrhages

Diabetic Retinopathy

1. Neovascularization
2. Microaneurysms
3. Abnormal vessels
4. Capillary dropout
Non proliferative Diabetic Retinopathy

Proliferative Diabetic Retinopathy

Factors that lead to progression

- Puberty and pregnancy
- Systolic and diastolic blood pressure
- Hyperlipidemia: hard exudates in the macula and high risk of visual loss.
- Poor control of serum glucose
- ASA and smoking has no effect.

Evaluation of Diabetics

- May progress without visual symptoms.
- Florid neovascularization and still maintain perfect 20/20 vision.
- Insulin dependent, juvenile onset:
  - Needs exam during first 4 years, then yearly
- Non insulin dependent, adult onset:
  - Needs exam at the time of diagnosis, then yearly
- Diabetes prior to pregnancy:
  - Needs exam prior to or early in first trimester, then every trimester

Ocular Circulation

Central retinal a.  Posterior ciliary a

Ischemic Optic Neuropathy

- Sudden, painless vision loss with signs of optic neuropathy.
- Non arteritic
  - HBP or DM
  - Blood loss / Anemia
- Arteritic
  - Cranial arteritis
Cranial Arteritis

- Carotid stenosis, cardiogenic emboli, vasculitis, and hypercoaguability
- Ophthalmic emergency: paracentesis
- Urgent evaluation to prevent further events

Retinal Artery Occlusion

- Carotid stenosis, cardiogenic emboli, vasculitis, and hypercoaguability
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Retinal Emboli

- Cholesterol (Hollenhorst plaque)
- Calcific
- Platelet – fibrin
- Carotid (aorta, heart valves)
- Cardiac
- Asymptomatic
- BRAO

Retinal Vasculitis

- Primary Ocular conditions
  - Diabetic retinopathy
  - Neovascularization
  - Hypertensive retinopathy
- Systemic associations
  - Stroke
  - Rheumatoid arthritis
  - Sarcoidosis
- Retinal angitis
  - Neovascularization
  - Inflammation

Retinal Vein Occlusion

- Branch vein occlusion
- Central retinal vein occlusion
  - Hypertension, glaucoma, hypercoaguability, anticoagulation antibody syndrome, hyperviscosity, myeloproliferative disorders, anemia

Transient Vision Loss

- Monocular
  - Transient Visual Obscuration
    - Optic disc edema (papilledema, uveitis, tumor)
- Binocular
  - Transient Monocular Blindness (Amaurosis Fugax)
    - Carotid stenosis
      - Cardiogenic Vasculitis
      - Hypercoaguability
  - Migraine
  - Vertebrobasilar TIA

Sibony 2012
45 yo WM with a visual disturbance.

Migraine

- Binocular
- Scintillations, hemianopic scotoma
- 15-45 minutes
- Fortification spectra
- "Spectral march" across the visual field
- With or without headache
- Prior history of migraine headaches
* Strong family history of headaches

Pituitary tumors

- ACTH: Cushing's
- GH: Acromegaly
- Prolactin:
  - F: amenorrhea, galactorrhea
  - M: impotence, gynecomastia

Orbital Lymphoma

- Extramedullary B cell lymphomas
- 50-70 yo
- Unilateral (bilateral)
- Proptosis, anterior congestion, ophthalmoparesis, ptosis
- 40% associated with systemic involvement.
- May infiltrate any of the orbital structures or present as a molding, non-displacing mass.
- Immunohistochemistry to distinguish from benign lymphoid hyperplasia

Orbital Myeloma

Orbital Lymphoma

Orbital Myeloma
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Tumors of the posterior pole

Choroidal metastasis: lung
Melanoma

Anterior segment tumors

Squamous Cell carcinoma
Metastatic carcinoma

Papilledema

- Idiopathic intracranial hypertension
- Brain tumors
- Venous sinus thrombosis
- Obstructive hydrocephalus
- Meningitis
- Cerebral edema
  - Subarachnoid hem

Pseudopapilledema
Ophthalmic - Systemic Disease

Part 2

Ophthalmic Manifestations of Systemic Disease

Cells and flare

Uvea

Keratic precipitates

Hypopyon

Complications of Uveitis

Posterior Uveitis
Sarcoidosis

- Infectious
  - Toxoplasmosis
  - Syphilis
  - Lyme
  - Viral
  - TB
  - Herpes zoster
  - Nematodes
  - CMV
  - Toxocara canis (dog roundworm)

- Autoimmune
  - Ankylosing spondylitis
  - Reiter's syndrome
  - MS
  - Inflammatory bowel disease
  - Sarcoidosis
  - Vogt-Koyanagi-Harada
  - Vasculitis
  - Behcets

- Idiopathic
  - Idiopathic syndrome
  - Lymphoma
  - Ocular ischemia
  - Retinoblastoma

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Scleritis

- 50% with systemic disease
- Rheumatoid arthritis
- Wegener's
- Polyarteritis
- Lupus
- Relapsing polychondritis

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Myasthenia Gravis

Pre & Post

Tensilon Test

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Pre Tensilon
**Orbital Cellulitis**

- Bacterial infection of the orbit secondary to sinusitis
- Erythema, swelling, proptosis, Ophthalmoplegia, vision loss
- Fever, leucocytosis

**Infection: sinusitis**

**Candida endophthalmitis**

- Opportunistic infection
- Diabetics, burn patients, chronic iv antibiotics, iv drug abuse, patients receiving parenteral nutrition

**Toxoplasmosis**

- Protozoan infection due to T. gondii, Host: cat
- Ingestion of contaminated uncooked meat or reactivation from prior transplacental in utero exposure

**HIV**

- HIV retinopathy
- CMV retinitis
- Usu seen with CD4 <50
- Rx. Gancyclovir, Foscarnet, Cidofovir

**Herpes Simplex Keratitis**

- Intracellular DNA virus
- Usually Type I
- Primary infection usually in children
- Neonatal (type II)
- Recurrent forms (type I), trigeminal ganglion reservoir
Zoster ophthalmicus

Optic Neuritis

Acute, painful vision loss with decreased acuity, abnormal color vision, APD and Central scotoma on visual fields. Fundus findings consist of three types:

- Retrobulbar Optic neuritis
- Papillitis
- Neuroretinitis

Keyser-Fleischer Ring

Wilson's disease
Inherited disturbance in copper metabolism
Neurological problems especially basal ganglia

Bulls eye maculopathy

Placquinil (chloroquine): risk of toxicity with cumulative doses of 300 gm

Toxic retinopathies

Phenothiazines
Tamoxifen
Toxic reactions

Genetic Disorders

- Phakomatoses
  - Neurofibromatosis
  - Tuberous sclerosis
  - Von Hippel Lindau
- Mitochondrial myopathies
- Coloboma
- Down’s syndrome
- Marfan syndrome

Neurofibromatosis

- Dominantly inherited
- 3:10,000
- Lisch nodules of the iris
- Café au lait spots (>5)
- Cutaneous neurofibroma
- Optic n. gliomas
- Intertriginous freckles
- Osseous lesions (sphenoid dysplasia)

von Hippel Lindau

- Dominantly inherited
- Capillary angiomas of the disc and retina
- CNS tumors: hemangioblastomas
- Abdominal or visceral tumors e.g. renal carcinoma, pheochromocytomas

Kearns-Sayre

- Chronic progressive external ophthalmoplegia
- Mitochondrial DNA disease
- Chronic progressive external ophthalmoplegia
- Cardiac conduction disturbance
- Retinitis pigmentosa

Coloboma

- Isolated, autosomal dominant
- Trisomy 13
- Aicardi’s syndrome
- CHARGE (coloboma, heart, atresia, retardation, genital, ear)
- Goldenhars syndrome
- Epibulbar dermoids, maldevelopment ear, mouth and jaw