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 heme
- 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
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 hypercoagulability
- Ophthalmic emergency: paracentesis
- Urgent evaluation to prevent further events

Retinal Emboli

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

Retinal Vasculitis

- Retinal periphlebitis
- Sarcoidosis
- Lupus

- Primary Ocular conditions
- Secondary systemic associations
- Systemic overlap

- Giant cell arteritis
- Rheumatoid arthritis
- Microangiopathy of retinal vasculature
- Inflammatory bowel disease
- Nephrotic syndrome
- Angiitis, Polyarteritis
- Toxoplasmosis
- Coxsakie (AVM)

Retinal Vein Occlusion

- Branch vein occlusion
- Central retinal vein occlusion
- Hypertension, glaucoma, hypercoagulability, anticoagulant antibody syndrome, hyperviscosity, myeloproliferative disorders, anemia

Transient Vision Loss

- Monocular
- Binocular

- Transient Monocular Blindness (Amaurosis Fugax)
- Carotid stenosis
- Cardiogenic Vasculitis
- Hypercoagulable

- Migraine
- Vertebrobasilar TIA

- Seconds
  - 2-10 m
  - 15-45 m
  - 2-10 m
45 yo WM with a visual disturbance.

Migraine

- Binocular
- Scintillations and 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, ophthalmoplegia, 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
Tumors of the posterior pole

- Choroidal metastatic 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
Part 2

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Uvea

Cells and flare

Complications of Uveitis

Posterior Uveitis

Keratic precipitates

Hypopyon

Sibony 2012
Sarcoidosis

- Autoimmune
  - Ankylosing spondylitis
  - Reiters syndrome
  - MS
  - Inflammatory bowel disease
  - Sarcoidosis
  - Vogt-Koyanagi Harada
  - Vasculitis
  - Behcets
- Idiopathic
- Masquerade syndrome
  - Lymphoma
  - Ocular ischemia
  - Retinoblastoma

Uveitis

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

Scleritis

- 50% with systemic disease
- Reumatoid arthritis
- Wegeners
- Polyarteritis
- Lupus
- Relapsing polychondritis.

Myasthenia Gravis

Pre

Post

Tensilon Test

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Pre Tension
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

- 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

Dysthyroid orbitopathy

Keyser-Fleischer Ring
Wilson disease
Inherited disturbance in copper metabolism
Neurological problems especially basal ganglia

Bulls eye maculopathy

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

Toxic retinopathies

Phenothiazines
Tamoxifen

Sibony 2012
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 angiomias 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)
- Goldenhar syndrome
  - Epibulbar dermoids, maldevelopment ears, mouth and jaw