Course Handout

Introduction

• The eye is an extension of the brain
• The anatomy of the eye is structured to serve the functions of the retina
• The primary reason for dilation is to detect systemic disease

Case Studies in Systemic Disease

• In this course, several patient cases will be presented. These cases will illustrate ocular complications of a variety of systemic diseases. The symptoms, clinical features, pathogenesis, diagnostic workup, and management of these conditions will be reviewed.

Connective Tissue Disorders

• Ankylosing Spondylitis
• Sjogren Syndrome
• Pseudoxanthoma elasticum
• Ehlers Danlos syndrome
• Paget’s Disease

Dermatologic Disorders

• Rosacea
• Basal Cell Carcinoma
• Squamous Cell Carcinoma

Endocrine Disorders

• Graves Disease
• Diabetes Mellitus
  o Some important systemic effects of diabetes that affect retinopathy and increase the risk of “Heart Attack”:
  o #1 Proteinuria
  o First sign of renal disease
  o As nephropathy increases…the glomerular filtration rate falls
  o American Diabetes Association (ADA) recommends yearly urinalysis
  o #1 Proteinuria (Cont’d.)
  o Random Spot Urine or 24 hour collection
  o Normal <30
  o Microalbuminuria  30 mg – 299 mg
    Albuminuria > 300 mg
  o #2 Anemia
o Gets more severe as renal disease worsens
o Kidney production of *Erythropoietin* decrease, which means that less reaches the bone marrow and less red blood cells are made
o Measured by Hematocrit (HCT) and hemoglobin levels in a CBC
o If hemoglobin levels are less than 11g/dl = anemia
o Anemia may actually be making the retinopathy worse!!!
o Treat the patient with “Procrit” if patient is not on dialysis
o also give iron
o Kidney Erythropoietin Bone Marrow RBC’s

#3 Hyperlipidemia
o Cholesterol and triglyceride healthy levels should be < 200 mg/dl
o PCP should consider Lipitor if cholesterol high

#4 Hypertension
o Target blood pressure for diabetics with nephropathy is 130 /80
o ACE inhibitors should be given if blood pressure is high
o ACE inhibitors are both renal-protective and anti-proteinuric

#5 Hyperglycemia
o Induces vasoconstriction = kidney (glomerular) damage
o The Hemoglobin A1C should be as close to 7 % as possible

**Pituitary Disorders**

- **Pituitary Adenoma**
  - **Anatomy**
    - The gland is divided into an “Anterior” Lobe and a “Posterior” Lobe
    - Pituitary gland sits within the sella tursica (depression in the sphenoid bone) at the base of the skull
  - The Optic Chiasm is ~ 10 mm above
  - Laterally the walls of the sella tursica are adjacent to the cavernous sinus
  - **Classification:**
    - Nonsecretory
    - Prolactin Secreting
    - Growth Hormone Secreting
    - Adrenocorticotropic Secreting
    - Leutinizing Hormone Secreting
    - Follicle Stimulating Hormone Secreting
    - Thyroid Stimulating
  - **Pituitary Adenoma**
    - Microadenomas - < 10 mm in diameter and confined to the sella tursica
    - Macroadenomas - > 10 mm in size and extends beyond the sella tursica
  - **Signs and Symptoms**
    - Headache – usually frontal in location with a “dull” pain
    - Loss of vision
    - Loss of visual field
    - Diplopia – secondary to cranial nerve palsy
    - Visual Hallucinations – tumor affecting temporal lobe
    - Infertility, galactorrhea, amenorrhea, impotence
  - **Ocular Emergency:**
    - A sudden onset of ophthalmoplegia, vision loss, nausea, vomiting, and severe headache = Pituitary Apoplexy
    - Pituitary Apoplexy = Acute hemorrhagic infarction of a pituitary adenoma
  - **Visual Fields:** Bitemporal Defect
  - Diagnosis made by MRI or CT scan
  - **Treatment:**
    - Surgery, Radiation, Medication
Hematologic and Cardiovascular Disorders

- Giant Cell Arteritis
  - Acute painless vision loss (VA loss is usually permanent)
  - Pale swelling of the optic nerve head with flame shaped hemes
  - Central retinal artery occlusion may occur
  - Cranial nerve palsy (CN 3,4,6) may also be present, CWS
  - Possible association with "Polymyalgia Rheumatica" (PMR)
    - Stiffness in the neck, shoulder, and hip
    - 50% of Giant Cell patients have PMR
    - Is there a link between GCA and PMR ???

- Hypertension
  - Malignant HTH: Blood Pressure > 200/120
    - Disc edema with or without exudate
    - Arterio-venous crossing changes
    - Nerve fiber layer infarcts (cotton wool spots)
    - Macular edema
    - Hard exudates / flame shaped hemes
    - Choroidal ischemia

- Leukemias

- Ocular Ischemic Syndrome
  - Hypoperfusion Retinopathy / Ocular Ischemic Syndrome:
    - Usually unilateral but may be bilateral in 20% of cases
    - Males > Females by a 2 to 1 ratio
    - Dot and blot hemes / microanuerysms found only in the mid-peripheral retina = Hypoperfusion Retinopathy
    - When the above is associated with neovascularization of the Disc, Retina, Iris or Angle = Ocular Ischemic Syndrome
  - Pathogenesis: Ocular Ischemic Syndrome:
    - Atheromatous ulceration and stenosis at the bifurcation of the common carotid artery (90% occlusion has to be present)
  - Symptoms: Ocular Ischemic Syndrome:
    - Ocular and periorbital pain in 40% of cases = "Ocular Angina"
    - Prolonged recovery of vision following exposure to bright light-known as "Light Induced Amaurosis"
    - Amaurosis Fugax (Transient Monocular Blindness) in 5% of cases
    - Transient Ischemic Attacks (TIA)
    - Vision Loss (90%) – Short Posterior Ciliary Arterial hypoperfusion
  - Ocular Signs: Ocular Ischemic Syndrome:
    - Dilated but not tortuous retinal veins
    - Retinal Hemorrhages in mid-peripheral retina (80%) of patients
    - Cotton Wool Spots (5%)
    - Neovascularization of the Disc (35%)
    - Neovascularization of the Retina (8%)
    - Rubeosis iridis (65%)
    - Uveitis – mild anterior (20%)
    - Emboli (retinal)
    - Lower IOP - initially
  - Work Up:
    - Carotid artery evaluation (Carotid – Duplex Scanning)
    - Possible MRA (Magnetic Resonance Angiography)
    - Cardiology work up (Echocardiogram) – Transesophageal
    - Lipid Panel
  - Treatment:
    - Consider carotid surgery if warranted (Endarterectomy)
Therapeutic approach – Aspirin (325 mg QD or BID)
- Panretinal photocoagulation (PRP) if neovascularization
- Stop smoking
- **Important Note:**
  - Leading cause of death = Ischemic heart disease
  - Second leading cause of death = Stroke
- Sickle Cell Disease

**Neoplastic Disease**

- Primary Ocular Tumors
  - Ocular melanoma
- Metastatic Tumors to the Eye, Orbit, and Visual Pathway
  - Lung cancer - #1 cause of death in males
  - Breast cancer - #1 cause of death in females
  - Prostate cancer – most common cancer in men
  - Colon cancer – 3rd most common in men, 2nd most common in women
  - Melanoma – skin most common site of cancer development (Males = trunk) (Females = extremities)
  - Ovarian cancer – disease of postmenopausal women
  - Pancreatic cancer – usually >65 years old, wt. loss, jaundice, anorexia
  - Uterine cancer – disease of postmenopausal women (abnormal vaginal discharge or bleeding)

**Inflammatory Conditions**

- Systemic Lupus
- Sarcoidosis
- Rheumatoid Arthritis

**Infectious Disease**

- Syphilis
- Chlamydia
- Cytomegalovirus
- Gonorrhea
- Hepatitis B
- Herpes (HSV / HZV)
- HIV
- Bartonella (Cat scratch)

**Other Disorders**

- Interferon Retinopathy
- Radiation Retinopathy
- Pupil-involved Third Nerve palsy
  - May be caused by the following:
    - Aneurysm
    - Microvascular disease (Infarction)
    - Tumor
    - Trauma
    - Infection (syphilis)