Pathology of the Eye
M2 Pathology Course
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Key Concepts

1. The retina and optic nerve are embryologic derivatives of the central nervous system and respond to disease like the brain.
2. There is a narrow margin of tolerance for injury to the eye.
3. Systemic diseases affect the eye.
4. Some diseases are unique to the eye.

Introduction

1. We devote a disproportionate amount of time in this course to ocular pathology
   a. Although we spend less than 1% of the course discussing pathology of the eye, it has been estimated that between 9% and 12% of all visits to a family practitioner or general internist involve the eye or visual system

2. Patients are extremely afraid of losing vision
   a. Examples

Key Concept #1: The retina and optic nerve are embryologic derivatives of the central nervous system and respond to disease like the brain.
1. Example 1: Temporal Arteritis
   a. Pathology
      i. This condition affects older patients. Recall the pathology of arteriosclerosis and calcification of the internal elastic lamina of arteries in older patients. These two findings are the baseline “normal” findings in the age group affected by temporal arteritis.
      ii. An arteritis is an inflammation within the vessel, frequently damaging the vessel wall.
      iii. in temporal arteritis, the inflammation may be especially intense at the level of the internal elastic lamina
      iv. may involve a component of granulomatous inflammation (another name for this condition is “giant cell arteritis”)
   b. Associated changes
      i. damage to the intima – resulting in
      ii. intimal hyperplasia - resulting in
      iii. narrowing of the lumen – resulting in
      iv. alterations in blood flow – predisposing to
      v. thrombosis – which leads to
      vi. either ischemia or infarct of tissue supplied by the artery
   c. Review …
      i. be certain you can distinguish between ischemia and hypoxia
      ii. be certain you can define what is meant by the term infarct
   d. Other important facts …
      i. The temporal artery is one of a number of cranial vessels affected
      ii. Vasculitis of the vessels supplying the optic nerve head (the posterior ciliary vessels) may lead to ischemia to the optic nerve and even infarct of the nerve
      iii. Because the optic nerve is a tract of the central nervous system, the infarcted optic nerve swells, as does the brain after an infarct
iv. Because the blood supply of the optic nerve is diminished in temporal arteritis, the *swollen* optic nerve also appears *pale*. Thus, the ophthalmoscopic *pale and swollen* optic nerve signifies either severe ischemia to the nerve or an optic nerve *infarct*.

1. compare with *papilledema*, in which the optic nerve is *hyperemic* and *swollen*
   a. another hint in the differential diagnosis: papilledema is usually bilateral

v. Because there is no regeneration within the central nervous system, an infarct of the optic nerve results in *permanent* visual loss!

e. Temporal Arteritis: Clinical Features
   i. Usually affects the elderly (>60 years old)
   ii. Symptoms: loss of appetite, fatigue, low-grade fever
   iii. Sometimes associated with polymyalgia rheumatica
   iv. Consequences of ischemia
      1. transient obscuration of vision
         a. *ischemia* of the optic nerve
      2. jaw claudication – often misdiagnosed as temporal mandibular joint disease
         a. *ischemia* of the masseter muscle
      3. scalp tenderness when combing or brushing hair
         a. *ischemia* of scalp tissues

f. Temporal Arteritis: Diagnostic Tests
   i. Erythrocyte Sedimentation Rate (ESR)
   ii. C-Reactive Protein
   iii. Temporal artery biopsy

g. Treatment: high dose corticosteroids

2. *Example 2: Non-arteritic Anterior Ischemic optic neuropathy (AION)*
   a. *Emboli* to the optic nerve circulation

3. *Example 3: Central Retinal Artery Occlusion*
   a. Occlusion of the central retinal artery may produce a diffuse *infarct* of the neurosensory retina
      i. As a consequence, the retina may swell
      ii. Because the histology of the macula is such that the retina is physiologically thin in the foveola, the center of the fovea, the retina is not opaque in the fovea, permitting the normal red-orange color of the underlying surrounding the foveola in which no ganglion cells are detected
      iii. the *cherry-red spot*
         1. Cherry-red spots also develop because of storage diseases in which a variety of substances deposit in the thick ganglion cell layer highlighting the foveola in which no ganglion cells are present and the red-orange color of the underlying choroid is allowed to show through
         2. Question: what is the most common cause of a “cherry-red spot” of the retina?

**Key Concept #2:** There is a narrow margin of tolerance for injury to the eye

1. Comparing the consequences of bronchopneumonia with endophthalmitis
   a. Bronchopneumonia may resolve without consequences
   b. The brain (and by extension, the optic nerve and retina) are exquisitely sensitive to acute inflammation

2. Consequences of disturbing the function of the eyelid
   a. Exposure of the cornea – pain and predisposition to infection
      i. Basal cell carcinoma
      ii. Remember – the roof of the orbit is the floor of the frontal cranial fossa
Key Concept #3: Systemic diseases affect the eye

Example 1: Sarcoid
a. Uveitis
b. Granulomas in the conjunctiva

1. Example 2: Diabetes Mellitus
a. Cataract
b. Microcirculation – Diabetic Retinopathy
   i. Thickening of basement membranes
      1. thickening of the basement membrane of the epithelium of the ciliary body may be analogous to mesangial changes in the kidney
      2. thickening of the basement membrane of the microcirculation
   ii. Loss of pericytes relative to the number of endothelial cells
      1. pathophysiological consequence: breakdown in the “blood-retinal barrier”, rendering the vessel leaky
         a. hemorrhages
         b. exudates
         c. macular edema
   iii. Intraretinal microangiopathy
      1. including microaneurysms
         a. the use of intravenous fluorescein angiography to visualize microaneurysms – they are “leaky”
      2. proliferative diabetic retinopathy: pre-retinal vascular proliferation
      3. targets for therapy
2. **Example 3: Retinopathy of Prematurity**
   a. The temporal (lateral) aspect of the newborn retina is not mature at term, and much less so in a premature infant
   b. In a low birth-weight infant who has respiratory distress syndrome, administration of O₂, the immature vessels in the lateral peripheral retina may go into spasm, producing retinal ischemia, and inducing both intraretinal microangiopathic changes and proliferative changes

3. **Example 4: Sickle Retinopathy**
   a. In S-C and S-Thalassemia
   b. A decreased O₂ tension in the retinal periphery may precipitate sickling of the erythrocytes in the microcirculation, retinal ischemia, etc.

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**Key Concept #4:** Some diseases are unique to the eye
1. Cataract
2. Glaucoma
3. Age-related macular degeneration
4. Retinal detachment