Anterior Segment Disease and the Systemic Link

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Summary

As optometry's role in health care increases, so does our responsibility to appropriately diagnose and appropriately educate them on the systemic link. This course will address three critical anterior segment findings along with their association with systemic conditions and appropriate action plans for the progressive optometrist.

Learning Objectives

1) Understand corneal verticillata and its potential association with system disease
2) Understand floppy eyelid syndrome and it's systemic link
3) Discuss strategies important for the diagnosis and management of anterior uveitis and Wilson's disease

Course Outline

1) Corneal Verticillata
    a. What is it?
        i. Whorl like deposits in the basal layer of the epithelium
        ii. Will not stain
        iii. Rarely causes any visual disturbance
    b. What Causes it?
        i. Medication
            1. Not a reason to discontinue medication
            2. Amiodarone
                a. Most common medication to cause this finding
                b. Anti-arrhythmic medication
            3. Chloroquine and Hydroxychloroquine
                a. Used to treat malaria
                b. Used for rheumatoid arthritis and lupus
            4. Indomethacin
                a. Non-steroidal anti-inflammatory
                b. Used to treat arthritis, gout
        ii. Fabry's disease
            1. Genetic lysosomal storage disease
                a. Deficiency of the enzyme alpha galactosidase A
                b. Causes glycolipids to accumulate within blood vessels, tissue and organs
            2. Symptoms can initially appear in early childhood
            3. Pain to the extremeties and/or GI tract

4. Kidney and cardiac involvement
5. Angiokeratomas, anhidrosis
6. Generalized fatigue and neuropathy
7. Diagnosed through measuring enzyme activity
8. Treated with Fabrazyme (alpha-galactosidase) delivered through intravenous infusion

c. What to do when we see it?
   i. Question about medications
   ii. In individuals not taking high risk medications, consider Fabry's
      1. Enzyme assay for alpha-GAL
      2. Is X linked so in females will often times require a genetic test

2) Floppy eyelid syndrome
   a. What is it?
      i. Extensive lid laxity secondary to decreased elastin content in tarsal plate
      ii. Spontaneous eversion of the lid can occur while sleeping
      iii. Can lead to chronic irritation of the lid
      iv. Critical to perform lid eversion on eye examination
   b. What causes it?
      i. Low elastin levels and spontaneous eversion
      ii. Will cause signs/symptoms of ocular discomfort
      iii. Has a strong association obstructive sleep apnea
      iv. Also associated with keratoconus, down syndrome
   c. What do we do when we see it?
      i. Question about sleep patterns
      ii. Question about sleep apnea or confirm diagnosis
      iii. Refer to physician for appropriate testing
      iv. Monitor optic nerves carefully for risk of glaucoma
3) Anterior Uveitis
   a. Clinical signs and symptoms
   b. Full medical history
      i. Medications
      ii. Special consideration
         1. Autoimmune conditions
         2. Systemic infections
         3. Recent history of herpetic conditions
         4. Systemic symptoms consistent with undiagnosed condition
      iii. Slit Lamp examination
         1. Cells and flare in anterior chamber
            a. Cells – graded on scale from 0 to 4
               i. Graded using a 1 to 3mm field of light
            b. Flare – graded on scale from 0 to 4
            c. Endothelial deposition – keratic precipitates

2. Hypopyon
3. Posterior synechia

iv. Posterior segment examination
1. Pupil dilation is critical for fundus examination
2. Rule out posterior uveitis

4) Pathophysiology
a. Receptors in the nucleus of the cell promotes production of inflammatory mediators
b. Phospholipase activation
c. Cyclooxygenase – 1 and cyclooxygenase – 2
1. Constitutively
a. Continues at a constant level
b. COX-1 – important for gastrointestinal function
2. Facultatively
a. Increases with inflammation
b. COX-2 – is up regulated with inflammation

ii. Phospholipid bilayer and phospholipase A2 – original molecules that begin the production of end products
iii. Arachidonic acid
iv. Prostaglandins and thromboxane – inflammatory end product

d. Treatment options
e. Topical corticosteroids
i. Binds to steroid receptor in cell cytoplasm
ii. Enters the nucleus of the cell and promotes production of anti-inflammatory mediators and inhibits the production of inflammatory proteins
iii. Promotes lipocortin-1 production inhibiting phospholipase A2
iv. Inhibits COX-1 and COX-2
v. Suppresses cyclooxygenase production
vi. Up-regulate anti-inflammatory proteins
vii. Down-regulate inflammatory proteins
viii. Ester vs. ketone steroids
ix. Side-effects – increased intraocular pressure and cataract formation
1. Treat intraocular pressure increases with

f. Topical corticosteroids
i. Frequency of dosing is dependent on severity
1. More severe case will require higher dosing schedule to remediate intraocular inflammation

ii. Prednisolone acetate
1. Discuss branded versus generic

iii. Difluprednate
1. Understand vehicle
2. Consistent concentration throughout the bottle
3. Dosing efficacy at qid in affected eye has been shown to be equal to eight times a day with prednisolone acetate

4. Be cognizant of IOP increase

g. Cycloplegics
   i. Inhibits response of ciliary body
   ii. Adds comfort to patient
   iii. Cyclopentolate, homatropine, scopolamine, atropine

h. Pain control
   i. Consider oral non-steroidal anti-inflammatory agents
   ii. Consider oral narcotics
      1. Codeine and hydrocone
      2. Understand how these medications are commercially available
      3. Discuss appropriate considerations when treating with oral pain medications

5) Further investigation
   a. Etiology
      i. Occurs approximately 8 per 100,000 in the population
      ii. 90% are acute anterior uveitis
      iii. Idiopathic
   b. Systemic association
      i. Consider if it is recurrent, unusual severity, unresponsive to treatment or bilateral
      ii. Rapid Plasma Reagin (RPR)
         1. Fluorescent treponemal antibody absorption (FTA-ABS)
         2. Serologic test for syphilis
      iii. Erythrocyte Sedimentation rate (ESR)
         1. Help evaluate for sarcoidosis when suspected
      iv. Serum lysozyme levels and Angiotensin-converting enzyme (ACE)
         1. Help evaluate for sarcoidosis when suspected
      v. HLA-B27
         1. Common in individuals with ankylosing spondylitis
      vi. Anti-nuclear antibody (ANA) and possibly rheumatoid factor (RF)
      vii. Chest radiography
         1. Rule out sarcoidosis and tuberculosis

6) Kayser-Fleischer Ring
   a. What is it?
      i. Brown/yellow ring around the periphery of the cornea
      ii. Copper deposits in Descemet's membrane – can extend into descemet's membrane
      iii. Usually seen bilaterally when associated with systemic disease
   b. What causes it?
      i. Copper deposition in descemet's membrane which fades as you move toward the central cornea
      ii. Often seen in Wilson's disease – excessive copper deposition in the body

1. Symptoms develop from excessive copper build up in the liver and brain

c. What do we do when we see it?
    i. Refer to physician to rule out Wilson's disease if is seen during an eye examination
        1. Treated with low copper diet and chelating agents such as prescription medications and zinc supplements