Lupus & the Eye

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Ocular Inflammatory Diseases

- 3rd Leading cause of blindness worldwide
- Incidence 15:100,000 U.S.
- Dry Eye Disease/Keratitis associated with Sjogren’s syndrome, SLE, RA, Scleroderma
- Uveitis associated with RA, Spondyloarthropathies, JIA, SLE
- Scleritis associated with Wegener’s, RA, SLE, JIA, Sarcoid
- Retinitis associated with Takayasu’s, PAN, Wegener’s Granulomatosis, Behcet’s
- Optic Neuritis associated with Giant Cell Arteritis
Dry Eye Disease

- Multifactorial disease of the tears and ocular surface
- Most common manifestation of lupus patients 39.5%
- Associated with underlying autoimmune disorder (i.e. Sjogren’s syndrome - chronic, systemic, progressive disease affecting 4 mil people in US)
- Symptoms include visual disturbance, discomfort, tear film instability
- Potential damage to the ocular surface
- Increased Tear Osmolarity
- Inflammation of the ocular surface


Dry Eye Disease

- Dry eye is also referred to as ocular surface disease, dysfunctional tear syndrome (DTS), or keratoconjunctivitis sicca
- Dry eye is accompanied by inflammation of the ocular surface
- Dry eye is common
  - (10% US Population; 23-30 Million Dry Eye patients)
  - Often under diagnosed
- Dry eye impacts vision quality and can cause blurred vision, fluctuations, light sensitivity, and glare.
- Quality of life and activities can be greatly impacted by dry eye symptoms, with significant psychological impact.
- Patients have reported willingness to trade years at the end of life to be free of dry eyes
Dry Eye Disease: Work-Up

- Review of Medical History
- Review of Medications for potential Side-effects
- Complete Eye Examinations
  - Schirmer Tear Testing
  - Include Tear Osmolarity
  - Ocular Surface Inflammatory Markers (MMP-9)
  - Serological Markers for Sjogren's disease, including Salivary Protein-1, Carbonic Anhydrase, Parotid Secretory Protein
Artificial tears, baseline therapy for ocular surface rehydration

Patients who test positive for inflammatory markers
- Restasis (topical cyclosporine)
- Topical Steroids
- Azithromycin
- Doxycycline
- Oral Secretagogues
- Oral Immunomodulation
- Amniotic Membrane Tissue
Uveitis associated with Lupus

- **Findings:**
  - Inflammation of the Iris/Uveal Tissue

- **Symptoms:**
  - Light sensitivity/Pain/Decreased Vision

- **Reported Prevalence of Lupus in patient with Uveitis varies between 0.1%-4.9%**

Retinal Vasculitis associated with Lupus

- Due to immune complex deposition of the blood vessels and basement membrane
- Leads to vasculitis and thrombosis
- Typical manifests with decreased vision
- 2nd most common presentation behind ocular surface disease (21%)
Complications related to specific treatments for Lupus: Represent the third most common ocular manifestation

Steroid-related Adverse Reaction

- Frequently employed for acute/chronic inflammation in context of auto-immune disease
- Can be seen with all routes of administration: topical/oral/inhaled/subcutaneous/intramuscular/intravenous/intraarticular
- Findings
  - Elevated Intraocular Pressure (IOP)
  - 31% patients exposed to steroid develop ocular hypertension
  - Typically seen weeks after institution of therapy
  - 5% of population care Myocilin Gene-associated with acute, malignant rise in intraocular pressure
  - Risk factor for Glaucoma Formation
- Cataractogenesis (Cataract Formation)
- Delayed Wound Healing of Ocular surface
- Increased Risk of Ocular Infection
- Thinning of Ocular Tissue; i.e. cornea / scleral thinning
- Secondary concerns related to increased Blood sugars

Steroid-use Screening Recommendations

- Baseline Evaluation
- 3-6 months depending findings
- Examination
  - Visual Acuity
  - Tear Film Evaluation
  - Biomicroscopy
  - Dilated Funduscopic Examination
  - Goldmann Applanation Tonometry
  - +/- Humphrey Visual Field Testing
Plaquenil Macular Toxicity: More Specifically

- Originally Reported in chloroquine-induced retinal toxicity in 1959
- 1967 hydroxychloroquine (Plaquenil®) first cases reported by Shearer et al.
- Rare: 6.8/1000
- Irreversible
- Associated with:
  - Dosage (recommended not to exceed 400mg daily)
  - Duration of treatment (> 5 years)
  - History of Renal insufficiency
  - History of Hepatic Insufficiency
  - Obesity (Increased BMI)
  - Age > 60 years
  - Concurrent retinal disease

Shearer RV, Dubois EL. Ocular changes induced by long-term hydroxychloroquine (plaquenil) therapy. Am J Ophthalmic1967;64(2):245-252
Plaquenil: Ocular Manifestations

- **Corneal Verticillata**
  - superficial whorl-like deposits of the corneal
  - visually asymptomatic
  - reversible with cessation

- **Bilateral Pigmentary Retinopathy**
  - Early
    - typically asymptomatic
  - subtle paracentral scotoma (loss of vision around the central area of focus)

- **Bull’s Eye Maculopathy** (ring of pigment loss around the central area of the retina)
  - Central vision depression

- **Wide Spread Retinal Pigment & Retina Cell Atrophy (Death)**
  - Central Vision Loss
  - Peripheral Vision Loss
  - Nyctalopia (Difficulty with Night Vision)
2011 American Academy of Ophthalmology Plaquenil Screening Recommendation

- Baseline exam prior to institution of therapy
- Screening after 5 years of treatment, if no high risk characteristics
- Screening sooner with high risk characteristics.
- Examination includes, but not limited to biomicroscope,
  - 10-2 Humphrey Visual Field
  - Fundus Autofluorescence
  - Spectral Domain-OCT imaging
  - multifocal-Electroretinogram


Marmor MF, Melles MB. Disparity between visual fields and optical coherent tomography in hydroxychloroquine toxicity. Ophthalmology 2014;121:1657-1662
2011 American Academy of Ophthalmology Plaquenil Screening Recommendation

- Newer studies demonstrate Plaquenil® at “safe doses” can still lead to toxicity
- Emphasis is on Prevention of Rare cases of Toxicity
- Identify Pre-existing Retinal Disease
- Early Identification
- Preservation of Function/limit potential visual deficit

Ask a Question!

- As part of the Ask the Expert series remember to submit your question on **Lupus & the Eye** by **July 15, 2016** to [www.lupus.org/resources/submit-a-question-for-lupus-expert-qas](http://www.lupus.org/resources/submit-a-question-for-lupus-expert-qas).
- Check back in **August** for answers to 15 selected questions.
- If you are viewing this at a later date we encourage you to call our Health Educators at 1-800-558-0121 for any further questions.
What do YOU think about the *Ask the Experts* series?

Please click on this link to take a short survey and share your thoughts!

Your feedback is valuable to us, thank you!