

Ocular Inflammation and Autoimmunity: Sjogren's Syndrome, Rheumatoid Arthritis, Lupus, Ankylosing Spondylitis, Giant Cell Arteritis, Grave's Disease

I: Autoimmunity 101: How does our body learn to destroy itself?

Let's use Sjogren's syndrome as an example of this pathophysiology.

1. The etiology of autoimmune diseases is still unknown. However, 2 major theories exist: a viral attack and a local loss of immune protection
2. Viral etiology: The ocular mucosal tissues are exposed to all forms of viral species. Viruses can travel from the surface to invade the lacrimal gland could certainly cause an autoimmune response. Changes in cytokine expression of lacrimal acinar cells would activate both T and B immune cells that are present in the healthy lacrimal gland.
3. Failed local immunity: Inflammation in and around the lacrimal gland from any cause could create a breakdown in immune protection. Sjogren's syndrome, then, becomes a disease of failed local immunity as stimulated acinar cells become antigen presenting cells. MHC class II positive epithelial cells initiate an autoimmune response by presenting their own internal proteins on their surfaces to reactive T lymphocytes and thus activate these cells at the local level. This form of inflammation has been noted in several ocular conditions including corneal transplant rejection, uveitis and proliferative vitreoretinopathy. Dendritic, T and B cells also play a role in this process.
4. This same sort of pathophysiology applies to our understanding of RA, SLE and autoimmune thyroiditis.

II: Classic Cases:

1. exophthalmos eyes, diplopia, abnormal visual fields and indistinct nerve margins
2. ocular pain, visual loss, headache
3. red eyes, dry eyes, pain
4. dry eye, fatigue
5. red eyes deep pain and blur

III. History followed by ocular and systemic workup:

1. Thyroid:

History: ask about fatigue, restlessness, sweat, hair loss, swallowing problems.
Ocular examination: lids and globe position, EOM's, use fluorescein to observe poor coverage, do visual fields, observe ON's

Lab tests: thyroid peroxidase

2. Ankylosing spondylitis:

History: back pain

Ocular tests: all normal uveitis tests including ruling out posterior uveitis.

Lab tests: HLA-DR, X rays

3. Rheumatoid arthritis:

History: joint pain and inflammation, fatigue, skin rash, myalgia, morning stiffness

Ocular examination: staining, Schirmers, corneal integrity, episcleritis, scleritis, uveitis anterior and posterior, vasculitis.

Lab tests: RA, anti-CCP

4. Systemic Lupus Erythematosus:

History: varied, fatigue, pain, myalgia, arthralgia, rash butterfly

Ocular tests: Stain, Schirmers, episcleritis, scleritis, uveitis, vasculitis

Lab tests: ANA, anti-dsDNA

5. Giant Cell Arteritis:

History: pain, fatigue, vision loss intermittent, pain with chewing

Ocular tests: VA, palpate temporal arteries

Lab tests: ESR, CRP, biopsy of temporal artery

6. Sjogren's Syndrome:

History: dry eye, dry mouth, fatigue

Ocular tests: Stain, Schirmers

Lab tests: Ro, La, ANA

New Classification:

American College of Rheumatology approved new SS Classification Criteria.

Symptoms of dryness accompanied by:

2 of the following 3

1. Positive anti-Ro and/or anti-La OR Positive RF and ANA $\geq 1:320$
2. Labial salivary gland biopsy with focus score of ≥ 1 focus/4mm
3. KCS with ocular surface staining score of ≥ 3

Note no Schirmer scores

Staining methods are as in past nasal/cornea/temporal

Cornea:

Fluorescein staining of cornea: to be graded between 4-8 minutes after staining:

- a. 1-5 punctate epithelial erosions =1
- b. 6-30 punctate epithelial erosions =2
- c. greater than 30 =3
- d. add 1 score if central stain (4mm)
- e. add 1 score if filaments anywhere
- f. add 1 score if confluent staining anywhere

Maximum score per cornea is 6

Conjunctiva:

Lissamine green 1% liquid, immediate observation

- a. 0-9 dots is grade 0
 - b. 10-32 dots is grade 1
 - c. 33-100 dots is grade 2
 - d. greater than 100 dots is grade 3
- (any confluent staining is considered grade 3)

Maximum score for each temporal and nasal conjunctiva is 3.

Abnormal score is 3 or more in combined scoring of fluorescein and lissamine green.

IV: Differential Diagnoses:

1. dry eyes: primary or secondary Sjogren's: secondary includes RA and SLE, CREST
2. red eyes: primary or secondary SS: secondary includes RA, SLE and CREST, RA
3. uveitis: RA, ankylosing spondylitis, SLE