Systemic Vasculitis in the Eye

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Introduction

• What is uveitis?
• The eye and special considerations
• What can happen in the eye?
• Ocular involvement in specific diagnoses
• What to look out for
The Eye

• Clear structure: we can SEE the inflammation
• May be a sign of disease activity/recurrence
• The eye is privileged: may need higher treatment dosage
• Also many forms of vasculitis limited to the eye
What does vasculitis do?

• Systemic vasculitis can affect different parts of the eye
  • Scleritis (wall of the eye)
  • Keratitis (cornea)
  • Orbital inflammation (behind the eye)
  • Uveitis (inside the eye)
  • Lacrimal gland, parotid gland
What is Uveitis?

- Part of ophthalmology that deals with inflammation and infection inside the eye
- From the Latin “uvea” meaning “grape”
- Can be related to systemic disease or isolated to eye
- May be limited to the front, middle, or back of the eye
- **Immune-mediated**, malignancy, infectious causes
- Can be blinding
Orbit

- The eye’s bony home
- Inflammation behind the eye can cause eye to bulge (proptosis)
- Can damage optic nerve
- Granulomatosis with polyangiitis
- Polyarteritis nodosa
- Churg-Strauss
Sclera

Scleritis

- Granulomatosis w/ polyangiitis
- Polyarteritis Nodosa
- Churg-Strauss
- Microscopic polyangiitis
- Connective Tissue Disease
- Idiopathic
Cornea

Peripheral Ulcerative Keratitis
Lens

Cataracts
Vitreous

Vitritis
Optic Nerve
Retina/Choroid
Optical Coherence Tomography

Normal retina
Inflammation of the retina

- Vasculitis in the retina can severely impact vision
- Can be isolated to the eye or associated with systemic disease
  - Any of the systemic vasculitis disorders can rarely involve retina
- Symptoms may be mild or severe
  - Flashes, floaters, blurred vision, blind spots
- Followed by examination and different types of imaging
- May need to be treated aggressively
What can happen in the retina?
Retinal Vasculitis
Vasculitis & Fluorescein angiography
Swelling in the retina
Central retinal vein occlusion
Arteriolar Occlusion
Lack of blood flow to retina
Neovascularization

- Vasculitis $\rightarrow$ blocked vessels $\rightarrow$ damaged retina
- Abnormal new vessels grows $\rightarrow$ bleeding in eye
Treatment Basics

- Treat the underlying disorder
  - Steroids first
  - Many require immunomodulatory therapy
    - Adalimumab (Humira), infliximab (Remicade), rituximab, tocilizumab (Actemra)
    - May require higher doses than for systemic disease

- Can supplement with directed therapy to eyes
  - Steroid injections behind the eye
  - Steroid injections and implants in the eye
  - Injections for abnormal blood vessel growth

- Close monitoring
The Specifics

• Large vessel vasculitis
  – Giant Cell Arteritis
  – Takayasu’s Arteritis

• Medium vessel vasculitis
  – Kawasaki’s Disease
  – Polyarteritis Nodosa

• Small vessel vasculitis
  – Granulomatosis with Polyangiitis
  – Churg-Strauss

• Others
  – Behcet’s Disease
  – Susac Syndrome
Giant Cell Arteritis

- Inflammation of larger arteries
- Temporal artery, branches of carotid artery, aorta
- Older people
- Jaw pain while chewing
- Pain in temples
- Double vision
- Loss of vision
Giant Cell Arteritis

Eye findings

- Inflammation of vessels supplying optic nerve/retina
  - Arteritic ischemic optic neuropathy
  - Central retinal artery occlusion
  - Loss of vision on one side, can progress to other side
  - Vision may not return
Giant Cell Arteritis

- Cranial neuropathy
  - Damage to cranial nerves
  - Control eye movement
  - Double vision
- High dose steroids
- Biopsy temporal artery
- Guarded prognosis
Takayasu’s Arteritis

- Inflammation of large arteries like aorta
- Can cause inflammation and blockage of vessels in the retina and optic nerve
- Artery & vein occlusions
- Optic neuropathy
- Inflammation in front, middle, and back of eye
Polyarteritis Nodosa

- Inflammation of small-medium arteries
- Eye findings in ~ 20%
- Scleritis
- Peripheral ulcerative keratitis
- Blocked vessels in retina and choroid
- Orbital inflammation
  - Behind the eye, can push eye out (proptosis)
- Can affect cranial nerves (double vision)
Granulomatosis with Polyangiitis

- Inflammation of small vessels
- Affects lungs, sinus, kidneys, eyes
- Ocular findings in 25-50%
- Scleritis
- Peripheral ulcerative keratitis
- Retinal vasculitis (5-10%)
  - Causes blocked vessels and decreased vision
- Inflammation of vessels supplying optic nerve & cranial nerves
- Orbital inflammation (behind eye)
Churg-Strauss Syndrome

- Inflammation of small arteries and veins
- Eye findings uncommon
- Peripheral ulcerative keratitis
- Inflammation in front and middle of the eye
- Blockage of retinal arteries and veins
  - Blind spots, decreased vision
- Optic nerve inflammation
- Orbital inflammation
- Damage to cranial nerves \(\rightarrow\) double vision
Behcet’s Disease

- 70% develop ocular findings
- Chronic disease affecting retina and front of eye
- Typically described among those of “silk road” ancestry
  - We have patients of all races and ethnicities
Behcet’s Disease

- Wall of the eye (scleritis)
- Front of eye (anterior uveitis)
- Retinal vasculitis
  - Can cause blockages
- Patches of retinitis
  - Inflammation of retina itself
- Optic nerve damage
Susac Syndrome

- Inflammation of small arteries
- Affects retina, inner ear, brain
- Multiple arteriolar occlusions in retina
Things to look out for

• Certain symptoms don’t necessarily mean a specific finding
• It never hurts to have an eye exam!
• Red and painful eye
• Floaters, flashes
• Eye bulging out
• Double vision
• Blind spots
• Decreased vision
• Distorted vision
Conclusions

• Eye involvement of systemic vasculitis can have serious effects
• Vasculitis can effect many parts of the eye
• Treatment is a team effort between ophthalmologist and rheumatologist
• Be evaluated if you have any concerning symptoms
  – Uveitis specialist is ideal, but any ophthalmologist is great!
Thank you’s

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Thank You—Questions?