Uveitis
Diagnosis & Management
Introductory Lecture Series
Part I

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Definitions

- **Uvea:**
  - Iris
  - Ciliary Body: Pars Plicatta, Pars Plana
  - Choroid

- The term “Uveitis” generally describes ocular inflammation that is somehow related to these ocular structures.
Anatomical Subtypes

- Anterior
- Intermediate
- Posterior
- Panuveitis
History

• Pain, Redness & Photophobia
  – Acute vs. Chronic

• Decreased Visual Acuity
  – CME, Cataract, Media Opacities, etc...

• Flashes & Floaters
  – Posterior & Intermediate Uveitis Respectively.
History

• Past Medical History
  – DM, HTN, Osteoporosis, Lipids, etc…
  – Rheumatological Diseases
  – Infectious Diseases, STDs
Review of Systems

- Oriented to the differential diagnosis of the particular uveitic syndrome present.
  - Ex. Intermediate Uveitis…
    - DDx: MS, Sarcoidosis, Lyme Disease, Luetic…
    - Ask for neurologic symptoms, etc…
  - If you are not sure ask for questions on all systems on a systematic fashion…
Physical Examination

• Cells
  – Occur due to the breakdown of the blood-ocular barrier.
  – Grading systems allow to assess the adequacy of treatment and presence of persistent inflammation.
Grading Systems: A/C Cells

3mm x 1mm slit beam:

- 0 = Quiet
- 1 - 4 = trace
- 5 - 10 = 1+
- 11 - 20 = 2+
- 21 - 50 = 3+
- > 50 = 4+

Avoid...
- Rare
- 1 to 2+
Grading systems: Flare

No Flare = 0
Faint = 1+
Iris and Lens Clear = 2+
Iris and Lens Hazy = 3+
Fibrin & Plastic Aq. = 4+

Note: There is no such thing as trace flare…!
Grading Systems: Vitreous Cells

3mm x 1mm slit beam:

0 = Quiet
1 - 10 = 1+
11- 20 = 2+
21- 50 = 3+
> 50 = 4+

Note: There is no such thing as trace vitreous cells...!
Grading Systems: Vitreous Flare/Haze

NFL clearly visible = 0
ON & vessels clear. Hazy NFL = 1+
Hazy ON & vessels = 2+
O N visible = 3+
ON not visible = 4+
Treatment

- Steroids
  - Topical
  - Periocular (Sub-Tenon’s)
  - Intraocular

- Steroid Sparing Agents (Immunomodulation)
  - Anti-Metabolic: Methotrexate, CellCept, Azathioprine
  - Anti T-Cell: Cyclosporin-A
  - Akylating: cyclophosphamide, Chlorambucil
Topical Steroids

- Prednisolone Acetate 1% (Pred Forte)
  - Maximum Dose is q1hr

- Side Effects:
  - PSC Cataracts
  - Glaucoma (takes 3 weeks to ensue)
Topical Steroids

- Rimexolone Acetate 1% (Vexol)
  - Almost as good as Pred Forte
  - Less likely to cause Ocular Hypertension
Periocular Steroids

- Posterior Sub-Tenon’s Kenalog
  - 40mg Dose
  - Use 25 G Needle Only
  - Half Life 6 weeks
Posterior Sub-Tenon’s Kenalog

- Good for induction treatment when systemic steroids are better avoided
  - Ie. Patients with uncontrolled DM
- Good for CME

- Irreversible:
  - Avoid if OTN is a concern
  - Avoid if Viral Etiology is being Considered
  - Avoid if patient has chronic disease
**Intraocular Kenalog**

- Dose is 4MG
- Higher doses can be achieved if concentrated steroid is used
- Good for CME
- Rarely Needed
- Irreversible:
  - Avoid if OTN is a concern
  - Avoid if Viral Etiology is being Considered
  - Avoid if patient has chronic disease
 Prednisone

- Start 1mg/kg po qd.
- Do not exceed 60 mg/day!
- **Always** prescribe along with Calcium supplementation!
Prednisone...

- Weight & arterial blood pressure on every visit...

- Monthly basic chemistry.

- Bi-annual serum lipids.
Prednisone...

- Yearly DEXA Scan...

- Remember...
  - Osteopenia and Osteoporosis are treatable conditions...
The “Standard” Prednisone Taper

Always on a weekly basis:

First by ten:

60, 50, 40…

Then by 5:

35, 30, 25, 20…

Then by 2.5:

17.5, 15, 12.5, 10, 7.5, 5, 2.5, stop

• F/U on a monthly basis or sooner if a threshold is reached!
Steroid Sparing Agents

• Very Strong:
  – Akylating agents: cyclophosphamide, chlorambucil.

• Moderate to strong:
  – CellCept, Imuran.

• Mild:
  – Cyclosporine, Methotrexate
  – Probably as effective as prednisone itself.
Case # 1

- 57 y/o woman presents with a 2 day history of pain, redness and photophobia OD.
- Had a similar episode 2 years ago OS which was treated with a “white drop”, her symptoms resolved without sequelae.
- 1ry ophthalmologist did a Rheumatoid factor and a ANA test, both were negative.
Case #1

**OD:**
- Va sc: 20/30
- Ta: 16
- A/C: 3+ C, 1+ Flare
- Lens: Clear
- Vitreous: Anterior +1 C
- Retina: Flat, no CME

**OS:**
- A/C: D/Q
- Focal posterior Synechia at 2 o’clock, trace PSC.
Case # 1

- What is the ocular diagnosis?
- What is the most likely etiology?
- What questions would you ask her?
- What other work-up should be ordered?
- Did the 1ry ophthalmologist order the correct work-up?
- How would you treat this patient?
- What complications may she develop?
Case # 1

- On further inquiry the patient had history of large joint, episodic, asymmetric, non-deforming polyarthritis which she ascribed to an old sports injury. She also had history of a heel spur, and recurrent cystitis with negative cultures.
  - What is the diagnosis now?

- The patient was treated successfully with q1 hr Pred Forte. Within 2 weeks she was quiet. She was then tapered off over 6-8 wks. She recovered 20/20 VA.

- She was HLA-B27 positive.
Acute Anterior Uveitis...

- Acute, Unilateral, Recurrent, Alternating, Anterior Uveitis...
  - If Acute Unilateral: ~50% HLA-B27 (+)
  - Acute, Unilateral, Recurrent: ~70% HLA-B27 (+), if Alternating >80%.
  - 50% of HLA B27 (+) anterior Uveitis have an underlying Seronegative Spondyloarthropathy.
  - The review of Systems Should be directed towards diagnosing Ankylosing Spondilytis, Reiters, Psoriatic Arthritis, IBD associated AS, etc…
Trivia Question!

• What is the most common cause of hypopion associated anterior Uveitis?
• HLA-B27
• Remember HLA-B27 uveitis is far more common than Beçet’s Disease...
• Beçet’s causes hypopion uveitis in a quiet eye, HLA-B27 eyes are generally angry!
Case # 2

• 33 y/o AA woman. 7mo h/o progressive VA loss. 5mo ago she noticed slight pain 3/10 on a sunny afternoon, but her eyes were never “really” red.

• She then went to a local ophthalmologist who prescribed a “milky drop” which he told her to take every 2 hours for a week, this made her eyes feel better, he then told her to taper down the drops over 2wks because “she could get a cataract”. She now comes for a second opinion.

• She said she was tested for “lupus and syphilis” and the tests were normal.
Case #2

- Va cc: 20/70
- Ta: 10
- A/C: 4+ C, 3+ Flare
- Lens: ASC +2, PSC + 3
- Vitreous: Anterior +2 C
- Retina: CME, otherwise wnl.

- Va cc: 20/200 ph 20/150
- Ta: 8
- A/C: 3+ C, 3+ Flare
- Lens: PSC + 2
- Vitreous: Anterior +2 C,
- Retina: Florid CME, otherwise wnl.
Case #2

- What is the ocular diagnosis?
- What are the most likely etiologies?
- What questions would you ask her?
- What other work-up should be **ordered**?
- How would you treat this patient now?
Case #2

- You called the previous ophthalmologist and received a fax with a (-) RPR and (-) ANA.
- On further questioning she admits to a “rash” which has not gone away (see Figure)…, otherwise ROS is (-).
- You order an FTA-Abs, a CXR and start Pred Forte (16,16) & AS 1%.
Case #2

• She comes in 1wk for F/U. She still has 2+ cells bilaterally, her cc Va is now 20/50 OD and 20/125 OS. CXR & FTA were (-). The dermatology consult was (+) for non-caseating granulomata. Her rheumatologist wants to start her on Plaquinil 200 po bid.

• How should you manage her ophthalmological treatment?
• What advice should you give regarding starting Plaquinil at this juncture?
Case #2

- She comes for her F/U after being on PF q1hr for one month. Her Va is unchanged, she still has trace cells OD and 1+ cells OS.
- The Plaquinil was held and the dermatologist opted for a topical treatment.

- How would you manage the patient now?
- Any supplements, monitoring?
Case # 2

- You treated her with 60mg oral prednisone, she was absolutely quiet after 4 wks, her Va was now 20/30 OD and 20/70 OS. You then started a “standard” taper and followed her on a monthly basis.

- On the second month, she comes at a dose of 17.5mgs of prednisone, she is asymptomatic but you notice 1+ cells OU. Her VA is now 20/40 OD and 20/100 OS.

- How would you manage her now?
Case # 2

- You upped the prednisone back to 60mgs and started CellCept at 1g bid. She becomes quiet at a week, you start to taper the prednisone and are able to taper her all the way to 10mgs in 3 months without a flare-up.

- What labs should you be getting at this juncture and at what frequency?
- What are the most common side effects of CellCept?
- How should you manage her now?
Case # 2

- She comes for a routine monthly F/U. She is now at 5mgs of prednisone daily. VA is 20/40 OU, she is quiet, no CME, there is PSC +2 OU. She complains of muscle stiffness and pains. Her AST is 55 (slightly elevated).

- What is the cause of her stiffness?
- How should you manage her now?
Case # 2

- It is a year after her initial visit with you, she is off the prednisone, and remains on CellCept 1g bid. LFTs are normal.
- She complains of glare impairing her driving at night, there is no CME, her BCVA is 20/50, J8 OU, she has bilateral PSC +3.
- What precautions should you have for her phaco?
- How would you manage her medications?
The “Standard” Prednisone Taper

Always on a weekly basis:

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A Word about Uveitis Workup

- **Syphilis and Sarcoidosis**
  - Both Syndromes can Imitate almost any type of Uveitis
  - **All Patients** must have:
    - RPR
    - FTA-Abs: 20% of Syphilitic Uveitis is RPR (-) and FTA-Abs (+)
    - CXR: 87.5% Sensitive in detecting Sarcoidosis.
  - All a other tests should be dictated by the PMHx and ROS
Conclusion

• Acute Uveitis Should be treated aggressively to avoid structural complications and should be expected to resolve in 6-8 weeks.

• Chronic Uveitis requires treatment “forever”. You only taper down to find thresholds and if these are found treatment should be held above the threshold in order to prevent complications from structural complications.

• Always remember to seek infectious causes, use common sense:
  – Unilateral follicular conjunctivitis with uveitis => HSV
  – Syphilitic uveitis is not treated adequately until the patient receives IV treatment.