Uveitis & Ocular Immunology
Introductory Lecture Series
Intermediate Uveitis

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Uveitis appetizer of the day!

- Not related to the conference but nonetheless extremely important!
- If a person comes to your office with history of unilateral uveitis that presented with increased IOP and later developed corneal (endothelial) decompensation…
- What is your working diagnosis?
- If the same person has a positive Rheumatoid Factor and Rheumatoid Arthritis what is the working diagnosis?
- If the same person had a positive ANA test, what is the working diagnosis?
**Herpes Simplex Virus Uveitis**

- Unilateral Granulomatous Uveitis with Iris transillumination defects in a patient that suffers recurrent cold sores who has corneal aesthesia and possibly, high IOP and a corneal stromal haze.
- **Treatment:**
  - Acyclovir 400mg 5times/d, Pred Forte and Atropine.
- **Note:** Acute if treated very early otherwise may become chronic.
HSV Uveitis

Iris Atrophy 360
Herpes Zoster Virus Uveitis

- Unilateral Granulomatous Uveitis with Iris transillumination defects in a patient that suffered V1 shingles that likely affected the nasociliary nerve, who may have corneal aesthesia and possibly high IOP and possibly a corneal stromal haze.

- **Treatment:**
  - Acyclovir 800mg 5times/d, Pred Forte and Atropine.

- **Note:** Look at the fundus and R/O Acute Retinal Necrosis.
Zoster Uveitis

Note: Segmental Iris Atrophy
Uveitis that may have high IOP upon presentation.

- DDX for the OKAPS:
  - HSV & VZV
  - CMV
  - Toxoplasma
  - Posner-Schlossman Syndrome (Dx of Exclusion)
    - Rare in real life, common in the OKAP!

After the initial presentation, the differential of glaucoma in uveitis patients includes: anterior synechiae, bombè, steroid response, and pigment dispersion (HSV/VZV).
Now back to Intermediate Uveitis!
History

- 1908: Fuchs - Cyclitis
- 1950: Schepens - Peripheral Uveitis
- 1960: Brockhurst - Peripheral Cyclitis
- 1960: Welch - Pars Planitis
- 1961: Gass - Vitritis
- 1987: IUSG* - Intermediate Uveitis

*International Uveitis Study Group
Definition

Syndromes of Predominantly Vitreous Inflammation
Differential Diagnosis

• Most Common:
  – Idiopathic (most common)
  – Multiple Sclerosis
  – Sarcoidosis

• Least Common:
  – Lymphoma
  – Toxocara canis
  – HTLV-1
  – Becets & Other Vasculitides

• Also Possible:
  – Syphilis
  – Lyme Disease
  – Bartonella

  – Resolved Iridocyclitis
  – Retinitis Pigmentosa
  – Undiagnosed (retinitis/choroiditis)
Epidemiology

- 10% of uveitis patients.
- Wide age range population
  - Childhood to Elderly population
- No true gender or race predilection.
- Bilateral 80% of time.
  - Often Asymmetric.
Presentation

• Most Commonly:
  – Floaters, Decreased Vision, Photophobia
  – White Eye, No Pain, Few KP, 1+ A/C Cells, No Synechiae, CME, Deep Vitreous Cells

• Occasionally:
  – Pain, Redness, Photophobia & Posterior Synechiae on First Episode
  – Then becomes chronic with predominant vitritis.
Clinical Findings

• Main Features:
  – Sine qua non: Vitreous Cells

• May or may not have:
  – Snow Balls
  – Snow Banking

• If all three present the diagnosis is likely either *Pars Planitis “syndrome”* which is often associated to multiple sclerosis or rarely CNS lymphoma.
Clinical Findings
Complications

- CME
- Cataracts
- Glaucoma
- Optic Disk Edema
- Optic Neuritis
- Retinal Detachment
- Venous Sheathing (may not have vasculitis)
- Vasculitis (may not have venous sheathing)
- Retinal Neovascularization
- Vasoproliferative Tumor of the Retinal Periphery
Complications: CME

- Up to 50% of patients
- Most common cause of visual loss in intermediate uveitis patients.
- Prevalence increases with the severity of inflammation.
Complications: Cataracts

- May be related either to the inflammation or corticosteroid use.
- Most often PSC.
- PE & IOL has good prognosis (if measures taken).
- Should **never** hold corticosteroids in an active eye to prevent cataract formation.
Complications: Glaucoma

If the IOP is increased:

1) Make sure to R/O Herpes & Toxoplasmosis. (not intermediate uveitides)
2) Do gonioscopy and make sure you are not dealing with synechial angle closure.
3) If the angle is open consider patient as steroid responder and switch to Vexol (rimexolone).
4) Never compromise adequate treatment for a high IOP, if needed “Filter, Shunt and Treat”!
Complications: Optic Nerve Swelling

- **Disk Edema**
  - Up to 20% of eyes
  - Most often secondary to intraocular inflammation.
  - If bilateral consider MRI

- **Optic Neuritis:**
  - 7.5% of patients
  - Patients may or may not have multiple sclerosis
  - Treat as in ONTT.
Complications: Retinal Detachment

- **Serous Retinal Detachment**
  - May be shallow and peripheral (benign).
  - May be aggressive, bilateral and associated with aggressive disease and poor prognosis, (malignant).
  - Must not confuse with VKH or sympathetic ophthalmia (IVFA, fundus findings help differentiate this entities).

- **Rhegmatogenous Retinal Detachment**
  - Often associated with dialysis at the snow bank
  - Poor prognosis (PVR).
Complications: Venous Sheathing

- If present must consider and IVFA to R/O vasculitis.
- Most often benign in nature.
- May point towards certain etiologies (ie. Candle wax drippings)
- Generally may be observed if < 270 degrees
Complications: Venous Sheathing

Vs.
An OKAP moment...
An OKAP moment...

If you get this picture on you OKAP…

What is your answer?
Sarcoidosis
Complications: Vasculitis

- Vessel wall inflammation with associated ischemia
- Often heralded by the presence of NV
- May or may not have associated sheathing
- Must consider a systemic vasculitis (ie. Becets).
Complications: NVD & NVE

- If associated with capillary drop-out treat with PRP.
- If no capillary drop-out is present immunosuppress the patient (ie. Corticosteroids, Cell Cept, etc...)
- If TRD is present consider PPV.
Complications: VTRP

- Vasoproliferative Tumor of the Retinal Periphery
- Described by Dr. Shields and Associates
- Among a series of 113 tumors of different origin 13% were associated with IU.
- Generally implies severe disease (IU).
Differential Diagnosis

- **Most Common:**
  - Idiopathic (most common)
  - Multiple Sclerosis
  - Sarcoidosis

- **Least Common:**
  - Lymphoma
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  - Becets & Other Vasculitides

- **Also Possible:**
  - Syphilis
  - Lyme Disease
  - Bartonella
  - Resolved Iridocyclitis
  - Retinitis Pigmentosa
  - Undiagnosed (retinitis/choroiditis)
**Work-Up**

- **Ophthalmological Exam**
  - Toxocara Canis, Toxoplasma gondi, Birdshot, AZOOR, Intraocular Lymphoma, Retinitis Pigmentosa.

- **History and Physical Exam:**
  - MS, Sarcoidosis, Becet’s, HTLV-1

- **Laboratories:**
  - FTA-Abs, RPR, CXR (Always)
  - Consider: MRI, Lyme Ab, Bartonella Ab.
Clinical Course

- 25% follow a uniphasic course.
  - Quiet after a single PSTK or Oral Steroid Course and never recurs.
- 50% follow an indolent chronic course.
  - Requires chronic suppression with prednisone 10 or immunomodulation.
- 25% follow a recurring course
  - Quiets for long after a single course of treatment but recurs later on.
Treatment

• Indications for treatment:
  – Vitritis
  – CME
  – “True” Vasculitis
  – Retinal Detachments
  – Neovascularization

• Factors to Consider:
  – Laterality
  – Severity
  – Clinical Course
  – Other Illnesses
    • DM, Obesity, Becets, CNS Lymphoma
Treatment

• General Rules:
  – **Always** suppress the anterior chamber reaction.
  – Always treat non 20/20 CME.
  – Avoid settling for partial results, always try to obtain a dry macula.

• Armament Arsenal Includes:
  – Topical, periocular, intraocular and oral steroids.
  – Antimetabolic Agents (CellCept, MTX, AZA)
  – Cyclosporin-A which may be added to the above.
  – Akylating agents:
    • Last resource for sight threatening disease
    • May consider for some vasculitides
Case # 1

- 28 y/o woman, presents complaining of floaters for one week. You notice inferior peripheral snowballs OU. 1+ vitreous cells OU. A/C is quiet. ROS is negative.
- BCVA is OD: 20/20 ; OS: 20/25
- OCT shows CME OS.

What is your work-up?
How do you manage her?
Case # 2

- 40 y/o obese and diabetic woman presents with floaters and photophobia OD. She has obesity and DM Type II. Exam shows BCVA OD: 20/80 ; OS: 20/20. OD has 1+ A/C cells, few NG KP. The lens is clear and there is florid CME (MS IV saw it!). Work-up and ROS are negative.

- How would you treat her?
- When should you consider systemic treatment?
- What is an adequate PSTK course of treatment?
Case #3

- Bilateral 20/30, 20/100 after referring MD treated with PF and PSTK x 5 (last one 6 weeks ago). IOP is 24, 32. Patient has active anterior and intermediate uveitis OU, Bilateral CME. Gonioscopy shows an open angle.

- How do you treat her?

- When should you consider CellCept.
• 34 y/o lawyer is referred to your practice due to “pars planitis”. He has a BCVA of OD: 20/100 ; OS: CF. He had a 6th nerve palsy, not completely recovered. ROS reveals history of mouth and genital ulcers, Erythema Nodosum, Arthritis & Diarrhea. He has active anterior and intermediate uveitis, the left retina shows foci of ischemic necrosis and NVE.

• What is the diagnosis?
• Is there a blood test for this?
• How would you treat him?
• A 29 y/o with a 7 year history of pars planitis comes for follow-up. She states that she had a seizure last month and that her family physician just started her on Dilantin. A HVF shows a left inferior quadrantanopsia.

• What is your next step of management?
• What is the likely diagnosis?
Case # 6

- A 18y/o boy is sent to you due to unilateral “pars planitis” because “he just does not seem to respond to Pred Forte”. See Fig.

- What work-up do you need?
- What is the diagnosis?
Case # 7

- Deer Hunter from West Virginia, presented with floaters and decreased vision one week ago. Lyme antibody testing is partially positive, he denies history of ECM.

- How would you treat him?
27 y/o woman complains of many flashes OS. On your exam OD has 1+ vitreous cells, an enlarged physiologic blind spot and a zone inferotemporal to the disk with Retinitis Pigmentosa like changes and vessel attenuation. BCVA is 20/20 OU.

What is your diagnosis?
What is your work-up?
Case # 9

- 50 y/o man is referred due to chronic vitritis on your fundus exam you notice the following changes OU:

- What is your diagnosis?
- How do you treat and monitor him?
Prognosis

• Depends on the severity of the inflammation as well as on the promptness and adequacy of treatment.

• Most series show 20-40% under 20/40 BCVA.

• Aggressive treatment has been shown to improve outcomes and prognosis in uveitis patients.