Case Presentation

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CC/HPI

- CC: eye pain OU, mild redness, headache, tinnitus, and change in color perception
- HPI
  - 34 yo F
  - Lebanese
  - Symptoms x 3mo
  - No improvement on Imuran 200mg qd (started at home)

Histories

- PMH: unremarkable
- SH: pharmaceutical engineer; ophthalmologist husband; two children; no EtOH/tobacco/illicit drug use
- FH: unremarkable

ROS

- Headaches
  - Frontal and retro-orbital
  - Throbbing quality
  - At times 7-8/10 in severity
  - Randomly throughout the day
  - Not relieved by Motrin or Tylenol
  - Not exacerbated by anything in particular
- Tinnitus
  - Bilateral
  - Almost constant
  - No change in hearing
- Physical exam unremarkable otherwise

Exam

Visual Acuity
OD Dva sc 20/20
OS Dva sc 20/20

Pupils
OD: equal, round, reactive, no APD
OS: equal, round, reactive, no APD

Cornea:
OD: clear and compact
OS: clear and compact

Iris:
OD: normal
OS: normal

Intraocular Pressure
OD 14mmHg
OS 14mmHg

Conjunctiva:
OD: trace injection
OS: trace injection

Anterior Chamber:
OD: 1.5+ cells
OS: 1.5+ cells

Lens:
OD: clear
OS: clear

Exam

Vitreous:
OD: 2+ vitreous cells
OS: 2+ vitreous cells

Optic Nerve:
OD: edema 4+
OS: edema 4+

CD Ratio v/h:
OD: difficult to assess
OS: difficult to assess

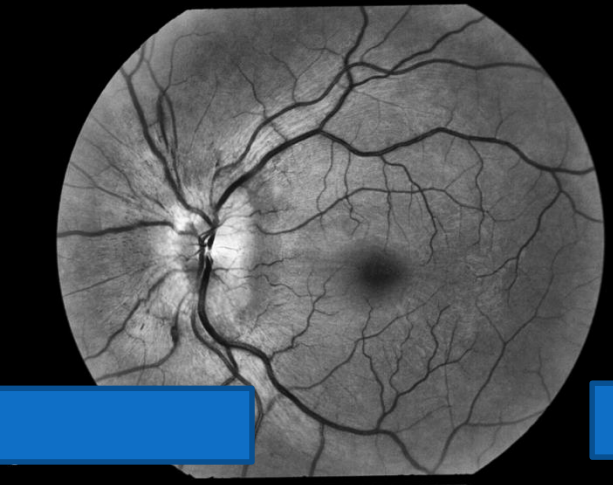
Macula:
OD: normal
OS: normal

Vessels:
OD: normal
OS: normal

Periphery:
OD: normal
OS: normal

Fluorescein Angiography

- Impressive late-phase disc staining OU
- No macular leakage
- No vasculitis
- Normal transit time
- Peripheral sweeps wnl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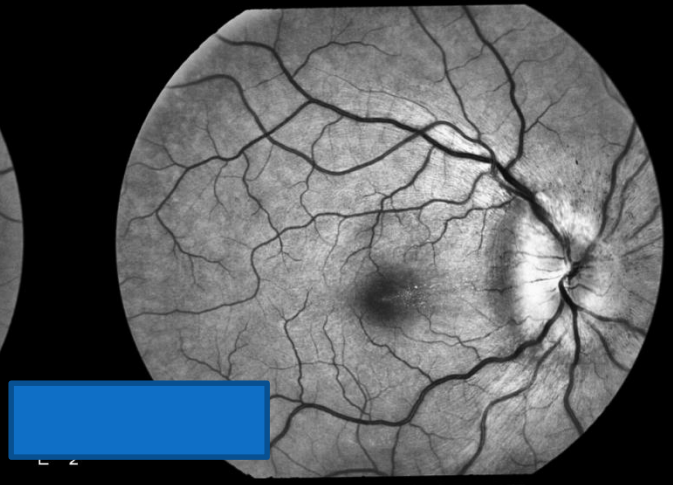
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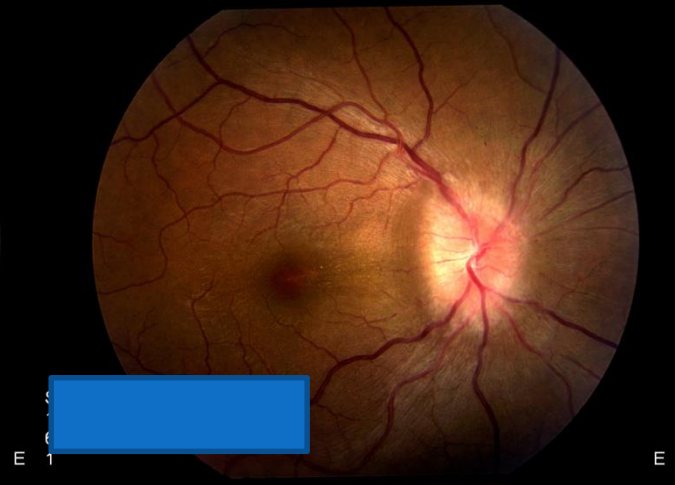
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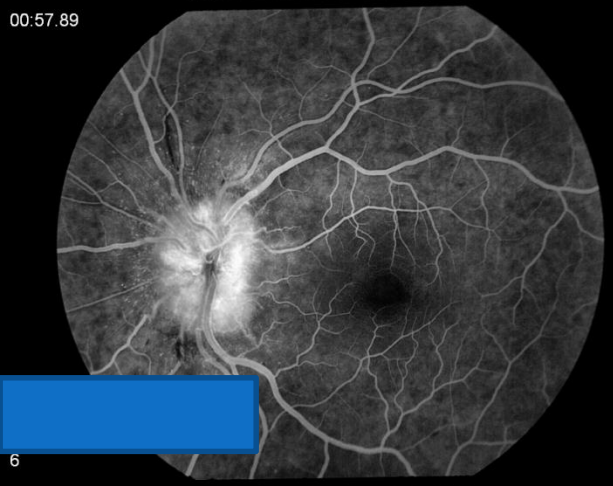
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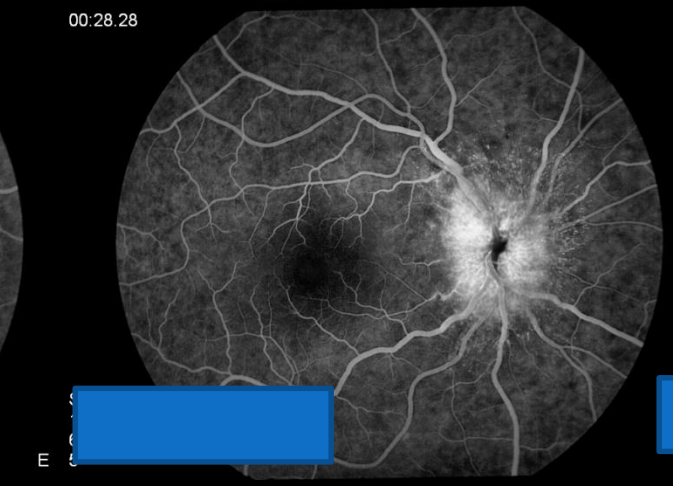
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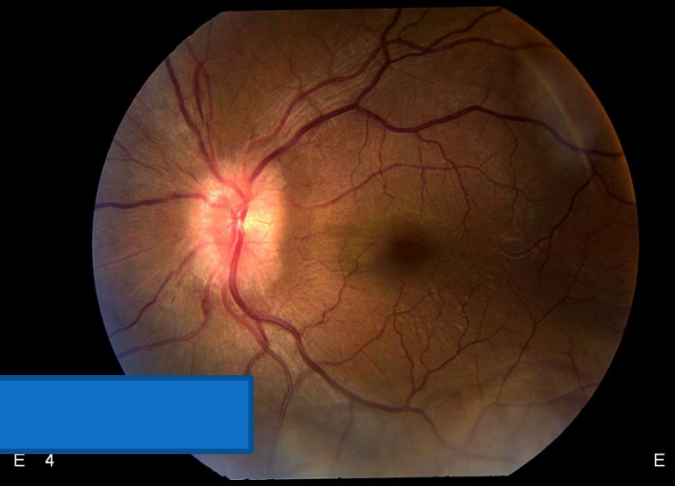
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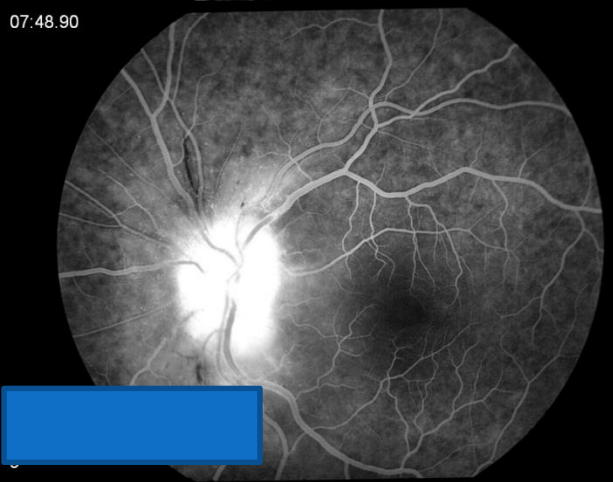

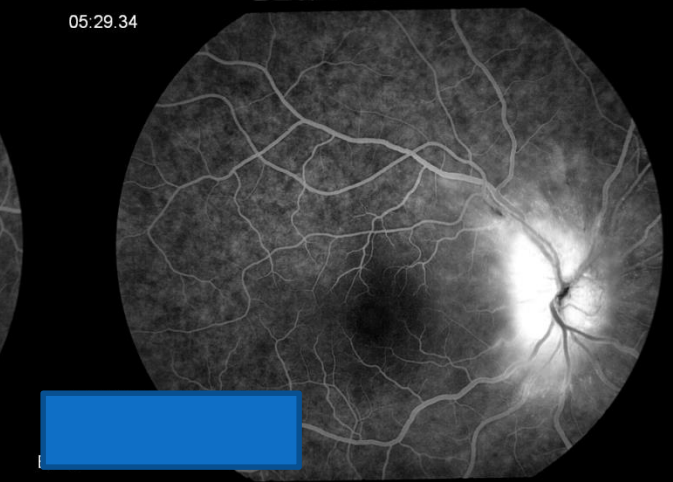

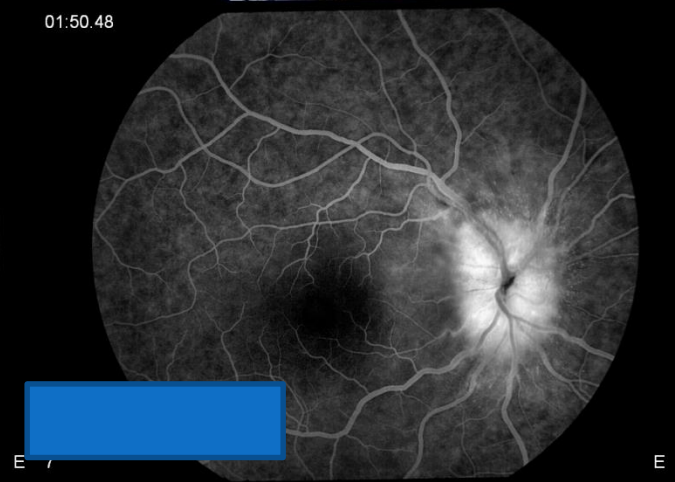
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Assessment

- Bilateral panuveitis with papillitis

Top Differentials?

- VKH
- ABD

Serologies

- ACE, lysozyme, ssDNA, dsDNA, ANA, ANCA, Bartonella, Brucella, leptospira, and FTA-ABS all negative
- Elevated C3 level (211)
- HLA-B51+

CSF Analysis

- Increased lymphocytes and macrophages with a few neutrophils (pleocytosis)

Treatment Plan

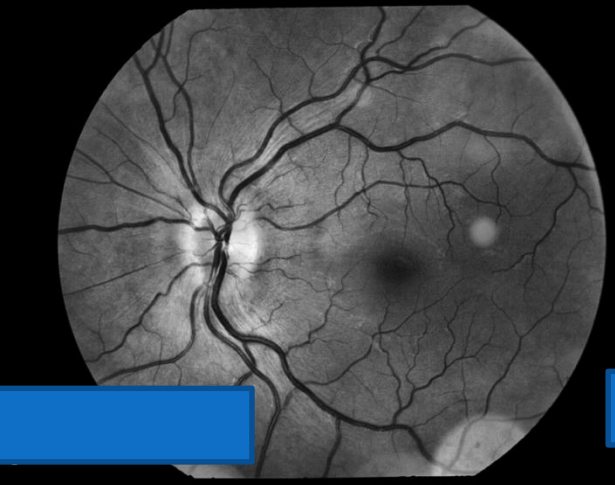
- In the absence of poliosis or hypopigmented skin lesions, **incomplete VKH** was diagnosed
- Cyclosporine 200mg added

Clinical Course

- 2 weeks later
  - Anterior and vitreous cells vanished
  - Headaches and tinnitus unchanged
  - FA: persistent ON inflammation
- CSA and AZA both boosted to 300mg

Clinical Course

- 6 weeks from 1st visit
  - SSx unchanged
  - Exam unchanged
  - FA: ON staining better but not completely normal
- IV solumedrol x 2
  - Initially improved but quickly worsened, both angiographically and symptomatically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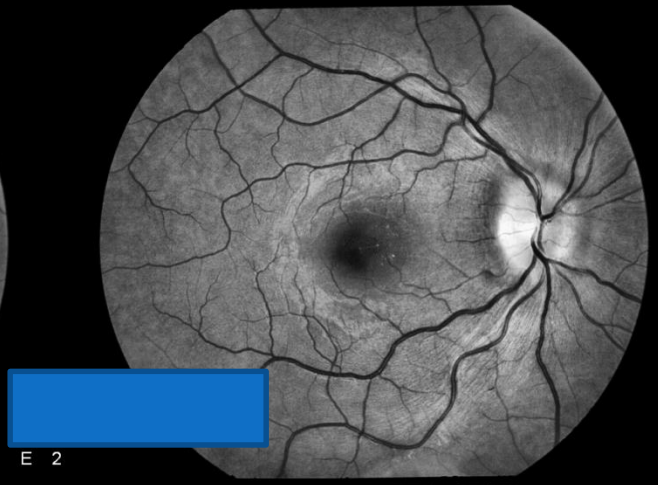
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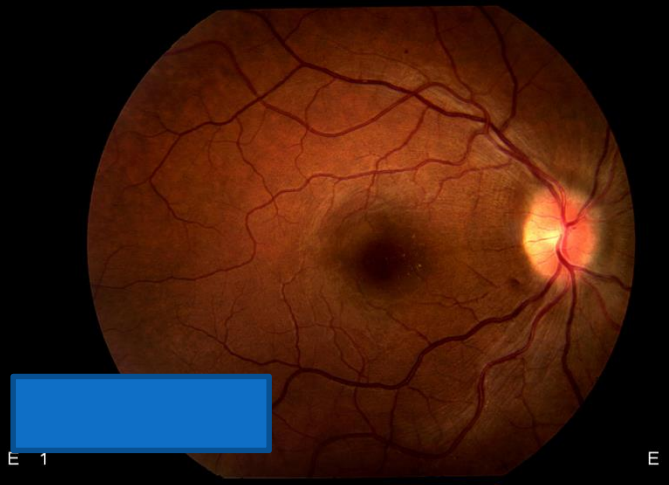
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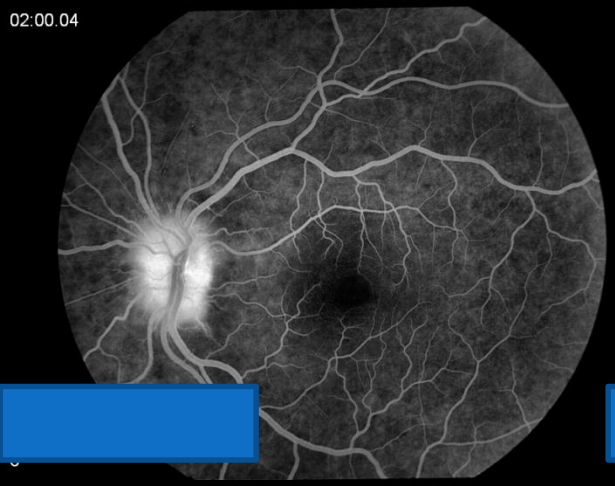
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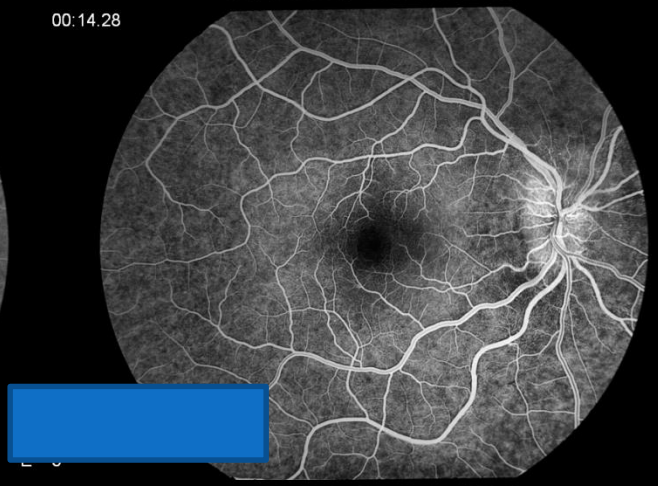
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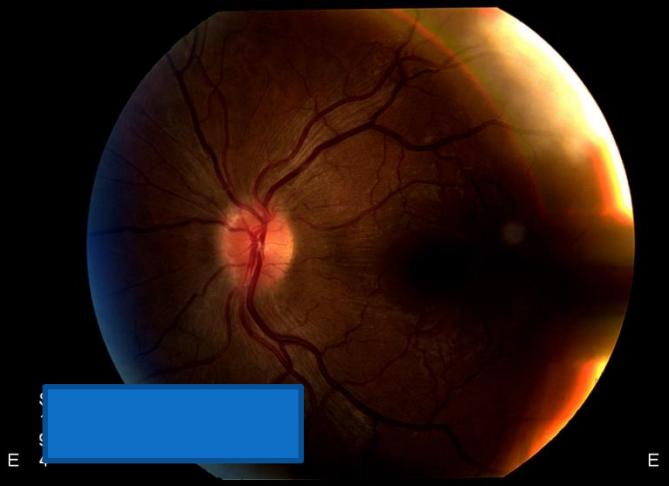
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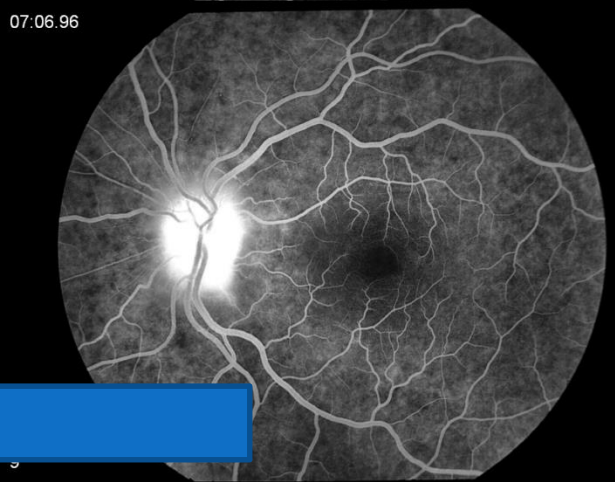

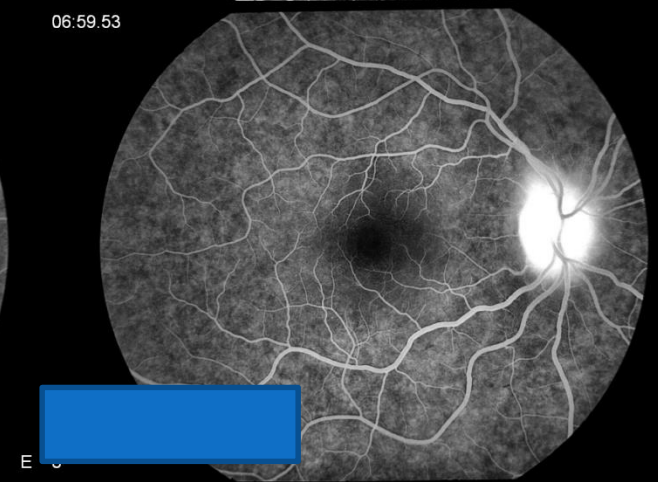
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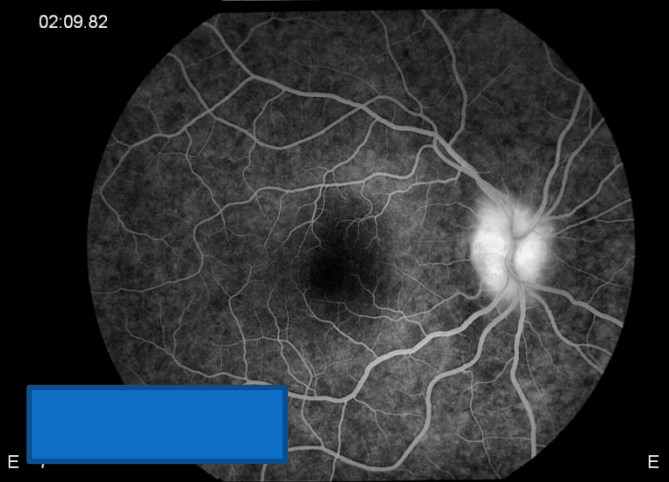
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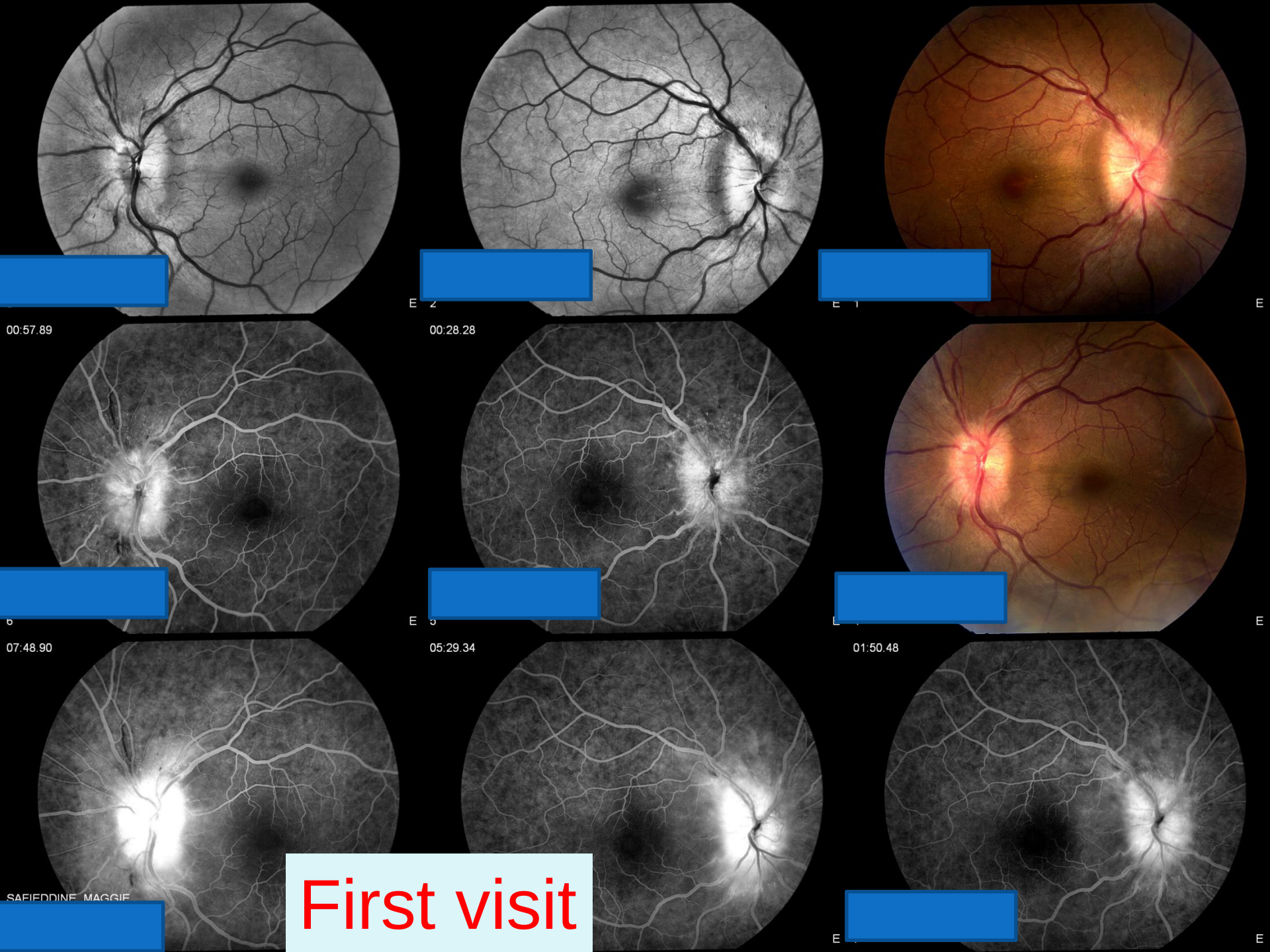
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First visit

Clinical Course

- Fatigue with IMT's
- Post-nasal drip, sinus trouble
- CSA and AZA withheld
- Serologies repeated
  - CMV IgG+
  - CMV IgM-
  - CMV DNA PCR+
- PO valganciclovir started

Clinical Course

- Headaches and tinnitus vanished following 2-week course of antiviral therapy
- Eye pain and redness much less frequent, relieved with PF 1-2x/day

Cytomegalovirus

CMV Diseases

- Fetus/Infant
  - Congenital CMV infection
  - Perinatal CMV infection
- Immunocompetent patient
  - CMV mononucleosis
  - Post-transfusion CMV (similar to CMV mononucleosis)
- Immunocompromised patient
  - CMV pneumonitis
  - CMV GI disease
  - CMV retinitis
  - Polyradiculopathy, transverse myelitis, and subacute encephalitis

CMV Retinitis

- Epidemiology
  - Most common ocular manifestation
  - In pre-AIDS era, mostly in immunocompromised
  - Most frequent cause of blindness in AIDS (pre-HAART)
  - In immunocompetent, most remain asymptomatic, while a minority develop mono-like symptoms
    - Age-dependent prevalence
  - Mode of transmission: body fluid
    - Reaches eye via bloodstream

CMV Retinitis

- Diagnosis
  - Ab of little value as 50% of normal population have positive value
    - False + IgM if RF or IgG is not removed properly
  - Serum DNA levels appear correlated with organ disease
  - Goldmann-Witmer coefficient may be useful in unclear cases
  - Viral PCR from ocular fluid or tissue
    - Virus may persist in tissues without causing disease
  - Clinical picture far more important than lab findings in distinguishing active and inactive diseases

CMV Retinitis

- Clinical presentation
  - Small, white, fluffy infiltrates in the peripheral retina that may be hard to distinguish from HIV microvasculopathy
  - Low-grade vitritis
  - Mild optic nerve involvement
  - Clinical course highly dependent on immune status

CMV Retinitis

- Treatment
  - The goal is to improve immune status with HAART therapy: >100 cells/µl CD4+ count
    - Good in disease prevention and regression
  - Virostatic agents
    - Ganciclovir (Cytovene): oral & IV & intravitreal
    - Valganciclovir (Valcyte): oral
    - Foscarnet (Foscavir): IV & intravitreal
    - Cidofovir: IV & intravitreal

Other Ocular CMV Infections

- Conjunctivitis
- Keratouveitis
- Corneal endothelialitis
- Anterior uveitis
- Posterior uveitis
- Panuveitis
- Papillitis

Take-Home Points

- Infections must always be kept in mind despite the fact that all else suggest an autoimmune etiology.
- While frequently the cause of retinitis in immunocompromised patients, CMV may be responsible for uveitis and other ocular inflammation in immunocompetent ones.

Thank You